

Topic: 1 - Epilepsy

Abstract—WCN 2013

No: 3

Topic: 1—Epilepsy

Nitrofurantoin-induced life-threatening seizures

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Background: Nitrofurantoin (NF) is a widely used antibiotic, used for treatment of cystitis caused by *Escherichia coli* (EC). With MEDLINE search we found no reports on seizures in patients treated with NF.

Objective: Drug-induced life-threatening seizure and discussion to avoid fatal outcome are reviewed.

Patients and methods: We report the case of an 87-year-old Slovenian female who was admitted in 2012 to a psychiatric department because of dementia. Her past history was unremarkable. She had no known history of seizures. Baseline laboratory results collected on admission were normal. Rivastigmine transdermal patch was introduced. After 10 days in hospital, patient was treated with NF 200 mg daily because of lower urinary infection with EC.

Results: After 3 days of treatment with NF seizures (grand mal) and convulsions were observed first time, consequently midazolam was introduced. Next morning patient was transferred to the Department of Internal Medicine. During transportation, seizures with convulsions occurred and patient received midazolam and oxygen immediately. NF was withdrawn from therapy and switching to amoxicillin and clavulanic acid 2 g daily was introduced. After seizures, laboratory tests results of serum electrolytes were as follows: hyponatremia (Na 110 mmol/L) with low plasma osmolality and elevation of urinary sodium and urinary osmolality. A normal baseline laboratory results were determined and polyuria was noted. Patient left hospital 2 weeks later with no symptoms of EC urinary infection and psychical status normalized.

Conclusion: NF-induced acute hyponatremia leading to seizures is seen rarely. Early psychiatric evaluation and another antimicrobial agent should be used in elderly psychiatric patients with lower urinary tract infections.

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Abstract—WCN 2013

No: 27

Topic: 1—Epilepsy

Improvement of quality of life in resistant epilepsy by combined therapy with repetitive transcranial magnetic stimulation

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Background: Some new technologies are develop today to reduce seizures and to improve quality of life in resistant epilepsy: DBS, VNS, tDCS, rTMS.

Objective: The aim of our study is the improvement of a quality of life in epilepsy patients by including low-frequency repetitive transcranial magnetic stimulation into combined therapy.

Methods: Thirty six patients with epilepsy (mean age = 28.1 ± 2.9 years) who take low doses of anticonvulsants (AED) were studied (EEG, neuropsychological tests, QOLIE-31, SSQ). Patients needed to decrease AED doses in consequence of expressed side effects. RTMS (1 Hz, 20% of big ring coil (Neurosoft) MMI intensity) was performed during ten consecutive days over the temporal lobe projection with focusing on hippocamp.

Results: Mean seizure frequency per week after 10 rTMS significantly decreased in the following 4-weeks after rTMS period compared with the pre-treatment period (2.3 vs. 0.17 per week; $P = 0.016$) which corresponds to 82.9% reduction. Anticonvulsive rTMS effect and seizure severity lowering persisted during 3 months ($p < 0.05$).

Depression was diagnosed in 42% of patients before complex therapy. Direct after rTMS course antidepressive and antianxious effect was obtained at 46% of depressive subjects and objectified by Beck and Spielberger–Hanin scales ($p < 0.004$). The common QOLIE-31 point significantly increased one and three months after rTMS ($p < 0.05$).

Conclusions: Our results indicate that including of 1 Hz low intensity rTMS into complex epilepsy therapy with low doses of anticonvulsants has a determining function and leads to a significant anti-epileptic effect, improves the psychological condition and quality of life of these patients.

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Abstract—WCN 2013

No: 30

Topic: 1—Epilepsy

Vagus nerve stimulation in 5 patients with drug-resistant epilepsy in western China

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Purpose: This study was designed to assess the efficacy and safety of vagus nerve stimulation in adults with drug-resistant epilepsy.

Methods: In this retrospective review, a database was prospectively created with 5 patients from western China, who underwent vagus

nerve stimulator implantation for drug-resistant epilepsy between July 2011 and March 2012; all of the patients were males aged 19 to 31 at the time of implantation. All patients in the primary implantation group had adequate follow-up (>3 months from implantation) and no patient had early device removal because of infection.

Results: Duration of vagus nerve stimulation treatment varied from 7 to 15 months. Seizure freedom was achieved in 3 patients, 80% seizure reduction in 1 patient, and no improvement in 1 patient. No permanent injury to the vagus nerve occurred to any patient.

Conclusion: Vagus nerve stimulation is a safe and effective palliative treatment for focal and generalized drug-resistant epilepsy in adults in western China. When this treatment was used in conjunction with a multidisciplinary and multimodality treatment regimen including aggressive antiepileptic drug regimens, 3 in 5 patients with drug-resistant epilepsy experienced seizure free. Long-term prospective randomized trials are needed for patients with drug-resistant epilepsy to potentially expand the number of patients who may benefit from this palliative treatment.

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Abstract—WCN 2013

No: 31

Topic: 1—Epilepsy

Seizure freedom of Epilepsia Partialis Continua (EPC) with vagus nerve stimulation (VNS) therapy: A case report

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Vagus nerve stimulation (VNS) is generally considered as a palliative treatment for patients with drug-resistant partial onset epilepsy. We report a case that a patient with drug-resistant generalized onset epilepsy, Epilepsia Partialis Continua (EPC) became seizure free for 13 months with VNS combined with antiepileptic medication regimens. A 21-year-old, right-handed man started having seizures at the age of 18. He reported countless limbs shaking for about 10 s, and occasional generalized tonic-clonic seizures at a frequency of 5–7 times per year. He was initially treated with valproic acid (VPA) and carbamazepine (CBZ) with minimal benefits. Then he was treated with lamotrigine (LTG) 200 mg daily, Levetiracetam (LEV) 1250 mg daily and topiramate (TPM) 200 mg daily with minimal benefits for the last several months. His long-term video-EEG monitoring revealed small spike wave discharges. Sometimes he couldn't say a word or have dinner for the twitch of the mouth. A vagus nerve stimulator was subsequently implanted in Sept 2011. Initial stimulation parameters were: current output 0.25 mA, frequency 30 Hz, pulse width 250 ms, 30 s signal on time, and 5 min signal off time. The patient had two wild twitches of the mouth per week. Then his stimulation current output was changed to 0.50 mA, and his seizure became countless as pre-operation. Fortunately, he became seizure free after his stimulation current reverted to 0.25 mA. After one month of seizure freedom, the patient withdrew his LEV and TPM. He has remained seizure free for almost 9 months, during which time he has been gainfully employed.

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Abstract—WCN 2013

No: 48

Topic: 1—Epilepsy

Temporal lobe epilepsy patients associated with mild focal cortical dysplasia. Quantitative assessment by cortical surface analysis

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Objective: To evaluate temporal lobe cortical surface through quantitative magnetic resonance imaging (MRI) and its relation to postsurgical evolution in patients with temporal lobe epilepsy associated with mild focal cortical dysplasia with MRI negative.

Methods and subjects: Measurements of cortical surface area (SA), cortical thickness (CT), folding index (FI) and volume were generated with FreeSurfer in 10 patients and 20 subject controls. High resolution pre-surgical T1-weighted volume scans 160 contiguous slices were obtained for each subject.

Results: The patient group displayed significantly reduced volume in entorhinal cortex, parahippocampus, superior and inferior temporal gyrus; SA was also greater in ipsilateral neocortical epileptogenic zone when compared to the control group. An increase of the CT was also shown in entorhinal cortex and parahippocampus. In addition, the ipsilateral neocortex showed an increase folding index ($p < 0.0001$ Test Mann–Whitney). Class 1 Engel's patient had greater volume of mesial structures and smaller volume in neocortex. CT was lower in entorhinal cortex and parahippocampus, whereas the SA has a tendency tended to be higher in best evolution patients ($p < 0.0008$ Test Mann–Whitney).

Conclusions: Patient's with TLE associated with mild FCD showed significant volume reductions and CT increase in entorhinal cortex and parahippocampus. There was also an increase of the SA and a significant volume decrease in ipsilateral temporal lobe epileptogenic zone. Patients who had better clinical evolution showed bigger volume in the mesial structures associated with smaller neocortical volume and SA increase (superior and inferior temporal lobes).

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Abstract—WCN 2013

No: 67

Topic: 1—Epilepsy

Disrupted migration of GABAergic interneurons lead to the epileptogenesis: Interneuron pathology associated with Arx mutation

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Aim: X-linked lissencephaly with abnormal genitalia (XLAG), showing severe neonatal seizure and developmental delay, is a rare disorder caused by mutations in the *aristaless*-related homeobox (*ARX*) gene, located in Xp22.13. *Arx*-null mice for human XLAG model showed loss of tangential migration of GABAergic interneurons. However, GABAergic interneuron distribution of XLAG brain has never been reported. In the present study, we investigated subpopulation of GABAergic interneurons in the brain of an infant with XLAG, who had a nonsense mutation of the *ARX* gene, compared with those of age-matched normal control and Miller–Dieker syndrome.

Methods: We performed immunocytochemistry for interneuron and migration markers.

Results: Glutamic acid decarboxylase (GAD)- and calretinin (CR)-containing cells were significantly very few in the neocortex and, interestingly, located in the white matter and neocortical subventricular

zone, while neuropeptide tyrosine and cholecystokinin positive cells were normal. From previous rodent studies, the imbalance of GABAergic interneurons may be derived from the caudal ganglionic eminence tangential migration. Also, in the neocortical subventricular region, the GAD- and CR-containing cells had Mash-1 protein, like a radial migration marker, and nestin protein.

Conclusion: ARX protein controls not only tangential migration of GABAergic interneurons from the ganglionic eminence, but also may serve to induce radial migration from the neocortical subventricular zone.

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Abstract—WCN 2013

No: 82

Topic: 1—Epilepsy

Neurometabolic effect of repetitive transcranial magnetic stimulation in epileptic brain: A sham-controlled proton magnetic resonance spectroscopy study

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Background: Repetitive transcranial magnetic stimulation (rTMS) is a perspective method to reduce seizures at resistant epilepsy. Mechanisms of rTMS action are active researched.

Objective: The study was designed to evaluate the neurometabolic effect of single and ten sessions of low-frequency repetitive transcranial magnetic stimulation over temporal lobe.

Patients and methods: Sixteen epilepsy patients (mean age = 28.1 ± 2.9 years) were enrolled in a prospective single-blind, randomized study [sham (n = 7) vs. real (n = 9)]. rTMS (1 Hz, 20% MMI intensity of ring coil) was performed during 10 min over the temporal lobe projection with focusing on hippocamp. Hippocampal brain regions bilaterally were investigated by single-voxel proton magnetic resonance spectroscopy (¹H MRS) before and after rTMS.

Results: Our results showed a significant reduction of seizure frequently after real 10 rTMS sessions that lasted for at least 3 months following treatment in 66.8% patients (p < 0.05). These clinical changes were correlated with increases in N-acetylaspartate (NAA)/Cholin (Cho) + Creatin (CR) ratio in the ipsilateral hippocamp region after single and ten rTMS sessions (p < 0.05). We found a trend for a positive correlation between increases NAA levels at both hippocampus and remission of epilepsy more than 3 months (r = 0.67, p = 0.05). For the sham group, there were no significant increases in NAA levels after rTMS (p > 0.1).

Conclusion: New technology of low-frequency rTMS delivered into temporal lobe leads to significant antiepileptic effect at patients with resistant epilepsy. That effect correlated with neurometabolic effect in deep brain regions and can predict course of epilepsy after complex therapy with low-intensity rTMS.

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Abstract—WCN 2013

No: 140

Topic: 1—Epilepsy

Investigation of patients with temporal lobe epilepsy by means of quantitative electroencephalography and magnetic resonance spectroscopy

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Objectives: Quantitative electroencephalogram (qEEG) plays a significant role in investigating the brain activity. To improve the spatial resolution, qEEG analysis has also been combined with medical imaging technology like magnetic resonance imaging. One of these medical imaging techniques is magnetic resonance spectroscopy (MRS) which is an MRI technique used to measure regional variations in neurochemistry and display concentrations of various brain metabolites in preset regions of interest in the brain. In this study we aimed to evaluate the correlation of qEEG abnormalities and MRS findings in patients with temporal lobe epilepsy (TLE).

Methods: Forty-four patients (29 women and 15 men) aged between 13 and 70 years were included to this study and the power spectrum analyses of each multi-channel EEG were performed to all patients with the concomitant MRS measures of Cho, total Cr, Naa concentrations, and ratios of Cho/Cr, Naa/Cr and Naa/Cho.

Results: Mean qEEG band powers of anterior alpha, anterior beta, central alpha, central beta, posterior alpha, and posterior beta on the right temporal area were significantly bigger in the patients whose Naa/Cho + Cr ratio was lower than 0.71 than the patients whose Naa/Cho + Cr ratio was equal and bigger than 0.71. In addition correlation analyses between Naa/Cho + Cr, Naa/Cho + Cr and Naa/Cho ratios and qEEG band power values showed low–medium correlations.

Conclusions: Our study's results suggest that there is not a strong but low–medium correlation between qEEG and MRS findings of patients with TLE that would be explained by concomitant but different pathological factors causing both MRS pathologies and qEEG abnormalities.

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Abstract—WCN 2013

No: 174

Topic: 1—Epilepsy

The effect of antiepileptic drugs on IQ in children with epilepsy

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Objective: Measures in this study focused primarily on IQ scores in children suffering from epilepsy with special emphasis on different antiepileptic medications.

Material and methods: Our study cohort included 56 patients (30 girls and 26 boys) with a mean age of epilepsy onset 8.9 ± 2.64 years, ranged from 5 to 15 years. The cognitive functions were analyzed every six months from January 2011 through February 2013 by using the batteries of neurocognitive tests.

Results: The duration of epilepsy was ranged from one to nine years, with the mean age of 4.18 ± 1.79 years. In the carbamazepine group the IQ scores decreased from 85.9 ± 9.48 to 79.45 ± 6.99 at last follow-up period (t = 3.129, p = 0.006). Patients who take valproic acid have demonstrated lower intelligence scores 75.25 ± 5.06 compared with baseline 80.95 ± 6.8 (p = 0.001, t = 3.77, 95% CI). In the topiramate group the IQ scores have also declined to 69.95 ± 9.01 compared with baseline 80.40 ± 6.36 (t = 8.09, p = 0.003, 95% CI). Patients on levetiracetam have shown improvement in intelligence scores that can be associated with significantly decreased seizure frequency in this group. Cognitive outcomes were defined as either measure was worse in patients with higher seizure frequency (more than one seizure per month), t = 2.16, p = 0.038.

Conclusion: Antiepileptic drugs as well as seizure frequency can impact on cognition in children with epilepsy. However, this association was dose dependent. An improved diagnostic and treatment methods of the consequences of epilepsy in childhood can increase the probability of a favorable prognosis.

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Abstract—WCN 2013**No: 176****Topic: 1—Epilepsy****Outcomes of using prednisolone and ACTH in infants with West syndrome**I. Khusainova, T. Khusainov, G. Sadikova. *Department of Pediatric Neurology, Tashkent Pediatric Medical Institute, Tashkent, Uzbekistan*

Objective: Hormonal treatment of infants suffering from West syndrome is widely used. The aim of the current study was to compare the efficacy and cost of oral prednisolone and synthetic ACTH (Synacthen Depot) therapy in the treatment of infants with West syndrome from Uzbekistan.

Material and methods: In this retrospective cohort study thirty-four infants were included with new-onset West syndrome who have been treated in the Republic Children's Hospital since September 2010. Inclusion criteria were presence of clinical spasms, hypsarrhythmic patterns on EEG, neuropsychic impairment and at least six months of follow-up period. Based on the 2004 United Kingdom Infantile Spasms Study (UKISS) we used high-dose of oral prednisolone (4 mg/kg). A single intramuscular injection of ACTH at doses of 40–60 IU/day has been used according to our protocol.

Results: Twenty two males and twelve females were participated in this study. Mean age of seizure onset was 5.7 ± 2.4 months. Twenty-one infants (61.7%) were identified as symptomatic whereas thirteen (38.3%) were cryptogenic. Fourteen (41.2%) infants received ACTH injection whereas twenty (58.8%) infants received oral prednisolone. Initial response to first line therapy was similar (71.4% for ACTH and 65% for prednisolone, $p = 0.24$). Adverse events were reported in fifteen (75%) infants who have received prednisolone compare with eleven (78.6%) infants who were on ACTH.

Conclusion: Prednisolone and ACTH showed no significant difference in the initial treatment of West syndrome. We are prescribing oral prednisolone as a less expensive alternative of ACTH therapy of West syndrome.

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Abstract—WCN 2013**No: 182****Topic: 1—Epilepsy****EEG phase relationship among electrodes for child absence epilepsy**Z. Meiyun^a, J. Nan^b, Z. Benshu^c, C. Ying^c, W. Fenglou^c. *^aDepartment of Neurology, Tianjin Union Medical Centre, Tianjin, China; ^bMechanics Department, Tianjin University, Tianjin, China; ^cNeurology Department, General Hospital of Tianjin Medical University, Tianjin, China*

Background: EEG phase relationship among electrodes in seizures of child absence epilepsy (CAE) is not conformed.

Objectives: To investigate EEG phase relationship among electrodes in seizures of CAE and compare it with that of normal controls.

Patients and methods: The transient EEG signals of 10 clinical seizures with 20 sub-clinical epilepsy-form discharge in 15 cases of children absence epilepsy and 12 normal controls are recorded at awake and eye-closed state. Wavelet transform is employed to unfold the digital EEG signals into multi-scale components. The phase-lock average waveforms (PLAW) of different electrodes are obtained by phase-lock conditional sampling and phase-lock average techniques. The phase differences among electrodes are determined by correlation functions for PLAW. The phase relationships among electrodes of PLAW in CAE are compared with that of the 12 normal children at the same age.

Results: There exists phase-difference in PLAW among electrodes of normal controls. A phase-difference of PLAW between forehead

lead and occipital lead is about $\pi/2$ for normal controls. There are two types of phase-difference for CAE. One type is the PLAW of all electrodes are synchronized while another type is that the phase-difference between forehead lead and occipital lead is about π .

Conclusions: Phase-lock conditional sampling and phase-lock average techniques are employed to obtain the phase-difference of the PLAW among electrodes. The study result shows that the phase-difference among electrodes for CAE is different from that of normal controls and there are two types of phase-difference for CAE, whose mechanism remains to be fatherly explored.

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Abstract—WCN 2013**No: 188****Topic: 1—Epilepsy****Cross-reactivity of anti-epileptic drugs and its association with the HLA-B*1502 allele in Han Chinese patients**W. Wang, F.Y. Hu, X.T. Wu, D.M. An, B. Yan, D. Zhou. *West China Hospital, Sichuan University, Chengdu, China*

Background: Recently, a few patient cases that were positive for the HLA-B*1502 allele have been reported to exhibit cross-reactivity to AEDs, suggesting that the HLA-B*1502 allele may contribute to the genetic susceptibility to the cross-reactivity of cutaneous adverse reactions (cADRs) to AEDs. However, there is limited convincing evidence, and this hypothesis is still controversial owing to the small sample sizes used in previous studies. Thus, it is necessary to further explore the potential association between the cross-reactivity of AEDs and the HLA-B*1502 allele.

Objective: The aim of this study was to investigate the cross-reactivity between different types of AEDs in Chinese patients and to detect its association with the HLA-B*1502 allele.

Patients and methods: A total of 107 subjects, including 37 patients and 70 healthy volunteers, were tested for the HLA-B*1502 allele. All of the patients had a history of cADRs induced by one AED.

Results: Twelve out of 37 patients had experienced cross-reactivity, of which only two patients carried the HLA-B*1502 allele. The allele frequencies of HLA-B*1502 in the cross-reactivity group ($2/12 = 16.7\%$), tolerant group ($5/25 = 20.0\%$) and healthy control group ($5/70 = 7.1\%$) were not significantly different ($P > 0.05$).

Conclusion: Physicians should avoid prescribing aromatic AEDs as an alternative drug in patients who experienced aromatic AED-induced cADRs. Although the HLA-B*1502 allele is not a universal risk predictor for the cross-reactivity of cADRs to AEDs, but it may be a specific marker for cross-allergic reactions between CBZ and OXC as well as between CBZ and PHT.

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Abstract—WCN 2013**No: 193****Topic: 1—Epilepsy****Predetermining risk factors for development of epilepsy in children with encephalitis**N. Sharipova, A. Khalilova. *Neurology, Child Neurology & Medicine Genetics, Tashkent Pediatric Medical Institute, Tashkent, Uzbekistan*

In some cases, seizures, occurring in acute encephalitis, disappear completely, but at the majority of patients is developing symptomatic epilepsy.

Purpose: Identify risk factors for the formation of the epileptic focus in encephalitis.

Materials and methods: The study involved 30 patients with symptomatic epilepsy at the age of 6 months till 5 years old who had encephalitis of various etiologies. Anamnesis of patients, the data of clinical research, electroencephalography, and MRI have been analyzed.

Results: The analysis of anamnesis of patients was identified cerebral ischemia in perinatal period in 18 (60%) cases. Inflammatory disease of the brain in patients has developed as a result of the intestinal infection in 11 (36.7%), of respiratory viral infection in 15 (50%), of vaccination in 4 (13.3%) cases. Seizures were generalized tonic-clonic character in 12 (40%), tonic in 11 (36.7%) cases and in 7 (23.3%) cases were generalized tonic-clonic seizures with focal component. Serological examination of blood showed increasing of antibody titer IgG fraction to herpes simplex virus in 18 cases and in 12 cases to cytomegalovirus.

On MRI in 7 (23.3%) cases were identified structural changes in the brain: gipogeneziya callosum in 4 (13.3%), shizentsefaliya in 2 (6.6%) and porentsefaliya in 1 (3.3%) case.

Conclusion: The formation of epileptic focus determines many factors, including intra-uteri infection, chronic hypoxia, hereditary factors, structural changes of the brain, and inflammatory diseases of the brain in most cases, are the impetus for the manifestation of epileptic seizures.

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Abstract—WCN 2013

No: 216

Topic: 1—Epilepsy

Review article: Epilepsy in Ethiopia

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Background: Chronic non-communicable diseases, such as epilepsy, are increasingly recognized as important health care problems in developing countries. The most prevalent neurological disorder identified was epilepsy.

Methods: A review of journals was done on papers: movement disorders in Ethiopia available until the end of June, 2011 at the main Library of Addis Ababa University, School of Medicine, Department of Neurology.

Results: It was estimated 360 to 400 thousand epileptic Ethiopians are living with poor medication. The prevalence of epilepsy was 5.2/1000 inhabitants at risk, 5.8 for males, and 4.6 for females. The highest age-specific prevalence was found for ages 10–19 years. The annual incidence of epilepsy was 64 in 100,000 inhabitants at risk, 72 for males, and 57 for females. Generalized tonic-clonic seizures were the most common seizure type and occurred in 69–81%. On clinical grounds, partial seizures occurred in 18–20% and in one-third of these secondary generalizations followed, unclassifiable seizures occurred in 11%.

Traditional treatment with local herbs, holy water and amulets was the most common. Only 1.6% had been treated with recognized antiepileptic drugs in rural part of the country and as few as 13% were treated with antiepileptic drugs in cities like Addis Ababa.

Conclusions: Despite health education on epilepsy given to the community, a minority of subjects were treated with AEDs, which may reflect the inadequacies of the health services and transportation difficulties faced by the patients.

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Abstract—WCN 2013

No: 243

Topic: 1—Epilepsy

Electro-clinical profile and identification of predictors of seizure control in adults with complex partial seizures in Kano, northwestern Nigeria

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Background: Ability to predict seizure control with future treatment can be very useful in the management of patients with CPS. However, there is paucity of data on CPS in northwestern Nigeria. The aim of the study was to profile the clinical and EEG characteristics of CPS as well as evaluate independent determinant of a 2-year seizure control of CPS.

Method: This was prospective systematic study of patients with CPS in two tertiary hospitals in over a period of 4 1/2 years. The patients were followed up for a minimum of a 2-year period to determine their seizure control status.

Results: A total of 158 (105 males and 53 females) were enrolled. Thirty (19%) were lost to follow up. Their age ranged between 15 and 85 (median = 30.5) years. Sixty six (41.7%) patients had aura and 65 (60.8%) had automatism. The most common aura and automatism were abnormal epigastric sensation and oro-alimentary respectively. Twenty eight (18%) had associated behavioral manifestations. EEG abnormality was recorded in 56 (53.9%). Sixty seven (42.4%) had tried traditional treatment before having been seen by a physician. On AED, 117 (55.7%) had adequate seizure control. Duration of epilepsy was identified as the only independent determinant of a 2-year seizure control.

Conclusion: Complex partial seizure was one of the most common seizure types in adult epilepsy occurring commonly between the second and fourth decade of life. Abnormal epigastric sensation and oro-alimentary automatism were the most common aura and duration of epilepsy > 2 years was the only independent determinant of a 2-year seizure control.

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Abstract—WCN 2013

No: 248

Topic: 1—Epilepsy

An assessment of clinical practice toward pharmacological treatment of epilepsy: A survey of adult neurologists in R. Macedonia

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Purpose: The aim of the study was to evaluate clinical practice toward pharmacological treatment of epilepsy among adult neurologists in Macedonia.

Methods: A questionnaire was developed that addressed decisions about when to start and to stop antiepileptic drug (AED) treatment, as well as the choice of the first and alternative drugs about different types of seizures.

Results: 24 neurologists answered the questionnaire. Half of them would administer antiepileptic drug after first unprovoked seizure considering EEG, MRI and other investigations, while the others will wait for second seizure. For most of them discontinuation of the treatment should be individualized, while the others would do that

after 3–5 or more than 5 years of seizure-free period. Most commonly used first-line drugs for partial seizures were carbamazepine and lamotrigine, while the second choices vary among a number of new generation and older AEDs. For generalized tonic–clonic seizures, absences and myoclonic jerks, valproate was by far the most commonly used AED (surprisingly, carbamazepine was drug of choice for absences for 16.5% of neurologists). Valproate is prescribed by almost all of neurologists when patients have several types of seizures. Alternatives for patients with mixed seizures were usually lamotrigine and topiramate. Commonly prescribed doses of AEDs tend to be lower to moderate and almost no one administers maximal tolerate doses.

Conclusion: A survey showed that prescription patterns in high percent were in concordance with current evidence about the spectrum of efficacy of individual AEDs in different types of seizures. Yet some of results are a cause of concern.

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Abstract—WCN 2013

No: 253

Topic: 1—Epilepsy

Socio-demographic profile and management of patients living with epilepsy in Dakar, Senegal

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Epilepsy remains a major public health problem especially in developing countries where access to new therapies remains limited. We conducted a cross-sectional study over a period of eight months from November 2009 to June 2010 at Fann Hospital and Health Center Pikine through research on adherence.

The objective was to describe the sociodemographic profile and management of patients living with epilepsy.

The study involved patients living with epilepsy aged over 15 years, diagnosed clinically with epilepsy and/or confirmed by an EEG and put under antiepileptic drug for more than 3 months.

We recruited 411 patients aged 15–74 years with a mean age of 28.93 years.

The age range was 15–24 years with 44.6% majority.

The male sex predominated with 52.3% and the sex ratio was 1.09.

Singles outnumbered with 64.7%.

The level of education was the most representative secondary with 29.4% in patients without profession were 35.5%.

Most of the patients were from semi-urban areas with 47.7%.

Generalized seizures were more frequent with about 70%.

Most of the patients were taken care of either by their family or by their own expense.

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Abstract—WCN 2013

No: 274

Topic: 1—Epilepsy

Peculiarities of the epilepsy immunopathogenesis

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Epilepsy remains to be one of the actual problems of the modern neurology. In the 60s of the 20th century Semenov V.F. laid the foundation of new direction in the study of epilepsy pathogenesis. Particularly, he proposed a hypothesis about the role of autoimmune mechanisms of epilepsy.

Purpose: To study immunological aspects of idiopathic and symptomatic epilepsy with determination of the role of immunological pathomechanisms of their development.

Material and methods: There 52 patients with symptomatic and idiopathic epilepsy that were studied. Thirty eight patients (group I) were with symptomatic epilepsy and 14 patients (group II) had idiopathic epilepsy. In this case we studied level of neurotropic autoantibodies to NF-200, GFAP, S100, OBM, Voltage-dependent Ca channels, glutamate receptors, GAMC-, dopamin, serotonin and n-choline-receptors.

Results: Neuroimmune change in epilepsy is accompanied by increase in autoantibodies to the receptors of choline, GAMC, dopamin, serotonin and borderline deviations to the protein B2 glycoprotein. However, more reliable in comparison with parameters of control group contents of autoantibodies was noted to such neurospecific proteins such as NF-200, S100 and to double-helical DNA.

Conclusion: The parameters of the protein S100 in both groups were higher and reliably differ from control group. In the patients from group II there was noted increase in autoantibodies to double-stranded DNA that in this case may be considered as the marker of the autoimmune process in the central nervous system.

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Abstract—WCN 2013

No: 314

Topic: 1—Epilepsy

Anthropological and epidemiological study of epilepsy in the region of Tangier (Morocco)

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Objectives: Determine the knowledge, attitudes and beliefs of the public towards epilepsy in Tangier region (northern Morocco).

Methodology: Performed in the Neurology Department at the Hospital Al Kortobi Tangier, via a standardized questionnaire about epilepsy, conducted on 180 participants (patients and family members).

Results: The study revealed that the disease is unknown in this region because 66.1% of respondents have not “read” or “heard”, about a disease called “epilepsy”, 60% did not know a person with epilepsy, yet 90% of the respondents attended convulsions. Concerning the public’s attitude towards epilepsy, the least we can say is that the rejection and marginalization watch with epilepsy: 70% do not allow their children to play with people who may suffer from convulsions. 80% of the participants do not allow their children to marry sometimes with people making seizures. 50.6% of respondents believed that epileptics cannot exercise any craft. Worse, 40% of respondents believed that epilepsy is a form of mental retardation. 15.6% of the people interviewed believed that the main cause of epilepsy is supernatural (jinn, witchcraft) and 20.6% think it’s caused by stress, 15.6% think it is related to the alcohol and drugs. As for treatment, 68.9% of our sample suggested to ask doctor advice to treat epilepsy, 46.7% using Fkih’s help, 8.3% see a psychologist. 11.10% of people with epilepsy use the plant *Ruta graveolens* to treat epilepsy.

Conclusions: Epilepsy remains a misunderstood disease and is still subject to wrong interpretation. Awareness campaigns should be conducted to clarify certain aspects of epilepsy.

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Abstract—WCN 2013**No: 313****Topic: 1—Epilepsy****Epilepsy: Analysis of electroencephalograms and cerebral maturation**

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The objective of this study was to emphasize why and how the change in brain electrical activity in children, adolescents, adults and old men, and take, therefore, a relationship between the evolution of the EEG and epilepsy related to cerebral maturation.

Methods: A clinical study based on analysis of 100 electroencephalograms (EEGs) of epileptic and normal subjects of different ages was carried out at the hospital Al-Kortobi of Tangier, Service of Neurology (Morocco).

Results: Analysis of electroencephalograms showed, first, that during childhood the EEG is characterized by the gradual transformation of brain waves (Delta–Theta–Beta–Alpha) in time reflecting the different stages of brain maturation. On the other hand, there is a regional component of brain maturation in a posterior–anterior gradient chronological. Completion maturation is set between 12 and 14 years.

Conclusions: One thus understands why the newborn babies make mainly motor seizures and why the occipital epilepsy begin preferentially in the very first months from life, while the frontal epilepsy appear only seldom before the 2 year age, and the temporal epilepsy seldom before 8–10 years.

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Abstract—WCN 2013**No: 373****Topic: 1—Epilepsy****Vascular seizures**

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Objective: 39 vascular seizure patients admitted on last 6 months 2012, aged from 20 to 88 years (mean 60.71 years), 18 M; 21 F were evaluated.

Previously hypertension prevailed in 32 patients isolated or associated with: atrial fibrillation, smokers, diabetes mellitus, cerebral palsy, thrombotic syndrome, post-traumatic vertebral dissection stroke, mitral stenosis and clipped aneurisma. **Etiology:** hemorrhagic (49%), ischemic (61%); post-ischemic 22, post-hemorrhagic 8, acute ischemic 3, acute repeated ischemic 4 and acute hematoma 3 patients. Seizures determining epilepsy followed stroke meanly 15 months/mostly 1–6 m after (74%), while immediate seizures occurred in (26%). Partial motor mostly secondary generalized prevailed (59%), generalized without evident laterality (18%), partial complex (18%), and partial sensory seizures (5%). 16% had secondary generalized SE from partial motor onset. **Stroke** lateralization; left (46%) to right sided (39%) while bilateral (15%). **Lobar localization;** F (44%) isolated or spread to P/T (9/8), T (41%) lying to P in 9 or T/T-insular in 7, while bilateral localization in 6 (15%). **EEG:** Ictal; F, F-T discharge (5%), right F dominated PLEDs (10%). Unrealized (5%) or interictal (80%) **in which** anterior bilateral unilaterally predominated (41%), focal (23%), bilaterally anterior / posterior to diffuse (16%). AEDs: VPA 13, CBZ 8, TPM 2, LEV 2, GBP 4, DHT 6, CNZ in 3 while two cases of repeated ischemic hemispheric stroke died.

Conclusion: Hypertension was the main cause of stroke determining seizures and women were more affected. Ischemic stroke slightly prevailed, while lobar F and T localization were mostly affected.

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Abstract—WCN 2013**No: 375****Topic: 1—Epilepsy****Seizure severity and health related quality of life of adult Nigerian patients with epilepsy**

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Backgrounds: There is paucity of information about the association of seizure severity and quality of life in PWE in sub-Saharan Africa. We hypothesized that seizure severity will significantly influence QoL of Nigerian PWE.

Objective: We evaluated the relationship of seizure severity to health related quality of life of patients with epilepsy being followed up in an outpatient neurology clinic in southwestern Nigeria.

Materials and methods: Eighty-eight consecutive patients with epilepsy who met the recruitment criteria completed the study questionnaire in company of an eyewitness. The study questionnaire comprised of the National Hospital Seizure Severity Scale (NHS3), the Quality of Life Inventory in Epilepsy (QOLIE-31) and the Beck's Depression Inventory–II (BDI-II).

Results: We found a minute association between seizure severity and QOLIE-31 total score ($r = -.262$, $p = .014$). This association was washed off in the context of depression. However, increased seizure severity predicted a worse QOLIE-31 seizure worry ($R^2 = 0.311$, $\beta = -.289$; $p = 0.003$). On a multivariate regression analysis, generalization of seizures and the presence of falls were items on NHS3 that predicted a worse QOLIE-31 seizure worry score and time to recover predicted a worse QOLIE-31 total score.

Conclusions: Reducing seizure severity may be an alternate endpoint in epilepsy care in Nigeria (particularly difficult to control seizures) because of its practical clinical relevance in view of the fact that state of the art epilepsy care is still farfetched. Mood stabilizing antiepileptic medications may be useful in this respect.

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Abstract—WCN 2013**No: 378****Topic: 1—Epilepsy****Symptomatic epilepsy as a result of stroke**

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Objectives: Cerebrovascular diseases are significant aetiological factors of epilepsy. Seizures may appear in the acute phase of the insult, or during the reconvalescence. Our goal was to analyse the frequency and correlation between epileptic seizures and the type and size of cerebrovascular lesion.

Methods: In this prospective study, we evaluated patients with stroke who were hospitalised at the Department of Neurology in Nis,

between January and December 2012. Witnessed epileptic seizures occurred in 129 patients. Patients were evaluated and had the same investigations with anamnestic, clinical, neurological, EEG and neuroimaging (CT, MRI) variables which were compared.

Results: Of the 1386 patients with stroke who were admitted to the hospital, 482 (34,78%) had haemorrhagic, and 904 (65,22%) had ischemic stroke. Mean age was 52 ± 30 years. Of a total of 129 patients with witnessed epileptic seizures, 76 were male and 53 female. 46 (35, 66%) patients with haemorrhagic and 83 (64, 34%) patients with ischemic stroke developed seizures after 14 days of the stroke. Partial motor seizures (PMS) were registered in 73 (59, 35%) patients, partial seizures with secondary tonic-clonic generalisation (GTC) in 39 (31, 70%) patients and primary GTC seizures in 17 (13, 82%) patients. Status epilepticus (SE) was registered in 9 patients. 69 (53, 49%) patients had EEG pathological changes (spike or sharp-waves) and 36 (27, 90%) patients had focal or diffuse Theta-Delta waves. Normal EEG patterns were registered in 24 (18,60) patients.

Conclusion: Patients with ischaemic stroke and cortical lesions are at a higher risk of developing seizures. Partial motor seizures and partial seizures with secondary tonic-clonic generalisation are a dominant feature. Our results differ from the ones in literature, in which patients with haemorrhagic stroke are more likely to develop seizures. It may be the consequence of the fact that the mortality in haemorrhagic stroke patients is higher.

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Abstract—WCN 2013

No: 381

Topic: 1—Epilepsy

The spectrum of fixation-off sensitivity and scotosensitivity: Typical and atypical forms

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Background and objectives: To define the spectrum of the epileptic syndromes and epilepsies that can be associated with fixation-off sensitivity (FOS) and delineate the electro-clinical types of FOS.

Design and methods: Clinical and video EEG data of all our patients with FOS over the last 7 years were reviewed using FOS technique described by Koutroumanidis and Tsipsios.

Results: From January 2005 to October 2012, 14 of about 1900 patients had one or more video-EEGs with FOS (0.7%). From the 14 patients with full clinical and EEG data available, 10 had various epilepsies that included: symptomatic or probably symptomatic focal (9), cryptogenic generalized (3), and two had no seizures. Two patients (33%) were photosensitive and one was scotosensitive. FOS EEG abnormalities were occipital in 8 patients, and generalized in two. Two showed atypical forms. One boy of normal intelligence showed abnormal behavior associated with disorientation and confusion and postictal amnesia. His video/EEG evaluation unexpectedly documented the presence of FOS. Another patient was diagnosed with atypical benign partial epilepsy, and his repeated video/EEG recordings showed FOS. His sister was diagnosed with epileptic encephalopathy with continuous spike and wave complexes. Three patients were diagnosed as childhood absence epilepsy.

Conclusions: Irrespective of classification, routine video-EEG monitoring for documenting FOS using Panayiotopoulos technique should be offered to selected patients with epilepsy. Unusual and rare cases within the spectrum of benign childhood seizure susceptibility syndrome can explain the atypical cases.

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Abstract—WCN 2013

No: 389

Topic: 1—Epilepsy

Ketogenic diet in Cdkl5-related epilepsy

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Purpose: To review efficacy and tolerability of a ketogenic diet (KD) in children with intractable epilepsy and CDKL5 mutations.

Methods: Children with a CDKL5 mutation initiated on the KD from 01/2009 to 12/2012 were identified from the KD diet database at Mayo Clinic. Charts were reviewed to determine seizure types and frequencies (a) prior to KD onset, (b) 3 months after KD onset and (c) at final follow-up on the KD. Significant improvement was defined as a >50% decrease in that seizure type.

Results: Seven children (4 female, median age at seizure onset 2 months) were initiated on a KD at median age 13 months (IQR 8, 19). Children were on a median of 3 AEDs (2, 3) and had previously failed 1 (0, 3) AED for lack of efficacy. Median duration of KD was 15 months (IQR 5, 27) and at follow-up, 5/7 remained on the diet. At 3 months after initiation, significant improvement was seen in 3/6 with spasms, 2/6 with focal, 2/3 with tonic and 1/1 with myoclonic seizures. At final follow-up on KD, significant improvement was seen in 3/6 with spasms, 3/6 with focal, 2/3 with tonic and 0/1 with myoclonic seizures. However, after initiation of the KD, new seizure types emerged in 4 children (1-focal, 2-tonic, 1-atonic/absence). The KD was well-tolerated, with 2/7 children developing mild and 1/7 severe adverse effects. Diet was discontinued in 2 (1-lack of efficacy, 1-adverse effects).

Conclusions: The KD was well tolerated and provided improved seizure control for the majority of children with CDKL5-related intractable epilepsy.

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Abstract—WCN 2013

No: 405

Topic: 1—Epilepsy

Major obstacles in the management of adult epileptics in a tertiary referral hospital in Northeast Nigeria

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Background: In Nigeria epilepsy affects hundreds of thousands of people with only a few neurologists available.

Objectives: We set out to assess the present state of epilepsy care in a district tertiary care hospital in northeast Nigeria, and to compare with modern management.

Materials and method: This was a retrospective study of hospital records of 200 adults with established epilepsy over the previous 4 years. We used a questionnaire to collect relevant data relating to the diagnosis and management of epilepsy.

Results: There were 150 (75%) males and 50 (25%) females with a mean age of 30.2 years (SD 5.8 years). The main aetiology was post traumatic (37%), cerebrovascular disease (22.5%), partially treated meningitis (17.5%), encephalitis (12%), and alcohol (10%). The majority of cases (85%) were generalized tonic clonic seizures. Less than a fifth had EEG and neuroimaging before commencement of AEDs. Patients referred had to travel long distances for EEG, CT and MRI brain and

most could not afford the high cost of these investigations. Majority of the patients (75%) were on phenytoin capsules, followed by phenobarbitone (10%), carbamazepine (7.5%), sodium valproate (5%), and ethosuximide (2.5%). There were no facilities to monitor blood concentrations of AEDs. There was lack of knowledge regarding causes and treatment.

Conclusion: There is paucity of older AEDs and unavailability of newer generation AEDs and most of them are not aware of the possibility neuroimaging or do not have money to travel to places where they are available. Cultural beliefs and preference for traditional healers delay treatment.

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Abstract—WCN 2013

No: 430

Topic: 1—Epilepsy

Evaluation of hormones in girls with epilepsy in dependence of receiving treatment

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Multiple changes in organism which are observed in prepubertal and pubertal age, create necessary not only of clear diagnostics, but also treatment of the disease in view of drug influence on hormonal functions. The aim of study was to investigate the influence of modern antiepileptic drugs on hormonal status in girls with epilepsy. In 50 girls aged 8–17 years with epilepsy, concentration in blood thyroid stimulating hormone (TSH), thyroglobulin antibodies (a/b TG), triiodothyronine (T₃), thyroxine (T₄), parathyroid hormone (P) and cortisol (C) was investigated. Treatment in most cases included valproic acid, carbamazepine and topiramate. Regardless of applied drugs significant differences in content of hormones were found in 2 (9,52%) cases, and in width of distribution of values — in 7 (33,33%) cases. Highest content of TSH found in girls 8–17 years treated with valproic acid, a/b TG — in ones who didn't got any antiepileptic drugs, T₃ — in ones taking barbiturates, T₄ — treated with topiramate, P — in girls taking valproic acid, C in girls 8–13 years receiving barbiturates and in girls 14–17 years — taking oxcarbazepine. Within 2–12 months after first study in 19 girls hormone levels were determined again. Levels of hormones in different treatment in relation to original average content of hormones in all girls in 64,86% cases change so as was in the first study. Significant difference in content was found in all hormones in girls with epilepsy treated with different antiepileptic drugs. Choice of antiepileptic drug, its dose and correction should be made considering its impact on children's hormonal profile.

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Abstract—WCN 2013

No: 431

Topic: 1—Epilepsy

Application of drug Levocarnitine in children with epilepsy

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Long-term studies established expressed disturbance of different brain energy links in ones with epilepsy.

The aim of the study was to identify metabolic disorders of 42 amino acids and carnitines and justify their correction by including the drug Levocarnitine in complex therapy of epilepsy in children. 32 children with epilepsy from 3 months to 14 years (12 boys and 20 girls) and

19 children of control group were examined. Blood content of 42 carnitines and amino acids was investigated. In treatment of children with epilepsy, valproic acid was used. The drug Levocarnitine was administrated to 14 children with epilepsy and months later concentration of carnitines and amino acids was examined again. It was found that in children with epilepsy, concentration of 8 amino acids and 6 carnitines was lower, and that of 4 amino acids and 26 carnitines was higher, than that in control group. Also the level of 10 amino acids and 19 carnitines in children with epilepsy significantly ($P \geq 0.90$) differs from control group. After taking Levocarnitine in 28 (66,67%) children with epilepsy, tendency to normalization of content of 8 amino acid and 20 carnitines was observed. Significant difference in the content of amino acids and carnitines in children with epilepsy and control group that was revealed before taking Levocarnitine in 12 (42,86%) cases, was absent after taking the drug. So using “Elcar” in complex treatment of epilepsy in children is safe and a quite effective method for correcting disorders of amino acids and carnitine metabolism.

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Abstract—WCN 2013

No: 443

Topic: 1—Epilepsy

Autoimmune epilepsy — Diagnosis, therapeutic remarks and outcome

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Background: Autoimmune epilepsy is a rare disorder, defined by epileptic seizures with a suspected autoimmune pathogenesis, based on the detection of neural antibodies, inflammatory CSF findings or MRI features of inflammation.

Objectives: We aimed to identify clinical characteristics and response to immunotherapy in our patients diagnosed with autoimmune epilepsy.

Patients and methods: We retrospectively reviewed 8 patients admitted in our clinic, diagnosed with autoimmune epilepsy. The clinical, imaging and immunological evaluations were recorded for each patient.

Results: All of the patients had complex partial seizures and tested positive for different types of antineural antibodies. The EEG pattern was modified, consistent with the epileptogenic lesion. The imaging studies showed variable degrees of involvement, ranging from multiple, disseminated lesions, to discrete areas of hypersignal. In two of the cases, the CT scans revealed multiple calcifications, independent of the lesions present on the MRI. Progressive cognitive impairment and neurological deterioration together with MRI lesion load were associated with an active process of inflammation. In the patients whose condition showed no signs of progression, immunotherapy brought no further benefit.

Conclusion: Patients diagnosed with autoimmune epilepsy usually have pharmacoresistant focal seizures. Early administered immunotherapy improved the seizure control, but it proved beneficial only in those patients who showed signs of active inflammatory disease.

In conclusion, it is important to consider the diagnosis of autoimmune epilepsy in patients whose status could be improved by well conducted immunotherapy. The advantages of immunotherapy should be weighed against the side effects, especially in patients who do not show signs of progression.

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Abstract—WCN 2013**No: 449****Topic: 1—Epilepsy****Driving and epilepsy; perspectives of patients with epilepsy in South Western Nigeria**

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Background: Patients with epilepsy (PWE) with poorly controlled seizures are restricted from driving in most developed countries but despite this many PWE still continue to drive.

Methods: This is a case–control study in which 42 patients with epilepsy (PWE) were studied along with 45 age and sex-matched normal control subjects. The adapted format of Driver perceptions and practices questionnaire (DPPQ) was verbally administered to the participants. DPPQ is a 46-item questionnaire that assesses the domains of safety concerns, Attitudes towards driving, Perceived susceptibility/severity, Perceived barriers, Helping relationship and Self-efficacy. The data obtained were collated and analyzed with the aid of SPSS version 15.0 statistical software.

Results: Twelve patients were driving regularly among PWE while 15 normal controls were driving regularly. The controls thought of ways to improve their safety and were less likely to have confidence to ask family or friends to drive them when compared to PWE. The PWE that were students or unemployed and those who have driving licence have lower scores on “self-efficacy” domains” when compared to PWE that were employed or possessed driving licence. PWE who had a high “Perceived barriers” to changing driving behavior and “Attitude towards driving” scores were more likely to drive when compared to PWE with lower scores.

Conclusion: There were minor differences in the driving behavior of PWE when compared to normal control subjects and PWE with high perceived barriers to changing driving behavior and poor attitude towards driving were more likely to drive.

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Abstract—WCN 2013**No: 475****Topic: 1—Epilepsy****Wada memory dominance in temporal lobe epilepsy**

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Background: The mesial temporal lobe epilepsy patients who undergo left temporal lobectomy and who have seizures postoperatively may have memory deficits. While formal neuropsychometric tests assess the material-specific memory functions, Wada test reveals a general hemispheric dysfunction against the risk of amnesia by giving stimuli that may be coded by different ways.

Objective: To examine the relationship between Wada memory dominance and verbal and nonverbal memory tests.

Materials and methods: Twenty-seven patients (11 males, 16 females, mean age 23.05 + 5.97) with selective amygdalohippocampectomy were included. All had undergone the intracarotid amobarbital test previously. Wechsler Test (immediate recall-WIR, delayed recall-WDR) and the Sozel Bellek Surecleri Testi (Turkish version of verbal memory test-delayed recall and recognition) were applied one month before and 12 months after the surgery.

Results: There was a significant difference between preoperative and postoperative WIR scores ($p = 0.045$). When we compare the results of preoperative and postoperative results of the memory tests, there was no significant difference between the scores according to the side of mesial temporal sclerosis (left/right), Wada language dominance (left/atypic) and Wada memory dominance (left/atypic). There was a significant positive correlation between the left memory dominance and verbal delayed recall test scores ($r = 0.829$, $p = 0.042$) and also between atypic memory dominance and verbal recognition test scores ($r = 0.731$, $p < 0.001$).

Conclusion: There was an improvement in visual memory tests after the surgery. In addition, although Wada memory test assesses both verbal and visual memory, Wada memory dominance can be effective in the evaluation of verbal memory, but not visual memory.

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Abstract—WCN 2013**No: 497****Topic: 1—Epilepsy****Treatment outcome of status epilepticus in Thammasat University Hospital, Thailand**

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Background: Status epilepticus (SE) is an emergency condition in epileptic patients. The morbidity and mortality are very high, particularly in those whose condition is underdiagnosed or improperly managed.

Objective: To describe clinical characteristics, treatment process, outcome and to determine clinical parameters which impact the SE outcome in Thammasat University Hospital, Thailand.

Material and methods: This was a descriptive study in 60 SE patients admitted between 2004 and 2011. Data included underlying etiologies, clinical course, type of antiepileptic drug (AED) treatment and outcome. Clinical characteristics were described in detail. The correlation between outcomes and clinical parameters was analyzed by Spearman's rank correlation coefficient.

Results: Sixty SE patients were included (38 men; 22 women), between 15 and 90 years of age, mean age 55 years. Generalized convulsive status epilepticus (GCSE) was most common (55 events, 91.6%). Forty-three patients (71.7%) had previous diagnosis of neurological disorders. AED withdrawal and old ischemic stroke were the common causes of SE. Eight patients (13%) developed refractory SE. Outcomes of SE included death (15 patients, 25%), complete recovery (16, 26.7%), partial recovery (12, 20%) and partial recovery with total dependent (17, 28.3%). The risk factors which correlated with death were old age, long duration of seizure and coma.

Conclusion: Most SE patients are old and have underlying neurological disorders. The mortality and morbidity rate were very high. Death is associated with old age, long duration of seizure and coma.

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Abstract—WCN 2013**No: 509****Topic: 1—Epilepsy****Pedunculopontine nucleus stimulation: A novel therapeutic technique in intractable epilepsy**

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Intractable epilepsy accounts for approximately 30% cases of epilepsy worldwide, and continues to pose a medical challenge even today.

Several studies have demonstrated the strong anti-epileptic influence of rapid eye movement (REM) sleep in humans, some researchers even claiming it as the most potent anti-epileptic state in human sleep–wake cycle. A reduction of REM sleep is a common feature reported in various forms of intractable epilepsy. Several studies have reported reduction to total disappearance of even the severe EEG abnormalities in West syndrome during REM sleep suggesting an explorable relation between the reduction of REM sleep and severity/intractability of seizures. In autopsy examination of the cases of West syndrome, the total number of neurons in pedunculopontine nucleus (PPN) and the number of acetylcholine neurons (AChN) in PPN in particular have been found to be reduced with comparative preservation of catecholaminergic neurons and GABAergic interneurons suggesting a specific change and displaying a strong indication of significance of lesions of AChN in epileptogenesis. Stimulation of AChN in PPN has been observed to induce REM sleep. Therefore, in view of the strong anti-epileptic influence of REM sleep, it is postulated that PPN may be electrically stimulated (with programmable and manual modes) for enhancing the genesis of REM sleep throughout the night sleep time when the susceptibility to seizure generation and occurrence is maximum. Thus, the postulated technique does hold promise as an effective anti-epileptic technique warranting an insightful study of its prospects and the success that may ensue from its preliminary trials.

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Abstract—WCN 2013

No: 518

Topic: 1—Epilepsy

Early onset of cortical thinning in children with Rolandic epilepsy

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Background: In clinical practice, there seems to be a correlation between language and Rolandic epilepsy. Epileptiform EEG discharges seem to be a cause, only morphological cortical differences in these children have never been proven.

Objective: Rolandic epilepsy is associated with language impairments. It is investigated whether Rolandic epilepsy is associated with abnormalities in cortical thickness, and whether these abnormalities are associated with declined language performance.

Patients and methods: 24 children (age 8–14 years) with Rolandic epilepsy were compared with 24 age-matched healthy controls. The Clinical Evaluation of Language Fundamentals (CELF) test was performed. Structural T1-weighted MRI was performed at 3 T to enable the assessment of cortical thickness. Linear regression was used to test for differences between patients and controls and to assess the effect of age and language indices on cortical thickness.

Results: For patients the core language index (mean \pm SD: 92 ± 18) was lower than that for controls (106 ± 11 , $p = 0.0026$) and below the norm of 100 ($p = 0.047$). Patients showed specific impairments with respect to the norm in receptive language (87 ± 19 , $p = 0.002$) and language content (87 ± 18 , $p = 0.0016$). Cortex was significantly thinner in patients in receptive language areas. Predominantly in left fronto-temporal regions, significant cortical thinning with age was found in patients only.

Conclusion: Reduced cortical thickness was found in Rolandic epilepsy in a language mediating area. Furthermore, early onset of cortical thinning was observed in multiple language mediating areas. These abnormalities represent subtle but significant pathomorphology of the language system and suggest that Rolandic epilepsy should not be considered merely as a benign condition.

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Abstract—WCN 2013

No: 392

Topic: 1—Epilepsy

Prenatal lesioning cortical dysplasia rat model with spontaneous temporal lobe seizures

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Background: Focal cortical dysplasia (FCD) is known to be a major cause of intractable epilepsy. We created a novel rat model of FCD by freeze lesioning during the late embryonic stage and examined the relationship between extratemporal FCD and epileptogenesis of temporal lobe seizures.

Objective: To investigate the epileptogenesis and histopathogenesis of FCD.

Material and methods: On embryonic day (E) 18, a frozen probe was placed on the frontal scalp of a Sprague-Dawley rat embryo through the uterus wall to produce multiple (two to four) freeze lesions on each hemisphere. EEG video monitoring for rat pups was performed from postnatal day (P) 35 to P77. Brain tissue was examined by immunohistochemistry, including semi-quantitative densitometry, at E17, E19, E21, P28 and P78.

Results: 68.8% of the rats with multiple FCDs showed spontaneous hippocampal seizures which began by movement cessation and culminated in wet-dog shakes during periods of hippocampal discharge, from P47 onwards. Microscopic findings revealed subarachnoid hemorrhage underneath the lesioned areas at E17, which developed into diffuse cortical necrosis at E19. FCD with severe disorganization and abnormal radial glial cells in the cortical layers appeared clearly after E21. Furthermore, immunoreactivities for N-methyl-D-aspartate receptors and glutamate transporters both in FCD lesions and in hippocampi were significantly enhanced compared to controls.

Conclusion: Prenatal freeze-lesioning of the brain during embryonic development produces a severe disorder of neuronal migration with increased excitability via glutamatergic receptors, and leads to spontaneous temporal lobe seizures in a rat model with multiple FCDs.

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Abstract—WCN 2013

No: 543

Topic: 1—Epilepsy

Quality of life in psychogenic nonepileptic seizures

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Background: Psychogenic non-epileptic seizures (PNES) are events resembling epileptic seizures (ES), which are not associated with ictal electrical discharges in the brain but by psychological processes.

Objective: In this study, we compared health-related quality of life (HRQOL) in patients with PNES with that of patients with ESs.

Methods: We evaluated 28 patients admitted to the Health Center between January 20, 2010, and January 20, 2012. 14 patients with epilepsy and 14 patients with PNES were recruited. Only patients with the definite diagnosis of ESs or PNESs were analyzed. All participants with ES were prescribed and actively taking antiepileptic medication(s). Patients completed an epilepsy-specific quality-of-life instrument (QOLIE-31), Beck Depression Inventory—II (BDI-II), Adverse Events Profile (AEP) and versions of the illness perception questionnaire revised (IPQ-R) adapted for epileptic or nonepileptic seizure disorders.

Results: Patients with PNES reported poorer HRQOL and greater depressive symptoms compared to ES participants. The depression and medication side effects have been shown to be important contributors to quality of life in patients with PNES. The overall HRQOL and scores on 13

of 19 QOLIE-31 subscales were significantly lower in PNES than in ES patients. AEP also were worse in PNES patients than in ES patients.

Conclusion: Patients with PNESs have a lower HRQOL and worse mood problems than do patients with ESs. This disadvantage is primarily due to depression and medication side effects. This result has important implications for psychotherapeutic treatment of PNES in that it provides a potentially modifiable target.

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Abstract—WCN 2013

No: 550

Topic: 1—Epilepsy

Epilepsy: A global cry...raising up the volume

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Background: Epilepsy is the most common serious neurological disorder and is one of the world's most prevalent noncommunicable diseases. Around 90% of people with epilepsy in developing countries are not receiving appropriate treatment.

Objectives: Epilepsy is common in sub-Saharan Africa but is poorly characterized. Most studies are hospital-based, and may not reflect the situation in areas with limited access to medical care. Examination was done, to determine if the clinical features could help elucidate the causes.

Patients and Methods: We conducted a detailed descriptive analysis of 445 epileptic patients identified through a community-based survey of 151,408 people in Kenya, including the examination of electroencephalograms.

Results: Approximately half of the 445 people were children aged 6 to 18 years. Seizures began in childhood in 78% of those diagnosed. An episode of status epilepticus was recalled by 36% cases, with an episode of status epilepticus precipitated by fever in 26%. Overall 169 had an abnormal electroencephalogram, 29% had focal features, and 34% had epileptiform activity. In the 146 individuals who reported generalized tonic-clonic seizures, only 22% had focal features on their electroencephalogram. Overall 71% of patients had evidence of focal abnormality, documented by partial onset seizures, focal neurologic deficits, or focal abnormalities on the electroencephalogram.

Conclusion: Children and adolescents bear the brunt of epilepsy in Africa Low Income settings. The predominance of focal features and the high proportion of patients with status epilepticus, suggest that much of the epilepsy in this region has identifiable causes, many of which could be prevented.

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Abstract—WCN 2013

No: 570

Topic: 1—Epilepsy

Status epilepticus in thrombotic thrombocytopenic purpura: A case report

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Background: Thrombotic thrombocytopenic purpura (TTP) is a condition characterized by an ischemic vasculopathy frequently associated with fluctuating neurologic symptoms, including confusion, stupor, and seizures.

Objective and methods: We describe a 30 year old man with TTP who developed seizures not well controlled with intravenous midazolam and oxcarbazepine. He had a progressive neurological

deterioration and EEG showed a non convulsive status epilepticus (NCSE). He was treated in intensive care unit with intravenous levetiracetam (LEV) after midazolam bolus for 48 than switched to oral therapy (total dose of LEV 3000 mg/die), in association with plasma-exchange and Rituximab. His neurological status improved in few weeks and he had a complete neurological recovery. He stopped oral LEV six months after hospitalization.

Conclusion: Fluctuating stupor in TTP has generally been attributed to microvascular occlusive disease, but NCSE is a treatable condition that can cause similar symptoms. Aggressive treatment with rituximab, plasma-exchange and antiepileptic drugs may prevent permanent neurologic deficits.

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Abstract—WCN 2013

No: 595

Topic: 1—Epilepsy

Role of polymorphism (rs2986017) and (rs11191692) in the calcium homeostasis modulator 1 (CALHM1) gene in temporal lobe epilepsy

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Background: Calcium homeostatic mechanisms play an important well known role in the development and maintenance of epilepsy in general.

Objectives: Investigate the role of calcium homeostasis modulator 1 gene, SNPs; rs2986017 and rs11191692 in temporal lobe epilepsy in Egyptian patients and its influence on clinical manifestations.

Patients and methods: A case-control study conducted on 55 Egyptian unrelated temporal lobe epilepsy patients and 31 healthy Egyptian control subjects, age and sex matched. Neurological examination, electroencephalogram and brain MRI were done to all patients to confirm temporal lobe epilepsy. Two single nucleotide polymorphisms of calcium homeostasis modulator 1 gene, rs2986017 and rs11191692 were genotyped using PCR-RFLP for both studied groups.

Results: The genotype and allele frequencies for SNPs; rs2986017 and rs11191692, did not differ between TLE patients and control group (P value = 0.435, 0.884, 0.621 and 0.690 respectively). Regarding SNP rs2986017, the CC genotype and the C allele were the most frequent while the GG genotype and G allele were the most frequent among SNP rs11191692 in both patients and controls. A significantly positive correlation between EEG changes and allele frequency of CALHM1 gene SNP rs11191692 was detected with P value = 0.034 but no correlation with CALHM1 gene SNP rs2986017 (P value = 0.369).

Conclusion: Our study did not support the hypothesis that CALHM1 SNP rs2986017 or rs11191692 is associated with the risk of TLE. Elucidation of the precise mechanism requires further functional study as well as genetic analysis in independent population samples and on larger scales.

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Abstract—WCN 2013

No: 622

Topic: 1—Epilepsy

Cerebral mechanisms of antiepileptic defense

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Background: Not every patient, even one with the most severe manifestation of epilepsy, status epilepticus, died in the pre-antiepileptic

drug era. Evidently, there are some cerebral mechanisms of antiepileptic defense.

Objective: To study the neurophysiological mechanisms of antiepileptic brain defense.

Material and methods: The data is based on 50-year-long investigations: 150 adult patients with convulsive status epilepticus (CSE) and a group of 100 patients, children and adults, with various forms of epilepsy with EEG absence patterns. Clinical investigation, EEG, including the MDL method, CT/MRI and an assessment of homeostatic indices was used. The study involved experiments on animals that were focused on creating acute and chronic foci in sensorimotor cortex and subsequent stimulation and destruction of orbitofrontal cortex. The cooling of the epileptogenic foci resistant to AEDs was examined.

Results: Stimulation of the orbitofrontal cortex suppresses epileptogenesis, while its destruction speeds it up. In patients with CSE, postspike slow wave transforms the life-threatening tonic phase of a fit into a more favorable clonic phase. A separate analysis of spike and inhibitory postspike slow wave by the MDL method showed a difference in their localization. Unlike spike, the slow wave, in 81% of patients, was localized in the orbitofrontal cortex and the frontal part of the gyrus cinguli. The cooling of the epileptogenic zone restored its sensitivity to AEDs.

Conclusion: The epileptic brain creates an antiepileptic system; its key elements are the orbitofrontal cortex and the frontal part of the gyrus cinguli; its basic neurophysiological mechanism is slow wave.

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Abstract — WCN 2013

No: 645

Topic: 1 — Epilepsy

Ketogenic diet efficacy in children with Lennox–Gastaut Syndrome

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Purpose: To review the efficacy and tolerability of a ketogenic diet (KD) in children with Lennox–Gastaut Syndrome (LGS).

Methods: Children initiated on KD for LGS from January 2009 through December 2012 were identified from the KD diet database at Mayo Clinic Rochester. Charts were reviewed to determine seizure frequency (a) prior to KD onset, (b) 3 months after KD onset and (c) at final follow-up on the KD, as well as concurrent anti-epileptic medication (AED), KD side effects, and epilepsy etiology. Significant seizure reduction was defined as >50% decrease in seizures.

Results: Nineteen children were identified (10 females). Etiologies included: genetic (3), structural (3), genetic and structural (3), unknown (10). Mean age at seizure onset was 18 months (range 0–74) and at KD initiation 54 months (range 7–166). At initiation, children were on a mean of 2.5 AEDs (range 2–4) and had previously failed 2.3 (range 0–8) AEDs for lack of efficacy. Mean duration of KD was 25 months (range 1–89).

At 3 months, significant seizure reduction was seen in 11/19; 2/3 genetic, 7/10 unknown, 1/3 structural, and 1/3 both genetic and structural etiologies. At final follow-up, 9/11 continued to have significant seizure reduction. Two discontinued diet due to recurrent vomiting, 5 had mild adverse effects (constipation, mild weight loss).

Conclusions: The KD was well tolerated and provided improved seizure control for the majority of children with Lennox Gastaut syndrome due to genetic or unknown etiology. Those with structural etiology may be less likely to respond to ketogenic diet.

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Abstract — WCN 2013

No: 647

Topic: 1 — Epilepsy

Features of myoclonic epilepsy of adolescence

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Background: myoclonic epilepsy of adolescence (MEA) is an idiopathic form of epilepsy, with occurrence of up to 11–12% of all epilepsy forms.

Material and methods: 42 clinical cases were analyzed (12 males, 30 females) using the psycho-neurological, neurophysiological, neuroimaging examination.

Results: The average onset of the disease was at 13.2 ± 0.5 years. The leading clinical symptom was epileptic myoclonus in various muscle groups. 5 (7.1%) patients' had also generalized seizure paroxysms, 2 (4.7%) patients developed absences. All patients experienced fits in the morning. The provocation factors were sudden awakening, sleep deprivation, alcohol consumption and stress. 2 patients had positive reflexes of oral automatism, myoclonic jerks in face, the rest of patients had no neurological deficiency out of epi-paroxysms. EEG showed the appearance of short discharges or generalized polyspikes or polysharp-wave patterns. The valproic acid therapy induced remission in 18 patients, 21 patients developed positive dynamics with reduction of myoclonic episodes and generalized seizure paroxysms frequency, 2 patients developed the disease aggravation, one showed no improvement.

Conclusions: Female patients dominate (2:1). The isolated myoclonic attacks prevailed, there was a combination with generalized seizure paroxysms and absences in 11.8% of cases only. Monotherapy with valproic acid medications was effective in major part of MEA cases; in certain cases aggravation of the disease is possible, that is a reason for dose reduction or medication substitution.

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Abstract — WCN 2013

No: 677

Topic: 1 — Epilepsy

The possibilities of neuroimaging in diagnostics of symptomatic epilepsy in children

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Background: Epilepsy is one of the most complex medical and social problems at present time. The high prevalence of the disease in paediatric population of Uzbekistan (10 per 1000) determines the importance to develop effective measures for early diagnosis, new approaches to correction of treatment and prevention of complications of epilepsy.

Objective: To identify MRI findings of symptomatic epilepsy in children with inflammatory etiology such as meningoencephalitis.

Materials and methods: MRI studies were conducted with 35 children with the diagnosis of symptomatic epilepsy after meningoencephalitis. Children ages ranged from 1 year to 14 years.

Results: in our study the main symptoms of epilepsy after meningoencephalitis were multiple lesions of white and gray matter, their predominant bilaterality and symmetry, a clear demarcation from the surrounding tissues. In the study of 35 children who recovered from meningoencephalitis following MRI signs were found, in 5 (14.3%) cases it was midline shift of the brain, in 12 (34.3%) cases it was asymmetry of the lateral ventricles. Subarachnoid perivascular space expansion was found in 22 (62.9%) cases, which often revealed in the fronto-temporal region of the

brain. Expansion of the subarachnoid space was revealed in 19 (54.3%) cases, mainly due to atrophy of the brain.

Conclusion: Magnetic resonance imaging has an important role in the clinical diagnostics of epilepsy, the use of which in the study of symptomatic epilepsy is one of the important conditions of adequate diagnosis, treatment and prognosis of the disease.

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Abstract – WCN 2013

No: 696

Topic: 1 – Epilepsy

Effect of AED on the EEG pattern in epileptic children

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Background: The value of qEEG analysis as predicting seizure exacerbation in children with new onset epilepsy is important. The aim of the work was to study the dynamics of EEG characteristics in epileptic children at different stages of antiepileptic drug (CBZ) monotherapy.

Objective: 78 epileptic children (4–10 years old, Males – 41, Females – 37) were analyzed. All patients have partial seizures with or without secondary generalization. The EEG performed in three times: at the first visit (before the administration of CBZ) and in 3–4 and 6–8 months after the beginning of the treatment.

Methods: Following characteristics of EEG were analyzed: absolute values of the power spectra (AVP), EEG mapping-topography and qualitative characteristics. The recordings were performed on wake state in the same time.

Results: The quantitative analysis of AVP dynamic revealed a reliable increase of this index 3 months after the beginning of the treatment. Elevation remained after 6 months of treatment. This effect most expressed in low frequency bands. CBZ therapy showed decreases an average frequency of alpha waves. The qualitative analysis revealed decrease in the density of spontaneous epileptiform graphoelements and generalized epileptiform bursts.

Conclusion: Any type of AED should be accomplished with maximal caution and under regular EEG control, due to in some cases worsening of EEG revealed (predict) before the onset of clinical signs of exacerbation.

The study of EEG pattern of background activity, spectral analysis, EEG mapping (topography) and qualitative analysis using qEEG approach during AED therapy allows to determine correctly the treatment strategy.

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Abstract – WCN 2013

No: 734

Topic: 1 – Epilepsy

Telemedicine: A great potential to be explored for epilepsy – Literature review

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Background: Telemedicine can be broadly defined as the use of telecommunications technologies to provide medical information and services. Although this definition includes medical uses of the

telephone, facsimile, and distance education, telemedicine is increasingly being used as shorthand for remote electronic clinical consultation. This technology facilitates early diagnosis in epilepsy, improves prognosis, and enhances patient care.

Objective: To review the literature that demonstrates the potential use of telemedicine in epilepsy diagnosis on emergency medical services, ambulatory follow-up, patient care and tele-education.

Material and methods: PubMed and Bireme databases and Scielo virtual library were used to select bibliography.

Results: Telemedicine enables the realization on medical actions at a distance through audiovisual and data communication, highlighting the teleconsulting. In primary health care, several physicians hold poor knowledge about epilepsy and new drugs, and that is why teleconsulting is an important tool. The insufficient number of neurologists requires training of general practitioners through tele-education. Residents of remote areas can be screened at a distance, reducing costs and relieving the centers. Patients with epilepsy should adhere to treatment, or orient themselves and learn to live with the disease, and informative sites and virtual communities can be helpful.

Conclusion: A Brazilian project facing epilepsy has great chances of achieving good results, as in other countries and even in Brazil as the Stroke Care National Project.

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Abstract – WCN 2013

No: 846

Topic: 1 – Epilepsy

EEG findings in post-stroke seizures

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Background: Stroke is an important cause of epilepsy especially in elderly. EEG findings are generally not considered to be very helpful for the diagnosis of post-stroke seizures.

Purpose: The aim of the study was to describe the EEG characteristics in patients who developed seizures after stroke.

Patients and methods: The study population consisted in 130 patients with ischemic or hemorrhagic first-ever stroke admitted at our Service from March to December 2010. Patients with SAH, Cerebral Vein Thrombosis and previous history of seizures were excluded from the study. All the patients underwent brain CT scan or MRI. 17 patients with post-stroke seizures had an interictal EEG after the first seizure. EEG electrodes were placed according to the International 10–20% system. Activation procedures such as hyperventilation and photic stimulation were done when it was possible.

Results: Ischemic stroke was diagnosed in 94 (72%) patients and hemorrhagic stroke in 36 (27.6%) patients divided in 76 (58.4%) males and 54 (41.5%) females and mean age 68 +/- 12 years. Periodic lateralized epileptic discharges (PLEDs) on the EEG after stroke were found in one patient, (5.8%). Frontal intermittent rhythmic delta activities (FIRDAs) were observed in 4 patients (23.5%). Diffuse slowing was found in 6 patients, 35.3% of the seizure group. Normal EEG findings were seen in 3 patients, 11.8% of the post-stroke seizures patients. Focal slowing was found in 3 patients, 17.6 % of seizures group.

Conclusion: FIRDAs and diffuse slowing are the most frequent EEG findings in patients who developed seizures after stroke.

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Abstract – WCN 2013**No: 856****Topic: 1 – Epilepsy****Clinical presentation and neurophysiological findings in patients with pineal region expansions – Our 17 year experience**

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Background: In the neurological and neurosurgical follow up of our patients with pineal region expansions, we have noticed certain clinical and neurophysiological regularities.

Objective: To perform retrospective study including 84 patients with pineal region expansions in the period from 1992 to 2009.

Patients and methods: The study included 55 women and 29 men. All patients had headache, while 32 patients (38%) had primary generalized seizures. Common EEG pattern showed paroxysmal discharges of 3 or more Hz spike-and-wave complexes. Brain MRI showed in 70 patients (83.4%) cysts, and 14 patients (16.67%) had expansive process with compressive effect. Based on the size of the cyst (15 mm or more) and signs of compression on the quadrigeminal plate and of the surrounding veins operation was performed in 70 patients.

Results: Pathohistological analysis revealed pineocytomas in 11 cases (15.71%), pinealoblastomas in 2 cases (2.86%), one case of teratoma (1.43%), while 56 patients had cysts (80%). Following surgery clinical condition improved in all patients – patients became seizure-free and headaches significantly decreased, and other symptoms (diplopie, nausea, vomiting, vertigo and blurred vision) disappeared.

Conclusion: Headache and also primary generalized seizures are often present in patients with pineal region expansions. Mass effect on the surrounding veins and compression on the quadrigeminal plate and the aqueduct, together with hemosiderin deposits can be involved in the pathogenesis of seizures. We suggest to perform high resolution brain MRI in all young patients that have seizures and specific EEG changes.

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Abstract – WCN 2013**No: 474****Topic: 1 – Epilepsy****Common reference-based indirect comparison meta-analysis of intravenous valproate versus intravenous phenobarbitone in generalized convulsive status epilepticus**

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Background: In the literature, only one randomized control trial (RCT) directly compared intravenous valproate (IV VPA) with intravenous phenobarbitone (IV PB) in the treatment of established generalized convulsive status epilepticus (GCSE).

Objective: To compare IV VPA with IV PB in the treatment of GCSE in patients of any age, indirectly estimating their efficacy and safety through a common-reference based indirect comparison meta-analysis (CRBMA) and to evaluate whether CRBMA is reliable and consistent with results of direct head-to-head RCTs.

Material and methods: RCTs of IV VPA and IV PB versus IV PHT for GCSE were systematically searched. A random effects model was used to estimate Mantel-Haenszel odds ratios for efficacy and safety of IV VPA versus IV PHT in a standard meta-analysis. Adjusted

indirect comparisons were then made between VPA and PB using the obtained results.

Results: The CRBMA showed that, compared with PB, VPA does not lead to significantly higher seizure cessation (OR 1.00; 95% CI 0.36–2.76), although it has lower adverse effects (OR 0.17; 95% CI 0.04–0.71). Results of CRBMA are consistent with those of a recently published head-to-head comparison between IV VPA and IV PB.

Conclusion: There is no evidence supporting a superiority of IV VPA over IV PB for the treatment of GCSE in terms of efficacy. Some data derived from direct and indirect comparisons suggest that VPA has a better safety profile than PB. In the absence of comparative clinical trials studies, a CRBMA provides some evidence of the relative efficacy and safety of competing AEDs.

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Abstract – WCN 2013**No: 899****Topic: 1 – Epilepsy****Anticonvulsant actions of Ska-31 in pilocarpine treated chronic epileptic rats**

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Purpose: Pilocarpine-induced status epilepticus or limbic seizure model is used to test activity of new molecules with anticonvulsant potential. In the present study we focused on anticonvulsant effect of SKA-31 (SK-channel opener) in pilocarpine-induced epileptiform activities.

Methods: Male Wistar rats (4–5 weeks) were pre-treated with methylscopolamine (1 mg/kg) followed by i.p. administration of pilocarpine (340 mg/kg). 90 min post status epilepticus, diazepam is injected intraperitoneally (10 mg/kg). Animals were observed under video recording for about 5–6 days. Age-matched pilocarpine control group received same protocol except saline instead of hippocampal slices (400 μM) from pilocarpine treated and pilocarpine control rats were prepared. Age-matched control rats were used that received saline instead of pilocarpine. To induce epileptiform activities in slices 4-aminopyridine (4-AP, 100 μM) is used. Local field potential recordings were performed from medial entorhinal cortex using glass microelectrode filled with ACSF.

Results: SKA-31 at 150 and 50 μM blocked SLE in 87% and none of slices respectively. Whereas, in control slices blockade of seizures is observed in 100% of slices. It is noticed that SKA-31 significantly decreased duration of SLEs in 23 ± 4 min of application as well as reduction in amplitude of SLEs. Whereas, complete seizure suppression is achieved in 33 ± 2 min.

Conclusion: These results suggest that SKA-31 suppressed ongoing propagation of epileptiform events in chronically treated pilocarpine rats.

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Abstract – WCN 2013**No: 908****Topic: 1 – Epilepsy****American Clinical Neurophysiology Society's standardized critical care EEG terminology: Interrater reliability and 2012 version**

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Background: Critical care EEG (ccEEG) monitoring has gained widespread use. However, uniformly accepted nomenclature for EEG patterns encountered in these patients has been lacking.

Objective: A group of electroencephalographers from the American Clinical Neurophysiology Society and the Critical Care EEG Monitoring Research Consortium developed standardized ccEEG terminology to describe rhythmic or periodic EEG patterns, avoiding terms with clinical connotations e.g. “triphase waves”.

Results: Rhythmic or periodic patterns are characterized by a Main term #1 to describe pattern location (Generalized, Lateralized, Bilateral Independent or Multifocal), followed by Main term #2 to describe pattern type (Periodic Discharges (PDs), Rhythmic Delta Activity (RDA), or Spike/Sharp-and-wave). Additional modifiers such as Prevalence, Duration, Frequency, Amplitude and Sharpness are added.

Another key modifier is the “Plus” term, indicating patterns which are more ictal-appearing. For PDs this includes superimposed fast or rhythmic activity and for RDA, fast activity or intermixed sharp waves or spikes. Characterization of background activity and standardized quantification of sporadic epileptiform discharges is also included.

Analysis of interrater agreement (IRA) for a 2010 version of this terminology showed good IRA for main terms 1 and 2 (Kappa values 0.87 and 0.92, respectively) and moderate IRA for “plus” modifiers. Further IRA analysis of the current version is underway. An online training module and neonatal version have been completed; certification testing and incorporation into the “SCORE” system are underway.

Conclusion: Adoption of standardized critical care EEG terminology will aid in communication and facilitate multicenter research in order to study the significance of these complex patterns.

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Abstract – WCN 2013

No: 964

Topic: 1 – Epilepsy

The neurological status of children with epileptic encephalopathy

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Background: Epileptic encephalopathy is a condition in which the pathologically changed electrogenesis of the brain causes disorders of the brain. The epileptic process itself leads to a progressive disorder of brain function. It is difficult to make correct diagnosis of neurological status of these patients because of they do not have specific complaints of seizures, although in some cases the survey can reveal seizures in patient history.

Objective: Main purpose was to study the features of neurological status in children with epileptic encephalopathy.

Material and methods: We studied 69 children aged from 3 to 14 years with a diagnosis of epileptic encephalopathy. In this study we used clinical, neurological and instrumental methods such as electroencephalogram (EEG).

Results: In the study of neurological status of patients with epileptic encephalopathy it was revealed the predominance of cognitive impairment, decreased intelligence, memory and thinking children. In the study of cognitive functions of children with epileptic encephalopathy Type 1 it was observed more severe intellectual disorders compared to patients with epileptic encephalopathy type 2. According to the study of analysis of electroencephalogram in children with epileptic encephalopathy it was revealed focus of epiactivities in brain without morphological changes. Finally we would like to emphasize that the EEG markers of epileptic encephalopathy in children are fundamentally different from normal EEG of children in different ages.

Conclusion: Thus, a detailed clinical and neurological analysis correlated with EEG data allowed to exclude errors in diagnostics of epileptic encephalopathy in children.

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Abstract – WCN 2013

No: 659

Topic: 1 – Epilepsy

Assessment of connectivity strength between the primary focus and remote cortical fdg pet abnormalities in children with non-lesional epilepsy

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Objective: Our objective was to quantitatively assess the connectivity strength (based on probabilistic DTI analysis) of fiber tracts that originate in the seizure onset region (as defined based on intracranial ECoG) and that project to remote cortical areas characterized by F18-deoxyglucose (FDG) PET abnormalities that are electrophysiologically normal.

Method: We developed a computational framework that allows quantitative assessment of the spatial relationship between multi-modality neuroimaging data (MRI, DTI, PET, and ECoG). The framework is based on the parcellation of the cortical surface in native space using landmark-constrained conformal mapping, which yields finite cortical elements (FCEs) that are homotopic across subjects. The FCEs were subsequently used as source/target regions for probabilistic fiber tracking (55 direction), allowing the calculation of a connectivity score. We applied this approach to 7 young children (3–12 years) with non-lesional epilepsy who underwent presurgical evaluation and compared the obtained connectivity pattern against 12 age-matched normal children.

Results: Only few FDG PET abnormalities were determined within the ECoG confirmed seizure onset area, although FDG PET abnormalities were frequently observed adjacent to this area, most likely as a result of functional disconnect of this area from ictal involvement. Connectivity analysis showed a decreased connectivity score (3–9%) in epilepsy patients compared to the normal group. Moreover, significant abnormalities in the connectivity pattern between the seizure onset and other cortical lobes were determined.

Conclusion: Our findings indicate that the connectivity strength between the primary focus and remote FDG PET abnormalities is decreased as compared to a normative group.

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Abstract — WCN 2013**No: 1016****Topic: 1 — Epilepsy****Infantile spasm in children: Clinical features and outcome**

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Objective: Clinical features and outcome of children with infantile spasms.

Study design: Interventional and observational study.

Place and duration of study: The Department of Neurology, Children's Hospital, Lahore, Pakistan, from April 2008 to March 2013.

Methodology: Children aged <2 years presented with history of infantile spasms were assessed. Clinical presentation, EEG findings and response of anti-epileptic drugs was analyzed.

Results: We enrolled 450 infants with infantile spasms at their first presentation. Out of 450 children, 76% presented at age of <6 month, 72% presented due to infantile spasms and 18% because of global developmental delay. Spasm types were mixed (38%), flexors (44%), extensor (16%) and asymmetric (2%). Hypsarrhythmia (67%) was the predominant EEG finding followed by modified hypsarrhythmia (24%) and other forms of epileptic discharges in 9% children. Majority of children were receiving oral Phenobarbitone, Carbamazepine or Valproate sodium. We initiate the management with oral Prednisolone followed by Clonazepam or valproate acid. ACTH therapy was administered in only 5 children.

Conclusion: Infantile spasms are one of the refractory epilepsy in children. Abnormal EEG findings predominantly the hypsarrhythmia or modified hypsarrhythmia are the hallmark. Majority of children received conventional AED with poor response. Oral prednisolone is proved to be the most effective AED. These children should be referred to the tertiary care pediatric neurology centers.

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Abstract — WCN 2013**No: 911****Topic: 1 — Epilepsy****Continuous EEG in critically ill patients: Experience at a single tertiary care center**

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Background: Continuous EEG (cEEG) can provide critical information about brain function including detection of causes of secondary injury such as seizures. Although recent advances have led to widespread use of cEEG in this population, there is limited data on current clinical practices.

Objective: To describe clinical findings in critically ill patients undergoing cEEG at a tertiary care center over a three year period.

Patients and methods: We retrospectively reviewed data from 1305 cEEG monitoring sessions from 2009 to 2011. Patients were selected to undergo cEEG at the discretion of the treating physician. Indications for monitoring, EEG findings, neurologic diagnosis and outcome at discharge were assessed.

Results: The primary indication for cEEG was the detection of subclinical seizures (72.3%), followed by characterization of spells (18.5%). The three most common neurologic diagnoses were subarachnoid hemorrhage (16.5%), altered mental status (16.0%) and new-onset seizure (10.1%). Seizures were reported in 17.9% (216/1202 patient encounters) and 51% (110/216) were exclusively electrographic. High rates of nonconvulsive seizures were seen in patients with CNS infection (36.4%, n = 4/11), CNS neoplasm (16.4%, n = 11/67), vascular malformation (16.7%, n = 1/6) and intracerebral hemorrhage (10.4%, n = 10/96). Good outcome (discharge to home or rehab) was seen in

46% of all patient encounters and 48% of patients who experienced seizures during cEEG.

Conclusion: Subclinical seizures are common in critically ill patients with a variety of underlying neurologic diagnoses. Prospective studies are needed to determine the incidence and predictors of seizures in a non-selected population as well as the impact of seizures on outcome.

doi:10.1016/j.jns.2013.07.070

Abstract — WCN 2013**No: 1045****Topic: 1 — Epilepsy****An epidemic of seizures and psychosis in a sudanese village — A challenging experience**

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Background: In January 2010, a wave of pathological laughter, crying and bizarre behavior affected about 122 people in a remote village in the far North West State of Kordofan in Sudan.

Patients, methods and results: Detailed investigation of the clinical presentations and possible underlying causes was done with provision of medical treatment.

Children constituted 52% and were more severely affected than adults. The main presentations were visual hallucinations, uncontrolled laughter, twisting movements, delirium, and convulsions. No vascular manifestations were detected. Males were affected more than females (60%).

In a few of the severely affected patients a lumbar puncture was performed (7/122). The patients were treated symptomatically with benzodiazepines. Carbamazepine was used in those presenting with recurrent seizures. Routine urine, blood and CSF basic parameters were within normal limits for routines, but the toxicology screen of urine missed the critical period for detection of the suspected toxic substances. Samples from the water sources were clear, but the wheat consumed by the villagers grew the fungus *Claviceps purpurea* in abundance. Further tests on the fungi revealed their production of very high level of LSD-like ergot alkaloids. No long term neurological sequelae were noticed on follow up.

The wheat came from stores in Darfur which is near the affected village. The epidemic was contained and a public education campaign was launched to avoid recurrence of the event.

This paper includes videos and a literature review.

Conclusions: This study draws attention to the importance of vigilance about neurotoxins as causes of bizarre presentations.

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Abstract — WCN 2013**No: 1047****Topic: 1 — Epilepsy****The sensitivity of the long term eeg and sleep eeg in the diagnosis of epilepsy**

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Background: Long term video-EEG monitorization (LTEEG) is a very valuable diagnostic tool in the differential diagnosis of epileptic and non-epileptic seizures, in the classification of epileptic seizures and in the evaluation of surgical candidacy in patients with medically refractory seizure disorders. When there is diagnostic uncertainty as to the diagnosis of epilepsy, LTEEG is used to detect interictal epileptiform abnormalities. Sleep-EEG may also reveal interictal epileptiform abnormalities in patients with an initial negative routine EEG.

Objective: We aimed to compare the sensitivity of the LTEEG and sleep-EEG in the diagnosis of epilepsy.

Patients and methods: We recruited 29 adult patients with epilepsy, hospitalized at least five days in the LTEEG laboratory without any observed clinical events during the test. We performed a sleep-EEG study of 30 min in the following week and we compared the result of the LTEEG with the result of the sleep-EEG.

Results: The results of the LTEEG and the sleep-EEG were same in 28 of the 29 patients (96%). In only one patient, generalized interictal epileptiform abnormalities were observed in LTEEG whereas sleep-EEG was normal.

Conclusion: In patients with epilepsy, sleep-EEG is as sensitive as the LTEEG to detect the interictal epileptiform abnormalities. While LTEEG will remain the gold standard for confirming the electrophysiology underlying a given event, sleep-EEG represents a practical and cost-effective alternative in the diagnosis of epilepsy.

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Abstract – WCN 2013

No: 1060

Topic: 1 – Epilepsy

White-matter tract integrity and seizure propagation in non-lesional temporal lobe epilepsy

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Background: Non-lesional temporal lobe epilepsy (TLE) patients refractory to antiepileptic drugs are candidates of presurgical evaluation. We explored diffusion-tensor imaging (DTI) and its utility in elucidating the white-matter tract integrity in relation to seizure-spreading and prolongation.

Methods: Fourteen patients with electrographic unilateral non-lesional TLE (7 left, 7 right) and sixteen normal subjects underwent DTI with 31 diffusion-gradient directions followed by computation of fractional anisotropy (FA)-values by voxel-based morphometry (VBM) and construction of two major temporal-lobe tracts: inferior-longitudinal-fasciculi (ILF) and uncinete-fasciculi (UF). Tract-based-spatial statistics (TBSS) offered calculation of FA-values for each tract using automated VBM-database. The averaged duration of hemispheric seizure-involvement was calculated for each temporal region of left/right TLE-groups.

Results: The FA-values of ILF and UF were significantly different between the temporal lobes in normal subjects ($p < 0.005$) and in non-lesional TLE patients ($p < 0.005$). Using ILF, both left/right TLE patients had significantly different ipsilateral FA-values compared with normal subjects ($p < 0.006$) and similar results were found with UF. Left TLE patients had significantly different ipsilateral FA-values in ILF when compared with right TLE patients (ipsilateral-higher-FA) ($p < 0.006$) and similar results were found with UF (ipsilateral-lower-FA). Using UF, multiple linear-regression on right TLE patients showed a significant correlation between duration of ipsilateral-onset and ipsilateral FA-values ($R^2 = 0.69$, $p = 0.02$) and a similar result for duration of contralateral-spreading and contralateral FA-values ($R^2 = 0.6$, $p = 0.04$). Corresponding analyses on left TLE with UF, or that using ILF, were not significant.

Conclusions: Left/right non-lesional TLE patients were functionally different categories and their side-to-side difference may be demonstrated on DTI. Frontal-temporal connections may underlie the association between seizure-spreading/prolongation and white-matter tract integrity in this group of patients.

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Abstract – WCN 2013

No: 813

Topic: 1 – Epilepsy

The study of mental disorders in children with age related epilepsy with centro-temporal (Rolandic) spikes

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Purpose: To examine the association between age at onset, massiveness of Rolandic spikes in sleep EEG, their localization in the beginning of the process, migration of spikes through the cerebral cortex during the follow-up and to examine mental disorders in children to address the question of the appropriateness of anti-epileptic therapy and its duration.

Material and methods: Children with early onset "Rolandic" epilepsy (up to 3) years, 10 people, with the debut of the children of 4–7 years, 10 people, and children with late onset Rolandic epilepsy (over 7 years) also 10. These children did not show any clinical neuroimaging and neurological disorders.

Evaluation of mental disorders (including speech) was carried out at the first call, and every year before Rolandic spikes reduction in EEG. In addition, academic performance (in school) was evaluated.

Results: For the younger age group in the first place on the representation of mental disorders detected tempo delayed development of speech skills, the second highest representation of symptoms in these children was ADHD; in the middle age group ADHD reaches a degree of psychopathic disorders, while speech disturbances were mild or not diagnosed. In the older age group mental disorders should not go beyond the mild forms, without changing the school and social adaptability.

Conclusions: Children with age related epilepsy with Rolandic spikes at an early onset are more in need of anti-epileptic therapy, which is pathogenic for such mental disorders.

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Abstract – WCN 2013

No: 837

Topic: 1 – Epilepsy

Clinical features of epileptic attacks in patients with acute stroke and rational approaches to its treatment

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Goal: To study epileptic attacks clinical features in patients with acute stroke and to develop approaches to rational therapy.

Materials and methods: Comprehensively examined 78 patients with acute ischemic stroke: men – 46 (59%), 32 (41%) – women (44 to 78 years). Neurological examination performed by standard technique using instrumental investigation methods – electroencephalography, Doppler ultrasound of head main arteries, and brain computer tomography.

Results: The analysis showed: the left and right hemisphere strokes were equally often, 37.4% and 39.1%, respectively, in 13% of patients with stroke in vertebrobasilar basin, 10.5% patients suffered from recurring cerebral stroke. 16% of patients had auras, in 22 (28.2%), patients epileptic attacks developed in stroke debut, in 7 days after stroke epileptic attacks were observed in 28 (35.9%) patients. 68 (87.2%) patients had the prevalence of focal attacks, and 15 (22%) had simple partial attacks, 21 (30.9%) patients – the second-generalized, and only 20 (29.4%) – polymorphic partial attacks. The status epilepticus developed in 2 (2.9%) cases at stroke debut. Primary generalized attacks were only observed in 7 (10.3%) patients.

The foci of epileptiform activity were revealed in 22 (28.2%) patients, and 56 (71.8%) patients had centers of slow-wave activity. Left-sided localization of focal activity predominated in 48 (61.5%) cases. 26 (33.3%) cases revealed a tendency to predominance of critical stenosis and occlusion of left carotid.

Conclusions: Rational approaches to treatment of these patients will improve the quality of life, and social and family adaptation.

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Abstract – WCN 2013

No: 1069

Topic: 1 – Epilepsy

Autosomal recessive cortical myoclonic tremor and epilepsy: Association with a mutation in the potassium channel associated gene *CNTN2*

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We characterize a consanguineous Egyptian family with an autosomal recessively inherited familial cortical myoclonic tremor and epilepsy. We used multipoint linkage analysis to map the causative mutation to a 12.7 megabases interval within *1q31.3–q32.2* with a LOD score of 3.6. For further investigation of the linked region in an efficient and unbiased manner, we performed exome sequencing. Within the suspected region we identified a homozygous single base pair deletion (c.503_503delG) leading to a frameshift in the coding region of the 6th exon of *CNTN2* alias *TAG-1* (p.Trp168fs), which segregated in the respective family. Many studies point towards an important role of the *CNTN2* product Contactin-2 in neuronal excitability. Contactin-2, a glycosylphosphatidylinositol-anchored neuronal membrane protein, and another transmembrane protein called Contactin-associated protein-like 2 are together necessary to maintain voltage gated potassium channels at the juxtaparanodal region. *CNTN2* knockout mice were previously reported to suffer from spontaneous seizures and mutations in the gene encoding Contactin-associated protein-like 2 (*CNTNAP2* alias *CASPR2*) have been described to cause epilepsy in humans. To further delineate the role of *CNTN2* in epilepsy patients, we sequenced the coding exons in 189 Caucasian epilepsy patients. No recessive mutation was detected, and heterozygote carriers of rare *CNTN2* variants do not seem to be predisposed to epilepsy. Given the severity of the mutation and the proposed function of the gene, we consider this mutation as the most likely cause for cortical myoclonic tremor and epilepsy in this family.

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Abstract – WCN 2013

No: 1041

Topic: 1 – Epilepsy

Subjective handicap in people with epilepsy in Georgia and Turkey

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Background: People with epilepsy have an impaired quality of life due to the impact of the condition on various domains of their life.

Objective: To assess epilepsy-related subjective handicap in people with epilepsy in two neighboring countries: Georgia and Turkey.

Methods: We distributed self-administrated questionnaires to consecutive people with epilepsy taking antiepileptic drugs attending outpatient clinics at I.U. Cerrahpasa Medical Faculty in Istanbul and at the Institute of Neurology and Neuropsychology in Tbilisi. The SHE questionnaire was used which contains 32 items in six domains. Low score indicates higher handicap. Probabilities of <0.05 were considered as significant.

Results: A total of 200 people took part (equal number at each site). Mean age was 31.7 years. 62% of responders were female. 24% had completed higher education. 27% had a “Work and activities” score below 50 points, 13% for “Social and personal”, 28% for “Self perception”, 30% for “Physical”, 19% for “Life-satisfaction” and 23% for “Change” domains. In ‘Soc. personal’ (p = 0.003), ‘Life satisfaction’ (p = 0.042) and ‘Change’ (p = 0.006) subdivisions mean rank among people in Georgia were significantly lower except in “Social and personal” domain in which the Georgian cohort had higher scores. Association was found between social class and subdivision variables such as ‘Physical’ (p = 0.036), ‘Self perception’ (p = 0.001), ‘Life satisfaction’ (p = 0.001) and ‘Change’ (p < 0.001). People in lower social strata had lower scores.

Conclusion: Epilepsy imposes a high social burden on people in both countries with people with epilepsy in the lower strata of society may have a high burden.

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Abstract – WCN 2013

No: 1091

Topic: 1 – Epilepsy

Status epilepticus in glioma

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Background: Symptomatic epilepsy is common in glioma, especially in low grade glioma. There are few studies of status epilepticus (SE) in glioma, but they are assumed to be relatively infrequent and usually at the time of diagnosis or progression. We wanted to look at the semiology, relation to cancer disease, treatment response and recurrent SE in this population.

Material and methods: Patients with glioma WHO grades II–IV and ≥ 1 epileptic seizure at the time of diagnosis or later, have since 2009 been included in a prospective clinical observational study at Haukeland University Hospital. N = 61 per 04.03.2013. SE is defined as seizure >30 min or repeated seizures without restitution.

Results: We identified 17 SE in 13 glioma patients. Nine patients had glioma grade IV, one patient grade III and three patients grade II. The semiology of 11/17 SE was secondary generalized. 9/17 SE occurred within six months of diagnosis. In half of the cases, SE was onset symptom or sign of progression. In 10/17 antiepileptic drugs (AEDs) were already in use at the time of SE. Intravenous diazepam was sufficient in 10/17 SE. Midazolam was given in 2/17, one AED in 3/17 and two AEDs in 2/17, in addition to diazepam. General anesthesia was not discussed. Recurrent SE was seen in 4 patients.

Conclusion: SE was more frequent in high grade glioma and the seizures were secondary generalized. Half of SE occurred in early phase of cancer disease. All SE responded to first or second line treatment. Most patients had not recurrent SE.

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Abstract – WCN 2013**No: 1102****Topic: 1 – Epilepsy****Epilepsy and Parkinson: Frequent combination?***H. Stefan, J. Winkler. Dept. of Neurology, University Hospital Erlangen, Erlangen, Germany*

Background: Between 2000 and 2010 a cohort of 612 patients with Morbus Parkinson were registered. 21 of those suffered from epilepsy and Parkinson. Male = 15 patients, age 52–91 years, mean 74 years; female = 6 patients, age 64–88 years, mean 80 years. Etiology of epilepsies of all was analyzed.

Objective: Epidemiological studies indicate a strong increase of elderly patients. Compared to 1985 in 2030 an increase of 30% of the population in Germany will be older than 65 years is estimated. The question arises: Which comorbidities of neurological disorders, especially epilepsy and Parkinson occur because in special syndromes (FTDP-17) a frequent combination of both is described. How frequently patients with Morbus Parkinson also suffer from cryptogenic epilepsies (unknown etiology).

Patients and methods: Most patients suffered from symptomatic epilepsies due to ischemia or intracranial bleedings (n = 11), while only two patients suffered from cryptogenic focal epilepsies (0.32%).

Results: Our findings indicate a rare coincidence of Morbus Parkinson disease and cryptogenic epilepsies. This is in agreement with observations that occurrence of Parkinson decreased seizure frequency. Concerning seizure inhibition reduced GABAergic and dopaminergic mechanisms are reported.

Conclusion: Anticonvulsant action of dopamine is attributed especially to D2 receptor stimulation in the fore brain. Because epilepsy treatment by means of selective D2 agonists is beneficial in animal studies a well designed clinical trial is warranted to test its efficacy.

doi:10.1016/j.jns.2013.07.079

Abstract – WCN 2013**No: 1128****Topic: 1 – Epilepsy****Prescription patterns and self-reported side effects of antiepileptic drugs in patients with epilepsy at tertiary referral center in Austria***E. Pataraja, R. Jung, K. Trimmel, S. Aull-Watschinger. Medical University of Vienna, Vienna, Austria*

Objective: The aim of the present study was to examine the trends in prescribing antiepileptic drugs (AEDs), seizure frequency and self reported side effects in patients with epilepsy at the tertiary referral center in Austria.

Methods: 530 consecutive patients with epilepsy who were examined at the specialized epilepsy outpatient clinic of the Department of Neurology, Medical University of Vienna, were enrolled in the study. Demographic data, current AEDs, seizure frequency, Hospital Anxiety and Depression Scale and self-report Adverse Events Profile were evaluated.

Results: Monotherapy was used in 55.3% of patients, 30.9% of patients had 2 AEDs, 13.8% of patients had 3 and more AEDs. 32.6% of patients were seizure free. 32.3% of patients were not-seizure-free and had monotherapy, the remaining patients had AED combination. The most frequent used drug was Levetiracetam (LVT) in focal epilepsies (37%) and Valproic acid (VPA) in generalized epilepsies (44%). Most frequently reported side effects were ataxia (p = 0.048), aggression (p = 0.014), nausea (p = 0.052), and cognitive impairment (p = 0.059). Most common combination of AEDs was LVT & CBZ (25.3%). The other combinations were LVT and Lamotrigine

(9.7%), LVT and Lacosamide (9.3%) and LVT & OXC (8.0%). Seizure-free patients had scored better in HADS (anxiety 8.9%, depression 5.8%) compared to not seizure-free patients (anxiety 16.6%, depression 12.5%).

Conclusions: The present retrospective study provides updated information on the patterns of prescription of AEDs in a large population of patients with epilepsies. There was a high portion of patients with more than one AED, which can be explained by increasing number of new AEDs, many of which are approved only for add on therapy.

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Abstract – WCN 2013**No: 1149****Topic: 1 – Epilepsy****A presentation of two clinic cases of Hypomelanosis of Ito and epilepsy***J. Zhang. Department of Neurology, Hunan Children's Hospital, Changsha, China*

Hypomelanosis of Ito, also known as the lack of incontinentia pigment achromians, is mainly with skin and nervous system symptoms and may be associated with many side symptoms such as head and facial dysplasia, congenital malformation of heart and reproductive systems, etc. Because it is related to multiple systems and organs it is now considered a multi-system involved skin disease.

Ito (Ito's) first reported this disease in 1951 and he considered it might be due to autosomal dominant inheritance [1]. Now people are inclined to think that the possible pathogenesis of the disease is mutations caused by the X chromosome inactivation or activation that caused abnormal transition of mesoderm and ectoderm precursors during embryonic development, and as a result, leading to skin section line development disorder, neuronal migration disorder, neurodysplasia, and forming multi-system abnormalities.

At present the available epidemiological data for such kind of neurocutaneous syndrome disease is very limited. Two cases were identified in Changsha, China and are reported here. At first the whole process of diagnoses and treatments for the patients will be described in detail. Then, discussions are presented on how to identify Hypomelanosis of Ito from another two similar neurocutaneous syndromes: incontinentia pigmenti and tuberous sclerosis. The last part of this paper is recommendations on general procedures of diagnosis and treatments for this disease.

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Abstract – WCN 2013**No: 1145****Topic: 1 – Epilepsy****Hot water epilepsy-seizure induction procedure during EEG recording***B. Labovic, R. Raicevic, T. Lepic, N. Rajsic. Clinic of Neurology, Military Medical Academy, Belgrade, Serbia*

Hot water epilepsy (HWE) represents a rare clinical entity, classified in a group of "reflex epilepsy" in which seizures are precipitated by hot water during bathing.

Case report: We report HWE in a 21-year-old male, with symptoms that appeared at the age of 7 years old. After initial provocative procedure we have applied a tea pot filled with warm water at 38 °C and wrapped it with towel and then was pressed against his back but with no effect. But when we used a shower of wider surface with hot

water of the same temperature 38 °C, this has been effective. It is possible that the stimulation of broad skin areas involves hyper synchronization of the parietal and frontotemporal regions that is critical to elicit seizure not the temperature alone.

Discussion: Findings of local epileptiform in frontotemporal regions lead to the assumption of structural lesion of temporal lobe but subsequent neuro-imaging studies failed to prove it. Experimental data derived from animal model mimicking HWE has shown ischemic changes in specific topographic areas, like Sommers' sector in hippocampus, layers 4 and 5 neurons of the cerebral cortex reticular neurons in brainstem a pathological feature reminiscent of the human epileptic brain. Autopsy findings of three HWE patients have suggested the most likely pathophysiologic mechanism in abnormal thermoregulation among genetically susceptible population with possible environmental influences.

Conclusion: It is possible that stimulation of the broad skin areas involves hypersynchronization on the parietal and frontotemporal regions that is critical to elicit seizure, not the temperature alone.

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Abstract – WCN 2013

No: 1170

Topic: 1 – Epilepsy

PRRT2 mutation in Japanese PKC/PKD cases

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Introduction: Paroxysmal kinesigenic choreoathetosis dyskinesia (PKC/PKD) is characterized by brief attacks of involuntary movement triggered by sudden onset of movement. PKC/PKD is commonly inherited as an autosomal dominant trait, and *PRRT2* gene was recently identified as the causative gene.

Objectives: To search for mutations in *PRRT2* gene in Japanese cases with PKC/PKD.

Patients and method: Two cases with a family history of PKC/PKD and three sporadic cases were studied. All cases developed symptoms during childhood. Blood samples were collected after obtaining informed consent, and genomic DNA was extracted using standard procedure. Three exons of the *PRRT2* gene were amplified by PCR, and sequencing was then carried out using ABI 310 automated sequencer.

Results: Truncating mutation in two familial cases was identified, but any mutations were not found in sporadic cases.

Conclusion: *PRRT2* gene mutation is also confirmed in Japanese PKC/PKD cases.

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Abstract – WCN 2013

No: 1175

Topic: 1 – Epilepsy

Pooled analysis of individual patient data from European observational studies on zonisamide use in clinical practice: Sub-group comparisons of efficacy

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Background: A pooled analysis has been implemented from 5 European observational studies on zonisamide in routine clinical practice: France, Germany/Austria, Spain, Nordic countries and Scotland.

Objective: To assess overall efficacy of zonisamide and explore sub-group comparisons in a larger sample of real-life observational studies.

Material and methods: Efficacy endpoints were assessed at 6 months according to: $\geq 50\%$ responder rate, seizure-free rate ≥ 2 months and retention rate. Sub-group comparisons were the number of previous antiepileptic drugs (AEDs) before zonisamide initiation and the main first line AED with zonisamide.

Results: A total of 988 patients with partial epilepsy were included: 427, 292, 136, 102 and 31 patients in France, Germany/Austria, Scotland, Nordic countries and Spain respectively. There was a significant association between the number of previous AEDs at zonisamide initiation and efficacy endpoints ($p < 0.01$). For patients without previous AED, the $\geq 50\%$ responder rate was 59% (versus 46% for patients with at least 3 previous AEDs); the seizure-free rate was 33% (versus 17%). Zonisamide–valproic acid seemed to be the best bitherapy associated with efficacy endpoints, compared to zonisamide–carbamazepine, zonisamide–lamotrigine and zonisamide–levetiracetam combinations, but not significantly ($\geq 50\%$ responder rate ($p = 0.12$); for seizure-free rate ($p = 0.11$)).

Conclusion: These preliminary results regarding efficacy sub-group comparisons show a trend in favour of a better responder rate for patients “newly” treated for partial epilepsy (i.e. with a lower number of previous AEDs at zonisamide initiation). Complementary analyses will be performed to take into account potential heterogeneity regarding confounding factors including baseline severity.

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Abstract – WCN 2013

No: 1074

Topic: 1 – Epilepsy

Precise detection of epileptic spikes in the EEG

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We present a novel method that automatically detects interictal epileptic spikes in the electroencephalogram (EEG). Conventionally, EEG experts visually analyze the EEG recordings in order to identify such patterns. This is time-consuming, laborious and prone to errors especially for long term recordings of many hours or days. For this reason automatic spike detection systems are of great interest in order to allow an objective and accurate investigation with significant time-savings.

Since it is often preferred to find clear spikes rather than to find all spikes, we developed a method maximizing the precision, i.e. the portion of true positives in the detections. Our system first detects spikes based on a non-restrictive definition of the spike morphology. Subsequently the spikes are grouped by clustering methods using morphological features and the results of a source localization. The clustering allows an immediate overview of the types of spikes contained in the EEG data and the identification of multiple sources. Representative group averages are presented to a user who is able to deselect artifact clusters containing false positive detections only.

Our results were obtained from the EEG of three patients with a recording duration totaling 6.5 h and containing 282 spikes, which were marked by experienced EEG experts. After the initial detection we achieved an average precision of 85% with a sensitivity of 48%. The subsequent clustering performed a very clear separation of true spikes and artifacts and could be used to increase the precision of up to 98%.

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Abstract – WCN 2013**No: 1187****Topic: 1 – Epilepsy****A paraneoplastic limbic encephalitis as the first manifestation of an plasmacytoma**R. Schimana. *Clinic Vilshofen, Vilshofen, Germany*

Background: A 72-year old woman had several drop-attacks and a neurological syndrome with aphasic symptoms, ataxic gait and mnestic deficits. The syndrome is getting better, but 7 days later she had a temporal lobe seizure. Is the syndrome the first sign of a epilepsy with a prolonged remission or a nonconvulsive status epilepticus or seizures are expression of an systemic CNS-disease.

Objective: Hemiparesis left with fine motoric skills deficits.

Neuropsychology: Deficits in short time memory and attentional functions.

Material and methods: The classical neurological diagnostic procedures.

Results: EEG: No slow activity. No seizure pattern. Theta-Delta focus right temporoparietal. MRI: no new ischemic sign. Hyperintensive lesions in T2 weighted images.

Leptomeningeal enhancement.

Liquor: cells: 4, protein: 120 mg/dl, glucose: lactate normal. Reiber diagram: blood-brain barrier disturbance. Ig G very increased, Ig M very decreased. Oligoclonal bands in serum and liquor positive. Laboratory: normal except for creatinine 1.5 mg/dl. Vasculitis-screening and thyreoid antibodies negative. Electrophoresis: monoclonal gammopathy.

Immunofixation (IF) serum: light chain type kappa.

IF urine: bence jonce proteinuria Paraneoplastic antibodies: CV2-positive, NMDA-AB, potassium-channel AB, Hu, Ri, Ma negative. Bone marrow biopsy: immunohistochemistry compatible with an plasmacytoma light chain type kappa.

Conclusion: Based on the results the syndrome is in the order of a paraneoplastic limbic encephalitis. Under a cortisontherapy the syndrome and blood-brain barrier disturbance is getting better. The neurological diagnostics contribute to the detection of the plasmacytoma. Hitherto we seen paraneoplastic neurological syndromes with positive CV2-antibodies in relation to thymoma, lung-cancer, but not with a plasmacytoma.

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Abstract – WCN 2013**No: 553****Topic: 1 – Epilepsy****Physical disabilities of epilepsy in poor resource countries**

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Background: Epilepsy is an important cause of disability and death. It has been suggested that 85% of children with disability live in resource poor countries (RPCs), but there is little data to support this.

Methods: We conducted a survey of epilepsy in children aged 6–9 years in at Kenyatta National Hospital. First, we screened for neurological disability by administering the Ten Questions Questionnaire (TQQ) to parents/guardians of the children. In phase two, we performed a comprehensive clinical and psychological assessment on children who tested positive on TQQ and on a similar number of children who tested negative.

Results: A total of 10218 children were screened, of whom 955 (9.3%) were positive on TQQ. Of these, 810 (84.8%) were assessed, and of those who tested negative 766 (8.3%) were assessed. The prevalence for moderate/severe epilepsy was 61/1000 [95% confidence interval (95% CI) 48–74]. The most common domains affected were epilepsy (41/1000), cognition (31/1000), and hearing (14/1000). Motor (5/1000) and vision

(2/1000) impairments were less common. Of the neurologically impaired children ($n = 251$), 56 (22%) had more than one impairment. Neonatal insults were found to have a significant association with moderate/severe epilepsy in both the univariate [odds ratio (OR) = 1.70; 95% CI 1.12–2.47] and multivariate analyses (OR = 1.30; 95% CI 1.09–1.65).

Conclusions: There is a considerable burden of moderate/severe epilepsy in Kenya hospitals, with motor, vision, epilepsy, cognition, and hearing being the most common domains affected. Neonatal insults were identified as an important risk factor.

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Abstract—WCN 2013**No: 504****Topic: 1—Epilepsy****Classification of focal epileptic seizure transformations in adults**

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Background: In the structure of clinical understanding of various focal forms of epilepsy both frequency and type of seizures, as well as seizure patterns change dynamics.

Objective: To study various seizure transformation types as an important indicator of the disease severity and to create transformations' classification, suitable to be employed in clinical practice.

Patients and methods: Seizure transformations were studied in 273 epileptic patients. The impact of seizure dynamics on functional status of the brain, quality of life, and emotional and cognitive status of patients was assessed. By transformation we understood complete change of the seizure type or its components.

Results: Full seizure type modification revealed positive and negative seizure transformations. Positive transformation manifested itself as modification of secondary generalized or complex partial seizure into simple partial seizure, as well as modification of secondary generalized seizure into complex partial seizure. With negative transformation, this group of patients displayed modification of complex partial and/or simple partial seizure into secondary generalized seizure. Also, addition of other simple and complex seizures was observed.

Positive transformation was usually accompanied by improved quality of life, emotional state, and cognitive functions. With negative transformation these indicators deteriorated.

Transformations with changing focal seizure component manifested in it being longer or shorter in the structure of secondary generalized or complex partial seizure.

Conclusion: The suggested classification of focal seizure transformations is a vital criterion in estimating clinical state of patients and types of disease progression. This is important for identifying adequate therapy.

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Abstract – WCN 2013**No: 1253****Topic: 1 – Epilepsy****Effect of anti epileptic therapy on serum homocysteine in children**

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Background: Elevated serum homocysteine concentration is associated with increased risk for vaso-occlusive disease like cerebrovascular stroke, coronary artery disease, and also the risk of resistance to anti-epileptics and refractory epilepsy. Hyperhomocysteinemia has been frequently associated with the administration of anti epileptic drugs (AEDs).

Objective: This study aims at evaluating the effect of anti-epileptic therapy on serum homocysteine levels in children.

Methods: 53 children (males—32, females—21) presenting to the Pediatric Outpatient and Inpatient Department of Holy Family Hospital, New Delhi with seizures in age group of 6 months–14 years were included in the study. Serum homocysteine (Hcy) levels of children already on AEDs for >6 months (Group A) were compared with children before initiation of anti epileptic drugs (Group B). These children were followed up after 6 months of anti epileptic therapy and serum homocysteine was compared (Group C).

Results: Average Hcy levels in subjects who had already received >6 months of antiepileptic drug therapy were $12.58 \pm 2.68 \mu\text{mol/l}$, compared to $8.83 \pm 2.82 \mu\text{mol/l}$, at recruitment ($p = 0.001$). Significant increased levels were also observed in children followed up after 6 months of AED— $10.27 \pm 3.06 (\mu\text{mol/l})$ compared to $8.63 \pm 2.90 (\mu\text{mol/l})$ at initiation of AED. 9 children who received >1 AED had significantly higher levels— $14.15 \pm 2.56 (\mu\text{mol/l})$ compared to children on monotherapy— $10.22 \pm 3.06 (\mu\text{mol/l})$. Carbamazepine therapy for 6 months caused significant increase in Hcy $10.78 \pm 2.82 (\mu\text{mol/l})$ compared to baseline of $9.30 \pm 2.70 (\mu\text{mol/l})$ ($p = 0.016$).

Conclusions: AEDs in children, especially those receiving multidrug or long duration treatment, cause significantly increase in homocysteine levels, a risk factor for vaso occlusive diseases.

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Abstract — WCN 2013

No: 1249

Topic: 1 — Epilepsy

Prescribing patterns of antiepileptic drugs and interaction risk in general practice

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Background: In the last years a growing trend in anti-epileptic drug (AED) use was observed, but few data concerning indication of use and drug interaction risk are available in general practice.

Objective: To analyze the prescribing pattern of newer and older AEDs in the period 2005–2011 and to assess the exposure to potential drug interactions in a general practice setting of southern Italy.

Patients and methods: We analyzed data of almost 150,000 individuals registered in 123 general practitioners' lists. One-year prevalence and incidence were calculated. The frequency of concomitant treatment with clinically relevant interacting drugs was also determined.

Results: Prevalence of AED use increased from 10.7/1000 to 13.0/1000 for old AEDs, and from 14.7/1000 to 16.2/1000 for new AEDs, with a peak of 22.3 in 2006. Among older AEDs, phenobarbital and valproate were the most widely used in 2011, accounting for 21.23% and 16.23% of total AED use. In the same year oxcarbazepine and lamotrigine were the most used new AEDs (10.91% and 10.79% of total, respectively), while gabapentin and pregabalin exhibited the higher incidence of use. A high number of patients treated with older AEDs received co-prescription at clinically relevant interaction risk. Lamotrigine also showed a high annual rate of possible interaction.

Conclusion: The increasing use of newer AEDs was mostly due to the prescription of gabapentin and pregabalin for neuropathic pain. The revised re-imburement criteria determined a fall in use in 2007. The large use of phenobarbital should be considered due to the risk of adverse drug reactions.

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Abstract — WCN 2013

No: 98

Topic: 1 — Epilepsy

Peculiarities of structural white matter abnormalities on clinical realization of epilepsy

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Objective: The current study examined patterns of white matter tract abnormalities and relationships with clinical and neurophysiologic factors in subjects with epilepsy.

Methods: DTI data with tractography were obtained in 46 epilepsy patients (36 with pharmacoresistant subjects and 10 at remission stage) and 10 age-matched healthy controls using 1.5-Tesla MR scanner (Philips). Analysis of fractional anisotropy (FA) and diffusion mean diffusivity (MD) for anterior and posterior hemispheric quadrants was performed. Correlation analysis for FA and MD with clinical course of disease and EEG-mapping, cognitive event potential parameters was carried out.

Results: Subjects with epilepsy, as compared to healthy controls, demonstrated four patterns of tracts reduced: in frontal lobe, lateral parts of hemisphere mono- or bilaterally, anterior and/or posterior commissure, complex of several tracts changes. Epilepsy subjects have reduced FA, predominately in the epileptogenic cerebral hemisphere ($p < 0.05$). MD increasing and cluster of tracts reduce in frontal lobe typified for pharmacoresistant course of disease ($p < 0.05$). Pattern of tracts reduced in lateral parts of hemisphere monolaterally correlated with focal onset of epilepsy, bilaterally — with generalized seizure onset ($R = 0.42$, $p = 0.03$). Anterior and/or posterior commissural reducing correlated with prolonged P300 latency ($R = 0.39$, $p = 0.029$). Mean FA and MD was positively correlated with Beck and Spilberger–Hanin scale data ($R = -0.2$, $p < 0.001$) and P300 latency ($R = 0.23$, $p < 0.001$). Presence of epileptic EEG activity was correlated with FA reduction ($R = 0.7$, $p = 0.01$).

Conclusions: Epilepsy is associated with widespread disturbances in white matter tracts. DTI data can be predictors of clinical course of disease and cognitive disturbances.

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Abstract — WCN 2013

No: 1280

Topic: 1 — Epilepsy

Clinical and epidemiological characteristics of epilepsy in children in Kazakhstan

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Objective: To study the epidemiological and clinical features of epilepsy in children in Kazakhstan.

Methods: Patients with epilepsy aged 0 to 14 years.

Results: The analysis of 1477 cases of epilepsy in children. The high prevalence of epilepsy (12.1–14.3%) was observed in regions with unfavorable environmental conditions. The frequency of epilepsy among boys was 58.6%, of girls — 41.4%. The age of onset of the disease: in the first year of life — 20%, 1–3 years — 27.5%, 4–7 years — 31.8%, 8–14 years — 20.7%. 378 children with epilepsy were subjected to in-depth examination. Analysis of the etiologic factors of symptomatic epilepsy ($n = 234$) showed the prevalence of perinatal factors — 65% (152), congenital malformations of the nervous system — 32.9% (77), genetic abnormality — 2.1% (5). 103 patients (27.2%) had generalized epilepsy, 177 (46.8%) — partial epilepsy, 98 (25.9%) — epileptic encephalopathy. Generalized epilepsy presented idiopathic forms:

generalized epilepsy with isolated attacks – 56.3% (58 children), childhood absence epilepsy – 24.3% (25), benign myoclonic epilepsy of infancy – 11.7% (12), epilepsy with myoclonic–astatic seizures – 7.8% (8). Partial epilepsy consisted of: occipital – 25 (14.12%), temporal – 7 (3.95%), frontal – 16 (9%), rolandic – 18 (10.16%), epilepsy with secondary generalized attacks – 111 (62.71%). Epileptic encephalopathy: syndrome Ohtahara – 4 (4.1%), WS – 67 (68.3%), LGS – 15 (15.3%), ESES – 3 (3%), pseudo-Lennox syndrome – 9 (9.1%).

Conclusion: The results were the bases for the formation epilepsy register in Kazakhstan.

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Abstract – WCN 2013

No: 1331

Topic: 1 – Epilepsy

Towards surgical treatment of status epilepticus complicating Lennox Gastaut syndrome: Place of the vagus nerve stimulation

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Introduction: About 50–70% of epileptic patient with Lennox Gastaut syndrome (LGS) present a status epilepticus (SE) requiring immediate treatment. Typically, refractory SE does not respond to anti-epileptic drug therapy. Vagus nerve stimulation (VNS) therapy seems to be an alternative for these patients.

Purpose: Our study consists in comparing the clinical evolution and quality of life of three patients followed for LGS in the department of neurology of Charles Nicolle Hospital before and after setting up a VNS.
Methods: We retrospectively reviewed three children with LGS and recurrent SE treated successfully with VNS. Neurological exam, electroencephalography (EEG), and cerebral MRI were made.

Results: The mean age was 10.6 years. The sex ratio was 2/1. Between the 12th and 48th month long-term follow-up, the decrease in seizure frequency was over than 50%. There was also a decrease of drowsiness on the post critical and improving awareness, attention and mood. There are no more electrical abnormalities.

These results are consistent with those described in previous studies about this topic. VNS has also an economic benefit with reduction in hospital costs and hospital stays in Intensive Care Unit. The precise action of VNS as a neuromodulatory treatment for epilepsy is still unknown, but efficacy seems based on an incremental effect. The main described effect of VNS on brain activity is a thalamic activation and inhibition of the solitary tract nucleus.

Conclusion: The management of LGS with refractory SE is very difficult. VNS can be a good alternative to prevent the recurrence of seizures.

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Abstract – WCN 2013

No: 1335

Topic: 1 – Epilepsy

Meropenem induced myoclonus: A case report

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Background: Among adverse drug reactions myoclonus is of rare occurrence.

Objective: To report a case of meropenem induced myoclonus.

Patients and methods: This patient was evaluated at the Clinical Hospital of Paraná Federal University, Brazil.

Results: A 61-year-old woman was seen because of myoclonic status. In the previous year she had been hospitalized due to dilated cardiomyopathy (ejection fraction = 21%) plus non-sustained ventricular tachycardia and managed with an implantable cardiac defibrillator. She was readmitted on a setting of precordial pain plus cardiac insufficiency profile requiring vasoactive drugs and an intra-aortic balloon. Her coronary angiography was unremarkable. In the following days her dyspnea worsened requiring non-invasive ventilation and her central catheter became colonized. She evolved with oliguria, vomiting, hypoglycemia, syncope and cardiac arrest successfully managed with cardiorespiratory resuscitation (CPR). Her renal function decreased and a urinary infection ensued, so intravenous meropenem 100 mg/day was started based on the antibiogram. On this very day she presented with intense and maintained myoclonic jerks involving her shoulder girdle, neck and head. The myoclonic jerks were attributed to renal impairment and she underwent two hemodialysis sessions without changes in myoclonic status. Her prolonged electroencephalography (EEG) recording revealed spikes and polyspikes and waves that were abolished by 2 mg of intravenous diazepam suggesting myoclonus of cortical origin. After diazepam clearance myoclonus recurred and was abated only when meropenem was withdrawn and substituted by cefepime. Myoclonus never relapsed thereafter.

Conclusion: Meropenem epileptogenicity should be remembered, especially for those patients bearing renal insufficiency and within a complex clinical scenario.

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Abstract – WCN 2013

No: 1363

Topic: 1 – Epilepsy

Neuron specific enolase is the most sensitive biomarker in acute seizure patients

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Objectives: Identification and diagnosis of seizure are difficult among patients with a loss of consciousness. Distinguishing only based on medical history obtained from the patient can be complicated as there may be no witnesses of the event. We evaluate several makers for effectiveness of diagnose seizure.

Patients and methods: Adult patients aged 19–79 years who admitted to Gangnam Severance Hospital were included. We only included the patients had blood test within 8 h after seizure attack. We analyze serum biochemistry including neuron specific enolase (NSE), prolactin, and ammonia.

Results: We enrolled 41 patients who had final confirmed diagnosis of a generalized tonic-clonic seizure and underwent analysis of serum prolactin, NSE and ammonia (29 males, 13 females). NSE is the most sensitive biological marker to diagnose seizure (sensitivity: NSE 86% > ammonia 69% > prolactin 39%).

Conclusion: Serum NSE during short post-ictal period may be useful tests for diagnosis of seizure. We consider to analysis NSE when the patient with loss of consciousness is uncertain to differentiate to other disease rather than seizure.

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Abstract — WCN 2013**No: 1378****Topic: 1 — Epilepsy****Zonisamide monotherapy in adult patients with partial, generalized & combined seizures: Interim analysis of a open-label, non-comparative, observational study**

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Purpose: To evaluate efficacy and safety of Zonisamide monotherapy in adult patients with partial, generalized & combined seizures.

Method: In this open, non-comparative, multi-center, observational study, 119 patients (73 males) having partial, generalized and combined seizures were treated with Zonisamide monotherapy (100–500 mg/day) for 24 weeks. Seizure frequency, clinician's global assessment of response to therapy (CGART) and patient's global assessment of tolerability to therapy (PGATT) were assessed every 4 weeks. Primary outcome was reduction in seizure frequency and secondary outcomes were responder rate (>50% reduction in seizure frequency) and seizure freedom over 24 weeks. Adverse events were recorded during the study period. Change in seizure frequency from baseline was analyzed by Friedman test (non-parametric repeat measures ANOVA).

Results: 97 patients completed 24 weeks study period, while 23 discontinued (12 lost to follow up, 3 due to medication error, and 1 withdrew due to adverse event). Mean percent reduction in seizure frequency at 24 weeks was 89.30%. After 24 weeks, overall responder rate was 66.18%, whereas 24 weeks seizure freedom was 45.59%. Adverse events were reported in 6 (5.04%) patients, of which 2 (1.68%) had loss of appetite, while 1 patient each reported dizziness, and aggressive behavior with sleep disturbance.

Conclusion: Zonisamide monotherapy demonstrates favorable efficacy and tolerability in adult patients with partial, generalized & combined seizures. However this is an interim analysis.

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Abstract — WCN 2013**No: 1369****Topic: 1 — Epilepsy****Efficacy and safety of Zonisamide as first add-on therapy in adult patients with partial, generalized and combined seizures**

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Purpose: To evaluate the efficacy and safety of Zonisamide in adult patients with partial, generalized & combined seizures.

Method: In this open, non-comparative, multi-center, observational study, 271 patients (151 males) having partial, generalized and combined seizures were treated with Zonisamide (100–500 mg/day) for 24 weeks as first add-on to the primary antiepileptic drug of clinician's choice. Seizure frequency, clinician's global assessment of response to therapy (CGART) and patient's global assessment of tolerability to therapy (PGATT) were assessed every 4 weeks. Primary outcome was reduction in seizure frequency and secondary outcomes were responder rate (>50% reduction in seizure frequency) and seizure

freedom over 24 weeks. Adverse events were recorded during the study period. Change in seizure frequency from baseline was analyzed by Friedman test (nonparametric repeat measures ANOVA).

Result: 231 patients completed 24 week study period, and 40 discontinued (30 lost to follow up, 7 due to medication error, and 1 withdrew due to adverse event). Mean percent reduction in seizure frequency at 24 weeks was 92.97%. After 24 weeks, the overall responder rate was 52.14%, whereas 24 weeks overall seizure freedom was 39.83%. Adverse events were reported in 6 (5.04%) patients, of which 2 (1.68%) patients reported loss of appetite.

Conclusion: Zonisamide demonstrates favorable efficacy and tolerability as first add-on in adult patients with partial, generalized & combined seizures.

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Abstract — WCN 2013**No: 1348****Topic: 1 — Epilepsy****Health economic consequences of epilepsy in Austria**

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Introduction: Even though it is one of the most common neurologic diseases, in Austria no cost-of-illness study exists for epilepsy. Health economic evaluations are important tools for decision makers, such as insurance carriers or physicians, to ensure fair distribution of limited resources.

Objective: The purpose of this study was to evaluate the direct and indirect costs of epilepsy.

Method: 131 consecutive patients attending the outpatient clinic of the tertiary epilepsy center in Vienna were prospectively evaluated. A patient diary and a questionnaire were used to record direct and indirect costs for a twelve-month period. 67 patients contributed at least three months, 46 patients twelve months of information. The human capital approach was used to calculate the indirect costs.

First results: The mean annual cost per person was 14,322 €. Direct and indirect costs contributed 50% to the total costs. Early retirement was the largest cost factor (6396 €/44.6%), followed by diagnostics (2527 €/17.6%), inpatient care (2272 €/15.9%) and antiepileptic drugs (2012 €/14%). Refractory epilepsy and partial seizures were cost driving factors.

Conclusion: In this study the total costs exceed those of comparable studies. This can be explained with differences in methodology. The pattern of cost-distribution in this study is found in most of the recent existing studies. However, studies including more patients and exact evaluation of cost driving factors are needed.

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Abstract — WCN 2013**No: 1415****Topic: 1 — Epilepsy****Brain MRI findings and case history in a boy with repeated epileptic seizures: Case report**

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Background: Epilepsy is a chronic neurological disorder characterized by recurrent and unprovoked seizures. Brain MRI is one of the most reliable neurodiagnostic methods.

Objective: We report unusual brain MRI findings in a child with epileptic seizures.

Patients and methods: We present a previously healthy 8-year-old Caucasian boy with clear personal and family history with repeated epileptic seizures and brain demyelinating-like lesions.

Results: Postictal neurological status was normal. Visual Evoked Potentials showed bilateral optic nerve central afferent dysfunction. Lumbar puncture was performed with no significant findings. There was no presence of oligoclonal bands either in cerebrospinal fluid or in blood serum. Postictal electroencephalogram showed slow background activity with groups of delta and theta waves over anterior regions; whereas control EEG was with better organized background activity, with groups of slow and sharp wave discharges. A head CT scan showed relative narrowing of subarachnoid space on brain convexity with suspected discrete brain edema. Brain MRI showed multifocal, supratentorial and subcortical demyelinating-like lesions. Demyelinating-like lesions were asymmetrical with cortical and cortical-subcortical border line distribution in both hemispheres. There was no significant association of deep white matter.

Conclusion: Full diagnostic assessment should be obtained for all patients with epileptic seizures. Considering the available references, we found such MRI findings unusual in a child with epileptic seizures.

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Abstract – WCN 2013

No: 1438

Topic: 1 – Epilepsy

The prognostic value of interictal scalp EEG to surgical outcome in patients with hippocampal sclerosis; influence of antiepileptic drugs

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Objective: We performed this retrospective study to determine if there are differences in interictal EEG's prognostic value during gradual reduction of antiepileptic drugs (AEDs) in relation to surgical outcome, defined by Engel's classification, in patients with refractory TLE with hippocampal sclerosis (HS).

Patients and methods: In a total number of 55 patients, we analyzed laterality and absolute number of IED's as an average value for the total time of monitoring (TT), and in three different periods of interest defined by the level of AEDs' reduction – full medication period (FMP), reduced medication period (RMP), and withdrawal medication period (WMP). Laterality was defined as unitemporal ($\geq 90\%$ of IEDs on the side of HS) and bitemporal ($< 90\%$ of IEDs on the side with HS).

Results: The laterality of IED's during TT ($p = 0.047$) and during FMP ($p = 0.005$) was a statistically significant predictor factor for surgical outcome. There were no statistically significant differences of laterality during RMP (0.074) and WMP (0.131) in relation to outcome. The absolute number of IED's on the side of HS was not predictive for the surgery either as an average value ($p = 0.502$), or during FMP ($p = 0.706$), RMP ($p = 0.709$) or WMP ($p = 0.719$). The absolute number of IED's on the contralateral side to HS was a predictive factor for surgery during TT ($p = 0.083$) and during FMP ($p = 0.01$). There were no statistically significant differences during RMP ($p = 0.081$) and WMP ($p = 0.083$).

Significance: The prognostic value to the surgical outcome has the laterality and the absolute number of IED's both during TT and FMP.

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Abstract – WCN 2013

No: 1463

Topic: 1 – Epilepsy

Retigabine as adjunctive treatment in adults with refractory partial onset epilepsy. Own experience at least a half-year follow-up

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Background: Retigabine (RTG) exhibits a novel mechanism of action opening neuronal KCNQ/Kv7 potassium channels leading to hyperpolarization of membrane potential and decreased neuronal excitability. RTG has been available in Slovakia since October 1, 2011.

Objective: Objective of the study was to monitor pharmacotherapy-resistant patients focusing on RTG efficiency and safety.

Patients and methods/material and methods: In the survey we monitored demographic data, etiology of epilepsy, response to therapy, co-mediation, and adverse side-effects of RTG treatment. Mean follow-up time period was 11.7 months.

Results: 33 men and 28 women of median age 39.4 years (range: 20–61) and a median duration of epilepsy of 26 years (range: 2–51) were included.

Median seizure frequency one month before RTG treatment was 8 (range 2 to 50).

Median RTG final dose was 600 mg/day (range 150 to 1150 mg/day). Most frequent co-medication: levetiracetam, carbamazepine, lamotrigine, valproate.

Most frequent etiology: 32.8% perinatal injury, 24.6% cryptogenic, 14.7% trauma, 8.2% cerebral infection.

Responder rate: 14.7% seizure free; 41.0% responders reporting 50-to-99% reduction of seizures; 19.7% reported seizure reduction below 50%; 16.4% reported no change; 3.3% reported deterioration; 21.3% of patients discontinued their treatment.

Side-effects were observed in 34.4% of patients – most frequently fatigue, drowsiness, vertigo and tremor.

Conclusion: RTG in our study demonstrated the efficiency and tolerability profile comparable to that in pivotal trials. RTG treatment was efficient and well tolerated in a highly therapy resistant population of partial-onset epilepsy patients.

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Abstract – WCN 2013

No: 1477

Topic: 1 – Epilepsy

Midazolam plasma levels after the administration of a galenic nasal spray formulation in healthy volunteers

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Background: The initial treatment of seizures is intravenous or rectal benzodiazepines. However, intravenous access during a seizure is difficult. Rectal administration is safer, but can be considered socially

unacceptable. Intranasal Midazolam (INM) would be a new approach to the management of seizures but this nasal formulation is not commercially available in Argentina.

Objective: To investigate MDZ plasma levels after intranasal administration (IN) of a galenic nasal formulation containing MDZ in healthy volunteers.

Patients and methods: 4 healthy volunteers (3 male, 1 female) were included. They received a single 4 mg MDZ dose intranasally administered. MDZ nasal spray formulation consisted of 2.5% (w/v) midazolam, propylene glycol, and water. Blood samples were drawn at 10 min, 1 and 2 h after MDZ IN. Volunteers were asked to classify local irritation immediately and 2 h after MDZ IN. Data of sedative effects were also recorded.

Results: 4 mg IN MDZ dose was well tolerated. After the administration low grade local irritation was reported by the volunteers. MDZ plasma levels were observed 10 min after IN (mean: 11.6; SD: 4.6 ng/ml). One hour after IN MDZ mean level was 8.9 (SD:7.7) ng/ml. Sedative effects were observed in 3 volunteers.

Conclusion: This study showed that MDZ levels could be measured after IN of the galenic nasal formulation developed by our group. Since MDZ base is a lipophilic compound with low solubility in water, further studies are needed to evaluate inter-individual pharmacokinetic variability or the utility of enhancers of transmucosal absorption to increase bioavailability.

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Abstract – WCN 2013

No: 1499

Topic: 1 – Epilepsy

Pharmacologic and genetic aspects of resistant epilepsy in children in Uzbekistan

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Objectives: To develop methods of detection for the frequency and influence of polymorphism of cytochrome P450 genes on pharmacologic and genetic action of antiepileptic drugs (AED). Identify the role of these genetic markers in individual responses to AED and development of pharmacologic resistance in epileptic patients.

Material and methods: The research is based on studying the results of treatment of patients (n = 20) with resistant epilepsy. The control group included 25 volunteers of Uzbek nationality adjusted by sex and age.

Results and conclusions:

1. Minimum level of gene variety of marker CYP2C9 in children was determined with not very high frequency of heterozygosis. Reduction of frequency of heterozygote genotype, absence of adverse genotype T/T and simultaneous increase in the share of individuals with normal homozygous C/C genotype among apparently healthy patients can be related to selective advantage of C allele.
2. Frequency of CYP2C9 polymorphism in pharmacoresistant patients considerably exceeded that in the control group indicating the association between development of resistance to AED and the genetic marker. The obtained statistically insignificant difference is most likely caused by the small amount of patients in the groups.
3. The genotypic variant distribution analysis of polymorphic marker 9896 C-G of CYP2E1 gene showed that among patients with the resistant epilepsy and the controls, the share of people with mutant genotypes had almost identical frequencies (20%). The risk of development of resistance to AED in carriers of the genotype was statistically insignificant (>0.05).

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Abstract – WCN 2013

No: 1513

Topic: 1 – Epilepsy

Symptomatic versus idiopathic temporal lobe epilepsies: Findings from a cohort of 75 patients

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Introduction: Temporal lobe epilepsy (TLE) is the most common form of focal epilepsy. Two main varieties have been described in TLE: mesial and lateral or neocortical temporal epilepsy.

Objective: To determine clinical, electrical and radiological differences between mesial and neocortical variants of temporal lobe epilepsies in idiopathic versus symptomatic groups.

Methods: Retrospective study of clinical, electrical and radiological features in four groups: mesial symptomatic (MSTLE), neocortical symptomatic (NSTLE), mesial idiopathic (MITLE) and neocortical idiopathic (NITLE).

Results: Total of 75 patients: 24 MSTLE, 33 NSTLE, 14 MITLE and 4 NITLE. The mean age of onset was respectively: 11.54, 22.63, 9.78 and 15.7 years. History of febrile seizures is found in 66% in MSTLE. The main types of seizures are automatisms and visceral sensory auras in MITLE symptomatic (75% and 58% respectively) and idiopathic (43% and 71% respectively). Psychic auras are the most frequent type in NSTLE (42%) and language alterations in NITLE (100%). spike-waves are common in mesial group. Pharmacoresistance is more frequent in MSTLE (54%) and NITLE (50%). In the symptomatic group, brain MRI shows association of other temporal lobe abnormalities to mesial sclerosis in 46%.

Conclusion: Our study shows earlier age of onset with a higher frequency of febrile seizures and pharmacoresistance in MSTLE group. The Symptomatic group of TLE is characterized by older age and higher frequency of background EEG abnormalities. Some features are shared between the symptomatic or idiopathic variants and others depend on the mesial or neocortical localization.

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Abstract – WCN 2013

No: 1511

Topic: 1 – Epilepsy

Multimodal presurgical evaluation in children with focal cortical dysplasia

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Objective: To evaluate the effectiveness of multimodal presurgical evaluation in patients with focal cortical dysplasia (FCD).

Methods and subjects: We performed pre- and postsurgical evaluation in 20 patients with malformations of cortical development (MCD) aged from 2 months till 17 years old. All of them underwent continuous video-EEG, MRI, 18 FDG-PET and SISCOM investigations.

Results: There were operated 20 patients with MCD with pre- and postsurgical evaluation. The mean age at neurosurgery was 6.5 years. 18 patients had different types FCD and 2 had hemimegalencephaly. The mean period between epilepsy onset and neurosurgery was 6 years. In 15 (75%) patients the presurgical MRI (1.5 T) detected FCD correlated with focal rhythmic epileptiform discharges during video-EEG register. In 5 patients (25%) with negative MRI, 18 FDG-PET and SISCOM were useful to complete the preoperative study. Surgical procedures included 6 temporal, 10 extratemporal, 2 multilobar resections and 2 functional hemispherotomies. Brain histological analysis confirmed the diagnosis of MCD in all cases. The outcome after 1 year of neurosurgery, according

to Engel classification, was IA in 16 patients (80%). Isolated FCDs type I were observed in 18% of patients and characterized by more frequent seizures, negative MRI and worse post-surgical seizure control. The outcome was better in patients with transferred extratemporal resections and functional hemispherotomy and worse in patients with multilobar resections.

Conclusions: The multimodal study with MRI, 18 FDG-PET and SISCOM was useful to complete the pre-surgical study in FCD, especially in patients with negative MRI.

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Abstract – WCN 2013

No: 1518

Topic: 1 – Epilepsy

Hemimegalencephaly in children – Some clinical and genetic aspects

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Purpose: To analyze the clinical features of hemimegalencephaly.

Methods: MRI, video-EEG and genetic analysis were performed on 8 patients with hemimegalencephaly.

Results: 3 patients have an isolated type of hemimegalencephaly and 5 patients a syndromic type, which associated with ipsilateral hemihypertrophy of the whole body. Six patients had seizures (severe refractory epilepsy – 5, febrile seizures – 1). The mean age of the epilepsy onset was 3 and 7 months. All patients had a contralateral hemiparesis and developmental delay. The seizure semiology ranged from febrile seizures to polymorphic types, like myoclonic seizures (2), complex partial seizures (3), asymmetric tonic spasms (2) and focal clonic seizures (1). Three cases registered continuous “burst-suppression” pattern in the side of hemimegalencephaly. According to genetic molecular analysis of 3 patients mutations of genes localized in the 17th chromosome were detected. In 2 patients with hemimegalencephaly accompanied by pachygyric changes and refractory seizures alteration of the gene LIS1 was found and in 1 patient well controlled by AED treatment mutation of the gene RPH3AL (rabphilin 3A-like) was found. In the 4 other patients genetic investigation was negative as in all family members. In 2 patients with severe progressive epileptic encephalopathy a functional hemispherectomy was performed resulting in 90% reduction of seizure.

Conclusions: In most cases, a clear correlation was found between the side of the hemimegalencephaly and seizure semiology which was also confirmed by the continuous video-EEG register. Genetic investigation of patients with hemimegalencephaly could be useful as a part of the complex diagnostic method and treatment prognosis.

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Abstract – WCN 2013

No: 1561

Topic: 1 – Epilepsy

Charaka and apasmara (epilepsy)

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Background: Ancient Indian medicine called Ayurveda is nearly 3000 years old, and has some amazing observations.

Objective: To provide an original study.

Material and methods: Life of Charaka and his observations on epilepsy.

Results and conclusions: Charaka was an excellent observer, teacher, scholar, healer of ancient India. Ayurveda is a science of life – one of the Vedas. Charaka lived around 1000 BC, in Taxilla in the Indus valley of Punjab. The sage Punarvasu Athreya is a pivotal figure of Charaka Samhita (Treatise). Charakas believed in discussions and seminars rather than in dogmatic teachings – thus he appears very modern.

Apasmara or epilepsy comes from the word Smara, which means “memory”; Apasmara is loss of memory. Smirti (memory) is recollection of past objects or events. Charaka defined Apasmara as a disease with loss of memory, which can lead to death. It is the loss of memory associated with darkness (unconsciousness), loathsome appearance due to derangement of intellect and mind. He associated Apasmara with a perverted mind, unclean food, anxiety, emotions, and agitations. He observed visual hallucinations, unawareness, falls, and convulsions followed by deep sleep. He classified 4 types of epilepsy. He advised emetics, enemata, ointments, milk, and many vegetable drugs. He advised proper diet, and promote memory and to lead a righteous life. Thus Charaka combined the science of philosophy and deep ethics of life.

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Abstract – WCN 2013

No: 1582

Topic: 1 – Epilepsy

Epilepsy and social security – General aspects of the insured claimants and medical decisions

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Objective: Provide a profile of individuals with a previous diagnosis of epilepsy that file claims for social security benefits and a profile of the medical advisory decisions that support the concession of these benefits.

Participants: Thirty individuals with illness-related problems due to epilepsy were selected from the claimants that receive Social Security Incapacity benefits.

Methods: An exploratory data analysis of the 188 Social Security medical files (between 2003 and 2008) of the thirty claimants was performed using the clinical and epidemiological information and the medical advisory criteria.

Results: The mean age of the claimants was 39 years and most of them were males in jobs that do not require a lot of schooling. The first claim was filed within an average of four years of employment. On average, each worker files a claim every three months, which entitles him/her to receive incapacity/sickness benefits for 17 months. The frequency of seizures and the medications used by the claimants were registered in 60% of the medical files. In addition, the description of the physical and neurological exam was incomplete in 50% of the files. Furthermore, 60% of the files did not include the argument or the clinical evidence that was used to justify the concession of the benefit.

Conclusion: The medical advisory decisions on epileptic workers tend to be inconsistent, lenient and generally lacking in clinical evidence. The disparities among the granted benefits indicate the need for the National Social Security System to review and draft specific guidelines for epilepsy.

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Abstract — WCN 2013**No: 1583****Topic: 1 — Epilepsy****The brain abscess associated with pulmonary arteriovenous malformation**

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Pulmonary arteriovenous malformation (pulmonary AVM) has direct links between an artery and a vein in the blood circulation of lungs in the human body. (1) Hereditary hemorrhagic telangiectasia (HHT), Rendu-Osler-Weber disease, is a genetic disease through autosomal dominant pattern having a feature of vascular malformations. (2) Pulmonary arteriovenous fistulas (PAVFs) are induced right to left shunt and if not treated properly, these may cause critical neurological problems such as brain abscess and meningitis. (3)

A 35-year-old man checked into an emergency room with complex periodontitis but no problems were identified in his brain images including cerebral vessels. A few days later, he suffered from convulsive movement with loss of consciousness after which brain images were taken to find a brain abscess in the right frontal region. Also, he had complained about swelling, redness as well as mild tenderness on the right side of the neck, which led to testing his neck and chest by a CAT scan. His CAT scan showed two large (>1 cm) AVM nidus in the right apex and right lower lung as well as a tiny lingular nodule, probably intrapulmonary lymph node.

We recognize through this case that we would proceed to operate his pulmonary arteriovenous malformations to prevent recurrent critical neurologic complications such as brain abscess or meningitis.

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Abstract — WCN 2013**No: 1562****Topic: 1 — Epilepsy****Noninvasive localization of nonconvulsive seizure with pulsed arterial spin labeling (PASL) MR imaging**

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Background & objective: Nonconvulsive seizures are very poorly localized seizure disorder due to variability of EEG abnormality and tiny pathology of ictal onset zone. The purpose of this study was to determine localizing value of pulsed arterial spin labeling MR imaging, which is a noninvasive method for calculating regional CBF. **Methods:** Five patients with nonconvulsive seizure in EEG monitoring are enrolled in this study retrospectively. EEG and brain SPECT, MR imaging, ictal PASL imaging, Postictal PASL imaging were examined in all patients. EEG localization and hyperperfusion of SPECT and PASL imaging were compared in patients with MRI abnormality and hyperperfusion of brain SPECT. Postictal PASL imaging was performed after normalization of EEG.

Results: In 4 patients of regional MRI abnormality, we found hyperperfusion at the same site with PASL MR imaging in all patients, which was identified with Brain SPECT, localization of ictal activity in EEG. Conversely, we found 1 minimal abnormal lesion in MRI, only T2 high signal intensity in lateral temporal cortex, whose location was found due to hyperperfusion in PASL MR imaging.

Conclusion: Pulsed arterial spin labeling MR imaging will be a useful noninvasive tool in detection of location of ictal zone in nonconvulsive seizure.

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Abstract — WCN 2013**No: 1600****Topic: 1 — Epilepsy****Metabolic syndrome in children with epilepsy on valproate and phenytoin monotherapy: A cross-sectional study**

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Background: Metabolic syndrome is a well recognized complication of valproate therapy. However there is paucity of data on the prevalence of metabolic syndrome in children with epilepsy on valproate monotherapy.

Objective: To compare the prevalence of metabolic syndrome between children on valproate and phenytoin monotherapy.

Methods: Children aged 3–16 years with epilepsy on valproate or phenytoin therapy for at least 6 months were enrolled. They were evaluated for the presence of abdominal obesity, dyslipidemia, glucose intolerance and hypertension. Metabolic syndrome was diagnosed when 3 of the following criteria were met: elevated fasting glucose (>100 mg/dl), hypertension, high triglyceride levels or abdominal obesity. The prevalence of metabolic syndrome and abnormalities in the individual parameters were compared between the two groups.

Results: Four out of 57 valproate-treated children (7%) and none of the 53 phenytoin-treated children had metabolic syndrome. Mean serum triglyceride levels were higher in the valproate group as compared to the phenytoin group (94.97 ± 38.58 mg/dl versus 77.60 ± 11.44 mg/dl, $p < 0.05$). The total cholesterol values were also significantly greater in valproate treated children as compared to the phenytoin group (148.28 ± 25.95 mg/dl versus 132.83 ± 23.51 mg/dl, $p < 0.05$). The serum fasting glucose was comparable in both groups.

Conclusion: The prevalence of metabolic syndrome and dyslipidemia was higher in valproate treated children as compared to phenytoin treated children with epilepsy. Children on valproate treatment need to be monitored for the development of metabolic syndrome.

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Abstract — WCN 2013**No: 1633****Topic: 1 — Epilepsy****The effectiveness of medical treatment of childhood epilepsy**

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Objective: To analyze the effectiveness of AED for children with epilepsy.

Material and methods: An analysis has been conducted of the effectiveness of treatment by AED based on the type of epilepsy in children at the age from 0 to 18. Idiopathic focal epilepsies — 51 children, idiopathic generalized — 89, symptomatic focal — 162, cryptogenic focal — 64 and epileptic encephalopathy (predominantly West Syndrome) — 28 children. AED prescribed to the children are as follows: Valproate, CRBM, Topiramate, Levetiracetam, Oxcarbazepine, Lamotrigine, Succinamide, Benzodiazines, Barbiturates, and Vigabatrin applied in monotherapy or in combination. The effectiveness level is based on the 4 criteria: seizure-free period during 6 months and over; seizure rate fall by 50% or over; no effect (no fall/decreasing or fall by less than 50%); seizure recommencing after drug-induced remission period and over.

Results: Idiopathic focal epilepsies: seizure-free period for 80%, seizure decrease — 8%, no effect — 6%, 6% — return to seizures. Idiopathic generalized epilepsies: seizure-free period for 75%, seizure decrease — 6%, no effect — 11%, seizure recommencing — 8%. Symptomatic focal: seizure-free period for 46%, seizure decrease — 21%, no effect — 25%, seizure recommencing — 9%. Cryptogenic focal: seizure-free period — 67%,

seizure decrease – 16%, no effect – 14%, seizure recommencing – 3%. Epileptic encephalopathy: seizure-free period – 32%, seizure decrease – 22%, no effect – 46%.

Conclusion: The study confirms that AED is more effective in treating idiopathic type of epilepsy and has less significant effect on treating symptomatic focal epilepsies and epileptic encephalopathy.

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Abstract – WCN 2013

No: 1584

Topic: 1 – Epilepsy

Familial cortical myoclonic tremor and epilepsy (FCMTE): Refinement of the *fcmt2* locus and confirmation of a founder haplotype

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Objective: To describe the electro-clinical and genetic features of a new large FCMTE family (A) that originates from the same area as another family (B), previously resulted unlinked to FCMTE2 locus (Gardella, 2006).

Methods: *Pedigree A:* 31 collaborative individuals were assessed directly. A polygraphic study was conducted in 15, back averaging analysis, somatosensory evoked potentials and C-reflex study in four. *Pedigree B:* Seven individuals were reviewed after a 10-year follow-up. The genetic study investigated 30 and 6 subjects of pedigrees A and B with 26 microsatellite markers at FCMTE1, FCMTE2, and FCMTE3 loci. We genotyped 12 markers in two affected individuals from two different families where a founder haplotype on FCMTE2 was described (Madia, 2008).

Results: *Pedigree A:* We studied 16/19 living affected members (M/F:5/11; mean age 47.8 years). Cortical myoclonic tremor (CMT) was associated with generalized seizures in ten patients (62.5%), with a mean age at onset of 28.1 and 33.8 years, respectively. Psychiatric disorders recurred in 37.5% of cases. Neurophysiological investigations confirmed the cortical origin of myoclonus in all the patients. *Pedigree B:* the status of one individual was changed to unaffected. Genetic analysis established linkage to FCMTE2 locus on chromosome 2p11.1–2q12.2 in both pedigrees and narrowed the critical interval to 10.4 Mb. Haplotype analysis identified a founder haplotype identical to that previously observed in families from the same area.

Conclusions: This study reduces the number of positional candidate genes in the FCMTE2 locus to 59 and confirms evidence of a founder effect in Italian families.

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Abstract – WCN 2013

No: 1739

Topic: 1 – Epilepsy

Predictive value of EEG in patients with West syndrome

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Background: West syndrome (WS) is an age-dependent epileptic encephalopathy characterized by a clinicoelectrical triad: infantile spasms (IS), hypsarrhythmia and delay and/or regression in psychomotor development. Although WS is considered one of the intractable epilepsies, the prognosis differs widely.

Objective: Determining the significance of electroencephalography (EEG) for long-term prognosis of WS.

Patients and methods: Our study group comprised 68 patients diagnosed with WS, aged 2 to 17 months, treated at the University Children's Hospital in Belgrade from 1987 to 2008. The outcome was assessed through the response to therapy (seizure control and EEG findings) at the follow-ups after 3, 6 and 12 months.

Results: After 3 months, seizure control was established in 50% and EEG improvement was observed in 52.9% of patients. At 6 months follow-up, 47.6% of patients were seizure-free and EEG improved in 42.9% of cases. Evaluation after 12 months showed 65.6% seizure-free patients and EEG improvement in 55.7% of patients. There was a statistically significant correlation between EEG after 3 and 6 months and seizure control after 12 months. The majority of patients with EEG improvement after 3 months (82.8%) and 6 months (95.8%) had no seizures at one year follow-up.

Conclusion: Our study has shown that in treated patients EEG improvement after 3 and 6 months can have a predictive value for long-term outcome of WS in terms of seizure control and favorable response to therapy.

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Abstract – WCN 2013

No: 1755

Topic: 1 – Epilepsy

The economic burden of epilepsy on a black South African community

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Background: The world's burden of epilepsy is carried mostly by the developing world (85%). According to the World Health Organization (WHO), Africa alone has 10 million people with epilepsy. There however, continues to be a paucity of studies on the economic impact. It is well established that epilepsy is a known cause of preventable unemployment.

Aim: To assess the economic impact of epilepsy on a black South African patient population and public healthcare system.

Method: 200 adult patients known to be with epilepsy were interviewed employing a questionnaire as our primary tool. The questionnaire investigated the financial, clinical and demographic parameters. All the patients included in the study had been assessed by a qualified neurologist. Patients with a single seizure or uncertain diagnosis were excluded from the study population.

Results: Of the respondents 95 were female, 105 male and a mean age of 35 years. Only 6.5% had acquired a tertiary qualification. The vast majority (86%) of the study participants are unemployed. This is much higher than the unemployment rate published by Statistics South Africa for the 3rd quarter of 2012 at 25.5%.

Conclusion: People with epilepsy face exceeding levels of stigmatization and thus limited opportunities in society. Extremely low levels of employment (3%) were noted. The level of education seems to be markedly hampered by the condition. More studies are needed on the economic impact especially in the developing world as this could influence government policy.

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Abstract – WCN 2013**No: 1759****Topic: 1 – Epilepsy****Analysis of efficiency and safety of oxcarbazepine in pediatric and adult patients with different forms of epilepsy**

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Background: Oxcarbazepine is an anticonvulsant used in therapy of focal seizures for patients from the age of 1 month and of GTCS from the age of 2 years.

Objective: The aim of the study was the analysis of efficiency and safety of oxcarbazepine in children and adult epileptic populations depending on the patient's age and form of epilepsy.

Patients and methods: 155 patients receiving oxcarbazepine (male 78, female 77) of the age from 5 months till 59 years were observed with dynamic video-EEG control during the period 2006–2012. The following were identified: IGE with isolated GTCS – 5, focal frontal lobe epilepsy – 40, temporal lobe – 63, parietal lobe – 6, occipital lobe – 13, idiopathic focal – 5, epileptic encephalopathies – 23 patients.

Results: Oxcarbazepine was effective in 59.3% of the patients (n = 92). Low efficiency was seen in 29.1% (n = 45) patients. The aggravation effect has been noted in 11.6% (n = 18) of patients. Drug compliance (for >1 year) was 56.8% (n = 88). Side effects were observed in 9.7% of cases. High efficiency in group <1 year (n = 14) was 35.7% (n = 5), low 35.7% (n = 5), aggravation 28.6% (n = 4); in group 1–3 years (n = 24) high efficiency 37.5% (n = 9), low 37.5% (n = 9), aggravation 25% (n = 6); in pediatric population >3 years (n = 78) high efficiency 62.8% (n = 49), low effect in 29.5% (n = 23), and 7.7% aggravation (n = 6), in adult population >18 years (n = 39) the efficiency was 74.4% (n = 29), low effect 20.5% (n = 8) and aggravation in 5.1% (n = 2).

Conclusion: Oxcarbazepine is highly effective drug in therapy of IGE with isolated GTCS, in symptomatic focal forms of epilepsy and Panayiotopoulos syndrome. With increase in patient's age the efficiency of oxcarbazepine rises, while aggravation risks decrease.

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Abstract – WCN 2013**No: 1748****Topic: 1 – Epilepsy****EEG as a prognostic tool for classic absence**

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Purpose: The main purpose of this paper is to offer some guidance to expect the prognosis of classical absence from the EEG.

Method: This was an observational study conducted from July 2006 to July 2012. Data of 42 newly diagnosed patients with typical absence seizures was collected retrospectively and analyzed.

Results: The mean time until seizure and EEG control for those with +ve 3 Hz SWC during routine EEG recording was (9.9 ± 14.4) months. While the mean total follow-up period for all cases was (19.3 ± 20.5). 100% of patients with +ve 3 Hz SWC during the 3rd minute and after HV were controlled and 80% of the overall no. of controlled patients in this study was found during 3rd min HV (p value = 0.049). The majority of the controlled patients with 3 Hz SWC after HV and during 2nd minute or 3rd minute HV was prescribed only monotherapy.

Conclusion: Overall prognosis of typical absence was good yet the presence of the 3 Hz SWC with late onset either during the 3rd minute HV or even after had a better prognosis as regard. All of

this group were controlled, monotherapy was used to control the majority of patients with late onset 3 Hz SWC. These data were only significant for the association between the 3Hz SWC and the percentage of control of absence seizures during the 3rd minute HV (80%) (p value = 0.049). A multicenter study should be done in the nearby future to confirm these results.

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Abstract – WCN 2013**No: 1809****Topic: 1 – Epilepsy****Headaches in patients with epilepsy**

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Background: Headache and epilepsy may coexist by chance, headache can be a part of seizure or the post-ictal state, but also both disorders can share a common underlying etiology.

Objective: To present coexistence of epilepsy and headaches and migraine-epilepsy syndromes characterized by a close temporal relationship between seizure and migraine attack.

Patients and methods: Adult patients (age range 18–65) with epilepsy, without any progressive neurological disease underwent detailed clinical examination and interview regarding occurrence and characteristics of headaches.

Results: Out of 203 patients with epilepsy 14.3% suffered from migraine and 30% from tension type headaches. Interictal headaches occurred in 39% and periictal headaches in 19.7% of patients. 5% of patients reported periictal headaches and 16.8% postictal headaches, that were migraine-type in one-third, and tension-type in two-thirds of cases. Two patients with idiopathic partial occipital epilepsy (Gastaut) reported migraine headaches during their partial seizures or during postictal period. Three female subjects with primary generalized epilepsy were classified as catamenial migralepsy since the onset of the menstruation activated both migraine attacks and seizures. Symptomatic etiology was diagnosed in three cases in which the clinical picture of migraine associated with epileptic seizures was imitated by arteriovenous malformation, aneurysm and occipital lobe astrocytoma. Migraine associated with epilepsy in 15 out of 29 patients required a prophylactic therapy. Postictal migrainous headache was relieved with sumatriptan, since postictal tension-type headache was treated by analgesics.

Conclusion: Classification of headaches, their temporal relation to seizures and etiologic diagnosis are mainstay for the treatment and prognosis of their comorbidity.

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Abstract – WCN 2013**No: 1799****Topic: 1 – Epilepsy****Metabolic bone disease in epilepsy patients**

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Background: Epilepsy is a chronic disorder of the brain that affects people of all ages. According to WHO, around 50 million people worldwide have epilepsy. Epilepsy patients have higher risk of bone fractures and bone metabolism disorders, that could potentially be prevented.

Objective: To characterize bone metabolism markers in patients with epilepsy of different age groups.

Patients and methods: 65 patients (age: 18–80 years) with previously diagnosed epilepsy were enrolled in the study. Patients were divided into two groups according to age: <50 and ≥50 years. Reviewing patients' medical charts we evaluated biochemical markers: PTH, vitamin D, calcium, phosphate, and creatinine levels. Bone mineral density (BMD) was assessed using DXA scanning. The data were analysed with SPSS 16.0.

Results: BMD was decreased in 55.6% of patients <50 years and 87.8% of patients ≥50 years. DXA scan T-score mean was –2.0 for the younger patients and –2.6 for older patients. Mean PTH level was 50.0 pg/ml in the younger patients and 65.0 pg/ml in the older patient group. Of all epilepsy patients in this study 30.8% had raised serum PTH levels: 20.0% in younger and 40.5% in older patients. Abnormal serum Ca and phosphate levels were detected in 10.2% and 5.2% of all patients, respectively. Vitamin D levels were on the lower reference level in both age groups.

Conclusion: Bone metabolism disorders are present in most epilepsy patients even before 50 years of age as shown by DXA scan and biochemical markers: PTH, Ca, and P levels.

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Abstract – WCN 2013

No: 1807

Topic: 1 – Epilepsy

Changes of amygdalae in systemic lupus patients with epilepsy

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Objective: To analyze slight changes of amygdalae and their volumetry in systemic lupus erythematosus (SLE) patients with neuropsychiatric symptomatology (NP-SLE), because disability of amygdalae and changes in size and activity had been reported in patients with depression and mood disorders, in some studies it even correlated with the severity of depressive episodes. NP-SLE includes mood disorders, cognitive deficits up to dementia, stroke, epilepsy etc. We performed amygdalar volumetry by semiautomatic method in special software, using manual contoured rims in sequence using thin slices.

Methods: We investigated 23 female patients with proven NP-SLE, aged 19 to 67 years, which were recruited for a prospective longitudinal neuroimaging study conducted at 1.5T MR device. Findings were statistically processed and compared with corresponding healthy persons.

Results: In our study we found, that in patients with active NP-SLE right amygdala correlated from all monitored parameters only with the left amygdala ($p = 0.01$), left amygdala correlated with right amygdala ($p = 0.01$) and with volumetry in flow attenuation inversion recovery (FLAIR) and T1 weighted images ($p = 0.05$).

Conclusions: We failed to demonstrate significant volume change in amygdalae in the sample of NP-SLE patients. We did not demonstrate significant difference of amygdalar volume due to the duration of NP-SLE or age of the patients. The authors' research was supported by the research project MZO 0006416 and 000 000 23728.

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Abstract – WCN 2013

No: 1824

Topic: 1 – Epilepsy

Excitatory and inhibitory neurotransmitters with interleukins correlations in neurodegenerative encephalopathies

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Proinflammatory interleukins (ILs) involved into microglial–neuronal interactions may explain how latent neuroinflammation can contribute to neuronal dysfunction in degenerative encephalopathies and their outcomes. Study aim was to determine IL-6, IL-8, IL-1b, glutamate, aspartate, GABA in patients with temporal lobe epilepsy (TLE) and hypoxic ischemic encephalopathy (HIE).

Methods: TLE ($n = 49$; 20–41 years), 8 – brain trauma (mean duration \pm SD: 6.74 ± 4.33), HIE ($n = 34$; 41–55 years), controls ($n = 16$; 25–35) were investigated. Serum ILs were measured by immunoenzyme assays using VectorBest reagents; neurotransmitters measured by ion-exchange chromatography.

Results: TLE showed high IL-6 level: Mean \pm SD 26.45 ± 6.67 vs. HC 4.44 ± 1.49 pg/ml ($P < 0.01$; $t = 2.69$). TLE and HIE showed no differences in IL-8 concentrations: 6.05 ± 1.8 and 7.1 ± 2.4 vs HC 4.46 ± 1.06 pg/ml. General IL-1b range in all TLE: 18.1–56.55 pg/ml where 23% TLE had higher IL-1b level with Mean \pm SD 24.35 ± 3.24 vs HC 19.3 ± 2.14 pg/ml ($P < 0.01$; $t = 6.4$): these patients had cognitive decline (10), neurological deficit (8) with partial epilepsy (6). ILs levels positively correlated with seizure frequency ($P < 0.05$), no differences between seizure types. TLE: glutamate was 167.89 ± 10.96 vs HC 124.37 ± 4.007 , mkmol/l; aspartate 12.6 ± 1.41 vs 8.02 ± 0.73 , mkmol/l, GABA 3.404 ± 1.22 vs 6.49 ± 0.96 , mkmol/l ($P < 0.01$); HIE: glutamate 146.27 ± 6.29 ; GABA 4.73 ± 1.69 . Low GABA with tendency to high glutamate was noted in each neurodegenerative encephalopathy occasions.

Conclusions: IL-6-mediated inflammation found in TLE was associated with high seizure frequency caused by seizure-induced microglial activation; discovered changes between neurotransmitters might underlie cytokine-mediated neuroinflammation suggesting their role in promoting seizures or establishing chronic epileptic focus. There may be potential role for antiinflammatory therapy targeting cytokines.

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Abstract – WCN 2013

No: 1839

Topic: 1 – Epilepsy

Assocation of effectiveness of epilepsy pharmacotherapy and patients' genealogical data

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About 30% of patients with epilepsy are likely to have seizures resistant to pharmacotherapy. The mechanism of pharmacoresistance is not fully understood, but clearly some factors, including family history of epilepsy, can predict pharmacoresistance.

The aim of the study was to determine the correlation between pharmacoresistant epilepsy and prevalence of relatives with epilepsy in the studied group of patients.

309 patients, all Caucasians from Transcarpathian Ukraine, mean age of 39.82 ± 8.63 , diagnosed with epilepsy participated in the study. All

were receiving antiepileptic therapy in standard therapeutical doses. Drug-resistant epilepsy was defined as uncontrolled seizures over a year despite attempts to treat with three or more different AEDs. We managed to build family tree of every studied patient viewing relatives up to 4th level of kinship, which included 6659 people.

According to genealogical research, 71 male probands had 192 relatives suffering with epilepsy and 50 female probands had 181 relatives with diagnoses of epilepsy at some point of their lives. Group with well-controlled epilepsy (group I) included 213 patients, while pharmacoresistant group (group II) was presented by 96 patients. Group I had 16.9% of probands whose relatives suffered with epilepsy, while group II had 88.5% of patients whose relatives were diagnosed with epilepsy ($p < 0.001$).

In the group of patients with pharmacoresistant epilepsy the prevalence of relatives suffering with epilepsy at some point of life was nearly 5 times higher than in non-resistant group. Family history of epilepsy can be important factor in predicting result of epilepsy pharmacotherapy.

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Abstract – WCN 2013

No: 1853

Topic: 1 – Epilepsy

Treatment outcome in epilepsy caused by intracranial vascular malformations

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Intracranial vascular malformations may be asymptomatic but may also cause partial onset seizures (POS) and lesional epilepsy, intracranial bleeding and different neurological deficits.

The aim is to evaluate the treatment outcome in epilepsy caused by intracranial vascular malformations.

Eleven patients with (POS), females/males – 8/3, aged 15–50 years, were evaluated clinically, electrophysiologically and with MRI. Four patients had childhood seizure onset, five adolescence and two adulthood. Cerebral cavernous malformation (CCM) was found in four, cerebral arterio-venous malformations (AVM) in four and Sturge–Weber syndrome (SWS) in three. Two patients with SWS and one with AVM developed hemiparesis contralateral to the vascular malformation, additional two patients developed hemiparesis after surgery (one after AVM embolisation, one after CCM lesionectomy) improving gradually.

Three of four patients with AVM underwent surgery, only one is seizure free with AED and one is only on pharmacological treatment with rare POS. Three of four patients with CCM underwent surgery, all are seizure free while still on AEDs, one of them after polytherapy with two AEDs. One patient with multiple CCM is on AEDs, with rare seizures during pregnancy with carbamazepine monotherapy when acetazolamide was withdrawn. Three patients with SWS are on AEDs without seizure control, but with rare seizures in two and frequent seizures in one, who developed hemiparesis and intellectual deterioration during the course of the disease from childhood to adulthood.

AEDs, surgery or both should be used for intracranial vascular malformations treatment to prevent seizure reoccurrence, neurological and intellectual deficits thus improving the life quality.

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Abstract – WCN 2013

No: 1926

Topic: 1 – Epilepsy

Epilepsy in African cultural approaches

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The countryside in Africa is occupied by more than 80% of the population and significant progress are made by individuals, families and the community in the management of epilepsy.

Several cross-sectional studies taking into account the qualitative aspects were conducted in several locations. These studies have focused on the general population, patients living with epilepsy, accompanying persons, traditional healers, health officials and notables. The knowledge and interpretation of epilepsy are wrong. Epilepsy is considered to be supernatural, scary, dishonourable, contagious, incurable disease, deadly whose cause may be witchcraft, heredity or the transgression of taboos and prohibitions. The consequences are failure to comply with the treatment and the resurgence of epileptic seizures. For example, Benin is a high prevalence area because the rate of epilepsy varies from 15 to 38.4% but the average national is valued at 8%.

Preventive treatment through the respect of banned cultural as sexual abstinence, not fresh meat and okra, the prohibition of drinking alcohol consumption and respect for taboos. Epilepsy therapeutic route begins at a traditherapeute upon the occurrence of the first crisis. He will later consult a nurse who issued the phenobarbital, only often available antiepileptic. But while effective this modern treatment will be stopped due to lack of financial means.

The implementation of the results and the recommendations of the various studies will help to positively change the habits and behaviour of populations.

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Abstract – WCN 2013

No: 1785

Topic: 1 – Epilepsy

Altered GABA_A receptor subunit expression at thalamocortical synapses in an animal model of absence epilepsy

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Absence epilepsy is a non-convulsive generalized form of epilepsy, characterized by spike-wave discharges, which predominantly affects children. It is accompanied by sudden impairment of consciousness and has an adverse impact on childhood learning. While disruption of the thalamocortical (TC) network is known to lead to generation of hypersynchronous intrathalamic oscillatory activity during absence seizures, the underlying cellular and molecular mechanisms remain largely unknown. Recent studies using human and animal models suggest region-specific changes in GABA_A receptors (GABA_AR) in the thalamus may underlie hypersynchronous oscillations in absence seizures. Therefore the aim of the current study was to investigate whether GABA_A $\alpha 1$ and $\beta 2$ subunits mediating phasic inhibition at TC synapses are altered in the well-established stargazer mouse model of absence epilepsy. Postembedding electron microscopy-immunogold cytochemistry was used to analyze density of GABA_AR subunits in epileptic stargazers compared to matched non-epileptic control littermates ($n = 6$, 200 synapses/subunit). In the epileptic mice, synaptic expression of both GABA_A $\alpha 1$ and $\beta 2$ subunits was significantly increased by 54% ($p < 0.01$) and 49% ($p < 0.01$), respectively. These findings suggest upregulation of phasic GABA_AR-mediated inhibition at TC synapses in the ventrobasal thalamus. Whether such changes occur pre- or post-seizure remains to be investigated. Enhanced phasic GABA_AR inhibition may be one of many factors contributing to increased inhibition recently reported in TC neurons of other absence models. Currently available anti-epileptics are associated with varying degrees of adverse side effects and inefficacy, thus further

studies to elucidate the mechanisms underlying hypersynchronous activity in absence seizures may highlight novel drug targets for absence epilepsy treatment.

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Abstract – WCN 2013

No: 1956

Topic: 1 – Epilepsy

Neural autoantibodies and immunotherapy-responsive epilepsy: A prospective study

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Background: Autoantibodies specific for neural antigens have been described in association with several encephalopathies which have seizures as a prominent feature. In these cases seizures are poorly controlled with conventional anti-epileptic drugs (AED). In contrast, a good response is often achieved with immunotherapies.

Objectives: To evaluate the frequency of autoantibodies specific to neural antigens in patients with epilepsy and their response to the immunotherapy.

Materials and methods: Two groups of patients were included in the study: 29 patients with epilepsy and other neurological symptoms and/or autoimmune diseases (group 1) and 30 patients with AED-resistant epilepsy (group 2). Forty-two age and sex-matched healthy subjects (HS) were included as controls. Patients' Serum and CSF, when available, were evaluated for the presence of neural autoantibodies by immunohistochemistry on frozen sections of mouse brain and by cell-based assays. GAD65-IgGs were tested by RIA.

Results: Neural-specific autoantibodies were detected in 11/59 patients (19%) but not in healthy controls. In particular, neural-specific IgGs were detected in two patients from group 1 (2/29, 7%) (anti-GAD65, 1; antibody specific for an unclassified intracellular antigen [AUIA], 1) and in nine patients from group 2 (9/29, 31%) (anti-LG11, 2; anti-GAD65, 1; AUIA, 4; antibody specific for an unclassified synaptic antigen, 2). In four patients from group 2, immunotherapy resulted in >50% seizure reduction at three months.

Conclusions: A significant percentage of patients with AED-resistant epilepsy harbor neural-specific autoantibodies and may respond to immunotherapy.

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Abstract – WCN 2013

No: 1948

Topic: 1 – Epilepsy

Sustained release forms: Application to antiepileptic drugs in developing countries

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Background: Epilepsy is a chronic neurological disease affecting more than 70 million people worldwide, among them nearly 90% in developing countries (DCs). There, primary healthcare is extremely limited, only four major antiepileptic drugs (AEDs) are used: phenobarbital, carbamazepine, sodium valproate and phenytoin. Furthermore, there is also a problem of accessibility, availability and quality of drugs.

Objective: The main hypothesis of the study was based on the fact that a “long” sustained release form that would reduce the number of drug's administrations would allow overcoming the problems of compliance and accessibility.

Methods: A literature review of sustained release forms applicable to an AED was performed and the primary endpoint was duration of active ingredient release longer than 24 h. We also assessed the feasibility and acceptability in DCs.

Results: Two AEDs were found not to be of conventional release: carbamazepine and sodium valproate. But they are not of sustained release. Several techniques were available including: esterification, transdermal devices, liposomes and polymeric devices preformed or *in situ* formed. *In situ* methods for the preparation of injectable biodegradable microparticles or implants for the controlled delivery seemed best suited to our objective. Furthermore, they seem to fulfill the requirements of feasibility and cost. Sodium valproate appeared to be the most suitable molecule by reducing its cost and its hepatotoxicity with a “long” sustained release form.

Conclusion: This new approach to the treatment of epilepsy would solve many problems in the DCs. However, preclinical and clinical studies are required to obtain an approval.

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Abstract – WCN 2013

No: 1961

Topic: 1 – Epilepsy

Preventive and treatment aspects of the post-traumatic epilepsy

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Background: Posttraumatic epilepsy is one of the serious consequences of traumatic brain injury (TBI). To achieve the timely prevention and control of paroxysms therapeutic measures should be directed to the correct selection of major antiepileptic drugs and dose titration to the initial therapy.

Objective: To study the effectiveness of antiepileptic drugs in prevention and treatment of post-traumatic epilepsy.

Material and methods: We examined 48 patients with different periods of TBI who were treated at the Emergency Unit Hospital. All patients were divided into 3 groups:

- I with acute TBI (18),
- II with intermediate period of TBI (15),
- III with distant period of TBI (15).

The control group was 15 almost healthy individuals. Were used clinical, neurological, instrumental, laboratory, and statistical methods.

Results: The patients of the first group received antiepileptic drugs with preventive aim after an epileptic seizure in anamnesis. The best results were obtained with lamotrigine ($p < 0.05$). Posttraumatic epilepsy patients in groups II and III revealed the following manifestations: generalized seizures (65.3%), focal seizures (24.2%), combined paroxysms (11.5%), paroxysmal activity on EEG with a predominance of polyphasic complexes (69.4%), spike-waves (63.1%), and sharp waves (78.3%). In these groups of patients was the most efficient use of combination therapy of valproate and lamotrigine ($p < 0.05$).

Conclusions: Thus, our findings demonstrate the effectiveness of lamotrigine for the prevention of post-traumatic epilepsy. And high efficiency combined therapy using valproate and lamotrigine in the treatment of post-traumatic epilepsy.

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Abstract — WCN 2013**No: 1991****Topic: 1 — Epilepsy****Normalized distribution of EEG frequencies within one map based on source density analysis for individual recognition of pathological features**

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Background: Subjective interpretation of EEG raw data is still the preferred method of evaluation in patients.

Objective: We now describe a method for objective quantitative EEG analysis, which allows online real time recognition of individual pathological deviations from normality.

Method: EEG data derived from 16 channels are processed online in real time using source density analysis. After non-linear LaGrange interpolation of virtual electrode positions a map corresponding to 64 channels is displayed (Software “neoCATEEM®” from Mewicon GmbH, 4164 Schwarzenberg, Austria). Conversion of frequency content into spectral colors and application of the RGB mode results in a single map. Spectral frequency maps are continuously displayed by means of a moving time average (up to 3 min).

Result: Normalization of EEG data within one frequency range is achieved by calculating median values of all corresponding electric power values from all electrode positions to give 100% reference values. Data are compared statistically online real time to a database containing data from 300 healthy brains. Focal aberrations of the frequency content were much better recognized and gave better statistics using source density in combination with individual normalization than using absolute power values. Patient data were compared suffering from epilepsy (enhanced theta power at T3), migraine (enhanced alpha1 and alpha2 power at F7 and T3) and psychosis (enhanced beta1 power at T6).

Conclusion: Individual normalization of quantitative EEG data in combination with an automatic comparison to a database might serve for individual rational pharmacotherapy by using drugs able to modulate the particular disturbed frequencies.

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Abstract — WCN 2013**No: 2003****Topic: 1 — Epilepsy****Oligohydrosis in epileptic patients treated with zonisamide**

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Background: Zonisamide is classified as a sulfonamide and is characterized having multiple antiepileptic action-mechanisms including inhibiting carbonic anhydrase, which may lead to the oligohydrosis.

Objective: The purposes of this study include the following: (1) to determine the incidence and (2) to reveal the risk factor of oligohydrosis-related symptoms in epileptic patients treated with zonisamide.

Patients and methods: I prospectively studied 153 patients under ages of 20 who were newly diagnosed with epilepsy or referred from other hospitals for controlling a seizure. The patients were treated with zonisamide as a monotherapy or adjuvant therapy. The data was collected by direct interview at least 3 months after taking zonisamide. Facial flushing, lethargy, itching sensation, irritability with hyperthermia, heat sensation and heat intolerance were considered as an oligohydrosis-related symptom.

Results: 24.8% of patients were treated by zonisamide as a monotherapy, and the other patients were treated by zonisamide as an adjuvant therapy. The oligohydrosis-related symptoms were observed in 11.1% of patients, and 2% of the patients have stopped taking zonisamide due to the symptoms. The oligohydrosis-related symptoms were observed more frequently in the patients between 15 and 20 years old than younger ages, and more frequently in the patients who had taken topiramate.

Conclusion: The frequency was significantly higher than the results from previous studies. Clinicians should monitor the patients who are taking zonisamide regarding the oligohydrosis-related symptoms. Especially, the patients between ages of 15 and 20 years old and the patients who have a drug history of topiramate should be observed carefully.

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Abstract — WCN 2013**No: 2006****Topic: 1 — Epilepsy****Effect of cognitive stimulation on hippocampal ripples in epileptic patients**

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Interictal HFOs (ripples and fast ripples) have been repeatedly identified in recordings from depth macroelectrodes in epileptics. In contrast to fast ripples, which are believed to reflect the neuronal substrates of epileptogenicity, ripples are considered to be a signature of both normal and epileptic brain processes. The differentiation of physiological and epileptic ripples in intracranial recordings remains unavailable.

We analyzed SEEG recordings in seven patients with intractable partial seizures in whom hippocampal activity was recorded in resting state and subsequently during simple cognitive task with randomly presented frequent and rare visual stimuli. Using automated detection of ripples based on length of power envelope, we analyzed potential differences in ripple rate (RR) in the cognitive versus resting period, within epileptic (EH) and non-epileptic hippocampi (NH). Further direct impact of cognitive stimuli on ripples (immediately after the stimulus) was investigated.

Ripples have been detected within hippocampal recordings in all the investigated subjects. Mean RR in resting periods was 11.36 ± 8.34 /min within EH, and 10.13 ± 8.75 /min within NH. In the cognitive task periods mean RR within EH and NH decreased to 7.16 ± 5.75 /min, and 8.75 ± 7.69 /min respectively. The reduction of RR during cognitive stimulation was significant in EH, but not in NH. Interestingly we observed a transient suppression of ripples in both EH (slight) and NH (significant) in the first second after the stimuli onset, followed by a significant increase in RR for both EH and NH.

Our results point to different reactivity of ripples within EH and NH to the cognitive stimulation.

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Abstract – WCN 2013**No: 2011****Topic: 1 – Epilepsy****Ketamine in refractory status epilepticus – A retrospective study on 16 patients**

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Background: Refractory status epilepticus (RSE) is defined as status epilepticus (SE) not responding to first- and second-line antiepileptic drug (AED) therapies. Ketamine is a noncompetitive NMDA receptor antagonist.

Objectives: Few case reports and retrospective studies on ketamine in SE are available. We aimed to evaluate safety, tolerability and outcome.

Patients and methods: We retrospectively analysed data of patients with RSE (n = 84) treated in our neurological intensive care unit from 01/2012 (n = 70) to 02/2013 (n = 14). We analysed aetiology, duration, type of SE, daily dose of ketamine, cotherapeutic agents, treatment response and disposition.

Results: In 16/84 (19%) patients (median age 68.5 years; range 50–80) with RSE, ketamine was used in combination with midazolam. Causes of RSE: 6/16 post-anoxic, 3/16 systemic infection, 3/16 stroke/intracerebral haemorrhage, 2/16 unknown, 2/16 preexisting epilepsy with low AED level. RSE types: 2/16 myoclonic SE, 4/16 convulsive SE, 10/16 nonconvulsive SE. Ketamine was initiated median on 4th day of SE (range 1–18); median duration of treatment 4.5 days (range 1–22). In 4 patients ketamine was started with a bolus of 200 mg, followed by median continuous infusion 187.5 mg/h (range 150–200); 12 patients received continuous infusion from the onset, median 187.5 mg/h (range 75–250). Median maximum dose was 175 mg/h (range 100–300). RSE was terminated in 10/16 patients; 11/16 patients died due to either RSE (6/11) or their main disease despite terminated SE (5/11).

Conclusion: Ketamine may be effective in later stages of SE when adequate first and second-line treatments fail, but overall outcome remains poor with 70% in-hospital mortality.

doi:10.1016/j.jns.2013.07.132

Abstract – WCN 2013**No: 1796****Topic: 1 – Epilepsy****Clinical utility of (¹⁸F)-florofluzazenil pet in presurgical evaluation of refractory focal epilepsy**

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Gamma-aminobutyric acid is the brain's main inhibitory neurotransmitter and has already been implicated in the pathophysiology of epilepsy by several investigations. A selective antagonist of GABA_A-benzodiazepine receptors, which is frequently used, is the radioligand ¹¹C Flumazenil. In patients with intractable focal epilepsy a localized reduction in (¹¹C) FMZ binding is demonstrated, which correlates closely with the area of seizure onset.

We used as one of the first programmes in middle Europe 185 Mbq-¹⁸F-Flumazenil in the preoperative evaluation of 37 patients with refractory focal epilepsy. ¹⁸F-Flumazenil has a longer half-life (2 h) compared to the more commonly used ¹¹C Flumazenil (20 min). 14 of

our patients had finally been operated. 11 patients had temporal lobe epilepsy, in 3 patients an extra temporal epileptogenic area had been identified.

Eight patients had concordant unilateral focal results. Interestingly, two patients had neither a focal lesion on MRI nor any lateralization or localization in the FMZ PET. Seizure semiology and ictal EEG were guidance for successful intracranial monitoring. The postoperative outcome was excellent (one reached Wieser1a, one Wieser1). Two patients showed a bilateral temporal abnormal FMZ PET result, both had unilateral MRI pathology. Postoperative outcome in these two patients was non favourable (one Wieser4, one Wieser5).

The general postoperative results showed that fortunately 78% had a favourable outcome (Engel I&II) and 22% had a non favourable outcome (Engel III&IV).

In summary, ¹⁸F-FMZ-PET is a helpful tool in the preoperative investigation of patients with refractory focal epilepsy. Discordant results may predict less favourable outcome.

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Abstract – WCN 2013**No: 2015****Topic: 1 – Epilepsy****A double-blind extension study to assess long-term safety/efficacy of zonisamide versus carbamazepine monotherapy for treatment of newly diagnosed partial epilepsy**

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Objective: To investigate the long-term safety and maintenance of efficacy of zonisamide versus carbamazepine monotherapy for partial seizures in adults with newly diagnosed epilepsy.

Patients and methods: A long-term, double-blind, extension study was conducted in patients completing a Phase III non-inferiority trial comparing zonisamide and carbamazepine monotherapy. Patients continued their randomised treatment, with dosing adjusted according to tolerability/response within permitted dose ranges (zonisamide 200–500 mg/day; carbamazepine 400–1200 mg/day). Safety assessments included treatment-emergent adverse events (TEAEs) and clinical laboratory parameters. Efficacy assessments included retention rate (primary assessment) and seizure freedom rate.

Results: Overall, 120/137 (87.6%) zonisamide patients and 134/158 (84.8%) carbamazepine patients completed the study. For zonisamide versus carbamazepine, incidences were similar for TEAEs (52.6% vs. 46.2%), serious treatment-related TEAEs (0.7% vs. 1.9%) and TEAEs leading to withdrawal (1.5% vs. 0.6%). Most TEAEs (>90%) were of mild/moderate intensity; the most frequently reported being decreased weight (5.8% vs. 0%) and headache (4.4% vs. 6.3%). Zonisamide was associated with small-to-moderate decreases in bicarbonate levels from baseline (mean –3.4 mmol/L). Vital signs and physical/neurological examinations revealed no safety concerns. For zonisamide versus carbamazepine, retention rates after 12, 18 and 24 months were 58.4% vs. 61.4%, 27.7% vs. 27.8% and 5.8% vs. 2.5%, respectively; and the proportion of patients remaining seizure free for ≥24 months was 32.3% versus 35.2% (intent-to-treat population).

Conclusion: Once-daily zonisamide monotherapy demonstrated favourable long-term safety and maintenance of efficacy in treating partial seizures in adults with newly diagnosed epilepsy. No new or unexpected safety findings emerged.

Supported by Eisai.

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Abstract — WCN 2013**No: 1265****Topic: 1 — Epilepsy****Cognitive malfunction impairment and stigma in HIV/AIDS epileptic patients in Kenya**

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Background: Epilepsy is one of the most prevalent neurological conditions and knows no age, racial, social class, geographic, or national boundaries. The impact of epilepsy rests not only on the individual patient, but also on the family and indirectly on the community. The burden of epilepsy may be due to the physical hazards of epilepsy resulting from the unpredictability of seizures; the social exclusion as a result of negative attitudes of others toward people with epilepsy; and the stigma.

Objective: The impact of the many infections on neurological morbidity and mortality is caused by bacterial meningitis, particularly that due to pneumococcal and meningococcal organisms, is still common. The neurological outcomes of these readily treatable infections depend on the availability of and access to health services, which differ tremendously across the region. The situation is made worse where populations are displaced by conflicts. Epidemics of meningococcal disease are particularly frequent in the meningococcal belt.

Results: Viral encephalitis is on the increase, where the prevalence of HIV infection is high as well. The HIV virus enters the nervous system within hours of an individual's becoming infected. Acute inflammatory demyelinating polyneuropathy (Guillain-Barré syndrome), which can cause paralysis leading to death from respiratory failure, often accompanies this initial HIV infection and occurs with greater frequency in those infected with HIV/AIDS than in uninfected populations.

Conclusions: Opportunistic infections of the nervous system occur in about 30 to 40% of those with AIDS. Neurological complications of HIV infection have become major, overwhelming components of the health burden.

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Abstract — WCN 2013**No: 2067****Topic: 1 — Epilepsy****Low concentrations of lead diminish the threshold of pentylenetetrazole induced seizure in mice**

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Background: Lead is one of the most common toxic metals and its exposure is associated with several neurological damages and behavioral disturbances. There is controversy in the results on the relation of lead exposure and seizure incidence.

Objective: We examined the effects of short-term exposure of low lead concentrations on the chemical seizure induced by pentylenetetrazole (PTZ).

Material and methods: Mice received lead acetate in drinking water for 30 days at concentrations of 50 and 100 ppm. Slow infusion of

PTZ (5 mg/kg, 0.3 ml/min) was utilized to induce seizure. The latency between the start of infusion and onset of convulsion signs including myoclonic twitch, face and forelimb clonus, running and bouncing clonus and tonic hindlimb extension were recorded and then converted to threshold convulsion dosage.

Results: Following 30 day exposure, blood lead level in both the experimental groups was significantly higher than the control group ($p < 0.01$). The exposure to 100 ppm of lead reduced seizure thresholds ($p < 0.001$) in all phases of convulsion while 50 ppm of lead did not ($p > 0.05$). Significant effect of 100 ppm lead acetate indicates that even this concentration of lead increases the possibility of seizure attacks in lead exposed populations. Lead acetate at a concentration of 50 ppm did not affect, thus 100 ppm maybe considered as minimal effective concentration of lead.

Conclusion: Several mechanisms such as Ca replacement or BBB damage are suggested for this effect but exact mechanisms are unclear. Nevertheless more well-designed studies may be executed to deliver further accurate information.

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Abstract — WCN 2013**No: 2076****Topic: 1 — Epilepsy****A model for an enhanced screening tool for depression in people with epilepsy**

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Background: Depression is a frequent comorbidity in people with epilepsy. To increase the detection of this frequent comorbidity various screening tools have been described, in particular NDDI-E. We have previously validated NDDI-E and a visual analogue scale, ET, in a multicultural London outpatient setting.

Objective: The aim of the current study is to create a new more efficient screening tool for depression in people with epilepsy.

Methods: Data from 250 patients was prospectively collected in London from 05/2009 to 02/2010. Logistic Regression models and recursive partitioning techniques (classification trees, random forests) have been applied to identify an optimal subset of the 13 items and provide a framework for the prediction of class membership probabilities (for the DSM-IV based Depression classification as "Major"/"Nil").

Results: Both logistic regression models and classification trees (respectively random forests), suggest the same choice of items for classification (NDDI-E 4, NDDI-E 5, ET-Distress, ET-Anxiety, ET-Depression). The most useful regression model includes all 5 mentioned variables and outperforms the NDDI-E as well as the ET with respect to ROC. Classification trees were grown by restricting the possible splitting variables to different subsets of items. In all considered situations the ROC curves indicated more precise classification than the original screening tests, though the trees had to be cut back to ensure comprehensible splitting rules in some situations.

Conclusion: For the first time, we have created a model of a screening tool for depression containing both verbal and visual analogue scales, which should be more precise than previous tools.

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Abstract – WCN 2013**No: 2071****Topic: 1 – Epilepsy****White matter abnormalities in patients with idiopathic generalized epilepsy**

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Background: In the past decades, several studies investigating idiopathic generalized epilepsies (IGEs) disclosed subtle brain abnormalities which may be involved in the pathophysiology of this group of diseases. Investigations focused mainly in the grey matter. Few studies using structural neuroimaging explored the white matter.

Objective: To evaluate the presence of focal abnormalities in the white matter of patients with IGE patients using voxel-based morphometry (VBM).

Patients and methods: 28 patients (18 women) with IGE and 21 controls were investigated. All participants were scanned using a 1.5 T MRI. Volumetric T1 (3D) weighted sequence with isotropic voxels (1 mm) was used for analysis. VBM was conducted with SPM8 and VBM8 toolbox. Processing involved normalization, automatic segmentation and smoothing of the grey matter and white matter separately. Statistical analysis was performed with two sample T tests to compare differences in the grey matter and white matter between patients and controls. Level of significance selected was $p < 0.05$ corrected for multiple comparisons.

Results: Significant focal increase in the grey matter was disclosed mainly in the right subcallosal gyrus (cluster size = 746, $p = 0.04$) and left orbital gyrus ($p = 0.006$). White matter analysis showed focal atrophy in the anterior cingulate limbic lobe and cingulate gyrus (cluster size = 5020, $p < 0.0001$).

Conclusions: This investigation supports structural abnormalities in the grey matter and white matter in the frontal and limbic lobes in patients with IGE. White matter abnormalities disclosed located in the mesial parasagittal structures are in accordance with previous investigations.

Study supported by: FAPESP.

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Abstract – WCN 2013**No: 2072****Topic: 1 – Epilepsy****Clinical and diagnostic features of migraine–epilepsy**

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According to modern concepts migraine and epilepsy are comorbid neurological diseases with similar pathogenesis and clinic. The prevalence of headache in patients with epilepsy is 59%, and a quarter of them had migraine.

The study features clinical neurology, EEG and neuroimaging indicators of migraine and epilepsy. Complex clinical neurology, EEG, neuroimaging (MRI, PET, NMRS and MR tractography), and psychological examination were performed in 47 patients with migraine, 50 patients with locally due to epilepsy and 17 – with a migraine–epilepsy.

We found that patients with migraine most frequently reported the distribution of a hemicrania with emphasis in the fronto-temporo-orbital region. The hemicrania was permanent, had a pulsating nature and was often in conjunction with accompanying symptoms. In patients with locally induced epilepsy in the interictal period, there are different types of headaches: 28% – migraine with aura and/or without, 33% – tension

headache, and 8% – unclassifiable headache. According to the video-EEG monitoring it is most common in patients with migraine (67%) and migraine–epilepsy (88%) to have a combination of focal and generalized epileptiform activity emanating from the back and/or temporal lobes. MRI in patients with migraine and migraine–epilepsy were discovered extension of the temporal horn of one of the lateral ventricles (33 and 25%), cranio-vertebral anomalies and other MP-changes.

Thus, the differential diagnosis of migraine and epilepsy locally by conducting a comprehensive examination of patients with the inclusion of clinical neurology, EEG and neuroimaging techniques, allows you to set the type of attack and objectify further medical therapy.

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Abstract – WCN 2013**No: 2073****Topic: 1 – Epilepsy****Neurophysiological study in patients with benign adult familial myoclonus epilepsy**

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Objective: The aim of the present study is to investigate cortical excitability in patients with benign adult familial myoclonus epilepsy (BAFME), using transcranial magnetic stimulation (TMS).

Methods: Single-pulse and paired-pulse TMS were performed in 6 BAFME patients to obtain multiple measures from the first dorsal interosseous muscle, including resting motor threshold (rMT), active motor threshold (aMT), contralateral silent periods (CSP), and short-interval intracortical inhibition (SICI) and intracortical facilitation (ICF). Age-matched twenty-one healthy subjects were also studied. Also all patients had recorded somatosensory evoked potential (SEP) elicited by digital nerve stimulation.

Results: rMT, aMT and SICI were significantly decreased compared to control group. And CSP was significantly shortened in the patients compared to control groups (rMT: $p < 0.01$, aMT: $p < 0.01$, CSP: $p < 0.001$, SICI-2 ms: $p < 0.05$). In contrast, there were no significant differences between patients and control groups in ICF. Significant correlations were not observed between SEP amplitude and TMS measures.

Conclusions: These findings suggested that there was motor cortical hyperexcitability secondary to the loss of intracortical inhibition, in addition to hyperexcitability of the sensory cortex in patients with BAFME.

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Abstract – WCN 2013**No: 2134****Topic: 1 – Epilepsy****The potential efficacy of intravenous lacosamide in epilepsy at hospital cases**

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Objectives: To appraise the efficacy of the lacosamide, third generation antiepileptic, for treatment of different cases of crisis, including status, with intravenous administration.

Material and methods: Patients with different types of crisis, who were assessed, while their hospitalization in the Lozano Blesa Clinic Hospital, Zaragoza. The cases were treated with intravenous lacosamide (ivLCM). The information was collected by retrospective analysis. The efficacy was defined as the cease of the crisis with the treatment. It could be in the first line or more. The data was recollected with standardized questionnaire by a neurologist.

Results: Nineteen patients were selected, the average age was 50 years, 68.4% men. 63.2% had a history of previous crisis, of them 41.7% were generalized, 33.3% complex focal and 25% simple focal. At the moment of the use of ivLCM 42.1% were in status (22.2% convulsive status, 66.7% no convulsive and 11.1% focal) and 57.9% no status. The ivLCM was used at the first instance in 31.6%, the second in 57.9% and both the third and fourth in 5.3%. Initially used doses were, 200 mg in 78.9%, 100 mg in 15.8%, and 400 mg in 5.3%. The maintenance doses were 200 mg/12 h in 72.2% and 100 mg/12 h in 27.8%. The obtained efficacy was in the second day, average, with a positive response of 89.5%. No side effect was registered.

Conclusions: The LCM has shown efficacy and good tolerance, even beyond use as a third-line treatment.

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Abstract — WCN 2013

No: 357

Topic: 1 — Epilepsy

Role of postoperative antiepileptic drug withdrawal in children: Long-term seizure outcome

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Background and aim: The effect of time to stop AED postoperatively in children on long-term seizure outcome is still not clear. We aimed to assess the effect of timing of AED withdrawal on subsequent seizure recurrence and long-term seizure outcome in children.

Methods: This retrospective study included 150 child patients who underwent cranial surgery between January 2007, and December 2011, and who started AED withdrawal after having reached postoperative seizure freedom. Patients had at least 1 year postoperative follow-up. Time intervals from surgery to start of AED reduction (Time to Reduction = TTR), and complete discontinuation (Time to Discontinuation = TTD) were studied in relation to seizure recurrence during or after AED withdrawal.

Results: Mean age was 8.3 year (range 2–15 years). Median TTR and TTD were 12 months and 35 months respectively. 71 patients had seizure recurrence during or after AED withdrawal. After a mean postoperative follow-up of 31 months, 85 patients were seizure free for at least 1 year. TTR and TTD showed statistically significant relation to regain seizure freedom after restart of drug treatment, to seizure freedom, and to cure (defined as being seizure free and off AEDs for at least 1 year) at final follow-up.

Conclusion: Early postoperative AED withdrawal dose affect long-term seizure outcome and cure. Also it might identify patients who need continuous drug treatment and preventing unnecessary continuation of AEDs in others.

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Abstract — WCN 2013

No: 2088

Topic: 1 — Epilepsy

Spectral analysis of EEG in patients with drug resistant epilepsy

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Objective: To study spectral characteristics of electric paroxysmal activity (PA) of the EEG in patients with localization-related drug resistant epilepsy (DRE) with structural changes in the hippocampus and evaluate their relationship with the occurrence of the phenomenon of secondary bilateral synchronization syndrome (SBS).

Materials and methods: 42 patients with DRE were studied, 23 — with MRI-detected hippocampus sclerosis (HS), and 19 — without HS. Spectral analysis (SA) was performed in EEG fragments with PA, average spectral parameters for 4 second recording periods were calculated.

Results: The data analysis of quantitative EEG revealed topographical differences in spectral power values of PA between the groups. The most pronounced results were obtained for the delta and theta rhythms. In the first group of patients average spectral power of delta waves in leads Fp1, Fp2, F3, F4, C3 and C4 was significantly lower than in second group ($p < 0.05$). A similar trend is observed in the range of the theta rhythm. Even distribution of spectral capacity of delta and theta rhythms with no significant fronto-occipital differences in all recorded leads found out in the first group. A gradient growth of spectral power of theta waves in the direction of the occipital lobes to the frontal determined in patients with HS.

Conclusion: Thus, the structural changes of the hippocampus influence on the severity of PA and on neurophysiological mechanisms of SBS occurrence. SA could be used for the early diagnosis of SBS phenomenon and selection of treatment strategy in patients with DRE.

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Abstract — WCN 2013

No: 2159

Topic: 1 — Epilepsy

A novel intronic variant of SCN1A gene responsible for severe epileptic encephalopathy with refractory status epilepticus

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Objective: To describe a 32 year old male with encephalopathy and drug resistant epilepsy with frequent status epilepticus.

Patient and methods: The patient was born at term by normal pregnancy after prolonged labor. A significant global developmental delay had been recognized since 6 years of age. At age 5 months he experienced the first epileptic seizure during hyperpyrexia characterized by a left hemiconic seizure during sleep followed, a few hours later, by a tonic-clonic seizure. Since then he had monthly febrile and afebrile tonic-clonic seizures, mainly arising from sleep. Despite trials of several antiepileptic drugs, seizures increased on frequency, becoming weekly. Recurrent episodes of intractable status epilepticus worsened by phenylhydantoin were also reported. At our first assessment (32 years of age) the patient underwent a comprehensive clinical, neurophysiological and neuroradiological study.

Results: Neurological examination showed craniofacial dysmorphism and severe delay in language and motor acquisition. Interictal EEG showed diffuse background slowing and independent epileptiform discharges over both the temporal regions. During prolonged video-EEG monitoring we recorded one of his typical status epilepticus clinically characterized by left arm clonic seizures and secondarily generalized seizures. Brain MRI disclosed a mild cerebellar atrophy with evidence of the cerebellar sulcus. Molecular analysis of SCN1A gene identified a de novo intronic mutation, causing nucleotide substitution c.338+3A>C.

Conclusions: We describe a new de-novo mutation of SCN1A gene responsible for severe early-infantile epileptic encephalopathy with recurrent status epilepticus resistant to sodium blocker AEDs.

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Abstract – WCN 2013

No: 1611

Topic: 1 – Epilepsy

Seizure due to imipramine discontinuation: A case report

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Imipramine serotonin, noradrenalin and lower level of dopamine block the reuptake pump. In particular, it is used in nocturnal enuresis treatment. It is known that high dose of imipramine causes epileptic activity. However, seizure because of depending on the discontinuance of imipramine is rarely seen. A 19 year old male patient had been taking 75 mg/day imipramine regularly for 1 year for nocturnal enuresis treatment. Patient had been contracted with seizure after sudden discontinuance of medicine. Routine biochemical inspections, EEG and cranial MRG findings were evaluated normal and it was considered that sudden discontinuance of imipramine caused seizure. Consequently; we would like highlight that in order to avoid seizure depending on discontinuance of imipramine, it is necessary to discontinue imipramine by decreasing it.

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Abstract – WCN 2013

No: 2197

Topic: 1 – Epilepsy

Personality characteristics of epileptic and non-epileptic using personality factorial inventory

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This article aims to demonstrate the comparison of personality characteristics between groups of epileptic patients and the general population. The survey was conducted with people aged between 18 and 60, living in Salvador, Bahia. We sought to identify the intraception, performance, autonomy, aggression and social stigmas. The methodology applied in this research work was to use the Personality Factorial Inventory (PFI), in a universe of 200 individuals, 100 epileptic patients and 100 general population (caregivers, staff and students). All completed the informed consent and provided answers about the socio-demographic information on the total of 10 items. Participants were interviewed at the Foundation of Neurology and Neurosurgery (FNN) and the University Hospital Professor Edgard Santos (HUPES)–UFBA. The test consisted of 155 questions, in order to measure 15 dimensions of personality. Among male patients with epilepsy, the analysis of data showed the following dominant factors in the personality profile of average behavior: a) high average level of denial, revealing individuals who indulge in resignation and accept the punishment and guilt; and b) low level of performance, indicating subjects with difficulty performing independent activities. While, in females patients with epilepsy showed a profile with a high concentration of anger, hatred and self-confidence. The study shows the characteristics of personality to an association between the dimensions of PFI in epileptics. They are more concerned with themselves, less

interactive and dominant. The results obtained in the general population present a strong rejection towards epilepsy due to ignorance and fear of contamination.

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Abstract – WCN 2013

No: 1119

Topic: 1 – Epilepsy

Contralateral blink inhibition is a possible pathomechanism for ictal unilateral eyelid blinking – A case report

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Purpose: Little is known about ictal unilateral eyelid blinking (UEB) as a lateralizing sign in focal seizures. We present a case study and a comprehensive review of the literature and the pathophysiology of UEB.

Methods: We report a 38 year old, right-handed woman with intractable epilepsy, who underwent prolonged non-invasive video-EEG monitoring.

Results: A total of four complex focal seizures (CFS) with UEB were recorded. Semiological signs in all CFS were bilateral eyelid blinking, occurring in median 29 s after EEG-onset (range 18–37 s, median duration 9 s, range 6–11 s), followed by UEB on the right, in median starting 51 s after EEG-onset (range 30–71 s, median duration 17 s, range 10–34 s). UEB did not evolve into any clonic activity of the face. During one CFS, bilateral eyelid blinking recurred after UEB with higher blink frequency on the right. Further lateralizing seizure phenomena were ipsilateral head turning (in 2/4), contralateral eye deviation (in 3/4), aphasia (in 2/4), postictal impaired figural memory (in 1/4) and postictal nose wiping (in 2/4 CFS, respectively). Ictal EEG showed a right fronto-temporal seizure onset over F8, F8–Fp2 or Fp2–F4. Interictal EEG showed right fronto-temporal sharp waves over F8, F8–T4, F8–Sp2 or T2 and left temporal focal slowing. **Conclusion:** UEB was ipsilateral to the right fronto-temporal EEG pattern in this patient occurring only in CFS. Due to the asymmetric blink frequency during bilateral eyelid blinking, we hypothesize ictal UEB to be a contralateral blink inhibition, caused by activation in fronto-temporal cortical areas and mediated by trigeminal fibers.

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Abstract – WCN 2013

No: 2230

Topic: 1 – Epilepsy

When epileptogenesis occurs in patients with neurocysticercosis?

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Background: Neurocysticercosis (NC) is the most frequent neurologic parasitosis. Its role is indisputable for the etiology of many neurological disturbances. Epilepsy is the most common clinical manifestation.

Objective: To determine when epileptogenesis occurs in patients with NC, according to the evolutionary phases of the disease.

Method: 157 patients diagnosed with NC diagnosis through anamnesis, cranium computed tomography (CCT), analysis of liquor and electroencephalogram were included in this paper. The cysticercus lesions in CCT were classified in four phases.

Results: Seizure was the most important clinical form, appearing in 80.3% of the patients. Statistical significance ($p = 0,001$) was shown

in correlation between the convulsive form and phase II in CCT, which was performed in the beginning.

Discussion: CCT is very helpful in NC. There are many controversies, concerning epilepsy related to NC, specially, due to different forms of pathological manifestations of the infection. The appearance of the clinical forms, mainly in the initial phases of the disease can be derived from the immune stimulus by the parasite with local inflammatory response and important perilesional edema.

Conclusion: Phase II was the most exuberant phase of the disease, being important to rise seizure.

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Abstract – WCN 2013

No: 1228

Topic: 1 – Epilepsy

Is serotonin receptor HTR1B implicated in mesial temporal lobe epilepsy development?

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Background: Evidences from animal models have demonstrated that depletion of brain serotonin (5-HT), a neurotransmitter with a pivotal role in neurodevelopment and brain plasticity, lowers the threshold to induced seizures. It was also demonstrated that anti-epileptic drugs increase endogenous 5-HT concentrations and that 5HT-1B receptors could have an anticonvulsant role. Association studies have demonstrated that a polymorphism (rs6296) in 5HTR-1B gene may be a susceptibility factor for with temporal lobe epilepsy (TLE) development. The rs6296 G allele has been associated with decreased receptor activity. Our aim was to analyse the association between rs6296 and the development and clinical features of Mesial Temporal Lobe Epilepsy (MTLE) in a Portuguese population.

Material and methods: A cohort of 121 MTLE patients (67 F, 54 M, mean age = 44 ± 11 years, age of onset = 13 ± 9 years) was compared with a cohort of 192 healthy individuals (HI). All patients had Hippocampal Sclerosis (MTLE-HS). Genotyping was performed by TaqMan real time PCR methodology.

Results: rs6296 G allele was overrepresented in MTLE patients compared to controls (80.2% in MTLE vs 72.1% in HI, $p = 0.029$ OR = 1.561 (1.060–2.298)). We constituted 2 MTLE-HS sub-groups, according to febrile seizure antecedents and no differences in rs6596 allelic or genotypic frequencies were found.

Conclusion: The rs6296 G allele may be a susceptibility factor to MTLE-HS development. Since these receptors have an anticonvulsant role, a reduction in their activity could lower the threshold for seizure development.

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Abstract – WCN 2013

No: 2240

Topic: 1 – Epilepsy

Neuropsychological outcome of seizure-free temporal lobe epilepsy patients after temporal lobe resection with and without antiepileptic drugs

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Objective: Temporal lobe epilepsy (TLE) is a common clinical syndrome in adult epilepsy population with most frequently underlying pathology of hippocampal sclerosis. There is a lack of data on influence of antiepileptic drugs (AEDs) on neuropsychological outcome of seizure-free patients. In the present study we compared the neuropsychological outcome of seizure-free patients after temporal lobe resection who were off or on AEDs.

Methods: We retrospectively reviewed the neuropsychological outcome of patients with temporal lobe epilepsy who underwent either anterior temporal lobectomy or selective amygdalohippocampectomy between 1995 and 2011 at the Department of Neurology, Medical University Vienna. Seizure-free patients at last clinical follow-up were subjected to neuropsychological testing. The results of seizure-free patients that were on AEDs were compared with patients without AEDs.

Results: One-hundred-eight patients met the inclusion criteria. 49 patients were off AEDs and 59 patients were on AEDs. The mean duration of follow-up was 67.1 months. Patients without AEDs performed significantly better in most neuropsychological tests compared to patients with AEDs (HAWIE: $p = 0.003$; Verbal fluency, phonemic: $p = 0.007$; Digit Span, forward: $p = 0.042$; Digit Span, backward: $p = 0.023$; Corsi, forward: $p = 0.000$; Corsi, backward: $p = 0.001$; VLMT, learning: $p = 0.003$; VLMT, delayed recall: $p = 0.004$; VLMT, recognition: $p = 0.020$; DCS, learning: $p = 0.000$; DCS, delayed recall: $p = 0.000$; DCS, recognition: $p = 0.035$; Mosaic test: $p = 0.003$).

Conclusions: Neuropsychological outcome of seizure-free patients after temporal lobe resection is significantly better in patients that are off AEDs. The long term intake of AEDs may have significant influence on neuropsychological performance. Early discontinuation of AEDs should be considered in seizure-free patients with TLE.

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Abstract – WCN 2013

No: 2241

Topic: 1 – Epilepsy

Epilepsies of child and adolescent

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Epilepsy is a public health problem in Senegal, with a prevalence of 8.3 to 14/1000. It mainly affects children.

Objective: Of this work is to study the biographical aspects, phenotypic and evolutionary of epilepsy in a cohort of children in Senegal.

Patients and methods: This is a retrospective chart review of children with epilepsy followed up regularly at Fann University Hospital and Children's Hospital Albert Royer, July 2003 to December 2010. Inclusion criteria were: epilepsy aged under 16 years, regularly monitored for at least 3 years, with appropriate treatment, effective dose, with good adherence.

Results: We collected 522 children, aged 3 months to 16 years, with a sex ratio of 1.7 in favor of boys. The epilepsy was idiopathic in 57% of children and non-idiopathic in 43% of patients. Etiological factors were dominated by parental consanguinity, abnormal pregnancy and childbirth, infections of the central nervous system. In the group of idiopathic epilepsies not, the signs associated with epilepsy were language disorders (15.70%), behavior (15%) and motor deficits (10.32%). 22.41% of school children had learning difficulties sometimes leading to repetition or school exclusion.

Conclusion: The fight against epilepsy in Senegal implies an effective prevention policy which is necessary in improving the socio-health and the fight against infections. This is the challenge of the Senegalese league against epilepsy.

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Abstract – WCN 2013**No: 1261****Topic: 1 – Epilepsy****Efficacy and safety of eslicarbazepine acetate in acute manic episodes associated with bipolar I disorder: BIA-2093-203 phase II study results**

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Background: Eslicarbazepine acetate (ESL) was approved in 2009 by the European Medicines Agency as adjunctive therapy in adults with partial-onset seizures (POS), with or without secondary generalization.

Objective: To evaluate the efficacy, safety and tolerability of two dose-titration regimens of ESL as therapy of patients with acute mania.

Material and methods: Multicentre, double-blind, randomised, parallel-group, placebo-controlled dose-titration phase II study in patients with a diagnosis of bipolar I disorder who experienced an acute manic (including mixed) episode. Patients were randomised to: group A – ESL 600–1200–1800 mg; group B – ESL 800–1200–2400 mg; and group C – placebo 1–3 tablets. Patients were started on the lower dosage and were up-titrated as clinically required 3 and 6 days after therapy initiation; the dose was kept at the maximum for up to 3-weeks.

Results: 160 patients were included in the intention-to treat (ITT) population and 161 were included in the safety population. The ANCOVA of absolute change in Young Mania Rating Scale total score was [least square mean (p-value versus placebo)]: group A = –12.5 (p = 0.348); group B = –14.2 (p = 0.053); placebo = –10.3. Secondary efficacy analysis showed similar results with a trend for increase in efficacy across the randomised dosage-titration regimens from placebo to group B. Most common TEAEs observed in ESL groups ($\geq 5\%$ of patients) were headache, dizziness, nausea, vomiting, and diarrhoea.

Conclusion: The primary treatment comparison was not statistically significant. There was a trend towards efficacy in the ESL 800–1200–2400 mg dose group. The two ESL groups exhibited a comparably favourable safety profile.

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Abstract – WCN 2013**No: 1272****Topic: 1 – Epilepsy****Efficacy and safety of eslicarbazepine acetate in acute manic episodes associated with bipolar I disorder: BIA-2093-204 phase II study results**

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Background: Eslicarbazepine acetate (ESL) was approved in 2009 by the European Medicines Agency as adjunctive therapy in adults with partial-onset seizures (POS), with or without secondary generalization.

Objective: To evaluate the efficacy, safety and tolerability of ESL as therapy of patients with acute mania.

Material and methods: Phase II, double-blind, fixed multiple-dose, randomised, placebo-controlled, multicentre study in patients with a diagnosis of bipolar I disorder who experienced an acute manic (including mixed) episode. Patients were randomised to ESL 600 mg, ESL 1200 mg, ESL 1800 mg, or placebo and were followed for up to 3 weeks.

Results: Due to a slow recruitment rate the study was terminated early and only 38 patients were randomised. The decrease in Young Mania Rating Scale total scores was [mean (standard deviation)]: placebo = –17.7 (7.34), ESL 600 mg = –16.9 (2.75), ESL 1200 mg =

–16.7 (9.11), and ESL 1800 mg = –11.3 (SD = 10.89). At the end of the 3-week treatment period most patients in all groups were responders and most patients in all groups except ESL 1800 mg were in full remission. Overall, 30/38 (78.9%) patients experienced treatment emergent adverse events, ranging from 4/8 (50%) in the ESL 600 mg group to 10/10 (100%) in the ESL 1800 mg group.

Conclusion: Due to the early termination of the study, no definitive conclusions can be drawn regarding the efficacy, safety and tolerability of ESL for the treatment of acute manic (including mixed) episodes in patients with bipolar I disorder.

doi:10.1016/j.jns.2013.07.153

Abstract – WCN 2013**No: 1273****Topic: 1 – Epilepsy****Efficacy, safety and tolerability of eslicarbazepine acetate in the recurrence prevention of bipolar I disorder: BIA-2093-205 phase II extension study**

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Background: Eslicarbazepine acetate (ESL) was approved in 2009 by the European Medicines Agency as adjunctive therapy in adults with partial-onset seizures (POS), with or without secondary generalization.

Objective: To evaluate the dose-dependent efficacy, safety and tolerability of ESL for the recurrence prevention of bipolar I disorder.

Material and methods: Two part extension study of two phase II studies evaluated the efficacy, safety and tolerability of ESL in patients with acute mania (BIA-2093-203 and -204). In part I (results not shown), participants received open-label treatment with ESL 900 mg QD for 2 weeks. Part II followed a double-blind, parallel-group design in which participants were randomly assigned to treatment with ESL 300 mg, ESL 900 mg, or ESL 1800 mg QD up to 6 months after the last patient entered part II.

Results: Patients showing no worsening were (ITT): ESL 300 mg = 26 (76.5%), ESL 900 mg = 14 (56.0%), and ESL 1800 mg = 16 (61.5%). Patients developing manic symptomatology (ITT) were: ESL 300 mg = 3 (8.8%), ESL 900 mg = 4 (16.0%), and ESL 1800 mg = 5 (19.2%). Patients developing depressive symptomatology were (ITT): ESL 300 mg = 5 (14.7%), ESL 900 mg = 0, and ESL 1800 mg = 3 (11.5%). Differences between groups were not statistically significant. Overall, 46 (52.9%) patients reported treatment emergent adverse events (TEAE) that were considered mild to moderate in 41 (89.1%) patients.

Conclusion: There were no statistically significant differences between the ESL dosage groups in the primary and secondary efficacy analyses. ESL was overall well tolerated with no major safety differences between the groups.

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Abstract – WCN 2013**No: 1275****Topic: 1 – Epilepsy****The effects of eslicarbazepine on transient Na⁺ currents in chronically epileptic human hippocampal cells**

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Background: Loss of use-dependent block of transient Na⁺ channels is a suggested key mechanism underlying pharmacoresistance to

carbamazepine (CBZ) at the cellular level, both in human and experimental epilepsy.

Objectives: This study aimed to determine the activity of eslicarbazepine, the major active metabolite of eslicarbazepine acetate, in modulating transient Na^+ channels in isolated cells from epileptic tissue from patients with pharmacoresistant seizures.

Material and methods: Whole cell patch-clamp recordings were performed on dissociated granule cells from the hippocampus obtained from pharmacoresistant epileptic patients ($n = 26$) under control conditions and after application of CBZ and eslicarbazepine.

Results: Consistent with previous published observations, only a slight but significant slowing of fast recovery was observed upon CBZ application (mean \pm SEM, in ms): $\tau = 11.0 \pm 1.1$ and 11.7 ± 1.7 before and during application of $100 \mu\text{M}$ CBZ, respectively, $n = 8$, $p = 0.05$. A subsequent application of eslicarbazepine exerted a pronounced and significant additional slowing of fast recovery rates $\tau = 21.8 \pm 3.0$ during application of $300 \mu\text{M}$ eslicarbazepine. The effects of eslicarbazepine were concentration-dependent: $57.9 \pm 8.6\%$, $120.0 \pm 23.5\%$ and $121.1 \pm 21.7\%$ increase following application of 30, 100, and $300 \mu\text{M}$ eslicarbazepine, respectively. Accordingly, eslicarbazepine significantly inhibited neuronal firing in human granule cells.

Conclusion: These results suggest that eslicarbazepine exerts use-dependent effects resulting in reduced firing frequencies of excitatory neurons in pharmacoresistant hippocampal granule cells from patients with epilepsy, and potentially overcomes a cellular resistance mechanism to conventional AEDs. This does not preclude additional effects of eslicarbazepine on other properties of sodium channels, i.e. slow inactivation processes.

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Abstract – WCN 2013

No: 1276

Topic: 1 – Epilepsy

The effects of eslicarbazepine on persistent Na^+ current and the role of the Na^+ channel β subunits

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Background: In mice lacking β_1 subunits, carbamazepine causes a paradoxical up-modulation of persistent Na^+ currents (I_{NaP}) in the subthreshold range, leading to a failure in affecting neuronal firing (J Neurosci, 23, 8489–501, 2010). Eslicarbazepine is the major active metabolite of eslicarbazepine acetate, a once-daily antiepileptic drug approved in Europe as adjunctive therapy in adults with partial-onset seizures, with or without secondary generalization.

Objectives: To determine the effects of eslicarbazepine on I_{NaP} and the role of β subunits on the Na^+ channel.

Material and methods: To study the role of β subunits of the Na^+ channel we used a mouse line lacking either the β_1 or β_2 subunit, encoded by the *Scn1b* or *Scn2b* gene, respectively. Whole cell patch-clamp recordings were performed on CA1 neurons in $300 \mu\text{m}$ hippocampal slices under control conditions and application of $300 \mu\text{M}$ eslicarbazepine.

Results: Eslicarbazepine caused a significant reduction of maximal I_{NaP} conductance and an efficient reduction of the firing rate in wild-type mice. Eslicarbazepine did not cause a paradoxical up-regulation of I_{NaP} in *Scn1b* null mice. Consequently, the effects of eslicarbazepine on repetitive firing were maintained in these animals. In mice lacking the β_2 subunit the voltage dependence of activation was not affected when correcting for the time-dependent shift.

Conclusion: Eslicarbazepine exerts effects on I_{NaP} similar to those known for carbamazepine. In animals lacking the β_1 Na^+ channel

subunit these effects are maintained. Therefore, eslicarbazepine potentially overcomes a previously described putative mechanism of resistance to established Na^+ acting antiepileptic drugs.

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Abstract – WCN 2013

No: 1277

Topic: 1 – Epilepsy

Efficacy of eslicarbazepine acetate as adjunctive therapy of adult patients with partial-onset seizures up to one year of follow-up

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Background: Eslicarbazepine acetate (ESL) was approved in 2009 by the European Medicines Agency as adjunctive therapy in adults with partial-onset seizures (POS), with or without secondary generalization.

Objectives: To assess the efficacy of once-daily (QD) approved dosages of ESL 800 mg and 1200 mg.

Material and methods: Data from three phase III (BIA-2093-301, -302 and -303) multicentre, double-blind (DB), randomized, placebo-controlled studies in adult patients with ≥ 4 POS per 4 weeks despite treatment with 1–3 AEDs were pooled and analyzed.

Results: The DB period ITT population included 1035 patients. Compared with placebo adjusted seizure frequency over the 12-week DB maintenance period (primary endpoint) was significantly reduced with ESL 800 mg and 1200 mg ($p < 0.0001$ for both groups). Responder rate was significantly higher with ESL 800 mg (36.3%, $p = 0.0001$) and 1200 mg (43.5%, $p < 0.0001$) than with placebo (21.5%). Other secondary analyses also showed a significant effect of ESL 800 mg and 1200 mg against placebo. In the 1-year open-label extension, responder rate was 46.1% during weeks 5–16 and increased to 50.1% in weeks 41–52; proportion of seizure-free patients increased from 6.3% in weeks 5–16 to 13.6% in weeks 41–52. Mean Quality of Life in Epilepsy-31 and Montgomery-Asberg Depression Rating scales overall and several subscales scores showed significant improvements at the last assessment compared to baseline.

Conclusion: Once-daily ESL 800 mg and 1200 mg adjunctive therapy was significantly superior to placebo in reducing POS in adult patients and the efficacy was sustained up to one year of open-label treatment.

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Abstract – WCN 2013

No: 1279

Topic: 1 – Epilepsy

Tolerability of eslicarbazepine acetate as adjunctive therapy of adult patients with partial-onset seizures up to one year of follow-up

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Background: Eslicarbazepine acetate (ESL) was approved in Europe in 2009 as adjunctive therapy in adults with partial-onset seizures (POS), with or without secondary generalization.

Objective: To assess the tolerability of once-daily approved dosages of ESL 800 mg and 1200 mg.

Material and methods: Data from three phase III multicentre (BIA-2093-301, -302 and -303), double-blind (DB), randomized, placebo-controlled studies in adult patients with ≥ 4 POS per 4 weeks despite treatment with 1-3 AEDs were pooled and analyzed.

Results: DB period safety population included 1049 patients. There was a dose-dependent increase in the incidence (%) of all treatment-emergent adverse events (TEAEs) (placebo = 46.4, ESL 800 mg = 62.7, ESL 1200 mg = 67.5), possibly related TEAEs (placebo = 24.9, ESL 800 mg = 47.2, ESL 1200 mg = 55.0) and TEAEs leading to study discontinuation (placebo = 4.5, ESL 800 mg = 11.6, ESL 1200 mg = 19.3). No dose-dependent trend was found for serious TEAEs (placebo = 1.4, ESL 800 mg = 3.5, ESL 1200 mg = 3.2). TEAEs reported by $\geq 10\%$ of patients were dizziness, somnolence, headache, and nausea. The majority of TEAEs were mild or moderate in severity. The frequency of TEAEs in the ESL and the placebo groups was similar after 6 weeks of treatment. In the 1-year open-label extension 63.7% of patients reported TEAEs, 40.5% reported possibly-related TEAEs, 7.1% reported serious TEAEs and 7.3% reported TEAEs leading to discontinuation. TEAEs affecting $\geq 10\%$ of subjects were similar to DB period (dizziness and headache).

Conclusion: In this pooled analysis, once-daily ESL 800 mg and 1200 mg was well tolerated in up to one-year of adjunctive therapy in adult patients with partial-onset seizures.

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Abstract – WCN 2013

No: 2223

Topic: 1 – Epilepsy

Drug resistant epilepsy

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Background: The persistence of any kind of epileptic seizures, sufficiently frequent, debilitating in compliant patient for at least two years with antiepileptic therapy (two anti-epileptic drugs), has not been a subject of study.

Methods: The objective was presented in the clinical and paraclinical characteristics of the study. The study was prospective. It took place, from the 1st of October 2008 to the 30th of September 2009, in the Neurology Department.

Results: 36 cases of drug-resistant epilepsy were observed. With a sex ratio of 1.76 of male predominance. The age group most affected was zero to 19 years with 72.33%. Extremes of age were five and 49 years. The most common etiology was infectious, parasitic diseases with 49.22%. In women, malaria pregnancy was the most common etiology. Partial seizures, secondarily generalized, were most frequent with 38.88%. The most refractory association was Phenobarbital and carbamazepine with 30.55%. Limitations of the study were the lack of MRT, PET, and Magneto-Electroencephalography, and dosage of carbamazepine.

Discussion: The most common seizure types in the study were secondarily generalized partial seizures. Results were consistent with those reported by Gueguen et al. who stated that 30% of refractory epilepsies are partial. The most common etiologies were infection and parasitic infestations, which is due to the high incidence of neuroinfectious pathologies in Tropical areas. Medical treatment was instituted for all patients boarded on Gardenal and or carbamazepine not only on the basis of its efficacy but also on its accessibility.

Conclusion: Our results are similar to the studies already made on this subject, which state that 30% of epilepsies are refractory partial epilepsies.

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Abstract – WCN 2013

No: 2292

Topic: 1 – Epilepsy

Effect of metformin on seizures in FeCl₃ induced epileptic animal models

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Epilepsy is a disorder of the Central Nervous System characterized by recurrent seizures unprovoked by an acute systematic or neurologic insult and the sequence of events that turns a normal neuronal network into a hyper excitable network.

The purpose of the study was to investigate whether dietary intake of metformin can inhibit the onset and progression of seizures and their associated pathophysiology in experimental FeCl₃-induced epileptogenesis. Metformin was considered for this study because it is also well known for its antidiabetic, antioxidative, anticancer, and anti-inflammatory properties.

In the present study, seizures were induced by intracortical injection of FeCl₃ into young rats. FeCl₃ injection model was used to induce post-traumatic seizures as this model closely resembles human post-traumatic epilepsy. Synchronized video-EEG monitoring was performed to diagnose manifestation of seizures in young (4 months) and old (18 months) rats. Study was carried on membrane linked enzymes, membrane fluidity, lipofuscin, and antioxidant enzymes to identify the antiepileptic role of metformin using biochemical and histochemical studies.

Treatment with a metformin-supplemented diet significantly inhibited the onset of seizures in rats with iron-induced epilepsy. The seizure-suppressing potential of metformin is explained by the observed biochemical, behavioral, and ultra-structural results.

Our results indicate that metformin significantly prevents generalization of electroclinical seizure activity as well as the pathogenesis associated with iron-induced epileptogenesis.

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Abstract – WCN 2013

No: 2226

Topic: 1 – Epilepsy

Cytogenetic evaluation in epilepsy patients correlated with MTHFR C677T gene mutation and frequency of homocysteine levels

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Epilepsy is one of the most common serious brain disorders. Many chromosomal abnormalities are associated with neurological alterations, among which seizures and epilepsy. The purpose of the study was to determine the chromosomal abnormalities in patients suffering from epilepsy through chromosomal study of metaphases by the use of Trypsin-G-Banding Technique. Furthermore frequency of occurrence of polymorphisms of MTHFR (C677T) gene by PCR-RFLP and to analyze the concentration of homocysteine (Hcy) in epileptics was measured in 18 patients with epilepsy; and compared to 18 healthy controls. We describe some epileptic syndromes frequently reported in chromosomal disorders. We observed chromosome abnormalities such as r (20); r (14); (5p⁻); and (1q⁻) and interestingly one patient had mosaics of trisomy 21. Homocysteine levels showed an association with the MTHFR C677T gene mutation. The DNA damaged cells in patients were higher than in controls ($p < 0.05$). The frequencies of the MTHFR genotype (CC, CT, and TT) among the patients showed 22.22%; 44.44%; and 33.33% whereas among controls 27.8%; 38.8%; and 33.33%. Combining the heterozygous and homozygous MTHFR variant

genotypes (CT + TT) showed a very significant increase in the prevalence of the C677T mutation in patients (77.77%). The prevalence of the CT heterozygous variant genotype alone was significantly higher in patients (44.44%) compared to controls. In conclusion, we suggest the interaction between the gene polymorphism of MTHFR C677T and increased homocysteine levels and the finding of a higher frequency of the CT genotype in Coimbatore population. However further studies are needed to understand the mechanism of epilepsy associated with chromosomal abnormalities.

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Abstract – WCN 2013

No: 2348

Topic: 1 – Epilepsy

Epilepsy in children with HIV/AIDS in Botswana: Prevalence, risk factors, and outcomes

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Background: Seizures are common among patients with HIV/AIDS in the developing world, but there are few published studies that address the prevalence, etiology, or optimal treatment of epilepsy in children with HIV.

Objectives:

- 1) To determine the prevalence of seizures and epilepsy among children with vertically acquired HIV in Botswana;
- 2) To determine risk factors for epilepsy in children with HIV/AIDS;
- 3) To determine whether treatment with anticonvulsants is a risk factor for virologic treatment failure.

Methods: We conducted a retrospective cohort study with a nested case control component to determine the prevalence of epilepsy among children with HIV/AIDS followed at a major referral center in Gaborone, Botswana during the years 2003–2012.

Results: We found a period prevalence of epilepsy of 2.5% in this population with a total of 6.8% of children in the population having had a seizure at some point during the period of the study. Risk factors for epilepsy included CD4 count <200, advanced clinical stage, history of HIV encephalopathy, and history of CNS infections. We found an increased rate of virologic treatment failure of 45% among children with epilepsy treated with enzyme inducing anti-epileptic drugs compared to the background failure rate in the cohort of 18%.

Conclusions: Risk factors for epilepsy emerge in more advanced disease and may be preventable by earlier initiation of antiretroviral treatment. Treatment with enzyme inducing anti-epileptic drugs put children at increased risk of virologic treatment failure, presumably through drug interactions mediated by the CYP-450 system.

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Abstract – WCN 2013

No: 2336

Topic: 1 – Epilepsy

Predictive value of epigastric aura and its evolution into other vegetative signs for temporal lobe epilepsy: A noninvasive video-EEG study

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The initial seizure semiology usually provides valuable information about the seizure onset zone. However, the evolution of auras into other seizure manifestations provides even more localizing information.

We analyzed localization value of epigastric aura (EA) and its evolution into other periictal vegetative signs (PIVS) in patients with temporal (TLE) and extratemporal (ETLE) pharmacoresistant epilepsies who had undergone presurgical evaluation.

We analyzed video-EEG recordings of 170 patients (82 men and 88 women), age ranged 13–66 years (mean 34.67 ± 11.10; Med = 35.00) with medically intractable focal epilepsy. All of the patients underwent a complete presurgical evaluation in the Belgrade Epilepsy Center, comprising long-term noninvasive video-EEG telemetry and neuroimaging findings. The diagnosis of lobar epilepsy was established according to the correlation of clinical seizure semiology, ictal EEG findings and neuroradiology findings. The patient's description of aura and video recordings of seizures were analyzed in order to determine the occurrence of EA and PIVS and its predictive value for the diagnosis of TLE vs. ETLE.

Epigastric aura alone occurred in 55% of patients with TLE vs. 11% with ETLE ($p < 0.001$). On the other hand, evolution of EA into some of PIVS was registered in 28.3% patients with TLE vs. 1.8% with ETLE ($p < 0.001$). The positive predictive value of this finding was 96.77% for TLE, and these patients had 22-fold increased the chance to have TLE rather than ETLE (odds ratio = 21.7).

We found that occurrence of EA and its evolution into PIVS has high positive predictive value for the diagnosis of TLE.

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Abstract – WCN 2013

No: 2290

Topic: 1 – Epilepsy

Myeloradicular schistosomiasis with extensive medullar involvement in the magnetic nuclear resonance: Case report

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Objective: To discuss the importance of findings of the magnetic nuclear resonance in the schistosomiasis diagnostic.

Case report: 2 patients, aged 41 years and 11 years, respectively, both displaying a very intense pain in the thoracic spinal cord, as well as progressive paraparesis. The spinal cord fluid exams showed a positive reaction for schistosomiasis. The magnetic nuclear resonance of the thoracic spinal cord showed an extensive hypersignal area of the T2 and STIR, extending along the C5 and T10 level.

Discussion: The classical abnormalities, seen on the MNR in cases of medullar schistosomiasis, are the increase of spinal medullar cord and/or of the roots of the equine tail in the images, pondered in T 1, hypersignal in T2 sequences and the heterogeneous enhancing during the contrastive phase. In the present case report, as it has been demonstrated in Vital and colleagues' article, published in the journal "Archives of Neuropsychiatry", in 2012, the image of MNR is more extensive than the ones normally described in the literature, related to medullar schistosomiasis. The article concluded that all segments of the spinal cord marrow must be investigated in the cases of neuroschistosomiasis but it stressed the need of further studies in order to confirm these findings.

Conclusion: This case strengthens the hypothesis raised by Vital's article and contributes for more continuity in the studies about the classical diagnostic for schistosomiasis.

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Abstract – WCN 2013

No: 2360

Topic: 1 – Epilepsy

Pregnant women with epilepsy: Is the risk of peripartum psychiatric symptoms increased? – A prospective, population based cohort study

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Background: Peripartum psychiatric disease may harm the mother-child relationship and cause adverse offspring outcome, but is insufficiently investigated in patients with epilepsy.

Objective: Risk estimation of depression, birth- and general anxiety before and after delivery in women with epilepsy compared to women with other chronic disorders and to the general population.

Patients and methods: 101,769 pregnancies from the prospective Norwegian Mother and Child Cohort Study were included. Mothers reported on fear of birth, anxiety and depression in the 3rd trimester and 6 months after delivery using items from validated screening tools. Additional information on maternal health was obtained from the Medical Birth Registry of Norway. Risk of adverse outcomes was estimated as odds ratios (ORs) with adjustment for age, parity, educational level, income, and repeated pregnancies.

Results: Epilepsy was present in 713 cases. In the 3rd trimester 14% reported symptoms of depression 9% anxiety and 13% fear of birth as opposed 9%, 6% and 8% of women from the general population (OR 1.5–1.7 CI 1.2–2.2) and 11%, 8% and 10% of women with other chronic diseases. Six months after delivery, depression was present in 13% and anxiety in 10% of women with epilepsy. In patients on antiepileptic polytherapy 19% reported peripartum depressive symptoms and the OR for postpartum anxiety was 3.3 (1.5–7.5).

Conclusion: Pregnant patients with epilepsy should be screened for depression and anxiety. Adverse drug effects or poor seizure control could cause psychiatric morbidity in patients on polytherapy.

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Abstract – WCN 2013

No: 2426

Topic: 1 – Epilepsy

Bone mineral density in epileptic adolescents treated with lamotrigin and valproate

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Background: Antiepileptic drugs can produce negative influence on bone mineral density in adolescents with epilepsy.

Objective: We evaluated influence of lamotrigin (LTG) and valproate (VPA) on lumbar bone mineral density (BMD L₁-L₄) in adolescents with epilepsy.

Patients and methods: Lumbar bone mineral density Z-score (BMD L₁-L₄ Z-score) was measured in 31 adolescents with epilepsy aged 13–8 years, both genders treated with lamotrigin (n = 15) or valproate (n = 16) monotherapy longer than 1 year. Patient lumbar

spine BMD Z-score values were compared with matched control group values (32 healthy adolescents, both genders). All patients were ambulatory and had similar physical activity and calcium intake. Patients and control group are gender, weight and height matched. For statistical analysis we used software SPSS version 15 (Mann-Whitney U-test and Pearson's correlation). Statistical significance was p < 0.05.

Results: The lumbar spine BMD Z-score values in epileptic patients treated with lamotrigin were not significantly lower compared with control group values (0.69 ± 0.93 vs. 0.96 ± 0.86; p = 0.37; n.s.), as well as in epileptic patients treated with valproate (0.75 ± 0.87 vs. 0.96 ± 0.86; p = 0.56; n.s.) Therapy duration had no negative influence on lumbar BMD in both patient groups (r_{xy} = 0.10; p > 0.05).

Conclusion: Lumbar BMD Z-scores were lower in patient group treated with lamotrigin or valproate compared with control, but not significantly, and they were not dependent on therapy duration.

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Abstract – WCN 2013

No: 1520

Topic: 1 – Epilepsy

Ictal fearful perception of the presence of another in frontal lobe epilepsy – Case report

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Epileptic autoscopic phenomena, including the presence of another, are reported mostly with temporal and parietal cortex involvement. Here we report single patient with frontal lobe epilepsy whose attacks begin with fearful perception of the presence of another.

Epileptic attacks of this patient were recorded during video-EEG monitoring, mainly during sleep, consisting of sudden awakening with paroxysmal fearful perception of the presence of unknown person behind him, accompanied by vegetative arousal, after that flexion-extension of legs and pelvic thrusting with partial clouding of consciousness with diminished responsiveness and amnesia for the period of the attack. Attacks lasted approximately 30 s, repeating in a stereotype way many times during the night. Ictal EEG finding consisted of diffuse beta 18–20 Hz, and postictal diffuse delta and theta 3–4 Hz with the highest amplitude over both frontal regions. Interictal EEG was normal. Neurological exam and brain MRI were normal. Interictal 18 F-DG PET/CT showed hypometabolism in both frontal lobes, more on the left side. Neuropsychological testing showed a slightly decreased verbal memory. Attacks were partially controlled with high doses of Carbamazepine and Levetiracetam.

We presented patient with frontal lobe epilepsy whose attack semiology suggests that frontal cortex may have a role in the autoscopic phenomena.

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Abstract – WCN 2013

No: 2464

Topic: 1 – Epilepsy

Reproductive endocrine complications of antiepileptic therapy at women's epilepsy

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Objective: To study the frequency of reproductive endocrine complications (REC) from different antiepileptic drugs (AEDs) epilepsy in females.

Patients and methods: 155 women at the age of 16–45 years were included in prospective observational uncontrolled comparative study of the AEDs' reproductive side effects into 3 groups: 1gr. – monotherapy AEDs, 2gr. – polytherapy, and 3 compared gr. – no AEDs used. The diagnosis was made based on a comprehensive exam in accordance with ICD-10. REC that lasted for 6 months were taken into account. Naranja algorithm was used to determine reliability of communication "AEDs–REC".

Results: There were 70 patients (45%) in 1 gr, 65 (42%) –2gr, and 20 (13%) –3 gr. 7 AEDs were used in monotherapy, their combinations – in polytherapy. The average duration of AED therapy was 9.3 years, maximum – 33 years. The average quantity of AEDs in the anamnesis was 2.9 in 1 gr., in 2 –4 (max.– 9). The overall incidence of REC was 53%, in 40% due to treatment of AEDs. REC were associated with the taking of AEDs for 21 women (30%) at the 1 gr, 2 gr– 38 patients (59%), 2 people (10%) – the 3 gr. There were no differences in the frequency of REC from different AEDs in the groups.

Conclusion: Methodology of assessment of AED side effects on reproductive health must include an AED therapy anamnesis. Reproductive disturbances are a frequent side effect of AEDs at woman's epilepsy above at polytherapy. It is necessary to monitor a condition of reproductive health during treatment with antiepileptic drugs.

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Abstract – WCN 2013

No: 2147

Topic: 1 – Epilepsy

Status epilepticus in the elderly on a neurological intensive care unit

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Background: Status epilepticus (SE) is a common neurological emergency in the elderly and is associated with high morbidity and mortality.

Objective: We aimed to compare clinical factors and outcome of SE in patients ≥ 60 and < 60 years.

Patients and methods: We analyzed 137 patients with SE admitted to our NICU between 1/2011 and 12/2012 retrospectively, grouped them according to age (≥ 60 and < 60) and compared clinical factors, etiology and outcome between the groups.

Results: Median age was 68 years (range 14–90), 62% were ≥ 60 yrs (85/137), and 38% (52/137) were < 60 yrs. Tonic–clonic SE was the most frequent type in both groups (48% ≥ 60 vs. 58% < 60), comatose NCSE occurred more frequently in older patients (25% ≥ 60 vs. 6% < 60) ($p = 0.466$). SE lasted < 30 min in 36% (49/137), 30–60 min in 26% (35/137) and > 7 d in 4% (5/137) and did not differ between the groups ($p = 0.97$). 52% (71/137) had preexisting seizures (41% ≥ 60 vs. 69% < 60) ($p = 0.15$). An acute symptomatic cause of SE was identified in 26% (36/137) with cerebrovascular diseases being more frequent in the elderly group (43% ≥ 60 vs. 10% < 60) ($p < 0.01$). Mortality was significantly higher in patients ≥ 60 (22% ≥ 60 vs. 6% < 60) ($p = 0.01$), moderate disability in younger patients (17% ≥ 60 vs. 42% < 60) ($p < 0.01$).

Conclusion: In the elderly population SE occurs more often in patients without preexisting epilepsy and is most frequently caused by cerebrovascular diseases. Non-convulsive SE with coma occurs

more frequently in patients ≥ 60 . SE in older patients is associated with poorer outcome and higher mortality.

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Abstract – WCN 2013

No: 2488

Topic: 1 – Epilepsy

Evaluation of cognitive deficit in patients with epilepsy measured with MMSE (mini mental state examination)

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Background: Cognitive deficit represents damage of intellectual abilities that manifest themselves in the form of disorientation in time and space, impaired attention and memory, inability to recognize, judgment and reasoning, and impairment of complex intellectual abilities such as analyzing and using information.

Objective: To determine the prevalence and major parameters of cognitive deficits in patients with epilepsy, depending on the type of anti epileptic therapy compared to the control group.

Patients and methods: Study enrolled 100 subjects of both sexes, the age structure of 18–65 years old who were divided into two groups: group with 60 patients suffering from epilepsy and a control group of 40 healthy subjects.

Results: Of the total number of patients with epilepsy, 55% had cognitive deficits, where the deficit of attention and concentration was present in about 57% treated with valproate, 53% treated with carbamazepine and 42% treated with phenobarbital. Phenobarbital affected the understanding of spoken words by 16.6%, and the difficulties in tracing the figures were most pronounced in patients taking carbamazepine (76.47%) and lamotrigine (47%).

Conclusion: The results showed significant incidence of cognitive deficits in patients with epilepsy compared to the control group. Patients with epilepsy had the most damaged domains of attention and concentration on the MMSE. Phenobarbital, carbamazepine and valproate are anticonvulsants that have led to the dominance of attention and concentration, while patients on carbamazepine and lamotrigine had major difficulties in tracing figures.

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Abstract – WCN 2013

No: 2505

Topic: 1 – Epilepsy

The situation of the EEG service in a developing country/Egypt

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Background: EEG is the most important investigation in all types of epilepsies; provided that it was properly performed and carefully interpreted in the context of a well-described clinical setting.

Objectives: This work aims at highlighting the situation of the EEG service in a developing country.

Methods: All consecutive EEG reports from 1232 patient done over a year at 3 locations were retrospectively reviewed. The following data were evaluated; EEG findings, clinical indications, requesting physician specialty, age of the patient, duration of the record, state of the patient whether awake or asleep, and the cost of the procedure.

Results: EEG in a developing country, besides being ordered mainly by neurologists and neurosurgeons, was noticed to be ordered by other

specialties in a significant percentage. Striking enough overt continuous seizures/movements and witnessed seizure without return to baseline were not the main indication for ordering an EEG from all physicians including neurologists and neurosurgeons. The majority of the EEGs undergone for children at the school age or below were done during induced sleep using chloral hydrate. Routine short term EEG remains the most common type of EEG ordered in Egypt compared to long term video/sleep EEG. The concept of the continuous EEG monitoring for an ictal recording is having many limitations in a developing country; some of them are financial and others are related to the professional awareness about its usefulness.

Conclusion: More awareness is required as regards the indications and value of all types of EEG in a developing country.

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Abstract – WCN 2013

No: 2493

Topic: 1 – Epilepsy

Nonconvulsive status epilepticus proper: A study of 220 cases

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Background: NCSE may occur de novo in critically ill patients and in epilepsy patients (NCSE-proper). Little is known about the frequencies of different types of NCSE in an epilepsy population.

Methods: We analyzed clinical and EEG data of 220 cases with NCSE in our epilepsy outpatient clinic (n = 5376) from 1970 to 2003.

Results: The incidence of NCSE was 3.6%. A striking preponderance of female patients (140 (63.6%) versus 80 (36.4%)). Continuous generalized epileptiform EEG pattern was observed in 46%, focal patterns in 40.4% and 13.7% had diffuse EEG changes. These patterns were of definite significance in 49.6%, of probable in 36.8%, and of possible in 13.7%. NCSE occurred as late complication of epilepsies with a mean time delay of 16 years after seizure onset. Acute symptomatic NCSE was rare (7.3%). In 55.9% of patients NCSE was a recurrent phenomenon. Psychic abnormalities were more frequently observed in the study group (p < 0.001). Lennox–Gastaut syndrome is more (p < 0.001) and idiopathic generalized epilepsies are less (p < 0.024) inclined to develop NCSE. In 44.6% NCSE occurred in close vicinity to self-limiting convulsive episodes. NCSE was typical (25.9%) and atypical absence status (22.3%), as focal simple (26.4%) and as complex focal status epilepticus (19.1%). In 6.3% a classification was not possible on the basis of electro-clinical symptoms.

Conclusions: Diagnosis of NCSE in patients with epilepsy is rare and occurs as a late complication often recurrent. EEG is crucial for its diagnosis.

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Abstract – WCN 2013

No: 1743

Topic: 1 – Epilepsy

Intracranial metallic body artifact in routine scalp EEG, a case series

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Background: EEG artifacts are considered a disturbance in a measured brain signal and can be divided into physiological and extra physiological. Routine scalp EEG with an impeded intracranial

metallic bullet represents a fusion of both types in the same instance and is expected to interfere with the signals.

Objectives: Searching online for literature on rare EEG artifacts with an emphasis on intracranial metallic body artifacts revealed to be very anecdotic. The aim of this report was to create awareness about what to expect during the EEG recording in this situation.

Patient and methods: Two patients were presented; one for stat and the other for routine EEG. The former was a 19 year old male with a deeply imbedded intracranial bullet presented with disturbed conscious level. The latter was a 26 year old male with an imbedded bullet in the right hemisphere presented with focal seizures with secondary generalization.

Results: Standard EEG recording was done for both cases; other than an initial generalized interference pattern that disappears with meticulous adjustment of the impedance, the presence of intracranial metallic foreign didn't interfere significantly. EEG record in this setting was very sensitive to any minor changes in the impedance.

Conclusion: Routine scalp EEG with an intracranial metallic foreign body was expected to show a difficulty in EEG interpretation, yet a good quality EEG signal can be achieved by a meticulous concern upon the impedance. Further work is required to identify if the material or the location of the metallic foreign body might have an influence.

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Abstract – WCN 2013

No: 2486

Topic: 1 – Epilepsy

Antiepileptic drug levels in patients admitted for epilepsy surgery evaluation

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Background: Patients admitted for epilepsy surgery evaluation are considered pharmacoresistant. However, due to missed drug dosages or pharmacological interference, blood levels of AEDs may be not be in the recommended range, possibly contributing to higher seizure frequency.

Objective: Determine the frequency of subtherapeutic AED levels in patients admitted for epilepsy surgery evaluation.

Patients and methods: We reviewed retrospectively our database between 1.1.2007 and 31.12.2012 and identified 196 patients, who were admitted for in-depth evaluation of their pharmacoresistant epilepsy (85 children < 16 years, 111 adults). Drug levels were tested on admission under full therapy in all patients, albeit not of each drug. A total of 283/468 (60%) antiepileptic drugs in this patient group was measured. 26 patients were on monotherapy, 80 on bi-therapy, and 90 patients on 3 AEDs or more.

Results: In the whole patient group, 58 patients (30%) presented at least one AED in a subtherapeutic level. 49 patients had 1, and 9 patients had 2 insufficiently dosed AEDs. Overall, children had more often subtherapeutic levels than adults (60% vs 23%, p = 0.02). The most frequently decreased AED was Valproate, followed by Topiramate and Lamotrigine (p < 0.001). Supratherapeutic levels were rare and noted in only 11 patients.

Conclusion: Subtherapeutic drug levels were found frequently in pharmacoresistant patients, albeit no patient had insufficient levels in his entire drug regimen. Children are significantly more affected,

probably due to infrequent blood monitoring. While a decrease of AED levels does not question the diagnosis of pharmacoresistance, it may contribute to inefficiency of therapy.

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Abstract — WCN 2013

No: 2502

Topic: 1 — Epilepsy

Epilepsy and electroencephalographic features in trisomy 13: A case report and review of literature

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Background: Trisomy 13 (T13) is associated with considerable phenotypic variability but there have been no detailed descriptions of the characteristics of T13-related epilepsy.

Objective: The aim of this study is to describe seizures and EEG features in a patient with T13.

Material and methods: Clinical analysis included phenotypic examination, brain MRI, and EEG studies, and karyotype study.

Results: She was a 2 year old girl. She showed an extremely delayed psychomotor development. Epilepsy takes place at 6 months; seizures were characterized by staring, a respiratory arrest and symmetric hypertonia of four limbs. Neurological examination at the age of 1 year, revealed a global hypotonia and dysmorphic features: trigonocephaly, microphthalmia, tall forehead, bulbous nose, and high arched palate. Interictal EEG shows right temporal spikes. MRI showed mild anoxo-ischemia lesion. Karyotype studies confirmed free trisomy 13 known as Patau's syndrome. A treatment including valproic acid was started. At the age of 2 years, she present with generalized spasms. Sleep EEG showed hypsarythmia. A treatment by vigabatrin was successful.

Conclusion: There was only isolated case reports described with epilepsy on chromosome 13. The relatively low incidence of epilepsy in patients with trisomy13 is probably explained by the poor survival rates of this aberration. Epilepsy is generally focal and began in the neonatal period or early infancy like our patient. But to our knowledge this is the first report of a late onset West syndrome in T13. T13 probably caused epilepsy due to a dosage effect resulting from the addition of normal genetic sequences.

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Abstract — WCN 2013

No: 2500

Topic: 1 — Epilepsy

Negative myoclonus of epileptic origin in association with bilateral subdural hematoma

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Background: Negative myoclonus (NM) is a motor phenomenon characterized by a jerky involuntary movement caused by a brief and sudden interruption of tonic muscle activity. It frequently occurs in association with metabolic encephalopathies.

NM of epileptic nature (ENM) is defined as a brief interruption of tonic muscle activity, time-locked to a spike or epileptic transient on the EEG, without evidence of an antecedent positive myoclonus.

Objective: Case report and review of the literature.

Results: 58 year-old man with history of intracranial hypertension submitted to ventriculoperitoneal shunting, was brought to the emergency room with somnolence and involuntary movements of the upper limbs. At examination the patient was drowsy, aphasic, with a right hemiparesis and right Babinski sign. He presented continuous distal bilateral myoclonic jerks, characterized by a sudden and brief drop of the upper limb, compatible with NM. Brain CT scan revealed 2 subdural hematomas — right fronto-temporal and left high convexity. Angiography confirmed the presence of several dural fistulae. The EEG detected signs of right hemispheric dysfunction, frontal and frontotemporal slow wave activity. The EMG recording confirmed the presence of atonia in relation to the myoclonus. At back-average analysis, these movements were preceded by a frontal and anterior temporal, bilateral, slow wave, suggestive of a cortical origin.

Conclusion: We present an uncommon case of ENM in association with a bilateral subdural hematoma, underlying the importance of distinguishing this type of NM and of a neurophysiological assessment in cases that raise diagnostic concerns.

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Abstract — WCN 2013

No: 2554

Topic: 1 — Epilepsy

“First seizure clinic”: The impact on patient care and adherence — A prospective study

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Background: Patients admitted after their first seizure(s) receive usually a neurological consultation, CT and often EEG before being discharged to their general practitioner (GP). However, in many cases, patients are lost to follow-up with significant consequences of further seizures.

Objective: Yield of comprehensive in-house management (IHM) after a first epileptic seizure compared to “classical” management.

Patients and methods: 167 patients after first presumably non-provoked seizures were evaluated prospectively. In the interventional group (IG, N = 97), patients received initial and follow-up epileptology consultation, standard EEG, 3 T MRI, and if necessary long-term EEG. The control group (CG; N = 70) received established standard care before being referred to a private neurologist or GP.

Results: In the IG, 98% had an EEG, 86% a CT-scan, and 71% an MRI. 85% were seen again for follow-up in our epileptology clinic. In the CG, these values were quite similar except that only 53% underwent MRI ($p = 0.01$) and 51% were seen by a neurologist for follow-up ($p < 0.001$). A diagnosis of epilepsy was obtained more often in the IG (50 vs 25 patients; $p = 0.04$) and time delay to consultation was shorter, 19 vs 50 days, respectively.

Conclusion: Initial in-house management of patients with a first epileptic seizure shortens the diagnostic delay and increases the number of follow-up consultations by 34%. In parallel, it raises the number of diagnosed patients with epilepsy by 15%. 3 T-MRI provided added value in 17% of patients with normal CT-scan. Our results suggest that patients benefit significantly from comprehensive IHM.

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Abstract – WCN 2013**No: 2549****Topic: 1 – Epilepsy****Surgical palliative care in Lennox–Gastaut syndrome**E. Hentati, N. Ben Ali, M. Kchaou, S. Belal. *Service de Neurologie, Hopital Charles Nicolle, Tunis, Tunisia*

Aims: To retrospectively review the efficacy of vagus nerve stimulation in three pediatric patients with Lennox–Gastaut syndrome and examine the seizure-frequency, intensity and duration outcomes.

Methods: It is a retrospective study of 3 patients followed in the Department of Neurology of the Charles Nicolle Hospital to which had been implanted a vagus nerve stimulation. The standard initial parameters were: A stimulation for 30 s followed by a stimulation-free period of 5 min.

Results: The population included 3 children: two girls and a boy. The mean age was 9.33 years and the mean age at implantation was 8 years. The mean of the follow-up duration was 18 months. The median seizure-frequency reduction was 23%. We have noticed a reduction of duration of seizures. The Status epilepticus disappeared in one patient.

Conclusion: The efficiency of vagus nerve stimulation was partial. Indeed, the number and the duration of the seizures decreased but any of our patients presented a complete cure. However the efficiency of this technique would be superior to the isolated medicinal treatment.

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Abstract – WCN 2013**No: 2558****Topic: 1 – Epilepsy****Adverse birth outcomes after prenatal exposure to antiepileptic drugs**D. Kilić^a, H. Pedersen^b, M.I.S. Kjaersgaard^c, E.T. Parner^c, M. Vestergaard^d, M.J. Sørensen^e, J. Olsen^f. ^aDepartment of Neurology, Aarhus University Hospital, Denmark; ^bResearch Unit for General Practice, Denmark; ^cDepartment of Public Health, Section for Biostatistics, Aarhus University, Denmark; ^dDepartment of Public Health, Section for General Medical Practice, Aarhus University, Denmark; ^eRegional Center for Child and Adolescent Psychiatry, Aarhus University Hospital, Risskov, Denmark; ^fDepartment of Public Health, Section for Epidemiology, Aarhus University, Aarhus, Denmark

Background: There is limited knowledge of the effects of prenatal exposure to antiepileptic drugs and birth outcome.

Objective: To study birth outcome in newborn children after prenatal exposure to antiepileptic drugs.

Patients and methods: From Danish registers, we identified all children born from 1997 to 2008 and linked this with information on the mother's prescriptions for antiepileptic drugs during pregnancy. We used linear regression to study birth weight, gestational age and head circumference at birth, and binominal regression to study preterm birth (<37 weeks) and "small for gestational age" (<10%). Estimates were adjusted for potential confounding factors. Furthermore, head circumference and birth weight were adjusted for gestational age, and gestational age was adjusted for birth weight.

Results: We identified 679,762 newborn singletons. After adjustment for confounders, antiepileptic drug exposure (n = 2928) was associated with reduced gestational age; –0.92 days (95% confidence interval (CI): –1.40 to –0.44), lower birth weight; –31.96 g (95% CI: –51.74 to –12.18) and smaller head circumference –0.07 cm (95% CI: –0.14 to –0.004) compared to non-exposed. There was a higher risk of being born preterm (<37 weeks) (adjusted Risk Ratio (aRR): 1.51 (95% CI: 1.32–1.72)) and a higher risk of being small for gestational age (aRR: 1.21 (95% CI: 1.10–1.34)).

Conclusion: Prenatal exposure to antiepileptic drugs was associated with lower birth weight, reduced gestational age, decrease in head circumference, and increased risk of preterm birth and being small for gestational age.

doi:10.1016/j.jns.2013.07.179

Abstract – WCN 2013**No: 2495****Topic: 1 – Epilepsy****Efficacy of chronic stimulation of nucleus accumbens (NAC) and anterior thalamus (ANT) in patients with pharmacoresistant focal epilepsy**F.C. Schmitt^a, J. Voges^b, L. Buentjen^b, T. Zaehle^a, K. Bohlmann^c, H. Stefan^d, F. Oltmanns^e, H.-J. Heinze^a, M. Holtkamp^{e,f}, A. Kowski^f. ^aDepartment of Neurology, University of Magdeburg, Medical Faculty, Magdeburg, Germany; ^bDepartment of Stereotactic Neurosurgery, University of Magdeburg, Medical Faculty, Magdeburg, Germany; ^cEpilepsy Hospital Tabor, Epilepsy Center Berlin-Brandenburg, Bernau, Germany; ^dDepartment of Neurology, University of Erlangen-Nuremberg, Erlangen, Germany; ^eKoenigin-Elisabeth-Herzberge Hospital, Epilepsy Center Berlin-Brandenburg, Germany; ^fDepartment of Neurology, Charité University Medicine, Berlin, Germany

Background: The SANTE study proved efficacy of ANT stimulation in pharmacoresistant epilepsy. The nucleus accumbens has a relay function for the frontal lobe and for the Papez circuit. It therefore could also be an effective target to suppress ictal activity in these areas.

Objective: This observational study summarizes clinical outcome of DBS in the NAC and compares results to subsequent ANT stimulation.

Patients and methods: Four patients had stimulation of the NAC and subsequently of the ANT over 6 months. Stimulation parameters and stimulated contacts for each target, as well as concomitant AEDs remained unchanged. Clinical parameters such as seizure frequency, the Liverpool-Seizure-Severity-Scale (LSSS) and Beck's depression inventory were assessed. Due to pronounced interindividual heterogeneity in baseline period, clinical variables were related to the individual baseline values after each stimulation period and expressed as fraction of 1.

Results: Compared to the baseline period the LSSS score significantly reduced to 0.81 (+/–0.11; p = 0.014) and the relative number of disabling seizures (i.e. complex-partial plus generalized seizures) to 0.66 (+/–0.34; p = 0.091) in the four patients studied. In one subject, reduction of disabling seizures of more than 50% (21%) was observed. The number of non-disabling seizures and other clinical variables remained unchanged. Results of ANT stimulation are in progress and will be presented at the conference.

Conclusion: Six months of NAC stimulation significantly reduced the seizure severity score. A larger cohort may show a significant reduction in seizure frequency and reveal specific responders for ANT and NAC stimulation.

doi:10.1016/j.jns.2013.07.180

Abstract – WCN 2013**No: 2151****Topic: 1 – Epilepsy****Lack of potassium current in novel mutations of KCNQ2 and KCNQ3 identified in benign familial neonatal epilepsy (BFNE)**Y. Sugiura^a, Y. Ihara^b, A. Ishii^b, Y. Ugawa^a, S. Hirose^b. ^aDepartment of Neurology, Fukushima Medical University School of Medicine, Fukushima, Japan; ^bDepartment of Pediatrics, Fukuoka University School of Medicine, Fukuoka, Japan

Background: Benign familial neonatal epilepsy (BFNE), a form of autosomal dominant epilepsy of infancy, is caused by gene mutations in the *KCNQ2* or *KCNQ3* which code the voltage dependent potassium channels $K_v7.2$ or $K_v7.3$ expressing in neurons.

Objective: We analyzed *KCNQ2* and *KCNQ3* gene in two Japanese families with BFNE, and studied the mutant *KCNQ2/KCNQ3* potassium channel property to elucidate the mechanism of BFNE.

Patients and methods: We investigated *KCNQ2* and *KCNQ3* gene sequence of two families with BFNE. Whole-cell potassium currents were recorded from HEK293 cells transfected with the plasmid vector encoding either wild type or mutant *KCNQ2/KCNQ3* gene. Current–voltage relationships were determined by using test pulses between -120 mV and 40 mV from the holding potential of -100 mV.

Results: The sequence analysis of *KCNQ2* and *KCNQ3* gene identified *R581X* mutation in *KCNQ2* and *S337Y* mutation in *KCNQ3*. Wild type *KCNQ2* and *KCNQ3* channels showed normal potassium currents, however, these mutant channels showed no significant potassium currents.

Conclusion: We identified the novel mutations *R581X* of *KCNQ2* and *S337Y* of *KCNQ3* gene in Japanese families with BFNE. These mutations cause a deficiency of the potassium conductance and impair the repolarization of neurons. Thus it may cause abnormal prolongation of the neuronal excitation, which finally induces epilepsy.

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Abstract – WCN 2013

No: 2588

Topic: 1 – Epilepsy

Robotics in neurosurgical stereotactic interventions: Oblique intrainsular electrodes implanted in patients with epilepsy

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Objective: This study is to investigate: The feasibility, the safety and the utility of chronic depth electrodes stereotactically implanted by a robotic arm in the insular cortex of patients suffering from drug refractory focal epilepsy.

Methods: A total number of 32 electrodes in 29 patients were successfully implanted within the insula. 220 contacts were available for insula recording. Electrode insertion was guided by a robotic arm (*NeuroMate, Renishaw Mayfield, Switzerland*) connected to the stereotactic frame and driven by stereotactic planning software. The targeting of the insula is planned on a pre-surgical T1-MRI. The fusion between the preoperative 3D MRI and the postoperative 3D CT scan enabled us to identify the contact location in 3D.

Results: No morbidity occurred during the surgical step and the chronic SEEG recording or stimulation procedure. Clinical responses have been identified in terms of gyral and sulcal anatomy. They were classified into: painful responses, sensorimotor responses, speech disturbance, oropharyngeal responses, auditory phenomena and neuro-vegetative phenomena.

Conclusion: The advantages of the oblique approach are:

1. The implantation of electrodes within the insula using robotic arm appears in our study to be safe.
2. This approach can explore all insular regions by avoidance of the sylvian vascular network.
3. This approach offers a better sampling of insular EEG activity (until 10 contacts/electrode) than that obtained by the classical lateral trans-opercular approach (1 1/2 contacts/electrode).

4. This approach has allowed us to develop the first anatomic-functional organization scheme of the insular cortex according to its gyri and sulci.

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Abstract – WCN 2013

No: 1159

Topic: 1 – Epilepsy

Absence seizures arising from a mutation that causes selective loss of AMPA receptors within thalamocortical networks

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Absence seizures are characterized by spike-wave discharges (SWD), which arise from disturbances within thalamocortical (TC) networks leading to the generation of hypersynchronous intrathalamic oscillatory activity. However, the specific underlying cellular and molecular mechanisms by which absence seizures are produced within TC networks are still unclear and likely to be multifactorial. Several rodent absence epilepsy models have been used to investigate the cellular mechanisms underlying the generation of SWD. In this study we use the well-established stargazer mouse model of absence epilepsy, which carries a mutation in the gene for stargazin, an AMPA-receptor trafficking protein. Previously, we demonstrated selective loss of AMPARs at corticothalamic synapses in the reticular thalamic nucleus (RTN) but not at corticothalamic synapses in the ventral posterior thalamic region in epileptic stargazers. To further investigate synapse-specific changes in AMPAR expression in RTN, we used immunogold cytochemistry to analyse the relative density of AMPAR subunits at thalamocortical-RTN synapses in stargazers compared to non-epileptic control littermates ($n = 5$ pairs, 500 synapses). While AMPAR GluA2/3 and GluA4 subunits were significantly decreased (35% and 46% respectively; $p < 0.01$) at stargazer RTN corticothalamic synapses, no significant differences in AMPAR subunit expression occurred at RTN thalamocortical synapses. The synapse-specific loss of AMPARs at corticothalamic but not thalamocortical synapses in the RTN of stargazers is consistent with other studies showing specific loss of GluA4 at cortico-RTN synapses in the *Gria4*^{-/-} model of absence epilepsy. Further studies are needed to determine the causal link between synapse-specific loss of AMPARs in RTN and the generation of absence epilepsy.

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Abstract – WCN 2013

No: 2643

Topic: 1 – Epilepsy

Nodding syndrome; a new (infectious?) disease entity of the CNS in Eastern Africa

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Nodding syndrome is a poorly understood epidemic neurologic disorder affecting thousands of children and adolescents in sub-Saharan Africa. To date the aetiology remains unknown. The objective of this paper is to provide an overview of studies of the syndrome and in particular to describe the epidemiology, risk factors, clinical features, treatment outcomes and pathogenesis. The paper will examine the relationship with infectious agents and interventions to address these

and provide an update into the investigation of the syndrome, gaps in knowledge and directions of ongoing studies. It will also identify some research questions and possibilities for investigation.

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Abstract – WCN 2013

No: 282

Topic: 1 – Epilepsy

Lesionectomy of the contralateral epileptogenic focus may help epilepsy control

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Background: Epilepsy is a common exhaustive and strenuous chronic neurological disorder. Epilepsy is usually controlled, but not cured, with medications. However, over 30% of people with epilepsy do not have seizure control with the best medications. Epilepsy surgery is an option for people with focal seizures that remain resistant to treatment.

Patient: We report an eight year old girl who had a road accident followed by right frontal encephalomalacia and resistant seizures. The EEGs confirmed contralateral seizures. She had been resistant to all AEDs. VNS did not make a difference. There was no full agreement that she would benefit from a lesionectomy on the other side of the seizure discharges. After the operation and for 18 months, the girl's seizures dropped from 3–6 daily to only 3 brief seizures for the whole post-operative period.

Results: Her seizures have been described as two different types, starting while awake with abnormal eye movements to many different directions and screaming with subsequent forward tonic movements of upper limbs. **Type 2** while awake, her eyes move upwards and to the right. She would be unresponsive. Interictal EEG revealed sharp wave activities on the left frontocentral and centroparietal regions with phase reversals at C3.

Ictal EEG showed evidence of epileptiform activities starting from the frontocentral and centroparietal regions with fast recruitment of the right hemisphere. MRI revealed right frontal lobe cystic encephalomalacia with gliosis.

Conclusion: Children with intractable epilepsy and focal brain lesions are candidates for surgery even when they have generalized or contralateral epileptiform discharges.

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Abstract – WCN 2013

No: 2651

Topic: 1 – Epilepsy

Immunoabsorption and cyclophosphamide in a relapse of an anti-NMDA-receptor-encephalitis

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An 18 year old woman was transferred to our neurologic intensive care unit with clinical signs of a relapse of a non-teratoma-associated anti-NMDA-receptor-encephalitis. The disease was diagnosed 4 months before and treated with plasmapheresis (PP), IVIG and prednisone. On admission she needed intubation due to psychotic behaviour and repetitive generalized tonic-clonic seizures.

We started daily PP for 5 days, then switched to immunoabsorption (IA) every other day for 2 weeks. Additionally, we gave cyclophosphamide 250 mg/m² weekly for 3 times and prednisone 50 mg daily. We found positive anti-NMDA-receptor-antibodies before and negative

antibodies after IA as well as PP. Two weeks after admission the patient was extubated but needed reintubation the other day because of status epilepticus and psychosis.

We decided to continue IA every 4th day for 3 weeks, followed by weekly IA for 2 months and bi-weekly IA since then. With modified anticonvulsive therapy and low dose antipsychotic therapy the patient could successfully be extubated four weeks after admission. Four months after admission, the patient is steadily recovering with an actual modified Rankin Score of 2, seizure-free under reduced anticonvulsive therapy with phenytoin and valproate and in a stable psychiatric condition. Prednisone could be reduced considerably.

Discussion: There are few suggestions for therapy in non-teratoma-associated anti-NMDA-receptor-encephalitis. To our knowledge only one recent paper reports the use of IA in this disease. Our report shows that a relapse can successfully be treated with cyclophosphamide, steroids and repeated IA. Further studies are needed to determine the best treatment.

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Abstract – WCN 2013

No: 2670

Topic: 1 – Epilepsy

Ca²⁺ channel-blockers counter with the damaging effect of repetitive spreading depression on memory

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Background and purpose: Spreading depression known by transient loss of spontaneous and evoked neuronal activity and changes in ionic, metabolic and hemodynamic characteristics of the brain. Many studies have focused on the role of Ca²⁺ channels in spreading depression, however this role is not completely clear yet. On the other hand it has proven that impairment of memory is one of the main effects of spreading depression. In our study we aimed at determining the role of Ca²⁺ channel-blockers on repetitive spreading depression in terms of its effect on memory.

Methods: Wistar rat (60–80 g) randomly chosen in 4 groups and Nifedipine 1 mg/kg were administered weekly after 3 mol/L KCl injection for induction of repetitive SD in rat for 4 week. The groups were evaluated by T-maze memory test and the SD group was compared with control groups.

Result: T-maze test data demonstrated that in repetitive spreading depression group memory was impaired during the weeks. In the group which Nifedipine has been administered, memory improvement has been significantly observed.

Conclusion: Our study showed that administration of Nifedipine as a Ca²⁺ channel-blocker could significantly reduce the level of memory impairments, which is naturally followed by repetitive spreading depression.

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Abstract – WCN 2013

No: 2683

Topic: 1 – Epilepsy

Epilepsy: A historical overview

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Background: Seizures have always been one of human afflictions and few diseases have attracted so much attention and generated so much controversy as epilepsy. Wrapped since ancient times by religious, magical and scientific stigmas, about 400 BC, Hippocrates recognized epilepsy as a brain disorder, and despite this, only in recent decades significant efforts have been developed against centuries of ignorance and discrimination.

Objective: To present an overview of the evolution of knowledge and of facts about epilepsy elapsing almost 2300 years from Hippocrates to middles of the XIX century.

Material and methods: Narrative review based on epilepsy historical facts by searching books and electronic databases.

Results: The word epilepsy was first used by Avicenna in the XI century, and meant “be taken, attacked or overwhelmed.” In ancient Rome, epileptics were avoided for fear of contagion, while in the Middle Ages, were persecuted as witches. During the Renaissance, epilepsy was seen as a manifestation of physical illness. However, it was in the Enlightenment, in the century of scientific revolution (XVII), that epilepsy started to be understood according to anatomy, pathology, physiology and chemistry.

Conclusion: During those 2300 years, the hypothesis of the supernatural with designations of “sacred disease” and “lunatics” has been abandoned and new modern theories came out. The knowledge about the history of epilepsy highlights the need for greater efforts for humanity to start a new millennium with this disease free from the shadows of the past.

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Abstract — WCN 2013

No: 2684

Topic: 1 — Epilepsy

Epidemiological aspects of epilepsy in Brazil from 2008 to 2012

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Background: Epilepsy is the commonest neurological condition affecting people of all ages, race and social class. There are about 50 million people with epilepsy in the world, 40 million in developing countries with little or no access to treatment. There are few studies on the epidemiology of epilepsy in Brazil, despite being an important public health problem.

Objective: To describe the profile of patients hospitalized due to epilepsy in Brazil, according to sex and age, in the period from 2008 to 2012, comparing to literature.

Material and methods: Data from hospital admissions in Brazil, in the period from 2008 to 2012, registered by Brazilian Health System based on database (DATASUS).

Results: The estimated incidence of epilepsy in Brazil was 190 per 100.000/year. The total number of hospitalization was 47,046, of which 27,468 (58.4%) in males and 19,578 (41.6%) in women. Of the total admissions, 12,431 (26.4%) occurred from 1 to 9 years and 10,416 (22.1%) from 30 to 49 years. The lowest rate was found in 80 years and more, which was responsible for 1333 (2.8%) hospitalizations.

Conclusion: The incidence of epilepsy in the world varies from 40–70 per 100.000/year in developed countries to 100–190 per 100,000/year in developing countries, as Brazil. It is more common in males than females but this difference is rarely significant. The highest incidence is in early childhood, with a decrease in adolescence, in both scenarios. In Brazil, the incidence is lower among elderly,

different from the world, where the highest incidence is above 80 years.

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Abstract — WCN 2013

No: 2681

Topic: 1 — Epilepsy

Clinical & electroencephalographic and cognitive function correlation in epileptic children

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Cognitive dysfunction is one of the major contributors to the burden of epilepsy. It can significantly disrupt intellectual development in children and functional status and quality of life in adults. In children, cognitive problems are more diffuse responsible for learning difficulties and behavior problems.

The aim of study was to assess to what extent cognitive function is affected by clinical features and EEG characteristic of the epilepsy in children whether newly diagnosed or on antiepileptic drugs.

Subject and method: The present study is a cross section case control study. It was carried out on 95 children selected from pediatric-neurology outpatient clinic and inpatient department of El-Zahraa University hospital and El-Hussein University hospital during the period from November 2010 to July 2012. All studied children were subjected to full history taking with special emphasis on seizure description. Laboratory investigation: including CBC, ESR, liver & kidney functions and serum electrolytes as well as serum antiepileptic drug level whenever necessary. Neuroimaging was done to exclude organic brain lesion. Assessment of cognitive functions was assessed using the Arabic version of the Stanford–Binet intelligence Test (4th edition) which assesses four main cognitive abilities (Verbal reasoning, Visual/Abstract reasoning, Short term memory, Quantitative reasoning).

We came to a conclusion that epileptic children have significant cognitive impairment even if their seizures were controlled. Epileptic children with normal composite IQ could have Verbal, Visual/Abstract, Quantitative or memory impairment. Frequency and generalized seizure, duration of epilepsy, AEDs intake and poor seizure control are important factors for poor cognitive function.

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Abstract — WCN 2013

No: 2697

Topic: 1 — Epilepsy

Epilepsy surgery in Morocco study and long-term follow-up in 51 cases

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51 patients with intractable lesional temporal partial epilepsy were operated between 2005 and 2011. 48 patients had an anterior temporal lobectomy and 3 lesionectomy. In 29 patients (57%) hippocampal sclerosis was found, dysembryoplastic neuroepithelioma in 13 (25%), 4 patients with cortical dysplasia, 2 with ganglioma, and one case of cavernoma and astrocytoma.

Presurgical evaluation was the same in all cases, based on non-invasive methods (detailed history, neurological and psychiatric evaluation, interictal EEG, MRI, video-EEG with ictal EEG and neuropsychological tests).

After 7 years of follow-up, 40 patients (78.4%) are in class I of Engel's classification, among them 31 (60.7%) are seizure free (class IA), 5 patients in class II, 6 patients in class III, and no patients in class IV. The outcome is almost identical to the group with hippocampal sclerosis and the other group with other types of lesions for the first four years, after seven years of follow-up, 50% of the first group are in class I and 66.7% for the second group. Our experience demonstrates the feasibility of epilepsy surgery in developing countries particularly in Africa and appropriateness of non-invasive method evaluation.

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Abstract – WCN 2013

No: 2711

Topic: 1 – Epilepsy

α 7-Containing nicotinic acetylcholine receptors on interneurons of the basolateral amygdala and their role in the regulation of the network excitability

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The basolateral amygdala (BLA) plays a key role in fear-related learning and memory, in the modulation of cognitive functions, and in the overall regulation of emotional behavior. Pathophysiological alterations involving hyperexcitability in this brain region underlie anxiety and other emotional disorders, as well as some forms of epilepsy. GABAergic interneurons exert a tight inhibitory control over the BLA network; understanding the mechanisms that regulate their activity is necessary for understanding physiological and disordered BLA functions. The BLA receives dense cholinergic input from the basal forebrain, affecting both normal functions and dysfunctions of the amygdala, but the mechanisms involved in the cholinergic regulation of inhibitory activity in the BLA are unclear. Here, we demonstrate that the α 7-containing nicotinic acetylcholine receptors (α 7-nAChRs) are present on somatic or somatodendritic regions of interneurons, in the rat BLA. These receptors are active in the basal state enhancing GABAergic inhibition, and their further – exogenous – activation dramatically increases spontaneous inhibitory postsynaptic currents (sIPSCs) in principal BLA neurons. When α 7-nAChRs are activated in all of the BLA network, both inhibitory and excitatory currents increase in BLA principal cells, but the inhibitory currents are enhanced significantly more than the excitatory currents; the result is reduced excitability, as suggested by the decrease in the amplitude of evoked field potentials. The anxiolytic effects of nicotine, as well as the role of the α 7-nAChRs in seizure activity involving the amygdala and in mental illnesses such as schizophrenia and Alzheimer's disease may be better understood in light of the present findings.

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Abstract – WCN 2013

No: 2536

Topic: 1 – Epilepsy

Postictal sneezing: A case study

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Objective: To describe a patient with epilepsy who had postictal sneezing during long-term video-encephalography (EEG) monitoring. In the literature, sneezing is reported to be an autonomic phenomenon

as an aura in epilepsy. One case of postictal sneezing was reported previously.

Method: The patient underwent video-EEG monitoring for probable focal onset epilepsy. Three episodes of postictal sneezing were recorded and studied clinically and electrographically.

Results: Three seizures were recorded which were followed by sneezing twice when he became responsive. For the first seizure, the patient was off-camera and clinical features could not be determined except for sneezing. The two visible seizures were characterized by oral automatisms, staring, head posturing to the right, unresponsiveness, and in one of the two, upper limb dystonia and bilateral bicycling movements. All seizures concluded with sneezing twice. The EEG record was characterized by a buildup in fast activity in the left temporal regions and slowing in the left hemisphere in all three episodes, with attenuation in amplitude in one. In one of the three episodes, the slowing spreads bilaterally with a left-sided predominance. In all three episodes, sneezing occurred after the patient was responsive, and in two of the three, while left temporal slowing or amplitude attenuation was still present on the EEG.

Conclusion: Two of the three episodes of sneezing occurred while EEG was not back to baseline but the patient was responsive. In our case sneezing appeared to be a postictal event, probably representing a release phenomenon.

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Abstract – WCN 2013

No: 2793

Topic: 1 – Epilepsy

Vagus nerve stimulation in 19 patients with drug-resistant epilepsy in Western China

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Objective: To assess the efficacy and safety of vagus nerve stimulation (VNS) in drug-resistant epilepsy in Western China.

Methods: In this retrospective review of 19 patients who underwent VNS for drug-resistant epilepsy, there were 17 (89.5%) males and 2 (10.5%) females ranging in age from 7 to 58 years at the time of implantation. The generator was turned on two or three weeks after the plantation operation. Stimulation parameters were adjusted gradually. The output currents were adjusted from 0.25 mA, stimulative time was 30 s ON and 5 min OFF, the frequency was 20–30 Hz, and the pulse width was 500 μ s. Then the change of the output currents was 0.25 mA every 1–3 months.

Results: Duration of vagus nerve stimulation treatment varied from 3 to 29 months. Seizure freedom was achieved in 10 patients (52.5%). Seizure control \geq 90% was achieved in 11 patients (57.9%), \geq 75% seizure control in 12 patients (63.2%), \geq 50% improvement in 16 patients (84.2%), and $<$ 50% improvement in 3 patients (15.8%). There is no change of antiepileptic drugs (AEDs) in 14 patients; reduction of AEDs in 4 patients; increase of AEDs in one patient. Three patients (15.8%) reported one transient mild adverse effect.

Conclusion: VNS is a safe and effective palliative treatment for drug-resistant epilepsy in Western China. The key to the effectiveness of VNS is adjusting the right parameters to each patient. We believe better seizure control can be achieved as long as allowing more titration of the stimulation parameters and further AED regimen adjustments over time by the qualified epileptologists.

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Abstract — WCN 2013**No: 2797****Topic: 1 — Epilepsy****Variations in temporal lobe volumetry in association with temporal lobe epilepsy in Sudan**

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Introduction: Temporal lobe volumetry varies in correlation with normal variables like age and gender and with conditions like schizophrenia and epilepsy. This study considered these variations in patients with temporal lobe epilepsy compared to a matching control in Sudan.

Methodology: 63 patients with temporal lobe epilepsy were studied and compared to 69 matching controls. Three dimension structural MRI scan is done in the National Ribat University teaching hospital using 1.5 T Siemens scanner. 3D slicer 2.6 was used in computing the volume of the regions of interest.

Results: In the control group volumetry of the temporal lobe showed statistically significant left lateralization in males besides larger volume in males than in females. The superior temporal gyrus also showed left lateralization in males but the difference in volume between males and females is statistically not significant. The hippocampus showed right lateralization with statistically significant larger volumes in males than in females. In the study group left lateralization in the temporal lobe was not detected in males. The superior temporal gyrus showed statistically significant left lateralization in females. The temporal lobe and the superior temporal gyrus showed larger volumes in males than in females without volume difference compared to the control group. Hippocampal volume was reduced in the study group. Only the right hippocampus showed larger volumes in males than in females.

Conclusion: Findings in correlation with temporal lobe epilepsy differ between males and females. Some variations could be considered as preexisting risk factor for temporal lobe epilepsy.

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Abstract — WCN 2013**No: 2798****Topic: 1 — Epilepsy****MRI volumetry of the temporal lobe in Sudan: Validated protocol and preliminary normative data**

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Introduction: Variations in temporal lobe volumetry are reported in correlation with various normal factors like age, gender and handedness. No consensus in defining the borders of the temporal lobe, in particular the posterior border. This study aimed to design and validate a protocol for measuring the volume of the temporal lobe and then apply it in obtaining preliminary normative data in Sudan.

Methodology: 3D structural MRI was obtained in the National Ribat University teaching hospital, Sudan using 1.5 T Siemens scanner. 69 right handed apparently healthy volunteers were scanned. A protocol is designed considering the most postero-superior point of the lateral fissure as the posterior border. It is validated applying Cronbach's alpha in SPSS 16. 3D slicer 2.6 is used in volume measurements. The absolute volume is compared between genders and across each gender using ANOVA.

Results: Protocol reliability is 0.984. Larger volumes and left lateralization were detected in males but not in females.

Conclusions: The posterior end of the lateral fissure is a reliable point for temporal lobe demarcation. Volumetry of the temporal lobe is associated with gender. It displayed larger volumes as well as left lateralization in males than in females.

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Abstract — WCN 2013**No: 2799****Topic: 1 — Epilepsy****Increased risk of atherosclerosis antiepileptic drug use**

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Recent reports suggest that chronic epilepsy and long term AED usage may be associated with atherosclerosis.

By current study, we aimed to investigate the possible relation between antiepileptic drugs and atherosclerotic risk factors and carotid intima media thickness (IMT) as a marker of atherosclerosis. 92 patients, who has used carbamazepine (CBZ) and valproic acid (VPA) for three or more years and who has used a second antiepileptic drug (politherapy) for two years in addition to CBZ or VPA monotherapy, were included in the study. Control group composed of thirty one subjects with similar demographics. Fasting blood glucose, liver and renal function tests, lipid profile (triglyceride, total cholesterol, HDL-c, LDL-c), homocysteine, Hs-CRP, uric acid (UA), Lp(a), folate and vitamin B12 levels were examined in both groups.

The measurement of carotid intima media thickness (IMT) was performed by cranial magnetic resonance imaging and by cranial computerized tomography in patients whom magnetic resonance imaging could not be performed.

Although there was no difference between patients and control group in terms of IMT increase, there was positive correlation between carotid IMT and body mass index, T-col, LDL-c, Hs-CRP and homocysteine levels in the patient group. The finding, that there was no correlation between IMT values and duration of disease, duration of drug usage and dose, suggests that long term AED usage does not increase the development of atherosclerosis. It would be more appropriate to conduct novel long term follow-up studies in order to investigate whether AED usage leads to atherosclerosis.

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Abstract — WCN 2013**No: 2737****Topic: 1 — Epilepsy****Epilepsy care in Armenia. Data of 3382 consecutive epileptic patients**

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Objective: To assess comprehensive epilepsy care in Armenia in the situation of limited sector of government-sponsored health care and lack of private or public insurance.

Methods: The dates of 3382 consecutive patients with epilepsy were collected using specially designed questionnaires, which were filled in by trained epileptologists.

Results: 1552 (45.9%) females and 1830 (51.4%) males were involved. 41.8% of patients were inhabitants of Yerevan; other 58.2% were from regions. Immediately after diagnosis no treatment was initiated in 31% or was inadequate in 29%, and in 29% the suboptimal dose of appropriate AED was established. EEG was available for the vast majority (89.8%) and for 18.98% of patients also long-term (15–20 hours) video-EEG investigation was performed. 30.7% cases were idiopathic, 29.15% patients were symptomatic, 15.5% cases were cryptogenic and 24.4% were undefined. More frequently used AEDs are CBZ and VPA, or their combination. 11.68% of patients were prescribed to last generation AEDs either in monotherapy or any polytherapy regimen. Overall 18% of patients were seizure free at the moment of investigation.

Conclusions: Overall epilepsy care in Armenia is far from perfect and anticonvulsant treatment effectiveness is rather low with poor availability of last generation AEDs, and lack of non-AED treatment options even in the presence of adequate diagnostic and treatment protocols.

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Abstract – WCN 2013

No: 2777

Topic: 1 – Epilepsy

Features of memory impairment in temporal lobe epilepsy (tle) and juvenile myoclonic epilepsy (jme) in adults

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Background: Cognitive impairment, including memory loss, is quite common in epilepsy. The most frequently suffers memory loss in temporal lobe epilepsy, but also describes memory loss in JME (Kim, 2007).

Objective: To specify features of memory impairment for TLE and JME in adults, using neuropsychological and neurophysiological testing.

Patients and methods: We examined 10 healthy, 11 TLE patients and 10 JME patients, mean age of epilepsy group was 26.4 years, mean age of healthy group was 27.1 years. Objects were assessed using digit span, word span and cognitive evoked potentials (P300). Testing was conducted in the first half of the day and the patients had no depression according to Beck Depression Inventory. The patients with epilepsy were on monotherapy with antiepileptic drugs.

Results: The patients with epilepsy performed tests worse than the healthy patients ($p < 0.05$). But in TLE patients the worst results in digit span and word span meet the increased latency of P300 ($p < 0.01$). However, in JME, we did not find significant differences between the neuropsychological tests and evoked potentials.

Conclusion: Based on our data, we can assume the heterogeneous nature of memory impairment in epilepsy. However, some limitations in the study (small sample, antiepileptic therapy) require further clarification.

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Abstract – WCN 2013

No: 2852

Topic: 1 – Epilepsy

Management and outcome of patients with Landau Kleffner and continuous spike-waves during slow sleep syndromes

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Background: LKS and syndrome of CSWS share clinical features. They can be associated with electrical status epilepticus during slow-wave sleep (ESWS).

Objectives: Present study describes clinical and electroencephalographic findings in a cohort of 12 patients with LKS and CSWS and modalities of management. It aimed to verify long term outcome.

Patients and methods: The study included 10 males and 2 females with suspected epileptic encephalopathy (mean age of 4.176 ± 2.25). It extended over a period of five years. Patients enrolled were evaluated at Division of Pediatric Neurology, KKHU, Saudi Arabia. All patients had seizures, of them, nine (75%) had language disorders. Ten patients (83.3%) had behavioral disturbances. Awake EEG was normal in 5 patients. When reaching deep sleep, five patients with LKS and two with CSWS had generalized discharges reaching 90% in patients with CSWS and ranging from 40 to 70% in others. Patients were treated by either; Intravenous methyl prednisolone, intravenous immunoglobulins (IVIG) or combined IVIG with steroids. In all patients, antiepileptic medications were adjusted individually. Outcomes range from recovery in 4 patients to permanent aphasia in one, seven improved with either residual language deficits or neuropsychologic impairment. Poor prognostic factors were onset ≤ 4 years of age, duration of aphasia \geq one year, and persistent of ESES.

Conclusion: Treatment requires reversal of ESES/CSWS pattern on EEG. Use of combination of corticosteroids and IVIG seems to be effective approach and it is important to use an appropriate dose and repeat a cycle of therapy whenever required.

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Abstract – WCN 2013

No: 2883

Topic: 1 – Epilepsy

Endothelial and inflammatory markers which play a major role in drug resistance in epileptic patients

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Background: There are few studies on endothelium dysfunction in epileptic subjects and the relation between the levels of inter-cellular adhesion molecule-1 (ICAM-1), vascular adhesion molecule-1 (VCAM-1), inflammatory response and blood brain barrier (BBB) dysfunction.

Objective: This study has investigated the level of ICAM, VCAM, IL-1 β and CRP as endothelial and inflammatory markers in epileptic patients.

Patients and methods: This research was a cross-sectional study comprising eighty subjects in four groups: uncontrolled, controlled epileptic, during seizure and normal control subjects. All subjects with structural lesion and focal neurological sign were excluded. Random blood samples were taken and then the level of ICAM, VCAM, IL-1 β and CRP of serum was measured using ELISA and compared.

Results: There were no significant differences between the baseline characteristics ($p > 0.05$). The mean level of ICAM and VCAM in controlled (ICAM: 85.708 ± 2.209 , VCAM: 69.0725 ± 5.715) and uncontrolled seizure (ICAM: 90.793 ± 3.810 , VCAM: 90.793 ± 3.810) subjects was significant ($p < 0.001$). However, the mean level of inflammatory cytokines (IL-1 β and CRP) just in epileptic subjects (controlled and uncontrolled seizure) (CPR: 0.528 ± 0.108 , IL-1 β : 0.155 ± 0.01) and during seizure group (CPR: 0.184 ± 0.008 , IL-1 β : 0.519 ± 0.105) was significant.

Conclusion: It was revealed that VCAM level in uncontrolled epileptic subjects was significantly lower than controlled epileptic subjects

leading to low penetrance of BBB and more drug resistance. Of inflammatory cytokines, IL-1 β during seizure group was significantly higher than that in each group. But CPR during seizure group was significantly lower than that in epileptic subjects but not in control subjects. Further studies are needed to support this hypothesis.

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Abstract – WCN 2013

No: 2905

Topic: 1 – Epilepsy

Early seizures in patients with acute ischemic stroke

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Background: The aim of this study is to establish the incidence of acute post-ischemic stroke seizures and to determine risk factors associated with early seizures in acute ischemic stroke.

Patients and methods: A total of 1370 consecutive patients with acute infarction were included. Onset seizures and early seizures were defined as seizures occurring within 24 h and 7 days from the onset of infarction, respectively. Patients with high fever, previous history of epilepsy, and metabolic encephalopathy were excluded.

Results: Eleven patients (0.8%) developed early seizures after acute infarction; and 6 patients (55%) had onset seizures. TOAST classification of the stroke among those patients was large artery disease in 4 (36%), cardio-embolism in 3 (27%), vasculitis in 1 (9%), and cryptogenic stroke in 3 cases (27%). All of the 11 patients developed generalized tonic and/or clonic seizures. All patients had cortical involvement with various size of the lesions; large in 3 (27%), moderate in 5 (45%), and small lesions in 3 (27%). There were 2 cases of hemorrhagic transformation of the lesions.

Conclusions: Since all patients had cortical lesions, the most important factor of early seizures may be the location of lesions. Patients with lacunar and infratentorial infarctions did not develop early seizures. Patients with onset seizures had more seizures during acute stage than those without onset seizure.

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Abstract – WCN 2013

No: 2944

Topic: 1 – Epilepsy

Usefulness of PET/CT in diagnosis of temporal epilepsy

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Aim: The aim of this study was to assess the usefulness of routine 18F FDG PET/CT examination in patients with temporal lobe epilepsy before surgery.

Material and methods: 41 patients with medically refractory temporal lobe epilepsy (21 women and 20 men, age 31 \pm 9) underwent interictal PET/CT examination approximately 30 min post-i.v. injection of 250 MBq 18F FDG and contrast enhanced brain MRI. All patients underwent surgery (temporal lobe with hippocampus resection). The diagnosis was verified by histopathology. PET and MRI were assessed by two independent physicians, who were blinded to clinical signs and results of EEG.

Results: In 34/41 (83%) of patients in PET/CT areas of hypometabolism including mesial temporal lobe, that correlated with histopathology,

were observed. Pathologies were present in 18/41 (43%) of patients in MRI.

In 2 patients hypometabolism was observed in both mesial temporal lobes without lateralization of changes.

In 1 patient area of hypometabolism was present in the contralateral temporal lobe, there was no pathology in MRI.

In 4/41 (10%) of patients there was no pathology in PET/CT and MRI.

Histopathological findings included tumours (ganglioglioma, oligodendroglioma and DNET), focal cortical dysplasia (FCD) type IB, FCD IIA, FCD IIIa and hippocampal sclerosis. There were no significant differences in FDG accumulation between these pathologies.

Conclusions: 18F FDG PET/CT shows markedly more often abnormal tracer distribution that could correspond to epileptogenic foci than MRI.

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Abstract – WCN 2013

No: 2924

Topic: 1 – Epilepsy

Insights gained through the China demonstration project – practical experience from Sichuan

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Epilepsy is affecting 50 million people in the world, 85% of whom live in resource-poor countries. Epilepsy also imposes an enormous physical, psychological, social and economic burden on individuals, families and countries. In 1997, the Global Campaign Against Epilepsy was launched in China. As a part of the campaign, a demonstration project on epilepsy management was carried out in rural China, also in Sichuan in 2005.

In the past 8 years, this project expanded to 9 countries and covered 4,937,000 populations in Sichuan province. 8128 patients were enrolled and 6547 received phenobarbital or sodium valproate treatment. After 8 years of follow-up, 79.8% of the patients had 50% or greater reduction in seizure frequency and 43.3% remained seizure free. The quality of life greatly improved in 72% patients. The treatment gap was successfully reduced to 13%. The main premature cause of death of epilepsy patients is accidental death (59%), especially drowning (45.1%) and then we did targeted prevention. We made the intervention package to enhance medical compliance and improve seizure control, which helped the proportion of patients with >50% seizure reduction to 79.8%. Besides, 837 physicians were efficiently trained to treat people with epilepsy. The most important, this project had successfully implemented a sustainable treatment and management model of epilepsy in rural areas.

This project not only completed its basic aim in promoting education, training, and treatment for epilepsy patients in rural China, but also helped to find an operational model to sustain and success in management of chronic disease in resource-poor regions.

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Abstract – WCN 2013

No: 2960

Topic: 1 – Epilepsy

The nature history of untreated epilepsy in rural areas in Tibetan areas

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Purpose: To evaluate the nature history of people with untreated prevalent epilepsy in rural Tibetan areas.

Methods: During 2006 to 2010, we carried out a door to door epidemiologic community survey in a sample of 60,466 subjects in 5 rural counties in both Tibetan Autonomous Regions and Sichuan province. People with suspicious symptoms are investigated retrospectively, but only active epilepsy has been registered and followed up regularly and prospectively.

Results: At the survey, we identified 473 (60.7% male) people with convulsive epilepsy, of whom 367 (61.9% male) were “active epilepsy” and followed up for mean 41 ± 8.6 months. In the retrospective survey, 22.4% patients with untreated become spontaneous remission for more than 5 years, 31.7% become spontaneous remission for more than 3 years, and 48.8% seizure free for only 1 year. During the follow-up, 79 (21.5%) patients were tried to treat by antiepileptic drug (AED), but only 34 continued the treatment for more than 3 years, 45 (12.3%) patients used traditional Tibetan medicine, and another 243 (66.2%) patients believe in God instead of AED. 12 (3.3%) subjects died. 174 patients are followed up more than 5 years, 48(27.6%) of them has spontaneous remission. For the 308 who were untreated and under treatment followed up, 96(31.2%) of them have spontaneous remission for 3 years. Besides, 55.9% (19/34) of those who take AED regularly have seizure free after 3 years of treatment.

Conclusion: Spontaneous remission of epilepsy occurs in a portion of untreated patients. The AEDs do have an effect to elevate the rate of seizure free.

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Abstract – WCN 2013

No: 2974

Topic: 1 – Epilepsy

Localization of focal delta activity in epilepsy patients after brain surgery using MEG

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Background: In this study we introduce an automatic method for source localization of focal slowing demonstrating the relation between spikes and delta activity in 9 patients after previous brain surgery.

Objective: In case of focal epilepsy, surgery can often improve seizure frequency in these pharmacoresistant patients. However, not all patients are completely seizure free thereafter. Residual epileptic activity which leads to persisting seizures can be localized by interictal spikes. Additionally slow wave brain activity can be used for focus localization.

Patients and methods: Nine patients with pharmacoresistant epilepsy after brain surgery were examined using MEG. 20 min of spontaneous brain activity at rest were recorded and sources of delta and theta activities were localized with Dynamic Imaging of Coherent Sources (DICS, 5). Additionally interictal spike localization results were used for setting up a spherical region of interest (ROI) with a radius of 2 cm.

Results: In 3/9 patients, the global maximum of the theta band hits the ROI, in another 3 patients it is the third or fourth local maximum, and in 3 patients no local maximum is found within the ROI at all. The localization results lie at the borders, not more than 2 cm away from the spikes.

Conclusion: The results show a strong relation between interictal spikes and delta activity in patients after brain surgery. The method runs automatic, so it is user-independent and faster than visual inspection of data to find segments with slowing of brain activity.

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Abstract – WCN 2013

No: 2978

Topic: 1 – Epilepsy

Functioning and disability in patients with epilepsy: psychosocial difficulties reported by patients

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Background: Although the burden and associated psychosocial difficulties (PSDs) in persons with epilepsy (PwE) are known and recognized, there is not a comprehensive understanding of those difficulties and the factors that have an impact on them.

Objective: This study aimed to identify and analyse psychosocial difficulties that adult PwE experience in everyday life, their changes and determinants.

Patients and methods: 80 adult PwE (ICD-10: G40.1; G40.2 and G40.3) were interviewed using the EU project PARADISE Protocol.

Results: Mean age of patients was 41 years, 50% were females, 53% were married, 70% had at least high school education, and 66% were working. Clinical rating of epilepsy severity was moderate or severe for 69% of PwE and on average patients took two anti-epileptic drugs.

PSDs more often reported by PwE were related to memory problems (58%), being emotionally affected by health condition (74%), anxiety (69%), depressive symptoms (66%), problems at work (55%), driving problems (60%), and restlessness (80%). Half of the sample reported that PSDs got better in time in comparison with the previous years while 27% considered that there was no change. Main determinants acting as moderate or strong environmental barriers were side-effects of medicines (59%) and lack of sensitivity towards PwE (52%).

Conclusions: People with epilepsy as main respondents reported their PSDs. Perceived improvement of difficulties over time and identified determinants that have impact on PSDs suggest that it is important to consider environment's impact on health and disability when planning personalised care programmes for PwE.

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Abstract – WCN 2013

No: 2985

Topic: 1 – Epilepsy

Imaging memory in temporal lobe epilepsy: reorganisation of verbal and visual memory function following anterior temporal lobe resection

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Background: Anterior temporal lobe resection (ATLR) controls seizures in 70% of patients with intractable temporal lobe epilepsy

(TLE) but may impair memory function, typically verbal memory following left and visual memory following right ATRL.

Functional reorganisation can occur within the unaffected ipsilateral and contralateral hemispheres. We investigated reorganisation of memory function in TLE before and after left or right ATRL and the efficiency of postoperative memory networks.

Methods: We studied 46 patients with unilateral medial TLE (26 left) on a 3T GE-MRI scanner. All subjects had neuropsychological testing and performed an fMRI memory encoding paradigm for words, pictures and faces preoperatively and four months after left or right ATRL.

Results: Event-related analysis revealed that left TLE had greater activation in the left posterior medial temporal lobe (MTL) for encoding words postoperatively than preoperatively.

Greater pre- than postoperative activation for encoding words in the ipsilateral posterior MTL correlated with better verbal memory outcome after left ATRL.

After left ATRL greater postoperative than preoperative activation in the ipsilateral posterior MTL correlated with less good postoperative verbal memory performance, an effect that was not observed for visual memory after right ATRL.

Conclusion: We found effective preoperative reorganisation of verbal memory function to the ipsilateral posterior MTL, suggesting that it is the capacity of the posterior remnant of the ipsilateral hippocampus rather than the functional reserve of the contralateral hippocampus that is important for maintaining verbal memory function after ATRL; early postoperative reorganisation to ipsilateral posterior or contralateral MTL structures are inefficient.

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Abstract – WCN 2013

No: 3018

Topic: 1 – Epilepsy

Increased seizure frequency due to the copper deficiency in Wilson's disease

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Background: Epileptic seizures have been reported in Wilson's disease (WD) in 6–7% of patients, mostly following the chelator therapy. Pyridoxine deficiency due to penicillamine, direct copper toxicity, pathological changes, metabolic encephalopathy and rarely copper deficiency are the attributed mechanisms for seizures.

Objective: To present a patient with WD and intractable seizures, taking chelator and antiepileptic therapy with a strict copper restricted diet.

Patients and methods: A-29-year old female patient admitted to emergency unit with recurrent seizures. She had been diagnosed as WD and epilepsy in another center previously with a treatment of penicillamine, zinc acetate and carbamazepine. Complex partial and less frequent secondary generalized seizures had been started after taking penicillamine and responded well to carbamazepine initially. She complained increased daily frequency of seizures ranging from 5 to 10 in recent months.

Results: Although effective serum carbamazepine level and levetiracetam add-on therapy, changed chelator therapy with trientine and oral pyridoxine, daily frequency of seizures did not reduce and low serum copper [0.3 µg/dl (70–140)] has been attributed for the seizures. Zinc acetate and strict copper restricted diet were ended. As the serum copper levels increased during follow-up [16 µg/dl (70–140)], seizure frequency was decreased.

Conclusion: Epileptic seizures respond well to antiepileptic treatment in WD. In this case, add-on antiepileptic and pyridoxine therapy did not reduce the seizure frequency. The relation between decreased seizure frequency and increased serum copper levels

shows that copper deficiency can cause intractable seizures in patients who are taking chelator therapy with a strict copper restricted diet.

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Abstract – WCN 2013

No: 3019

Topic: 1 – Epilepsy

Attentional deficit in childhood benign epilepsy with centrotemporal spikes (beets) and absence seizure epilepsy

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Objective: There is controversy regarding the neuropsychological profile of children with epilepsy. The aim of this study is to analyze the attentional profile of children with benign epilepsy with centrotemporal spikes (BECTS) compared with children with absence seizures.

Methods: Sample: 34 children from 7 to 12 years (18 with BECTS and 16 with absence seizures). We get the attentional profile by applying the WISC-IV (Wechsler Intelligence Scale for Children-IV), Battery Luria-DNI (Manga and Ramos, 1991) and Attention Test d2 (Brickenkamp, 2002).

Results:

- We haven't found any differences in IQ between children with BECTS and absence seizures.
- We haven't found any differences in attentional profile when we use WISC-IV.
- Nevertheless, when we use specific Attentional Test d2, we have found a significant ($p < 0.05$) deficit in children with BECTS compared with absence seizure children in attentional control, inhibitory control, working capacity, concentration, and selective and sustained attention.

Conclusions: We must consider, in children with this diagnosis, the neuropsychological profile described to strengthen deficient neuropsychological and psychoeducational areas. In addition, we must continue research in future studies on the role of the attention in children with epilepsy.

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Abstract – WCN 2013

No: 2953

Topic: 1 – Epilepsy

Expression of adenosine kinase in human mesial temporal lobe epilepsy with hippocampal sclerosis: A preliminary study

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Background: Adenosine is a ubiquitous homeostatic molecule that acts as an "endogenous neuromodulator". Adenosine attenuates neuronal activity either presynaptically by inhibiting neurotransmitter release or by controlling neurotransmitter responsiveness at post-synaptic sites. Unbalanced adenosine metabolism has been

implicated in pathological conditions such as epilepsy. Adenosine kinase (ADK), synthesized by astrocytes, is the key regulator of extracellular adenosine levels in the brain. Evidences from experimental studies support a role for ADK in brain injury associated with astrogliosis, a morphological hallmark of Mesial Temporal Lobe Epilepsy with Hippocampal Sclerosis (MTLE-HS). In fact, expression of astrocytic ADK was found to be increased in the hippocampus and temporal cortex of MTLE-HS patients. Overexpression of ADK decreases extracellular adenosine and consequently may cause seizures. The aim of this study was to characterize ADK gene expression in MTLE-HS patients.

Methods: Previous studies used immunohistochemistry and Western blot analysis to investigate ADK expression. Here we quantified the expression levels of ADK by Real-Time PCR in the hippocampus (lesional and peri-lesional cortical area) of 10 MTLE-HS patients submitted to surgery as compared with 9 autopsy controls with no history of neurological disorders.

Results: Our results showed that ADK expression levels were similar in the hippocampus and temporal cortex of MTLE-HS patients when compared to healthy controls.

Conclusion: Our preliminary data demonstrate that ADK expression levels are not altered in MTLE-HS. These results do not preclude post-transcriptional ADK abnormalities at both protein and functional levels. Our results should be confirmed in a larger cohort as well as with complementary methodologies.

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Abstract – WCN 2013

No: 3057

Topic: 1 – Epilepsy

Proposal of a staging system for risk stratification and cross sectional reporting of status epilepticus

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Background: Definitions for status epilepticus (SE) vary regarding their time points and -windows of treatment options. Clinical phenomena are highly dynamic and symptoms may gradually disappear, as status advances (electromechanical dissociation). Pragmatically, this may cause problems, when transferring a patient in SE from one to another unit (e.g. emergency department to ICU). To date no standardized system exists for acquisition, documentation and communication of clinical core parameters to stage SE. Such a system would make research results better comparable for meta-analyses and serve as a tool for risk stratification.

Methods: Fifty consecutive patients with various types of status epilepticus of any cause were classified according to a proposed staging system for status epilepticus (SSSE) in a tertiary neurological university clinic in Salzburg, Austria. A data sheet was designed and explained to responsible physicians in charge. The SSSE uses a letter code comparable to the pTNM-system in tumors. A “D” denotes the definition based on clinical grounds, “S” for semiology, “N” for level of neurophysiological evidence, “L” for laboratory changes and “I” for results of neuroimaging.

Results: SSSE was easily applicable and well accepted by physicians, did not interfere with patient management and was suitable for communication in medical records.

Conclusion: The SSSE is applicable to various forms of SE in any clinical setting with low time consumption and no interference with acute patient management. Larger prospective studies are needed to perform risk stratification and to determine test characteristics such as inter-rater reliability.

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Abstract – WCN 2013

No: 2951

Topic: 1 – Epilepsy

Clinical and CT-scan aspects of neurocysticercosis (NCC) at teaching hospital Yalgado Ouédraogo concerning 35 cases

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Background: The clinical and the CT-scan features of NCC are poorly studied in sub-saharian Africa. This study aims to emphasize the clinical and CT-scan characteristics of NCC at teaching hospital Yalgado Ouédraogo.

Patients and methods: We conducted a transversal prospective study from February 2001 to April 2006. The diagnosis has been established on the basis of clinical and cerebral CT scan examinations. All the patients received an antihelminthic drug.

Objective: This study focused on the clinical and CT features of NCC at Teaching Hospital Yalgado Ouédraogo, Ouagadougou, Burkina Faso.

Results: The most common clinical event in our study was seizure (94.3%) and generalized seizure was the most found type. Seizures were followed by headache with or without ICP (62.9%) and by focal deficit (17.1%). All the patients had a parenchymal location associated to extraparenchymal locations in the rate of 20%. There were 85.7% of isolated supratentorial lesions. The number of lesions among patients ranged from 1 to 67. The calcification and the vesicle were the most frequent lesions. Two solitary cerebral cysticercus granuloma and two giant cysts have been brought out. 94.6% of the patients with parenchymal location had seizures. NCC lesions were associated to cerebral infarction in two patients.

Conclusion: The diagnosis of NCC has been revolutionized by CT scan. MRI and immunologic tests which are unavailable in Burkina Faso will help in the diagnosis and the management of this infection.

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Abstract – WCN 2013

No: 3088

Topic: 1 – Epilepsy

Views of young people toward epilepsy in Novi Sad

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Epilepsy is a significant health problem especially prominent in underdeveloped countries. Quality of life in epilepsy is affected not only by the subjective experience of the illness it manifests, and long-lasting therapy, but also by the inappropriate attitude of the society that is mostly uniformed about the real nature of epilepsy.

In this investigation it was analyzed how well young people are informed about epilepsy and its characteristics as well as first aid to epileptic. A special questionnaire for investigation was used. The first part of the questionnaire was used to determine the socio-demographic characteristics of the study participants, and the second part dealt with student knowledge of epilepsy.

In this study 432 students were included from different schools of University in Novi Sad (excluding students from medical schools), ages 20–24, both sex.

The results show that the young people are familiar with epilepsy, and they have a basic knowledge regarding nature of the disease, but due to different subjective attitudes towards epilepsy and epileptic patients, it can be said that the attitude of the young population

toward epilepsy is still based on a low level information and prejudice. Students from the countryside were better informed than the students from the city. The knowledge about epilepsy was not dependent on sex. We can conclude that there is an adequate level of awareness and knowledge toward epilepsy among the young people in Novi Sad. However, there is still a need for further improvement of certain aspects of knowledge and understanding of the epilepsy.

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Abstract – WCN 2013

No: 3097

Topic: 1 – Epilepsy

Source analysis in patients with idiopathic generalized epilepsy

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Generalized spike and wave (GSW) discharges is the main neurophysiological signature of idiopathic generalized epilepsies (IGE). Investigations support that the pathophysiology of GSW and IGE includes a subtle cortical focal abnormality.

Investigate the relation between focal findings in the EEG and MRI of patients with IGE using quantitative techniques.

4 patients with IGE were investigated. EEG was recorded with 22 channels and source analysis was conducted using BESA software. GSW discharges were centered and averaged. Equivalent dipole and CLARA algorithms were used for source localization. All patients and 13 healthy controls were submitted to 1.5T MRI. Volumetric (3D) T1 sequence was used for structural evaluation of the cortical surface. Cortical reconstruction and statistical analysis was performed with Freesurfer. All images were submitted to the same processing. Using GLM the cortex of each patient with IGE was individually compared with the control group.

Subject 1: 10 discharges. GSW onset involved the posterior portion of the right parahippocampal gyrus. MRI showed reduced cortical thickness in the right supramarginal gyrus.

Subject 2: 4 discharges. GSW onset involved the left occipital lobe. MRI showed increased curvature in the left precuneus.

Subject 3: 80 discharges. GSW onset involved the right inferior frontal gyrus. MRI showed decreased curvature in the right insula.

Subject 4: 6 discharges. GSW and MRI (reduced curvature) showed involvement of the right anterior cingulate.

Cortical analysis and EEG source imaging showed an interesting correspondence. This study supports that focal onset and fast spreading networks may be involved in IGE mechanisms.

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Abstract – WCN 2013

No: 3051

Topic: 1 – Epilepsy

Dermatological manifestations of epilepsy among adult Sudanese epileptic patients

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Introduction: Epilepsy is a clinical syndrome characterized by increase electrical excitability of cortical neurons with or without loss of consciousness.

Objective: To study the pattern of dermatological changes associated with epilepsy among adult Sudanese epileptic patients.

Methods: This non-interventional descriptive study, 360 adult Sudanese epileptic patients were included in the study which was conducted at El shaab Teaching Hospital during the period from Feb 2004 to Aug 2007. Full detailed history and proper clinical examination were performed by

the authors. Dermatological changes were assessed by dermatologist. List of investigations was done including EEG, CT of the brain and serial of drug serum level.

Results: Out of 360 patients 31 were found to have scars due to repeated attacks of convulsions, one patient was found to have neurofibromatoma, one had tuberous-sclerosis, one had Sturge-Weber syndrome, one had Kaposi sarcoma, one had SLE, one diabetic patient had skin atrophy, one patient use to take phenobarbitone had skin eruption, and one patient on carbamazepine had skin change while five patients on phenytoin had skin manifestations.

Conclusion: Skin changes can occur in epileptic patients as part of drugs toxicity, or as part of clinical manifestations of certain diseases that can cause secondary epilepsy e.g. neurofibroma.

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Abstract – WCN 2013

No: 3096

Topic: 1 – Epilepsy

Hail Mary prayer – An ictal manifestation of non-convulsive status epilepticus

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Introduction: Epilepsy can be associated with religious manifestations in the form of experiential auras, post-ictal and interictal religious experiences. Prayers have not been described as possible ictal manifestations. In addition, ictal speech manifestations are common but ictal aphasia is a less frequent phenomenon. Specifically, the occurrence of ictal fluent aphasia is seldom reported. We describe a patient in which non convulsive status epilepticus was associated with continuous fluent jargon aphasia resembling a prayer.

Case report: A previously independent 81-year-old woman was admitted to our hospital with delirium apparently due to hyponatremia (121 mEq/L), a urinary infection and hypothyroidism. Four days after admission, she developed fluent jargon aphasia resembling “Hail Mary” prayer. There were no more new findings on neurological examination. EEG revealed multiple epileptic seizures beginning in left central leads with fast propagation to frontal and fronto-temporal areas, in accordance with non-convulsive status epilepticus. Head MRI revealed small restriction to diffusion concerning the left thalamus. Lumbar puncture was normal and anti-neuronal antibodies were negative. The patient was treated with valproate, levetiracetam, phenytoin, topiramate and levothyroxine with progressive clinical and electroencephalographic improvement. Two months later, there was recurrence of epileptic status and progressive cognitive deterioration, partially reversed with further anti-epileptic and corticosteroid treatment. The etiology of the status epilepticus remains elusive.

Conclusion: To our knowledge, this is the first reported case of non convulsive status epilepticus presenting as fluent jargon aphasia resembling a prayer. Clinicians should be aware of this possible presentation of status epilepticus.

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Abstract – WCN 2013

No: 3085

Topic: 1 – Epilepsy

Stigma of epilepsy among patients and their relatives attending charity clinic, Omdurman – Sudan

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Introduction: Epilepsy is the world's most common and most serious chronic neurological disease affecting near 50 million cases. Epilepsy has well recognized stigma which is defined by Link and Phalen as a status which exists when elements of labeling, stereotyping, separation, status loss, and discrimination occur together in a power situation that's allows them to unfold. Stigma has a major contribution to the reduction of quality of life in people with epilepsy.

Objective: The purpose of this study was to determine the types of stigma in PWE and to determine its frequency as well as its association with demographical factors and to determine the coping ability of PWE.

Method: Health facility-based cross-sectional analytical study was conducted in charity clinic on 80 people with epilepsy aged from 8 to 70 years. Standardized questionnaire was used for the interview of the patients and assistance was asked from their relative occasionally and the data was analyzed by SPSS16.

Results: 16.3% was found to have positive Felt stigma score. Nearly half of PWE had courtesy stigma also coaching stigma affected half of PWE. Fifth of PWE had poor coping score. Age below forty was a determinant factor for the coping of people with epilepsy as well as parent courtesy stigma. Frequency of seizures of more than 3 per month reduced coping score of PWE.

Conclusion: Results suggest the great need for psychological counseling for high risk group PWE as shown by the study.

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Abstract – WCN 2013

No: 3122

Topic: 1 – Epilepsy

Epilepsy in the Amazon: An invisible disease in the rain forest

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Background: Epilepsy is a serious disease with a lifetime prevalence of 1–2%. Epilepsy can be cost-effectively treated, yet the treatment gap is still shameless high even in mainstream parts of the globe. One can just wonder how epilepsy is managed in non-easily-accessible regions like Amazon rain forest.

Objective: To assess the perception epilepsy prevalence by the primary care staff working in Amazon region.

People and methods: We applied a home-made questionnaire to staff from three primary health care units in the city of Parintins located in the Oriental Amazon. This is part of the Strategy and Plan of Action on Epilepsy of PAHO-WHO/ILAE/IBE carried out by ASPE in Brazil.

Results: One-hundred-six people (91 women) answered the questionnaire (3 physicians, 4 nurses, 28 professional-allied-medicine, 60 health agents, and 11 others). When asked about: the prevalence of epilepsy, 76 (71%) did not know; whether they received any training on epilepsy, 41 had training, but only 11/41 (38%) pointed 1–2% as prevalence rate. None of the physicians and nurses knew how many patients should be under their care. When inquired about how many were they have currently, three did not know and the remaining four claimed to have between 2 and 10 patients.

Conclusion: The primary health care in Amazon does not have a planning for epilepsy management, as epilepsy seems to be an invisible disease. Actions to increase detection as well as provision of treatment are required in the region to bring epilepsy out of the shadows without cutting a tree.

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Abstract – WCN 2013

No: 3143

Topic: 1 – Epilepsy

HLA-B*1502 genotyping in carbamazepine induced Stevens Johnson's syndrome and toxic epidermal necrolysis

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Background: Stevens Johnson's Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are potentially fatal drug reactions caused by carbamazepine (CBZ). A strong association is reported between HLA-B*1502 allele and CBZ-induced SJS in Han Chinese. The prevalence of HLA-B*1502 and incidence of CBZ induced SJS/TEN in HLA-B*1502 positive seizure patients is reported.

Objective: To study the association of HLA-B*1502 and carbamazepine induced SJS/TEN in epileptic patients from South India.

Methods: Single center, prospective, case control study over four years. 7 ml of blood in ethylene diamine tetra acetic acid (EDTA) was collected from 352 patients on CBZ and 100 controls. After DNA separation, HLA-B*1502 genotyping was performed by Polymerase Chain Reaction amplification and gel electrophoresis, using Olerup HLA-B*1502 kits by Sequence Specific Oligopeptide method.

Results: 352 seizure patients (165 men and 187 women, mean age: 31.77 years) and 100 controls (50 men and women each, mean age: 32.05 years) were tested for HLA-B*1502. Out of 330 patients on CBZ tested for HLA-B*1502, 66 tested positive and 54 developed SJS/TEN. 2 patients on carbamazepine who tested negative for HLA-B*1502, developed SJS/TEN. Positive Predictive Value for development of SJS/TEN on exposure to CBZ: 81.82%, Negative Predictive Value: 99.30%, Sensitivity: 96.43%, and Specificity: 95.95%. The prevalence of HLA-B*1502 in the epileptic and control groups was 18.20% and 7% respectively.

Conclusion: There is a high prevalence HLA-B*1502 allele in South Indian epileptic population on Carbamazepine (18.20%) when compared to the non-epileptic population (7%). There was a strong association of HLA-B*1502 and the development of SJS/TEN in South Indians.

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Abstract – WCN 2013

No: 3139

Topic: 1 – Epilepsy

Autoantibodies to neuronal antigens in children with new onset seizures classified according to the revised ILAE classification

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Background: Autoantibodies against neuronal antigens are increasingly recognised in many CNS disorders including epilepsy. This includes neuronal surface proteins such as VGKC complex and its components (LG11, CASPR2 and contactin2), NMDAR, GABA_BR, AMPAR and glycine receptor (GlyR), as well as intracellular proteins such as GAD.

Objectives: We aimed to investigate neuronal antibodies in a cohort of paediatric patients with new onset seizures.

Methods: We prospectively recruited 114 children (2 month to 16 years) with new onset seizures presenting between September

2009 and November 2011 to the Children Hospital at Westmead, Australia, as well as 65 age-matched controls. Patients were clinically assessed and classified according to the revised ILAE classification 2010. Sera were tested blinded to a range of antigens by radioimmuno- and cell-based assays.

Results: Eleven out of 114 (9.7%) patients were positive for one or more autoantibodies compared to 3 of 65 controls (4.6%, $p = ns$). Patients had antibodies to VGKC-complex ($n = 4$), CASPR2 ($n = 3$), NMDAR ($n = 2$), VGKC-complex and NMDAR ($n = 2$). The proportion of patients with epilepsy of unknown cause was higher in the antibody positive group (7/11; 63%) compared with the antibody negative subjects (23/103; 22%; $p = 0.007$, Fisher's exact test). Furthermore, 4 of 11 antibody-positive patients had focal epilepsy of unknown cause (36.4%) compared to only 12/113 of the antibody-negative patients (11.7%).

Conclusions: Specific neuronal antibodies are present in children with new-onset epilepsy of "unknown cause", often with focal epilepsies, this group of children may benefit most from autoantibody screening and consideration of immune therapy.

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Abstract – WCN 2013

No: 3150

Topic: 1 – Epilepsy

Serum trace element levels in children receiving antiepileptic drug therapy: A cross-sectional study

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Purpose: Few studies have evaluated the effects of antiepileptic drug (AED) therapy on trace element status in children and their results have been conflicting. The newer AEDs are considered to have a more acceptable safety profile, but this confidence is somewhat guarded in the absence of long-term data. This cross-sectional study was conducted to analyze the serum trace element levels in epileptic children treated with conventional and newer AEDs and compare them with healthy controls.

Methods: The study included 92 epileptic children and 28 healthy controls. The participant distribution was as follows,

Group I Phenytoin (PHT) monotherapy ($n = 35$),
Group II Valproate (VPA) monotherapy ($n = 30$),
Group III Valproate plus Levetiracetam (VPA + LEV) ($n = 27$),
Group IV Healthy controls ($n = 28$).

Serum levels of seven trace elements i.e. zinc, copper, magnesium, manganese, iron, selenium and strontium were determined using inductively coupled plasma-atomic emission spectrometry (ICP-AES).

Results: Phenytoin monotherapy was associated with increased copper (1568.8 $\mu\text{g/L}$ vs. 1053.6 $\mu\text{g/L}$, $p = 0.009$) and strontium (37.0 $\mu\text{g/L}$ vs. 30.7 $\mu\text{g/L}$, $p < 0.001$) concentrations & decreased manganese levels (1.5 $\mu\text{g/L}$ vs. 1.9 $\mu\text{g/L}$, $p = 0.04$). Valproate monotherapy treated children had decreased serum zinc (1010.5 $\mu\text{g/L}$ vs. 1242.9 $\mu\text{g/L}$, $p = 0.01$) and selenium levels (67.0 $\mu\text{g/L}$ vs. 84.7 $\mu\text{g/L}$, $p = 0.02$) as compared to healthy controls. However, in VPA + LEV group no significant differences were observed in trace element profile as compared to healthy children.

Conclusions: A significant difference in trace element levels in VPA and PHT treated epileptic children as compared to controls suggests a possible association between AED therapy and trace element alterations. However, levetiracetam when used in combination with

valproate was not associated with these alterations. These findings further support its favorable adverse effect profile as compared to conventional AEDs.

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Abstract – WCN 2013

No: 3177

Topic: 1 – Epilepsy

West syndrome: A case report

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West Syndrome is age-dependent epileptic encephalopathy of infancy, characterized by:

1. The infantile spasms – massive myoclonic, tonic, propulsive, retropulsive, symmetric, asymmetric, and serial isolated spasms of axial and limb muscles.
2. The EEG pattern hypsarrhythmia.
3. Psychomotor retardation.

Case: The child was delivered on 42–43 weeks of gestation by cesarean section due to fetal hypoxia. Apgar score was 4/5.

The first attack of tonic spasms with turning the head to the right at 3,5 month. On Brain MRI: multiple cysts and encephalomalacia, mixed hydrocephalus. Was appointed *konvulex* 25 mg/kg/day. Attacks continued.

At the age of 6 months. EEG: saved atypical hypsarrhythmia with a regional accent. Sabril added 50 mg/day, the attacks were reduced to 20–30 per day.

At the age of 9.5 months. Received synacthen-depot 0.125 mg/day every day 1 week, then every other day 2 weeks. On the 4th day of treatment, the child began to laugh, pronounce sounds, trying to get up on all fours.

The age of 1 year and 2 months. Psychomotor retardation, observed dextral asymmetric spasms with turning the head to the right and making pose "swordsmen", single flexor and extensor spasms Taking Sabril 5 mg/kg/day, topiramate 3 mg/kg/day, *konvulex* 15 mg/kg/day.

Conclusion: This case is a symptomatic form of WS, which leads to resistance in the appointment of AEDs. Appointment of hormones led to a temporary reduction in seizure frequency. The prognosis of West syndrome depends on the time of diagnosis of the disease and prompts adequate therapy.

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Abstract – WCN 2013

No: 3172

Topic: 1 – Epilepsy

Investigating the gap in the pharmacological treatment of epilepsy in low- and middle-income countries

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Epilepsy affects more than 50 million people worldwide, with many more affected by its consequences. Nearly 80% of the people with

epilepsy live in developing regions and the majority of them do not receive appropriate treatment. This treatment gap is partly attributable to the lack of availability of antiepileptic drugs (AEDs). The WHO has developed a Model List of Essential Medicines (WMLEM) designed to prioritize health care needs, improve the drug supply and lower the costs in health care to assist countries in procurement and supply of medicines prioritization.

Aim: To assess to what extent essential AEDs, as recommended in the WMLEM, are present in countries' Essential Medicine Lists (EML) and review their accessibility at the health system level to develop a comprehensive view of treatment gaps.

A comparative analysis was performed on 109 EMLs of member countries and the corresponding chapter 5 WMLEM to assess quantitative and qualitative differences concerning the essential AEDs.

Phenobarbital was included in the EML of 96% of the responding countries, carbamazepine in 95%, phenytoin in 83%, and valproic acid in 92%. The medications were provided as recommended by the WMLEM in a fraction of these cases: phenobarbital in 12%, carbamazepine in 5%, phenytoin in 3%, and valproic acid in 11%. First-line AEDs were least likely to be included in low-income countries. All medications were more likely to be available in the hospital compared to the health center.

Consistent use and application of WMLEM in guiding national EML development can help improve availability of AEDs.

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Abstract – WCN 2013

No: 3207

Topic: 1 – Epilepsy

Psychiatric comorbidity and social aspects in men with epilepsy – The Norwegian mother and child cohort study

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Objectives: To investigate psychiatric disease and social aspects in men with epilepsy.

Method: This study was based on the Norwegian Mother and Child Cohort Study (MoBa). Information included self-reported diagnoses and validated diagnostic instruments from 71,700 men, who were fathers of children registered in MoBa. Questionnaires were answered around week 18 of pregnancy.

Results: 658 men (mean age 31.8 years) reported epilepsy, 36.9% using antiepileptic drugs (AEDs). Compared to the rest of the MoBa cohort, untreated men with epilepsy more often screened positive for present depression (15.3% vs. 10.3%, $p = 0.001$), while AED treated men more often had a history of Lifetime Major Depression (15.1% vs. 10.1%, $p = 0.011$). Self-reported ADHD was associated with untreated epilepsy (3.4% vs. 0.4%, $p < 0.001$), as was bipolar disorder (2.2% vs. 0.3%, $p = 0.003$), and unspecified psychiatric disorders (5.6% vs. 2.3%, $p = 0.008$). Unemployment due to disability was linked to both AED treated (9.1% vs. 1.4%, $p < 0.001$) and untreated

epilepsy (2.9% vs. 1.4%, $p = 0.009$), as were low income (10.3% vs. 5.4%, $p = 0.031$ and 9.7% vs. 5.4%, $p = 0.011$). Smoking was associated with untreated epilepsy (60.2% vs. 51.8%, $p = 0.001$). No difference was found for lower education level, alcohol use or narcotic use.

Conclusion: Epilepsy in young men is associated with a higher frequency of psychiatric disorders and adverse social aspects, including both AED treated and untreated epilepsy. Our findings indicate that untreated men with epilepsy are a more vulnerable group that may be in need of patient follow up even if they are not using AED.

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Abstract – WCN 2013

No: 3219

Topic: 1 – Epilepsy

Safety and efficacy of eslicarbazepine acetate (zebinix) in everyday clinical practice using a retrospective multicentre audit

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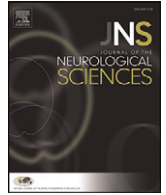
Background/Objective: We report on the safety and efficacy of Eslicarbazepine acetate (ESL) in routine clinical practice.

Method: A retrospective multicentre audit of outcomes following treatment with ESL for localisation-related epilepsy across 7 UK sites (2009–2013).

Results: 202 patients with median values for age 42.5 (17–83) years; duration of epilepsy 16.5 (0–65) years; 2 (0–4) concomitant AEDs, 12 month (2 days–53 months) duration of ESL treatment and 0–12 (64% \geq 2) previous AED exposures. ESL dosage ranged from 600 to 1600 mg/day. Baseline seizure types comprised secondarily generalised tonic-clonic seizures (78.2%), complex partial seizures (74.3%) and simple partial seizures (23.8%). Psychiatric comorbidity was reported in 30% of patients, mainly depression and anxiety. 105 patients (52%) experienced \geq 50% seizure frequency reduction. 40 subjects (19.8%) became seizure free, of whom 26 (65%) had 0–1 previous AED exposures. ESL was discontinued in 70 patients (34.7%) for reasons related to tolerability ($n = 43$), efficacy ($n = 7$), both ($n = 4$) or other ($n = 15$). Adverse events (AEs) were fatigue (18.8%), dizziness (10%) and disturbance in attention/concentration (9%); all were observed with AED polytherapy. Psychiatric and behavioural AEs ($n = 6$, 3.0%), included suicidal ideation ($n = 1$), and led to ESL withdrawal in 2 patients (1.0%). Hyponatraemia was reported ($n = 14$, 6.9%) and led to discontinuation in 4 patients (2.0%).

Conclusions: ESL has comparable efficacy to other AEDs for partial-onset seizures with or without secondary generalisation. Discontinuation due to tolerability was related to AED polytherapy. Reported AEs were consistent with ESL's known safety profile. The benign neuropsychiatric profile with once-daily dosing may convey some advantage in AED selection.

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Topic: 2 – Movement Disorders

Abstract – WCN 2013

No: 3195

Topic: 2 – Movement Disorders

Effect of HU-308 in mice models of Parkinson's disease

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Introduction: Parkinson's disease (PD) is characterized by the progressive loss of nigrostriatal dopamine neurons. Current pharmacotherapies fail to prevent or slow down the disease progression. Although the etiology of PD is presently unknown, major pathogenic processes, which trigger the progressive loss of nigral dopaminergic neurons, are oxidative stress, mitochondrial dysfunction, and inflammatory stimuli. There is less evidence that CB2 receptor agonists can have protective role in brain in response to neuronal damage during PD. In this study we investigated effect of CB2 receptor agonist (HU-308) in 1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine (MPTP) induced PD model in male mice to assess the neuroprotective role of HU-308 in PD progression.

Materials and methods: Male NMRI mice were divided to different groups. The first group received MPTP as control group. The second group was injected with MPTP and then with HU-308 to assess preservation of dopamine function in brain. In the third group, after pretreatment with HU-308, MPTP was injected to assay neuroprotection effect of HU-308.

Result: Behavioral assessment showed significant improvement in the pretreatment group with HU-308. Also Neurochemical analysis that was used to assess dopamine metabolites and immunohistochemistry studies after brain resection showed significant rise in dopamine metabolites, CB2 receptors up-regulation and survival of dopaminergic neurons in the first and second groups as compared with controls in mice's brain.

Conclusion: Our data indicate that CB2 receptors agonist protects against MPTP-induced nigrostriatal degeneration and suggest that CB2 receptors represent a new therapeutic target to slow the degenerative process occurring in PD.

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Abstract – WCN 2013

No: 3197

Topic: 2 – Movement Disorders

Social and medical dilemma of treating l-dopa induced dyskinesia in patients suffering from the PINK1 juvenile onset Parkinson disease

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Background: The PINK1 type of young onset Parkinson disease is characterized by rigidity, bradykinesia & rest tremor, and is very difficult to differentiate from idiopathic Parkinson disease. It may present with lower limb dystonia. Onset is between 30–40 years of age. It is autosomal recessive. The gene encoding protein PINK1 is the only gene where mutation occurs. l-dopa induced dyskinesia can be treated by lowering l-dopa dose and by deep brain stimulation (DBS).

Objective: The dilemma of trying to find a prompt treatment for l-dopa induced dyskinesia in PINK1 juvenile onset Parkinson disease in a poor family which is the second largest family in the Arabian countries.

Methodology and results: We are reporting l-dopa induced dyskinesia in 5 members out of 8 in 2 generations of high rate of consanguinity suffering from a proven PINK1 juvenile onset Parkinson disease in Sudan. The problem we have faced is that the smallest dose of l-dopa required to treat the crippling symptoms of rigidity and lower limb dystonia of those patients usually results in disabling dyskinesia. Deep brain stimulation (DBS) is not available in Sudan.

Conclusion: We've been very successful in diagnosing genetically juvenile onset PINK1-gene Parkinson disease in Sudan. Although we are dealing with the second largest family in the Arab world yet our poor patients cannot afford DBS treatment, a big social question for WFN.

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Abstract – WCN 2013

No: 3208

Topic: 2 – Movement Disorders

Risk of dyskinesia in Parkinson's disease patients who already have developed wearing-off: A secondary analysis of STRIDE-PD study

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Background: Levodopa/carbidopa/entacapone (LCE) increased the risk of dyskinesia compared to levodopa/carbidopa (LC) in the STRIDE-PD study. The risk of developing dyskinesia in Parkinson's disease (PD) patients after onset of wearing-off is not known.

Objective: To analyse the risk of dyskinesia in patients who have developed wearing-off in the STRIDE-PD study.

Materials and methods: 745 PD patients were randomized to LC or LCE in a 134–208-week double-blind trial. Subset of patients who developed wearing-off before dyskinesia was used for present analysis. Time of onset of wearing-off was used as baseline. Patients were divided into 4 dose groups based on levodopa dose at the time of onset of dyskinesia (or end of study if no dyskinesia); <400 mg, 400 mg, >400–600 mg, and >600 mg. The relationship between levodopa dose and time to dyskinesia was analysed using log-rank test. Hazard ratios (HRs) between LC and LCE were calculated using Cox proportion hazards model.

Results: 244 patients (105 (28%) and 139 (37%) on LCE and LC, respectively) developed wearing-off before dyskinesia. Median time from wearing-off to dyskinesia was 75 weeks among the 244 patients. Time from onset of wearing-off to dyskinesia was correlated with levodopa dose groups (Log-rank trend test $p = 0.003$), patients with high doses having higher risk. However, there was no difference in time to dyskinesia between patients receiving LCE and LC (HR = 1.11, $p = 0.618$).

Conclusion: Time from onset of wearing-off to dyskinesia was correlated with daily levodopa dose. However, when patients have already developed wearing-off, there was no difference between LCE and LC in time to dyskinesia.

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Abstract – WCN 2013

No: 3126

Topic: 2 – Movement Disorders

The effect of rTMS on the executive function and bradykinesia in Parkinson's disease

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rTMS can effect more than one symptom of PD in the same time. The motor symptoms of PD can be influenced by both high and low frequency stimulation. However, the effect on the executive function is not well known, which parameters are optimal.

The goal of the study was to examine the effect of rTMS (5 Hz + 1 Hz) on the executive function and bradykinesia. The rTMS was applied over the dorsolateralprefrontal (DLPF) areas for 7 days. We tested patients (20 PD and 20 control) with a Trail Making Test, double-test, and UPDRS before treatment and one month after treatment. The executive function was measured by the Trail Making Test, which showed a slight correlation with the results of double-test ($p = 0.30$) in controls. The patients with PD improved after one month in the total and motor score of UPDRS ($p < 0.05$) and they presented improvement in Trail Making Test and double-test. It seems the stimulation of the DLPF areas by rTMS may improve the executive function parallel with the motor function.

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Abstract – WCN 2013

No: 232

Topic: 2 – Movement Disorders

Safety issues of deep brain stimulation therapy for movement disorders: An analysis of a 10-year experience

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Background: In recent years, DBS became an approved effective treatment for movement disorders, foremost dystonia, Parkinson's

disease (PD), and tremor. DBS commonly presents as quite safe compared to lesioning procedures. However, DBS could be related to different, even life-threatening, complications, limiting usefulness of the therapy.

Objective: To assess the safety of DBS therapy in patients with movement disorders.

Methods: Until 2013, 255 patients were operated for continuous high-frequency DBS – 123 with primary or secondary dystonia, 129 with PD, and 3 with tremor. Age at surgery ranged from 9 to 72 years. Comprehensive analysis of related complications was performed.

Results: In all patients, 579 electrodes were implanted – 279 in globus pallidus internus, 226 in subthalamic nucleus, and 74 in ventrolateral nucleus of thalamus. There were 2 cases of intracranial hemorrhages; one patient remained with a permanent neurological deficit. In two cases, intraoperative seizures occurred. Among technical complications, electrode or connector fractures (7 cases), migration of pulse generator (6), and postsurgical infections (9) were observed. In 38 patients, correction of electrode position was required, and in 5 cases additional electrodes were necessary for optimizing the clinical effect. The most frequent adverse effect of DBS was dysarthria (11%, 28 patients). In 11 cases, the DBS system was permanently explanted due to different reasons. Several correlations were traced.

Conclusion: DBS therapy appears to be a relatively safe procedure for the treatment of movement disorders. Overall level of permanent operation-related disability is low, though neurologist inevitably confronts with unpredictable hardware-related, as well as stimulation-related complications.

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Abstract – WCN 2013

No: 2566

Topic: 2 – Movement Disorders

Iatrogenic parkinsonism: The role of flunarizine and cinnarizine

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Background: Flunarizine and cinnarizine are often associated with parkinsonian symptoms and many patients may be misdiagnosed with idiopathic Parkinson's Disease (PD). Clinical pattern and natural course after drug withdrawal need further clarification.

Objectives:

1. Compare demographic and motor features of patients with flunarizine/cinnarizine parkinsonism-induced (FC-PD) and PD at presentation;
2. Evaluate FC-PD patients outcome.

Methods: This was a retrospective study, based on clinical file review. Patients with parkinsonian signs who were on flunarizine/cinnarizine at first visit were compared to patients with idiopathic Parkinson's disease, regarding: symptom duration, age and parkinsonian features at presentation; gender; age of onset; motor outcome and dopamine dose increase (final dopamine equivalent dosis/follow-up time).

Results: We selected 31 FC-PD and 83 PD patients. FC-PD group presented with a significantly higher prevalence of bilateral tremor (vs. unilateral). FC was suspended in all patients. Twelve patients improved spontaneously (9 became asymptomatic). Fourteen patients were started on dopaminergic treatment – all improved (6 became asymptomatic). FC patients who did not improve spontaneously did not differ significantly from those who did, and from PD patients. FC patients who improved spontaneously presented with a significantly lower prevalence of unilateral tremor, bradykinesia and rigidity,

compared to PD patients. Dopamine dose increase was significantly lower in FC patients.

Discussion: FC differed from PD on the higher prevalence of bilateral tremor. FC-PD patient outcome was globally favourable (spontaneous improvement or improvement with lower dopamine doses) but heterogeneous. Spontaneously improved FC patients showed a diverging parkinsonian symptom profile, suggesting a different physiopathology.

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Abstract – WCN 2013

No: 3109

Topic: 2 – Movement Disorders

Early postoperative MRI is more sensitive than CT to detect intracerebral hemorrhage in deep brain stimulation surgery

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Introduction: Early postoperative magnetic resonance imaging (MRI) was performed to assess the incidence of intracerebral hemorrhage as a complication in stereotactic functional neurosurgery.

Methods: 25 patients undergoing deep brain stimulation (DBS) for Parkinson's disease, essential tremor, or dystonia were included in the study. Target structures were the subthalamic nucleus in patients with Parkinson's disease, the globus pallidus internus in dystonia patients, and the nucleus ventralis intermedius thalami in tremor patients. Multiple tract microelectrode recording and test stimulations were performed before implantation of DBS electrodes. Electrodes were implanted bilaterally in 24 patients. Unilateral implantation of DBS electrodes into the globus pallidus internus and the nucleus ventralis intermedius thalami was performed in one patient with hemidystonia and action tremor. All patients underwent postoperative computed tomography (CT) on postoperative day one. Early postoperative MRI was performed using T1 weighted, T2 weighted, and diffusion weighted imaging (DWI) on postoperative day 2 to 30.

Results: In all patients postoperative CT was negative for signs of intracerebral hemorrhage. Early postoperative MRI showed intracerebral hemorrhage along four of fifty implanted electrodes. The patients with intracerebral hemorrhage did not show any neurological deficit or conspicuity in their postoperative clinical course.

Conclusion: Early postoperative MRI is highly sensitive to intracerebral hemorrhage. Due to metal artifacts CT cannot detect small hemorrhages in the vicinity of the implanted electrodes. Clinically silent perioperative intracerebral hemorrhage may presently be an underreported complication of DBS surgery, because postoperative MRI is not routinely used in the postoperative follow-up of DBS surgery.

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Abstract – WCN 2013

No: 3067

Topic: 2 – Movement Disorders

Preliminary evaluation of Sensorfoot V1 and Senshand V1 in assessing motor skills performance of Parkinson's disease patients

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Background: Up to 10 millions of people are living worldwide with Parkinson's Disease (PD). To improve diagnosis and treatment, two ICT

wearable devices have been developed and tested to achieve an objective assessment of motor performance.

Objective: To evaluate the tasks of the Movement Disorder Society- Unified Parkinson's Disease Rating Scale (MDS-UPDRS) through biomechanical exercises objectively measured by means of a wireless and wearable device for motion recognition, composed by SensorFoot V1 (placed on the foot) and SensHand V1 (fixed on fingers and forearm).

Patients and methods: Ten PD patients (9 men, 1 woman; 67.9 ± 6.9 years; mean disease duration: 4.1 ± 2.8 years; mean Hoehn and Yahr stage: 2.1 ± 1.04; mean MDS-UPDRS score (III): 16.5 ± 6.6; mean l-dopa equivalent daily dose: 513 ± 287 mg) and 5 controls (2 men, 3 women, 64.8 ± 9.1 years) were included. They underwent tremor and motion analysis through exercises for both upper limbs (forearm pronation-supination, hand opening/closing, finger tapping) and lower limbs (foot tapping, rotation and gait). Biomechanical features (frequency, velocity, and angle) have been extracted for each exercise and compared to clinical assessment based on MDS-UPDRS.

Results: Generally, healthy people showed significantly ($p < 0.05$) more constant repetitive movements and better performances in comparison to patients; good correlations (Pearson's index $k > 0.7$) with PD patients' clinical scores were found.

Conclusion: The technological system has shown a clear potential to positively support neurologist in early diagnosis and clinical assessment of PD patients overtime.

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Abstract – WCN 2013

No: 3076

Topic: 2 – Movement Disorders

Quantitative gait analysis in Parkinson's disease using MS Kinect: Entropy and fractal dimension

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Background: Gait disturbances form a frequent reason of disability and impairment in patients with Parkinson's disease (PD). Gait hyperkinesia is the first to appear and it is responsible for the decrease in velocity. Stride length, gait variability, walking velocity and fractal scaling are impaired. Microsoft Kinect is a peripheral for personal computers that, via an integrated depth camera, enables skeleton-detection and people tracking in real time.

Objective: Non-linear gait parameters in Parkinson's disease (PD) were compared with those of healthy age-matched control subjects.

Patients and methods: Patients with PD ($n = 16$) and healthy age-matched controls ($n = 16$). Permutation entropy and the Sevcik's fractal dimension were estimated using the Matlab and Simulink computational environments. Data were collected using MS Kinect for PC in the skeleton mode.

Results: Compared to healthy controls, non-linear gait parameters of PD patients showed a significant increase in fractal dimensions and entropy.

Conclusion: In addition to time-distance, kinematic and kinetic gait variables, abnormal fractal dimension and entropy play an important role in characterizing gait in PD patients. Hence, these parameters could be used to quantify gait impairment in PD patients and document the effects of gait disorder therapies in PD.

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Abstract – WCN 2013**No: 3089****Topic: 2 – Movement Disorders****Electric cardioversion in a patient with deep brain stimulation**

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Deep brain stimulation (DBS) is an established treatment for motor complications in Parkinson's disease (PD). Many patients with DBS suffer from cardiac comorbidity. Concurrent use of DBS and cardiac pacemakers is reasonably safe when minimizing electromagnetic interference. Electric cardioversion and defibrillation probably confer a higher risk of electromagnetic interference with DBS systems. We report on the effects of external electric cardioversion in a PD patient with bilateral subthalamic DBS.

This 59 year old male patient had a 12-years history of PD. In 2007 he underwent DBS surgery with bilateral implantation of quadripolar electrodes (Medtronic 3389) into the subthalamic nucleus and placement of the neurostimulator (Kinetra) into the left abdominal region. In January 2013 the patient underwent aortic valve replacement with the DBS device turned off before and switched on immediately after surgery. On the second postoperative day the patient developed atrial fibrillation which was successfully treated with a bolus of amiodarone and electric cardioversion by a physician unaware of ongoing neurostimulation. Subsequent to progressive worsening of parkinsonism the DBS device was interrogated. The neurostimulator output was found switched off with amplitude set at 0 V and polarity, frequency and pulse width altered. Electrode impedances were unchanged. The neurostimulator could be switched on and reprogrammed. The patient recovered without neurological sequelae.

Although defibrillation or cardioversion may damage or cause reprogramming of a neurostimulator, these procedures have to be applied in emergency situations. According to the manufacturer the affected neurostimulator contains filtered feedthroughs and diodes which probably protected the device from damage.

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Abstract – WCN 2013**No: 1390****Topic: 2 – Movement Disorders****Use of a respiration sensor to record blink reflex to glabellar tapping in Parkinson disease patients with Myerson's sign**

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Background: Myerson's sign (also known as the "glabellar reflex") is a universally accepted sign of Parkinson's disease (PD), but its mechanism is still poorly understood.

Objective: In order to investigate the blink reflex in Myerson's sign-positive and -negative subjects, we used conventional finger tapping of the glabella in conjunction with surface electromyograms (EMGs) of the orbicularis oculi muscle, which were recorded for quantitative electrophysiological assessment.

Patients and methods: The subjects consisted of 9 PD patients with Myerson's sign (PD/M+), 7 PD patients without Myerson's sign (PD/M-), and 9 normal controls (NC). A quantitative electrophysiological assessment was conducted on the characteristics of Myerson's sign in Parkinson's disease. Surface EMGs of the orbicularis oculi were triggered by a respiration sensor, which detected tapping pressure on the glabella,

and were recorded continuously during tapping. Changes in surface EMGs over time with 20 repeated stimuli at a frequency of 2 Hz were compared among the PD/M+, PD/M-, and NC groups. We measured the mean latency and mean R1 and R2 areas in the PD/M+ Group.

Results: In the PD/M+ Group, reflex blinking caused by glabellar tapping tended to continue, and both the R1 and R2 components tended to continue on the surface EMGs.

Conclusion: The present study confirms that glabellar tapping is a useful diagnostic technique for PD and seems to support previous reports that interneuronal pathways in the brainstem are involved in reflex blinking.

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Abstract – WCN 2013**No: 2984****Topic: 2 – Movement Disorders****Frequency of buccopalpebral reflex according to the stage of Parkinson disease**

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Background: In our observation, eye blinking together with shrinking of lips was observed by the percussion of the upper lip in neurodegenerative diseases such as Parkinson's disease. This reflex is named as "Bucco-palpebral reflex" (BPR) by us.

Objectives: To investigate frequency of BPR according to the stage of Parkinson Disease and to compare findings of age-sex matched control group.

Patients and methods: 115 patients with Parkinson Disease and 107 control subjects were investigated prospectively. Demographic data and information about Parkinson disease were collected for each patient. Unified Parkinson's Disease Rating Scale (UPDRS) was used for staging of Parkinson's disease and BPR was evaluated.

Results: The mean age in patients with Parkinson's disease was 69.8 ± 8.6. It was 66.8 ± 8.4 in control subjects. BPR frequency of the control group was 3.7%, while those with parkinson's disease were 13.9%. This difference was statistically significant (p < 0.05). In the analysis of UPDRS sub-groups, activities of daily living score were 13.3 ± 7.1 in reflex positive group and 9.6 ± 7.7 in reflex negative group. According to the motor score evaluation of BPR positive and negative patients, the motor score was 26.2 ± 13.8 and 19.2 ± 13.0 respectively. These differences were statistically significant (p < 0.05).

Conclusion: BPR mainly observed in neurodegenerative disease such as Parkinson Disease may be a result of the removal of cortical inhibition; therefore it should be primitive reflex. Since UPDRS scores were higher in BPR positive groups, BPR is frequently seen in more severely affected patients.

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Abstract – WCN 2013**No: 3009****Topic: 2 – Movement Disorders****Spinocerebellar ataxia type 36 (SCA36): Expanding the Genotype and Phenotype**

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Spinocerebellar ataxia type 36 (SCA36) is an autosomal dominant neurodegenerative disorder caused by a hexanucleotide GGCCTG repeat expansion in intron 1 of the NOP56 gene. SCA36 has been reported from some regions of Japan and Spain, but its occurrence in different populations is unknown. We here performed a large-scale trans-continental assessment of this repeat in a cohort of 676 families excluded for previously known SCAs and originating mostly from France, Germany and Japan. We found SCA36 expansions in nine French and five Japanese index patients, as well as one each from China, Portugal and Spain, thus accounting for 1.9% of all French, 1.5% of Japanese and 0% of German SCAs. Besides long expansions ranging between approximately 800 and 2000 repeats, we found short GGCCTG expansions ranging between 23 and 30 repeats. The expansion was highly unstable as two individuals, one with a long and the other with a short expansion, existed in the same family. Clinically, the cardinal feature was slowly progressive cerebellar ataxia, frequently accompanied by hearing and cognitive impairments, tremor, ptosis, reduced vibration sense and sensory axonal neuropathy with the age at onset ranging between 39 and 65 years. Neuropathology in one asymptomatic French individual disclosed mild losses of Purkinje cells and hypoglossal neurons, suggesting that these neurons are consistently affected from an early stage. We conclude that SCA36, caused by a highly unstable GGCCTG repeat expansion in NOP56, is a rare but global disease with various extra-cerebellar symptoms.

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Abstract — WCN 2013

No: 2947

Topic: 2 — Movement Disorders

Safinamide is effective as add-on treatment in both early and Advanced PD

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Background: The mechanism of action of safinamide, a new chemical entity, combines both dopaminergic and non-dopaminergic activity in producing benefit in patients with Parkinson's disease (PD).

Objective: The safety and efficacy of safinamide have been evaluated in patients with Early PD (MOTION trial) as add-on to dopamine-agonist monotherapy, and Advanced PD fluctuators (SETTLE trial) as add-on to l-dopa and other PD medications.

Patients and methods: MOTION study, a 24-week randomized, placebo-controlled trial evaluating efficacy and safety of 50 mg/day and 100 mg/day of safinamide as add-on to a single dopamine-agonist at a stable dose, enrolled 679 patients, within 5 years of diagnosis, who never received l-dopa. SETTLE study, a 24-week randomized, placebo-controlled trial evaluating efficacy and safety of 50–100 mg/day of safinamide as add-on to l-dopa and other PD medications, enrolled 549 patients, with a PD diagnosis of more than 5 years, who presented motor fluctuations. The results for the MOTION study indicated that safinamide of 100 mg/day significantly improves motor symptoms (UPDRS III, $p = 0.040$), and quality of life (PDQ-39 and EQ5D) compared with placebo in patients taking a single dopamine-agonist ($N = 666$). The results for the SETTLE study indicated significant superiority of safinamide over placebo for ON time without troublesome dyskinesia ($p < 0.001$), OFF time ($p < 0.001$), UPDRS III, PDQ-39, EQ5D, CGI, PGI, and OFF time post first morning dose of l-dopa.

Conclusion: This new results from MOTION and SETTLE studies confirm previously available results from studies in Early PD patients,

and Advanced PD patients with motor fluctuations indicating that safinamide is efficacious as add-on treatment at any stage of the disease.

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Abstract — WCN 2013

No: 1587

Topic: 2 — Movement Disorders

Reduced penetrance alleles of TBP (SCA17) in Indian ataxia cohort: Homozygous mutation and ambiguity of 41–44 CAG/CAA for pathogenic association

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Background: CAG/CAA expansion mutation in TBP causes spinocerebellar-ataxia 17 (SCA17), characterized by wide spectrum of neurological impairments including cerebellar-ataxia, psycho-cognitive disturbance, dementia and extrapyramidal signs. The repeats > 48CAG/CAA produce typical SCA17 manifestations whereas 41–44(CAG/CAA) repeats are reduced penetrant alleles and have been documented both in the cases and healthy-controls.

Objective: To analyze the frequency of SCA17 in Indian ataxia cohort. **Subjects and methods:** 565 unrelated genetically-uncharacterized spinocerebellar ataxias (UC-SCAs), 355 healthy controls and 228 cases of known SCA-subtypes (Kn-SCA) were analyzed for SCA17-CAG/CAA(TBP) repeat-length. Detailed neurological and psychiatric evaluation was conducted in selected cases with abnormal repeat-length.

Results: We identified 39 UC-SCAs harboring CAG/CAA in the range of 41–46 including a homozygous TBP-CAG/CAA(45/46) expansion mutation in a 43.5 year old female with mild gait-ataxia and dysarthria, prominent blepharospasm, cervical-dystonia, limb-rigidity, dystonia, bilateral nystagmus, pyramidal, and multiple psychiatric impairment with 1.5 years of disease duration. In another case of sporadic ataxia, a 60 year old male with CAG/CAA-37/44 had only moderate degree of cerebellar-ataxia and mild psychiatric impairment. Notably we observed repeat length in the range of 41–44 with 1.6% frequency in pool of controls and Kn-SCA samples whereas UC-SCA category had higher frequency of 3.45% (P value = 0.005).

Conclusion: We report fifth case of homozygous-(TBP-CAG/CAA)_{45/46} mutation in SCA17 world-over. In our patient-population pathogenic significance of 41–44 is somewhat dilemmatic, although previous studies reported broader disease onset age (19–69 years) in this range of repeats and are associated with variable phenotypes. Further follow-up studies may confirm the pathogenic significance of 41–44 repeats in our study population.

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Abstract — WCN 2013

No: 3021

Topic: 2 — Movement Disorders

Apomorphine for Morning Akinesia Trial (Am Impact)

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Background: Morning akinesia, with unreliable or delayed onset of effect of first morning l-dopa dose is a frequent manifestation of motor fluctuations in Parkinson's disease (PD).

Objective: The primary objective of this study is to assess the effect of subcutaneous bolus apomorphine HCl injection in patients with morning akinesia with lack of predictable effect of first morning l-dopa dosing. A secondary objective is to assess apomorphine's gastric motility effect. The primary outcome compares patient self-reported, diary-recorded Time to On (TTO) following first l-dopa morning dose compared to TTO when using apomorphine each morning. The study will additionally provide data on apomorphine's effects on gastroparesis, an often underdiagnosed, non-motor PD symptom.

Methods: This study will enroll approximately 100 PD subjects across 12 sites. Subjects complete a 7-day Baseline Period recording daily TTO following their regular l-dopa morning dose. Following the Baseline Period, subjects start trimethobenzamide antiemetic therapy and begin APOKYN titration. Once optimal dose is identified, subjects self-inject apomorphine at regularly scheduled l-dopa morning dose time (first l-dopa morning dose is delayed by 40 min) during a 7-day Treatment Period and record TTO following apomorphine injection. Gastric motility sub-study subjects participate in gastric emptying imaging following a standardized meal during the Baseline Period and during the Treatment Period.

Results: Interim analysis of the primary TTO change from baseline endpoint, key secondary endpoints and gastric emptying time results from the sub-study participants will be available in October.

Conclusion: Apomorphine bolus injection may offer an effective treatment for morning akinesia and underlying gastroparesis.

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Abstract – WCN 2013

No: 3032

Topic: 2 – Movement Disorders

Spinocerebellar ataxia 7 associated with myopathy of tubular aggregates

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Introduction: SCA 7 is a spinocerebellar ataxia whose cardinal features are progressive cerebellar ataxia and macular degeneration. Pyramidal signs, ophthalmoplegia and parkinsonism might also be present, however, the association with myopathy is not described.

Case report: A 32 year-old Mozambican male presented with an 8 year history of stable decreased visual acuity associated with progressive dysarthria and appendicular and gait incoordination for the last 2 years. Son of non-consanguineous parents, his past medical and family histories were irrelevant even though the current health status of his father is unknown. Head titubation, slow and interrupted saccades at horizontal and vertical gaze, ocular apraxia, moderate dysarthro-hypophonia, severe, appendicular and gait ataxia, dysdiadochokinesia, bilateral positive Holmes sign, negative Romberg test, pathologic exaggerated reflexes without other pyramidal signs nor clinical evidence of myopathy were observed. Ophthalmologic examination showed bilateral maculopathy with bilateral atrophy of macular neuroretina. Brain MRI documented pancerebellar and brainstem atrophy while muscle biopsy of deltoid muscle revealed myopathy of tubular aggregates. The genetic test for SCA 7 was positive with 49 repeats.

Conclusion: We describe the first case of SCA 7 associated with myopathy of tubular aggregates, expanding the classic phenotype of SCA 7. The present case strengthens the importance of considering SCAs in differential diagnosis of sporadic ataxias with adult onset.

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Abstract – WCN 2013

No: 3030

Topic: 2 – Movement Disorders

Reversible Pisa Syndrome in a patient with Parkinson's disease on rasagiline therapy

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Pisa Syndrome (PS), reported also in patients with Parkinson's disease (PD), is clinically defined as the sustained lateral bending of the trunk, worsened by a prolonged sitting position or by walking. Its pathophysiology is still unexplained.

We report a case of reversible PS in a patient with PD improved after rasagiline withdrawal.

A 73-year-old woman with idiopathic PD, diagnosed 6 years before (H&Y stage = 1.5), started to complain of a trunk inclination towards the left that completely relieved by passive mobilization. This was compatible with the diagnosis of PS. This symptom started approximately one year before, when rasagiline was started because of mild motor worsening and wearing-off phenomena. At that moment, the patient was also on pramipexole 1.57 mg/day and levodopa 200 mg/day. EMG in the standing position showed a muscular hyperactivity on bilateral paravertebral muscles, prevailing on the leaning side (left). Muscular hyperactivity did not decrease during contralateral flexion, resulting in a co-activation of left and right paravertebral muscles. No myopathic pattern was observed. As PS started approximately at the same time rasagiline was first prescribed, according to previous reports¹, we withdrew the MAO-B inhibitor. An improvement of posture was observed within 4 weeks from withdrawal, confirmed by a normalization of EMG traces.

The recognition of reversibility of PS during the initial stages of its appearance is of considerable clinical importance, as it facilitates the rapid withdrawal or reintroduction of dopaminergic treatment, avoiding an initial veering towards the chronic irreversible variant.

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Abstract – WCN 2013

No: 2961

Topic: 2 – Movement Disorders

Vascular factors contribute to motor and cognitive dysfunction and mortality in Parkinson's disease

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Background: To study the impact of brain vessel pathology on the clinical status and mortality of PD.

Methods: The clinical and neuropsychological data were compared with clinical and MRI signs of vascular impairment and with the ultrasound brain vessel investigations in 57 consecutive patients with PD. In the second part, the baseline data obtained from patients who died (n = 18) during a four-year follow-up period were compared with the data of patients who survived.

Results: There was a significant correlation between clinical, cognitive status, mortality and intimomedial thickness (an indicator of large vessel impairment). Cognitive status was significantly related also to the pulsatility index and mortality to the resistance index (indicators of small vessel impairment).

The MRI data displayed a more severe vascular impairment in the deceased patients. The sum score of white matter hyperintensities was

significantly higher among decedents. A cluster analysis displayed two clusters that differed in the two parameters, i.e. in the age and in the sum score.

Conclusions: Our studies provide evidence that co-morbid atherosclerosis and otherwise subclinical impairment of brain vessels may contribute to motor and cognitive dysfunction and mortality in Parkinson's disease. The vascular pathology may act in association with other co-morbidities on the terrain of the progressive neurodegenerative pathology. This is a reason to be more attentive to the therapy of vascular pathology in PD patients.

Rektor I et al. *Parkinsonism Relat Disord* 2009; 15 (1):24-19.

Rektor I et al. *Mov Disord* 2012; 27(9):1169-72.

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Abstract – WCN 2013

No: 2937

Topic: 2 – Movement Disorders

Chlorophyllin: A possible new therapeutic agent for increasing frataxin levels in Friedreich's ataxia patients

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FRDA is an autosomal recessive neurodegenerative disease characterized by multiple symptoms such as progressive spinocerebellar ataxia, sensory nerve affection, diabetes mellitus and life-limiting hypertrophic cardiomyopathy. FRDA is caused by a GAA-trinucleotide expansion in the frataxin gene located on chromosome locus 9q13 which results in a markedly reduced expression of frataxin, a small mitochondrial protein. Decreased frataxin levels cause iron accumulation in mitochondria, oxidative stress and cell damage. Searching for compounds which could possibly influence frataxin expression we found that the chlorophyllins can significantly increase frataxin expression by still an unknown mechanism. Chlorophyllin (CHL) is a water-soluble mixture of salts of the green plant pigment chlorophyll. CHL currently has a wide range of uses, dietary and medicinal, including being a food colouring agent, a health food additive, an accelerant of wound healing and controlling urinary and faecal odours in geriatric patients. In a wide array of in vitro and in vivo assays, CHL has also been shown to exhibit antimutagenic, antigenotoxic and anticarcinogenic activities against numerous carcinogens.

We show for the first time that additional to its protective properties chlorophyllin can increase frataxin expression in vitro and in vivo. Our results provide a scientific basis for a clinical pilot trial examining the effectiveness of this agent for the treatment of FRDA patients.

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Abstract – WCN 2013

No: 2896

Topic: 2 – Movement Disorders

Cross-cultural adaptation for use in Brazilian patients and analysis of the psychometric properties of the brief ataxia rating scale

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Background: Assessment of the intensity of ataxia is essential in the clinical setting and research. Ataxia rating scales are commonly used for this purpose. The Brief Ataxia Rating Scale (BARS) has the advantage of being a short instrument.

Objective: Perform the cross-cultural adaptation of BARS for the Portuguese language spoken in Brazil (BARS-B) and test its psychometric properties.

Patients and methods: We performed translation, cross-cultural adaptation and analysis of the psychometric properties of BARS-B in a sample of 68 adults with ataxia and 38 healthy controls. Analyses of psychometric properties included: structural analysis, reliability analysis (internal consistency and reproducibility) and analysis of the validity of the instrument.

Results: Cross-cultural adaptation resulted in an instrument similar to the original. Structural analysis identified that BARS-B is organized in two factors: gait, limbs and speech scores (one factor) and oculomotor score (a second factor). Reliability analysis showed satisfactory internal consistency (Cronbach's alpha = 0.84); satisfactory intra-rater and inter-rater reproducibility (intraclass correlation coefficients = 0.96 for both). Content and face validities: the BARS-B's items contemplated what was intended to be measured. Criterion validity (Pearson's correlation of BARS-B with the scores of International Cooperative Ataxia Rating Scale–ICARS and Scale for the Assessment and Rating of Ataxia–SARA), showed positive and satisfactory correlations ($r = 0.87$ and 0.90 , respectively). Construct validity (Pearson's correlation between BARS-B and functional capacity estimated by Barthel index) was satisfactory ($r = -0.74$). Known-group validity was confirmed by Student's *t* test in 68 patients and 38 controls ($p < 0.001$).

Conclusion: BARS-B is a reliable and valid instrument for the assessment of ataxia.

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Abstract – WCN 2013

No: 2839

Topic: 2 – Movement Disorders

Clinical profiles of motor system of patient with Parkinson's Disease (PD) at tertiary hospital of north India

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Background: PD is a degenerative disease that occurs as a result of the loss of dopamine-producing brain cells. The four primary symptoms of PD are tremor, rigidity, bradykinesia and postural instability. PD usually affects people over the age of 50–60, although there are young-onset cases. Early symptoms of PD are subtle and occur gradually. Parkinson's disease affects movement, producing motor symptoms.

Objective: To know the clinical characteristics of PD patients visiting this tertiary care hospital at north India.

Methods and observations: This was an observational, cross sectional study of patients with Parkinson's disease attending this institute (Indira Gandhi Institute of Medical Sciences, Patna, Bihar, India) either as outpatient or indoor admission.

Results: Total 1141 patients with Parkinson's disease were analyzed. 649 were male and 492 were female. Age at onset—mean of 54.7 years (range = 24–82 years). Disease duration—mean of 3.8 years (range = 6 months–10 years). Place of birth—1059 village and 82 town. Residence (maximum)—801 village and 340 town. Family history—2 cases. Unilateral onset—1141, hands—1138, legs—3, right side—789, and left side—352. First symptom—tremor = 831, bradykinesia = 159, rigidity = 149, and hemiparetic = 2989 patients have bilateral involvement with persistent asymmetry and 152 have still unilateral disease. Mean time of bilateral involvement is 1.6 years (range—11 months–6 years). 1013 were on levodopa, 919 have marked improvement, and 597 have levodopa induced dyskinesia.

Conclusion: This is the first study of this kind performed in this region. Parkinson's disease affects both sexes, mainly males. Its main presentation is unilateral tremor of hand, then bradykinesia followed by rigidity. Right hand is more affected than left. It can rarely start from leg.

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Abstract – WCN 2013**No: 2848****Topic: 2 – Movement Disorders****Postencephalitic parkinsonism – case report**C. Falup-Pecurariu^{a,b}, M. Moarcas^a, M. Bustan^a, R. Chiperi^a, R. Alexandru^b.^aDepartment of Neurology, County Emergency University Hospital, Romania;^bFaculty of Medicine, Transilvania University, Brasov, Romania

Background: Infection is a rare cause of parkinsonism. Postencephalitic parkinsonism is encountered very rare nowadays and may appear as an early or late complication of encephalitis.

Objectives: The aim is to present a case report of a 45 year old woman with postencephalitic parkinsonism.

Method: The patient was diagnosed at the age of 26 years with midbrain encephalitis, that had an acute onset with gait disturbance, loss of balance, somnolence, vomiting, oculogyric crises and inflammatory CSF. After 4 weeks there was an onset of bilateral rest hand tremor and bradykinesia. Neurological exam showed bilateral parkinsonism. The lab exams that were performed and CT-scan were repeatedly normal. There was an initiated antiparkinsonian treatment with Trihexyphenidylum, followed by Amantadine with mild improvement of tremor.

The evolution over several years was marked by cognitive decline, the appearance of dysarthria. During evolution there were trials with Ropirinolum, Pramipexolum, and Rasagiline with no improvement. Levodopa gave only a mild improvement of bradykinesia. After 19 years of evolution the bradykinesia is quite stable without significant worsening. The patient is independent in most of the daily activities.

Conclusion: Postencephalitic parkinsonism in this case report had low or no response to antiparkinsonian treatment. Long-term follow-up shows no significant motor worsening.

doi:10.1016/j.jns.2013.07.527

Abstract – WCN 2013**No: 2801****Topic: 2 – Movement Disorders****Helicobacter pylori and small intestinal bacterial overgrowth in Parkinson's disease: Prevalence and clinical significance**A.H. Tan^a, S. Mahadeva^a, A.M. Thalha^a, C.K. Kiew^a, C.M. Yeat^a, S.W. Ng^a, S.P. Ang^a, S.K. Chow^a, K.M. Than^a, N.S. Hanafi^a, N.M. Ibrahim^b, P.R. Gibson^c, S.H. Fox^d, S.Y. Lim^a. ^aUniversity of Malaya, Kuala Lumpur, Malaysia; ^bUniversity Kebangsaan Malaysia, Kuala Lumpur, Malaysia; ^cMonash University, Victoria, VIC, Australia; ^dUniversity of Toronto, Toronto, ON, Canada

Background: A high prevalence of *Helicobacter pylori* infection (HPI) and small intestinal bacterial overgrowth (SIBO) has been reported in Parkinson's disease (PD), but with unclear clinical significance.

Objective: To study the prevalence and impact of gut infections on gastrointestinal (GI) symptoms and motor function in PD patients.

Patients and methods: 74 patients (with and without motor fluctuations) underwent 13C urea and lactulose-hydrogen breath tests. Upper and lower GI symptoms were evaluated using the Leeds Dyspepsia Questionnaire and the Global Symptomatic Score. On-medication motor function was assessed using the Unified Parkinson's Disease Rating Scale (UPDRS), and timed tests of gait and upper limb function (14-metre fast walk and Purdue pegboard).

Results: HPI and SIBO were detected in 32% and 21% of patients, respectively. There were no between-group (breath test positive vs. negative) differences in GI symptoms scores. HPI + patients had worse UPDRS scores (total = 61.1 vs. 47.2, $p = 0.004$; part III = 34.4 vs. 27.4, $p = 0.019$) and worse scores on the timed tests (38.1 vs. 27.9 steps, $p = 0.025$; 6.2 vs. 8.2 pins, $p = 0.013$) compared with HP-patients. A similar (non-significant) trend was seen with SIBO.

Conclusion: HPI was detected in one-third of the patients, but we found a lower prevalence of SIBO than what was reported previously. These infections were not associated with GI symptoms. The association between HPI and worse motor function has important clinical implications and should be confirmed with a larger sample size, and by studying the effects of eradication therapy.

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Abstract – WCN 2013**No: 2805****Topic: 2 – Movement Disorders****Spinal cord stimulation is effective to gait disturbances with Parkinson's disease**T. Ichikawa^a, H. Oshima^b, Y. Nishida^a, Y. Fumimura^a. ^aNeurology, Saitama Prefectural Rehabilitation Center, Ageo City, Japan; ^bNeurosurgery, School of Medicine, Nihon University, Tokyo, Japan

Backgrounds: Spinal cord stimulation (SCS) is indicated for pain from various origins. In patients with Parkinson's disease (PD), pain is a common non-motor symptom, which is sometimes severe to be treated with SCS. We studied the effect on gait in PD patients after SCS for pain.

Patients and methods: Six patients with PD (average disease period = 12.2 ± 5.5 years) underwent SCS between August 2012 and March 2013. We use two electrodes with eight stimulation contacts in one procedure. The electrodes were inserted by paramedian approach into the epidural space. We put the electrodes parallel to spinal column and the top of the electrodes was at the level of T6–T8 in five patients and C4 in one patient. After stimulation trial for a week, we placed the implanted pulse generator at the abdomen subcutaneously. Stimulation settings are 60–90 bps in frequency and 100 to 200 μ s in pulse width. Motor function was measured by Movement Disorder Society – Unified Parkinson's Disease Rating Scale (MDS-UPDRS) 1.9 (pain and other sensation) and part III before SCS and also three to five weeks after SCS. **Results:** MDS-UPDRS and part III are improved (before = 44.0 ± 4.4 , after SCS = 29.8 ± 3.0). The improvement is mainly due to Sections 3.7–3.13 which are evaluating gait and lower limb functions (before = 23.8 ± 6.1 , after SCS = 12.7 ± 3.5).

Conclusions: SCS can improve motor symptoms of Parkinson's disease, especially gait and lower limb function.

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Abstract – WCN 2013**No: 2657****Topic: 2 – Movement Disorders****Apoptosis in peripheral blood lymphocytes of patients with LRRK2-associated Parkinson's disease**T. Usenko^{a,b}, A. Yakimovskii^b, A. Emelyanov^{a,b}, S. Pchelina^{a,b}. ^aPetersburg Nuclear Physics Institute, RAS, Russia; ^bPavlov's State Medical University of Saint Petersburg, Saint Petersburg, Russia

LRRK2 mutations are the most frequent cause of familial Parkinson's disease (PD). Although the precise physiological and pathological role of *LRRK2* is unclear, direct link between mutant *LRRK2* and apoptosis has been suggested.

The aim of our work was to estimate apoptosis rate, *FAS* and *BCL-2* mRNA levels and the caspase-8 and caspase-9 activation in peripheral blood lymphocytes (PBLs) of patients with *LRRK2*-associated PD.

Apoptosis of PBLs was estimated using flow cytometric analysis stained by annexin V-FITC-PI; *FAS* and *BCL-2* mRNA levels were estimated by RT-PCR with TagMan probes (internal control – the level of G protein

(*GNB2L1*) mRNA); the caspase-8 and caspase-9 activations were examined by western blot (internal standard – β -actin) after 1 h, 24 h, and 48 h of incubation (37 °C, 5%CO₂) in PBLs of four patients with *LRRK2*-associated PD (mean age = 65 ± 10) and nine controls (mean age = 61 ± 7).

Apoptosis rate was higher in patients with *LRRK2*-associated PD (n = 4) compared to controls (n = 9) at 24 h and 48 h (p < 0.02, p < 0.03, respectively).

FAS mRNA level was higher in patients with *LRRK2*-associated PD (n = 4) compared to controls (n = 9) at 1 h and 24 h (p < 0.03, p < 0.05, respectively). Difference in *BCL-2* mRNA level between both groups was not statistically significant.

The caspase-8 activation was observed in all investigated groups and the caspase-9 activation was detected in all patients with *LRRK2*-associated PD (n = 2) and in one from all controls (n = 4) after 24 h of incubation.

Our results suggest that *LRRK2* mutations may lead to the activation of apoptosis pathway with predominant activation caspase-9 and increased *FAS* expression in patients with *LRRK2*-associated PD.

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Abstract – WCN 2013

No: 2778

Topic: 2 – Movement Disorders

Functional evaluation of central cholinergic circuits in patients with REM sleep behavior disorder and Parkinson's disease: A TMS study

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Background: Central cholinergic dysfunction has been reported in patients with Parkinson's disease (PD) and hallucinations by evaluating short latency afferent inhibition (SAI), a transcranial magnetic stimulation protocol which gives the possibility to test an inhibitory cholinergic circuit in the human brain. REM sleep behavior disorder (RBD) was also found to be associated with cognitive impairment in PD patients.

Objective: The objective of the study was to assess the cholinergic function, as measured by SAI, in PD patients with RBD (PD-RBD) and PD patients without RBD (PD-nRBD).

Materials and methods: We applied the SAI technique in 10 PD-RBD patients, in 13 PD-nRBD patients and in 15 age-matched normal controls. All PD patients and control subjects also underwent a comprehensive battery of neuropsychological tests.

Results: Mean SAI was significantly reduced in PD-RBD patients when compared with PD-nRBD patients and controls. Neuropsychological examination showed mild cognitive impairment in 9 out of the 10 PD-RBD patients, and in 5 out of the 13 PD-nRBD. SAI values correlated positively with neuropsychological tests measuring episodic verbal memory, executive functions, visuoconstructional and visuo-perceptual abilities.

Conclusion: Similar to that previously reported in the idiopathic form of RBD, SAI abnormalities suggest a cholinergic dysfunction in PD patients who develop cognitive impairment, and present findings indicate that RBD is an important determinant of MCI in PD.

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Abstract – WCN 2013

No: 2759

Topic: 2 – Movement Disorders

Mesocortical dopaminergic dysfunction in Parkinson's disease-depression: Evidence from a ¹²³I-FP-CIT SPECT investigation

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Depressive symptoms frequently occur in patients with Parkinson's disease (PD) but their pathogenesis is not fully understood although neurochemical deficits and structural changes have been suggested. Both striatal and extra-striatal dopaminergic deficits could play a major role although results on striatal dopamine transporter (DAT) density are conflicting.

To assess without an a priori hypothesis the extrastriatal DAT availability by using SPM on ¹²³I-FP-CIT SPECT images in a population of PD patients and depression (PD-d), and a population of PD patients without depression (PD-nd).

20 PD-d (mean age = 63.7 years ± 11.0; UPDRS III at SPECT scan = 17.6 ± 7.4); 35 PD-nd (mean age = 67.6 years ± 9.0; UPDRS III at SPECT scan = 19.6 ± 9.07) performed a ¹²³I-FP-CIT SPECT. Depression was diagnosed according to DSM-IV criteria and assessed by Beck Depression Inventory (BDI) (PD-d: 15 ± 2.9; PD-nd: 2 ± 3.4). SPM2 and VOI analyses were then used for group comparisons and correlations. A cluster, with statistically significant (p < 0.05) lower binding FP-CIT in PD-d with respect to PD-nd patients was found in right cingulate cortex, persistent after correction for age, disease severity and duration. When VOIs were correlated with BDI scores in the whole group, an inverse correlation in the right cingulate was observed (r = -0.367, p < 0.01).

Our data indicate no differences in striatal dopaminergic denervation in PD with and without depression. As regards to extra-striatal areas a significant association between depression and cingulate dopaminergic denervation was observed, confirming the dopaminergic hypothesis of PD-depression.

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Abstract – WCN 2013

No: 2710

Topic: 2 – Movement Disorders

Incomplete suppression of head stabilization in idiopathic spasmodic torticollis and Parkinson's disease

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Objective: Sensorimotor integration may be impaired in idiopathic spasmodic torticollis (ST) and other focal dystonias. Also, voluntary interaction with postural reflexes has been shown to be disrupted in ST patients. The way these deficiencies may contribute to the impaired head position control remains however unidentified.

Patients and methods: Thirteen ST patients and 23 healthy controls were subjected to unpredictable, transient, low amplitude, low velocity head-on-trunk, trunk-under-head and whole-body rotations in the horizontal plane. With the instruction not to resist the imposed displacements, resistance to horizontal neck deflections was evaluated.

Results: Patients exhibited a torque offset (bias) in the direction of torticollis before stimulus application. While controls reduced the initial resistance to head-to-trunk rotations within a few hundreds of milliseconds, torque exerted by ST patients increased throughout displacements. The increase of resistance relative to baseline in patients was, however, symmetrical, i.e. independent of the torticollis direction.

Spontaneous torque variations were significantly larger in patients. Strong correlations existed among these abnormal findings.

Conclusions: The deficient ST patients' ability to manipulate normal postural reactions is attributed to impairments of a long-latency slow feedback mechanism. Modeling of this suppressive mechanism suggested further that patients may also have difficulties in switching between feedforward and sensory-driven motor control. Similar findings have been obtained from studying parkinsonian patients with axial rigidity. Differences in impairment patterns between the two groups help to understand altered sensorimotor control in basal ganglia disease leading to rigidity and abnormal postures.

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Abstract – WCN 2013

No: 1456

Topic: 2 – Movement Disorders

Non-motor symptoms in patients with early stages of Parkinson's disease

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Parkinson's disease (PD) – one of the most frequent neurodegenerative diseases.

Objectives: To determine the influence of non-motor symptoms on quality of life and daily activities on PD patients in early stages.

Material and methods: 80 patients with PD on the early stages were included (male:female = 30:50). 20% of our patients had 1–1.5 stage of modified H&Y scale, 35% – 2nd and 45% – 2.5 stage. The mixed form of PD predominated over others (75%). We used Hoehn and Yahr scale modified by Lindvall to assess PD severity (Hoehn M., Yahr M., 1967, O. Lindvall, 1989); assess affective disorders – Hamilton Rating Scale – (M. Hamilton, 1959, 1999); non-motor symptoms – scale non-motor symptoms PD NMS (Chaudhuri K. R. et al., 2004).

Results: Non-motor disturbances were observed in 80%. Hyposmia was observed in 30%; constipation – in 58.6% of patients; violation of urination – in 73% of patients; 33% of patients reported increased sweating; hypersalivation was observed in 27% of patients. Sleep disorders were observed in 75% of patients: insomnia disorders in 44% of patients, excessive daytime sleepiness – 34%, pathological behavior in REM-sleep phase – in 45.7% of patients. Muscle and joint pain in 33% of patients in the early stages. Affective disorders were diagnosed in 80%. There was a positive correlation ($p = 0.047$) of the quality of life with non-motor symptoms, affective and sleep disorders.

Conclusions: Non-motor symptoms in the early stages of the disease significantly deteriorate the quality of life and daily activities of patients with PD.

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Abstract – WCN 2013

No: 2245

Topic: 2 – Movement Disorders

Sensitivity, specificity, positive and negative predictive values and accuracy of datscan™ for prediction of clinical diagnosis of early parkinsonian syndromes

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Objective: To assess the diagnostic efficacy data from clinical trial Kupsch et al, 2012 (data not previously published) conducted using DaTSCAN™ ([¹²³I]loflupane Injection).

Patients and methods: Study imaging group (n = 92) was used to assess the diagnostic accuracy of DaTSCAN™ in subjects with early, clinically uncertain Parkinsonian syndromes (PS) after 1 year follow-up. The reference standard was final clinical diagnosis 1 year after imaging (with DaTSCAN™ results available), and it was compared to baseline clinical diagnosis (without DaTSCAN™) and to baseline imaging diagnosis. Visual assessment of DaTSCAN™ images was performed by local nuclear medicine physicians, which is consistent with current clinical practice. Acquisition of SPECT data with DaTSCAN™ and their reconstruction were performed using a standardized imaging protocol.

Results: The sensitivity of clinical diagnoses at baseline using clinical data was 92% when compared to final clinical diagnosis at 1 year, but the specificity was only 52.4%. For the comparison of baseline DaTSCAN™ images to the clinical diagnosis at 1 year, the sensitivity was 93.9%; specificity was 95.4% ($p = 0.0005$ as compared to baseline clinical diagnosis). The PPV, NPV and diagnostic accuracy for baseline clinical diagnosis or baseline imaging diagnosis vs. final clinical diagnosis at 1 year (reference standard) were respectively: 69.7% vs. 95.8% ($p < 0.0001$), 84.6% vs. 93.2%, and 73.9% vs. 94.6%.

Conclusion: High sensitivity and specificity, PPV, NPV and diagnostic accuracy of DaTSCAN™ in diagnosis of early clinically uncertain PS were demonstrated. Performance of DaTSCAN™ compares favorably in this study to the performance of clinical diagnosis relative to final clinical diagnosis.

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Abstract – WCN 2013

No: 1563

Topic: 2 – Movement Disorders

Pure ataxia associated with N-methyl-d-aspartate receptor antibodies

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Introduction: Several movement disorders previously considered idiopathic or degenerative are now recognized as immune-mediated. The N-methyl-d-aspartate receptor antibody (NMDA) encephalitis has been associated with movement disorders, frequently hyperkinetic. It is estimated that pure monosymptomatic syndromes occur in about 5% of patients. Ataxia is a rare manifestation of this type of encephalitis.

Case study: Male, 73 years old, Caucasian, resident in Mozambique. He presented with clinical picture of fever, anorexia and cough lasting two weeks and treated with gentamicin, azithromycin and metronidazole for seven days with remission of complaints. Ten days after he started a rapidly progressive gait imbalance, being admitted 3 weeks later. Neurological examination showed slight horizontal nystagmus, bilateral appendicular ataxia and ataxic gait with loss of postural reflexes. Psychiatric disorder or hyperkinesias were not observed and patient and caregivers denied any other changes in previous weeks. Examination of cerebrospinal fluid and cranial MRI were unremarkable. The titer of anti-NMDAR antibodies in serum was high (1/1000), while infectious serologies, other autoimmunity studies and search for occult neoplasm, including positron emission tomography scan, were negative. Remarkable clinical improvement followed treatment with high-dose corticosteroids, with complete remission of ataxia within 3 months and reduction of anti-NMDAR

titer to 1/320. The patient remains asymptomatic six months afterwards.

Conclusion: We describe for the first time the association between pure ataxia and anti-NMDAR antibodies preceded by an infectious prodrome. Remission of the severe and rapidly progressive ataxia after corticosteroid therapy supports a potential pathogenic role of these antibodies in a presumably post-infectious setting.

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Abstract — WCN 2013

No: 1616

Topic: 2 — Movement Disorders

Voiding function is correlated with motor severity in patients with Parkinson's disease?

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Background: Patients with Parkinson's disease (PD) have not only motor impairment but also lower urinary tract dysfunction. Among lower urinary tract symptoms, storage symptoms/disorders are the most common and well known. However, the detail of voiding symptom/disorder is less clear.

Objective: We present a detailed data from an urodynamic-pressure-flow measurement which can evaluate both bladder contractility and outlet obstruction during voiding in PD patients, with particular attention to the interrelation between the questionnaire items, the urodynamic parameters, and the clinical characteristics.

Patients and methods: 114 consecutive patients with PD were recruited. The studied patients consisted in 48 de novo patients, 30 patients without motor fluctuations, and 36 patients with motor fluctuations.

Results: Lower urinary tract symptoms were found in 86.0% patients and abnormal UDS findings in 91.2%. During storage, detrusor overactivity (65.8%) and increased bladder sensation (13.2%) were shown. During voiding, 55.3% showed detrusor underactivity (DU), 18.4% showed bladder outlet obstruction (BOO), resulting in decrease in maximum flow rate (38.6%) and presence of 50–100 ml post-void residual (PVR) (13.2%). In multivariate analyses, urinary urgency, urge incontinence and hesitancy correlated with disease severity. DU, decreased maximum flow rate and PVR correlated with disease severity, while detrusor overactivity and BOO with gender. Night-time frequency, urinary urgency or urge incontinence impaired quality of life (QOL) in cooperation with disease severity, while prolongation or feeling of incomplete affected QOL by itself.

Conclusion: In PD patients, detrusor underactivity and related voiding parameters were correlated with motor severity and some voiding symptoms affected QOL by itself.

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Abstract — WCN 2013

No: 1205

Topic: 2 — Movement Disorders

Subthalamic deep brain stimulation can improve gastric emptying in Parkinson's disease

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Background: It remains unclear as to whether subthalamic deep brain stimulation (STN-DBS) would be effective in improving gastrointestinal dysfunction in patients with Parkinson's disease (PD).

Objective: To investigate the effects of STN-DBS on gastric emptying in PD patients.

Patients and methods: Sixteen patients with PD who underwent bilateral STN-DBS were enrolled. Gastric emptying was expressed as the peak time of ¹³C₂ excretion (Tmax) in the ¹³C-acetate breath test and was assessed in patients with and without administration of 100–150 mg levodopa/decarboxylase inhibitor (DCI) before surgery, and with and without STN-DBS at 3 months post-surgery. To evaluate potential factors related to the effect of STN-DBS on gastric emptying, we also examined the association between gastric emptying, clinical characteristics, equivalent dose of levodopa and serum ghrelin levels.

Results: Tmax values for gastric emptying in patients without and with levodopa/DCI treatment were 45.6 ± 22.7 min and 42.5 ± 13.6 min, respectively (not significant). Tmax values without and with STN-DBS after surgery were 44.0 ± 17.5 min and 30.0 ± 12.5 min (P < 0.001), respectively, which showed that STN-DBS was effective. The difference in Tmax values without levodopa/DCI before surgery and without levodopa/DCI and STN-DBS after surgery was not significant, although motor dysfunction improved and the levodopa equivalent dose decreased after surgery. There was little association between changes in ghrelin levels and changes in Tmax values in the STN-DBS trial after surgery.

Conclusion: These results showed that levodopa/DCI did not influence gastric emptying and that STN-DBS improves the gastrointestinal dysfunction in PD patients.

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Abstract — WCN 2013

No: 2637

Topic: 2 — Movement Disorders

Nocturnal blood pressure, non-dipping and psychosis in Parkinson's disease

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Objective: The aim of this study was to determine the relationship between 24 h ambulatory BP measurements (ABPM), e.g., absence or presence of nocturnal dipping, and psychosis in PD.

Methods: 21 patients with PD (15 male, 6 female, disease duration 9 ± 7.4 years) underwent 24 h ABPM using an autonomic protocol and self-report diary. A decrease in nocturnal (time asleep, as indicated in diary) mean arterial blood pressure (MAP) of less than 10% was defined as non-dipping. Patients were interviewed (including the Brief Psychiatric Rating Scale; BPRS) for the assessment of psychosis. Patients had a mean levodopa equivalent dose (LED) of 422. All patients slept at least 4 h through the night.

Results: 11 patients were dippers and 10 were non-dippers. BPRS scores were higher in non-dippers vs dippers and non-dippers on average met the criteria for psychosis (mean non-dipper BPRS: 34.3 ± 7.3 vs mean dipper BPRS: 27.5 ± 5.3 ; cut off for “mildly ill” 31). There was a correlation ($r = .488$) between BPRS scores and non-dipping (percentage of day/awake and at night/asleep MAP difference) indicating that those patients who had a blunted nocturnal fall in BP were more prone to psychosis or psychotic symptoms. (Pearson's Correlation = .423, $p = .025$).

Conclusion: These results suggest that PD patients who show a blunted circadian BP rhythm are more likely to develop psychosis than those whose BP decreases normally at night. Reasons for the pathological loss of nocturnal BP fall and psychological symptoms warrant further evaluation.

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Abstract – WCN 2013

No: 2594

Topic: 2 – Movement Disorders

Orthostatic tremor, progressive external ophthalmoplegia and twinkle mutation

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Background: Orthostatic tremor (OT) is characterized by 13–18 Hz tremor occurring in standing position and has unknown pathophysiological mechanism. OT is thought to be sporadic, but siblings with OT from 3 unrelated families have been reported. Mutations in *TWINKLE* can result in autosomal dominant progressive external ophthalmoplegia (PEO) and have been associated with Parkinsonism in rare cases.

Objective: To identify the molecular defect of a patient with OT and PEO.

Patient and methods: A 69-year-old man manifested ptosis and OT at age 37 years, followed by PEO and limb fatigue. His father had ptosis; a sister and a son have ptosis and PEO. EMG/nerve conduction and multichannel surface EMG recordings, muscle biopsy, brain MRI and MR spectroscopy (MRS), sequencing of all coding exons and flanking introns of *TWINKLE*, *POLG* and *ANT1* were performed.

Results: Patient had electrophysiological evidence for a mild myopathy and sensorimotor peripheral neuropathy. Polygraphic recordings showed orthostatic synchronic 17.5 Hz tremor in the leg muscles spreading to lumbar paraspinal and arm muscles. There was no rest tremor and no evidence for parkinsonism. Brain MRI/MRS showed mild generalized atrophy. Scattered ragged-red fibers were present on muscle biopsy. Gene analysis revealed a novel heterozygous missense mutation at an evolutionarily conserved residue of *TWINKLE*. The affected sister and son carry the same variant.

Conclusions: Although one cannot exclude the incidental association of OT and *TWINKLE* mutation, the simultaneous onset of OT and

ptosis at a younger age than usually observed raises the possibility of mitochondrial dysfunction in the pathogenesis of OT.

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Abstract – WCN 2013

No: 2544

Topic: 2 – Movement Disorders

Axial symptoms and cognitive functioning in Parkinson's disease

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Background: The association between motor and nonmotor symptoms in Parkinson's disease (PD) is still poorly understood.

Objective: To explore the association between axial symptoms and cognitive functioning in PD.

Patients and methods: Eighty-three consecutive patients with idiopathic PD stage 1–3 Hoehn & Yahr (57% men; mean age = 66.6, sd = 9.9; mean education = 6.7 y, sd = 4; mean disease duration = 6.9 y, sd = 4.4; mean levodopa equivalent dose = 722 mg, sd = 460) performed the Dementia Rating Scale-2 (DRS-2). This cognitive score was adjusted for age and education. Hoehn & Yahr and Unified Parkinson's Disease Rating Scale-III (UPDRS-III; mean = 30.8, sd = 9.7) were applied after 12 h without antiparkinsonian medication. Two indexes were derived from UPDRS-III: axial symptoms (items 18, 27–30) and other motor symptoms (remaining items). Pearson's correlation and multiple linear regression were used for data analyses.

Results: DRS-2 was significantly correlated with levodopa equivalent dose ($r = -.242$, $p = .028$), UPDRS-III total ($r = -.437$, $p < .001$), axial symptoms ($r = -.573$; $p < .001$), and other motor symptoms ($r = -.345$; $p = .001$). No significant association was found with disease duration ($r = -.133$; $p = .230$). Multiple linear regression analysis was conducted with DRS-2 as the dependent variable and the following as independent variables: levodopa equivalent dose, axial symptoms, and other motor symptoms. Only axial symptoms remained statistically associated with DRS-2 (adjusted $\beta = -0.415$; $p < 0.001$), while adjusting for other covariates.

Conclusion: This study confirms previous reports that cognitive functioning is related with motor symptom severity, but not with disease duration. Axial symptoms are independently associated with poorer cognition in mild to moderate PD, when other motor symptoms and levodopa equivalent dose are taken into account.

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Abstract – WCN 2013

No: 2601

Topic: 2 – Movement Disorders

Neurological disorders of HTLV-1 carriers patients taken care of on the clinic of neurology of the FBDC HTLV center

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In this study they described the neurological disorders of 116 carrying patients of the Human T Cell Lymphotropic Virus type 1 (HTLV-1) taken

care of in the Clinic of Neurology of the FBDC HTLV Center, from the following question: which were the disorders and neurological states found in the patients of the Clinic of Neurology of the FBDC HTLV Center between September of 2005 and August of 2006? To reach this purpose was carried through, using medical record analysis (documentary analysis) and the neurological examination. The more frequent neurological disorders had been paresthesia, hyperactive patellar reflex, lumbar pain, intestinal dysfunction, muscular weakness of lower limbs and urinary disturbances. Considering these disorders, 50 patients with HAM/TSP, 39 oligosymptomatic and 27 asymptomatic had been identified. The diagnosis of most of the patients was made between 1 and 8 years after the beginning of the symptoms. With these results, the expectation of the authors consists on the propagation of attendance programs to the HTLV-1 carrying patients with neurological disorders.

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Abstract – WCN 2013

No: 2610

Topic: 2 – Movement Disorders

Disautonomic disturbances as the main initial clinical presentation of multiple system atrophy syndrome

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Background: It has been previously reported a case in which the main initial presentation of multiple system atrophy syndrome is preceding cognitive impairment and extrapyramidal signs.

Objective: To present a series of cases of 3 patients, with multiple system atrophy syndrome who developed prior to cognitive impairment and extrapyramidal signs recurrent syncope as initial clinical feature.

Material and methods: Neurophysiological (autonomic function) tests, magnetic resonance image and neuropsychological examinations were required.

Results: Each patient's clinical history bore a common feature, a history of recurrent syncope and fainting. This lasted ten to fifteen years prior to mild cognitive impairment and in one case two extrapyramidal signs rather than cognitive impairment. These late cases proved to have both parasympathetic and sympathetic function alterations. It is extremely rare that these dysautonomia present as an initial clinical feature multiple system atrophy.

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Abstract – WCN 2013

No: 2633

Topic: 2 – Movement Disorders

Cerebellar dysfunction is closely related to cognitive decline in Uner Tan syndrome

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Background: Uner Tan syndrome (UTS), first reported in 2005, consists of habitual quadrupedalism, mental retardation and dysarthric or no speech, accompanied with cerebellar hypoplasia and mild gyral simplification of the cerebral cortex.

Objective: To investigate the relation of the cerebellar dysfunctions to cognitive functions in UTS cases.

Subjects and methods: The patients (n = 6) were recruited from a family with 17 siblings, constituting a homogeneous group. Cognitive ability was assessed using the Wechsler Adult Intelligence Scale-Revised (WAIS-R) and the Minimal State Examination Test (MMSE). To evaluate cerebellar ataxia symptoms, the ICARS (International Cooperative Ataxia Rating Scale) including subscales postural, kinetic, speech, and oculomotor disorders was used, being examined in relation to IQ test scores.

Results: Age was not significantly associated with ICARS, WAIS-R, and MMSE test scores. Higher ICAR scores indicated higher degrees of cerebellar impairment including the postural, kinetic, speech, and oculomotor disorders. The total ICARS ranged between 36 and 85, being close to maximal ICARS of 100. The scores for the ICARS subscales were also within the highest ranges. The Pearson correlation coefficients indicated highly significant negative linear correlations of the WAIS-R and MMSE test scores with ICARS including its subscales, the postural, kinetic, speech, and oculomotor disorders.

Conclusion: The results suggested that the impaired cognition in UTS cases may be caused by the cerebellar ataxia symptoms, such as postural, kinetic, speech, and oculomotor dysfunctions.

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Abstract – WCN 2013

No: 2503

Topic: 2 – Movement Disorders

Alcohol challenge and the sensitivity to change of the essential tremor rating assessment scale (TETRAS)

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Background: The performance scale of The Essential Tremor (ET) Rating Assessment Scale (TETRAS) has excellent inter- and intrarater reliability. Information on the ability to detect change is scarce.

Objective: We evaluated the sensitivity to change during a standardized ethanol challenge by comparing TETRAS with standard postural tremor accelerometry.

Patients and methods: A single oral ethanol dose calculated to reach 0.05 g/dl breath alcohol content (brAC) was given to 15 adult ET patients on two separate days. Postural tremor accelerometry was assessed on one day, TETRAS on the other. Measures including brAC readings were taken at 8 time-points (2 time-points pre, 6 time-points, every 20 min, up to 2 h post ethanol).

Results: Accelerometry data was log-transformed, and a cumulative score $\log\text{ACC}(r + 1)$ was calculated. Correlation between $\log\text{ACC}(r + 1)$ and TETRAS was significant. *rmANOVA* of TETRAS and $\log\text{ACC}(r + 1)$ showed a significant effect of time-point before and after ethanol. There was no difference between the two baseline measures with corrected post-hoc tests, whereas there was a difference between baseline and each following time-point. TETRAS and brAC were significantly correlated. BrAC measurements of the two study days showed neither a significant difference over time nor at the time-point of maximum effect.

Conclusions: Sensitivity to change of the TETRAS performance scale was shown after a pharmacologic intervention. Furthermore, the excellent reproducibility of the alcohol challenge could be of importance for further studies. The ability of TETRAS in demonstrating response after a treatment intervention establishes its potential as valid instrument for both clinical and research settings.

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Abstract – WCN 2013**No: 2533****Topic: 2 – Movement Disorders****Diagnostic exome sequencing in neurological disorders**

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Background: Next generation sequencing (NGS) or high throughput sequencing has for the last years been used routinely in genetic diagnostics at Telemark Hospital. Targeted NGS with panels for several genes contributes diagnostically to inherited neurological disorders such as Charcot–Marie–Tooth disease, distal Hereditary Motor Neuronopathy, Hereditary Spastic Paraplegia and Neurofibromatosis. In addition, NGS exome sequencing, i.e. sequencing of all exons, may identify pathogenic, disease causing mutations.

Material and methods: Exome NGS sequencing was performed in a patient with movement disorder to identify pathogenic mutations. A probable clinical diagnosis and mode of inheritance is a necessary tool for successful exome NGS. This case was sporadic. Hence, the alternatives are *de novo* mutation or autosomal recessive inheritance. Inheritance and phenotype aid the bioinformatic analysis to more easily elaborate the genes that are most relevant regarding the symptomatology.

The bioinformatic platform consists of CASAVA, bwa, GATK, BEDtools and Annovar. Normal sequence variants from dbSNP129 are removed before annotation. Variants described in Exome Sequencing Project, 1000 genomes and databases over variants detected in normal in-house controls are used to clarify individual variant pathogenicity.

The remaining, unexplained sequence variants are elaborated manually with Alamut, Human Genome Mutation Database and other prediction tools. All assumed pathogenic mutations identified by exome NGS are confirmed with Sanger sequencing.

Results: The identified mutation confirms the clinical neurological diagnosis.

Conclusion: The case is presented to elucidate the utility and versatility of exome NGS as a method also in more ordinary neurological disorders and not only restricted to syndromes or intellectual disability.

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Abstract – WCN 2013**No: 700****Topic: 2 – Movement Disorders****The role of liver transplant in the treatment of acquired hepatocerebral degeneration and hepatic encephalopathy**

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Background: Encephalopathy (HE) and acquired hepatocerebral degeneration (AHD) have been described in patients with advanced liver disease (ALD).

Objective: We aimed to describe two patients' evolution pre and post-liver transplant (LT).

Patients and methods: We reviewed clinical, neuropsychological and neuroimaging data (pre/post LT) from 2 ALD patients.

Results: 1. 67 year-old male, HCV cirrhosis, cognitive complaints since the age of 61. At 63 yo he had cognitive impairment, parkinsonian and dystonic syndrome and later oculomotor dysfunction. Brain MR showed discrete symmetric T1 hyperintensity in globi pallidi and putamina and bilateral T2 hyperintensity along the corticospinal tract. At 65 yo he was submitted to LT with progressive improvement. Seven months later he had no extrapyramidal signs, cognition improved and MR became normal.

2. 49 year-old female, alcoholic cirrhosis admitted with severe acute HE (altered consciousness – drowsy – without focal neurological signs). Brain MR showed multiple areas of T2 hyperintensity in fronto-parieto-occipital cortex, with restriction in water diffusion, cortical expansion and T1 hyperintensity in globi pallidi and red nuclei. Emergent LT was performed. She became awake. Extrapyramidal features and cognitive impairment took long to recover. One month later MR showed marked improvement of cortical T2 hyperintensities, persisting slight pallidal T1 hyperintensities. Four months later a complete resolution of T1 and T2 grey matter hyperintensities occurred with residual peritrigonal white matter T2 hyperintensity with cortical atrophy.

Conclusion: AHD and HE are entities with distinct clinical and imagiological features that might be reversed by LT.

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Abstract – WCN 2013**No: 1140****Topic: 2 – Movement Disorders****IRX4204 As a novel disease-modifying compound for treatment of Parkinson's disease**

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Background: IRX4204 is a second generation retinoid × receptor (RXR) agonist currently being tested for prostate cancer treatment with an excellent safety record. Nurr1-RXR heterodimer selective agonists have been actively pursued as a potential pharmacological target for Parkinson's disease (PD) due to their neuroprotective effects in models of PD.

Objective: The objective of our study is to evaluate the effects of IRX4204 as a potential novel therapeutic in PD neuropathology.

Materials and methods: Primary rat mesencephalic cultures were used to study the role of IRX4204 on Nurr1-mediated neuroprotection. The 6-hydroxydopamine (6OHDA) induced rat model of PD was used to examine the potential neuroprotective role IRX4204 on PD pathology. Nuclear magnetic resonance (NMR) and photo-induced cross-linking of unmodified protein (PICUP) were used to evaluate the role IRX4204 on prevention of α-synuclein binding and oligomerization.

Results: We found that IRX4204 can selectively promote dimerization of the nuclear factor Nurr1-RXR at nM concentration in vitro and can promote expression of neurotrophic factors for the survival and maintenance of nigral dopaminergic (DA) neurons in a dose-dependent manner in vivo. This evidence is consistent with a significant attenuation of PD-type motor impairments following 6-OHDA lesions in response to IRX4204. Using a combination of NMR spectroscopy and PICUP assays, we found that IRX4204 shifts and prevents oligomerization of α-synuclein.

Conclusion: Our data suggest that IRX4204 may benefit PD by providing neuroprotective support for DA neurons and by protecting DA neurons from α-synuclein neurotoxicity.

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Abstract — WCN 2013**No: 2532****Topic: 2 — Movement Disorders****The missing tau mutation: +15 exon 10/intron 10 boundary of MAPT stem loop structure in an Irish family with FTDP-17**

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Background: Up to 40% of families with autosomal dominant fronto-temporal dementia with parkinsonism (AD FTDP-17) carry pathogenic 5' splice site mutations in the Microtubule-Associated Protein Tau (MAPT) gene. Several stem-loop mutations of tau exon 10 were identified, but the +15 mutation remained elusive.

Aim: We present a family with the 'missing' +15 mutation.

Case: A 44 year old man developed progressive disinhibition, apathy and simultaneous amnesic syndrome. Neurological examination normal (primitive reflexes/disinhibition). Pedigree: autosomal dominant young-onset dementia.

Investigations: MRI brain — moderate bitemporal atrophy; PET CT — low anterior temporal activity, CSF — raised protein: phosphorylated tau/beta-amyloid normal. Fluorescent sequencing analysis for familial tau variant: sequence variant c.915 + 15A > C, at exon 10/intron 10 boundary of MAPT gene. Identical variant in first cousin.

Discussion: In 1998, Hutton described 5' splice site mutations in tau, associated with FTDP-17, destabilising a stem-loop structure, causing alternate splicing of exon 10. The predicted +15 A > C mutation at intron 10 was 'missing' until now. In this unique family, the proband had profound short-term amnesia and behavioral change, suggesting atypical familial Alzheimer disease or atypical FTDP-17. Detection of novel +15 mutation clarified diagnosis of FTDP-17 and informed genetic counselling. Finding this mutation 14 years after prediction highlights value of pursuing complete family history to guide appropriate molecular genetic testing.

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Abstract — WCN 2013**No: 2483****Topic: 2 — Movement Disorders****Orthostatic tremor responding to Rasagiline**

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Background: Orthostatic tremor is known as a lower extremity tremor occurring when standing. When a person stands, typical high frequency (13–18 Hz) tremor develops, and disappears with walking, sitting or continuous volatile leg movement.

Objective: In this report we presented a patient with tremor on the lower limbs diagnosed with Parkinson's disease.

Patients and methods: A 58-year-old female patient presented to our outpatient clinic with complaints of trembling in the body when standing, lasting for 1 year. The neurological examination revealed bradykinesia of the lower and upper right limbs, rigidity and rest tremor of the right hand and trembling of the lower limbs when standing.

Results: The brain MRI and the routine nerve conduction studies were normal. The superficial electrode recordings of femoris, anterior tibialis, biceps femoris muscles documented a tremor of 13 Hz. The patient was diagnosed with Parkinson's disease accompanied by orthostatic tremor and was started 2 mg of Rasagiline. Significant improvement was noted.

Conclusion: We found this case worth reporting because of the rare presentation of Parkinson's disease in the literature with orthostatic symptoms as a first sign.

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Abstract — WCN 2013**No: 2514****Topic: 2 — Movement Disorders****Extrapontine myelinolysis presented with sudden-onset generalised chorea**

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Objective: Pontine myelinolysis has been well described in the presence of rapid correction of hyponatraemia for decades. Both pontine and more rare extrapontine myelinolysis is considered as an iatrogenic syndrome following rapid correction of hyponatraemia leading to rapid changes in osmolality. However, it occurs also in various other medical conditions. Patients with chronic hyponatraemia are more prone to develop myelinolysis. It is also not so rare complication of haemodialysis.

Patients and methods: We present a case of 67-year old male with the history of diabetic nephropathy and nephrosclerosis on peritoneal dialysis with progressively worsening generalised chorea. Laboratory results showed mild hyponatraemia and severe uraemia. CT of brain was inconclusive. MRI of brain revealed symmetrical hypersignal lesions in both basal ganglia (putaminal regions) probably of osmotic demyelination origin. Patient was treated with hemodialysis and hyponatraemia was gradually normalised.

Results: According to laboratory and MRI results, diagnosis of EPM was established. Generalised chorea was treated with clonazepam with no effect. Switch to haloperidol was effective in treatment and chorea gradually disappeared. After 6 month since onset patient stopped taking Haloperidol and choreatic dyskinesias did not re-occur.

Conclusion: Generalised chorea with sudden onset is extremely rare presentation of EPM. We suggest tight cooperation of neurologists and nephrologists treating peritoneal dialysis patients to prevent permanent brain damage.

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Abstract — WCN 2013**No: 2520****Topic: 2 — Movement Disorders****Improved intraoperative therapeutic window with directional DBS compared to omnidirectional DBS using a novel lead design**

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Introduction: Deep brain stimulation (DBS) is currently carried in all directions around the lead. Directional leads may improve focalization of stimulation in the brain but their clinical effect has not yet been investigated. We report five cases of an on-going intraoperative study using directional DBS.

Methods: Three males with PD underwent STN DBS and two males with ET underwent Vim DBS. At the target determined for the permanent lead, directional stimulation was assessed using a novel lead (directSTIM™, Aleva Neurotherapeutics). It features two rings of three independent electrodes each. The therapeutic window (TW) is defined as the electrical current at which side effects occur minus the current at which a significant therapeutic effect is observed (low TW

boundary = TWLB). TW was measured in each direction (TWdir) and best TWdir was compared to omnidirectional stimulation (TWomni).

Results: A range of anticipated side effects presented and receded as we selected the direction of stimulation. 3/5 patients showed a ratio TWdir/TWomni ≤ 0.30 . 1/5 patient did not show a ratio < 1 . The ratio could not be measured on 1/5 patient. The best directional TWLB was 0.66 mA in average (0.3–1.0 mA) whereas omnidirectional TWLB was 1.14 mA (0.6–1.95 mA).

Conclusions: These observations suggest that directional TW is increased compared to omnidirectional TW. The side effects appeared in accordance with the spacial position of the directional electrodes. TWLB is significantly lower in the best direction compared to omnidirectional stimulation. Therefore, directional stimulation may increase effectiveness of DBS. Further studies are needed to confirm these results.

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Abstract – WCN 2013

No: 2458

Topic: 2 – Movement Disorders

Clinical and biological contribution in Parkinson's disease

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Introduction: Parkinson's disease (PD) represents the main cause of parkinsonian syndrome. The objective of this study was to identify the characteristics of patients with PD in the center of Tunisia.

Methods: A study was performed in patients with PD followed between 2006 and 2009 in Department of Neurology, Hospital Sahloul of Sousse. Demographic and clinical data at presentation were documented for all patients.

Results: We identified 300 patients with PD. The mean age at onset was 58.6 and the duration was 5.5 years. The frequency of young onset PD was 11.6% and late onset was 9.4%. A positive family history of PD was present in 14% of cases. The trembling form was 49.3%. Hypertension and diabetes were associated with PD in 25% and 15%, respectively. Median Hoehn–Yahr scale of PD patients at presentation was a median 2.0. A significant correlation was observed between severity of PD and the duration of the disease. L-DOPA is the most prescribed for treatment of PD (88.8% in first-line) and its motor complications were observed in 43%. A positive correlation was found between homocysteinemia and the dose of L-Dopa as well as the Hoehn–Yahr stage.

Discussion: There were no differences between our study and literature with regard to clinical forms. PD was significantly associated with a decreased risk of hypercholesterolemia and increased risk of diabetes. Our therapeutic attitude was different from the therapeutic strategy of the consensus established in 2000. The hyperhomocysteinemia seems to be related to the use of L-Dopa.

Conclusion: Our study provides data on the clinical profile of Tunisian patients with PD and demonstrates that it is similar to that from other populations.

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Abstract – WCN 2013

No: 2496

Topic: 2 – Movement Disorders

Fungal volatile organic compounds: Biogenic toxins as etiological agents for Parkinson's disease

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Background: Many volatile organic compounds (VOCs) are found in indoor environment as a by-product of microbial metabolism. In damp indoor environments, fungi are associated with poor indoor air quality and some epidemiological studies have suggested that exposure to microbial VOCs has potential to cause movement disorders thereby causing an adverse health effects.

Objective: We used *Drosophila melanogaster*, a simple genetic model as a reductionist approach to determine the causal relationship between fungal VOCs and reported movement disorders.

Material and methods: We used wild type *Drosophila melanogaster* strains and other mutant flies and exposed them to different concentrations of fungal VOCs. We assessed for survival, behavioral and immunohistochemical assays in order to determine the neurotoxic effects of fungal VOCs.

Results: We found that exposure to different fungal VOCs leads to loss of dopaminergic neurons and Parkinson's disease-like manifestations in *Drosophila* model. Furthermore, we observed gene-environmental interaction implying the alteration of dopamine homeostatic and signaling pathway upon exposure to fungal VOCs.

Conclusion: Our reductionist model demonstrates that VOCs emitted by fungi are neurotoxic to *Drosophila* and may provide insights into neurological health related problems reported from damp indoor environment and reported health issues after Hurricane Katrina and Sandy.

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Abstract – WCN 2013

No: 2231

Topic: 2 – Movement Disorders

Trust and risk behavior in Parkinson's disease: Experimental evidence and clinical implications

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Background: Trust is essential for social interactions. The basal ganglia, the frontal cortex, and the limbic system are known to regulate trust behavior. Parkinson's disease (PD) affects these brain regions. Moreover, dopamine and serotonin, neurotransmitters related to trust, are reduced in PD, and various neuropsychiatric phenomena of PD may be related to trust.

Objective: We designed an experiment to investigate whether trust behavior is altered in PD.

Patients and methods: We compared the behavior of 20 PD patients on dopaminergic treatment (age 72.4 ± 9.2 , motor symptom duration 39.2 ± 41.8 months, MMSE 28.4 ± 1.6 , 10 female) with the behavior of 20 healthy controls (age 68.4 ± 10 , MMSE 28.9 ± 1 , 10 female) in a computerized trust game, where subjects (trustors) entrusted money (0 to 10 Euros) to 16 emotionally-neutral faces (trustees). The invested amount was used as a measure for trust. To control for altered risk-taking, a computerized game-of-dice task was used. Patients suffering from neuropsychiatric co-morbidities (depression, anxiety, psychosis, impairment of impulse control, and compulsive behavior) were excluded.

Results: In the trust game, PD patients exhibited significantly less trust compared to controls (mean investment 3.431 ± 1.996 versus 5.528 ± 1.556 Euros; $p = .001$, two-sided Wilcoxon-test). In the game-of-dice task, risk-taking was higher in PD than in controls (number of risky choices 10.2 ± 3.9 versus 7.1 ± 3.6 ; $p = .009$, two-sided Wilcoxon-test).

Conclusion: Trust was reduced in persons with moderately advanced PD without psychiatric co-morbidities. This finding cannot be explained

by PD patients' risk behavior, because it was higher in the PD group. Results are discussed with respect to pathophysiological aspects and clinical implications.

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Abstract – WCN 2013

No: 2470

Topic: 2 – Movement Disorders

Central extrapontine myelinolysis appearing as acute parkinsonism

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Background: When facing an acute parkinsonism, we must take into account not only the secondary and most frequent etiologies, such as the pharmacological, vascular or toxicological ones, but also others which are less common.

Case report: We present the case of a 29-year-old man, who is admitted in our Neurology department with high frequency, constant and incapacitating rest and postural tremor, which was present for 5 days prior to the admission. It was accompanied by rigidity and bradykinesia – all of it with predominance in the right side of the body – as well as hypomimia, dysprosody and abundant sialorrhoea. Postural reflexes were also seriously damaged.

As a precedent, he had been hospitalised that month with an Addisonian crisis, and his sodium levels had increased from 109 to 138 mEq/L in a day.

MRI evidenced a bilateral hyperintensity of the basal ganglia, which is compatible with central extrapontine myelinolysis. The DaTSCAN was normal.

Conclusion: We would like to express the importance of treating hyponatremia in a slow way, because of the risks of osmotic demyelination of the central nervous system, not only distinctly at the pontine level, but also in other locations.

Additionally, we would also like to emphasize the importance of the careful analysis of the patient's history, searching for processes with a possible damage of the basal ganglia. Thus, we will be able to find the diagnosis of more infrequent etiologies of parkinsonism, as was the case here.

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Abstract – WCN 2013

No: 2476

Topic: 2 – Movement Disorders

Impulse control disorder in psychogenic parkinsonism

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Psychogenic movement disorders (PMD) appear in 1.9% of general population and in 2.25% patients with neurological diseases. Psychogenic parkinsonism (PP) is rare and accounts for less than 10% of PMD. The psychogenic parkinsonism is estimated of 0.17% of all cases of parkinsonism.

The authors present the case of 53-year old woman with long-standing recognition of Parkinson disease (PD) with motor fluctuations and dyskinesias, treated with L-Dopa, ropinirol and amantadine. She was admitted to the Neurological Department of Wroclaw Medical University in order to confirm the PD diagnosis, estimate the appropriate treatment or eventually qualify to the operation. The medical history indicated to the impulse control disorders (ICD) – compulsion for the internet usage, shopping (she bought the same, useless things repeatedly). The analysis

of the medical history, observation of the motor symptoms and their changes, nonspecific influence of dopaminergic treatment, psychological and psychiatric evaluations allowed us to recognize psychogenic parkinsonism (the Jankovic criteria 1, 2, 6, 7, 8) for the certain recognition of PP.

Despite of any changes of the dopaminergic treatment, the compulsive actions disappeared spontaneously. Slow discontinuation of the dopaminergic treatment did not enhance the typical for PD motor disturbances.

The influence of dopaminergic agonists and L-Dopa treatment as well as familiar, psychiatric disorder (patient's mother suffered from bipolar disorder) or conversion with taking the pattern from the internet, was thought to be important in the etiology of ICD in our patient.

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Abstract – WCN 2013

No: 2274

Topic: 2 – Movement Disorders

Novel mutations in cathepsin D in juvenile onset neuronal ceroid lipofuscinosis

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Background: Mutations in cathepsin D (CTSD) have been previously reported in patients with neuronal ceroid lipofuscinosis (CLN10). To date only 7 mutations have been reported in four families with an NCL-like illness. We report the identification of two novel homozygous mutations in two families with juvenile onset ataxia and retinitis pigmentosa.

Methods: Exome sequencing combined with homozygosity mapping was employed to identify a novel CTSD mutation in a single patient. This patient presented age 14 years with retinitis, progressive ataxia and cognitive impairment and had previously been screened for the known lysosomal storage disorders. The coding exons of CTSD were subsequently screened in 95 patients with complex ataxia and retinitis pigmentosa. One additional patient was identified with a further novel CTSD mutation. catD activity was then assayed in patient fibroblasts and leucocytes using a fluorometric assay.

Results: 2 novel homozygous mutations were identified in exons 4 and 9 in the respective patients. Both presented with a juvenile onset of retinitis pigmentosa, ataxia and cognitive impairment. Fluorometric assay of the catD function revealed significantly reduced activity compared to controls in both individuals. The assay was also optimised for use in homogenised leucocyte pellets in addition to fibroblast samples.

Discussion: This study reports two novel mutations that have been demonstrated to result in significant reduction in catD activity. This study expands range of mutations in CLN10 and also provides further insight into the range of phenotypes associated with the disorder. Patients with complex ataxia and retinitis should be screened for CTSD deficiency.

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Abstract – WCN 2013

No: 2465

Topic: 2 – Movement Disorders

Self-assessment of dysarthria in Parkinson's disease verified by use of computer acoustic speech analysis

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Speech disorders in Parkinson's disease (PD) have a complex character, depending on many factors influencing speech, phonation and articulation programming. There are no studies defining self-assessment of speech disturbances in PD patients.

The studies covered 50 patients diagnosed with PD according to the clinical criteria determined by the United Kingdom Parkinson's Disease Society Brain Bank. The control group comprised 30 participants, not diagnosed with any neurological disorders or other illnesses that might have influenced speech. In the II part of the UPDRS (point 5) the median result was 1 (in 66% of patients). This point assesses speech problems based on the patient's medical history and our results mean insignificant speech disorders not causing understanding difficulties. In the point 18 of the III part of the UPDRS, the median result was 2 (in 48% of patients), which stands for a moderate dysarthria. In computer acoustic analysis we revealed the changes of speech parameters in all our patients. The differences between PD and control groups were statistically significant. Speech disorders in PD are not usually exposed in the patients' self-assessment. Self-assessment of speech disturbances depends on the precise sounds differentiation, mostly in high frequency range. These abilities diminish within age and it could be the reason of the impairment of speech problems' perception. The computer acoustic speech analysis is objective and precise method of assessment of speech disorders in PD patients.

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Abstract – WCN 2013

No: 2457

Topic: 2 – Movement Disorders

Motor and non-motor determinants of chronic fatigue in idiopathic Parkinson's disease (IPD)

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Background: Fatigue is one of the most common non-motor symptoms in patients with Parkinson's disease (PD) with significant impact on quality of life. Fatigue has received increasing attention during recent decade; however, few studies have examined the determinant factors of fatigue and little is known about the relationship between fatigue and other motor and non-motor symptoms.

Objective: The aims of this study were to evaluate determinant factors relating to fatigue in PD and to compare the strength of measure of association for different non-motor and motor symptoms with fatigue severity.

Patients and methods: This cross-sectional study was performed in a movement disorder clinic in Tehran, Iran during 2011–2012. A total number of 54 PD (38 males and 16 females) patients with the mean age of 62.5 (SD = 9.1) yr were recruited. Fatigue was scored using the Fatigue Severity Scale (FSS). Other scales were used including PDQ39 (PD-specific QoL), HADS (anxiety & depression), MNA (mini-nutritional assessment), Schwab & England (activity of daily living) and Hoehn & Yahr scales (level of disability).

Results: Patients' score in fatigue had significant correlation with anxiety ($r = 0.32$, $P = 0.020$), depression ($r = 0.41$, $P = 0.002$), disease severity measured by Hoehn & Yahr scale ($r = 0.36$, $P = 0.007$) and activity of daily living described by Schwab & England scale ($r = -0.35$, $P = 0.011$).

Conclusion: Our findings showed that more severe depression is accompanied with more severe fatigue in PD patients. In addition to motor symptoms of PD, anxiety and depression have noticeable relationship with fatigue, which must be considered in management of fatigue in PD patients.

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Abstract – WCN 2013

No: 1914

Topic: 2 – Movement Disorders

Parkinsonian syndrome revealing a Systemic Lupus Erythematosus disease one case study (displayed communication)

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Introduction: The Systemic Lupus Erythematosus (SLE) has many neuropsychiatric manifestations. Rarely Parkinson's syndrome has been reported in this condition.

Objective: To report a new case of parkinsonian syndrome revealing a SLE.

Patients and methods: A 56-year-old woman who presented clinically since 08 years of a parkinsonian syndrome trembling form, has a walking disorder (freezing) with postural instability worsening rapidly and unresponsive to dopaminergic therapies.

The blood tests revealed leukopenia 3100/mm³ with elevated proteinuria 24 h (3 g/24 h) without alteration of renal function.

The evaluation of auto-immunity has revealed the presence of high levels of antinuclear 1/320, and antinative DNA antibodies at 1/100UI/ml with negative antiphospholipids antibodies.

The brain MRI showed abnormal signal intensity on T2 and FLAIR in basal ganglia (putamen).

According to the diagnostic criteria defined by reference SLE American College of Rheumatology (ACR), the diagnosis is in favor of a parkinsonian syndrome secondary to SLE.

The evolution was favorable with corticosteroids therapy which is another criterion of this diagnosis.

Results: The mechanisms underlying neurological manifestations of SLE are not yet elucidated, and two assumptions are made, one leading to immunological direct neuronal damage, the other explained by vascular damage of small vessels.

There are 30 cases of Parkinson's disease as a complication of lupus reported in the literature.

Conclusion: Parkinsonism in young women can hide various etiologies including SLE, that we need to know and seek to establish appropriate care.

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Abstract – WCN 2013

No: 2449

Topic: 2 – Movement Disorders

Biballismus due to thyrotoxicosis

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Background: Bilateral ballism, characterized by continuous, coarse, flinging involuntary movements involving the limbs with proximal dominance is extremely rare. Hyperthyroidism is usually accompanied

by nervous system dysfunctions. Here, we present a patient in whom biballism occurred d thyrotoxicosis.

Case: A 62-year old right-handed male with mild tachycardia has noted abrupt onset of involuntary movements 1 week prior to admission. He was alert and well orientated. Constant ballistic movements involving proximal musculature were seen on both arms and legs. These movements disappeared during sleep. No involuntary movements were seen in his face. Cranial nerve examination and strength of extremities were normal. Tendon reflexes were symmetric and slightly brisk. The hematological examination, biochemical examination including blood glucose, electrolytes, renal and liver function tests, copper studies were normal. Laboratory tests for thyroid function were as follows: total serum T3:3.72 ng/ml, total serum T4:14.5 ng/ml, free T3:8.5 pg/ml, free T4:4.3 ng/dl, TSH: 0.001 mu/ml, thyroglobulin: 230 ng/nl. Cranial MRI demonstrated no abnormalities. The abnormal movements suppressed over the next week following administration of propylthiouracil 100 mg three times a day. Two weeks later when free T3 and T4 were decreased, the ballistic movements dramatically resolved. Another episode occurred during a 9-month follow up. Since the biballism disappeared with euthyroidism but reappeared whenever there was a further increase in T3 and T4, its relationship on hyperthyroidism and its reversibility are clearly demonstrated.

Conclusion: Hyperthyroidism-related biballismus was thought to be a result of a transient metabolic disturbance of the basal ganglia circuits rather than a permanent and irreversible change.

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Abstract – WCN 2013

No: 2387

Topic: 2 – Movement Disorders

Psychotic symptoms overshadow motor dysfunction and dementia in normal pressure hydrocephalus – A case report

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Background: The characteristic symptoms of normal pressure hydrocephalus are dementia, urinary incontinence and gait disturbance. Psychiatric symptoms may also occur but appear usually after the characteristic symptoms.

Objective and methods: In our case report we present a 63 year old woman that had been diagnosed with a bipolar affective disorder at the age of 61 with an acute onset of psychosis. She had been treated with several neuroleptics and anticonvulsants before being admitted to a neurological department. The reason for the admission was extrapyramidal signs, which had been explained as a side effect of the neuroleptic use. In the physical examination the patient showed in addition to the motor dysfunction, a massive cognitive decline. Although a MRI of the brain and a lumbar puncture had been already preformed in the past, we decided to repeat those. Now signs of normal pressure hydrocephalus were visible in the MRI. Also the patient significantly improved in walking and in cognitive testing after the second spinal tap.

Results: A surgically implanted ventriculoperitoneal shunt to drain excess cerebrospinal fluid has stabilized the patient. Neuroleptics and anticonvulsant medications have been stopped. No further signs of a psychosis have been reported since then.

Conclusion: This case demonstrates the need to consider NPH when older patients present with psychotic symptoms, particularly in the presence of cognitive impairment, gait disturbance, or incontinence.

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Abstract – WCN 2013

No: 2395

Topic: 2 – Movement Disorders

Transcranial direct current stimulation for treatment of freezing of gait in Parkinson's disease. A cross-over study

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Background: Progression of Parkinson's disease (PD) is frequently characterized by the occurrence of freezing of gait (FOG), a disabling motor complication which generally does not improve with dopaminergic therapy. Recent evidence shows that non-invasive brain stimulation techniques, such as repetitive transcranial magnetic stimulation (rTMS) and transcranial current direct stimulation (tDCS), could improve motor performances in PD patients.

Methods: In this cross-over, double-blind, sham-controlled study we investigated the safety and efficacy of tDCS of the primary motor cortex of ten PD patients with FOG in "on" state. The patients underwent anodal and sham stimulation in a randomized balanced order for five consecutive days. Clinical assessment over a 1-month period included the revised version of the UPDRS (MDS-UPDRS), Stand Walk Sit (SWS) test, video-analysis of gait, Freezing of Gait Questionnaire (FOG-Q) and Gait and Falls Questionnaire (GFQ).

Results: A significant improvement of MDS-UPDRS score and gait, with reduction in the number and duration of FOG episodes, was observed in patients who received anodal tDCS as compared to the sham intervention. The beneficial effects were observed after the first tDCS intervention, became more evident after the end of the whole session, and persisted for all the observation period.

Conclusion: Anodal tDCS of the motor cortex is safe and has therapeutic potential in PD patients with FOG in "on" state. Induction of dopamine release in the basal ganglia and modulatory effects on abnormal patterns of cortical activation in PD may account for the observed effects.

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Abstract – WCN 2013

No: 2409

Topic: 2 – Movement Disorders

Myocardial 131 I-meta-iodobenzylguanidine (MIBG) scintigraphy in the differential diagnosis of Parkinson's disease

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The aim of our investigation is to evaluate how myocardial 131 I-meta-iodobenzylguanidine (MIBG) scintigraphy may assist in the establishment of final diagnosis in PD.

Materials and methods: Eighteen patients clinically diagnosed as idiopathic PD (IPD) and fourteen patients with extrapyramidal symptoms (EPS) underwent MIBG scintigraphy. Planar chest MIBG imaging was performed 15 min, 2, 3, 4 and 24 h after tracer injection. Myocardial MIBG activity was quantified by means of a heart to mediastinum ratio (H/M ratio). A ratio over 1.8 was considered normal, between 1.31–1.79 decreased, and less than 1.3 severely decreased.

Results showed that the H/M ratio from early and delayed MIBG scintigraphy was pathologically decreased (<1.3) in fourteen patients diagnosed as IPD and in six patients with EPS. One patient with EST had decreased H/M ratio (1.3–1.79). Three patients initially diagnosed with IPD as well as seven patients with EPS had normal H/M ratios. In total, we confirmed initially established clinical diagnoses of IPD in 14 of 18

patients and changed the final diagnosis from EPS to PD in 7 of 14 patients.

Conclusions: According to our findings, it can be concluded that cardiac dysautonomia is common in IPD, confirming that myocardial MIBG imaging can be a powerful but simple and affordable method in differential diagnosis of PD, thus greatly assisting in diagnosing impaired adrenergic activity present in PD. Also, myocardial MIBG imaging can be of great help in establishing the final diagnosis in patients with EPS, especially when DaTSCAN is not available.

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Abstract – WCN 2013

No: 2342

Topic: 2 – Movement Disorders

DaTSCAN™ (ioflupane I 123 injection), a radiopharmaceutical indicated for visualization of the striatal dopamine transporter using SPECT: Safety profile review

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Objective: To assess DaTSCAN™ ([¹²³I]ioflupane injection) safety data from clinical trials (Phases I–IV) in clinical development and post-marketing experience.

Patients and methods: In GE-sponsored clinical trials, DaTSCAN™ was administered to 1236 subjects (1171 patients and 65 healthy volunteers). Clinical trials collected adverse events (AEs), laboratory parameters, vital signs and electrocardiograms (ECG). Administered activity of [¹²³I]ioflupane ranged from 3 to 5 mCi (111 to 185 MBq), comparable to administered activities for other commercially available ¹²³I-labeled products. The calculated whole body effective dose was 4 to 6 mSv, comparable to 1 year of natural background radiation (3 to 4 mSv).

Results: Review of trial data on non-serious AEs, vital signs, laboratory parameters, and ECG identified no safety issues. There were mild and infrequently reported AEs and no SAEs or deaths that were considered related to DaTSCAN™ administration. The most common AE ascribed to DaTSCAN™ by the investigator was headache (1%), followed by nausea, and vertigo, dry mouth, hunger, dizziness, and formication (<1% each). Most of these AEs were mild. The safety profile established in clinical trials is supported by limited AE reports (including hypersensitivity reported as rash and pruritis shortly after dosing) from post-marketing exposure in over 300,000 patients who have been imaged.

Conclusions: Comprehensive review of the safety data from clinical trials and 11 years of post-marketing use show that AEs associated with DaTSCAN™ are mild and infrequently reported, and DaTSCAN™ is a safe product to use. DaTSCAN™ is also considered to be low risk from a radiation dosimetry perspective.

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Abstract – WCN 2013

No: 2346

Topic: 2 – Movement Disorders

Gating of sensory evoked potentials in upper limb dystonia

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Background: Primary focal hand dystonia is a condition characterized by abnormal postures in the limb. Previous studies have identified abnormalities of sensory discrimination, worsening of dystonic postures by vibration of the limb and deficits in sensori-motor integration.

Objective: To compare the normal attenuation (gating) of somatosensory evoked potentials (SEPs) caused by movement in healthy people with that of patients with dystonia. We were also interested in the impact of vibration on SEP attenuation in healthy people and those with dystonia.

Methods: 6 patients with focal/segmental dystonia, 7 healthy controls. SEPs were obtained by stimulating the median nerve at the onset of a voluntary self-paced movement of the right thumb and at rest. We also recorded SEPs at the onset of movement while the subject was also receiving vibration on the palmar surface of the hand.

Results: SEP amplitude at the onset of movement was suppressed in controls compared to baseline SEP amplitude (ratio SEP amplitude at onset to baseline SEP amplitude = 0.67). However SEPs were not suppressed in patients with dystonia (ratio: 0.97). When the limb was vibrated during movement we did not find SEP suppression in either group: healthy controls' ratio: 0.92, dystonia patients' ratio: 1.1.

Conclusion: These preliminary results suggest that sensory afferents from the dystonic limb are not attenuated during movement. Vibration causes reduction in SEP suppression in both healthy people and in patients with dystonia. These data provide new information with regard to deficient sensori-motor integration in dystonia and a possible mechanism for the known worsening of dystonia caused by vibration of the limb.

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Abstract – WCN 2013

No: 2362

Topic: 2 – Movement Disorders

Impulse generator replacement in deep brain stimulation

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Background: Deep brain stimulation (DBS) has become an option for several movement disorders refractory to medical therapy. The finite life of non-rechargeable batteries powering implantable pulse generators (IPG) implies a periodic replacement. Patients undergoing DBS may require multiple substitutions and recent studies point to an increased rate of infection compared to the primary surgical procedure.

Objective: Determine the average duration of the pulse generator battery; analyze the rate of complications associated with the replacement procedure.

Material and methods: Retrospective review of medical and surgical records of 74 consecutive patients who underwent DBS and at least one impulse generator replacement in the context of “end of life” of the battery (between 2005 and 2012).

Results: 74 patients (95.9% Parkinson's disease, 4.1% secondary generalized dystonia) underwent surgery to replace the IPG (Kinetra/Activate PC-Medtronic®); 5 patients made two substitutions. The average duration of the batteries was 52.8 ± 3.2 months (12–82 months). There were no significant differences in the duration of subsequent vs primary generators. In 4 patients, a system infection occurred with an infection rate of 5.1%. In all cases it was necessary to remove the DBS system completely.

Discussion: Ten years after the beginning of DBS as a therapeutic option in Portugal, it became necessary for us to evaluate the complications associated with impulse generator replacement. At present, the possible use of rechargeable batteries in order to avoid the complications

inherent to a new procedure is discussed. In our center, the rate of infectious complications of replacement procedure is less than 6%.

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Abstract – WCN 2013

No: 2377

Topic: 2 – Movement Disorders

Clinical abnormalities in Niemann–Pick type C-carriers – A family case report

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Background: Niemann–Pick type C (NPC) is an autosomal recessive, neurodegenerative lysosomal storage disorder. It occurs at a frequency of approximately 1:120,000 live births, but is probably underdiagnosed, especially in the case of adult manifestation. NPC disease shows variable clinical pictures (neurological, psychiatric and systemic symptoms) and manifestations in different age groups. In teenagers and adults, dementia and psychiatric presentations may overshadow motor dysfunction. Vertical supranuclear gaze palsy (VSGP) is a very specific sign, but often missed, if the oculomotor system is not examined carefully. There have been no investigations about clinical abnormalities in heterozygote carriers of NPC so far.

Objective: In our family case presentation we report neurological, psychiatric and systemic symptoms in NPC carriers.

Patients and methods: Family members of a patient with genetically proven NPC underwent physical examination. Additionally, genetic and neuropsychological testing was performed.

Results: The father of the patient showed mild bradykinesia and hepatomegaly. The mother suffers from cervical dystonia and anxiety disorder. A grandmother of the patient is in treatment for paranoid dementia. The sister is healthy so far. All of them are NPC carriers.

Conclusion: There is a possibility that NPC carriers show mild neurological, psychiatric and systemic symptoms. Further investigations are necessary and would improve the understanding of the pathophysiology of NPC.

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Abstract – WCN 2013

No: 2192

Topic: 2 – Movement Disorders

Opioid treatment is effective in patients with severe RLS after failure of previous medications – Results of a 1-year study

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Background: Opioid use may provide a new treatment approach for patients with severe restless legs syndrome (RLS) after failure of mainly dopaminergic medications.

Objective: The primary objective was to demonstrate the superior efficacy of oxycodone/naloxone prolonged-release fixed-combination (OXNPR) versus placebo in reducing RLS symptom severity using the International RLS Study Group Rating Scale (IRLS).

Patients and methods: After a 7-day washout, patients (N = 304) with severe RLS (IRLS total score ≥ 21) inadequately controlled on previous medication were randomised to OXNPR bid (mean oxycodone dose 21.9 ± 15.0 mg/day) or placebo in the 12-week double-blind study. 197 patients participated in a 40-week open-label extension (mean oxycodone dose 18.1 ± 10.5 mg/day).

Results: Superior efficacy of OXNPR versus placebo was shown (clinically relevant treatment difference in IRLS total score at Week 12 of 8.15 units (95% CIs [5.46; 10.85]; $p < 0.001$, $n = 276$ full analysis population). Beneficial effects of OXNPR on RLS symptom severity were observed within 1 week and continued over long-term. Mean IRLS total score under OXNPR was 31.6 ± 4.5 at randomisation (indicating severe symptom load), reducing to 21.0 ± 9.1 at Week 1, 15.1 ± 10.6 at Week 12, and 9.7 ± 7.8 (representing mild RLS symptom severity) at end of extension ($n = 152$). Results of the IRLS were supported by significant improvements in RLS-6 Rating Scale and CGI responder rate results for OXNPR versus placebo, which were maintained for over 1 year. OXNPR treatment was well tolerated in accordance with the expected safety profile. No case of augmentation was reported during the study.

Conclusion: Prolonged-release oxycodone/naloxone is an effective and well-tolerated short- and long-term treatment for patients with severe RLS.

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Abstract – WCN 2013

No: 1502

Topic: 2 – Movement Disorders

Efficacy, safety and tolerability of rasagiline as an add-on therapy to dopamine agonists in early Parkinson's disease: The ANDANTE study

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Background: Dopamine agonists (DAs) are often used as initial PD monotherapy. However with disease progression DA monotherapy alone becomes sub-optimal and patients often require additional dopaminergic therapy to maintain symptomatic efficacy. Traditional options at this stage include levodopa (associated with risk of dyskinesia) or increasing DA dose (associated with higher risks for ICDs and other AEs). Rasagiline is a selective, irreversible MAO-B inhibitor providing a distinct rationale as add-on therapy to DAs for additional symptomatic benefit.

Objective: Determine the efficacy, tolerability and safety of add-on rasagiline therapy for early-PD patients sub-optimally controlled by DA monotherapy.

Methods: 18-weeks, placebo-controlled (randomized 1:1) study of PD patients (Hoehn & Yahr 1–3) aged ≥ 30 years taking stable DA dosages with suboptimal control. Primary outcome: change from baseline in total-UPDRS score. Secondary outcomes: changes from baseline in UPDRS motor and ADL scores and CGI. Safety was assessed by AE frequency/severity. Out of 328 patients randomized, 321 (mean age 62.6; duration PD 2.13 years) were included in the efficacy analysis.

Results: Treatment with add-on rasagiline significantly improved total-UPDRS scores versus placebo (treatment effect \pm SE: -2.4 ± 0.95 ,

$p = 0.012$). Significant improvements in UPDRS-motor scores were also observed ($p = 0.007$). There were no significant differences between groups for ADL or CGI scores. Rasagiline was well-tolerated, with no significant difference in percentage of patients with AEs (64.2% vs. 61.0%) or serious AEs (4.9% vs. 3.0%) versus placebo. Only 11 patients required rescue levodopa during the study.

Conclusions: Addition of rasagiline significantly improved motor symptoms in patients sub-optimally controlled with DA monotherapy, and was well-tolerated.

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Abstract – WCN 2013

No: 2183

Topic: 2 – Movement Disorders

Oxycodone/naloxone prolonged-release has positive quality of life outcomes in patients with severe RLS – Results of a 1-year study

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Background: Off-label use of opioids for severe restless legs syndrome (RLS) after failure of mainly dopaminergic medications is common, but multicenter trials are lacking.

Objective: Efficacy and safety of oxycodone/naloxone prolonged-release fixed-combination (OXNPR) versus placebo in RLS. The primary endpoint was the International RLS Study Group Rating Scale (IRLS). Disease-specific QoL was assessed using the QoL-RLS-Scale as the secondary endpoint.

Patients and methods: Patients (N = 304) with severe RLS (IRLS total score ≥ 21) inadequately controlled on previous medication were randomised to OXNPR bid (mean oxycodone dose 21.9 ± 15.0 mg/day) or placebo for 12 weeks, after a 7-day down-tapering phase. 197 patients participated in a 40-week open-label extension (mean oxycodone dose 18.1 ± 10.5 mg/day). The QoL-RLS-Scale comprised 12 questions, each scored on a 6-point scale.

Results: Median scores at baseline (mostly 4) reflected a moderate impairment of QoL in RLS patients. A positive effect of treatment with OXNPR was seen across all 5 dimensions of burdens from RLS, including effects of RLS symptoms, effects of disturbed sleep, efforts to handle RLS symptoms, effect of pain caused by RLS and overall QoL, with median scores of 2 to 3 for all questions at Weeks 4 and 12 (indicating little remaining QoL impairment from RLS symptoms). The treatment difference was statistically significant in favor of OXNPR in the QoL-RLS total score ($p \leq 0.003$; ANCOVA). Further small improvements in the QoL-RLS were seen during the extension phase.

Conclusion: Prolonged-release oxycodone/naloxone is an effective treatment for severe RLS, with associated improvements in patients' burdens of daily activities and QoL.

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Abstract – WCN 2013

No: 2250

Topic: 2 – Movement Disorders

Cognitive deficits in multiple system atrophy (MSA): Comparison with sporadic adult onset ataxias of unknown aetiology (SAOA) and longitudinal decline

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Multiple system atrophy (MSA) is a sporadic, progressive, adult-onset neurodegenerative disorder characterized clinically by a variable combination of autonomic, parkinsonian, cerebellar, or pyramidal signs. Sporadic adult onset ataxias of unknown aetiology (SAOA) denote all non-hereditary degenerative adult onset ataxias distinct from MSA. SAOA is less progressive, but early differential diagnosis is often difficult. We compared cognitive functions in both disorders and studied the putative progression of cognitive dysfunction in MSA over time.

At baseline, 25 MSA patients (male 17, age 60.8 ± 6.8 years (y), disease duration (dd) 3.6 ± 1.8 y), 21 SAOA patients (male 8, age 51.5 ± 13.1 y, dd 7.3 ± 5.4 y) and age-matched control groups were neuropsychologically assessed and statistically compared (multivariate analysis of covariance, MANCOVA). MSA patients and controls were in part re-assessed at annual follow-up (repeated MANOVA).

MSA and SAOA patients had significant deficits in reasoning, word fluency, visual thinking, working memory, reaction time, choice reaction time and interference compared to controls. With dd as moderator MSA patients significantly differed from SAOA patients in interference and as a tendency also in working memory. During a follow-up of 15.7 ± 7 months, no decline was observed in the MSA patient group ($n = 13$). Cognitive dysfunction in both patient groups comprised mainly executive functions allocated to the frontal lobe. In spite of partially different neuropathology, MSA and SAOA had similar deficits, but MSA patients seem to have these deficits earlier in disease course than SAOA. Cerebellar atrophy, shared by both, may contribute via cerebello-cerebral disconnections to an impaired supportive role of the cerebellum in cognitive function.

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Abstract – WCN 2013

No: 2267

Topic: 2 – Movement Disorders

Non-motor manifestations in Parkinson's disease: 5 year follow-up

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Background: Parkinson's disease (PD) is often associated with numerous psychiatric and behavioral disorders and mild cognitive impairment and dementia.

Apathy, anxiety and depression are among the most frequently reported neuropsychiatric features in PD. The point-prevalence of dementia is 30% and patients with PD have an almost sixfold increased risk of developing dementia compared to healthy controls.

Objective: The objective of this study was to examine the prevalence and clinical correlates of behavioral changes, mild cognitive impairment and dementia in a clinical population-based sample of patients with Parkinson's disease.

Methods: A series of 360 PD patients; the assessment included comprehensive neurological examinations (motor scoring with Hoehn and Yahr staging and the Unified Parkinson's Disease Rating Scale); psychiatric examinations with the Apathy Scale (AS), Hamilton Depression Rating Scale (HDRS-17), Hamilton Anxiety Rating Scale (HAMA-S); and cognitive screening with the Mini-Mental State Examination (MMSE) and Clinical Dementia Rating (CDR) scale.

Results: Apathy coexisted with depression in 133 (36.9 %) of PD patients, depression without apathy in 16 (4.4 %), apathy without depression in 84 (23%), and neither apathy nor depression in 127 PD patients (35.2%). A total of 172 patients were cognitively intact; whereas 114 (36%) were diagnosed with MCI. Thirty patients (9.4%) were diagnosed with dementia.

Conclusion: These findings suggest that apathy and depression may be separable in PD, although both are common in patients with PD; and that PD patients with more prominent axial signs and older age are in higher risk for developing cognitive impairment.

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Abstract – WCN 2013

No: 2102

Topic: 2 – Movement Disorders

Non-motor features and motor complications in IPD: Are UPDRS parts I and IV relevant evaluation tools?

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Background: The UPDRS is the most widely used scale in Parkinson's disease. However despite the fact that non-motor symptoms and motor complications are common in IPD with prevalence figures of up to 98% and 40%, parts I and IV which focus on these symptoms were previously infrequently used.

Objective: As during the last decade research has particularly focused on and has made progress in elucidating various non-motor features and motor complications of PD we examined whether this reflects now in an elevated use of the respective UPDRS parts.

Methods: A systematic literature search using the database "PubMed" to identify randomized clinical trials on IPD, published between 1998 and 2011 was performed. Identified abstracts were hand searched and analysed regarding usage of clinical scales and particularly UPDRS and its parts.

Results: Almost all of the 163 studies (97.6%) that had included the UPDRS applied part III alone or in combination with other parts. In contrast part I or part IV alone or in combination with another part (except part III) was utilized in only 0.6% and 1.8% respectively. One of these studies used part I of the MDS-UPDRS the revised version of the UPDRS.

Conclusion: Despite the recent focus of PD research there is still a big discrepancy between high prevalence of non-motor features and motor complication and low frequency of use of respective UPDRS parts. It seems that for reason of comparability of results researchers when investigating these symptoms are better off in using other evaluation tools.

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Abstract – WCN 2013

No: 2326

Topic: 2 – Movement Disorders

POLG-1 related SANDO and levodopa-responsive parkinsonism

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Background: Mitochondrial dysfunction has recently emerged as one of the relevant mechanisms associated with the etiology and pathogenesis of neurodegenerative parkinsonism. Mutations in *POLG1*, the nuclear gene encoding the catalytic subunit of mitochondrial DNA polymerase gamma, have been associated with a wide phenotypic spectrum, including inherited parkinsonism usually in combination with progressive external ophthalmoplegia.

Clinical case: A 52-year-old man presented with progressive bilateral ptosis, unsteadiness of gait and muscle weakness at the age of 39 and developed dysphagia and diplopia three years later. At age 44, neurological examination revealed fatigable bilateral blepharoptosis, external ophthalmoparesis with diplopia on horizontal gaze, fluctuant dysarthria and dysphagia. Peripheral neurophysiologic examination was compatible with axonal sensory neuropathy. Sequencing of the *POLG1* gene revealed compound heterozygotic missense substitutions: p.P648R and p.R807C. This case was initially reported as a typical SANDO syndrome. During the last 3 years, the patient developed right-dominant parkinsonism, with hand rest tremor, moderate limb bradykinesia, cogwheel rigidity and hypomimic face. ¹²³I-ioflupane DaTscan demonstrated a bilaterally reduced dopamine uptake in the corpus striatum. The patient was treated with coenzyme Q 30 mg tid, levocarnitine 1 g tid and levodopa (up to 400 mg/day). At 3 years of follow-up, he sustains a positive dopaminergic response, with a decrease of 17 points in the MDS-UPDRS-III score (53% improvement) after a 300 mg l-dopa challenge test.

Discussion: To our knowledge, this is the first case of parkinsonism reported in *POLG1*-positive SANDO patients. We think that this case expands the clinical and molecular findings of *POLG1*-related parkinsonism to include SANDO phenotype.

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Abstract – WCN 2013

No: 96

Topic: 2 – Movement Disorders

Neuroprotective effects of 17-beta-estradiol: Relevance to depressive symptoms in Parkinson's disease

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Parkinson's disease (PD) is a movement disorder characterized by the loss of dopaminergic neurons in the substantia nigra causing dopamine depletion.

The objective of this study was to investigate the neuroprotective potential of 17β estradiol (E2) treatment on the activities of acetylcholinesterase (AChE) and monoamine oxidase (MAO), membrane fluidity, neurolipofuscin, genomic DNA degradation, protein oxidation levels and testing learning memory, occurring in the brains of female rats of the 3 month (young), 12 month (adult) and 24 month (old) age groups, and to see whether these changes are restored to normal levels after exogenous administration of estradiol.

The aged rats (12 and 24 months old) (n = 8 for each group) were given subcutaneous injection of 17β-estradiol (0.1 μg/g body weight) daily for one month. After 30 days of hormone treatment, the experimental animals of all groups were sacrificed and the brains were isolated for

further study. Learning was tested in a Morris water maze and ultrastructural studies of brain region by MRI.

The results obtained in the present work revealed that normal aging was associated with significant increases in the activity of MAO, genomic DNA degradation and protein oxidation levels in the brains of aging female rats, and a decrease in AChE activity and membrane polarization. It can therefore be concluded that E2's beneficial effects seemed to arise from its antioxidant and antilipidperoxidative effects, implying a therapeutic potential drug for age related changes. Based on our studies and those of others, we conclude that E2 has therapeutic potential for adjunctive therapy of PD.

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Abstract – WCN 2013

No: 1913

Topic: 2 – Movement Disorders

Initiation of pharmacological treatment in Parkinson's disease in the UK: A Clinical Practice Research Datalink retrospective study

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Background: In UK guidelines, levodopa (LD), dopamine agonists (DA) and monoamine-oxidase-B (MAO-B) inhibitors are recommended as initial options for the pharmacological treatment of Parkinson's disease (PD). The choice for these treatments should take into account patient clinical profile, lifestyle characteristics and patient preferences.

Objective: To describe patient profile according to the initial treatment for PD and describe early treatment patterns using general practitioner medical files.

Material and methods: Retrospective database analysis in the UK using the Clinical Practice Research Datalink (CPRD). Medical records of patients aged 40 years or above, with an incident prescription for a PD drug between 2005 and 2011 were extracted. Patient characteristics, comorbidities, healthcare provider and second-line treatment were described per treatment group.

Results: 9231 fulfilled the inclusion criteria; 58% were male and the mean age was 74 years (SD 10). The majority of patients started with LD monotherapy (74%), followed by DA (14%), and MAOB-I (5%). The proportion initially treated with LD was stable over time, while it increased for rasagiline and slowly decreased for selegiline and DA. 81.4% of patients were 70 years or more in LD group, vs. 31% for DA, 40% for rasagiline and 57% for selegiline patients. When second-line treatment was initiated, it was mainly addition of LD or DA in proportions of 40/41% respectively for rasagiline, 52/32% for selegiline and 66%/na for DA.

Conclusion: This retrospective analysis shows that patient profiles differ according to index drug and confirms that age is a key characteristic in the choice for PD treatment.

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Abstract – WCN 2013

No: 2275

Topic: 2 – Movement Disorders

Functional connectivity in early Parkinson disease: A resting-state fMRI study

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Background: Parkinson disease (PD) is characterized by dopamine depletion in the nigrostriatal system that leads to an abnormal modulation of cortico-subcortical networks.

Objective: This study aims to evaluate the functional connectivity (FC) of the basal ganglia circuit in early PD.

Methods: We enrolled early, treated (tPD = 44) and drug-naïve (nPD = 26) PD patients and 27 healthy controls. Considering that all PD showed lateralized parkinsonian features, the images from 24 patients whose symptoms were on the left (L) side were left–right (R) flipped so that the hemisphere contralateral to the affected limbs was on the L for all patients. A seed-based FC analysis from resting-state data was performed using bilateral caudate, putamen, pallidum and thalamus as seed regions of interest ($p < 0.05$, multiple comparison corrections).

Results: In nPD patients, caudate and putamen showed increased connectivity with their contralateral homologous regions, R-thalamus showed increased connectivity with L-thalamus, and L-caudate and L-thalamus presented decreased connectivity with homolateral prefrontal regions. tPD patients showed reduced connectivity between L-caudate and anterior cingulum, while an increased connectivity was found between R-putamen and the contralateral one. In both nPD and tPD, L-thalamus was hypoconnected with homolateral prefrontal regions. Only in tPD patients, L-thalamus was hyperconnected with contralateral sensorimotor cortices.

Conclusions: nPD patients and tPD showed different connectivity profiles compared to controls, a finding which raises the hypothesis that L-DOPA treatment might play a role in the modulation of brain connectivity.

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Abstract – WCN 2013

No: 2259

Topic: 2 – Movement Disorders

Functional connectivity in patients with focal dystonia

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Background: Primary dystonia is a hyperkinetic movement disorder which has been traditionally conceptualized as a disorder of the basal ganglia. However, recent data suggest a widespread brain pathology involving motor control pathways.

Objective: This study aims to investigate the functional connectivity of the sensorimotor and bilateral frontoparietal networks in patients with focal dystonia using resting state (RS) functional MRI (fMRI).

Methods: Fifty patients with dystonia and 36 healthy controls were studied. Patient group included 17 subjects with blepharospasm (mean Blepharospasm Disability Index 10.6), 15 subjects with spastic torticollis (mean Toronto Western Spasmodic Torticollis Rating Scale 22) and 18 subjects with writer's cramp (mean Writer's Cramp Rating Scale—part B 10.7). RS fMRI data were analyzed using a model free (MELODIC) approach in FSL ($p < 0.05$ family-wise error).

Results: Compared with controls, patients with blepharospasm showed an increased functional connectivity of the sensorimotor network in the supplementary motor area bilaterally and left primary sensorimotor cortex, frontal eye field, and superior parietal lobule. Blepharospasm showed no RS fMRI changes in the frontoparietal network. Patients with writer's cramp showed a decreased functional connectivity of the bilateral superior parietal gyri within the frontoparietal networks, while

they did not show altered functional connectivity of the sensorimotor network. No RS fMRI abnormalities were detected in patients with spastic torticollis.

Conclusions: This study showed specific patterns of altered functional connectivity in patients with focal dystonia. These findings add novel elements to the pathophysiological substrates of these conditions.

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Abstract — WCN 2013

No: 2244

Topic: 2 — Movement Disorders

The achievements of I-123 ioflupane (DatSCAN) in a diagnosis of parkinsonism

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Background: The diagnosis of various parkinsonian syndromes is primary clinical, but sometimes require more time to final diagnosis.

Objective: I-123 ioflupane brain SPECT is considered a biomarker for differentiating conditions with and without nigrostriatal neurodegeneration. Reduced striatal binding of I-123 ioflupane is in correlation with lower concentration of presynaptic dopamine.

Materials and methods: We performed a I-123 ioflupane brain SPECT using standard protocol in 218 patients clinically diagnosed to have different parkinsonian syndromes (84 pts with IPD, 102 pts with ET and 24 pts with other forms of parkinsonism). A dose of 185 MBq of 123I-ioflupane was administered and brain SPECT imaging was conducted 3–6 h after radiotracer injection. Images were analyzed visually and for this purpose patients were classified into two groups — with and without nigrostriatal deficit.

Results: We confirmed a clinical diagnosis in 40 patients (21.5%) clinically diagnosed as ET and in 68 patients (36.6%) clinically diagnosed as IPD. In 62 patients (33.3%) the clinical diagnosis of ET changed in condition with nigrostriatal neurodegeneration. In 16 patients (8.6%) clinically diagnosed as IPD we didn't find any striatal deficit. The group of 24 patients with other different forms of parkinsonism was excluded in this analysis.

Conclusion: Our experiences show that 123I-ioflupane brain SPECT has an important role in detecting and monitoring nigrostriatal dopaminergic neurodegeneration and could be routinely used in clinical practice to support or change the clinical diagnosis. Also, we emphasize that a good collaboration between neurologist and nuclear medicine physician is useful in reaching final diagnosis.

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Abstract — WCN 2013

No: 198

Topic: 2 — Movement Disorders

Mapping PDQ-39 and PDQ-8 scores onto EQ-5D utility index in patients with Parkinson's disease

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Background: Parkinson's disease (PD) is a chronic neurodegenerative condition which can severely affect patients' QOL. QOL in PD can be measured using disease-specific or generic measures.

Objective: This study aims to develop an algorithm for mapping of disease-specific QOL scores to the EQ-5D utility index for patients with PD.

Patients and methods: EQ-5D and PDQ-39/8 data were obtained for 117 UK PD patients (HY stages 1–4) via a cross-sectional survey of practicing clinicians. The EQ-5D utility index was regressed on PDQ-39/PDQ-8 summary indices using OLS regression. Analysis was repeated using domains of PDQ-39 or questions of PDQ-8. Mapping equations with and without adjustment for age and self-perceived disease status were used to predict utilities which were subsequently compared to those observed.

Results: All models were statistically significant. For unadjusted and adjusted models R² ranged from 0.57 to 0.69 and 0.63 to 0.72 respectively. Predictive power of the models using PDQ-39 domains and PDQ-8 questions was higher [RMSE 0.168 and 0.169 respectively] compared to those using summary indices [RMSE 0.192 and 0.191 respectively]. There was no statistically significant difference between the means of the observed and predicted EQ-5D utilities when any of the models were used.

Conclusion: All mapping algorithms estimated utilities for patients with HY stage 1–4 PD with good accuracy. Therefore mapping functions reported here could be used for predicting EQ-5D scores from either PDQ-39 or PDQ-8 when EQ-5D data was not collected. Further validation is needed to ascertain whether the mapping function performs well for patients in HY stage 5.

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No: 2255

Topic: 2 — Movement Disorders

Brain structural changes in primary focal dystonia

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Background: Studies using structural and diffusion tensor (DT) MRI reported inconsistent differences in grey (GM) and white matter (WM) relative to controls.

Objectives: This study aims to investigate if there are GM and WM abnormalities in patients with primary focal dystonia and if such changes are related to clinical features.

Methods: 3D T1-weighted and DT MRI scans were obtained from 51 patients with focal dystonia (17 blepharospasm [BL], 16 spasmodic torticollis [ST], 18 writer's cramp [WC]) and 36 healthy controls. Voxel-wise analyses of GM volume and WM microstructural abnormalities were performed.

Results: Compared with controls, BL patients showed decreased GM in the right precentral gyrus and rolandic operculum, bilateral postcentral gyri, cerebellum and right hippocampus. This pattern of GM atrophy was associated with clinical severity. ST patients compared with controls showed decreased GM density in the left precentral and postcentral gyri, and right rolandic operculum and lingual gyrus. A correlation was found between ST severity and GM density of the middle cingulate gyrus. Relative to controls, WC patients showed GM density increases in the right middle frontal gyrus and left superior frontal gyrus. DT MRI analysis showed a widespread pattern of WM damage in BL patients, a decreased fractional anisotropy in the right internal capsule in patients with WC, and no changes in ST.

Conclusions: Patients with dystonia exhibit GM and WM alterations in regions relevant to motor function, sensory processing, and cognitive

modulation of motor behaviour, which likely contribute to the clinical features of focal dystonia.

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Abstract – WCN 2013

No: 2220

Topic: 2 – Movement Disorders

Instrumented timed up and go test in progressive supranuclear palsy

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Background: To diagnose progressive supranuclear palsy (PSP) is challenging. Its cardinal features are postural instability and gait disturbance besides vertical gaze palsy. Instrumented timed up and go (ITUG) test was developed to evaluate gait and balance quantitatively.

Objective: Our aim was to find characteristic mobility parameters during ITUG-test, which may help the early diagnosis of PSP.

Patients and methods: Six patients with PSP (age: 66 ± 7.3 years; disease duration: 37 ± 12.1 months) and seven age-matched control subjects (C) performed ITUG-test consisting of gait, turning, sit-to-stand and turn-to-sit items three times. Six Opal monitors (APDM Inc.) with three-dimensional gyroscope and accelerometer were placed to the chest, trunk and limbs. Data were analyzed with Mobility Lab software and compared with Mann–Whitney test.

Results: The duration of the whole ITUG-test (median/quartile range: PSP: 29.1/19.1s, C: 18.4/4.37s; $p = 0.01$) and of the turning (PSP: 4.1/1.77s, C: 2.4/0.62s; $p < 0.01$) and turn-to-sit items (PSP: 5.5/0.88s, C: 3.6/1.30s; $p < 0.01$) were significantly longer in PSP compared to controls. Peak velocity of turning (PSP: $98.7/24.99^\circ/s$, C: $149.2/82.73^\circ/s$; $p < 0.01$) and of turn-to-sit (PSP: $93.8/25.95^\circ/s$, C: $148.5/72.33^\circ/s$; $p < 0.01$) and sit-to-stand item (PSP: $51.7/23.2^\circ/s$, C: $71.8/15.92^\circ/s$; $p = 0.05$) were significantly smaller in the PSP group. The range-of-motion of the trunk in the turn-to-sit item was smaller in PSP than in controls (PSP: $17/6.28^\circ$, C: $31.3/6.95^\circ$; $p = 0.02$).

Conclusion: During the whole ITUG-test we detected signs of bradykinesia. Parameters of sit-to-stand and turn-to-sit items may be influenced by the axial rigidity in PSP. Further studies are needed, if they may be early indicators of the disease.

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Abstract – WCN 2013

No: 1103

Topic: 2 – Movement Disorders

Patient survey evaluating botulinum toxin type A treatment for blepharospasm

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Introduction: Two recent patient surveys in cervical dystonia and post-stroke spasticity showed that satisfaction with botulinum toxin treatment varies over the treatment cycle. Patient satisfaction with botulinum toxin regimens in blepharospasm, which is a disabling condition that has a negative impact on patients' quality of life, has not yet been evaluated in a clinical study.

Patients and methods: This non-interventional survey has collected self-reported information from patients (aged 18–80 years) who have received ≥ 2 botulinum toxin treatments for blepharospasm at

5 medical centers in the USA. Information collected includes demographic and baseline disease characteristics and details of previous botulinum toxin treatments (injection intervals, physician's rationale for intervals used, time course of treatment effects, treatment satisfaction, and current health status). Treatment effects were evaluated via patient-reported Jankovic Rating Scale and Blepharospasm Disability Index scores at the time of interview and at the time of the peak effect of the last treatment cycle (as remembered by patients at the time of interview).

Results: By 27 March 2013, 123 patients had been recruited; the maximum expected enrollment is 130 patients (ClinicalTrials.gov identifier: NCT01686061).

Conclusion: This survey will provide valuable insight into patient needs and treatment satisfaction with current botulinum toxin treatment regimens, which will inform future management of blepharospasm and help improve outcomes.

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Abstract – WCN 2013

No: 2213

Topic: 2 – Movement Disorders

A clinical and imaging profile of 35 patients with movement disorders associated with infection

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Background: Movement disorders associated with infections remain an important debilitating disorder in Asian countries.

Objectives: This study aims to report the clinical and imaging profile of a large cohort of patients with movement disorders (MD) associated with infection.

Methods: This was a chart review of 35 patients (F:M=15:20) presenting with MD in the Neurology services of National Institute of Mental Health & Neurosciences, India. The demographic profile, type of infection, time from infection to MD, phenomenology of MD and MRI findings were reviewed.

Results: The mean age at presentation was 22.6 ± 13.3 years, (5–60), age of onset of MD was 15.7 ± 15 years, and duration of symptoms was 6.9 ± 8.1 years (42 days to 32 years). The mean latency of onset of MD after the infection was 5.9 ± 4.2 weeks. The phenomenology of MD includes the following:

- (i) pure dystonia—28.6%,
- (ii) dystonia with choreoathetosis—22.9%,
- (iii) parkinsonism—14.6%,
- (iv) pure tremor, hemiballismus, myoclonus and chorea—2.9% each, and
- (v) mixed MD—22.9%.

Most often the MD was generalized (60%), followed by right upper limb (31.4%) and left upper limb (8.6%). Encephalitis was the most common infection (85.7%) which was associated with MD. Abnormalities of brain MRI, seen in 79.2%, included signal changes in

- (i) thalamus—52.0%,
- (ii) putamen and subcortical white matter—16% each,
- (iii) pons—12%
- (iv) striatopallidum, striatum and gray matter—8% each, and
- (vi) caudate, cerebellum, lentiform nucleus, midbrain and subthalamic nucleus—4.0% each.

Conclusions: Movement disorders associated with infection were most often post-encephalitic. Dystonia was the most common movement disorder, and thalamus was the most common anatomical site involved.

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Abstract – WCN 2013**No: 1644****Topic: 2 – Movement Disorders****Motor and cognitive dysfunction in Parkinson's disease patients with impulse control disorders**

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Background: Impulse Control Disorders (ICD) are frequent and incapacitating disorders in Parkinson's Disease (PD).

Objectives: To investigate the relation between ICD and:

1. Cognitive dysfunction;
2. Motor phenotype;
3. Motor side effects of dopaminergic treatment.

Methods: ICD were diagnosed with the Questionnaire for Impulsive-Compulsive Disorders. Patients with at least one type of ICD were compared to a group of PD patients without ICD, matched for gender, education age, age of onset, duration of disease, Hoehn and Yahr stage and dopamine equivalent doses (DED). Patients were assessed with the UPDRS and the following neuropsychological tests: FAS test, Stroop test, digit span test, Frontal Assessment Battery, Mini-Mental State Examination, Rey Auditory Learning Verbal Test, semantic fluency test, and Raven Colored Progressive Matrices.

Results: We selected 14 patients with ICD, 6 with more than one type. ICD and non-ICD (n = 14) patients did not differ significantly regarding neuropsychological tests, motor scores or the prevalence of motor complications of dopaminergic treatment. Patients with multiple ICD presented with a significantly (p = 0.002) higher prevalence of motor fluctuations (4, 67%) than patients with one type of ICD (0) and patients without ICD (1, 7%). They also had significantly longer disease duration, higher motor stage and DED.

Discussion: Our results suggest that ICD are not related to cognitive dysfunction or the non-tremor motor subtype. Multiple ICD could represent a worse behavioral phenotype, bearing a stronger relation with motor fluctuations and a higher susceptibility to chronic dopamine stimulation. However, differences in DED and disease duration could have biased these results.

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Abstract – WCN 2013**No: 537****Topic: 2 – Movement Disorders****Latent trigger point platelet rich plasma (PRP) injections under ultrasound guidance for chronic myofascial pain**

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Background: Dry needling (DN) under ultrasound guidance is an effective method for trigger point (MTrP) inactivation. Latent MTrPs, local dystonia, insensitive to DN, cause postural imbalance, decrease

long term outcome. Injections of platelet rich plasma (PRP) may become beneficial method for treatment latent MTrP. The aim was to test feasibility of PRP therapy for incurable latent MTrP.

Materials and methods: We included 20 patients with chronic myofascial pain, with poor results after four DN sessions. All patients were randomly assigned to dry needling therapy, either to PRP injections under ultrasound guidance. Latent MTrPs were visualised as stiff, hyperechoic areas. We evaluated pain according to visual analogue scale data, movement volume, activity of MTrP and US criteria of muscle restoration at 1, 7, 28 days after start of treatment.

Results: Pain relief after DN was revealed superior vs group B (P < 0.01). Level of local twitch response was elicited in 33% of patients after DN versus 19% in group B (P < 0.05). Restoration of muscle structure and elimination of MTrP were registered after 7 days after PRP injection. After 28 days, movement volume increased by 40% after PRP versus 25% after DN (P < 0.05), pain and MTrP recurrence was significantly lower in group A: 36 ± 2% compared to 58 ± 3% in group B (P < 0.05).

Conclusions: Injections of PRP under ultrasound guidance are effective for inactivation latent MTrP, improve long term outcome, and can be used as supplement for myofascial pain treatment.

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Abstract – WCN 2013**No: 1125****Topic: 2 – Movement Disorders****Effects of active-assisted cycling on motor function and balance in Parkinson's disease**

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Background: Parkinson's disease (PD) is a progressive neurodegenerative disorder that results in tremor, rigidity, bradykinesia and postural instability. With the progressive nature of PD, it is critical to identify optimal rehabilitative interventions to minimize disease symptoms. We have recently developed an active-assisted cycling (AAC) intervention, using a commercially available motorized cycle trainer, which is well tolerated by individuals with PD.

Objective: This study aims to determine if a four-week interval active-assisted cycling (AAC) protocol improves motor function and balance in individuals with PD.

Patients and methods: Individuals (N = 27) were randomly assigned to an AAC or a control group. The AAC group completed 12 sessions over four weeks on the Motomed Viva 2 Parkinson movement trainer. Interval AAC included bouts of cycling where the motor assisted individuals to pedal up to 90 revolutions per minute. The control group did not cycle but completed functional assessments at baseline and after 4 weeks. Assessments included motor function (UPDRS Motor III) and balance (Biodex Balance System SD) testing.

Results: UPDRS motor function scores improved in the AAC condition but not in the control group. Specifically, bradykinesia and tremor were reduced after this intervention. Limited effects on balance were noted.

Conclusion: A four week interval active-assisted cycling intervention improves UPDRS scores but does not affect balance in individuals with PD. However, these results suggest that interval AAC could be an important adjunct to medication for improving motor function in Parkinson's disease.

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Abstract – WCN 2013**No: 1869****Topic: 2 – Movement Disorders****Parkinson disease's epidemiology in Goiás and Brazil**H.H.S. Matozinho, N.S. Dias, M.O.F. Iwamoto, T.A.G.J. Ribeiro, F.O. Gomes.
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Background: Parkinson Disease (PD) is a result of complex interaction between genetic and environmental factors. It is characterized by 4 clinical signs: resting tremor, rigidity, bradykinesia, instability, gait and postural difficulty.

Objectives: This study aims to check the prevalence of PD in Goiás, comparing it with Brazil and data in literature.

Methods: Analysis of the numbers of PD hospitalizations through the Unified Health System (UHS), focusing on aspects as: gender, age, race, year of internalization (2000–2011), in Goiás and Brazil. The Hospital Information System (HIS) – Unified Health System Informatics Department (UHSID) provided data.

Results: The gender number of hospitalizations in Goiás was: 57% 'male' and 42% 'female'; Brazil: 51% 'male' and 48% 'female'. The most common ages affected with PD in Goiás were: 30% '70–79 years old', 26.5% '60–69 years old', 23.5% 'above 80 years old'; Brazil: 32% '70–79 years old', 24.4% '60–69 years old', 19.4% 'above 80 years old'. The race prevalence for PD in Goiás was: 44% white and 15% grayish-brown; Brazil: 50% white and 12.5% grayish-brown.

Conclusion: Although some articles say that PD is more prevalent among men, this study found no prevalence difference between genders in Goiás, neither in Brazil. The most common affected ages and races by PD in literature are the same found in this study.

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Abstract – WCN 2013**No: 2111****Topic: 2 – Movement Disorders****Consideration of mechanisms of muscular rigidity in Parkinson's disease**M. Nagaoka^a, N. Kakuda^b, Y. Hayashi^a, G. Futatsubashi^c, T. Fukushima^a, K. Watanabe^a. ^a*Rehabilitation Medicine, Juntendo University, Tokyo, Japan;* ^b*Neurology, Higashiyamato Hospital, Tokyo, Japan;* ^c*Comprehensive Educational Science, Tokyo Gakugei University, Tokyo, Japan*

Background: Rigidity is one of the cardinal symptoms in Parkinson's disease. Based on the findings of microneurography, Burke D et al. (1977) suggested that, in Parkinson patients, the spindle afferent responses look like those of normal subjects who are not fully relaxed and in whom there is a co-activation of the skeletomotor and fusimotor activity. In previous paper (J Physiol, 1998), we showed the existence of dynamic and static fusimotor actions on the human muscle spindle during voluntary contraction.

Objective: In this experiment, we tested whether increased spindle activities during weak isometric contraction of forearm muscles may enhance the stretch reflex as seen in Parkinson's disease.

Subjects and methods: Thirteen healthy subjects (8 males and 5 females, age between 26 and 50 years old) were recruited. The details of methods were the same as appeared in previous paper. A tungsten micro-electrode was inserted into the left radial nerve and spindle activities were recorded from extensor carpi radialis brevis (ECRB). The wrist joint was moved 25° with 5–40°/s by DC-Torque motor. Spindle activities and EMG were recorded and analyzed by SC/ZOOM system (Umeå University).

Results: Total number of 43 spindles was recorded from ECRB. All spindles increased the firing rate by lengthening of ECRB. There was

no stretch reflex in ECRB at rest and during isometric contraction except one subject.

Conclusion: These findings suggest that increased stretch reflex was exceptional in normal subjects even with α - γ co-activation. There might be substantially increased excitability in α motoneurons in Parkinson's disease.

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Abstract – WCN 2013**No: 323****Topic: 2 – Movement Disorders****Pathophysiological analysis of dropped head syndrome caused by various diagnoses – Based on surface EMG findings and responses to physiotherapy**M. Nagaoka^a, H.-N. Lin^a, Y. Hayashi^a, I. Yonezawa^b. ^a*Rehabilitation Medicine, Juntendo University, Tokyo, Japan;* ^b*Orthopedic Surgery, Juntendo University, Tokyo, Japan*

Background: Dropped head syndrome is seen in various diseases. Its pathophysiological mechanisms are unknown.

Patients and methods: We investigated its pathophysiological mechanisms with physical and radiological examinations including MRI, surface EMG and responses to physiotherapy. Subjects had dropped head as a complaint, but their primary diagnoses were various. We investigated 16 cases: 10 cases of Parkinson disease and related disorders, 3 cases of cervical spondylosis and 3 cases with other diagnoses.

Results: We found that patients had common findings such as bulging of cervical muscles, and tonic EMG activities mainly in the extensors in the sitting and standing position, but in the flexors of the neck only in the supine position. Of the 16 cases, 14 were treated with physiotherapy to improve the alignment of the pelvis and whole vertebral column; 6 of the 14 cases (42%) showed remarkable improvement.

Conclusion: We conclude that the primary reason of dropped head syndrome is unknown in Parkinson disease and cervical spondylosis, but also that many of the patients have secondary changes in alignment of the skeletomotor system which could be treated with physiotherapy. For 8 cases with no improvement by physiotherapy, plausible factors relating to prognosis will be discussed.

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Abstract – WCN 2013**No: 2122****Topic: 2 – Movement Disorders****"Restless bladder" and the boundaries of the restless legs syndrome**E. Antelmi^a, G. Coccagna^b, L. Ferini-Strambi^c, S. Marelli^c, F. Provini^a. ^a*IRCCS Institute of Neurological Sciences and Department of Biomedical and Neuromotor Sciences, University of Bologna, Italy;* ^b*University of Alabama at Birmingham Bologna, Bologna, Italy;* ^c*Department of Neurosciences, Sleep Disorders Center, Università Vita-Salute San Raffaele, Milan, Italy*

Background: Recently different reports on atypical localization of Restless Legs Syndrome (RLS) symptoms (namely one arm, tongue, thorax, and abdomen) beg the question of the boundaries of RLS.

Objective: This study aims to present two patients with a new atypical localization of restlessness sensation.

Patients and methods: A 43-year-old woman and a 47 years old man, with a positive family history for RLS, came to our attention

complaining of unpleasant sensations localized in the inguinal and bladder region, arising only in the evening while resting, especially when falling asleep. In both patients symptoms started in the adulthood and progressively got worse. Symptoms were intense enough to force void (usually few drops). Voiding yielded transient benefit so that micturition was repeated even up to 20 times per night, ceasing in the morning. There was no urgency during the daytime. Both patients undergone many repeated clinical and diagnostic investigations (laboratory tests, cystoscopy, urography, urodynamic tests, pelvis and spinal MRI).

Results: All investigations were unrevealing. Various treatments with antispastic, analgesic, antibiotic or disinfectant drugs, and antidepressants had no effect. Finally, the administration of pramipexole 0.18 mg before bedtime determined a complete cessation of symptoms, confirmed also after one year of follow-up.

Discussion: Without considering symptom localization, both cases present disturbing similarities with RLS: the classical circadian presentation of symptoms, the symptom's release with muscle contractions (leading to repeated voiding) and the striking response to dopaminergic medication. Thus, we emphasize the need to be aware of this atypical presentation in order to institute appropriate treatment.

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Abstract – WCN 2013

No: 2079

Topic: 2 – Movement Disorders

Non-motor symptom differences according to Parkinson's disease phenotype

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Introduction: Non-motor symptoms (NMSs) such as sleep disorders, fatigue, cardiovascular, neuropsychiatric, gastrointestinal and others are noticed in most of Parkinson's disease (PD) patients, at all stages and phenotypes with an important impact on quality of life. The study objective was to assess the NMSs in a hospital PD sample and differences according to phenotype.

Methods: We have included 140 consecutive PD patients from a tertiary care center. All patients were assessed with Non-motor Symptoms Questionnaire, other variables being gender, age, treatment duration, PD phenotype and H&Y staging. The patients were classified into a tremor predominant phenotype (30, 21.4%, group 1), akinetic-rigid (55, or 39.3%, group 2) and mixed one (55, 39.3%, group 3). Windows Office Excel 2007 program was used for statistical analysis.

Results: The mean patients' age was 58.42 ± 7.8 years, 79 men (56.4%), with an average Hoehn and Yahr stage 1.95, UPDRS part III off score 25.3 ± 12.7 , mean disease duration was 4.07 (0.5 to 16 years). All the motor scores presented more severe values in the akinetic-rigid group. The average score of NMSs was 29.8, being significantly higher in akinetic-rigid type of PD vs. tremor predominant vs. mixed one (23.28 ± 3.3 vs. 40.9 ± 3.8 vs. 21.9 ± 2.3 , $p_{1-2} < 0.01$, $p_{1-3} > 0.05$, $p_{2-3} < 0.001$). The mean NMS number per patient was higher in the akinetic-rigid group (4.26 ± 0.36 vs. 5.8 ± 0.24 vs. 4.74 ± 0.25 , $p_{1-2} < 0.001$, $p_{1-3} > 0.05$, $p_{2-3} < 0.01$).

Conclusion: Akinetic-rigid phenotype of PD has a more severe expression of non-motor symptoms both in the frequency and intensity, adding to clinical heterogeneity of this particular disorder.

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Abstract – WCN 2013

No: 2129

Topic: 2 – Movement Disorders

Arylsulphatase A (ASA) activity in familial Parkinsonism: A pathogenetic role?

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Background: Lysosomal impairment is one of the pathogenic pathways identified in Parkinson's disease (PD). Arylsulphatase A (ASA) is a lysosomal enzyme. It's still controversial whether ASA partial deficiency, caused by the pseudodeficiency allele (ASAp), represents a non-pathogenetic condition, although different neurological diseases have been linked with it.

Objective: We analyzed the pedigrees of three patients with PD and ASA partial deficit.

Materials and methods: We performed clinical, laboratory, neuroimaging and neuropsychological investigations, ASA activity dosage and genetic analysis for ASA alleles in three pedigrees.

Results: In family # 1 we found three subjects with MD (two sisters: a 65-index case- and a 68 years old lady with PD and a 26 years old boy-index case's son – with essential tremor). In family # 2 we detected two sisters with PD (a 78 and a 67 years old lady). Finally, in family # 3 we had a 63 (index case) and her sister, a 72 years old lady, both with PD. Patients showed a mild form of disease (6/6) with frequently atypical features, i.e. postural tremor (4/6), tremor of the head (1/6), orthostatic tremor (1/6), cognitive impairment (4/6) and poor L-dopa response (6/6). ASA activity was reduced in all the examined family members, due to the presence of ASAp.

Conclusion: Our report describes three families with ASA pseudodeficiency and high prevalence of movement disorders. ASA partial deficit could act at the level of the converging pathway between lysosomal impairment and neurodegeneration, increasing aSyn protein aggregation and accumulation.

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Abstract – WCN 2013

No: 2070

Topic: 2 – Movement Disorders

Autonomic dysfunction in Tunisian patients with Parkinson's disease

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Background: Parkinson's disease (PD) is a progressive neurological disorder characterized by a large number of motor and non-motor symptoms such as olfactory impairment, sleep problems, cognitive problems, psychiatric symptoms and autonomic dysfunction (AD). The autonomic domain is broad including symptoms and signs that relate to cardiovascular, gastrointestinal, urinary, thermoregulatory, pupillomotor, and sexual functioning.

Objective: The aim of this study was to assess the prevalence and the clinical characteristics of autonomic dysfunction in Tunisian patients with PD.

Patients and methods: We conducted a prospective study in 361 patients with PD (187 males, 174 females), at the National Institute of Neurology in Tunis. Demographic and clinical data were collected and included motor scores (MDS-UPDRS, Schwab and England and Hoehn and Yahr scores). The autonomic function was evaluated using a reliable and valid instrument, the SCOPA-AUT test.

Results: Autonomic dysfunction was frequent in our population. 37.7% of our PD patients showed cardiovascular dysautonomic symptoms,

48.7% had gastrointestinal disorders and 33% had urinary problems. AD was correlated with age, disease motor severity, depression, cognitive dysfunction, sleep disorders and high doses of dopaminergic medication.

Conclusions: AD was an important feature of PD in our patients, warranting increased clinical awareness and highlighting the need for efficacious therapies for the wide spectrum of problems related to this domain of PD.

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Abstract – WCN 2013

No: 1059

Topic: 2 – Movement Disorders

Polyneuropathy in idiopathic Parkinson – An analysis of risk factors in a mixed Parkinson population

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Background: Several studies have shown a high prevalence of peripheral neuropathy (PN) in idiopathic Parkinson's disease (IPD), suggesting it was levodopa induced or related to a vitamin B12 deficiency on the basis of cumulative levodopa exposure. However, existing studies included no or only small numbers of patients without PD medication.

Objective: This study examined the prevalence of PN in a cohort of consecutive IPD patients of a general neurology outpatient clinic with a substantial number of de novo patients and analyzed the prevalence of potential risk factors.

Patients and methods: 20 IPD patients, Hoehn and Yahr stage 1–4, were examined for the presence and severity of PN, using a definition of distal symmetric polyneuropathy, including electrophysiological measures, the Utah Early Neuropathy Scale (UENS) and the Toronto Clinical Scoring System (TCSS). UPDRS examinations determined the severity of IPD. Potential risk factors, including alcoholism, cobalamin deficiency, high fasting-homocysteine and -methylmalonic acid levels, abnormal fasting glucose, HbA1c levels, and other PNP related serum parameters were analyzed.

Results: PN was present in 55% of IPD patients. T-tests and chi square tests didn't show a significant difference of age, UPDRS or sex between PN-positive and PN-negative patients. An analysis of risk factors didn't show a significant cumulation of risk factors or even of cobalamin-associated laboratory parameters. UPDRS levels significantly correlated with TCSS-levels.

Conclusion: A high prevalence of PN in IPD patients, independent of levodopa exposure or other known risk factors, was shown, but further studies regarding etiology of PN are warranted.

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Abstract – WCN 2013

No: 2021

Topic: 2 – Movement Disorders

Blunted responses to emotional stimuli in Parkinson's disease: An EEG study of visual affective word processing

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Background: Parkinson's disease (PD) is associated with abnormal processing of emotions, including blunted physiological arousal and startle reflex when responding to threatening negative stimuli, and deficits in the production and recognition of facial expression and emotional prosody.

Objective: To identify neurophysiological abnormalities of emotional processing in PD using event related potentials generated from EEG while processing emotional information.

Patients and methods: We recruited 17 non-demented PD patients without affective disturbances and 19 healthy adults. ERPs were recorded from 128 channels, while participants performed a visual word affective priming task. This task elicited information on abnormalities in automatic emotional processing, and consecutively presented negatively or neutrally valenced two words in a 150 ms interval. Participants evaluated the valence of the second word (target word) and amplitudes of ERPs were examined. Repeated measures ANOVA were computed to examine congruency and target valence effects on the ERP amplitudes for within groups and also between groups.

Results: In healthy adults, larger amplitudes for P300 ($F = 8.7$, $p < 0.01$) and late positive potential (LPP) ($F = 15.3$, $p < 0.01$) were observed when negative in comparison to neutral stimuli was presented at frontal (Fz) and right parietal (P4) regions, respectively. These effects reflecting biases towards negative stimuli were lost in PD patients ($F_{Fz} P300: F = 0.002$, $p = 0.97$; $F_{P4} LPP: F = 3.84$, $p = 0.07$).

Conclusion: Neurophysiological deficits in automatic emotional processing are present in PD patients without affective disturbances. These deficits in automatic emotional processing in PD can be identified by the frontal P300 and the right parietal LPP ERP components.

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Abstract – WCN 2013

No: 2032

Topic: 2 – Movement Disorders

Chorea-athetosis as the first clinical manifestation in HIV patient

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Objectives: There are many reports in the bibliography that commend the clinical association of infection with HIV with movement disorders.

Case report: 35 years old female was introduced to our hospital because of hyperpyrexia and abnormal involuntary movements in the extremities of the left side (choreo-athetotic).

The first MRI examination of the head revealed in the right thalamic area pathological signal in T2w and T1w images. GD-enhancement was marked. Lumbar puncture was negative.

PCR for virus in the CSF was negative. IgG Toxoplasma titre was elevated in blood examination. HIV test was positive. The choreo-athetotic movement was the first clinical manifestation to our patient. She received therapeutic dose of toxoplasmosis (co-trimoxazole) IV for 6 weeks. The clinical status of the patient has improved.

We do follow-up to the patient with MRI and clinical examination monthly.

Conclusion: Solitary lesions with mass effect or contrast enhancement were seen in many HIV patients that have been studied for possible focal brain lesions. Most of them were caused by cerebral toxoplasmosis.

In other patients PML or HIV encephalitis or lymphoma has been diagnosed. The issue of increased vulnerability of the basal ganglia to HIV infection is discussed.

Evidence of dysregulation by SPECT has been reported.

WE conclude that dystonia, athetosis or chorea could be the first ever clinical sign of HIV patients.

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Abstract – WCN 2013

No: 2002

Topic: 2 – Movement Disorders

Botulinum toxin in focal hand dystonia – Reasons for drop-out from the treatment

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Objective: The goal of the study was to assess the reasons for discontinuation of long-term treatment of writer's cramp in patients treated with botulinum toxin (BTX). We also observed the long term BTX treatment outcomes.

Introduction: Botulinum toxin A is considered to be an effective treatment for the patients suffering from focal hand dystonia (such as writer's cramp) but its long-term usefulness may be limited by frequent discontinuation of the treatment for various reasons.

Methods: We reviewed the records of 58 patients who had more than 400 applications of BTX. We also conducted surveys via mail or telephone interviews with the patients who were lost to regular follow-up.

Results: Each patient had at least 2 application sessions, median treatment duration was 82 months. 83% of treated patients reported at least a partial improvement of their condition. There were no serious adverse events.

Most frequent side effect was the weakness of muscles adjacent to the injection site.

Altogether 32 (55%) patients chose to discontinue the treatment.

Conclusions: High proportion of the patients chose to discontinue the treatment with BTX. The main reasons for discontinuation were side effects (33%), low efficacy (30%), relocation (7%), change of writing habits (6%), new symptoms and change of treatment (6.5%), other and unknown reasons (17.5%). While BTX remains an effective treatment, drop-out rate appears to be higher than in other focal dystonias, especially due to the side effects and subjectively assessed low efficacy in some patients.

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Abstract – WCN 2013

No: 1994

Topic: 2 – Movement Disorders

Protocol for the microelectrode recording of neuronal activity in head-restrained, alert animals

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Background: Neuronal activity microelectrode recording (MER) is the standard means of intrasurgical target localization for Deep Brain Stimulation electrode implantation. Since MER is usually only obtained for patients with Parkinson's disease under local anesthesia,

an important question is how the neuronal discharge characteristics of the Basal Ganglia vary with the level of alertness.

Objective: Designing a protocol for MER in head-restrained, alert animals.

Material and methods: Thirty-six adult Sprague–Dawley rats were assigned randomly to two groups: control and 6OHDA-lesioned animals. Between 21 and 28 days after the lesion procedure, animals went through stereotactic surgery with the objective of registering spontaneous activity of the medial Globus Pallidum (GPi). Animals were placed in an ad-hoc built restraining device that did not allow any spontaneous movements during the whole recording time. The eyes of the animals were covered; all recordings were obtained in conditions of environmental silence. The alertness level was characterized periodically with a standardized non-painful stimulus.

Results: We were able to record neuronal activity in 38 single-neurons uninterruptedly during the awakening process, and for long periods of thereafter. The mean length time of the recordings was 58.98 ± 30.82 min. After the surgery, wellbeing and alertness level were confirmed by letting the animals explore the laboratory.

Conclusion: We described a protocol for recording in relaxed, head-restrained animals the activity of single deep-brain neurons, under stationary conditions during the transition from deep anesthesia to full alertness. The protocol enables to evaluate the impact of the level of alertness at the cellular scale.

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Abstract – WCN 2013

No: 1924

Topic: 2 – Movement Disorders

Hypercapnic respiratory failure in patients with progressive supranuclear palsy

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Objective: We investigated the frequency of the hypercapnic respiratory failure in progressive supranuclear palsy (PSP). We also discussed the clinical meaning of such hypercapnaemia in PSP.

Background: Progressive supranuclear palsy is a well recognized atypical parkinsonian syndrome. Although nonmotor complications, such as aspiration, dysphagia, and autonomic impairment appear with advance, there is no report of hypercapnaemia in PSP. Since we experienced a case of PSP who showed remarkable hypercapnaemia in his terminal state, we hypothesized that hypercapnaemia could be a common condition in advanced stage of PSP.

Patients and methods: We extracted consecutive 6 cases of PSP who died at our hospital from January, 2008 to December, 2011, and their medical records were reviewed retrospectively about the physical conditions.

Results: The mean age of PSP cases at death was 79.7 (70–86) years old. The mean age at onset was 71.5 (59–79) years old, and average disease duration was 8.2 (3–11) years. Two of those were classical Richardson's syndrome. Five cases were pathologically confirmed as PSP. Hypercapnaemia was observed among five cases within three months before they died.

Conclusions: Hypercapnic respiratory failure is often seen in patients with advanced PSP. Therefore, such hypercapnic condition can be one of the common clinical features in advanced PSP. Although there is some influence of respiratory complications, such as pneumonia, it may reflect the central hypoventilation in endstage PSP.

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Abstract – WCN 2013**No: 2000****Topic: 2 – Movement Disorders**
Isolated focal tongue dyskinesia

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Background: Focal dyskinesias affect a restricted region of the body in isolation and may exhibit a broad range of abnormal movements. **Objective:** To describe a patient with isolated focal tongue dyskinesia. **Patients and methods:** A 71-year-old woman was referred because of a two-year history of continuous movements of her tongue. Spontaneous tongue movements were described as fluctuating but did not fully disappear.

Results: At neurological examination continuous movements with a waxing and waning U-shaping of the tongue were observed at rest. These movements quite abated while tongue protrusion and speaking, did not interfere with feeding, could become more vigorous in response to head position change and stress but were not distractible. The patient was given a placebo injection but her tongue movements were unaffected. Brain MRI, dopamine transporter single photon emission CT, electroencephalography, and laboratory investigations were normal. A video-polysomnography documented a normal macro- and micro-structure of sleep and the absence of abnormal movements of the tongue during sleep. No psychiatric disturbances were detected. Clonazepam at a dose of 0.5 mg per day conferred significant improvement.

Conclusions: Focal tongue dyskinesia is rare and can be misdiagnosed as having a psychogenic cause. Our patient had some unique features: tongue movements were present at rest, were not distractible, and did not interfere with feeding and speech. These clinical features and normal psychiatric evaluation support the tongue dyskinesia in our patient that is "symptomatic" and the need of an ongoing follow-up.

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Abstract – WCN 2013**No: 1977****Topic: 2 – Movement Disorders****Quality of life in patients with Parkinson's disease and impulsive control disorders**

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Dopaminergic replacement therapy (DRT) is effective in treating the motor symptoms of Parkinson's disease (PD), but can lead to impulsive control disorders (ICDs) in some patients. ICDs include pathological gambling, hypersexuality, compulsive shopping, binge eating, punding, and compulsive use of DRT (dopamine dysregulation syndrome).

Objectives: To survey quality of life in patients with PD and ICD.

Material and methods: 247 patients with PD were questioned by Questionnaire for Impulsive-Compulsive Disorders in Parkinson's Disease – Short (Weintraub D et al., 2009). For assessment quality of life in PD patients with ICD, PDQ-39 (Peto V et al., 1995) was used; for assessment quality of life in caregiver, Burden Interview (Zarit SH et al., 1980) was used; and for assessment daily activities, the scale of Schwab & England (1967) was used. 55 patients (23%) (aged from 42 to 82 years, with 28 men) were recruited to this study. Average disease duration was 11.2 ± 5.56 years (from 2 to 24 years). L-dopa equivalent dose was from 900 to 2000 mg/day.

Results: Depression of daily activities was averaged $67\% \pm 19.67\%$. The quality of life in PD patients with ICD was 25% to 89% ($52.4\% \pm 19.97$). Caregivers noticed high burden in 7.7% cases, moderate – in 61.5% and low burden – in 30.7% cases.

Conclusions: ICD has significant influence to daily activities in PD patients and reduces quality of life in PD patients and their caregivers.

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Abstract – WCN 2013**No: 1978****Topic: 2 – Movement Disorders****Myoclonus after treatment of cobalamin deficiency: An unusual complication in adult**

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Introduction: Myoclonus is not a classic feature of vitamin B12 (vit B12) deficiency in adults, though they are frequent in infant cases.

Objective: We report an adult patient with cobalamin deficiency that developed myoclonus exaggerated after the initiation of vit B12 treatment.

Case report: A 56 years old woman presents an acute mental confusion with walking and urinary difficulties. Neurological examination found a disoriented and hallucinated patient with pyramidal and cordonal syndromes, associated to peripheral sensory signs. Mild myoclonus of the upper limbs was noted initially. The vit B12 blood level was low. The MRI showed an extensive posterior cervical and dorsal myelitis. 48 h after the initiation of vit B12 therapy, she developed head myoclonus persisting even during sleep, and contrasting with an improvement of the mental confusion. The electroencephalogram was comital. The myoclonus improved after the administration of low doses of clonazepam.

Discussion: In infantile cobalamin deficiency, variable movement disorders were reported (tremors, chorea, dystonia, myoclonus) usually occurring early in the disease and worsening after treatment. In the literature, there were only two reported adults with cortical and spinal myoclonus, one of them appearing after treatment. This phenomenon may be explained by the sudden stimulation of folate and cobalamin pathways and producing a temporary imbalance of the complicated metabolic pathways of cobalamin.

Conclusion: To our knowledge, this is the third report of myoclonus in an adult patient with vit B12 deficiency. Myoclonus should be considered as one of the extraordinary neurological manifestations of vit B12 deficiency in adults.

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Abstract – WCN 2013**No: 1930****Topic: 2 – Movement Disorders****Detection of hypokinetic dysarthria at early motor signs of Parkinson's disease by acoustic analysis and statistical methods**

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Background: Parkinson' disease (PD) is a slowly progressive and highly debilitating CNS disease. By the time it is firmly diagnosed via routine neurological examination, there is already substantial damage to the CNS. Dysarthria is present in 70–90% of individuals with PD. It has been suggested that subtle signs of the dysarthria,

detectable only by acoustic methods, might serve as biomarkers to help detect the presence of the disease in its early stages.

Purpose: To detect voice and speech abnormalities at early motor signs of PD (EMSPD).

Methods: The speech (reading a paragraph) of 49 individuals with EMSPD (25 M, 24 F; age = 63.18 ± 9.55 years; H&Y = 1.66 ± 0.40 ; UPDRS = 18.65 ± 8.36 ; years since diagnosis = 2.61 ± 1.44) and 84 healthy controls (HC, 40 M, 44 F, age = 64.87 ± 8.45) was analyzed using acoustic measures of vowels, voice quality, pitch prosody, and pauses and rhythmic aspects of speech. Statistical methods were then applied to determine the acoustic predictors for the diagnostic model (these methods included a backward elimination effect selection logistic regression procedure; model diagnostic procedures and ROC analyses; and P values, odds ratios and associated 95% confidence intervals, along with optimized model cutoffs).

Results: ROC area under the curve (AUC) was 0.8657. Optimization of the cutoff for maximum overall correct prediction (at probability 38% or higher) yielded 81.6% sensitivity and 73.3% specificity, with overall 78.5% correct prediction.

Conclusions: Dysarthria is present in at least 78.5% of patients with EMSPD. It is yet to be determined whether dysarthria is present and detectable by acoustic and statistical analyses at preclinical stages of the disease.

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Abstract – WCN 2013

No: 1932

Topic: 2 – Movement Disorders

Clinical features of G_{M1} gangliosidosis type 3

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Background: G_{M1} gangliosidosis is a rare inherited lysosomal storage disorder due to deficiency of the β -galactosidase, resulting in accumulation of G_{M1} gangliosides and other glycoconjugates in the brain and visceral organs. GM1 gangliosidosis type 3 is a late-onset chronic form, and this form is very rare.

Objective: To describe clinical features of G_{M1} gangliosidosis type 3 from genetically proven three cases.

Methods: The patients were 2 males and 1 female. Average age of the patients was 36 yo (18–58). Description of clinical features, imaging, biochemical and genetic analysis was done.

Results: All patients were born of a consanguineous marriage. All patients were born normally and had normal developmental milestones. Average age of the disease onset was 7.8 yo (3–11). Neurological examination showed generalized dystonia, gait disturbance and speech difficulty. Dystonia was especially prominent in face and neck, which were previously reported in only few cases as blepharospasm or jaw-closing impairment. Two patients had mental retardation. No patients had facial dysmorphic features or cardiomyopathy. Bone radiography revealed scoliosis, hip dysplasia, and flattening and anterior beaking of the vertebral bodies in all cases. Brain MRI showed slight abnormality of basal ganglia in all cases, and cerebral atrophy in 2 cases. Average leukocyte β -galactosidase activity was $7.8\% \pm 2.0\%$ of normal. All patients had homozygous I51T mutation in *GLB1* gene.

Conclusion: In comparison with prior cases, facial and neck dystonia, and speech difficulty were distinctive features in our cases. This might be a characteristic of I51T mutation in G_{M1} gangliosidosis type 3.

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Abstract – WCN 2013

No: 1922

Topic: 2 – Movement Disorders

The application of L-DOPA-containing polymeric nanoparticles provides motor function recovery in 6-OHDA-induced Parkinson's disease model

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Background: The inclusion of L-DOPA into the PLGA nanoparticles increases effective half life time, drug's bioavailability and efficacy. Furthermore, nasal administration is an advanced brain delivery route for the Parkinson's disease (PD) treatment.

Objective: To detect the efficacy of nano-DOPA nasal administration in comparison with standard drug using PD model.

Materials and methods: L-DOPA-containing nanoparticles (250 ± 50 nm) were synthesized with double emulsion method. The therapy's efficacy was examined using Wistar rats (320 ± 20 g) with 6-OHDA induced PD. Animal's motion coordination and behavior were analyzed using forelimb placing, open field etc. tests. The drugs were administered daily, 0.35 mg/kg (by L-DOPA).

Results: Significant coordination performance improvement was detected in the nano-DOPA-treated group by the second week of therapy. By the therapy start the results of PD animal groups in the placing task reached $81 \pm 15\%$ (by the intact control). L-DOPA showed significant effect ($91 \pm 8\%$) at the first administration only. After the 17 weeks of treatment coordination performance was $94 \pm 12\%$ in nano-DOPA-treated, $62 \pm 13\%$ in L-DOPA-treated and $51 \pm 28\%$ in non-treated group of animals. A week after the drug cancellation the results were $77 \pm 15\%$, $60 \pm 15\%$ and $47 \pm 29\%$ respectively. * $p < 0.05$.

Conclusion: Nano-DOPA nasal administration provides lasting motor function recovery and allows to considerably lower the effective drug dose, keeping the effect even a week after the drug cancellation.

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Abstract – WCN 2013

No: 1181

Topic: 2 – Movement Disorders

Automatic detection of freezing index of Parkinson's disease using a portable gait rhythmogram

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Background: Freezing of gait is one of the most important motor symptoms in Parkinson's disease. However, we do not have a good method to evaluate accurately the daily profile of freezing in patients with Parkinson's disease. The aim of this study is to develop a new method of automatic detection of daily profile of freezing of gait on Parkinson's disease using a portable rhythmogram.

Patients and methods: Firstly, by using a portable gait rhythmogram, we examined gait acceleration and rhythm in 30 patients compared with 27 normal control subjects on 5 m or 10 m walking. Secondly, we studied daily profile of gait in 20 patients with Parkinson's disease using the rhythmogram during 2 or 3 whole days.

Results: The gait acceleration as strength of parkinsonian gait was apparently weak compared as normal control. We also could detect

freezing gait by consecutive changes of gait rhythm and rapidly changes above 3 Hz (cadence: 180 steps/min), which was also confirmed by video camera on 5 m or 10 m walking. Furthermore, we also could make a whole day profiling of gait and an automatic detection of freezing index (freezing frequency a day) on parkinsonian gait by studying gait rhythm with the portable gait rhythmogram.

Conclusion: In this study, we could extract the characteristic of the parkinsonian gait and evaluate especially freezing events more objectively. This method may bring us to evaluate severity of parkinsonian gait not only in a consulting room but also daily profile even not to see directly using the portable gait rhythmogram.

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Abstract – WCN 2013

No: 1009

Topic: 2 – Movement Disorders

Comparison of smooth pursuit eye movement (SPEM) in patients with idiopathic Parkinson's disease (PD) and progressive supranuclear palsy (PSP)

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Background: SPEM is impaired in most patients with PD and PSP, though the nature of the impairment is poorly understood.

Objective: To examine whether the impaired SPEM in PD/PSP involves impaired visual motion working memory and/or movement-preparation.

Patients and methods: Twenty-one PD patients, 6 PSP patients and 10 controls completed a memory-based SPEM task (Shichinohe et al., 2009). Cue 1 indicated visual motion-direction while cue 2 instructed the subject to prepare to pursue (go) or not to pursue (no-go). Subjects were required to pursue correct spot from 2 oppositely moving spots or not to pursue by selecting a 3rd stationary spot. For comparison, simple ramp-pursuit was also examined.

Results:

(1) In the memory-based SPEM task, error rates of all PD were low (<5%), similar to controls, but all PSP had higher error rates.

(2) During go trials, controls exhibited SPEM followed by corrective saccades. Most PD (12/21) and PSP (3/3) tracked correct spot with saccades; SPEM was absent before the saccades. Mean latencies of the first saccades in PD and PSP were longer than those of controls ($p = 0.02$).

(3) During simple ramp-pursuit, most PD (15/20) and PSP (5/6) exhibited initial SPEM followed by corrective saccades, and their latencies were similar to those of controls.

(4) In both tasks, peak SPEM velocities were lower in PD and PSP than those in controls ($p < 0.01$).

Conclusions: The working memory of motion-direction and go/no-go selection was normal in PD but not in PSP. Both had difficulty in initiating SPEM using the cue information.

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Abstract – WCN 2013

No: 519

Topic: 2 – Movement Disorders

Association of idiopathic polyneuropathy and incident Parkinson's disease: A population-based observational study

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Background: Studies have reported an association between levodopa exposure in Parkinson's disease (PD) and an increased incidence of idiopathic polyneuropathy (IPNP).

Objective: To investigate if newly diagnosed PD patients have a higher prevalence of IPNP than the general population.

Patients and methods: Newly diagnosed PD patients without previous exposure to antiparkinsonian drugs and age-, sex-, and practice-matched controls were identified in the UK General Practice Research Database. They were compared for the presence of IPNP within 3 years preceding the index date, i.e. date of PD onset in PD patients or matched date in controls. PD patients and controls were ≥ 18 years of age and had ≥ 3 years of person-time in the database. Patients with risk factors for neuropathy (e.g. diabetes, alcohol abuse) or prescriptions of drugs known to cause neuropathy or parkinsonism were excluded. Conditional logistic regression analysis was used to calculate odds ratios for the association between IPNP and PD.

Results: 5089 PD patients and 19,897 controls were included. 20 PD patients (0.39%) and 40 controls (0.20%) had a diagnosis indicating IPNP within 3 years prior to the index date. After manual record review, 15 PD patients and 24 controls had confirmed IPNP. The prevalence of IPNP was increased 2.4-fold in PD patients compared to controls (OR = 2.41; 95% CI 1.17–4.81). There was no indication that age or sex acted as effect modifiers.

Conclusions: In this observational study, patients with incident PD had a higher prevalence of pre-existing IPNP that cannot be explained by adverse effects of antiparkinsonian drugs.

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Abstract – WCN 2013

No: 1890

Topic: 2 – Movement Disorders

A substantial investigation on psychiatric symptoms in patients with Parkinson's disease (PD)

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Objective: To investigate a situation of psychiatric conditions in patients with PD.

Methods: Our survey was carried out after our Hospital Ethics Committee approved our clinical research program. We conducted the survey of 351 patients with PD in our out-patient clinic who assented to our clinical research program. Used test batteries were bellows, for cognition were MMSE and WAIS-III, for frontal lobe function were FAB, word fluency test and apathy test, for depression were HAM-D, BDI and SDS, for memory function was RBMT, for hallucination was NPI, and for sleep function was ESS. At the same time, we checked their motor function (UPDRS), their QOL (PDQ-39) and imaging studies (MRI, ECD-SPECT, MIBG cardiac spectroscopy). These data were analyzed by adequate statistical methods.

Results: Patient background: M:F = 160:191, examined age: 69.6 ± 7.7 y-o, age of onset: 63.0 ± 4.5 y-o, and duration of illness: 4.5 ± 6.6 years. H-Y stage: Y-2, 36%; Y-3, 44%; and Y-4, 19%. Results of score in the first year were as follows: In MMSE score <23 points, 30.3%; in RBMT score <15 points, 32.3%; and detected depression state by BDI, 48% (however, no one in depression state in HAM-D). Frontal lobe function is impaired in proportion to other lobes from our results.

Conclusions: In this investigation, characteristics of psychiatric symptoms in PD become identified. It is important to continue with such investigations for the purpose of improving QOL in PD patients.

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Abstract — WCN 2013

No: 1833

Topic: 2 — Movement Disorders

Transcranial sonography and pocket smell test in differing Parkinson's disease from essential tremor

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Introduction: We wanted to determine the specificity and sensitivity of transcranial sonography (TCS) and Pocket Smell Test (PST) in differing Parkinson's disease (PD) from essential tremor (ET). We also wanted to compare these methods with the specificity and sensitivity of DaTSCAN used as a gold standard.

Patients and methods: In this study we included 110 patients with the symptomatology of parkinsonism present in the last 6 to 12 months. According to the DaTSCAN results, patients were divided into two groups. The PST was performed to evaluate the olfactory dysfunction. Using TCS the possible substantia nigra hyperechogenicity was encircled and measured. The sensitivity and specificity of PST and TCS were calculated.

Results: From 110 patients, 51 (46.4%) patients had ET and 59 (53.6%) patients suffered from PD. Olfactory dysfunction was present in 44 (74.6%) patients with PD, and in 10 with ET. The specificity of PST is 80.39%, and the sensitivity is 74.58%. The hyperechogenicity of the substantia nigra was predominantly found in patients with Parkinson's disease. The specificity of TCS is 88.2%, and the sensitivity is 94.9% in confirming Parkinson's disease.

Conclusion: We suggest that it would be useful to perform these methods routinely in the everyday practice as a good screening protocol before indicating DaTSCAN.

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Abstract — WCN 2013

No: 1831

Topic: 2 — Movement Disorders

Eye of the tiger sign in a patient with levodopa-induced motor complications

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Background and objective: An important differential diagnosis of Parkinson Disease (PD) is Progressive Supranuclear Palsy-parkinsonism (PSP-P). The most useful clinical features to distinguish PSP from PD are early falls due to postural instability and supranuclear gaze palsy or slowed vertical saccades. We report a parkinsonian patient with levodopa-induced motor complications referred for DBS.

Methods: Case report and review.

Results: A 56 year-old man with a 5 year history of a levodopa-responsive akinetic-rigid parkinsonism (improvement by 57.4% on the UPDRS-III on a levodopa challenge test) was referred for DBS due to severe levodopa-induced motor fluctuations and dyskinesias, punting and pathologic gambling. A symmetric akinetic-rigid parkinsonism with important axial involvement was noted, including dysarthria and "on" freezing of gait. Also, frontalis overactivity, apraxia of eyelid opening and slowed vertical saccades were observed. No postural instability or supranuclear gaze palsy was present. A clinical diagnosis of PSP-P was made and the patient was excluded from DBS. T2-weighted MRI disclosed hypointensity with central hyperintensity of the globus pallidus (eye of the tiger sign). Laboratory evaluation was normal. Pantothenate kinase 2 gene mutation screening was requested.

Conclusion: The eye of the tiger sign, as well as the presence of significant levodopa-induced motor complications, has been rarely reported in PSP. This case highlights the difficult differential diagnosis between PD and PSP-P early in the beginning of the disease, and shows that severe levodopa-induced motor complications, although uncommon, may be present in PSP-P.

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Abstract — WCN 2013

No: 1829

Topic: 2 — Movement Disorders

Restless legs syndrome in pregnancy: A case responding to glucocorticosteroids

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Background: Several studies have related hypothalamic-pituitary-adrenal system activation and restless legs syndrome (RLS). Pregnant women did not participate in them. There are no published cases of RLS in pregnancy responding to glucocorticosteroids either, as they are usually avoided due to potential teratogenicity.

Objective: To describe the effect of glucocorticosteroids over RLS in a pregnant woman treated with a single dose of 20 mg of intravenous methylprednisolone followed by oral prednisone for 3 days because of an asthmatic attack.

Patients and methods: Prospective follow-up during 2nd, 3rd trimester and 12 weeks postpartum. Assessment included familial and past medical history, interview about RLS symptoms, International Restless Legs Syndrome Scale (IRLSS), and blood tests including hemoglobin and ferritin.

Results: A 45 year-old pregnant woman was diagnosed with severe RLS as she met the diagnostic criteria (IRLSS: 38). She had positive family history, was asthmatic and RLS symptoms had started at the 22nd week. Hemoglobin value was 12 g/dL and ferritin 11 µg/L. Initial treatment with oral iron and clonazepam 0.5 mg at night induced a mild amelioration of symptoms (IRLSS: 22). There was a sudden important improvement after iv methylprednisolone which was maintained for two weeks after completing oral treatment (IRLSS: 12). Then symptoms went back to the previous situation (IRLSS: 22) although hemoglobin had risen. At twelve weeks postpartum, she was asymptomatic.

Conclusion: Placebo effect is not a probable explanation to her improvement because glucocorticosteroids were given as a treatment to another disease. Further studies on hypothalamic-pituitary-adrenal activity in pregnant women with RLS would be useful to understand RLS pathogenesis.

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Abstract – WCN 2013**No: 1745****Topic: 2 – Movement Disorders****Posterior SMA syndrome after subcortical stroke: Contralateral akinesia reversed by visual control**

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Background: The Supplementary Motor Area (SMA) plays a key role in motor programming and production, and is involved in internal-cued movements. SMA syndrome after lesion of the “SMA-proper” is characterized by transient impairment of voluntary movements and motor sequences. This syndrome is assumed to follow from interruption in the motor cortico-subcortical loop. A few case-reports indicate that such syndrome occurs after a lesion isolating SMA from subcortical structures.

Aim: To characterize the pattern of motor impairment in a patient with SMA-subcortical motor loop disconnection after stroke.

Method: The present case developed a transient left hemiparesis following subcortical stroke in the territory of the right anterior cerebral artery, which disconnected the SMA from the basal ganglia. Eight days after the stroke, when the hemiparesis had regressed, the patient presented a specific SMA motor disorder of the left hand manifesting as an akinesia exacerbated when visual attention was not directed towards his hand. We assessed finger tapping with the left and right hands and closed and open eyes, in the left and right hemifields. We indexed movement speed as the number of tapping in 5-second periods.

Results: Ideomotor, ideational gestures and motor sequences were preserved. On the tapping task, the left hand was slower than the right hand. Critically, visual feedback increased tapping speed for the left but not the right hand. The hemispace of the task execution had no effect on tapping performance.

Conclusion: Our results suggest that SMA-basal ganglia disconnection decreases contralateral movement initiation and maintenance and visual cues can partly compensate this effect.

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Abstract – WCN 2013**No: 1803****Topic: 2 – Movement Disorders****An epidemiological study of neurodegenerative parkinsonism prevalence in the population of “Upper Land” in the south-eastern Moravia, Czech Republic**

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Objective: To assess the prevalence of neurodegenerative parkinsonism in the rural population of the “Upper Land” region in the south-eastern Moravia, Czech Republic.

Background: It has been recently described that the prevalence of neurodegenerative parkinsonism in isolated European communities might be higher than estimated in the general population. We have recently described this phenomenon in one of the villages (population 1500) of the region of south-eastern Moravia, where we have found the prevalence of 2.5% (in the population older than 50 years). The presented study involved the whole region, i.e., 10 villages (population approximately 9000).

Methods: In the first stage, each patient visiting GP for any reason over the study period of three months completed the screening questionnaire for parkinsonism. In the second stage, neurologists identified all persons with positive signs of parkinsonism among those screened positive. In the third stage, all identified persons were admitted to tertiary movement disorder centers to confirm or exclude the diagnosis of parkinsonism.

Results: The sample consisted of 1039 villagers; 193 persons were screened positive, and 69 of them were identified as manifesting signs of parkinsonism; the diagnosis was confirmed upon examination in the tertiary movement disorder centers.

Conclusion: The prevalence rate of 2.8% in the population older than 50 years substantially exceeds the estimated prevalence in the common Central-European population, future research will focus on the heredity in the families in which parkinsonism has been identified.

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Abstract – WCN 2013**No: 1802****Topic: 2 – Movement Disorders****Characteristics of motor complications in patients with Parkinson's disease in southern Tunisia (ST)**

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Introduction: Motor fluctuations remain often underdiagnosed in the first years of treatment. Despite the importance of this problem, little was known about the cause of motor complications.

Objective: To estimate the prevalence, characteristics and factors that correlated with the occurrence of motor complications in patients with PD in ST.

Patients and methods: 102 unrelated patients were recruited. The mean age at onset was 64.9 years (range 40–86) and 57.8% were men. The severity of motor fluctuations and dyskinesias was assessed by the Unified Parkinson Disease Rate Scale IV “UPDRS IV”. Abnormal Involuntary Movement Scale “AIMS” was used to assess the severity of abnormal movements.

Results: We identified 44 patients (n = 44/102, 43.13%). Motor fluctuations were reported in 24.50% and occur after a mean duration of evolution of 10.66 years (range 3–24). The mean age at onset of PD in patients with motor fluctuations was 52.48 years (range 33–72). An early age at onset (≤40) of PD was present in 16% of cases (n = 4). Based on the UPDRS IV, the duration of episodes was variable: 25% of the day in 60%, half of the day in 28% and up to 75% of the day in 12% of cases. Dyskinesias were present in 33.33% and occur after an average duration of the disease of 9.5 years (range 2–31). An early age at onset (≤40) of PD was frequently associated with abnormal movements (20.58%). The average score of the AIMS was 10.58/40 (range 4–29).

Conclusion: The prevalence of motor fluctuations increased with increasing disease and treatment duration as well as advancing disease severity.

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Abstract — WCN 2013**No: 1805****Topic: 2 — Movement Disorders****Parkinson's disease as a protective factor of atherogenesis**

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Background: Parkinson's disease (PD) is one of the most common neurodegenerative diseases. Both genetic and environmental factors are discussed in the pathogenesis PD. Role of hyper- and hypo-cholesterolemia remains poorly understood.

Material and methods: 46 patients with PD were included, mean age 63.1 ± 7.2 years and disease duration 3.4 ± 3.3 years. 30 people of the same ages without neurodegenerative diseases and significant vascular disorders completed the control group. We investigated blood lipid spectrum and results of Doppler ultrasound scan.

Results: Patients were classified using the Hoehn Yahr scale: 1 stage — 5 (10.9%) patients, 2 st. — 20 (43.5%), 3 st. — 19 (41.3%) and 4 st. — 2 (4.3%) patients.

PD patients had a lower level of triglyceride (1.3 ± 0.5 vs 2.0 ± 0.9) and more thin intima-media according to the results of the Doppler ultrasound scan (0.8 ± 0.1 mm vs 1.0 ± 0.3 mm) without differences in total cholesterol level. In groups according to the Hoehn Yahr stages (3st. vs 2st.) level of total cholesterol was significantly lower in PD patients with more severe stages (4.4 ± 0.7 vs 5.1 ± 1.4), $p = 0.011$.

Conclusion: PD patients had lower level of total blood cholesterol, triglycerides and intima-media thickness with more significant changes in severe stages of PD which may be considered as lower risk of atherogenesis.

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Abstract — WCN 2013**No: 1804****Topic: 2 — Movement Disorders****Clinical experience with continuous intestinal dopaminergic therapy: A perspective from Hungary**

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The continuous intestinal levodopa/carbidopa gel (LCIG) therapy in advanced Parkinson's disease (PD) was introduced in Hungary in 2011 July.

Here we report our two-year experience with LCIG therapy. In three clinical centers we tested 37 and include 30 patients. We analyze the gender and age distribution, PD disease, levodopa therapy and motor fluctuation duration. Inclusion criteria were advanced Parkinson's disease (PD), lack of sufficient response to oral medication, clinically significant fluctuations in motor and non-motor symptoms and a good clinical response on test phase.

We observed a significant improvement in not only motor but also non-motor symptoms, reduced fluctuations, less OFF time, increased ON time and better quality of life in patients.

Failure of LCIG therapy was predicted by cognitive decline and low psychosocial support.

Side effects and technical complications (PEG/J system problems, pump problems, surgical procedures and wound problems) were tolerable. Treatment was terminated in 4 patients.

In conclusion CDS therapy is a favorable preference in advanced PD with poor response on oral medications and serious motor fluctuation.

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Abstract — WCN 2013**No: 1795****Topic: 2 — Movement Disorders****Severe nigrostriatal degeneration without clinical parkinsonism in patients with POLG mutations**

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Background: There is increasing evidence linking mitochondrial dysfunction to degeneration of the substantia nigra and Parkinson's disease. Mutations of the DNA polymerase of mitochondria (POLG) are common causes of mitochondrial dysfunction and disease. Parkinsonism is however rarely seen with POLG defects.

Objective: To investigate the substantia nigra of patients with POLG encephalopathy.

Patients and methods: We studied the substantia nigra in eleven patients with POLG encephalopathy. Mitochondrial and histological changes were characterized in six patients and in vivo nigrostriatal integrity and cerebral metabolism were investigated using dopamine transporter imaging and positron emission tomography in five.

Results: At the molecular level, patients showed mitochondrial DNA depletion, high levels of deletions and selective, progressive complex I deficiency in nigral neurons, which paralleled progressive neuronal loss. Histology and imaging showed that the degree of nigral degeneration and nigrostriatal denervation in our patients were even more pronounced than in idiopathic Parkinson's disease. Despite this, our patients did not have parkinsonistic features. The additional presence of thalamic and cerebellar dysfunction in our patients suggested that these may play a role in counteracting the effects of basal ganglia dysfunction and prevent the development of clinical parkinsonism.

Conclusions: Our findings confirm the vulnerability of nigral neurons to mitochondrial dysfunction. Moreover, we show that severe nigrostriatal depletion can occur without the clinical correlation of parkinsonism. These results raise fundamental questions about our current understanding of the pathophysiological model of parkinsonism and suggest that other and yet unknown mechanisms contribute to the generation of the parkinsonistic syndrome.

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Abstract — WCN 2013**No: 1777****Topic: 2 — Movement Disorders****Substantia nigra hyperechogenicity and apparent diffusion coefficient (ADC): Relationship to clinical features of the Parkinson's disease**

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Background: Among the new neuroimaging methods, transcranial sonography (TCS) and diffusion tensor imaging (DTI) offer a unique opportunity to visualize changes in the substantia nigra in vivo.

Aim: To investigate whether SN hyperechogenicity correlated with DTI findings and other clinical data.

Methods: 69 patients (30 males and 39 females, mean age 65.5 ± 8.1 years) diagnosed with PD were examined by TCS using Toshiba Aplio XG system. Echogenicity of SN was assessed. 18 of these patients were studied with a DTI protocol at 1.5 T scanner. Regions of interest (ROIs) were drawn in the ventral, middle, and caudal SN. Clinical data were collected: disease duration, UPDRS, MMSE, FAB, Parkinson's Disease-Cognitive Rating Scale (PD-CRS), Beck Depression Inventory (BDI).

Results: The significant correlation was found between DTI findings (sum of ADC of ventral SN on both sides (mean 1.62 ± 0.15) and sum of bilateral SN echogenic sizes (mean $0.63 \pm 0.19 \text{ cm}^2$)), measured by TCS (Spearman $r = 0.69$, $p = 0.001$). TCS data were related to cognitive impairment in PD patients. Diameter of the third ventricle was correlated with MMSE score ($r = -0.44$, $p < 0.001$), FAB and PD-CRS scores ($r = -0.51$, $p < 0.001$ and $r = -0.48$, $p = 0.001$), depression according to BDI scores ($r = 0.33$, $p = 0.022$), age ($r = 0.41$, $p = 0.001$), and age of onset ($r = 0.42$, $p = 0.001$). Bilateral mean SN area correlated with age ($r = 0.31$, $p = 0.009$), diameter of the third ventricle ($r = 0.47$, $p < 0.001$), MMSE ($r = -0.34$, $p = 0.004$), FAB ($r = -0.32$, $p = 0.006$), PD-CRS ($r = -0.29$, $p = 0.015$), and UPDRS ($r = 0.25$, $p = 0.035$).

Conclusions: Our data suggest that SN hyperechogenicity correlates with ADC of the ventral SN measured by DTI. Results indicate that SN hyperechogenicity reflects not only nigrostriatal degeneration but also neurodegeneration of the brain as a whole.

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Abstract – WCN 2013

No: 1772

Topic: 2 – Movement Disorders

Stimulation related side effects in ten patients with subthalamic deep brain stimulation

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Objective: We wanted to present stimulation-related side effects in our series of subthalamic deep brain stimulation (DBS) patients derived retrospectively from patient's records.

Background: The appropriate placement of the electrodes to the exact target is very important in the subthalamic nucleus DBS. Microelectrode recordings and stimulation during surgery are very important to accomplish this task. Arrangement of stimulation parameters can prevent stimulation related side effects. Paresthesias, eye opening apraxia, dysarthria, diplopia, hypomania, gait disorders and ataxia can be seen during stimulation.

Patients and methods: In 10 idiopathic Parkinson patients bilateral subthalamic DBS were performed. Mean age of the patients was 58 ± 12 . During and after surgery within 3 months stimulation-related side effects were recorded.

Results: In 9 patients side effects were developed during stimulation. They were prominent on the most affected side. Paresthesias, diplopia, dysarthria and gait disorders were the most prevalent side effects. In two cases hypomania and in one case dopamine dysregulation syndrome developed. In all cases these effects vanished in the stimulation off state. Side effects other than dysarthria and postural

impairment were improved after the arrangement of stimulation parameters.

Conclusions: Our results of 10 patients were in accordance with the literature. Stimulation related side effects of DBS of the subthalamic nucleus, though seen quite frequently, can be handled mostly by close supervision of the stimulation parameters during and after surgery.

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Abstract – WCN 2013

No: 1731

Topic: 2 – Movement Disorders

Reflex studies in patients with essential tremor–Parkinson's disease: Comparisons to Parkinson's disease and essential tremor

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Introduction: The association between essential tremor (ET) and Parkinson's disease (PD) is an ongoing controversy. Although coexistence of ET and PD is considered as a coincidence, it is now more widely accepted that ET may evolve into PD (ET–PD). Here, we aim to analyze functional similarities and differences by means of electrophysiology.

Patients and method: Age- and gender-matched 22 ET, 15 PD, and 25 ET–PD patients and 20 healthy subjects were enrolled in the study. Diagnosis was established according to the clinical criteria. Electrophysiological studies included polygraphic analysis, blink reflex (BR), auditory startle reaction (ASR) and long latency reflex (LLR).

Results: Rest tremor was observed in the ET group: mostly on distal extremity with the highest frequency. Postural tremor was seen in all ET and in almost all ET–PD whereas it was present in only half of PD cases. Myoclonus-like activities were observed in ET and ET–PD. Tremor was mostly alternating and asymmetrical in PD and ET–PD whereas it was more or less synchronous in ET. BR probabilities were similar between all groups, however, total ASR probability was reduced in all patient groups ($p < 0.001$). LLR2 was more common in ET and ET–PD with higher amplitudes (679.8 ± 318.8 ; 814.0 ± 717.6 ; 370.5 ± 316.3 ; $p = 0.048$).

Conclusions: Our study shows that there exists a distinct group of tremor patients displaying some features like ET and some like PD. Despite the integrity of BR pathway, decreased probability of ASR in ET and ET–PD similar to PD suggests involvement of at least ASR pathway. Myoclonus like activity and high amplitude LLR suggest cortical involvement. Therefore, we may propose that ET–PD is probably a neurodegenerative disease with cortical involvement and some PD-like features.

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Abstract – WCN 2013

No: 1466

Topic: 2 – Movement Disorders

Evaluation of an Arabic version of the non-motor symptoms scale in Parkinson disease

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Background: Although non-motor symptoms of Parkinson disease (PD) are common and can cause severe disability, they are often under-recognized. Non-motor symptom assessment scale of PD (NMSS-PD) is

a specific validated scale in different populations for the screening and the assessment of these symptoms.

Objective: To evaluate an Arabic version of the NMSS-PD as an instrument for measuring non-motor symptoms in Tunisian patients.

Methods: We conducted a psychometric analysis of the Arabic version of the NMSS using an observational study of 62 patients with PD. The battery also included the scales for outcomes of PD-autonomic (SCOPA-AUT), the Pittsburgh sleep quality index (PSQI), Beck depression inventory, the geriatric depression scale, the MMSE, visual analogical scale for pain and neuro-psychiatric inventory. The correlation between these tests and the NMSS was studied. The reliability of NMSS was explored.

Results: Mean age in our population was 63 ± 10 years, sex-ratio was 1.95 and disease duration was 6.6 ± 5.2 years. 89% of them were on l-dopa. All patients were clustered around Hoehn&Yahr Stages 2–3. The mean NMSS score was 82 ± 56 . The mean Cronbach's alpha of the NMSS Arabic version was 0.87 and showed a satisfactory internal consistency. There were highly significant correlations between the NMSS and the SCOPA-AUT as well as the NMSS and PSQI scores. There was a significant negative correlation between the attention/memory domain and MMSE.

Conclusion: The Arabic version of the NMSS should be considered a comprehensive, useful measurement for non-motor symptoms in Tunisian PD.

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Abstract — WCN 2013

No: 1737

Topic: 2 — Movement Disorders

Drug-induced extrapyramidal syndromes

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Background: There are drug-induced extrapyramidal syndromes leading frequently to diagnostic challenges.

Objective: To correctly diagnose and determine the moment of occurrence (patient's age and days of treatment) and the drugs which induce extrapyramidal syndromes.

Patients and methods: We conducted a prospective study on 34 patients (10 women, 24 men). We used a specially designed questionnaire, the drug-induced extrapyramidal symptoms scale (DIEPSS) and a complete neurological examination.

Results: The drug-induced extrapyramidal syndromes are polymorphic regarding the clinical manifestations, the moment of occurrence and the dosage and type of medication used. The distribution by age showed two peaks, 20–29 years of age (29%) and 70–79 years of age (15%). The majority of drug-induced extrapyramidal syndromes occurred with typical neuroleptics (haloperidol, zuclopenthixol) as well as atypical neuroleptics (risperidone, olanzapine, clozapine) and antidepressants. Most of the extrapyramidal symptoms (58.82%) occurred in days 0–9 of treatment. The clinical manifestations were parkinsonism (35.29%), tremor (29.41%), neuroleptic malignant syndrome (20.58%), dystonia (8.8%) and akathisia (5.88%). The first line treatment was to stop or reduce the medication that induced them, hydration, administration of Diazepam, Trihexyphenidyl or l-Dopa.

Conclusions: Although the number of cases analyzed was not large enough to be statistically representative, it was found that males are more frequently affected than women and that young people showed a higher risk of developing drug induced extrapyramidal

syndromes. We found no demonstrable risk factors for developing drug induced extrapyramidal syndromes. The patient's medical history is very important to avoid diagnosis errors (example Parkinson's disease, dystonia).

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Abstract — WCN 2013

No: 1711

Topic: 2 — Movement Disorders

The efficacy of botulinum toxin injections in deep muscles of upper limb with and without using needle electrical stimulation

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Background: Upper limb spasticity after stroke often required the injections of botulinum toxins. However, injections into the deep muscles are not always effective. This may be due to the difficulty in finding target muscles.

Objective: To determine the efficacy of botulinum toxin for injection in muscles, pronator teres and digitorum profundus, with and without using needle electrical stimulation.

Patients and methods: Patients with spasticity of the upper limb (required obligatorily treatment of pronation patterns and clenched fist) and severe spasticity of 2 or more points by the modified Ashworth scale (mAs). In group-A, 23 patients were treated by experience injectors without electrostimulation. In group-B, 18 patients were injected under the control of electrostimulation. Both groups were treated by the same experienced injector. We used botulotoxin (analyzed only for target muscles), Dysport (120–320 U) or Xeomin (40–100 U). A control visit was carried out after 1 month to assess spasticity (mAs).

Results: In group-A, baseline spasticity was 2.48 ± 0.60 ; in group-B, 2.56 ± 0.63 (not-significant differences). The average dose of Xeomin for group-A was 85.7 ± 15.1 U while that for group-B was 89.3 ± 14.8 U (not-significant differences). The average dose of Dysport for group-A was 194.4 ± 44.6 U while that for group-B was 183.3 ± 41.1 U (not-significant differences). After 1 month, average score for mAs was 1.52 ± 0.87 in group A and the change compared with baseline spasticity was 0.95 ± 0.80 . Average score of mAs in group B was 1.19 ± 0.75 , and the change compared with baseline spasticity was 1.38 ± 0.72 . Differences between the groups were on average 46% and are valid ($p < 0.05$).

Conclusions: The use of electrostimulation improves the efficiency of botulinum toxin injection into the deep muscles of the upper limb.

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Abstract — WCN 2013

No: 1747

Topic: 2 — Movement Disorders

l-DOPA and attentional boost in Parkinson's disease: a clue to subthreshold impulsivity and psychosis

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When participants perform an attention-demanding task (i.e., remembering white target letters and ignoring black distractor letters in briefly presented sequential stimuli), they also encode the background

context. This phenomenon is called attentional boost, which is impaired in individuals with hippocampal atrophy. In this study, we examined the effect of L-DOPA in attentional boost. We report data from 20 patients with Parkinson's disease (PD) before and after L-DOPA therapy. Participants received a letter detection task, while also viewing a series of briefly presented natural and urban scenes in the background of the letters. Before L-DOPA treatment (unmedicated state), the performance of PD patients was similar to that of the controls: they exhibited higher levels of scene recognition performance when scenes were presented with target letters relative to baseline (scenes alone) and distractors. After L-DOPA treatment, target-associated scene recognition was further enhanced, but PD patients also improved recognition when scenes were presented with distractor letters outperforming controls. L-DOPA did not affect baseline scene recognition. Changes in subthreshold impulsive traits and psychosis-like experiences (follow-up minus baseline) correlated with paradoxically enhanced scene recognition at distractor letters. These results suggest that L-DOPA provides an attentional boost at both behaviorally relevant and irrelevant points of time when distractors are presented. A behaviorally irrelevant boost may account for subthreshold psychiatric symptoms detectable in PD.

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Abstract – WCN 2013

No: 889

Topic: 2 – Movement Disorders

Relapsing polychondritis and lymphocytic meningitis with varied neurological symptoms

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Background: Relapsing polychondritis is a rheumatological disease characterized by bilateral auricular chondritis, vestibular compromise and varied neurological symptoms.

Objective: To report the case of a patient with relapsing polychondritis and several neurological manifestations.

Patient and method: We report the case of a patient with relapsing polychondritis with several neurological manifestations.

Results: A 69 year-old male was admitted with a 20-day history of ataxia, paraparesis, tinnitus, vertigo and confusion. Two months before he started with bilateral auricular chondritis and arthritis of metacarpophalangeal joints and ankles. He had been previously seen at another hospital, where he had been treated for herpetic encephalitis, with improvement of confusion. On examination he had downward nistagmus, rigidity of upper limbs, paraparesis, absent reflexes, tactile hypoesthesia, dimetric movements, gross postural and action tremor, bradykinesia and truncal ataxia. He also had swelling and a purplish erythema of both ear lobes and arthritis in the metacarpophalangeal joints of the right hand. Brain and cervical MRI disclosed a mild thickening of the dura. A new lumbar puncture confirmed the presence of elevated leukocytes and laboratory exams disclosed augmented inflammatory activity. A diagnosis of relapsing polychondritis was made based on the association of chondritis, arthritis and vestibular ataxia with predominant neurological symptoms. Following a course of Prednisone 1 mg/kg qid there was major improvement of chondritis, arthritis, ataxia and paraparesis, but the tremor remained unchanged.

Conclusion: Relapsing polychondritis is a multisystemic disease that can manifest not only with bilateral auricular chondritis and vestibular compromise, but also with varied neurological symptoms.

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Abstract – WCN 2013

No: 1699

Topic: 2 – Movement Disorders

Effects of botulinum toxin treatment on subjective visual vertical perception in cervical dystonia

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Background: Previous data suggest that we might abandon the idea of a single “reference frame” for verticality perception. When the head or body is tilted, otolithic and somatosensory signals can have opposite sign effects during perceiving verticality. When cervical dystonia (CD) is treated with botulinum toxin, abnormal head posture improves within 3 weeks.

Objective: This dynamic alteration of head posture is a unique model to study effects of altered somatosensory and otolithic input on static graviceptive function.

Patients and methods: Static graviceptive function was assessed by means of subjective visual vertical (SVV) judgement. Thirty patients suffering from idiopathic CD and, for control, thirteen healthy individuals were investigated. At baseline and 3 weeks after injection, patients were investigated at 6 different head positions (no fixation, fixed upright (0°), fixed deviation at 15° and 30° to the left and right, respectively). The control group was investigated at 5 head positions (0°, 15° and 30° to the left and right, respectively).

Results: At baseline, CD's SVV judgement in habitual head position vs. control's at 0° was significantly deviated ($p = 0,0166$). Three weeks after injection, there was no significant difference. Thus, disease severity and SVV aberration correlated positively (Pearson correlation: 0.61). We noticed a general trend of major SVV errors of CD patients in contrast to healthy subjects, and a trend of contraversive deviation of SVV in fixed head positions (at 30°: $p = 0,052$).

Conclusion: Verticality perception of CD differs from healthy subjects and improves after BoNT therapy. Thereby, increased neck muscle proprioception has a major influence on verticality perception.

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Abstract – WCN 2013

No: 1674

Topic: 2 – Movement Disorders

Anatomic localization of secondary dystonias: Analysis of 21 patients

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Objective: To investigate anatomical distribution, etiology and symptomatology of secondary dystonia.

Background: Although lesions of basal ganglia, especially putamen and globus pallidus are widely accepted sites for the development of secondary dystonias, structural lesions located in the centrum semiovale, thalamus, brainstem, cerebellum and peripheral nervous system are also reported sites. Analysis of lesion localization and correlation of symptoms may contribute to the relation of phenotype and pathophysiology of dystonia.

Patients and methods: 21 patients with structural lesions verified with magnetic resonance imaging (MRI) or computerized tomography were selected among 365 dystonic patients referred to our movement disorder clinic between 2001 and 2013. The patients were classified according to age onset, distribution and etiology.

Results: There were 13 patients with childhood onset and 8 patients with adult-onset. 4 patients had focal, 4 patients had segmental, 3

patients had multifocal, 6 patients had generalized and 3 patients had hemidystonia. Etiologic analysis revealed 7 patients with perinatal cerebral injury, 5 patients with infection of CNS, 4 patients with trauma, 4 patients with stroke and 1 patient with peripheral injury. 8 patients had putaminal, 6 patients had pallidal, 2 patients had putaminal and cortical, 3 patients had hemiatrophy of cerebral hemispheres. 1 patient had brainstem and 1 patient had ulnar nerve injury verified with EMG.

Conclusion: Basal ganglia are the site of predominant localization for secondary dystonia but different sites of CNS especially white matter adjacent to basal ganglia and brainstem are also involved in secondary dystonias.

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Abstract – WCN 2013

No: 1722

Topic: 2 – Movement Disorders Vascular Parkinsonism

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Aim: Identification of clinical features that suggest for vascular Parkinsonism.

Methods: In this study we included 86 patients with Parkinsonism, who were examined in the Extrapyramidal Unit of University Neurologic Service, in Tirana during 2006–2009. Clinical examination highlighted that 71 patients were with Parkinson's disease and 17 were with Parkinsonic syndrome.

We analyzed age, sex, duration of symptoms from the first visit, evidence of vascular disease, and distribution of asymmetry of the signs. We used UPDRS to evaluate the clinical signs. Response to levodopa therapy was determinant in visit of control.

The evidence of vascular disease was assessed by these indices: installation of clinical Parkinsonism signs at least one month after stroke, and personal history for last stroke before neuroimaging evidence for stroke in 2 or more territories. The data were analyzed in SPSS 12.1 software. The standard deviation of ages and duration of symptoms were calculated using student's t-test. The results were statistically significant for p value < 0.05.

Results: The vascular Parkinsonism and Parkinson's disease can differentiate easily clinically (p = 0,001 to p = 0,0001).

The patients with vascular Parkinsonism were more elderly and present more walking disorders than rest tremor and hadn't a good response to levodopa therapy compared with Parkinson's disease patients. (P = .00001).

The patients with vascular Parkinsonism present more falling, dementia, sphincter incontinence (p = 0.001) and pseudobulbar syndrome (p = 0,005), compared with Parkinson's disease patients.

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Abstract – WCN 2013

No: 1588

Topic: 2 – Movement Disorders Are there any correlations between the length of “ephedrone-abuse” and severity of depression in patients with “ephedrone”-induced Parkinsonism?

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Introduction: Severity of depression is different in patients with similar age, similar length of “ephedrone” abuse and similar severity of movement disorder. Causes of variability are unclear.

Objective: To identify the relationship between the severity of depression and length of “ephedrone”-abuse.

Patients and methods: We examined 33 male and 1 female patients with a mean age of 40.2 years (SD ± 3.8 years) and with 0.5–1.5 years history of “ephedrone”-abuse and movement disorder. We formed four groups of patients: group I – 9 patients with minimal depression; II – 5 patients with mild depression; III – 10 patients with moderate depression; and group IV – 10 patients with severe depression. Depression was evaluated according the Beck depression inventory. Movement disorder was assessed with UPDRS.

Result: In our study, a clear relationship between the severity of depression length, severity of Parkinsonian syndrome and length of “ephedrone”-abuse was not found.

Conclusion: A relationship between the severity of depression, severity of Parkinsonian syndrome and length of “ephedrone”-abuse was not shown in examined patients. Our study has limitations: the numbers of patients was small and controls of same age were not examined. Further studies are planned to assess the role of “ephedrone”-abuse in the severity of depression and movement disorder in patients with ephedrone-induced Parkinsonism.

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Abstract – WCN 2013

No: 1634

Topic: 2 – Movement Disorders Hemorrhagic syndrome in Wilson's disease patients

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Introduction: Severity of hepatic impairment due to Wilson's disease (WD), and angiotoxic and hemolytic effects of copper can significantly influence hemostasis and lead to hemorrhagic syndrome (HS). The incidence of HS and disturbances of hemostasis have not been studied before.

Objective: To investigate the incidence of HS and disturbances of platelet hemostasis and plasma coagulation in patients with WD.

Methods: 68 patients with WD and 65 patients with other liver diseases (LDG) were examined retrospectively for different signs of HS they had since the birth till the time when diagnosis was established and chelating treatment was started. 65 normal individuals were examined as the control group (CG). Parameters of platelet and plasma coagulation hemostasis were evaluated in 48 patients with WD.

Results: 52% of WD patients at the age of 3–9 years presented hemorrhagic signs and 77% at the time when diagnosis was established. The result was significantly higher than that in the CG and LDG group. The most typical hemorrhagic signs were nasal and gingival bleeding and bruises and in 68% of patients they repeated daily or weekly. 95% of patients with WD had abnormal laboratory parameters of hemostasis: thrombocytopenia, increase of intravascular platelet activation and aggregation, increased vWF activity, prolongation of APTT, decrease of PT (because of reduced activity of factors V, VII and X), decreased activity of antithrombin and plasminogen level and diminished time of factor XII-dependent fibrinolysis, and elevated levels of d-dimer.

Conclusion: Patients with WD demonstrate the high incidence of HS in comparison with patients with other liver disorders.

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Abstract – WCN 2013**No: 1632****Topic: 2 – Movement Disorders****The role of postoperative levodopa test in combining deep brain stimulation programming and medication therapy in Parkinson's disease**

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Background: In Parkinson's disease, the combination of subthalamic deep brain stimulation (STN-DBS) and medication therapy need to be adjusted to treat the clinical symptoms. This strategy is assisted by a drug challenge test, when motor symptoms are evaluated by clinical rating scales during continuous bilateral stimulation, in medication ON and medication OFF states.

Objective: We investigated if quantitative motion analysis during a postoperative levodopa test can help the therapy setting.

Patients and methods: Eleven patients (age: 61.7 ± 9.54 years, disease duration: 18 ± 6.11 years) performed 14 s of finger tapping, hand grasping and pronation–supination of the arm in three conditions (STIM ON–MED OFF, STIM OFF–MED OFF and STIM ON–MED ON) one year after bilateral STN-DBS implantation. A motion sensor consisting of a three-dimensional gyroscope and accelerometer was worn on the index finger to measure limb movement. Speed as root mean square angular velocity and amplitude as root mean square excursion angle were calculated from the signal of the gyroscopes and compared with ANOVA for repeated measures ($p < 0.05$).

Results: In case of optimal DBS settings, speed and amplitude of the three tasks did not differ significantly in the STIM ON–MED OFF and STIM ON–MED ON states and they were significantly higher than in the STIM OFF–MED OFF condition. In the single subject level, further DBS programming was needed when levodopa administration improved the speed and amplitude of the movement during bilateral STN-DBS.

Conclusion: The postoperative levodopa test assisted by quantitative motion analysis facilitates the optimal setting of DBS, so medication dose can be reduced to avoid its side effects.

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Abstract – WCN 2013**No: 1627****Topic: 2 – Movement Disorders****Pharmacokinetics of levodopa and 3-omd in patients with Parkinson's disease and motor fluctuations infected with *Helicobacter pylori***

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Background: Recent investigations suggest that eradication of *Helicobacter pylori* (*H. pylori*) may influence levodopa (LD) pharmacokinetics and improve the motor function of infected patients with Parkinson's disease (PD). Our study was aimed at comparison of pharmacokinetics of LD and its metabolite 3-O-methyldopa (3-OMD), between *H. pylori*-positive and -negative patients with Parkinson's disease and motor fluctuations.

Materials and methods: Seventy-three patients with the clinical diagnosis of PD, under stable LD therapy, reporting daily motor fluctuations and who had no history of previous eradication treatment, were screened for the infection with an antigen stool test. Thirty seven patients: 18 with *H. pylori* infection and 19 noninfected, matched both demographically and clinically, were selected for the examination of LD and 3-OMD pharmacokinetic parameters. Blood samples were collected immediately before and after morning LD dose 13 times: at 15 min intervals up to 2 h and then at 30 min intervals up to 4 h. LD and 3-OMD concentrations were determined using a high-performance liquid chromatography method (HPLC) coupled with electrochemical detection.

Results: The analysis of the pharmacokinetic parameters of LD and 3-OMD showed no significant differences between *H. pylori*-infected and noninfected groups.

Conclusions: There is no evidence for malabsorption of LD in *H. pylori*-infected patients.

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Abstract – WCN 2013**No: 1682****Topic: 2 – Movement Disorders****Incidence, prevalence and clinical pathway of Parkinson's disease among Pakistani population**

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Introduction: Parkinson's disease is an idiopathic disorder of the extrapyramidal system characterized by rigidity, akinesia and tremors. The aim of this study was to describe the clinical spectrum of the disease from Pakistan, a developing country in south-east Asia.

Methods: This study was conducted at the teaching hospitals of Peshawar Medical College, Pakistan over a period of 1 year from August 2011 to July 2012. Patients with Parkinson's disease were identified by an ICD-9 coding system of the hospital medical records. An informed consent was obtained from all the participants. Demographical characteristics, clinical features, laboratory investigations and radiological investigations were recorded and analyzed.

Results: A total of 48 patients were identified. 30 were males and 18 were females. Mean age of onset of the disease was 51 years. 28 patients had onset of illness during the sixth or seventh decade of life. Mean duration of illness at the time of presentation was six years. Rigidity, bradykinesia, tremors, primitive reflexes, and difficulty in performing fine work and walking were the most common clinical features. 36 patients had predominantly unilateral symptoms. 12 patients had cognitive impairment. Cognitive decline was more common in the elderly and in patients with disease duration of longer than 8 years.

Conclusion: Parkinson's disease is more common in males. Tremor, rigidity, walking difficulty, bradykinesia and difficulty in performing fine work are the commonest clinical features. Disease severity increases with duration of the disease. Cognitive impairment is not uncommon in these patients and is associated with disease duration and age of onset of the illness.

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Abstract – WCN 2013**No: 1685****Topic: 2 – Movement Disorders****Transcranial sonography in movement disorders**

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Background: So far there is no reliable test that can clearly distinguish between various movement disorders and therefore the diagnosis is still based on clinical examination. However, the misdiagnosis rates for the most common movement disorders: Parkinson's disease (PD) and essential tremor (ET) in the early stages is as high as 20–30% for PD and in about one of three patients in ET. We initiated this study to assess the possibility of transcranial sonography (TCS) to help differentiate PD from ET by measuring echogenicity of the substantia nigra (SN).

Patients and methods: Our study included 60 patients with PD, 30 patients with ET and 60 healthy controls. The TCS recordings were done in the axial plane by a standardized protocol by two independent investigators. SN was displayed, encircled, and measured two times. Mean area was calculated. Mann–Whitney *U* test for inter-group comparison was applied.

Results: Patients with ET and PD had a mean SN size of 0.15 cm² (± 0.04) and 0.27 cm² (± 0.06), which showed a significant difference ($p < 0.001$). In the control group bilateral combined mean SN size was 0.17 cm² (± 0.06), which was significantly different from the PD group ($p < 0.001$), but not from the ET group ($p = 0.240$).

Conclusion: The measurement of SN by means of TCS is a valuable tool in the differentiation of movement disorders. Due to portability, lack of invasiveness and ease of reproducibility, TCS might help in diagnosing PD or in differential diagnosis of doubtful clinical cases.

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Abstract — WCN 2013

No: 1573

Topic: 2 — Movement Disorders

Spinocerebellar ataxia 7 (sca7) in India: Genotype–phenotype correlation and insight into origin of mutation in a predisposed endogamous population

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Background: Spinocerebellar ataxia (SCA7) is characterized by neuro-ophthalmic degeneration and occurs due to CAG repeat expansion in exon 3 of ATXN7 in the region of chromosome 3p12-p21.1. The SCA7 mutation has been identified in various ethnic and geographical regions with highest prevalence in South Africa and in the Scandinavian region.

Objectives: To describe the clinical-genetic characteristics of nine SCA7 families of Indian origin and cross comparison with worldwide studies.

Methods: 33 individuals including 22 affected and 11 unaffected from nine SCA7 families were clinico-genetically characterized. CAG repeat distribution analysis was carried out in 375 control DNA samples from 20 diverse Indian populations based on ethnicity, linguistics and geographical location.

Results: Genotype to phenotype studies show correlation of CAG length with age at onset ($R^2 = 0.844$) and CAG repeat adjusted SARA scores with duration ($y = 0.0234x + 0.1798$; $R^2 = 0.591$). Meta-analysis of CAG repeats with disease onset reveal association of repeats <49 with earlier age at onset in Southeast Asian compared to European populations. Analysis of CAG repeats from 20 diverse Indian populations represented in the Indian Genome Variation Panel showed the presence of pre-mutable repeat (28–34) alleles in the same population (IE-N-LP2) from which SCA7 families were derived.

Conclusion: We observed quite a homogeneous phenotypic expression of SCA7 mutation with subtle differences in age at onset mediated by

CAG repeat in various geographical populations. We also speculate that there is recurrent generation of predisposed intermediate permutable 28 CAG repeats in ATXN7 across various world populations determining the occurrence of SCA7.

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Abstract — WCN 2013

No: 1542

Topic: 2 — Movement Disorders

The effect of DBS on cortico-muscular coherence in advanced Parkinson's disease

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Background: DBS is a well-established treatment for advanced Parkinson's disease (PD) even though its mechanism of action is not precisely known. Cortico-muscular coherence (CMC) is used to measure the connectivity between the motor cortex and muscle during motor activity.

Objective: Our hypothesis was that DBS would strengthen CMC when ameliorating motor functioning.

Patients and methods: We measured 19 patients with advanced PD, treated with bilateral STN-DBS, with MEG (magnetoencephalography). Simultaneously with MEG, EMG was recorded from the activated extensor carpi radialis longus muscle during wrist extension for coherence calculations. The measurements were done with both DBS on and off. UPDRS motor scores were used to evaluate the motor performance. Antiparkinsonian medication was continued during the study. For artifact removal, the spatiotemporal signal space separation was applied.

Results: 13 out of 19 patients showed CMC between 10 and 18 Hz. The effect of DBS on CMC was inconsistent; eight patients had enhanced CMC when DBS was off and five when DBS was on. The peak frequency of CMC correlated negatively with the rigidity scores of the active hand when DBS was off.

Conclusion: DBS alters CMC with great individual variety in advanced PD. We did not find linear correlation between CMC amplitude and motor performance. The peak frequency of CMC correlated negatively with the rigidity of the active hand when DBS was off.

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Abstract — WCN 2013

No: 1546

Topic: 2 — Movement Disorders

Analysis of alpha-synuclein ala53thr mutant: a computational approach

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Background: Parkinson's disease (PD) is a progressive neurodegenerative disorder due to loss of dopaminergic neurons from the substantia nigra pars compacta of the CNS. Mutations of alpha-synuclein (AS), a protein involved in the formation of synaptic vesicles of dopamine, are a cause of PD. AS has a native disordered state that undergoes α -helical conformation upon interaction with membrane lipids. The neurophatological hallmarks of PD are "Lewy bodies". They are made of aggregated AS that are potentially toxic agents.

Objective: Investigate with computational approach AS mutants.

Material and methods: We performed a statistical study of a known natural variant and other experimental mutations of SNCA gene coding for AS linked with familial PD using PASTA software. Among them we selected the Ala53Thr missense mutation and performed a molecular dynamics (MD) investigation.

Results: The prediction with PASTA correlates with literature. In particular, this approach reveals that the Ala53Thr mutation increases the tendency of AS to fold in a β -secondary structure altering the functionality of the protein. The MD analysis shows that in the cytoplasm the AS structure is unstable.

Conclusion: Wild-type AS presents an α -helical conformation in membrane lipids but is disordered in cytoplasm. The mutation Ala53Thr increases the probability that AS fibrillizes impairing the binding to membrane lipids. This data may support key neurophatological events in PD: the lack of dopamine release because of the absence of AS mutant in the membrane lipids; and the presence of Lewy bodies due to AS mutant aggregation in the cytoplasm.

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Abstract – WCN 2013

No: 1532

Topic: 2 – Movement Disorders

Functional imaging of the default mode network in cognitively impaired patients with Parkinson's disease

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Background: While there is a good evidence for an altered default mode network (DMN) in both Alzheimer's disease (AD) and amnesic mild cognitive impairment preceding AD, there is rather conflicting data available for changes in the DMN in Parkinson's disease with cognitive impairment (PD–CI).

Objective: We studied changes in the DMN connectivity when switching from baseline to the task condition using fMRI and 2 groups of participants: 18 PD–CI and 18 age-matched healthy controls (HCs).

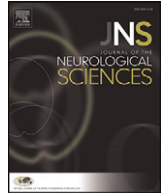
Patients and methods: Based on our previous work in PD and PD–dementia, the cognitive task was designed to test visual memory encoding. A connectivity analysis was conducted using the psychophysiological interaction (PPI) method with the posterior cingulate cortex (PCC) as a seed. The threshold was set at $p < 0.05$, FWE corrected.

Results: In the HC group we found greater connectivity between the PCC and both posterior and anterior nodes of the DMN during the baseline than during the task. PD–CI showed increased connectivity with the left premotor cortex and bilateral parietal cortices during the task as compared with baseline. Furthermore, a reversed pattern of correlations/anti-correlations between PCC and bilateral superior/inferior parietal lobules was observed in PD–CI as compared to that in HC.

Conclusion: We demonstrated that PD–CI expresses task-specific changes in the DMN connectivity that differ from HC and involve the regions specifically engaged in the studied task, i.e. attentional and visual processing.

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Topic: 2 - Movement Disorders

Abstract – WCN 2013

No: 1527

Topic: 2 – Movement Disorders

Idebenone in treatment of patients with Huntington's chorea and hereditary myopathy

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Introduction: The treatment of Huntington's chorea and hereditary myopathy is absent. We studied the efficacy of energotropic therapy.

Materials and methods: A total of 18 adult patients with the Huntington's chorea, 5 Huntington's chorea gene carriers, and 12 with hereditary myopathy were studied. Cytochemical analysis of the peripheral blood lymphocytes was performed.

Results: In patients with Huntington's chorea and gene carriers the levels of lactate and mitochondrial enzymes were changed. Idebenone 90 mg a day was prescribed. 1–6 months later the mean MMSE score grew from 21.1 to 23.9. In mild hyperkinetic syndrome, its slight decrease was noted. Medication was also prescribed to the carriers of the Huntington's chorea gene to postpone the time of the clinical manifestations of the disease and diminish the disease severity. 12 patients with hereditary myopathy took Idebenone in the dose of 90 mg/day. The power growth of the deltoid muscle and that of the shoulder biceps muscle were statistically reliable. The power of the other muscles also increased but less significantly. Before treatment with drug, the patients could squat, on the average, 5.1 ± 3.1 times; and 1 month later – 6.0 ± 3.1 times.

Conclusion: In patients with Huntington's chorea, Huntington's chorea gene carriers and hereditary myopathy changes in mitochondrial function are revealed which requires energotropic therapy. Idebenone is an effective drug to treat cognitive disturbances in patients with Huntington's chorea and muscular strength in patients with hereditary myopathy.

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Abstract – WCN 2013

No: 1534

Topic: 2 – Movement Disorders

Analysis parameters of cerebral morphometry in cerebrovascular and Parkinson's Diseases

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Objective: Cerebral volumetric morphometry was studied for revealing the signs of the neurodegenerative process in PD and cerebrovascular disease (CVD) patients using 3 T MRI.

Patients and methods: Volume values of cerebral substance and liquor, substantia nigra (SN), and red nucleus (RN) were determined. Under examination there were 10 PD patients with I stage (by Hoehn and Yahr's scale) (PD1), 10 PD patients with III stage (PD2), 10 patients with early CVD (CVD1), 10 – with expressed post-stroke CVD (CVD2), and 10 persons – control (C).

Results: Analysis of supratentorial structures revealed significant decrease of cerebral volume in CVD and PD2. An increasing volume of the subarachnoid space was found in both CVD and PD as compared to the control (C). The volume of cerebral ventricles in CVD exceeded C values. Investigation of midbrain structures showed that SN volume in PD1, PD2, and CVD1 (on T2) didn't differ from C, but in CVD2 it was less than in C. While in CVD progression SN volume decreased, in PD – it increased. RN volume was decreased in all patient groups as compared to the control (C).

Conclusion: In PD and CVD, the signs of neurodegenerative process were revealed manifesting themselves in decrease of the cerebral substance volume, extension of liquor spaces, and decreasing the midbrain structures. But it has significant differences of PD and CVD. Increase of the SN volume in PD is likely due to deposition of ferritin. The data can be helpful for the early diagnostics of the PD.

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Abstract – WCN 2013

No: 1531

Topic: 2 – Movement Disorders

What influence most on the quality of sleep in Parkinson's Disease patients?

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Introduction: Sleep disturbances are one of the most common nonmotor symptoms of Parkinson's Disease (PD) and its prevalence vary from 60 to 98%.

Aim: To investigate incidence of sleep disturbances in patients with idiopathic PD and to evaluate what kind of sleep disturbance influence most on the quality of the sleep.

Patients and methods: We analysed 104 PD patients treated on the University Hospital Centre Osijek and General Hospital Nasice. All patients were asked if they have sleep disturbance or not. To evaluate sleep disturbances we used Parkinson's Disease Sleep Scale (PDSS).

Results: Sleep disturbance was reported by 58.7% patients, while 41.3% did not have sleep disturbance. Mean total PDSS score was 94.6 ± 30.1 . The lowest scores were on items evaluating nocturnal awakenings (2.8) and nocturia (2.6). In a group of patients reported not to have sleep disturbance total PDSS score was 104.4 ± 28.8 (lowest scores were on items evaluating nocturnal awakenings (3.9) and nocturia (3.3)), while in a group that had a disturbance the score was 87.7 ± 29.3 (lowest score were also on items evaluating nocturnal awakenings and nocturia (2.0)). We found statistically significant difference between two groups of patients on this items: nocturnal awakening ($t = -2.869$; $p < 0.005$), restlessness ($t = 1.941$; $p < 0.01$), distressing dreams ($t = -2.947$; $p < 0.005$), distressing hallucinations ($t = -1.956$; $p < 0.05$), tremor on waking ($t = -2.135$; $p < 0.05$) and morning tiredness ($t = -2.494$; $p < 0.01$).

Conclusion: Among sleep disturbances nocturnal awakening, restlessness, distressing dreams, distressing hallucinations, tremor on waking and morning tiredness influence most on the quality of sleep in patients with Parkinson's Disease.

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Abstract – WCN 2013

No: 1539

Topic: 2 – Movement Disorders

Treatment of diphasic dyskinesias in Advanced Parkinson's Disease with Duodopa

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Objective: This is an overview of two patients with Advanced Parkinson's disease (PD) and diphasic dyskinesias that have been treated with Duodopa.

Patients and methods: A clinical study has been carried out by presenting the history and the progress of the disease, the emergence of movement complications and dyskinesia. An attempt for clinical analyses of the diphasic dyskinesias was made, as well as for their recognition and differentiation from the peak-dose dyskinesias. A graphic illustration of the daily rhythm of the patients' condition is shown herein. The way the patients walk is demonstrated with short videos.

Results: The diphasic dyskinesias affect mainly the lower limbs and often change the way the patient walks, which sometimes may look 'strange' and may be associated with psychogenic movement disorders. The diphasic dyskinesias differ from the peak-dose dyskinesias. The two types of dyskinesias are often mixed and a different approach must be applied to each one of them. The general rules and strategies for treatment of the diphasic dyskinesias with Duodopa are outlined herein.

Conclusion: As evident from the clinical study, the diphasic dyskinesias are much more serious, as well as much more difficult to recognize and to treat.

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Abstract – WCN 2013

No: 1509

Topic: 2 – Movement Disorders

Fahr's syndrome: A Tunisian cohort and review of the literature

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Background: Fahr's syndrome is characterized by symmetrical and bilateral intracerebral calcifications, located in the basal ganglia. It's

mostly associated with a phosphorus calcium metabolism disorder, but could have other etiologies.

Objective: To report clinical and paraclinical manifestations in patients with Fahr's syndrome, to determine the etiologies and to describe their treatment and outcome.

Patients and methods: We conducted a retrospective study over an 11-year period including all patients diagnosed with Fahr's syndrome and their first degree relatives followed for neurologic manifestations. Demographic data, neurologic examination, imaging, and treatment were analyzed.

Results: We identified 14 patients diagnosed with Fahr's syndrome, and two first degree relatives with neurologic manifestations (sex ratio = 0.6, mean age = 47.9 years, mean age of onset = 38.8 years). Family history revealed neurologic troubles (10) and endocrine diseases (10), with consanguinity in 9 patients. Movement disorders were the most frequent inaugural manifestation (5) and finding on examination (7). Neuropsychological assessment revealed mainly dysexecutive troubles (7). Brain imaging showed calcifications mainly in pallidum (6). Fahr's syndrome was secondary in 11 patients, mainly due to dysparathyroidism (8). Two families with Fahr's disease were identified.

Conclusion: The high consanguinity rate and the frequent neurologic and endocrine family history in our series suggest a genetic origin of Fahr's disease or endocrine diseases that could lead to Fahr's syndrome. Further genetic studies are needed to elucidate the causative genes implicated in this rare disease. However, a battery of biological tests should be systematic in order to rule out a secondary origin of Fahr's syndrome.

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Abstract – WCN 2013

No: 1501

Topic: 2 – Movement Disorders

Behavioral impairment in Parkinson's Disease in the Siberian region

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Background: A recent large study found that Impulsive behavioral disorders (IBDs) occur in approximately 15% of patients with Parkinson's disease (PD). IBDs may lead to significant impairments in psychosocial functioning, interpersonal relationships, physical health, and life quality. Patients and relatives may not report such behaviors to a treating physician, perhaps due to embarrassment, not suspecting an association with PD treatment, or ambivalence regarding ceasing the behavior. Hence, there is evidence that IBDs aren't always recognized in routine clinical PD care.

Objective: Examination of tools for screening IBDs in PD patients at the common practice in Movement Disorders clinics.

Material and methods: Data was collected from 3 centers across the Siberian region of Russia (Tomsk, Kemerovo, Novokuznetsk). 834 medical records were surveyed for IBDs screening and the tools used.

Results: 106 of 834 patients with movement disorders (12.7%) were screened for behavioral impairment, 98 of whom (11.75%) had QUIP-RS, 8 – other tests (including QUIP). As a result 56 of 98 patients (57.14%) had IBDs, 27 (27.55%) – 2 or more IBDs. 83 of 426 PD patients (19.48%) were screened for behavioral impairment. As a result 51 of 83 patients (61.14%) had IBDs.

Conclusions: IBDs screening was done only in 19.48% of PD patients. QUIP-RS being the most commonly used screening tool. Due to the fact that the IBDs aren't always recognized in routine clinical PD care and its association with low quality of life, we recommend routine behavioral screening with a sensitive tool like QUIP-RS to aid in the comprehensive management of all PD patients.

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Abstract – WCN 2013

No: 1498

Topic: 2 – Movement Disorders

Clinical and genetic aspects of torsion dystonia in the Moscow region

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Objective: Studying specific weight of the family cases of torsion dystonia (TD) among all TD cases in the Moscow Region and phenotypical features of patients according to dermatoglyphics data.

Materials and methods: The register of patients with TD (n = 336) was created for the Moscow Region population. A contact method of dermatoglyphics was conducted.

Results: Among 336 patients with TD, 4 patients with repeated familial disease were revealed. In the first family, it was father and son, and in the second it was mother and daughter. In the first family, dystonic dysphagia was found in the father, and spastic torticollis in the son. In the second family, both mother and daughter had spastic torticollis. Specific weight of family cases accounted for 0.12% among the whole number of involved patients. The other family was revealed also, in which the son had generalized TD. DYT1 form of the disease was diagnosed based on DNA analysis. The father also had a mutation of the DYT1-gene but clinical manifestations were absent. Dermatoglyphics (studying skin patterns on the palm surface) was performed in 14 patients with TD. The rate of simple patterns on the fingers (A + T arches) decreased, and the rate of more complex patterns increased (radial loops R and curls W).

Conclusion: Thus, the specific weight of family cases among all TD patients was 0.12%. TD is inherited by an autosomal dominant type. It was revealed that there was incomplete penetration of the pathological gene in DYT1 patients. Dermatoglyphics showed an increased rate of more complex patterns (radial loops R and curls W) on the patients' fingers.

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Abstract – WCN 2013

No: 1497

Topic: 2 – Movement Disorders

Mindfulness in Parkinson's Disease leads to increases in gray matter density

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Background: PD consists of motor and non-motor symptoms. There is a need to objectively analyze non-pharmacological interventions used to help alleviate symptoms in PD.

Objective: This study was conducted to evaluate structural brain gray matter changes following a mindfulness based intervention (MBI) in Parkinson's disease (PD).

Methods: Twenty-seven of 30 PD patients completed this longitudinal RCT comparing MBI, consisting of a structured 8-week program of mindfulness training (n = 14), with usual care (UC) (n = 13). MRI 3D T1 weighted (1 × 1 × 1 mm³ resolution, Siemens 3 T System) data sets were obtained at baseline and after 8 weeks. Voxel based analysis was performed using DARTEL from the SPM8 software. Segmented gray matter baseline images were subtracted from the follow-up scan. Resulting difference maps of MBI and UC groups were statistically compared, to examine gray matter density differences in time between both groups. Results were reported at p < 0.001, uncorrected for multiple comparisons.

Results: Increased gray matter density was found in the MBI group compared to the UC group over time in the right caudate, left and right hippocampus and right amygdala. On the other hand, increased gray matter differences were found in the UC compared to the MBI group in the left cerebellum.

Conclusions: This is the first quantitative analysis of neurobiological effects of mindfulness in PD to the best of our knowledge. Increased gray matter density was found in the MBI group in the neural networks that have been postulated to play a role in mediating benefits of meditation.

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Abstract – WCN 2013

No: 1492

Topic: 2 – Movement Disorders

Excessive daytime sleepiness in patients with Parkinson's disease

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Background: Excessive daytime sleepiness (EDS) is one of the most commonly reported sleep complaints in patients with Parkinson's disease (PD), affecting between 15% and 50% of PD patients. There are conflicting reports about the association of Hoehn and Yahr (H&Y) stage and the degree of EDS.

Objective: To determine the stage related distribution of EDS in PD patients.

Material and methods: 426 PD patients are registered in electronic database of movement disorders of the Siberian region. The diagnosis of PD was made using the criteria established by UK Brain Bank. The severity of PD was estimated using H&Y Scale. The Montreal Cognitive Assessment (MoCA) was used for evaluating cognitive impairment, the 39-Item Parkinson's Disease Questionnaire (PDQ-39) – for quality of life, the Hospital Anxiety and Depression Scale (HADS) – for evaluating anxiety and depression, the Epworth Sleepiness Scale (ESS) – for quantifying EDS.

Results: We selected 76 patients with the equivalent mean daily dose of medications, normal cognition, without clinically significant anxiety and depression. All patients were divided into three homogeneous groups: I – 26 (H&Y 2.0), II – 25 (H&Y 2.5), III – 25 (H&Y 3.0). Sleep-related problems were most prominent in patients in advanced disease (p < 0.05). Among the III group 56% of patients had EDS, II – 36%, I – only 19%. A lower quality of life was found in patients with EDS when compared with patients without sleepiness on the following indexes of the PDQ-39: mobility, activity, cognitive (p < 0.05).

Conclusions: Sleep-related problems were most prominent in patients with severe, advanced disease.

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Abstract – WCN 2013**No: 720****Topic: 2 – Movement Disorders****The contemporary challenges and clinician-scientists' perspective in translational medicine neuroscience research 'new pathways of discovery': From bench to bedside**

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Neurodegenerative diseases affect millions of people worldwide leading to a growing concern over the public health implications. Major gaps in our knowledge base hinder the delivery of optimal medical care to affected individuals. Translational Medicine research (TMr) may fill the gaps at the point of development of modern treatment for not only morbidities but also comorbidities. Antioxidants are revolutionizing TMr contemporary and its future directions towards promising therapy. Conventional medicinal researches and use of antioxidants have highlighted the progressive need to restructure approaches in order to facilitate the efficient integration and translational of new technologies into novel effective therapeutic. Conventional scientists-clinicians have been driving research but cultural differences between these two groups of investigators – practitioners largely stem from less information about TMr, the lack of communication, differences in education and training from developed to developing countries. Contemporary gaps outline the recent initiatives to overcome challenges that expected to strengthen TMr, catalyze the generation of innovative treatment, revise diagnosis, drive rapid benefits from the lab-bench to the patient-bedside and generate tremendous research foci.

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Abstract – WCN 2013**No: 1491****Topic: 2 – Movement Disorders*****Nigella Sativa* oil controls astrogliosis, parkinsonism and oral dyskinesia in haloperidol-treated rats**

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Background: The symptoms of parkinsonism and oral dyskinesia have been showing to be induced by neuroleptics in the striatum.

Objective: This study deals with the effects of haloperidol (HAL) on *Nigella sativa*-oil (NS) treated rats in duration of 6 weeks.

Results: HAL treated rats showed movement disorders followed by oral dyskinesia. The cytoarchitectonic pattern of striatum showed astrogliosis. Astrogliosis was observed in caudate and accumbens nuclei. Such effects of HAL was absent in rats treated with NS-oil showing it normal and glial cells protective.

Conclusion: It may be concluded that symptoms of parkinsonism, oral dyskinesia can be prevented by treatment of rats with NS-oil. How far this study can be appreciated to humans needs to be elucidated.

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Abstract – WCN 2013**No: 1468****Topic: 2 – Movement Disorders****Rifampicin inhibits neuron apoptosis through suppression of the TLR4 pathways in LPS-stimulated co-cultured BV2 Cells**

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Agents suppressing microglial activation are attracting attention as candidate drugs for neuroprotection in neurodegenerative diseases. Recently, researchers have focused on the immunosuppression induced by rifampicin. Our previous study has shown that rifampicin inhibits microglial inflammation and improves neuron survival against inflammation; however, the mechanism through which rifampicin inhibits the microglial inflammation and its neuroprotection are not completely understood. This study we examined the effects of rifampicin on morphological changes induced by LPS in murine microglial BV2 cells. Then we investigated the effects of rifampicin on signaling pathways stimulated by lipopolysaccharide (LPS) including Toll-like receptor-4 (TLR-4) and nuclear transcription factor kappa B (NF- κ B) in BV2 microglia. In addition, we cocultured BV2 microglia and neurons to observe the indirect neuroprotective effects of rifampicin. Rifampicin inhibited LPS-stimulated expression of TLR-4. When neurons were cocultured with LPS-stimulated BV2 microglia, pretreatment with rifampicin increased neuronal viability and reduced the number of apoptotic cells. Taken together, these findings suggest that rifampicin, with its anti-inflammatory properties, might be a novel treatment for neurodegenerative diseases.

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Abstract – WCN 2013**No: 1446****Topic: 2 – Movement Disorders****Compassionate use of doxycycline in****Gerstmann–Straeussler–Scheinker syndrome.****Case report on a descendant of the original family**

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Background: GSS is a rare, autosomal dominant inherited prion-disease, first reported in an Austrian pedigree in 1936. It is characterized by progressive cerebellar ataxia and late onset of dementia, mainly caused by a point mutation at codon 102 of the prion protein (P102L).

This patient descends from the originally described Austrian GSS family. His mother had suffered from progressive dementia and died at the age of 41 years. Genetic analysis showed a P102L mutation with homozygosity for methionin at codon129. At the age of 27 years our patient developed progressive unsteadiness of gait, followed by mild dysarthria, ataxia, areflexia and pos. pyramidal signs after 18 months. CMRI showed moderate cerebellar atrophy, FDG PET revealed cerebellar hypometabolism. CSF showed a weak positive signal for 14–3–3 protein and genetic testing a P102L mutation with heterozygosity at codon 129. Over the following 2 years ataxia progressed rapidly, with an increase of the SARA score from 11 (12/08) to 20.5 (3/10).

Methods: Based on preclinical evidence that tetracyclines show anti-prion activity, a trial of doxycycline(100 mg/d) was started.

Results: The following 33 months, doxycycline was tolerated without side effects. Symptom progression appeared to slow during the first 10 months of treatment (SARA 22.5; 12/10) but was similar to the pretreatment period thereafter (SARA 33; 12/12).6 years after onset, the patient is wheelchair-bound with a severe flaccid paraparesis, upper-limb-ataxia, dysarthria and dysphagia.

Conclusion: Based on the quite linear progression of SARA scores, a substantial therapeutic effect of doxycycline in our patient seems unlikely. The treatment with low-dose doxycycline was well tolerated. The SARA appears to be useful to monitor the progression of ataxia in GSS, though assessment of lower-limb-ataxia in advanced disease was confounded by muscle-weakness which is often observed in advanced GSS.

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Abstract – WCN 2013

No: 1448

Topic: 2 – Movement Disorders

Facial movement disorders due to posterior fossa lipoma: Report of 3 cases

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Objective: To demonstrate rare but important cause of refractory hemifacial spasm and blepharospasm. Blepharospasm, an involuntary blinking and inability to keep eyes open and Hemifacial spasm which is defined as irregular involuntary painless clonic contractions of hemifacial muscles are both not rare conditions. Both conditions are more frequent in females. Common cause of hemifacial spasm is an aberrant loop branch of basilar artery that compresses facial nerve and causes focal demyelination which results to ephaptic transmission and spasms and other etiologies are multiple sclerosis-regeneration phase and sequel of Bells palsy-tumors of 8th nerve-aneurysm of basilar artery. Most cases of blepharospasm are idiopathic. Here we describe 2 patients with hemifacial spasm and one case of blepharospasm which all 3 cases had parapontine lipoma at posterior fossa.

Patients: Two patients were female with long standing hemifacial spasm and other was male with chronic refractory blepharospasm. All 3 patients were treated with anticholinergics – Tegretol–gabapentin–baclofen–phenytoin and multiple injections of Dysport with incomplete responses. Serum biochemistry, ESR, CBC, CT of brain without contrast and EEG all were normal. Brain MRI with and without GD demonstrated extra-axial parapontine masses with signal intensity consistent for lipoma. Patients referred to neurosurgeon.

Conclusion: These cases reveal the importance for attention to any patient with hemifacial spasm or blepharospasms for performing brain MRI to rule out CPA and posterior fossa masses as a cause of hemifacial spasm. Thus these syndromes should not necessarily be considered idiopathic anytime.

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Abstract – WCN 2013

No: 591

Topic: 2 – Movement Disorders

Pseudo-dominant' inheritance in Friedreich's ataxia: Clinical and genetic study of a Brazilian family

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Objective: To evaluated a family with pseudo-dominant inheritance in Friedreich's ataxia (FRDA).

Background: FRDA is an autosomal recessive inherited disorder characterized by progressive ataxia, hypertrophic cardiomyopathy, skeletal abnormalities, areflexia, loss of vibratory and position sense. A rare pattern is the pseudodominant pattern with phenotypic variation.

Methods: We evaluated a Brazilian family of Italian descent with marked variation of phenotype.

Results: The father developed progressive ataxia at 30 years of age; at 68 years he had dysarthria, dysphagia, generalized ataxia, dysmetria, loss of deep tendon reflexes, was unable to sit without support, walk or stand. MRI was normal and echocardiogram showed cardiomyopathy. DNA analysis showed two expanded alleles with more than 700 GAA repeats in each one. Of three children, two were affected. A 39-year-old man began to have difficulties with balance at the age of 22, followed by impaired speech, dysphagia, horizontal nystagmus, head tremor, loss of vibratory sensation and tendon reflexes, truncal ataxia and couldn't stand even with support. He also had scoliosis and cardiomyopathy. He had one allele with 700 GAA repeats and another with 900 repeats. The index case first exhibited clumsiness in handwriting at the age of 26, followed by progressive unsteadiness of gait, slurred speech, square wave jerks, postural tremor in the hands and loss of knee reflex. MRI and echocardiogram were normal. DNA analysis showed one allele with 350 GAA repeats and another with 1000 repeats.

Conclusions: We present a family with pseudodominant inheritance of FRDA, focusing on the molecular basis of intra-familial clinical polymorphism.

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Abstract – WCN 2013

No: 1403

Topic: 2 – Movement Disorders

Spinal involvement in cerebrotendinous xanthomatosis

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Background: Cerebrotendinous xanthomatosis (CTX) is a rare, AR metabolic disease originating from deficiency of sterol-27 hydroxylase which results in storage of cholestanol and cholesterol in lens, CNS and tendons. Most common neurological manifestations are MR, pyramidal signs, cerebellar ataxia, dysphagia, dysarthria and PNP. Usual cranial imaging findings include cerebral atrophy and signal changes in cerebellum and less commonly in basal ganglia.

Objective: We aim to present and emphasize spinal involvement in CTX.

Patient: We report a 30 year-old woman with truncal ataxia since childhood. Other neurological findings are pyramidal findings in lower extremities and loss of vibration sense distal to the patella. She had previously undergone surgery for bilateral early-onset cataracts and tumor-like enlargement of Achilles tendon. Pathological diagnosis of the latter material was xanthoma. There was no similar family history. MRI showed increased signal intensity in basal ganglia, dentate nucleus, pons, medulla oblongata and posterior and lateral columns of spinal cord. History of juvenile cataracts and tendon xanthoma suggested CTX and she is still under the genetic assessment.

Conclusion: CTX is a rare disorder and spinal cord involvement is a rarer presentation. Verrips and colleagues (1999) reported seven patients and suggested use of term, 'spinal xanthomatosis'. Those patients had predominant chronic myelopathic findings with no or mild cerebellar findings. However, our patient had both cerebellar and spinal cord findings. Despite the presence of early onset cataracts and pathological confirmation of xanthomas, diagnosis was still delayed. Spinal variant should be included in differential diagnosis in patients with signal changes of posterior and lateral columns.

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Abstract – WCN 2013**No: 1420****Topic: 2 – Movement Disorders****Interleukin-6 and fatigue syndrome in Parkinson's disease**

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Introduction: Fatigue syndrome is one of the non-motor symptoms of Parkinson's disease (PD) that significantly reduces the quality of life. The pathophysiology of this symptom is poorly studied.

Objective: To assess the potential role of proinflammatory cytokine interleukin-6 (IL-6) in the development of fatigue syndrome in PD.

Methods: We determined the level of IL-6 in the serum of 65 patients with Parkinson's disease by immunosorbent assay. To determine the severity of fatigue syndrome, we used Multidimensional Fatigue Inventory (MFI-20). Operation compared with the concentration of IL-6 in serum was compared with the severity of fatigue.

Results: Fatigue syndrome was diagnosed in 30 patients (46%). Mean severity of fatigue in patients with PD was 58 (49–66) points, which was significantly ($p = 0.000$) higher than in the control group (33, 30–36 points). The value of IL-6 was significantly ($p = 0.012$) higher in patients with the FS (1.0, 0.5–1.9 pg/ml) than without this syndrome (0.35 0.0–0, pg/ml). There was a significant correlation between IL-6 with physical fatigue ($R = 0.42$; $p = 0.000$), reduced activity ($R = 0.46$; $p = 0.000$), reduced motivation ($R = 0.24$; $p = 0.051$), and general fatigue ($R = 0.52$; $p = 0.000$).

Conclusion: Fatigue syndrome is present in 46% of patients with PD, being a very common and important symptom. Patients with fatigue syndrome have higher levels of IL-6 in serum. The higher level of serum proinflammatory cytokines in PD patients leads to the increasing of the fatigue symptoms.

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Abstract – WCN 2013**No: 1367****Topic: 2 – Movement Disorders****Autonomic dysfunction in patients with early untreated Parkinson's disease**

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To determine whether the autonomic nervous system is involved in the early stage of Parkinson's disease (PD), we evaluated orthostatic blood pressure regulation, heart rate variability, skin vasomotor function and sympathetic sweat responses (emotional sweating) in 50 untreated PD patients (age 64.3 ± 8.8 years, disease duration 1.8 ± 1.4 year) and compared them with 20 healthy control subjects (age 63.6 ± 7.9 years). Five PD patients fulfilled the criteria for orthostatic hypotension. The mean decrease in systolic blood pressure during head-up tilt in PD patients was mildly but significantly lower than in controls ($p = 0.0001$). There were no differences between the 2 groups in heart rate variability, with analysis of low frequency (LF; mediated by baroreflex feedback), and high frequency (HF; an index of parasympathetic vagal activity) modulation. However, LF/HF, an index of cardiac sympatho-parasympathetic balance, was significantly lower in the PD group than in controls ($p = 0.02$). Amplitudes of sympathetic sweat responses were significantly lower than in controls ($p < 0.01$), with negative correlations with modified Hoehn & Yahr stage. Amplitudes of skin vasomotor reflexes in PD patients did not differ from controls. Our study in early untreated PD patients, without the confounding effect of drugs, suggests impairment of sympathetic cardiovascular and

sudomotor function with orthostatic dysregulation of blood pressure control, reduced LF/HF (parasympathetic dominance of cardiac sympatho-parasympathetic balance) and reduction in sympathetic sweat responses.

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Abstract – WCN 2013**No: 1365****Topic: 2 – Movement Disorders****Total antioxidant activity, glutathione and lipid peroxidation in neurologic Wilson disease: Clinical, biochemical and MRI correlation**

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Background: Increased free copper in neurologic Wilson disease (WD) is a potent oxidant but there is no study evaluating the role of glutathione, total antioxidant activity (TAC) and lipid peroxidation in them.

Objective: To evaluate TAC, glutathione and malondialdehyde (MDA) in neurologic WD and correlate their levels with clinical, MRI and biochemical changes. Subjects and methods: 28 patients with neurologic WD whose median age was 16 years were included. Neurological severity was categorized on a 0–3 grade. Cranial MRI, blood counts, hemoglobin, liver and kidney function tests, ultrasound abdomen, serum ceruloplasmin, serum free copper and 24 hour urinary copper were done. Plasma glutathione (GSH), TAC and malondialdehyde (MDA) were measured in the patients and 64 matched controls.

Result: The plasma GSH (2.26 ± 0.49 vs 2.6 ± 0.39 mg/dl $p < 0.001$) and TAC levels were reduced (1.63 ± 0.02 Vs 2.09 ± 0.03 mmol Troloxequivalent/l; $P < 0.001$) and MDA increased (4.90 ± 0.74 vs 3.21 ± 0.69 nmol/ml $p < 0.001$) in the neurologic WD compared to controls. The GSH ($r = 0.54$) level positively correlated with serum ceruloplasmin and negatively with free serum copper ($r = -0.52$) and urinary copper ($r = -0.75$). TAC correlated with duration ($r = -0.39$) and ceruloplasmin ($r = 0.48$). MDA level correlated negatively with serum ceruloplasmin ($r = -0.61$) and positively with serum free copper ($r = 0.41$) and urinary copper ($r = 0.61$). The clinical severity and hematological, liver, renal and MRI findings however did not correlate with GSH, TAC and MDA levels.

Conclusion: Lower GSH, TAC and higher MDA levels suggest copper induced oxidative injury in neurologic WD. The role of adjunctive antioxidant in the treatment of neurologic WD needs to be explored.

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Abstract – WCN 2013**No: 1381****Topic: 2 – Movement Disorders****A rare case of osteodural arteriovenous fistulas with focal hand myoclonous presentation**

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Introduction: Dural arteriovenous fistulas (DAVFs) are found approximately 10–15% of all vascular abnormalities. The most of DAVFs have leptomeningeal reflux and vascular congestion. Symptoms of DAVFs may be characterized either nonaggressive (e.g. tinnitus) or aggressive symptoms (e.g. intracerebral, subarachnoid hemorrhage

and many neurological deficits). The focal hand myoclonus is a rare presentation of neurological deficit.

Case: A 58 year-old female presented with weakness of the right arm with myoclonus of the right hand for 1 month. The other extremities were normal. She didn't have symptom of headache. The MRI brain showed cytotoxic edema at the left superior fronto-parietal area. The cerebral angiography showed moderate hypertrophy of the left middle meningeal artery shunting into the left fronto-diploic vein, which drain into the posterior left frontal cortical vein. The study was compatible with osteodural arteriovenous fistula and focal venous congestion at the left fronto-parietal region. She took valproic acid and clonazepam but was not able to relieve the focal hand myoclonus. She developed progressive weakness of the right arm, then the urgent transarterial glue embolization was performed. The procedure showed successful glue embolization with nearly complete obliteration of the arteriovenous shunt and less venous congestion. The several days later, the power motor of the right arm was improved and the myoclonus of the right hand disappeared.

Conclusion: DAVFs with cortical venous congestion demonstrates the variety of neurological manifestation. The lesions are aggressive and need prompt diagnosis and treatment.

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Abstract – WCN 2013

No: 1357

Topic: 2 – Movement Disorders

Effectiveness of acupuncture and bee venom acupuncture in idiopathic Parkinson's disease

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Background: In light of the significant limitations of conventional therapy, interest is increasing in complementary and alternative therapies in Parkinson's disease.

Objective: This study aimed to explore the effectiveness of acupuncture and bee venom acupuncture as adjuvant therapies for idiopathic Parkinson's disease (IPD).

Methods: We recruited 43 adults with IPD who had been on a stable dose of antiparkinsonian medication for at least 1 month. They were randomly assigned to 1 of 3 groups: acupuncture, bee venom acupuncture (BVA), or control. All participants were assessed using the Unified Parkinson's Disease Rating Scale (UPDRS), the Parkinson's disease quality of life questionnaire, the Beck Depression Inventory (BDI), the Berg Balance Scale (BBS), and the time and number of steps required to walk 30 m. Treatment groups underwent acupuncture or bee venom acupuncture twice a week for 8 weeks. The initial assessment was repeated at the completion of treatment. The control group did not receive any treatment.

Results: Participants in the BVA group showed significant improvement on the UPDRS (total score, as well as parts II and III individually), the BBS, and the 30 m walking time. When compared to the control group, the BVA group experienced significantly greater improvement on the UPDRS. In the acupuncture group, the UPDRS (part III and total scores) and the BDI showed significant improvement. The control group showed no significant changes in any outcome after 8 weeks.

Conclusion: In this pilot study, acupuncture and bee venom acupuncture showed promising results as adjuvant therapies for Parkinson's disease.

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Abstract – WCN 2013

No: 1361

Topic: 2 – Movement Disorders

Triple stimulation technique could be useful differential marker between vascular parkinsonism and Parkinson's disease

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Objective: Since the concept of vascular parkinsonism (VP) had been introduced, many studies have been performed to identify some instrumental tools and clinical features useful to distinguish VP from Parkinson's disease (PD). However, VP remains difficult to distinguish from PD. In this study, we aimed to identify the quantitative difference of upper motor neuron impairment between two disease entities as a differential marker.

Methods: We studied 20 patients with VP diagnosed by the eligible criteria, 21 patients with PD, and 11 healthy controls. We performed transcranial magnetic stimulation (TMS) and triple stimulation technique (TST) in these patients and analyzed the results of TST in association with clinical aspects.

Results: In VP patients, mean TST amplitude ratio was 64.7 (\pm 23.9). While, PD group revealed 90.2 (\pm 9.8) of TST amplitude ratio. Other parameters showed no significant difference.

Conclusions: TST amplitude ratio can be used to differentiate VP and PD with a good degree of certainty.

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Abstract – WCN 2013

No: 884

Topic: 2 – Movement Disorders

Effect of opicapone and entacapone on levodopa pharmacokinetics when administered with immediate release 100/25 mg levodopa/carbidopa in healthy subjects

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Background: Opicapone (OPC) was developed to fulfil the need for more potent, safer and longer acting COMT-inhibitors.

Objectives: The objective of this study is to investigate the effect of once-daily 25, 50 and 75 mg OPC on the levodopa pharmacokinetics (PK), in comparison to placebo and 200 mg entacapone (ENT).

Methods: Single-centre, randomized, double-blind, gender-balanced, placebo-controlled study in 4 groups of 20 subjects each. This study consisted of a once-daily administration of OPC or placebo for 11 days and 100/25 mg levodopa/carbidopa, 200 mg ENT or placebo three times per day for 12 days.

Results: OPC at steady-state significantly increased the levodopa-AUC up to 78.9% and 73.74% with 75 mg OPC in comparison to placebo and ENT, respectively. Levodopa-AUC₀₋₂₄ was higher when levodopa/carbidopa was administered with any OPC dose than when administered concomitantly with ENT. A marked increase (>30%) in peak of exposure (C_{max}) to levodopa occurred with 75 mg OPC following levodopa/carbidopa administrations. OPC presented a significantly long-lasting and sustained S-COMT inhibition. Maximum S-COMT inhibition ranged from 67.1% (200 mg ENT) to 94.2% (75 mg OPC) and was higher than ENT for all OPC doses. The 50 and 75 mg OPC were somehow similar

(75 mg was slightly superior), thus, the 75 mg OPC may not bring a significant advantage over 50 mg OPC with regard to S-COMT inhibition. The tolerability profile of OPC was favourable.

Conclusion: OPC may offer a therapeutic advantage in relation to ENT as levodopa sparing agent in patients with Parkinson's disease receiving levodopa therapy. The dosages of 25 and 50 mg OPC likely provide the most adequate enhancement in levodopa availability as adjunct to levodopa/carbidopa therapy.

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Abstract – WCN 2013

No: 1034

Topic: 2 – Movement Disorders

Effect of opicapone multiple-dose regimens on levodopa pharmacokinetics, motor response, and erythrocyte-COMT activity in Parkinson's patients co-administered with levodopa/dopa-decarboxylase inhibitor

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Background: Opicapone (OPC) was developed to fulfil the need for more potent, safer and longer acting COMT inhibitors.

Objectives: Investigate the effect of once-daily 5, 15 and 30 mg OPC, a new COMT inhibitor, on the levodopa pharmacokinetics, in Parkinson's Disease (PD) patients with motor fluctuations treated with standard-release 100/25 mg levodopa/carbidopa (LC) or levodopa/benserazide (LB).

Methods: This was a multicentre, double-blind, randomised, placebo-controlled study in four parallel groups of PD patients treated with LC or LB and with motor fluctuations ("wearing-off" phenomenon). Subjects were sequentially and randomly assigned to be administered, once-daily, during the 21 to 28 day maintenance phase with placebo or 5, 15 and 30 mg OPC. Subjects performed two levodopa tests, one on the morning of the day after admission and another following the maintenance phase. Subjects also kept a diary to record ON/OFF periods.

Results: In relation to placebo, levodopa exposure increased 24.73%, 53.93% and 65.61% following 5, 15 or 30 mg OPC. Maximum S-COMT inhibition (E_{max}) ranged from 52% (5 mg OPC) to 80% (30 mg OPC). The study was not designed to detect any significant differences in motor performance, but the exploratory analysis performed shows improvement in various motor outcomes, including a dose dependent change in absolute OFF time corresponding to a percentage decrease of 0.77%, 4.16%, 29.55% and 32.71% with placebo, 5 mg, 15 mg and 30 mg OPC, respectively.

Conclusion: OPC is a promising new COMT inhibitor and deserves further clinical evaluation in larger samples of patients with PD on levodopa treatment with motor fluctuations.

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Abstract – WCN 2013

No: 1038

Topic: 2 – Movement Disorders

Efficacy and safety of opicapone, a new COMT-inhibitor, for the treatment of motor fluctuations in Parkinson's Disease patients: BIPARK-II study

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Background: Opicapone (OPC) was developed to fulfil the need for more potent, safer and longer acting COMT inhibitors.

Objectives: The objective of this study was to investigate the efficacy and safety of 25 and 50 mg-OPC administered once-daily, compared with placebo, in patients with Parkinson's Disease (PD) on levodopa treatment and with end-of-dose motor fluctuations.

Methods: Pivotal phase-III, multinational, multicentre, double-blind (DB), placebo-controlled and parallel-group study. Subjects were randomized to placebo (n = 135), 25 mg-OPC (n = 125) or 50 mg-OPC (n = 147). The DB phase lasted 14–15 weeks. Primary efficacy variable was the change from baseline in absolute OFF-time, based on patient diaries. Secondary endpoints include proportion of responders, course of OFF/ON-time, UPDRS-III, quality-of-life (PDQ-39), NMSS, PDSS, tolerability and safety (including mMIDI, C-SSRS and clinical laboratory tests) assessments.

Results: Mean reduction in absolute OFF-time in both 25 and 50 mg-OPC groups was considerably greater than in placebo (1.7, 2.0 and 1.1 h, respectively). Despite the high placebo response, 50 mg-OPC was significantly better than placebo (p = 0.0084), but 25 mg-OPC missed statistical significance by only 0.1 h. Significantly more patients receiving either 25 or 50 mg-OPC achieved the OFF-time responder endpoint than with placebo (62.4%-[p = 0.0405], 66.0%-[p = 0.0088] and 50.4%, respectively), and also achieved greater OFF-time reductions (11.0%-[p = 0.0297], 12.1%-[p = 0.0044] and 6.7%, respectively). Furthermore, mean increase in absolute ON-time without or with non-troublesome dyskinesias was considerably greater in both 25 and 50 mg-OPC groups than in placebo (1.4, 1.43 and 0.8 h, respectively). Opicapone was safe and well tolerated.

Conclusion: OPC once-daily was safe, well tolerated, and effective in reducing the OFF-time in patients with PD on levodopa treatment and with motor fluctuations.

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Abstract – WCN 2013

No: 914

Topic: 2 – Movement Disorders

Camptocormia in Parkinson's disease: A multicenter study in Japan

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Background: Camptocormia (CC) is an abnormal flexion of the thoracolumbar spine during standing and walking that abates in the

recumbent position, and may be one of the most disabling symptoms in Parkinson's disease (PD).

Objective: The objective of our study is to describe the clinical features of PD patients with CC, comparing with those without CC.

Patients and methods: Seventy-eight PD patients with CC underwent a multidisciplinary clinical examination (neurological, neuropsychological), blood test, and spine MR scan were compared with age/sex-matched 78 PD patients without CC.

Results: Patients with CC showed significantly higher frequencies of spinal operation and compression fracture ($P < 0.05$). CC was found in patients with more severe PD, as clinically assessed by the Hoehn and Yahr staging, the motor Unified Parkinson's disease Rating Scale part III, and presence of dementia. The serum creatine kinase levels of PD patients with CC were statistically higher than those of without CC ($P < 0.05$). We found more abnormalities of MR findings in spine and paravertebral muscles in PD patients with CC than those without CC ($P < 0.05$).

Conclusion: CC is associated with the history of spinal operation, compression fracture, and disease severity. The MRI findings and difference of CK levels between the groups suggest that CC in PD is attributed to a focal myopathy in spine and paravertebral muscles.

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Abstract – WCN 2013

No: 1349

Topic: 2 – Movement Disorders

Parkinson's disease and autonomic symptoms

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Introduction: There is a wide range of autonomic symptoms (AS) in Parkinson's disease (PD) but their occurrences are still a subject to discrepancies in the literature.

Objectives: The objectives of this study are to evaluate the occurrence of AS in a cohort of patients with PD and control subjects, to assess the relations with demographic, disease-related, and clinical variables and to qualify the impact of this trouble on the quality of life.

Methods: A cohort of 102 patients with PD was evaluated for the occurrence of AS, motor and non-motor symptoms, as well as for demographic and disease-related characteristics. The study was conducted in the Department of Neurology of Sahloul University Hospital between June 2011 and November 2011. 95 control subjects were recruited from the consultations and other departments. We used the following scales: SCOPA AUT, SF12, SCOPA Sleep and Beck Scale.

Results: All PD patients reported more problems in all in autonomic domains compared to control subjects with the largest differences in the gastro-intestinal tract. The prevalence of autonomic disorders was correlated with older age, great disease severity and high doses of dopaminergic medication. The severity of the disorder was associated with depressive symptoms, cognitive disorders, and sleep disorders with $p < 0.05$.

Discussion: Autonomic symptoms were assessed using a reliable and valid instrument. Our results show that, compared to a control group, Parkinson's disease expresses more gastro-intestinal, urinary, cardiovascular, thermoregulation, and sexual troubles; indicating that the entire autonomic system is affected by this disease, which is consistent with findings of the literature.

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Abstract – WCN 2013

No: 1337

Topic: 2 – Movement Disorders

Wilson's disease and psychiatric disorders: Is there comorbidity or indicator of relapse?

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Introduction: Wilson's disease is an infrequent pathology resulting from a change of the ATP7B gene on the long arm of chromosome 13. This involves a reduction of the transport of copper in the bile and its accumulation in the body, notably the brain.

Objective: The objective of this study is to illustrate the significance of psychiatric disorders in WD.

Case report: A 25-year old woman with a personal history of Wilson's disease (WD) presented acute psychotic manifestations. The psychiatric interview finds a state of psychomotor agitation, aggressiveness and depressed mood concept with suicidal intentions. Moreover, she has a bilateral extra pyramidal syndrome. The blood levels of copper and ceruloplasmin are collapsed. The patient confirmed that she had stopped her treatment with D-penicillamine for two months ago.

Discussion: WD begins in the form of a hepatic, neurological, or psychiatric disease in at least 90% of the patients. In some rare cases, the first manifestations of the disease can be psychiatric which accounts for only 10%. The disease can include later isolated behavioral problems, a schizophrenic syndrome, or a manic-depressive syndrome. The main difficulty was to establish whether psychiatric disorders are included in the WD array or not. Indeed neuropsychiatric co morbidity in the context of WD is significant. Poor adherence to treatment, cupric balance sheet and the improvement after the reintroduction of D-penicillamine confirm a psychiatric relapse of WD.

Conclusion: In WD, psychiatric manifestations can precede, complicate the somatic disorders or be independent. An early diagnosis is necessary to provide a specific treatment.

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Abstract – WCN 2013

No: 269

Topic: 2 – Movement Disorders

Cervical dystonia patient registry for observation of onabotulinum toxin A efficacy (CD Probe): Pain at baseline

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Background: Cervical dystonia (CD) is the most common focal movement disorder and is often associated with pain.

Objective: The objective of this study is to report the presence of pain associated with CD upon enrollment in subjects participating in a large registry.

Methods: CD PROBE is a prospective, observational, multicenter, registry designed to assess safety, effectiveness, and treatment utilization within clinical practice. CD subjects were naïve to botulinum toxin, new to the physician's practice, or had ≥ 16 weeks since last treatment if in a previous clinical trial. Factors delineated by the presence (score > 0) or absence (score = 0) of pain at baseline, as measured by the Pain Numeric Rating Scale (PNRS), were baseline demographics, Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS), and CD Impact Profile-58 (CDIP-58).

Results: CD PROBE enrolled 1046 subjects (74% female), with a mean age 58.0 (± 14.7 y), between 1/9/09 and 8/31/12. 88.9% ($n = 922$) of subjects reported pain at baseline (PNRS > 0). Compared to those

without pain at baseline, subjects reporting pain had significantly younger symptom onset (48.4 ± 16.5 y vs 53.9 ± 17.9 y; $p = 0.0026$), and significantly higher scores for TWSTRS total and subscales ($p \leq 0.0002$) and CDIP-58 subscales ($p \leq 0.0008$).

Conclusion: CD-PROBE is the largest registry ever conducted for subjects with CD and will provide real world experience of treatment response of repeat injection of onabotulinum toxin A. Pain is present in the majority of subjects with CD and is associated with increased severity across a range of measures.

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Abstract – WCN 2013

No: 1281

Topic: 2 – Movement Disorders

The sequence effect is not a defining characteristic of Parkinson's disease

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Background: The sequence effect (SE) describes progressive decrements of amplitude and speed with repetitive movements and is considered characteristic of Parkinson's disease (PD). It is associated with functional disability and freezing of gait but the pathophysiology remains unclear.

Objective: The objective is to objectively assess finger tapping (FT) in order to evaluate

- (i) what proportion of PD patients and age-matched healthy controls (HC) exhibit the SE and
- (ii) whether the MDS-UPDRS standard of ten taps is enough for the SE to manifest.

Patients and methods: Sixty-two PD patients and 47 age-matched HC from centres in the UK and USA performed FT whilst wearing electromagnetic sensors. Movement data from the first ten taps was used to calculate amplitude, speed, rhythm, halts and decrementing speed and amplitude (SE). Longer FT sequences were also examined. **Results:** After 10 taps all of the movement components differed significantly between the PD and HC groups (amplitude, speed and rhythm $p < 0.001$; halts $p = 0.03$) except for the SE ($p > 0.1$). Decrementing amplitude occurred in 61% and 59% and decrementing speed in 52% and 49% ($p = 0.9$), of PD and HC data respectively. An additional 23% of PD and 27% of HC ($p = 0.7$) exhibited the SE when FT was performed for 30 s (mean 79 taps).

Conclusions: The SE is not specific for PD. A similar proportion of PD and HC subjects exhibited the SE. For some subjects 10 taps is not long enough for the SE to manifest.

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Abstract – WCN 2013

No: 1308

Topic: 2 – Movement Disorders

Clinical hallmarks and epidemiology of stiff person syndrome in Tanzania

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Background: Stiff person syndrome (SPS) is an extremely rare anti-GAD antibody related disorder, with prevalence estimates ranging from 0.5 to 2/1,000,000. Its diagnosis is clinical, especially in most of Africa where anti-GAD titres are unavailable.

Objective: The objective of this study is to clinically delineate a series of stiff person syndrome patients, and to compare the prevalence in Tanzania to that in scientific publications.

Patients and methods: Patients were ascertained through neurological consultation in Kilimanjaro Christian Medical Centre during the period 2008–2013. Clinical diagnosis was established by means of neurological examination and exclusion of other amenable causes. Anti-GAD antibody testing or EMG is presently unavailable in Tanzania.

Results: A total of nine SPS cases are reported (three females and six males, aged 13–64 years). The estimated prevalence in this patient population was 0.5–6/1,000,000, depending on the estimated catchment area. One patient was referred abroad for intravenous immunoglobulin therapy, with marked clinical improvement, whereas the others showed some response to baclofen and benzodiazepines. Two patients presented with femoral fractures secondary to violent muscle spasms.

Conclusions: Stiff person syndrome is a rare auto-immune disorder, but is shown to occur in Tanzania. A literature search yielded comparable studies only from other parts of the world: we present the first SPS prevalence rates in Africa. Even though the sample is small and estimated prevalence form a wide range, this is comparable to estimates from other ethnic groups. Yet underestimation due to underreporting by patients as well as doctors is not unlikely.

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Abstract – WCN 2013

No: 1310

Topic: 2 – Movement Disorders

Clinically 'slight' bradykinesia is accurately detected using a novel device that requires a one-minute test period

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Background: Bradykinesia is the fundamental clinical feature of Parkinson's disease (PD) but when it is very mild it is difficult to detect. Dopamine transporter imaging is a useful diagnostic tool but it is expensive, not specific for PD, time-consuming and exposes the patient to ionizing radiation. A simple cheap non-invasive test that provides rapid objective measurement of bradykinesia could aid accurate early diagnosis of PD.

Objective: The objective of this study is to assess whether an objective measurement of 'slight' (MDS-UPDRS grade one) bradykinesia can be obtained from a novel device employing electromagnetic (EM) tracking sensors and evolutionary algorithm analysis.

Methods: Forty-nine PD patients and 41 age-matched healthy controls (HC) performed finger tapping (FT) for 30 s with each hand separately whilst wearing EM tracking sensors on the index finger and thumb. The kinematic data sets were analysed by custom-written computer evolutionary algorithms and compared to MDS-UPDRS clinical grade

and diagnostic group using receiver operator characteristic (ROC) curves. The composite algorithm was validated on an independent sample of FT data collected from 13 PD patients.

Results: PD FT assessments with grade one MDS-UPDRS were accurately distinguished from controls dominant hand FT with area under ROC curve of 0.952 ($p < 0.0001$). The algorithm correctly classified 14/15 of the PD FT data sets in the validation sample that had clinically slight bradykinesia.

Conclusions: This technology is able to provide an objective measurement of the most subtle clinical grade of bradykinesia. Potential applications include supplementing clinical diagnoses, screening and objective assessment of response to treatment.

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Abstract – WCN 2013

Topic: 2 – Movement Disorders

No: 1320

Choreoathetosis caused by glycemic decompensation in pancreatic neoplasm

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Background: Choreoathetosis is a neurologic disorder, related to a basal ganglia lesion, where two types of involuntary movements coexist. Chorea consists of continual, rapid, brief, irregular movements that flow randomly from one body part to another. Athetosis is a slow form of chorea, with writhing movements that mainly affect to distal regions of the limbs.

Objectives: We present a 72-year old woman with bilateral choreiform movements and diabetic ketoacidosis.

Patients and methods: Our patient had a longstanding diabetes type II with poor metabolic control, worse during the last month. She was hospitalized to control her blood glucose levels.

Results: Symmetric bilateral hyperintense putamen nuclei were found at MRI, related to maintained hyperglycemia. A hypertransaminasemia led us to complete the abdominal examination, and a body TC and a biopsy confirmed the diagnosis of pancreatic adenocarcinoma. Choreoathetosis was treated with clonazepam and pimozide, with no efficacy; then we tried haloperidol and tetrabenazine, improving the symptomatology.

Conclusion: Choreoathetosis is caused by very diverse etiology disorders. Long-evolution nonketotic hyperglycemia has been described to produce involuntary movements as choreoathetosis. Typically MRI shows hyperintense putamen on T1-weighted images, contralateral to the side of chorea, or bilateral if it is generalized.

This rare complication of DM type II is generally reversible with an adequate metabolic control. But if the movements are severe and disabling, it can be successfully treated with drugs that interfere with central dopaminergic function, such as the dopamine receptor-blocking drugs (neuroleptics), reserpine, and tetrabenazine. Other effective treatments can be benzodiazepines or amantadine.

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Abstract – WCN 2013

No: 181

Topic: 2 – Movement Disorders

A phase 2, placebo-controlled, randomized, double-blind trial of tozadenant (Syn-115) in patients with Parkinson's disease with wearing-off fluctuations on levodopa

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Background: Tozadenant is an oral, selective, adenosine 2-alpha receptor antagonist.

Objective: The objective of this study is to evaluate the safety and efficacy of tozadenant as an adjunct to levodopa in PD patients with wearing-off fluctuations.

Methods: In this international, 12-week, double-blind, phase 2 trial patients on stable doses of levodopa with ≥ 2.5 h of OFF time/d were randomized to tozadenant 60, 120, 180 or 240 mg BID or placebo. Primary endpoint: change from baseline to Week 12 in h/d spent in the OFF state. An MMRM ANCOVA was used for analyses with a prespecified hierarchical step-down approach to test multiple dose groups.

Results: 337 of 420 randomized pt completed treatment (mITT population): mean age, 63.3 yr; PD duration, 8.7 yr; baseline OFF time, ~6 h. Mean placebo-corrected change from baseline in OFF time was significantly reduced with tozadenant 120 mg BID (-1.1 h, $p = 0.0039$) and 180 mg BID (-1.2 h, $p = 0.0039$). ON time with troublesome dyskinesia did not increase significantly in any tozadenant group. Mean placebo-corrected scores of the following significantly improved: UPDRS III with tozadenant 120 mg BID (-2.2 , $p = 0.0325$) and 180 mg BID (-2.5 , $p = 0.0325$); UPDRS I-III in all tozadenant groups (all groups, $p \leq 0.03$); CGI-I/CGI-S scores; and PGI-I in the 120 mg BID tozadenant group. Most common AEs in the combined tozadenant groups: dyskinesia, nausea, dizziness, constipation, PD worsening, insomnia, falls.

Conclusion: Tozadenant, 120 or 180 mg BID, was generally well tolerated and efficacious in reducing OFF time and improving motor signs without significantly increasing troublesome dyskinesia.

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Abstract – WCN 2013

No: 1150

Topic: 2 – Movement Disorders

Antipsychotic efficacy and motor tolerability in a phase III placebo-controlled study of pimavanserin in patients with Parkinson's Disease psychosis (Acp-103-020)

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PD psychosis (PDP) is frequent, distressing and a leading cause of institutionalization. It complicates PD management and is linked to increased morbidity, incident dementia and mortality. Current antipsychotics lack efficacy and/or have serious tolerability and safety concerns. Pimavanserin, a selective 5-HT_{2A} antagonist, has shown antipsychotic effects and good tolerability in previous PhIII trials, but a robust placebo effect precluded statistical separation. A PhIII outpatient study, optimized to reduce placebo response, was conducted to assess the efficacy and safety of pimavanserin in PDP. Following screening, in which brief (non-pharmacologic) psychosocial therapy (BPST-PD) was offered, 199 non-demented patients with moderate-severe psychosis (on stable PD medication) were randomized to oral q.d. doses of 40 mg pimavanserin or placebo for 6 weeks. Pimavanserin met the primary endpoint using SAPS-PD (PD-adapted

version of the Scale for Assessment of Positive Symptoms, assessed by independent raters): -5.79 PIM vs -2.73 PBO (Baseline to Day 43 LSM $\Delta = -3.06$; $p = 0.001$). These results were supported by highly significant improvement in the secondary efficacy measure, CGI-I (LSM $\Delta = -0.67$; $p = 0.001$), assessed by Investigators blind to the SAPS-PD. Clinical benefits were also seen in all exploratory measures with significant improvements in nighttime sleep, daytime wakefulness, and caregiver burden. Consistent with previous studies, pimavanserin met the key secondary endpoint for noninferiority to placebo on motor function (UPDRSII + III) and was otherwise safe and well tolerated. Most common AEs were UTI (11.7%-PBO, 13.5%-PIM) and falls (8.5%-PBO, 10.6%-PIM). SAEs in >1 patient were UTI (1-PBO, 3-PIM) and psychotic disorder (0-PBO, 2-PIM). Pimavanserin appears effective, safe and well-tolerated for PDP.

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Abstract – WCN 2013

No: 1066

Topic: 2 – Movement Disorders

Incobotulinum toxin A demonstrated therapeutic equivalence to onabotulinum toxin A in the treatment of blepharospasm and cervical dystonia

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Introduction: Equivalent efficacy of incobotulinum toxin A (Xeomin®) to onabotulinum toxin A (Botox®) using a 1:1 unit dosing ratio was demonstrated in two multicentre, double-blind, randomised studies in blepharospasm and cervical dystonia, respectively.

Patients and methods: Both study protocols pre-specified the two-sided 95% confidence interval (CI) method for testing therapeutic equivalence of incobotulinum toxin A to onabotulinum toxin A for each study's primary efficacy outcome (blepharospasm: change in Jankovic Rating Scale [JRS] sumscore from baseline to 3 weeks post-injection; cervical dystonia: change in Toronto Western Spasmodic Torticollis Rating Scale [TWSTRS]–Total score from baseline to 4 weeks post-injection). Differences between treatment groups were determined by least square (LS) means and corresponding two-sided 95% CIs in the per-protocol population. Clinically irrelevant difference was defined as $\Delta = 0.8$ points in the blepharospasm study and $\Delta = 1.3$ points in the cervical dystonia study, predefining the therapeutic equivalence ranges as $[-0.8, 0.8]$ and $[-1.3, 1.3]$, respectively.

Results: In the blepharospasm study (per-protocol population $n = 256$), incobotulinum toxin A and onabotulinum toxin A both significantly ($p < 0.0001$) improved JRS sumscores 3 weeks post-injection (incobotulinum toxin A: -2.90 ; onabotulinum toxin A: -2.67). The LS mean difference was -0.23 (favouring incobotulinum toxin A), the 95% CI $[-0.68, 0.22]$ was within the predefined equivalence range $[-0.8, 0.8]$. In the cervical dystonia study (per-protocol population $n = 420$), both treatments significantly ($p < 0.0001$) improved TWSTRS–Total score 4 weeks post-injection (incobotulinum toxin A: -6.6 ; onabotulinum toxin A: -6.4). The LS mean difference was -0.33 (favouring incobotulinum toxin A); the 95% CI $[-1.05, 0.38]$ was within the predefined equivalence range $[-1.3, 1.3]$.

Conclusion: Incobotulinum toxin A and onabotulinum toxin A were therapeutically equivalent in treating blepharospasm and cervical dystonia at a 1:1 unit dosing ratio in the administered dose ranges.

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Abstract – WCN 2013

No: 1070

Topic: 2 – Movement Disorders

Long-term treatment of blepharospasm and cervical dystonia: Incobotulinum toxin A is well tolerated when injected at flexible intervals based on patient needs

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Introduction: In clinical practice, botulinum-toxin injections for blepharospasm or cervical dystonia (CD) are administered at ~3-month intervals; there is concern that shorter intervals might increase the risk of adverse events (AEs) and neutralizing antibodies. This post-hoc analysis investigated flexible incobotulinum toxin A (Xeomin®) injection intervals (6–20 weeks) in the long-term management of blepharospasm and CD.

Patients and methods: In the blepharospasm study, a randomized, double-blind, placebo-controlled Phase III study with an open-label extension period, patients could receive ≤ 6 incobotulinum toxin A treatments (flexible doses ≤ 50 U/eye). Patients in the CD study, a randomized, double-blind, placebo-controlled Phase III study with a randomized, double-blind extension period, could receive ≤ 6 incobotulinum toxin A treatments (fixed doses, 120 or 240 U). In both studies, injection intervals were flexible (≥ 6 weeks) and determined as clinically indicated. Standard safety assessments were performed throughout.

Results: In blepharospasm, 461 incobotulinum toxin A treatments were analyzed: 207 (44.9%) treatments were given at 6–11-week intervals and 254 (55.1%) at 12–20-week intervals. The most frequent AEs were eyelid ptosis and dry eyes. In CD, 821 treatments were evaluated: 369 (44.9%) were administered at 6–11-week intervals and 452 (55.1%) at 12–20-week intervals. The most frequent AEs were dysphagia and neck pain. In both studies, AE frequency, overall and for each treatment cycle, was similar for injection intervals < 12 weeks and ≥ 12 weeks, with no cumulative effects following repeated treatment.

Conclusion: Incobotulinum toxin A, administered with flexible intervals according to patients' clinical needs, is an effective and well tolerated long-term treatment for blepharospasm and CD.

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Abstract – WCN 2013

No: 1097

Topic: 2 – Movement Disorders

Satisfaction with botulinum toxin treatment in post-stroke spasticity: Results from two cross-sectional surveys of patients and physicians

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Introduction: There is little information available regarding patient's and physician's satisfaction with botulinum toxin treatment for post-stroke spasticity.

Patients and methods: Structured patient and physician surveys were conducted in Canada and the USA. Patients had post-stroke spasticity and had received ≥ 2 treatments with onabotulinum toxin A or abobotulinum toxin A; information on current and prior

botulinum toxin treatment cycles and treatment satisfaction was collected. Physicians included were experienced in treating post-stroke spasticity, with ≥ 3 -year history of injecting botulinum toxin for medical purposes; information regarding satisfaction with botulinum toxin treatment was collected.

Results: Most of $N = 76$ patients with post-stroke spasticity were very (42%) or somewhat (46%) satisfied with their current treatment. Satisfaction was highest at the time of peak effect and lowest just before re-injection. While 78% of patients stated they would prefer injection intervals ≤ 12 weeks, only 45% received such intervals. Intervals ≤ 10 weeks were preferred by 43% of patients but received by just 6.3%. Most of $N = 105$ physicians were moderately (58%) or very satisfied (37%) with botulinum toxins. Physicians felt that on average 16% of patients would benefit from shorter injection intervals and 25% would benefit from higher maximum doses than those currently permitted.

Conclusion: Patient satisfaction with botulinum toxin therapy for post-stroke spasticity varies over the treatment cycle and is lowest just before re-injection; many patients would prefer injection intervals shorter than the standard 12 weeks. While most physicians are satisfied with botulinum toxin therapy for post-stroke spasticity, many feel that some patients would benefit from shorter injection intervals and higher doses.

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Abstract – WCN 2013

No: 1278

Topic: 2 – Movement Disorders

Impaired activation of somatosensory cortex as fMRI correlate of reduced dexterity in PD

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Background: Previously (1ab), the DOPA-resistant component of decreased dexterity in PD was called limb-kinetic apraxia (LkA). Our recent fMRI pilot study (2) investigating PD patients OFF medication suggested impaired activation of primary somatosensory cortex (SSC) as closely related to LkA. Both in the behavioral (1ab) and fMRI studies (2), coin rotation (CR, LkA task, target condition) was contrasted with simple finger tapping (FT, bradykinesia task, reference condition). fMRI signal changes were hence corrected for basic motor skills and represented activation particularly associated with dexterity.

Based on the DOPA resistance of CR performance in (1ab), we hypothesized a reproducibility of impaired activation of the SSC ON medication.

Objective: The objective of this study is to confirm dysfunction of SSC as closely related to DOPA-resistant dexterity deficits in PD.

Patients and methods: According to the experimental setup (CR vs. FT) mentioned above, 10 PD patients performed the fMRI experiment in the ON as well as the OFF condition. 14 healthy controls performed the same experiment. Patients were compared to controls (SPM8).

Results: Confirming our pilot study, we assessed decreased activation of the SSC in patients OFF medication compared to controls. In line with our study hypothesis, this finding was reproducible in patients ON medication compared to controls ($p < 0.01$, uncorr., $k = 25$ vox.). Comparing fMRI signals in the SSC between ON and OFF conditions, no differences could be assessed (lowering even to $p < 0.05$).

Conclusions: Dexterity deficits in PD are linked to a dysfunction of the SSC.

(1a) Gebhardt-et-al_MovDisord 2008;23 :1701-6.

(1b) Quencer-et-al_Neurology 2007;68:150-1.

(2) Foki-et-al_ExpNeurol 2010;225:416-22.

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Abstract – WCN 2013

No: 1269

Topic: 2 – Movement Disorders

Subjective and objective alcohol responsiveness in different tremor disorders

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Background: A few studies exist on the effect of alcohol in essential tremor (ET) and Parkinson's disease (IPD), but so far no systematic studies have been performed in other tremor syndromes. Nevertheless, a positive response to alcohol – commonly simply inquired by history – is used as supportive diagnostic criterion for ET.

Objective: The objective of this study is to investigate if subjective/objective alcohol responses distinguish a broad range of tremor disorders.

Methods:

- (1) We assessed drinking habits and response of tremor to alcohol intake in 100 consecutive patients with arm tremor.
- (2) For a double-blind placebo-controlled oral alcohol test we recruited 12 patients with ET, 10 IPD, and 7 dystonic tremor (DT).

The effect to alcohol/placebo was assessed at baseline and after 30/60/120 min by clinical rating scales and accelerometry.

Results:

- (1) We investigated 44 patients with IPD, 22 ET, 20 DT, and 14 others. Of 77 patients drinking alcohol, 23 had no response, 26 had improvement, and 28 never paid attention. None of the evaluated parameters differed between the groups.
- (2) ET, IPD, and DT had a significant tremor improvement with placebo and alcohol. There were no significant between group differences. However, in the ET group there was a stronger objective effect of alcohol compared to placebo.

Conclusions: Subjective alcohol response inquired by history may not be suitable as supportive diagnostic criterion for ET. Objectively, all investigated patients improved with alcohol. This effect seems to be truly alcohol related in ET only.

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Abstract – WCN 2013

No: 1258

Topic: 2 – Movement Disorders

Exome sequencing analysis in familial progressive supranuclear palsy

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Background: Progressive supranuclear palsy (PSP) is a clinicopathological syndrome characterized by akinesia, supranuclear gaze palsy,

rigidity, axial dystonia, gait disturbance, frontolimbic dementia and other clinical deficits related to neuronal loss, gliosis, and presence of neurofibrillary tangles and neuropil threads in different brain areas, mainly in the basal ganglia, diencephalon, brainstem, and frontal and temporal lobes. Although PSP is mostly considered a non-familial or sporadic tauopathy, rare multiplex kindreds have been described. A genetic basis for familial clustering has been demonstrated in a Spanish family with PSP linked to chromosome 1, for which the causal gene is still unknown, and in families with causal mutations in the microtubule-associated protein tau gene (*MAPT*).

Objective: To identify the causal gene linked to 1q31.1.

Patients and methods: We have studied the Spanish family for which the genetic locus was mapped to a 3.4 cM region at 1q31.1. This family presents a pathology-proved PSP with a pattern of transmission compatible with an autosomal dominant inheritance. We have sequenced the whole-exome from one patient of this family, and DNA from 30 additional family members is available for segregation analysis. We have also checked the exome sequences of a cohort of about 50 PSP patients.

Results and conclusion: The exome analysis of the Spanish family is presently on-going. For the PSP cohort no variants were detected in the linkage region of chromosome 1. Therefore, this locus should not constitute a common cause of PSP.

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Abstract – WCN 2013

No: 1256

Topic: 2 – Movement Disorders

Tremor associated with Klinefelter syndrome – A case series and review of the literature

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Background: Previous case series suggested a link between Klinefelter syndrome (KS) and essential tremor (ET) or an ET-like syndrome.

Methods: We investigated three KS-patients with tremor including tremor-analyses and discuss our data in context to findings from a literature review. The clinical outcome after deep brain stimulation (DBS) is also reviewed.

Results: Tremor in KS is predominantly a postural and kinetic tremor that strongly resembles ET. However, KS patients with tremor differ from ET in several findings such as a less frequent family history, lack of alcohol responsiveness, and missing benefit to drugs used in ET. One of our patients and two cases from literature improved after DBS of the ventral intermediate nucleus (VIM) of the thalamus.

Conclusions: Tremor in KS seems to differ from ET. While anti-tremulous drugs exert no or little effect, first observations suggest that VIM-DBS may be efficacious.

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Abstract – WCN 2013

No: 1237

Topic: 2 – Movement Disorders

Assessment of thyroid function in spinocerebellar Ataxia type 2 Cuban patients

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Background: Dysfunction of thyroid axis has been linked to neurodegenerative conditions. Spinocerebellar Ataxia type 2 (SCA2) is a polyglutamine disorder caused by abnormal expansion of a CAG repeat sequence on ATXN2 gene.

Objective: To assess the association between thyroid function and SCA2.

Method: A case-control study was carried out with 41 SCA2 affected individuals and 41 healthy controls matched by age and sex. Serum levels of TSH, T3 and T4 were determined by radioimmunoassay. CAG repeat number on ATXN2 gene was assessed by PCR and gel electrophoresis. STRING algorithm was used to predict protein network interactions.

Results: T3 and T4 serum levels were significantly increased in affected individuals related to controls ($p < 0.05$) and this is dependent on sex. No significant associations were found between thyroid hormones and body mass index, abdominal circumference, resting metabolic rate, CAG repeat number, age at onset, disease duration or SARA score ($p > 0.05$). However, TSH serum levels significantly correlated with disease progression rate ($r = -0.32$; $p = 0.045$). Evidences were obtained for interactions between ATXN2 and thyroid hormone networks, although the relative weights for such interactions were low.

Conclusion: Spinocerebellar Ataxia type 2 is associated with sex-specific hyperactivity of thyrotropic axis; however, more work has to be done to clarify the clinical consequences of such an association.

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Abstract – WCN 2013

No: 1221

Topic: 2 – Movement Disorders

Anti-TNF-alpha inhibitors therapy and non infectious neurologic adverse effects

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Background: Infections are the most common adverse effects associated with TNF-alpha-inhibitors (TNF-alfa-I) therapy. Non-infectious neurologic complications are infrequent (several events suggestive of demyelination have been reported).

Objective: Present four cases of neurologic disorders (central and peripheral demyelination, pseudotumoral lesion, parkinsonism (2)) in four patients treated con distinct TNF-alfa-I.

Patients and methods:

Case-1: Patient (F-63-y) with rheumatoid arthritis (RA), treated with infliximab; she developed a clinical picture of brachial plexitis and, nine months later, myelitis and lumbar plexitis.

Case-2: Patient (M-58-y) with RA, treated with adalimumab; he consulted because of headache, weakness and numbness in his right body side.

Case-3: Patient (M-63-y) with psoriasis, treated with ustekinumab; he presented a left parkinsonism (PD).

Case-4: Patient (F-57-y) with RA, treated with adalimumab; she presented a right PD.

Results:

Case-1: Cerebral-MRI study was normal; spinal-MR-T2-WI study showed some hyperintense lesions in dorsal spinal cord. CSF study showed lymphoid pleocytosis. EMG study revealed denervation in the right deltoid and brachial biceps and in the left quadriceps. The evolution was favorable after the drug was discontinued.

Case-2: Cerebral-MRI study showed left frontal lesion with mass effect and Gadolinium enhancement. Histopathologic study revealed necrosis and mononuclear cells infiltration. The drug was discontinued and the evolution was favorable.

Cases-3-4: Cerebral-MRI study was normal in both patients. DAT-scan resulted abnormal in both patients. The biologic therapy was

discontinued. The PD progresses (like common PD) and responds to levodopa.

Conclusions: TNF- α -I therapies are related with distinct types of central and peripheral demyelinating lesions; less clear seems their risk of induction or acceleration of PD.

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Abstract – WCN 2013

No: 1209

Topic: 2 – Movement Disorders

Exercise training – Effects of MOTomed® exercise on typical motor dysfunction in Parkinson's disease

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Objective: Additionally to regular medication, alternative drug-free treatments, such as exercise or physical therapy, do not cause any side effects and play an important role in Parkinson's treatment. Recent scientific findings suggest very positive effects of movement training at high cadence, the so-called "Forced Exercise" (FE). The present paper investigates the effects of a home-based passive FE cycling training on general motor function and quality of life in Parkinson's patients.

Patients & methods: 44 Parkinson's patients (68.5 ± 7.8 years) were randomized to control group ($n = 23$; age: 71.3 ± 4.0 years) and intervention group ($n = 21$; age: 67.5 ± 7.8 years). The intervention group completed a 10 week FE cycling program with a motor-assisted movement therapy device. The subjects were encouraged to perform a daily 40 minute MOTomed® training session, at up to 90 revolutions per minute, in addition to their regular therapy (medication and physical therapy). Motor function and quality of life measures were assessed three times during the study period, a total of 25 test items were recorded (TMTBattery = 15 items, tremor spiral test = 2 items, PDQ-8 = 8 items). Subjects of the control group continued their standard therapy.

Results: The results of the study show significant improvements in walking ability (walking time: $F = 13.31$; $p = .000$; $p.Eta2 = .241$; walking steps: $F = 6.44$; $p = .000$; $p.Eta2 = .133$) and hand coordination (diadochokinesia of the right arm: $F = 3.76$; $p = .03$; $p.Eta2 = .082$).

Conclusion: Device-supported FE movement training of the lower extremities leads to improvements in walking ability and hand motor function, which suggest that FE may be affecting central motor control processes. To proof these findings the authors recommend further studies.

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Abstract – WCN 2013

No: 1232

Topic: 2 – Movement Disorders

Clinical cerebral dysfunction sclerosis in Africa

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Objective: To report on the occurrence of clinical cerebral dysfunction sclerosis.

Methods/subjects: All the patients referred to the clinic by neurologists and other specialists for electrophysiological tests with diverse neurological complaints. The patients were examined and diagnosed as having sclerosis on clinical grounds and established criteria are reported.

Results: Out of 2831 patients referred for electrophysiological tests over a ten year period, nine patients were diagnosed as having definite sclerosis on clinical grounds. Four of these had supporting laboratory findings (MRI scans, CSF studies and visual evoked responses). The presenting symptoms were predominantly visual disturbances and somatic sensorimotor disturbances which were seen in all the patients. Cerebellar dysfunction was observed less frequently, in less than half of the patients while sphincter disturbances were conspicuously rare. The sex distribution was overwhelmingly in favour of the female at a ratio of 7:2.

Conclusions: Cerebellar dysfunction sclerosis occurs most in African countries and may not be as rare as previously suggested and its prevalence is certainly on the increase. The development of higher incidence rates in communities where the illness was previously unknown may present opportune settings for the study of aetiological factors of this illness as it emerges. There is a need therefore for proper epidemiological studies to evaluate these factors, especially environmental factors, as the new disease continues to appear.

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Abstract – WCN 2013

No: 343

Topic: 2 – Movement Disorders

A long-term, open-label study of levodopa-carbidopa intestinal gel in advanced Parkinson's disease patients: Functional and health-related quality-of-life endpoints

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Background: Some Parkinson's disease (PD) motor complications are associated with pulsatile dopaminergic stimulation. Levodopa-carbidopa intestinal gel (LCIG) provides continuous drug infusion via an intrajejunal percutaneous gastrostomy tube.

Objective: To evaluate the effects of LCIG on functional and health-related quality-of-life (HRQoL) measures in patients with advanced PD.

Patients and methods: PD patients experiencing severe motor fluctuations (≥ 3 h/d OFF-time) despite optimized medical therapy were enrolled in an international, 54-week, open-label study. Individualized LCIG dosing was permitted up to 16 h/d; other PD medications were allowed after 28 days. Functioning and HRQoL were assessed at baseline and weeks 4, 12, 24, and 54 by the PDQ-39, UPDRS, EQ-5D, EQ-VAS and CGI-I.

Results: 272 (76.8%) of 354 patients completed the study. Total exposure to LCIG was up to 521 days (mean [SD] = 328.5 [132.3], $n = 350$). Mean [SD] PDQ-39 summary index was significantly improved at each visit (baseline = 42.8[15.1], $n = 320$; Endpoint = 35.8[16.8], $n = 317$ $P < .001$). UPDRS scores were improved at each visit ($P < .001$ all; Part II: baseline = 17.4[6.6], change at Endpoint = -4.4[6.5]; Part III: baseline = 28.8[13.7], change = -7.4[13.2]). Both EQ-5D and EQ-VAS improved from baseline through Endpoint ($P < .001$ all; EQ-5D: baseline = 0.588[0.195], Endpoint = 0.652[0.174]; EQ-VAS: baseline = 50.2[21.0], Endpoint = 64.1 [19.4]), as did CGI-I scores (Endpoint = 2.10[0.95]). AEs occurred in 323 (91.2%) patients; most were mild or moderate and transient.

Conclusion: In this large, open-label study in patients with advanced PD, LCIG provided significant improvements in functional and HRQoL measures that appeared as early as week 4 and were sustained through Endpoint.

Support: AbbVie.

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Abstract – WCN 2013

No: 1235

Topic: 2 – Movement Disorders

Ablation of rat substantia nigra is a good acute model of Parkinson's disease for cell transplantation therapy

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A valuable rat model of Parkinson's disease (PD) to study cell therapy was established. Forty-five female rats were examined. Using a stereotactic technique, we created an acute model of PD by high-frequency electric ablation of bilateral substantia nigra *pars compacta*. The ablation probe used was a 100 mm-long rigid bipolar needle electrode (1.0 mm in diameter). The rats developed persistent symptoms of PD, especially akinesia and postural instability. We tried all sorts of surgical protocols. The size and shape of the lesion that were ablated in a typical setting were evaluated. Then we determined that the optimal surgical parameter to place a lesion was 1 W of power for 15 s. The long-term viability of transplanted neural stem/progenitor cells was also examined and confirmed to be viable for more than 60 days following surgery. As the animal model was designed to suit the study of cell transplantation therapy, it did not utilize neurotoxic chemicals to make lesions and could avoid the harmful effects of them. The benefits of our ablation model of PD were the rapidity and uniformity of appearance of the characteristics of Parkinsonism. Therefore we can identify the onset of PD strictly. This feature is a unique merit that is not shared by the other methods. The technique described here may provide a good acute model of PD for the research of intracerebral cell transplantation. This model will contribute to the progress in the field of cell transplantation therapy.

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Abstract – WCN 2013

No: 1201

Topic: 2 – Movement Disorders

Distribution of wild and mutant torsinA in living cells

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Background: Mutation of torsinA is responsible for DYT1 dystonia. TorsinA is one of the AAA + proteins and is thought to result in synaptic pathology. Membrane distortion derived from mutant torsinA thought to cause disturbance of membrane trafficking and poverty of synaptic vesicles.

Objective: To analyze the distribution of wild type and mutant torsinA in living cells, we employed new fluorescent fusion proteins.

Materials and methods: We newly cloned wild type torsinA from cDNA library of human brain and obtained GAG deleted (deLE) clone by mutagenesis. Green fluorescent protein pEGFP (Clontech) was used for the production of fusion proteins with torsinA. All were transfected into Cos7 cells and SHSY5Y cells.

Results: TorsinA-pEGFP both wild type and deLE mutant localized endoplasmic reticulum (ER) and nuclear envelope in Cos7 cells. Wild type torsinA easily moved to perinuclear region with dense aggregation, probably aggresome. Mutant torsinA stayed ER and nuclear membrane. Cloned torsinA head portion remained ER and nuclear envelope, which is similar to mutant torsinA.

Conclusion: Previous reports revealed the different distribution of torsinA between wild type and deLE mutant protein similar to this result with different time range and time course. This study suggests that torsinA inserts ER and traffics to aggresome with binding proteins with torsinA instead vesicle movement to peripheral portion.

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Abstract – WCN 2013

No: 1196

Topic: 2 – Movement Disorders

The causes and route of admission to institutional care of people with Parkinson's disease

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Background: Institutional care is a major component of the cost of care for people with Parkinson's disease (PD) in the UK.

Objectives: To investigate the reasons for, and route of, admission to institutional care for people with PD.

Patients and methods: All patients known to the Northumbria Healthcare NHS Foundation Trust PD service with idiopathic PD, Parkinson's plus syndromes and vascular PD, who were living in institutional care on 1 January 2013 were included. Disease severity (Hoehn and Yahr stage) and demographic data were collected.

Results: Ninety people were included in the study (51 female). Patients were admitted from hospital (40%), home (58%) and sheltered housing (2%). On care home admission, mean age was 79.6 years (range 60–94) with average disease duration of 5.6 years. The reasons stated for admission to institutional care were patient inability to cope (41%), repeated falls (21%), cognitive impairment (20%), decreased mobility (20%), hallucinations (11%), spouse unable to cope (11%), delirium/confusion (8%), impaired swallow (3%), and stroke (2%). Many people had multiple reasons. Hoehn and Yahr stage was V in 48%, IV in 20%, III in 31% and I or II in only 1%.

Conclusion: Difficulty coping with activities of daily living was the most common cause of institutional care admission. Those admitted from hospital will have had a full multidisciplinary assessment of their needs, though for those admitted from home this may not be the case. More support in the community may prevent, or delay, institutional care admission.

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Abstract – WCN 2013

No: 1082

Topic: 2 – Movement Disorders

Intergenerational CAG repeat instability and mutational flow in Cuban families with spinocerebellar ataxia type 2

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Background: Spinocerebellar ataxia type 2 (SCA2) is a neurodegenerative disorder caused by the unstable expansion of a CAG repeat tract in ATXN2 gene.

Objective: To characterize CAG repeat variability in Cuban SCA2 population.

Patients and methods: A large dataset of 1745 individuals was assembled. The CAG repeat number was assessed by PCR.

Results: In 74 parent-to-child transmissions of large normal alleles (>24 repeats), a 37.8% of instability that was not associated to the gender of the transmitting parent ($p > 0.05$) was found. Two highly probable de novo mutations were identified in one of these families. In the 225 parent-to-child transmissions of expanded alleles studied a 76.4% of instability was found. The instability was highly significantly associated to the age at conception and gender of the transmitting parent, and to the size of the transmitted expanded allele ($p < 0.01$). A highly significant correlation was found for instability in sib-to-sib pairs after adjustment for age at conception and gender of the transmitting parent, and the size of the transmitted expanded allele ($r = 0.44$; $p < 0.001$). Intraclass correlation coefficient was 0.214, and familiarity of 42.8% for CAG repeat instability. The mutational flow for expanded alleles showed negative values in the range of 33 to 35 repeats and also for alleles greater than 57 repeats; the peak was found at 40.5 units.

Conclusions: There is an interplay of physiological and molecular factors in determining intergenerational instability at ATXN2 locus. These results have profound implications for genetic counseling services.

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Abstract – WCN 2013

No: 1182

Topic: 2 – Movement Disorders

Magnetic resonance imaging and proton spectroscopy in the Huntington disease

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Background: Structural abnormalities on MRI are found in carriers of Htt mutation, decades before manifest disease. Striatal atrophy correlates with a number of triplets, clinical symptoms and cognitive decline. In contrast to this, magnetic resonance spectroscopy (MRS) of high resolution show decreased concentration of N-acetyl-aspartate (NAA) and creatinine (Cr) in striatum and increased lactate (LL) and choline (Cho) in putamen in HD, not in preHD.

Report of cases: A 40-year-old-male and a 37-year-old-female patient presented with choreatic movements, psychiatric disturbances and cognitive decline. Genetic test revealed increased length of CAG triplets (male 24/40; female 20/54). In both patients coronal T2W1 MR showed striatal atrophy producing increased intercaudal distance and ventricular enlargement (“box-shape”) characteristic for HD. There is no putaminal hypointensities on T2, neither increased diffusion coefficient (ADC) in striatum on DWI, indicating hyperkinetic form of HD. MRS revealed depletion in NAA and Cr, and increased LL and Cho in caudatus. Increased LL and reduced NAA and Cr were found in putamen. Reduction in NAA was determined in prefrontal cortex. There were no metabolic alterations in thalamus.

Conclusion: Striatal atrophy, bicaudal diameter and intercaudal distance on structural and volumetric MRI may be used as biomarkers in preHD and HD, and correlates with pathology and clinical symptoms. Striatal atrophy on MRI is an early sign in preHD and HD and may be used for selection of asymptomatic carriers, assessment of conversion of preHD to HD and initiation of therapy. MRS detects metabolic changes in manifest HD and may be used in follow-up studies.

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Abstract – WCN 2013

No: 1189

Topic: 2 – Movement Disorders

Memory and attention in patients with Parkinson's disease

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Patients with Parkinson's disease (PD) have a high risk of developing Parkinson's disease dementia (PDD) with an accentuated decline in mental speed and executive functions and comparatively little deficits in short-term memory. PD patients without dementia may show these impairments to a smaller extent.

We determined the neuropsychological profile of PD patients not suffering from dementia.

In a multicentric study, the cognitive performance of PD patients was examined by means of the computer-based Memory and Attention Test (MAT). By means of the MAT, selective attention as well as working and short-term memory for verbal, figural and episodic material are assessed. The findings in the PD patients were compared with those of an age-, sex- and education-matched control group by means of unpaired Student's t-tests.

The study was conducted in 40 non-demented PD patients (26 men, 14 women) at ages between 48 and 82 years (mean/SD: 68.2/8.3 years). The PD stage according to Hoehn and Yahr was “1” in 2 patients, “2” in 21 and “3” in 7. MMSE scores ranged between 25 and 30 (mean/SD: 28.7/2.1). Neuropsychological testing revealed no impairments of short-term memory, a slight impairment of verbal working memory ($p \leq .05$) and a substantial impairment of selective attention, which increased with task complexity ($p < 0.01$).

In these non-demented PD patients we observed a decline of selective attention, which increased with task complexity, and a decrease of working memory. Both of these capacities are dependent on mental processing speed. Thus, mental slowing may have caused the deficits.

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Abstract – WCN 2013

No: 866

Topic: 2 – Movement Disorders

An open-label study in healthy male volunteers using positron emission tomography to assess brain adenosine A_{2A} receptor occupancy by V81444

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Background: V81444 is a selective non-xanthine adenosine 2A receptor (A_{2A}R) antagonist of potential utility in neurological disorders.

Objectives: We characterised the relationship between plasma concentration of V81444 and brain A_{2A}R occupancy following single oral doses.

Patients and methods: [¹¹C]SCH442416 PET scans were acquired before and at 2–3 h and 21–27 h after a single oral dose of V81444 (50, 100 or 250 mg) in 6 healthy male volunteers. A_{2A}R availability in the striatum was estimated using a simplified reference tissue model with cerebellum as reference tissue, and related to measured V81444 plasma concentration data.

Results: The images displayed a heterogeneous signal, consistent with the known regional distribution of A_{2A}R. Tissue time-activity curves were consistent with those described previously, with an early component (0–15 min) characterised by a rapid peak followed by washout, and a later component (15–90 min) dominated by a slow accumulating signal that was not regionally specific. Quantification of

data from the 0–15 min period produced binding potentials which were used to estimate $A_{2A}R$ occupancy. The relationship between V81444 plasma concentration and $A_{2A}R$ occupancy was consistent with a direct relationship, and yielded an estimate of EC50 (V81444 plasma concentration corresponding to 50% occupancy) of 320 ng/ml (95% CI: (198,442) ng/ml).

Conclusion: V81444 enters the brain following oral administration, and binds to $A_{2A}R$ in a dose dependent manner. V81444 displays a direct relationship between plasma concentration and brain $A_{2A}R$ occupancy. V81444 plasma concentrations >960 ng/ml (95% CI: (594,1326) ng/ml) are expected to lead to $A_{2A}R$ occupancy > 75%.

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Abstract – WCN 2013

No: 1183

Topic: 2 – Movement Disorders

The use of DaT-SPECT scanning in the differential diagnosis of parkinsonism within the Northumbria Parkinson's disease service

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Background: Dopamine transporter (DaT) scanning enables the determination of dopaminergic function within the basal ganglia. Dopaminergic function is decreased in Parkinson's disease (PD). UK NICE guidelines recommend that DaT scanning is used to differentiate between PD and essential tremor (ET).

Objectives: To determine whether referrals for DaT scanning follow NICE guidelines and how results affect management.

Patients and methods: Patients under the care of the Northumbria Healthcare NHS Foundation Trust PD service in the UK who had scans between 2010 and 2012 were included. Retrospective data collected included reason for scan, expected scan result, actual scan result and post-scan management.

Results: Two hundred and thirty-five people had a DaT scan, representing 24% of all referrals to the service during the study period. NICE guidelines were followed fully in only 73 (31%) of scans. The most common reasons for the scan were differentiation of PD from ET (31%) and confirmation of PD diagnosis (29%). One hundred and one scans (43%) were positive for PD, of which 85 (84%) were expected to be positive. Fifty-eight (57%) of those with positive scans were started on treatment immediately. Of 134 negative scans, 99 (74%) were expected, of which 9% led to PD treatment discontinuation and 18% to commencement of treatment for ET.

Conclusions: DaT scan can help direct the management of people with PD, although it should only be used where there is significant doubt over diagnosis. Future guidelines for use should consider amendments to widen the acceptable scope of use.

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Abstract – WCN 2013

No: 930

Topic: 2 – Movement Disorders

Saccade-related modulation of beta oscillation in the human internal globus pallidus

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Background: Studies in monkeys report more prominent pallidal cell firing during antisaccades than prosaccades (Yoshida 2009). We

reported saccade-related beta-band desynchronization (beta-SRDs) in the subthalamic nucleus (STN) of Parkinson's disease patients. We found more prominent beta-SRDs during antisaccades than prosaccades, like due to additional neural processing required. Since the STN projects to the internal globus pallidus (GPi), we expected enhanced beta-SRDs to antisaccades in the GPi.

Objective: To investigate whether oscillations in the GPi are modulated by saccades, and to compare the responses to prosaccades and anti-saccades. We hypothesized that antisaccades would produce earlier and/or more prominent beta-SRDs than prosaccades.

Patients and methods: We recorded the local field potentials from deep brain stimulation electrodes implanted in the GPi in 7 primary focal dystonia patients. The patients performed prosaccade and anti-saccade tasks. We analyzed the wavelet power spectrum averaged on saccade onset.

Results: The beta-SRDs were observed in the GPi just before and during saccades in 6 of 7 patients. The onsets, durations, and amplitudes of beta-SRDs were not different between prosaccades and antisaccades.

Conclusions: Beta-SRDs were observed in the GPi of dystonia patients. They were not related to inhibition of unwanted saccades, because they were not different between prosaccades and antisaccades. While it is well known that the basal ganglia output from the substantia nigra pars reticulata strongly inhibits the superior colliculus, our results suggest that the basal ganglia output from the GPi may contribute to the control of saccades, but are not a major contributor in dystonia patients.

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Abstract – WCN 2013

No: 1148

Topic: 2 – Movement Disorders

Is there a correlation between plasma homocysteine and Parkinson's disease associated with polyneuropathy?

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Background: Polyneuropathy has recently been described in higher proportions for patients with PD than in normal population. This finding was hypothesized to be related to the elevation of plasma homocysteine (Hcy), following the management of PD with levodopa. We conducted this study to clarify the clinical value of elevated plasma Hcy in PD patients.

Methods: A total of 37 PD patients without neuropathy (PDWo) and 41 PD patients with polyneuropathy (PDP), who were recruited for this study, were compared with 48 healthy controls. All PD patients performed electrophysiological tests, including nerve conduction study, to diagnose polyneuropathy. Plasma Hcy levels were measured in all subjects and compared with each groups.

Results: The Hcy of PDP showed higher Hcy level than those of PDWo, as well as healthy controls, but there was no significant difference between those of PDP and that of the healthy controls. In considering each group of PDWo and PDP, there were no intercorrelations between daily levodopa dose, duration of PD symptoms and PD treatment or motor severity.

Conclusion: We could cautiously assume that plasma Hcy level may be related with the involvement of peripheral nerve of PD patients in this study. However, we were unable to confirm pathophysiologic role of Hcy and the relationship between plasma Hcy level and levodopa in PDP.

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Abstract – WCN 2013**No: 382****Topic: 2 – Movement Disorders****Long-term complications of Parkinson's disease – 15th year, 20th year, and beyond; A hospital-based observational study**H. Shibayama, S. Kaji, D. Nishida, S. Hirata, F. Katada, S. Sato, T. Fukutake. *Neurology, Kamada Medical Center, Kamogawa, Japan***Background:** Long-term complications of Parkinson's disease (PD) based on regular follow-up in a local neurological center is insufficiently appreciated.**Patients and methods:** From registered PD patients, we selected those who have been examined in 15th and/or 20th year of the disease (YD) and information about motor status, levodopa equivalent daily dosage (LEDD), hallucination, cognitive disturbance, and dysphagia in each year was collected.**Results:** Of all PD patients between April 1995 and December 2012 ($n = 538$, male/female 222/316), 72 patients (13.4%) were examined in their 15th YD (Group PD ≥ 15 ; age at onset (AO) 58.1 ± 8.1 year-old; modified Hoehn and Yahr stage (mHY) at on/off, $3.3 \pm 0.9/3.9 \pm 0.9$; LEDD 567 ± 239 mg/day) and only 18 patients (3.3%) have taken their 20th YD examination (Group PD ≥ 20 ; AO 53.4 ± 6.6 , mHY at on/off, $3.8 \pm 0.9/4.4 \pm 0.7$; LEDD 703 ± 208 mg/day). Groups PD ≥ 15 and PD ≥ 20 have significantly younger AO ($p = 0.0001$, $p < 0.0001$) than patients who have less than 15th YD history (PD < 15 , $n = 466$, AO 67.4 ± 8.8). In Group PD ≥ 15 /PD ≥ 20 , troublesome hallucination (mainly visual, $n = 36/13$, 50.0/72.2%) appeared in $15.1 \pm 2.8/17.7 \pm 2.6$ th year, cognitive disturbance ($n = 27/10$, 37.5/55.6%) in $16.0 \pm 2.9/18.0 \pm 2.5$ th year, and dysphagia ($n = 29/13$, 40.3/72.2%) in $18.1 \pm 3.4/21.0 \pm 2.3$ th year. Follow-up periods after appearance of hallucination, cognitive disturbance, and dysphagia in Group PD ≥ 15 are 4.2 ± 2.7 , 3.2 ± 2.6 , 1.7 ± 2.1 years, respectively. The longest treated patient had AO at 54 year-old and was in mHY 3/0/4.0 in 29th YD without these 3 complications.**Conclusions:** During long-term treatment of PD, hallucination, cognitive disturbance and swallowing problem appear in near half or more of the patients after 15th YD and seem to contribute to lost follow-up.

doi:10.1016/j.jns.2013.07.424

Abstract – WCN 2013**No: 995****Topic: 2 – Movement Disorders****Role of magnetic resonance spectroscopy in differentiating parkinsonian syndromes of various etiologies**I. El-Banhawy^a, M. El-Toukhy^b, I. Fahmy^a, N.M. Shalaby^a, M.A. Nada^a, H.S. Shehata^a, I. Ismaiel^a. ^aNeurology, Cairo University, Cairo, Egypt; ^bRadiology, Cairo University, Cairo, Egypt**Background:** Parkinsonism is not a single disease entity as it has a number of causes that differ in the pathology according to the affected brain region. To our knowledge, no previous studies have been published to investigate the role of magnetic resonance spectroscopy (MRS) in the differentiation between idiopathic Parkinson's disease (IPD), drug-induced parkinsonism (DIP) and Wilson's disease (WD).**Objective:** Investigating the role of MRS in the differentiation between various parkinsonian syndromes.**Subjects & methods:** The study was conducted on 40 parkinsonian patients: 27 patients with IPD, 6 patients with DIP, and 7 patients with WD, and 15 normal control subjects. All were examined using single voxel proton MRS study of striatum, frontal cortex and subcortical white matter. Measured MRS parameters were N-acetylaspartate (NAA), choline-containing compounds (Cho) and creatine-phosphocreatine (Cr) and with peak area ratios (NAA/Cr, Cho/Cr, NAA/Cho ratios).**Results:** Patients with parkinsonism had significantly more cortical Cr and less NAA/Cr ratio and more subcortical NAA, Cr, Cho, and Cho/Cr ratios compared to control subjects. DIP patients showed significantly lower striatal Cho/Cr ratio and significantly higher subcortical Cho/Cr ratio compared to IPD. While comparing WD and IPD patients, a significantly lower cortical Cho/Cr ratio and significantly higher subcortical NAA/Cr and cortical NAA/Cho ratios were found in WD. Patients with WD had significantly higher cortical and subcortical NAA/Cho and striatal NAA/Cr ratios compared to DIP patients.**Conclusion:** MRS can be used as a useful in vivo investigative tool in differentiating between parkinsonian syndromes of various etiologies.

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Abstract – WCN 2013**No: 203****Topic: 2 – Movement Disorders****A long-term, open-label study of levodopa-carbidopa intestinal gel in advanced Parkinson's disease patients: safety and motor-symptom endpoints**A.J. Espay^a, P. Odin^{b,c}, H.H. Fernandez^d, R.A. Hauser^e, D.G. Standaert^f, A.E. Lang^g, V.S. Fung^h, F. Klostermannⁱ, W.Z. Robieson^j, K. Chatamra^j, J. Benesh^l. ^aUniversity of Cincinnati Academic Health Center, Cincinnati, OH, USA; ^bKlinikum-Bremerhaven, Bremerhaven, Germany; ^cSkane University Hospital, Lund, Sweden; ^dCleveland Clinic, Cleveland, OH, USA; ^eUniversity of South Florida, Tampa, FL, USA; ^fUniversity of Alabama at Birmingham, Birmingham, AL, USA; ^gUniversity of Toronto, Toronto, ON, Canada; ^hWestmead Hospital and Sydney Medical School, Sydney, NSW, Australia; ⁱCharite-University Medicine Berlin, Berlin, Germany; ^jAbbVie Inc., North Chicago, IL, USA**Background:** Motor complications in Parkinson's disease (PD) are associated with pulsatile dopaminergic stimulation. Levodopa-carbidopa intestinal gel (LCIG) provides continuous drug infusion via an intrajejunal percutaneous gastrostomy tube.**Objective:** To present safety and efficacy results of an international, 54-week, open-label study of LCIG in patients with advanced PD.**Patients and methods:** PD patients experiencing severe motor fluctuations (≥ 3 h/d OFF-time) despite optimized medical therapy were enrolled. Individualized LCIG dosing was permitted up to 16 h/d; other PD medications were allowed after 28 days. Efficacy outcomes included OFF-time, ON-time with- and ON-time without troublesome dyskinesias (TSD) (patient-diary-assessed), and UPDRS. AEs were monitored.**Results:** 272 (76.8%) of 354 patients completed the study. Mean (SD) exposure to LCIG was 328.5 (132.3) days. Mean [SD] OFF-time (baseline = 6.8[2.4] h/d, endpoint = 2.3[2.1], $P < .001$) and ON-time with TSD (baseline = 1.6[2.0], endpoint = 1.2[2.1], $P = .002$) were reduced, while ON-time without TSD (baseline = 7.6[2.5], endpoint = 12.4[2.6], $P < .001$) was increased. UPDRS scores were improved (Part II: change at endpoint = -4.4[6.5]; Part III: change = -7.4[13.2]; $P < .001$ both). Mean total daily dose of levodopa (mg) was steady (Week 4 = 1613.7; Endpoint = 1620.9). Ninety subjects (27.8%) took only LCIG; 158 (48.8%) took only concomitant oral levodopa formulations. While AEs occurred in 323 (91.2%) patients, most were mild or moderate and transient, and only 27 (7.6%) withdrew due to an AE. Complication of device insertion (34.9%) and abdominal pain (31.2%) was most common.**Conclusion:** In this large, long-term, prospective study, LCIG provided advanced PD patients with significant and clinically meaningful improvements in motor function and acceptable safety.**Support:** AbbVie.

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Abstract—WCN 2013**No: 1088****Topic: 2—Movement Disorders****N-Methyl-d-aspartate receptor (NMDAR) antibodies in post herpes simplex virus encephalitis (HSVE) neurological relapse**

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Background: Patients with HSV encephalitis can present with a second phase of encephalopathy often predominated by a movement disorder. These clinical relapses are commonly treated as a viral reactivation, although frequently no evidence of a viral reactivation can be identified, and post-infectious autoimmune mechanisms appear likely. There are now a number of distinct neurological syndromes in which brain directed antibodies have been identified, with a significant number of conditions featuring movement disorders. In particular, the movement disorder seen in NMDAR-antibody encephalitis has phenotypic similarities to the choreoathetosis in relapsing HSVE.

Methods: From a total of 20 cases of HSVE, we identified seven children who had a neurological relapse following their initial encephalitis. We tested their sera and CSF (n = 1) for autoantibodies reported in CNS autoimmune conditions.

Results: All children had presented with a typical history of HSVE confirmed by clinical, laboratory and radiological findings, and with improvement following intravenous acyclovir. At the time of their relapses, viral reactivation was identified only in one patient (14%), and no identifiable cause was found for the other six. On retrospective analysis of their sera, three out of six (50%) children, who had choreoathetosis, seizures and/or cognitive regression at relapse, had NMDAR antibodies. In two of the NMDAR antibody positive patients who were treated at relapse, a beneficial response was observed.

Conclusion: NMDAR-antibodies appear to be relatively common in children who often exhibit choreoathetosis at relapse. Immunotherapy should be considered in children who relapse after HSVE.

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Abstract—WCN 2013**No: 1115****Topic: 2—Movement Disorders****Diplopia in Parkinson's disease: Prevalence and associations with other motor and nonmotor symptoms**

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Background: In Parkinson's disease (PD) a range of visual symptoms occurs and little is known about diplopia which may impact on the patient's quality of life (QoL). The PD nonmotor symptoms questionnaire (NMSQ) and scale (NMSS) offer a quick way of self declaration (NMSQ question 29) and grade rating of severity and frequency (NMSS question 15) of diplopia in PD.

Objectives: In this retrospective analysis we report the prevalence of diplopia in 169 cases of PD evaluated at Kings College and Lewisham University hospitals (as part of a nonmotor natural history global study) as well as its relation to motor and other nonmotor symptoms (NMS), in particular cognitive impairment, depression, falls and nocturnal vivid dreams.

Methods: We analysed the NMSS records of 169 PD patients for diplopia, which was classified as mild (score 1–4), moderate (5–8) and severe (9–12). We then sought correlations with other relevant NMS and motor symptoms.

Results: 23 PD patients (13.6% of 169) reported diplopia ranging from mild to severe. Out of these 6 (26%) had moderate diplopia and 1 (4.4%) severe. 91.3% were between Hoehn and Yahr stages 2 and 3 and no significant correlations with cognitive impairment, falls, depression or intrusive vivid dreams were found although this may reflect the small sample size.

Conclusions: Our ongoing study suggests that 13.6% of our cohort of unselected PD cases developed diplopia (over 25% with moderate to severe diplopia). We are now expanding this study to a large European cohort to explore causations and correlations.

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Abstract—WCN 2013**No: 945****Topic: 2—Movement Disorders****Glutathione S-transferase omega 2 influence the clinical phenotype of spinocerebellar ataxia type 2**

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Background: Spinocerebellar ataxia type 2 is a neurodegenerative disorder caused by expansions of a CAG repeat sequence in ATXN2 gene. The CAG repeat number is the major genetic factor influencing disease onset and severity, but there is still wide phenotypic variation.

Objective: To assess potential modifier effects of rs4925 GSTO1 and rs2297235 GSTO2 polymorphisms on age at onset and disease severity in spinocerebellar ataxia type 2 affected individuals.

Patients and methods: Polymorphisms rs4925 GSTO1 and rs2297235 GSTO2 were genotyped in 124 patients with SCA2 and 100 controls.

Results: There were highly significant differences between affected and control individuals for rs2297235 GSTO2 genotypes under additive and dominant models (p = 0.01). Allelic frequencies were significantly different only for rs2297235 GSTO2 polymorphism (p = 0.046). In affected individuals, expanded CAG repeat number explained 37.1% of log transformed age at onset variability. rs2297235 GSTO2 genotypes, under additive and dominant models, significantly influenced the residual age at onset after adjusting for the expanded CAG repeat number and explained 3.0% of total variation and about 4.8% of the unexplained variation in age at onset. SARA scores were significantly influenced by CAG repeat number and disease duration. After adjusting for these two factors by multiple linear regression, neither rs4925 GSTO1, rs2297235 GSTO2 genotypes nor GSTO1–GSTO2 haplotype combinations significantly influenced SARA scores (p = 0.05).

Conclusion: rs2297235 GSTO2 polymorphism is a genetic modifier for age at onset in spinocerebellar ataxia type 2 affected individuals.

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Abstract—WCN 2013**No: 401****Topic: 2—Movement Disorders****Correlation between cognitive function and motor symptoms in Parkinson's disease**

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Background: In conventional management of Parkinson's disease (PD), treatment of motor dysfunction normally precedes that of cognitive impairment. Recent research suggests anti-dementia drugs may help cognitive impairment in PD. Thus, both motor and cognitive impairment will be treated simultaneously.

Objective: We examined the relationship between the cognitive and motor dysfunction in patients with PD treated with dopaminergic drugs for motor dysfunction.

Patients and methods: 49 patients (27 male, 22 female) with PD, and PD with dementia (PDD), taking dopaminergic drugs but no anti-dementia drugs (acetylcholinesterase inhibitor and NMDA receptor antagonist) participated. All patients were assessed on the Unified Parkinson's Disease Rating Scale (UPDRS) (Parts II and III), the Montreal Cognitive Assessment (MoCA) and the Frontal Assessment Battery (FAB). The MoCA and FAB scores were compared with UPDRS (II plus III) for each patient.

Results: Both MoCA and FAB scores showed highly significant negative correlation with UPDRS (II plus III) ($p \leq 0.0001$). The correlations were stronger than those with current age and duration of PD. They also showed significant negative correlation with UPDRS subscores of postural instability and akinesia, but not tremor. The MoCA score had a weak negative correlation with UPDRS subscore of rigidity, but the FAB score did not.

Conclusion: In patients with PD on dopaminergic agent, cognitive impairment, especially frontal lobe dysfunction, correlates with motor dysfunction such as postural instability and akinesia but not tremor. Pathophysiology of tremor is probably different from other motor symptoms.

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Abstract—WCN 2013**No: 1087****Topic: 2—Movement Disorders****Drug induced movement disorders—Pathophysiology, diagnosis and management**

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Drug induced movement disorders are generally caused by dopamine receptor blocking drugs. There is a great variety of drug induced acute and tardive movement disorders. Early onset drug induced movement disorders include akathisia, neuroleptic malignant syndrome, parkinsonism and dystonic reactions. They appear within hours to days after treatment initiation.

Late onset category of induced movement disorders include dyskinesia, dystonia, akathisia and chorea. They manifest months after exposure to drugs or when the dosage is increased or treatment is discontinued.

Pathophysiology of these drug induced disorders refers to many hypotheses: neurotoxicity, hypersensitivity of postsynaptic dopamine receptor, hyperactivation of the cholinergic system and also genetic predisposition.

The symptomatology of drug induced movement disorders is sometimes hard to differentiate from that of an idiopathic one and may resemble several other medical conditions.

We will discuss about the epidemiology, mechanism, differential diagnosis, risk factors, prevention and management of these syndromes as well as diagnostic procedures (PET, SPECT, MRI scans with T2 diffusion-weighted, DAT-scan) and therapeutic goals.

Treatment of these disorders inconsistently has a benefit and therefore the primary prevention is essential.

Movement disorders are also associated with antiemetics that block central dopamine receptors, lithium, selective serotonin reuptake inhibitors, stimulants, tricyclic antidepressants, organophosphates, organic solvents, carbon monoxide, and cyanide.

Conclusions: Incidence and prevalence of drug induced movement disorders are not entirely known due to a lack of recognition and a lack of validated criteria for the diagnosis of drug induced movement disorders.

This article focuses on diagnosis and mechanisms of these drug induced disorders and also on treatment and prevention.

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Abstract—WCN 2013**No: 1065****Topic: 2—Movement Disorders****Adverse drug reactions with Selegiline and Rasagiline compared to Levodopa and Ropinirole: A study in the French Pharmacovigilance Database**

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Background: The safety of Rasagiline has not been as extensively explored as Selegiline.

Objective: To compare spontaneous adverse drug reaction (ADR) reports to the French Pharmacovigilance Database (FPVD) between Selegiline and Rasagiline, taking Levodopa and Ropinirole as references.

Methods: We retrieved all ADRs in which Selegiline, Rasagiline, Levodopa or Ropinirole were considered as "suspected" drugs. ADRs were classified according to system-organ class (MeDRA).

Results: We retrieved 199 ADR reports for Selegiline, 132 for Rasagiline, 1851 for Levodopa and 432 for Ropinirole. Confusion, hallucination or agitation, were more frequent with Selegiline compared to the rest of the drugs (respectively, 19% vs 10%, 12% vs 8% and 6% vs 2%). Impulse control disorders were more frequent with Ropinirole vs Rasagiline (12% vs 4%) and less frequent with Selegiline (1%). Somnolence was less frequent with Selegiline and Rasagiline vs Ropinirole (1% and 4% vs 16%). Headache was more frequent with Rasagiline compared to rest of the drugs (5% vs 1%). Orthostatic hypotension was less frequent with Rasagiline vs Selegiline (1% vs 9%). Weight loss was more frequent with Rasagiline or Selegiline. Renal ADRs were more frequent with Rasagiline compared to the rest of drugs (6% vs 1%). Musculoskeletal ADRs were more frequent with Rasagiline compared to the rest of drugs (11% vs 2%).

Conclusion: Compared to Selegiline, Rasagiline was associated with a higher risk of renal or musculoskeletal ADRs, headaches or impulse control disorders. Conversely, the risk of orthostatic hypotension, confusion, hallucinations or agitation was higher with Selegiline.

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Abstract—WCN 2013**No: 1055****Topic: 2—Movement Disorders****Freezing of gait in patients with Parkinson disease is a kind of kinetic subcortical apraxia**

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Background: Freezing of gait is one of the late signs associated with apractic features which may be considered as a subcortical kinetic apraxia.

Objective: To investigate the correlation between the frequency and severity of freezing of gait and apractic disorders.

Material and methods: We examined 70 patients with PD: 39 men and 31 women (mean age—64.5 ± 8.5 years; mean disease duration—5.1 ± 3.5 years, Hoehn–Yahr stages from 2 to 4, mean UPDRS (part III) score—43.2 ± 11.6). The Gait and Balance examination was carried out with GABS (Jankovic et al., 2001) and FOG-Q (Gurevich et al., 2003) in combination with a comprehensive neuropsychological study including a 72-item apraxia scale. The apraxia scale consists of the oral, arm, leg and trunk apraxia examinations tests, and each of them includes imitation tests and tests on command. Patients were divided into 3 groups according to their FOG-Q score.

Results: Freezing of gait was found in 24 (34.3%) of the PD patients. Patients with freezing had advanced stages of PD, a higher UPDRS score and more severe axial symptoms ($p < 0.0001$) compared with the non-freezing patients. Patients with a higher FOG-Q score performed worse in the executive and visuospatial cognitive tests and they had a higher leg apraxia score ($p < 0.05$). Their performance in the apractic tests on command was worse than in the imitation tests.

Conclusion: Pathophysiology of FOG in PD is unclear but a subcortical apractic defect should be considered among the possible mechanisms of its origin.

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Abstract—WCN 2013**No: 1058****Topic: 2—Movement Disorders****WOQ-19 improves satisfaction of patients with Parkinson's disease**

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Objective: To analyse whether a 19-item Wearing-Off (WO) Questionnaire (WOQ-19) improves the sense of satisfaction in patients with Parkinson's disease (PD).

Background: It is very difficult to comprehend all complaints of PD patients in outpatient practice. The WO symptom is particularly under-recognized and associated with patient's quality of life and sense of satisfaction. WOQ-19 has 19 simple questions and is designed to detect WO.

Methods: This study included PD patients treated with L-dopa. We compared treatment satisfaction using WOQ-19 to that without using it. The sense of satisfaction was measured using a modified questionnaire for patient–physician communication, which is designed by the Office of Pharmaceutical Industry Research and can evaluate patient's factors, physician's factors and the patient–physician relationship. If WO is recognized using WOQ-19, then entacapone treatment is initiated.

Results: Seven PD patients enrolled for this study (two males and five females; age range, 54–88 years; mean, 75.1 years). Among them, two patients were administered entacapone on the basis of WOQ-19 results. Treatment satisfaction was improved using WOQ-

19, particularly for the question 'satisfaction of the patient-centred approach'.

Conclusion: WOQ-19 may improve the sense of satisfaction in PD patients and be useful for patient education.

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Abstract—WCN 2013**No: 1006****Topic: 2—Movement Disorders****Circadian blood pressure and heart rate variations in *de novo* Parkinson disease**

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Background: Altered blood pressure (BP) regulation and heart rate variations are characteristic findings of cardiovascular Dysautonomia in patients with Parkinson's disease (PD). However, these variations had not been adequately investigated.

Objective: The aim of this study was to investigate the patterns and characteristics of 24-hour BP variations and heart rate variations in patients with PD.

Patients and methods: Case–control comparisons of 142 consecutive newly diagnosed patients with PD and 57 age-matched controls were performed. All cases underwent clinical assessments and 24-hour ambulatory BP monitoring. The associations between BP and heart rate variations and parkinsonian motor symptoms were investigated.

Results: There were significant differences in the distribution of non-dipping, the percent of nocturnal BP decrease, the standard deviation of heart rate and nocturnal decrease of heart rate between patients with PD and controls. However, these abnormal diurnal BP and heart rate patterns were not associated with parkinsonian motor symptoms and not related to age, gender, or disease duration.

Conclusion: In conclusion, this result suggests that non-dipping and decreased nocturnal heart rate may be one of the cardiovascular autonomic dysfunctions in patients with PD, irrespective of age, disease severity, or motor symptom phenotype.

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Abstract—WCN 2013**No: 1027****Topic: 2—Movement Disorders****Motor sequence learning and motor adaptation in primary cervical dystonia**

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Background: Motor sequence learning and motor adaptation rely on overlapping circuits predominantly involving the basal ganglia and cerebellum. Given the importance of these brain regions to the pathophysiology of primary dystonia, and the previous finding of abnormal motor sequence learning in DYT1 gene carriers, we explored motor sequence learning and motor adaptation in patients with primary cervical dystonia.

Methods: We recruited 12 patients with cervical dystonia and 11 healthy controls matched for age. Subjects used a joystick to move a cursor from a central starting point to radial targets as fast and accurately as possible. Using this device, we recorded baseline motor performance, motor sequence learning and a visuomotor adaptation task.

Results: Patients with cervical dystonia had a significantly higher peak velocity than controls. Baseline performance with random target presentation was otherwise normal. Patients and controls had similar levels of motor sequence learning and motor adaptation.

Conclusion: Our patients had significantly higher peak velocity compared to controls, with similar movement times, implying a different performance strategy. The preservation of motor sequence learning in cervical dystonia patients contrasts with the previously observed deficit seen in patients with DYT1 gene mutations, supporting the hypothesis of differing pathophysiology in different forms of primary dystonia. Normal motor adaptation is an interesting finding. With our paradigm we did not find evidence that the previously documented cerebellar abnormalities in cervical dystonia have a behavioral correlate, and thus could be compensatory or reflect “contamination” rather than being directly pathological.

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Abstract—WCN 2013

No: 1017

Topic: 2—Movement Disorders

An autopsy case of Perry syndrome with DCTN1 156 T>A mutation

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We report the first autopsy case of Perry syndrome with a DCTN1 156 T>A mutation. A 74-year-old woman had the onset of resting tremor and bradykinesia at the age of 70. She was diagnosed with Parkinson disease and treated with l-dopa. Sudden onset of respiratory failure appeared at the age of 72 and the disease progressed with repeated remission and exacerbation of respiratory failure. Autonomic dysfunctions such as bradycardia and prominent changes in blood pressure appeared over the course. She died at the age of 74 because of aspiration pneumonia. Autopsy study revealed slight neuronal loss and reduction of melanin granule in substantia nigra without apparent gliosis and TDP-43-positive inclusion body.

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Abstract—WCN 2013

No: 976

Topic: 2—Movement Disorders

Evolution of movement related cortical potentials in early stage Parkinson disease

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Objective: The purpose of this study was to evaluate the role of cortical potentials related to movement in diagnostic stage and physio-pathology of movement disorder in the cases of early-stage Parkinson disease.

Methods: The data were acquired from forty patients with idiopathic Parkinson disease who did not have dementia and psychiatric disorders. A survey was performed among Parkinson disease and movement disorder polyclinics in Konya, Turkey.

Symptoms: The average age of patients was 66.6 ± 7.2 years and their ages were between 52 and 81 years. The period of the beginning of the disease's lower limit was 1 year, the upper limit was 6 years and the average was 3.15 ± 2.94 years. The age of the beginning of the disease's lower limit was 52, the upper limit was 72 and the average was 63.57 ± 7.23 . While the Hoehn & Yahr score's lower average was 1.37 ± 0.49 , UPDRS's lower limit was 15, the upper limit was 62 and the average was 29.32 ± 11.47 . DIP records were obtained from

triggering the right and left hand thumbs with random and counting patterns. When Parkinson disease group's and control group's DIP records were compared, N0 (msn), N1 (amp), N500 (amp), N650 (amp) values were plausible.

Results: Parkinson disease can be exactly diagnosed via only pathological examination but it is not used for diagnosis because it is an invasive method. Therefore, clinical diagnosis criteria are used. Cortical potentials related to movement in Parkinson disease can be used for examination as a non-invasive method and a companion to objective diagnosis. Furthermore, this method proves that it can aid the enlightenment of the neural mechanism of movement.

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Abstract—WCN 2013

No: 960

Topic: 2—Movement Disorders

Validation and cross-cultural adaptation of the self-assessment disability scale in patients with Parkinson's disease in Serbia

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Background: The symptoms of Parkinson's disease (PD) deteriorate over time affecting performance and causing disability.

Objective: The purpose of this study was to translate the Self-Assessment Disability Scale in patients with parkinsonism (SADS-PD) into Serbian language and assess its validity and reliability.

Methods: From January to July 2012, 114 consecutive PD patients were recruited at the Neurology Clinic in Belgrade. The inclusion criteria were: ability to walk independently for at least 10 m and ability to stand for at least 90 s. The exclusion criteria were: cognitive impairment, the presence of other major neurologic, psychiatric, visual, audio-vestibular, and orthopedic disturbances. The 25-item SADS-PD was translated according to an internationally-accepted methodology. The internal consistency of the scale was evaluated using Cronbach's alpha coefficient. Test–retest reliability was evaluated using Kendall's concordance coefficient for total scores. To evaluate construct validity, an exploratory factor analysis (principal component analysis, varimax rotation) was performed.

Results: Cronbach's alpha coefficient was 0.984. Kendall's concordance coefficient was 0.994. Duration of the disease, Hoehn & Yahr (H&Y) stage, Unified Parkinson's Disease Rating Scale (UPDRS) motor score, history of falls, and Hamilton's Depression and Anxiety Rating Scales (HDRS and HARS) scores were significantly correlated with the total SADS-PD score. On factor analysis the 25 items in the SADS-PD questionnaire were separated in 2 clusters with a total matrix variance of 79.7%.

Conclusion: The psychometric properties of the cross-culturally adapted SADS-PD questionnaire (Serbian version) have outstanding validity and reliability as an instrument for evaluation of the extent of disability in patients with PD.

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Abstract—WCN 2013

No: 961

Topic: 2—Movement Disorders

Pharmaceutical quality of seven generic levodopa/benserazide products compared with original Madopar®/Prolopa®

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Objective: To compare the pharmaceutical quality of seven generic levodopa/benserazide combinations with the original product (Madopar®/Prolopa® 125, Roche).

Introduction: Madopar®/Prolopa® is a combination of levodopa (l-Dopa) and benserazide. Generics assume the same composition and presentation as the original product.

Materials: Madopar®/Prolopa® 125 tablets and capsules were used for reference. Generics tested included four tablet (Betapharm, CT, MYLAN and ratiopharm) and three capsulated formulations (HEXAL, STADAPHARM and TEVA).

Methods: Shape, colour, appearance and hardness (tablets), resp. surface, colour, imprint and content (capsules), mass, degradation products, impurities, disintegration and dissolution were all tested.

Results: For all seven generic products at least one of the parameters fell outside the specifications. Content requirements were unmet by two tablet formulations for l-Dopa (specs: 100 mg ± 5%; test results: -5.6% and -7.6%) and for benserazide (specs: 28.5 mg ± 5%; -6.6% and +8.4%). Average mass (specs: 275 mg ± 3%) was not reached for two tablet formulations (+3.3%), and average fill mass (specs: 150 mg ± 5%) was not reached for three capsule formulations (+48.2%, +99.1%, +99.3%). The level of two benserazide degradation products (Ro 04-1419 and Ro 08-1580; specs: max. 0.49%) was off-limits (0.62%) for one capsule.

Conclusions: All seven analysed generic l-Dopa/benserazide products exhibited deviations when compared to Madopar®/Prolopa® 125. Deviations ranged from +8.4% (benserazide) to -7.6% (l-Dopa) in two tablets, with potential clinical consequences. Two of the degradation products were 26.5% above specifications, with potential safety concern. This should lead to conservative prescribing and invite further investigations, both pharmaceutical and clinical.

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Abstract—WCN 2013

No: 885

Topic: 2—Movement Disorders

The presence of RWA and RBD in patients with different types of MSA—A pilot videopolysonnographic study

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Background: Patients with multiple system atrophy (MSA) can present with different sleep problems. There is sparse data in the literature comparing polysomnographic parameters of the two types of the disease, namely MSA cerebellar type (MSA-C) and MSA Parkinson type (MSA-P).

Objective: To analyze and compare REM sleep without atonia (RWA) and REM sleep behavior disorder (RBD) in patients with the two subtypes of MSA.

Patients and methods: Over a course of two years 11 MSA-C and 27 MSA-P patients were studied with video-supported polysomnography (vPSG) for one night. 37 valid vPSGs were available for analysis. RWA was measured in 3-second mini-epochs as any chin EMG activity in REM sleep according to the method published by Frauscher et al. Severity of RBD was quantified using the RBD severity scale (RBDSS).

Results: RBD was detected in 8/11 (73%) patients with MSA-C and 19/25 (76%) patients with MSA-P ($p = 0.849$). RWA values were measured at 46.79% for MSA-C patients with RBD and 51% for MSA-P patients with RBD ($p = 0.208$). In the group without RBD, RWA was measured at 7.35% for MSA-C patients and 17.72% for MSA-P patients ($p = 0.354$).

Conclusion: Even though the pathology of the two subtypes of MSA is somewhat different, we found no significant difference in RBD and RWA measurements in patients with MSA-C and MSA-P.

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Abstract—WCN 2013

No: 592

Topic: 2—Movement Disorders

The Dystonia Coalition: Four years of progress

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Background: Clinical research in rare disorders, such as dystonia, is challenging because of limited access to large numbers of patients or patient materials.

Objective: The objective is to advance the pace of clinical and translational research in primary dystonias by fostering a collaborative atmosphere.

Methods: The Dystonia Coalition (DC) is a collaboration of medical researchers and patient advocacy groups that is funded by the National Institute of Neurological Disorders and Stroke and the Office of Rare Diseases at the National Center for Advancing Clinical and Translational Research. The DC has funding to support 4 large clinical research projects, a pilot project program to provide seed funding for new projects, career development awards to encourage new investigators to conduct dystonia research, and regular meetings to delineate important research problems.

Results: The DC has 47 clinical centers in North America and Europe. As of March 29, 2013, these centers have recruited 1044 participants with different primary dystonias into a biorepository study, 169 participants into a longitudinal natural history study, 210 participants into a study aimed at revising and validating the TWSTRS scale for cervical dystonia, and 132 participants into a study developing a new rating scale for spasmodic dysphonia. The DC has provided 11 additional grants for pilot projects proposed by its members and 10 career awards for junior investigators.

Conclusions: The DC provides an effective forum for the scientific community to define and address common goals in a collaborative manner with the direct input and support of patient advocacy groups.

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Abstract—WCN 2013

No: 955

Topic: 2—Movement Disorders

Orthostatic tremor heralding the onset of stiff-person syndrome

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Background: Orthostatic tremor (OT) is a rare movement disorder triggered exclusively by standing and accompanied by 13–18 bursting EMG patterns. Exaggerated startle, stiffness, and spasms of the axis characterize stiff person syndrome (SPS).

Objective: To report two patients with abnormal muscle tone control heralding as OT and briefly transforming into SPS.

Patients and methods: A 77-year-old and a 55-year-old female developed disabling unsteadiness with clinical and electrophysiological evidence of OT and benefit from clonazepam. Surprisingly the clinical picture underwent a change in a short time with progressive stiffness of the axial and mainly lower limb muscles with spasms and falls.

Results: MRI of the brain, cervical, thoracic and lumbar spine was normal. Results of antibody testing revealed anti-glutamic acid decarboxylase (anti-GAD) antibodies in the blood (>2000 IU/mL), whereas amphiphysin antibodies were absent. A search for an underlying neoplasm revealed no abnormalities. They were now diagnosed as having classic SPS and treatment with intravenous immunoglobulin (IVIg) conferred significant improvement.

Conclusion: Brainstem and spinal cord hyperexcitability conditions give rise to progressive alterations of muscle tone, posture and voluntary movement which are potentially immunotherapy-responsive. In this regard, OT observed in our patients can be considered as a “prodromal and soft sign” of a more complex and ongoing muscle tone dis-control. A search also for autoimmunity in a specific patient with “isolated OT” can be used to prompt for the most accurate clinical investigation and to choose the most timely and appropriate therapy.

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Abstract—WCN 2013

No: 634

Topic: 2—Movement Disorders

The disruption of ventral striatum and amygdala activity to financial cues in never-medicated Parkinson disease patients

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Background: The study of emotional processing in Parkinson disease (PD) has been of great interest with the increased incidence of impulse control disorders in patients with dopaminergic medications. However, it is still controversial whether the impulsive decision-making represents a behavioral endophenotype for PD.

Objectives: This study investigated the altered neural processes underlying behavioral responses in never-medicated PD patients by using a neuroeconomics task in conjunction with functional magnetic resonance imaging.

Methods: The de novo PD patients (n = 9) and age matched healthy controls (n = 8) performed a modified monetary incentive delay task, in which motor response was required for incentive cues that predicted the subsequent delivery of a financial reward or punishment. We analyzed the brain activities between incentive presentations and motor responses.

Results: In untreated PD patients, the activation in the ventral striatum was significantly reduced under reward anticipation, compared to the control. In addition, the activation in the amygdala was also significantly decreased under punishment prediction in PD patients.

Conclusions: The activity of the ventral striatum and amygdala to financial cues was reduced in untreated de novo PD patients. PD patients in the early stage might show the dual reduced brain activities, including the hypoactivity of the ventral striatum constituting the brain reward system and the dysfunction of the amygdala that may play a key role in loss aversion. In PD, the dysfunction in

the ventral striatum and amygdala may be present from the onset, and represent a behavioral endophenotype, prior to dopaminergic treatment.

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Abstract - WCN 2013

No: 909

Topic: 2 - Movement Disorders

Rapid eye movement sleep behavior disorder in patients with Parkinson's disease: Using rapid eye movement sleep behavior disorder screening questionnaire

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Background: Recent evidence suggests a strong link between rapid eye movement sleep behavior disorders (RBD) and Parkinson's disease (PD).

Objective: To evaluate the clinical features determining the co-morbidity of RBD and PD.

Patients and methods: The characteristics of nocturnal disturbances and other motor and non-motor features related to RBD in patients with PD and the impact of RBD on their quality of life were evaluated. Probable RBD (pRBD) was evaluated using the Japanese version of the RBD screening questionnaire (RBDSQ-J).

Results: A significantly increased frequency of pRBD (RBDSQ-J \geq 5) was observed in PD patients than in the controls (29.0% vs. 8.6%). After excluding restless legs syndrome and snorers in the PD patients, the pRBD group (RBDSQ-J \geq 5) showed higher scores compared with the non-pRBD group on the Parkinson's disease sleep scale-2 (PDSS-2) total score. Early morning dystonia was more frequent in the pRBD group. The Parkinson's disease questionnaire (PDQ-39) domain scores for cognition and emotional well-being were higher in the patients with pRBD than in the patients without pRBD. There were no differences between these two groups with respect to the clinical subtype, disease severity or motor function. The stepwise linear regression analysis showed that the PDSS-2 domain “motor symptoms at night”, particularly the PDSS sub-item 6 “distressing dreams”, was the only predictor of RBDSQ-J in PD.

Conclusion: Our results indicate a significant impact of RBD co-morbidity on nocturnal disturbances and quality of life in PD, particularly on cognition and emotional well-being.

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Abstract—WCN 2013

No: 931

Topic: 2—Movement Disorders

Effect of dopaminergic drugs on neuronal activity of substantia nigra in patients with Parkinson disease

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Background: Our previous ¹H-MRS study showed the decrease of SNc NAA/Cr in PD patients. It was correlated with age severity and duration. The decrease of SNc NAA/Cr in de novo PD seemed to become more prominent with the increase of disease duration compared to medicated cases.

Objective: To evaluate the effect of levodopa and dopaminergic medication on neuronal activity of SNc in PD patients.

Patients and methods: 204 PD patients and 150 PD patients treated with levodopa and dopaminergic agents were investigated between 2007 and 2012. Patients were further classified into 2 groups, levodopa alone and plus group. Multi-voxel $^1\text{H-MRS}$ examination of SNc was performed (1.5 T, TR/TE = 1000/144 ms). Single voxel examination was performed which placed ROI on the posterior half of the putamen (TR/TE = 1500/144 ms). NAA/Cr ratio was calculated and the difference was analyzed using Student's t-test.

Results: The NAA/Cr ratio of SNc between de novo and treated with levodopa alone PD showed significant differences of 1.63 and 1.43 ($p < 0.02$), respectively.

Conclusion: Levodopa seemed to restrain SNc neuronal activity. The phenomenon may be only a manifestation of the effect of age and disease severity, however, another explanation should be in consideration. The decrease of SNc NAA/Cr with the increase of NAA/Cr of the putamen in levodopa treated patients and the associated increase of NAA/Cr of the SNc and putamen in de novo PD patients suggest that the putamen receives enough dopamine derived from levodopa in the former and the putamen receives insufficient dopamine (denervated state) to promote SNc neuronal activity in the latter.

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Abstract—WCN 2013

No: 936

Topic: 2—Movement Disorders

Clinical correlates of serum insulin-like growth factor 1 in patients with Parkinson's disease, multiple system atrophy and progressive supranuclear palsy

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Background: Recently, increased serum insulin-like growth factor 1 (IGF-1) levels have been reported in patients with Parkinson's disease (PD) and multiple system atrophy (MSA) in small cohort studies; however, the clinical significance of serum IGF-1 in these disorders has not been fully clarified.

Objective: To assess a correlation between serum IGF-1 levels and clinical background factors in PD and related disorders and to investigate whether serum IGF-1 levels can be useful in differentiation of PD from MSA and PSP.

Patients and methods: A total of 67 patients with PD, 24 MSA and 14 PSP, and 49 healthy controls were included in this study. Serum IGF-1 and growth hormone (GH) were measured in the fasted state.

Results: Serum IGF-1 levels were significantly increased in MSA patients when compared with PD patients and controls, after adjusting age, gender and body mass index. No significant differences were observed in serum GH levels among patients and controls. In patients with PD, serum IGF-1 levels negatively correlated with disease severity and motor function. In contrast, patients with MSA showed an inverse correlation of serum IGF-1 levels with motor function and disease duration. There was a negative correlation between serum IGF-1 levels and motor function in patients with PSP.

Conclusion: The difference in the change and correlation of serum IGF-1 with clinical variables between these disorders may reflect different ongoing disease processes in each disorder.

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Abstract—WCN 2013

No: 940

Topic: 2—Movement Disorders

A semi-quantified evaluation of substantia nigra hyperechogenicity in Parkinson's disease and Parkinsonian syndrome

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Background: In Parkinson's disease (PD), transcranial sonography (TCS) has been used to evaluate substantia nigra (SN) hyperechogenicity as a diagnostic tool.

Objective: To quantify SN hyperechogenicity in patients with PD and Parkinsonian syndrome (PS), we applied semi-quantified evaluation method.

Patients and methods: Hospitalized patients with PD ($n = 28$; age, 65.0 ± 10.5 years) and patients with Parkinsonian syndrome (PS) ($n = 17$; age, 68.8 ± 9.6 years) and in-hospital controls ($n = 10$; age, 63.3 ± 8.7 years) were included. The PS group consisted of patients with progressive supranuclear palsy ($n = 7$) and multiple system atrophy ($n = 10$). TCS was performed using a conventional transcranial Doppler sonography equipped with 2.5 MHz transducer. The SN was identified within midbrain, and then the area of echogenic signals was circled and measured according to Berg et al. Next, echogenic signal of SN and dorsal midbrain were converted into grayscale using Adobe Photoshop and a median value of SN and dorsal midbrain on histogram was obtained. The SN to dorsal midbrain ratio was calculated.

Results: The PD group (4.1 ± 3.7) showed increased SN to dorsal midbrain ratio compared with that in PS group (1.7 ± 0.9) and controls (1.8 ± 1.1). The area of SN hyperechogenicity was larger in PD group ($0.19 \pm 0.11 \text{ cm}^2$) than in PS group ($0.07 \pm 0.07 \text{ cm}^2$) and controls ($0.05 \pm 0.07 \text{ cm}^2$).

Conclusion: Semi-quantified evaluation of SN echogenicity using SN to dorsal midbrain ratio is comparative to the previously reported method by Berg et al., measurement of the SN hyperechogenic area, and it may be useful in differential diagnosis of PD from PS.

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Abstract—WCN 2013

No: 944

Topic: 2—Movement Disorders

High resolution optical coherence tomography of the retina demonstrates selective thinning in individual retinal layers in Parkinson's disease

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Background: Parkinson's disease (PD) involves alteration of synaptic activity including dopaminergic neurons in the retina. Recent reports showed a decrease in retinal thickness (RT) and retinal nerve fibre layer (RNFL) peripapillary thickness as well as RT asymmetry measured by optical coherence tomography (OCT). However, individual retinal layers have not been analysed.

Objective: To investigate optic nerve (ON) and macular morphology in PD.

Methods: High-resolution spectral domain OCT was used to image 10 patients with PD and 10 healthy controls. RT and macular and

peripapillary RNFL thickness were measured in inner and outer annuli divided into quadrants. Retinal layers in the macular region were measured individually using ImageJ.

Results: RNFL was significantly thinner in PD compared to controls ($p < 0.01$ for all quadrants). Mean peripapillary RNFL thickness positively correlated with RT in the nasal and superior segments ($p < 0.01$). In PD, ON had normal configuration including disc areas, cup and rim areas and volumes. RT was significantly reduced in PD in all segments ($p < 0.05$) mainly due to changes in the inner plexiform layer (IPL, $p = 0.001$), outer plexiform layer (OPL, $p = 0.014$), outer nuclear layer (ONL, $p = 0.016$) and retinal pigment epithelium (RPE, $p < 0.01$). Duration of disease correlated with the thickness of IPL ($p = 0.003$). There were no inter-eye asymmetries in the ON and macula.

Conclusion: Our study shows for the first time that RNFL, IPL, OPL and RPE are the main affected structures in PD retina. This indicates that retinal changes are selective and may serve as biomarker to differentiate PD from other neurodegenerative disorders.

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Abstract–WCN 2013

No: 951

Topic: 2–Movement Disorders

A phase I single and repeated dose pharmacokinetic study of oral V81444, a selective non-xanthine adenosine A_{2A} receptor antagonist

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Background: V81444 is a selective non-xanthine adenosine A_{2A} receptor antagonist of potential utility in neurological disorders.

Objectives: This study evaluated its pharmacokinetics (PK) after first administration to man.

Methods: In a double-blind, randomised, placebo-controlled study, 18 healthy men in 2 alternating cohorts each received 2 single oral doses of V81444 and 1 of placebo. Doses were 30, 100, 200 and 300 mg. Thereafter, 31 healthy men received placebo or 50, 100 or 200 mg once daily orally, for 14 days. V81444 was assayed in plasma and urine, and PK parameters derived by non-compartmental analysis. Safety variables were also assessed.

Results: V81444 was absorbed promptly (lag time ≤ 0.5 h) after single doses. The maximum observed concentration was ≤ 4 h after dosing. V81444 exposure was less than proportional to dose (slope for area under curve (AUC) extrapolated to infinity: 0.88 [90% confidence interval: 0.81–0.95]). Geometric mean terminal elimination half-life was about 5–7 h (range: 3.29–11.4 h). After repeated dosing, steady-state was reached by Day 3 with little accumulation (accumulation ratio for AUC over the dose interval 1.19 [1.05–1.35]). The elimination half-life was somewhat longer (7–10 h). Less than 0.12% of the dose was excreted in urine. V81444 was well tolerated with no safety concerns at the doses studied.

Conclusion: Orally administered V81444 was rapidly absorbed with little drug accumulation at steady-state, a half-life compatible with twice daily dosing, and minimal urinary excretion.

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Abstract–WCN 2013

No: 874

Topic: 2–Movement Disorders

Correlation of striatal dopamine transporter binding with substantia nigra cell counts

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Background: Dopamine transporter (DAT) imaging is widely used for the differential diagnosis of parkinsonism. However, only limited data is available on the relationship between striatal DAT binding and dopaminergic cell loss in the substantia nigra (SN).

Objective: We retrospectively analyzed SN cell counts in patients who had ante mortem undergone DAT single photon emission computerized tomography (SPECT).

Patients and methods: Pathological diagnoses included Parkinson's disease ($n = 1$), dementia with Lewy bodies ($n = 2$), multiple system atrophy ($n = 1$), corticobasal degeneration ($n = 2$), atypical parkinsonism with multiple pathology ($n = 1$), Alzheimer's disease (AD, $n = 1$) and Creutzfeldt Jacob disease (CJD, $n = 1$). DAT SPECT with [¹²³I]β-CIT was performed using a standardized protocol on the same triple-head gamma camera. The density of neuromelanin-containing SN neurons/mm² was evaluated by morphometric methods using paraffin-embedded tissue sections.

Results: Mean disease duration at the time of DAT imaging was 2.3 years, followed by a mean interval of 29.3 months (range: 4–68 months) until death. Visual analysis of DAT images showed reduced striatal uptake in all 7 patients with neurodegenerative parkinsonism, but not in AD and CJD cases. Averaged (left + right / 2) striatal uptake was highly correlated with averaged SN cell counts (Spearman's rho 0.98; $p < .0005$). Similar high correlations were found in separate analyses for the right and left nigrostriatal system.

Conclusion: The present study shows a high correlation of striatal DAT binding in vivo with post mortem SN cell counts and confirms the validity of DAT imaging as an excellent in-vivo marker of nigrostriatal dopaminergic degeneration.

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Abstract–WCN 2013

No: 853

Topic: 2–Movement Disorders

Impaired cognitive performance and Hippocampal atrophy in Parkinson's disease

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Introduction: Since MRI has been used, hippocampal atrophy has been shown in demented and non-demented patients with PD. We aimed to assess the level of cognitive decline and the severity of hippocampal atrophy by measuring bilateral hippocampal volume in patients with PD.

Methods: Thirty three patients with idiopathic PD and 16 healthy subjects were enrolled to our study. All of these patients were under follow-up in movement disorders outpatient clinic, Department of Neurology, Uludağ University School of Medicine. Cognitive functions were tested with mini-mental state examination (MSE). The patients were classified as having normal cognitive function (PD-NC) or mild cognitive impairment (PD-MCI). Glucose, hemoglobin, TSH, total cholesterol, homocystein, vitamin B12, and folic acid levels were tested. Absence of depression was confirmed by Beck depression test. The cases were also evaluated with modified Wechsler memory scale, Luria draw test, Paven's test (RSPM), Stroop test, RUFF shape fluency test and KAS word fluency test, Trails A and B, forward and backward number repeating tests. Hippocampal volumes were calculated using Leonardo workstation.

Results: Mean ages of the PD-NC, PD-MCI, and healthy groups were similar. There was no difference between the hippocampal volumes of the groups. Right and left hippocampal volume was negatively correlated with repeats in word fluency test in healthy subjects. Right and left hippocampal volumes were positively correlated with cognitive functions in patients with PD-NC. Higher right hippocampal volume was associated with better performance in forward number test in patients with PD-MCI.

Conclusion: The findings of our study indicate that hippocampal atrophy is associated with cognitive decline in patients with PD.

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Abstract—WCN 2013

No: 854

Topic: 2—Movement Disorders

REM sleep behavior disorder and motor dysfunction in Parkinson's Disease—A longitudinal study

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Objectives: Longitudinal assessment of a Parkinson's Disease (PD) cohort, to investigate the evolution or REM sleep behaviour symptoms (RBD) over time and to test the relation between RBD at onset and motor and cognitive dysfunction progression.

Methods: An early stage, nondemented PD cohort (n = 61) was assessed at two time points, separated by a two year interval. Motor function assessment: Unified Parkinson's Disease Scale parts II and III (total and partial scores for bradykinesia, rigidity, gait/postural instability and dysarthria). Cognitive assessment: Frontal Assessment Battery (FAB) and Mini-Mental State Examination (MMSE). Dementia was diagnosed according to the DSM-IV-R criteria. RBD was defined as: presence of minimal clinical criteria for RBD and at least six affirmative answers in the REM Sleep Behaviour Disorder Screening Questionnaire.

Results: 25 patients had RBD at baseline, vs. 35 at follow-up. Three RBD patients did not present symptoms at follow-up, while 10 non-RBD patients developed RBD at follow-up (annual incidence of 12.5%). RBD and non-RBD patients did not differ significantly at baseline and follow-up. RBD was neither related to FAB or MMSE variation nor to dementia. The presence of RBD at onset was significantly related to an UPDRS total and bradykinesia scores increase over time.

Discussion: RBD symptoms can vary over time and have a tendency to increase during the early stages of disease. The presence of probable RBD could be a risk factor for motor function deterioration and particularly for bradykinesia worsening.

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Abstract—WCN 2013

No: 620

Topic: 2—Movement Disorders

Is live face-to-face UPDRS part III training more effective than remote training methodology in Parkinson's disease clinical trials?

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Background: Training and certification of raters' performance on the UPDRS part III in Parkinson's disease (PD) clinical trials are difficult. It is essential to implement a meticulously structured training/

certification programme prior to commencing such studies, to ensure reliability and consistency of performance.

Objective: In providing rigorous training to raters we analysed training data from two Phase III PD trials, to assess whether the method of training delivery (live versus remote) affected overall rating concordance.

Methods: Rater training consisted of didactic and interactive UPDRS presentations. Videotaped patient vignettes were scored and feedback provided. The chi-square test was used for testing differences of discrepancy by certification status. The effect of training delivery on rating agreement was examined by Fisher's exact test.

Results: A total of 617 potential raters submitted scores, of which 489 qualified to rate based upon initial score.

Raters initially qualified on scoring reported significantly more experience with the UPDRS scale, with PD patients and clinical trials than initially questionable raters.

Raters were trained either live at an Investigators' Meeting (IM) or in-study via online training systems. A significantly higher certification rate among IM-trained raters was seen (p = 0.012). Regional stratification demonstrated either a trend towards, or obvious, statistical significance favouring face-to-face training in most areas studied.

Conclusions:

- Experience levels of investigators impact significantly on initial certification status.
- Live training resulted in an overall significantly higher certification rate versus remote training.
- Differences were largely driven by regional variation, necessitating ongoing investigation into factors influencing these disparities.

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Abstract—WCN 2013

No: 876

Topic: 2—Movement Disorders

Essential tremor as a predictor of Parkinson's Disease

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Background: The aim is to present patients with long term of essential tremor, who later developed motor and nonmotor signs of Parkinson's Disease.

Objective: Essential tremor can precede as the only condition in the period of 5–6 years before development of motor and nonmotor signs of Parkinson's Disease. Persistence of the long term essential tremor can be predictor of the later development of Parkinson's Disease.

Material and methods: We examined 28 patients with an essential tremor, 21 women and 7 men, at the age between 55–62 years in the period of 5 years. The patients were hospitalized at the Movement Disorders Department at Neurology Clinic in Skopje, Republic of Macedonia and then were controlled on every 2 to 3 months. The following controls were consisted of detailed neurological examination. EEG, Evoked potentials, CT scan and MRI were also made.

Results: Two women (at the age 58 and 60 years) and one man (at the age 61 year) developed motor and nonmotor signs of Parkinson's Disease. On detailed neurological examinations two patients had rigidity and tremor and one has bradykinesia as well. All three patients had symptoms of depression, insomnia and sleeping disturbances as well. EEG was normal in ten patients and in two we notice nonspecific changes. One patients had prolonged VEP latency and SEP was normal in all patients. CT scan showed only slight cortical atrophy, such as MRI as well.

Conclusion: 12% of our patients with essential tremor developed Parkinson's Disease after 5 years of follow-up period.

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Abstract—WCN 2013

No: 886

Topic: 2—Movement Disorders

Investigation of cortical brain damage in patients with Machado–Joseph disease

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Background: Machado–Joseph disease (SCA3/MJD) is the most frequent spinocerebellar ataxia, and characterized by brainstem, basal ganglia and cerebellar damage. There are few MRI-based studies that investigate damage in the cerebral cortex.

Objective: To investigate damage to cerebral cortex and subcortical structures in SCA3/MJD through MRI-based volumetry of high resolution images.

Patients and methods: We included 44 patients with SCA3/MJD (mean age 49.0 ± 12.5 , 22 men) and 44 healthy controls (mean age 48.7 ± 12.3 , 22 men). Demographic/genetic data: CAG expansion (68.9 ± 5.1); SARA score (14.7 ± 7.2); Age at onset (40.5 ± 13.0); and disease duration (9.1 ± 4.0). All subjects underwent MRI scans in a 3T device, and 3D T1 images were used for volumetric analyses (slices of 1 mm, TE = 3.2 ms, TR = 7.1 ms, flip angle 8, isotropic voxels of 1 mm^3 , FOV = 240×240). Measurement of cortical thickness and volume was performed using FreeSurfer software v.5.1. We performed ANCOVA using subject's age as a covariate to compare groups and performed a GLM regression to assess correlations. In all analyses, we used an uncorrected $p = 0.001$.

Results: Group comparison showed reduction of cortical volume and thickness at left superior parietal, precentral and middle occipital gyri, as well as right paracentral sulcus and gyrus in SCA3/MJD. We also found volumetric reduction of cerebellar gray and white matter, thalamus, caudate, putamen, pallidum and ventral diencephalon. We then performed correlation analysis between clinical data and volume/thickness for those structures found to be atrophic in patients. Right pallidum volumes correlated with SARA scores ($r = -0.512$, $p = 0.001$).

Conclusion: SCA3/MJD patients have extensive subcortical and cortical damage. Basal ganglia damage is related to disease severity.

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Abstract—WCN 2013

No: 674

Topic: 2—Movement Disorders

Safety and efficacy of recombinant human platelet derived growth factor (Rhpdgf) in Parkinson's disease

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Objective: To evaluate safety, tolerability and exploratory efficacy of intracerebroventricular (ICV) administration of rhPDGF in patients with Parkinson's disease (PD).

Background: ICV infusion of rhPDGF significantly and long-lastingly reduces PD-like behavior and increases dopamine-transporter (DAT) binding in PD models. This effect is dependent on rhPDGF-induced stem/progenitor cell proliferation in the lateral ventricular wall. These findings have prompted a clinical study in patients with moderate/severe PD.

Methods: 12 PD patients were implanted with a drug pump and investigational catheter (Medtronic Inc.) leading into the lateral ventricle. Three dose cohorts (0.2, 1.5 or $5 \mu\text{g}$ rhPDGF/day) received either rhPDGF or placebo (buffer, 1 patient/cohort) for 12 days, after which all received buffer. Follow-up time was 85 days. Objectives included safety and tolerability assessment, and UPDRS, MADRS, MMT, EQ5-D and DAT-binding (PET).

Results: There were no unresolved adverse events related to the drug, infusion system or implant procedure. All patients improved in motor symptoms with no significant differences between dose cohorts. There were no significant therapeutic effects as assessed with MADRS, MMT or EQ5-D. Placebo patients displayed an expected reduction in DAT-binding over time. Patients in the highest dose group showed not only stabilization, but an increase in DAT-binding in regions of dopaminergic denervation with a maximum in the putamen ($P = 0.002$).

Conclusions: The ICV delivery of PDGF was safe and well tolerated and resulted in a significant dose-dependent positive effect on DAT-binding. The data support further clinical studies as PDGF may potentially slow down or reverse the nigrostriatal degeneration in PD.

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Abstract—WCN 2013

No: 858

Topic: 2—Movement Disorders

Brief assessment of cognitive decline in the early stages of Parkinson's Disease—A two year longitudinal study

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Introduction: Cognitive decline influences the outcome of Parkinson's Disease. Our objectives were to perform a longitudinal assessment of cognitive dysfunction in early stage patients, with brief neuropsychological tests.

Methods: Early stage, non-demented patients were assessed twice, over a 2 year interval, with the Frontal Assessment Battery (to detect frontal cognitive decline), the Mini-mental state examination (to detect global cognitive decline) and motor function scales. Dementia and hallucination were diagnosed according to DSM-IV criteria. We tested the relation between the clinical variables at baseline and the change in cognitive tests scores and the presence of dementia at follow-up.

Results: Of an initial 75 patients cohort, 61 were reassessed. Mini-Mental State scores did not progress significantly. Frontal Assessment Battery lexical fluency score decreased significantly. Four patients presented with dementia at t1. Mini-Mental State Examination score below cut-off, higher gait dysfunction, speech, rigidity scores, the non-tremor motor subtype and hallucinations were significantly related to dementia. Frontal dysfunction was related with a decrease in MMSE scores. Rigidity and speech dysfunction were related with a decrease in FAB scores.

Conclusions: Decline in Mini-Mental State Examination and FAB scores is small and heterogeneous in the early stages of Parkinson's Disease. Scores below cut-off in the Mini-Mental State Examination

Scores could be helpful to predict dementia. Non-tremor motor deficits could be predictive factors for frontal cognitive decline and dementia.

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Abstract—WCN 2013

No: 796

Topic: 2—Movement Disorders

Structural changes of raphe nuclei detected by brain parenchyma sonography and diffusion tensor imaging in Parkinson's disease associated with depression

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Objectives: Transcranial parenchymal sonography (TCS) showed decreased echogenicity of the brainstem raphe (BR) in depression in Parkinson's disease (PD). Magnetic resonance imaging and histopathological studies confirmed the hypothesis of a structural changes of the BR in depression in PD. We compared fractional anisotropy (FA) values and apparent diffusion coefficients (ADC) of the BR in depressed and non-depressed PD patients in comparison to TCS findings, to investigate possible structural changes of BR.

Methods: 53 PD patients with depression (PD + D+), 51 PD patients without depression (PD + D-) and 50 healthy individuals (PD - D-) were included. PD patients with and without depression were matched for age, disease stage and duration. Echogenicity of BR was rated using TCS with a two point scale (grade 1: normal BR echogenicity same as red nuclei, grade 0: hypoechogenic, invisible or interrupted BR). ADC and FA values were calculated using SPM7 software.

Results: Significant increase of mean ADC (t-test, each, $P < 0.0001$) and significant decrease of mean FA values (t-test, each, $P < 0.0001$) were found in patients with hypoechogenic BR compared with patients with normal raphe echogenicity. Mean FA values were significantly decreased in dorsal and ventral part of the BR in PD + D+ patients but only in ventral part of the BR in PD + without depression in comparison with healthy subjects (t-test, each, $P < 0.0001$).

Conclusion: Our findings confirm structural changes of the BR in depression associated with PD which may reflect the pathogenic role of the basal limbic system and its projections in the pathogenesis of depression in PD.

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Abstract—WCN 2013

No: 816

Topic: 2—Movement Disorders

Carpal tunnel syndrome in patients with Parkinson's Disease: An electrodiagnostic and ultrasonographic study

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Introduction: Tremor is one of the cardinal symptoms of Parkinson's Disease (PD) and may cause cumulative traumatic injury to nerves of the hands. Carpal tunnel syndrome, in particular, may be caused by median nerve trauma. The aim of this study is to analyze the electrodiagnostic and ultrasonographic results of patients with PD and to verify the effect of hand movement in PD on the median nerve.

Methods: We recruited 31 hands of healthy controls (60.25 ± 14.67 years) and 81 hands of patients with PD (64.95 ± 11.13 years). The patients with PD were divided by Hoehn and Yahr (H&Y) clinical stage into two groups: mild and advanced. Clinical symptoms of patients with PD were measured by Unified Parkinson's Disease Rating Scale (UPDRS), and quality of life was measured by Parkinson's Disease Questionnaire. Median nerve conduction and cross-sectional area by ultrasonography were also evaluated.

Results: Eighteen patients were classified in the mild group and 23 in the advanced group by H&Y stage. Median nerve cross-sectional area in patients with PD was significantly larger than that of the control group. Electrophysiologic findings of the median nerve were not significantly different between patients with PD and the control group. Total UPDRS score was not significantly correlated with cross-sectional area of the median nerve, but the severity of tremor was significantly correlated with cross-sectional area of the median nerve.

Conclusions: Carpal tunnel syndrome diagnosed by ultrasound was frequent in PD, and tremor may be a risk factor for the development of median nerve pathology.

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Abstract—WCN 2013

No: 821

Topic: 2—Movement Disorders

An electrophysiological and kinesiological method for assessing spasticity

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This study defines the electrophysiological characteristics of Patellar T reflex. Normal and spasticity changes by utilizing movement features of knee joint during the reflex and it also attempts to develop a measurement method to be used in spasticity.

Normal and spasticity behaviors were examined with the electrophysiological records obtained from m. Quadriceps femoris and m. biceps femoris muscles. Medium and late latency responses in relevant muscles were examined with kinesiological parameter in subgroups in which the spasticity of patients was evaluated in accordance with clinical Ashworth scale.

In the normal group, angular velocity was 57.95 ± 16.64 °/s. Latency of Patellar T reflex was 17.93 ± 1.94 ms and its amplitude was 6404 ± 2859 μV. In cerebral spasticity group, angular velocity was 61.32 ± 20.61 °/s. Latency of Patellar T reflex was 17.6 ± 2.45 ms and its amplitude was 7551 ± 3939 μV. While BF-MLR could not be obtained in normal cases, it was established to be 54.73 ± 10.18 ms latency in cerebral spastics.

It seems possible to produce a parameter that would assess and rate spasticity. Electrophysiological and kinesiological parameters failed to separately differentiate normal and spastics in compliance with Ashworth, but they could manage to differentiate between normal and spastic cases together. The parameter that was 69.61 ± 32.56 in normal cases decreased to 50.52 ± 28.98 in Ashworth 1, to 32.00 ± 14.90 in Ashworth 2, and finally to 14.48 ± 22.83 in Ashworth 3, and these changes was statistically significant.

Consequently, it is possible to note that the pendulum of patellar T reflex could differentiate between spasticity and normal by electrophysiological and kinesiological evaluation.

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Abstract—WCN 2013**No: 729****Topic: 2—Movement Disorders****Parkinson's disease Sleep Scale-2 is more specific for PD than the Epworth Sleep Scale**

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Objectives: To compare the specificity and sensitivity of Parkinson's disease Sleep Scale-2 (PDSS-2) and Epworth Sleep Scale (ESS) in Parkinson's disease (PD).

Background: Sleep problems are one of the most burdensome non-motor features of PD.

Methods: 60 patients fulfilling the UK Brain Bank criteria for PD were enrolled to the study. Each patient kept a patient diary for at least 7 days. Besides, ESS, PDSS-2, Hoehn–Yahr and UPDRS were obtained to describe the sleep-related problems.

Results: Severity of sleep problems was quantified by the total score of PDSS-2 and ESS. Out of the two sleep scales, only PDSS-2 demonstrated significant correlation with the sleep hours measured by the patient diary ($r = 0.61$, $p < 0.01$), UPDRS part 2 and HYS. The correlation between PDSS-2 and ESS was not significant ($r = 0.12$, $p > 0.05$).

Conclusions: For quantifying sleep disturbances in PD, the PDSS-2 seems to be more specific than ESS.

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Abstract—WCN 2013**No: 730****Topic: 2—Movement Disorders****Comparison of double monopolar and interleaving stimulation modes in the treatment of primary generalized and segmental dystonia**

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Objective: To systematically compare the efficacy and side-effect profile of double monopolar stimulation mode to those of interleaving stimulation mode in a prospective, randomized, double-blind and cross-over study.

Background: For the treatment of drug-refractory dystonia, bilateral pallidal deep brain stimulation (GPi-DBS) is proven to be an efficient option. On average, 40–55% improvement on dystonia rating scales (DRS) could be achieved according to the results of multicenter trials lasting for years. However, a considerable portion (10–25%) of patients experience minimal alleviation despite of good electrode placement. These patients can be regarded as non-responders to GPi-DBS defined as having either limited improvement (less than 25% on DRS) or worsening.

Methods: This randomized, cross-over, double-blind study was initiated in 2012 and is expected to be completed by 2015. An estimation of 20 patients with primary generalized or segmental dystonia is planned to be enrolled.

Design: Double-blind, cross-over.

Results: The interim analysis of 9 patients is presented. Average age was 33.5 ± 16.2 years. The size of improvement during the double monopolar phase was $43.7 \pm 9.3\%$, whereas in the interleaving phase it was $55.6 \pm 7.5\%$ ($p = 0.13$).

Conclusions: Although our preliminary findings are promising, more data is required to reach the statistical power and draw the conclusion that interleaving stimulation might be superior to double monopolar stimulation in the treatment of primary dystonia.

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Abstract—WCN 2013**No: 686****Topic: 2—Movement Disorders****Can repetitive transcranial magnetic stimulation (RTMS) help on-freezers with advanced PD?**

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Background and objectives: Freezing of gait (FOG) or “motor block” is experienced in about 30% of PD patients within 5 years, and nearly in 60% after 10 years. Treatment of “off” time FOG is relatively straight forward, but “on-freezing” is a difficult to treat scenario. This issue is rarely addressed in RCTs to assess investigational tools or treatment options.

Methods: This was a randomized, double-blinded, placebo-control study evaluated the efficacy and safety of rTMS in management of “on-freezing” in patients with advanced PD. Twenty two Egyptian patients received 12 rTMS sessions over 4 weeks (either real or sham) using figure-of-8-shaped coil over the leg area of motor cortex contralaterally to the more affected side in addition to rehabilitation program involving specific gait training techniques. Primary efficacy variables are FOG Q (SF), motor section and total score of UPDRS, 2ry outcomes are gait variables (Cadence, number of falls, stride length, stride time and turn time), and on-time.

Results: Improving of FOG Q (SF) ($P < 0.05$), together with significant decrease in number of falls and widened stride length ($P < 0.001$) was detected in patients receiving real rTMS. The total score of UPDRS and other gait variables was not significantly changed. No adverse events were recorded apart from mild and transient headaches.

Conclusions: These results indicate that rTMS has a positive effect in on-freezers with advanced PD with subsequent decrease of number of falls.

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Abstract—WCN 2013**No: 664****Topic: 2—Movement Disorders****fMRI, DTI, and MRS for characteristics of the level of Cognitive Impairment (CI) in patients with Parkinson's Disease (PD)**

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Background: Bringing together fMRI, DTI-, and MRS-data in substantia nigra (SN) we obtain biomarkers of neuronal dysfunction in patients with PD and different levels of CI.

Objective: We found regional specificity MRI-data for characteristics of CI.

Patients and methods: Three groups of PD-patients with various cognitive statuses are studied with 1.5T SIGNA (GE). The 1st group (DPDG)-13 PD-patients with dementia (MMSE < 25), 2nd group (CIPDG)-23 with mild CI ($30 > \text{MMSE} \geq 25$), 3rd group (NPDG)-18 PD-patients with normal cognitive function (MMSE > 30).

Materials and methods: fMRI-data are obtained by: TR/TE = 3000/60 ms. DTI-data (25dir) are obtained in anterior, medial, posterior SN (ASN, MSN, PSN).

Spectra are recorded with SVSSTEAM:TR/TE = 1500/144 ms.

Results: From analysis of fMRI (FSL5.0 FEAT, MELODIC) we found that in the NPDG connections between the APCG and PPCG, and inferior parietal gyrus bilaterally exist. In the NPDG activation of anterior (APCG), and of the posterior portion of cingulate gyrus (PPCG) decreased. In CIPDG activated clusters were found precuneally, and in PPCG, no connection to the parietal lobe or APCG. In DPDG no activation in PPCG was found. In anterior part of SN (APSN) NAA/Cr in DPDG, CIPDG, and NPDG: (1.68 ± 0.02), (2.04 ± 0.03), (2.32 ± 0.05), Cho/Cr: (0.84 ± 0.02), (0.81 ± 0.05), (0.53 ± 0.03). In posterior part of SN (PPSN) NAA/Cr in DPDG, CIPDG, and NPDG: (1.14 ± 0.12), (1.81 ± 0.02), (1.98 ± 0.04), Cho/Cr: (0.96 ± 0.02), (0.77 ± 0.03), (0.68 ± 0.03). We have found the progressive decreasing NAA/Cr in the PPSN and increasing of Cho/Cr for the patient of NPDG, CIPDG, and DPDG, that is associated with poorer cognitive function. MD in DPDG: (0.82 ± 0.05) $\times 10^{-3}$ mm²/s, in CIPDG: (0.74 ± 0.05) $\times 10^{-3}$ mm²/s, in NPDG: (0.71 ± 0.05) $\times 10^{-3}$ mm²/s. FA: (0.41 ± 0.05), (0.43 ± 0.05), (0.47 ± 0.05) in DPDG, CIPDG, and NPDG.

Conclusion: These data give us new approach for understanding pathophysiological changes in PD-patients associated with CI.

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Abstract—WCN 2013

No: 649

Topic: 2—Movement Disorders

Clinical variants of vascular parkinsonism

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Background: 1–15% cases of parkinsonism are caused by cerebral vessels' lesions.

Objective: To investigate the ethiological factors and clinical features of vascular parkinsonism.

Materials and methods: 100 patients with vascular parkinsonism were examined (males—80%, females—20%). The main part of patients was over 60 years old (65%). Diagnosis was determined on the basis of anamnesis data and the results of clinical, neurological, neurophysiological and neuroimaging tests.

Results: The following clinical forms were defined: extrapyramidal–pyramidal (41%), extrapyramidal–cerebellar (25%), extrapyramidal–dement (18%), hemi–extrapyramidal (6%), parkinsonism of lower part of body (10%). 68.7% of patients had the acute cerebral ischemic event at the onset of the disease, and 31.2% of patients developed parkinsonism as a result of chronic cerebral ischemia, caused by arterial hypertension and atherosclerosis. The movement impairment was accompanied by the depressive mental disorders in 40% of cases. Other nonspecific disorders (memory loss (26%), headache (24%), dizziness (21%), sleep disruptions (15%)) were present. Cerebral CT/MRI revealed the low density cortical and subcortical spots. The multiple cerebral infarctions were found in 22% of patients, solitary lacunar infarctions—in 14%, hemorrhage—in 6%, brain atrophy with substitutional hydrocephalus, periventricular and subcortical leukoaraiosis—in 58 % patients. Levodopa medications were of low effectiveness.

Conclusion: The main reason of vascular parkinsonism is cerebral atherosclerosis combined with arterial hypertension (68%). The clinical variant of vascular parkinsonism-plus (66%) was predominant.

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Abstract — WCN 2013

No: 640

Topic: 2 — Movement Disorders

d-Penicillamine versus zinc sulfate as first-line therapy for Wilson's disease

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Background: Wilson's disease (WD) is a treatable inherited copper metabolism disorder. However, there is no international consensus about the first-line therapy.

Objective: Our aim was to compare the course of treatment in patients with symptomatic WD that started d-penicillamine (DPA) or zinc sulfate (ZS) as a drug of choice.

Patients and methods: We included 143 consecutive, new diagnosed patients with symptomatic WD between 2005 and 2009. The decision about the first-line therapy drug was made at the physician's discretion. Data were analyzed in subgroups with predominantly neuropsychiatric and hepatic WD.

Results: Neurological and enzymatic improvements were achieved with similar frequency. In patients with neuropsychiatric WD, the probability of remaining on first-line therapy was similar for DPA and ZS (20% vs. 24% at the end of follow-up). In patients with hepatic WD, it was significantly higher for ZS (31% vs. 12%). After adjusting for type of WD, sex, age at diagnosis, and the presence of at least severe hepatic symptoms, patients treated with DPA were more likely to experience worsening (OR 3.84, 95% CI: 1.15–3.85) during the first 6 months of treatment. Patients on DPA were significantly more compliant (94% vs. 81%), but experienced more adverse effects (15% vs. 3%).

Conclusions: DPA and ZS are effective in the majority of WD patients. Despite certain differences, none of them appears clearly superior. Therefore, ZS may be considered a reasonable alternative to DPA as a first-line therapy.

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Abstract — WCN 2013

No: 582

Actigraphic study of gait disturbance in patients with Parkinson's disease

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Objectives: We performed actigraphy to quantitatively analyze gait disturbances in Parkinson's disease (PD).

Subjects: The study group comprised 11 healthy subjects (controls) (mean age, 74.3 years) and 34 PD patients (74.4 years; severity on Yahr stage scale, 2.2; disease duration, 8.0 years). The patients were divided into the following three groups: Group I (GI), almost normal gait; GII, mild gait disturbance; and GIII, apparent gait disturbance (including frozen gait and gait disturbance due to wearing off).

Methods: An actigraph was attached to the ankle to evaluate motor count (MC) and its coefficient of variation (CV) during usual gait and stamping for 60 s each. The mean MC per 5-second interval was calculated for each 60-second test. MC and CV were compared between controls and PD. Student's t-test was used for statistical analysis.

Results: 1) In controls, MC was 28.9 times during usual gait and 28.2 times during stamping. CV was 5.2% during usual gait and 8.3%

during stamping. 2) In PD during usual gait, MC was 29.0 times (GI, 28.6; GII, 29.1; GIII, 29.5) and CV was 9.3% (GI, 7.1%; GII, 9.0%; GIII, 11.9%). 3) In PD during stamping, MC was 30.1 times (GI, 30.6; GII, 30.9; GIII, 29.1) and CV was 11.8% (GI, 7.5%; GII, 10.8%; GIII, 17.2%). 4) MC did not differ significantly between controls and PD. In contrast, CV differed significantly between controls and PD during usual gait and stamping, respectively.

Conclusion: Actigraphy is useful for quantitatively evaluating gait disturbances in PD patients.

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Abstract – WCN 2013

No: 583

Topic: 2 – Movement Disorders

Impulsive compulsive behaviors in Japanese Parkinson's disease patients and utility of the Japanese version of the Questionnaire for Impulsive–Compulsive Disorders

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Background: Recently, non-motor syndrome in Parkinson's disease (PD) patients has attracted attention. Impulsive compulsive behaviors (ICBs), such as pathological gambling, compulsive sexual behavior, compulsive buying, compulsive eating, punding, and dopamine dysregulation syndrome (DDS) are one of the non-motor syndromes in PD, but they aren't very well identified. Whereas the prevalence and the relative factors of ICBs were estimated in previous reports, it hasn't fully understood.

Objective: In order to evaluate ICBs in Japanese PD patients, we constructed a Japanese version of the Questionnaire for Impulsive–Compulsive Disorders in Parkinson's disease (J-QUIP) and evaluated the utility of J-QUIP in Japanese PD patients.

Methods: J-QUIP was administered to 121 PD patients. Diagnoses of ICBs were made via interview of patients or their caregivers. Subsequently, in order to evaluate factors related to these conditions, we evaluated demographic and clinical characteristics, clinical features, and medications utilized.

Results: J-QUIP was able to accurately evaluate symptoms in 118 of 121 PD patients (97.5%). Sensitivity and specificity of J-QUIP were similar to that reported for the original version of QUIP. In our study, the prevalence of each disorder was as follows: pathological gambling (6.5%), compulsive sexual behavior (3.2%), compulsive buying (3.2%), compulsive eating (3.2%), punding (6.5%), DDS (2.2%). Significantly associated factors for these conditions were younger age ($p = 0.047$), earlier age of disease onset ($p = 0.015$), longer PD duration ($p = 0.001$), total levodopa equivalent dose ($p = 0.006$), and dosage of levodopa ($p = 0.019$).

Conclusion: We evaluated the prevalence of ICBs in Japanese PD patients along with factors associated with these behaviors via J-QUIP.

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Abstract – WCN 2013

No: 512

Topic: 2 – Movement Disorders

Randomized double-blind, placebo-controlled multi-center trial on molecular hydrogen water in Parkinson disease

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Background: Our previous randomized double-blind study showed that drinking 1 L of hydrogen (H₂) water for 48 weeks significantly improved ($p < 0.05$) the total Unified Parkinson disease rating scale (UPDRS) score of patients with Parkinson's disease (PD) receiving l-dopa. We aim to confirm these results by conducting a longer and more powered trial, which also includes patients not receiving l-dopa. Here, we present the design rationale and describe the study cohort for this trial.

Methods: The change in total UPDRS score (I–IV) from baseline to the 72th week will be used as the primary endpoint. The other analyzed parameters will be as follows: the change of total UPDRS and part II, III, each scores, and modified Hoehn and Yahr staging from baseline to 8th, 24th, 48th, and 72th weeks and post the 8th week; the change of total PDQ39, each scores, and subtotal scores; and the duration till the protocol was terminated because of addition of levodopa or disease progression. Safety analyses will include tests for adverse events, except dyskinesia, and laboratory examinations. Patients in the hydrogen water group will drink 1 L of H₂ water made by “Suisosui 5.0” and supplied by Ecomo International Co., Ltd., and those in the placebo water group will drink water filled with nitrogen. This study started in March 2013, and we will register 200 patients with PD from 15 centers.

Discussion: This study will confirm whether H₂ water can modify PD progression.

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Abstract – WCN 2013

No: 506

Topic: 2 – Movement Disorders

Degeneration of the cerebellum and brainstem in Huntington's disease (HD)

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Background: Huntington's disease (HD) is a progressive polyglutamine disease which is characterized neuropathologically by severe neuronal loss in the striatum and select layers of the neo- and allocortex. The cerebellum and brainstem are among the brain sites whose neuropathological state in and relevance for the clinical picture of HD is still controversial.

Objective: To analyze the pathoanatomy of the cerebellum and brainstem in HD patients.

Patients and methods: Serial thick tissue sections through the cerebellum and brainstem of eight clinically diagnosed and genetically confirmed HD patients and twelve control individuals underwent pigment–Nissl staining for neuronal lipofuscin pigment and Nissl material or were immunolabeled for the marker of cerebellar Purkinje cells (i.e. calbindin).

Results: Our study revealed a consistent neurodegeneration of the cerebellar Purkinje cell layer and cerebellar fastigial, globose, emboliform and dentate nuclei. An additional neuronal loss was present in the following brainstem nuclei: substantia nigra, pontine nuclei, reticulotegmental nucleus of the pons, superior and inferior olives, area of the excitatory burst neurons for horizontal saccades, raphe interpositus nucleus, and vestibular nuclei.

Conclusions: HD is associated with a widespread neurodegeneration of the cerebellum and brainstem and represents a multisystem neurodegenerative disease. Damage to the cerebellum and brainstem contributes to poorly understood HD disease symptoms (i.e. impaired rapid alternating movements, dysarthria, ataxia and postural instability, gait and stance imbalance, broad-based gait and stance, oculomotor dysfunctions).

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Abstract – WCN 2013

No: 494

Topic: 2 – Movement Disorders

Fatigue syndrome and white matter changes in patients with Parkinson's disease: Does link exist?

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Background: White matter changes (WMC), caused by concomitant cerebrovascular disease (CVD), appear on MRI in part of elderly patients with Parkinson's disease (PD). However clinical significance of WMC in genesis and severity of non-motor symptoms, including fatigue syndrome (FS), remain unclear.

Objective: To define association between FS and WMC in patients with PD.

Materials and methods: 20 PD patients with FS were compared with 18 PD patients without it. Groups were compared for presence of WMC. Following methods were applied: UPDRS, PD's fatigue scale (PFS), Autonomic impairment scale (Levin O.S., 2003), and Fazekas Scale (FaS). The patients underwent brain 1.5-T MRI (Avanto Siemens; Siemens AG, Erlangen, Germany). Middle ages of the patients were 64.5 ± 6.5 and 63.9 ± 7.5 . PD's stages were 2.6 ± 0.5 and 2.45 ± 1.1 , disease's durations were 5.9 ± 4.04 and 4.4 ± 2.85 correspondingly.

Results: In the 1st group of PD patients with FS WMC were marked in 65% cases, in the control-in 47%. PFS scores were 3.88 ± 0.36 in the 1st group and 2.37 ± 0.6 in the control. FaS was 0.9 ± 0.7 in the 1st group, 0.47 ± 0.51 in the control. Autonomic disturbances in the 1st group were 7.1 ± 2.9 , in the control 4.2 ± 2.4 . Among them cardiovascular disturbances were 3.15 ± 1.59 in the 1st group, 1.15 ± 1.1 in the control. UPDRS scores were 84.4 ± 21.5 in the 1st group and 68.0 ± 40.3 in the control.

Conclusions: WMC and autonomic disturbances occur more often among PD patients with FS. This fact can imply that orthostatic hypotension influences both neuroimaging data and genesis of FS. Orthostatic hypotension can be accompanied by hypertension in supine position, which promotes WMC.

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Abstract – WCN 2013

No: 478

Topic: 2 – Movement Disorders

Levosulpiride induced common and uncommon movement disorders—Parkinsonism and truncal akathisia: Three case reports

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Background: Levosulpiride is a substituted benzamide that is widely used for the management of dyspepsia and emesis. Because it exerts its pharmacologic activity mainly by blocking dopaminergic D2 receptor activity, levosulpiride can cause extrapyramidal symptoms, most of which are generalized parkinsonism followed by lower face dyskinesia. We report three cases of levosulpiride induced common and uncommon movement disorders.

Objective: To highlight levosulpiride induced movement disorders especially rare one like akathisia.

Patients and methods: We report three patients of levosulpiride induced movement disorders seen in our centre during July 2010 to September 2012. Two were male and one female with average age of 75.33 years. Two of these patients presented with symmetric parkinsonism and one with truncal akathisia. The longest duration of levosulpiride exposure at a dose of 75 mg/day, was one year and shortest one week. The onset was sub-acute in two patients and acute in one patient.

Results: Two patients exhibited resolution of parkinsonism upon cessation of levosulpiride. The interval from cessation of levosulpiride to disappearance of parkinsonism was 10 days and 30 days. One patient who had truncal akathisia continued to have symptoms despite drug withdrawal.

Conclusion: Although generalised parkinsonism is most frequently reported adverse effect of levosulpiride as seen in our patients also, awareness of rare movement disorder like truncal akathisia is crucial for management.

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Abstract – WCN 2013

No: 450

Topic: 2 – Movement Disorders

Genetic deconvolution of cerebellar ataxias in India through next generation sequencing: Novel mutations in atypical and typical genetic loci

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Introduction: Genetic characterization has led to the identification of nearly 60 causal loci in ataxias. The prevalence of ataxia types with respective genetic defect varies geographically. The genetic testing of each known ataxic associated variation for uncharacterized phenotype is challenging and cost/time expensive.

Objective: In our ataxic cohort (n = 1000) majority being North-Indian families, more than half of the cases (~57%) are genetically uncharacterized (UC). We aimed to delineate genetic defect in uncharacterized cases using next generation sequencing platform.

Methodology: We carried out exome-sequencing for 12 individuals from three uncharacterized families of recessive inheritance manifesting typical FRDA or unique phenotype such as infantile onset ataxia with hearing loss or seizures.

Results: Clinical investigations and exome sequencing in one family reveal association of GAA expansion negative FRDA phenotype with novel homozygous frame-shift mutation in *SACS* (Autosomal recessive ataxia of Charlevoix-Saguenay) locus. In second family with infantile onset ataxia and hearing loss we identified a novel homozygous missense mutation in *c10orf2*. In remaining family with juvenile onset ataxia and generalized seizures we identified novel compound heterozygous mutations in *CLN6* (Ceroid-Lipofuscinosis Neuronal 6).

Conclusion: We have identified novel mutation loci in typical known recessive ataxic phenotype and compound novel heterozygous mutations in *CLN6* a known locus for seizures–dementia–visual failure being associated with atypical manifestation of predominant ataxia and seizures. Further, this study emphasizes the prudent role of exome sequencing technology in delineation of genetic etiology in a rare and other genetically heterogeneous ataxic disorders.

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Abstract – WCN 2013**No: 446****Topic: 2 – Movement Disorders****New treatment for clinical stabilization in Parkinson's Disease (Cervô): Results in 32 patients at 29 months of follow-up**

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It is mandatory to have a safe and effective treatment to stop the neurodegenerative disease's progression. Previous clinical trials designed to stop Parkinson's Disease (PD) progression have failed to demonstrate significant clinical stabilization. We administrated to PD patients the first oral treatment designed to stop the disease's progression. This treatment called Cervô, contains four substances that have effect in controlling the most important known mechanisms of disease progression as: aberrant apoptosis, oxidative damage, mitochondrial degeneration, caspases and Mitogen-Activated Protein-Kinases (MAPK) activation, among others. We had previously demonstrated that it is safe to use Cervô in humans. Before beginning the clinical trial, we administrated the best medical symptomatic therapy to each patient and they remained with the same prescription during the observation period. Then, we added Cervô twice per day to all the patients.

Results: We included 31 patients with PD. Age: 32 to 86 years old (mean 66.3 years, SD \pm 13.5), 16 female (51.6%), 15 male (48.4%). Initial United Parkinson's Disease Rating Scale (UPDRS) score: 1–15 (mean 5, SD \pm 3). Follow-up period: 12–57 months (mean 29 months, SD \pm 14.5).

Clinical evaluations: There was no increase in the UPDRS score in 30 patients (96.77%) in the follow-up period, and 25 (80.64%) patients improved their basal UPDRS score. The mean UPDRS scores were 3.5 at 12 months and 2.5 at 24 months.

Conclusions: Cervô is a new and promising medication, specifically designed to stop neurodegenerative disease's progression. This needs to be confirmed with a randomized multi-centric study.

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Abstract – WCN 2013**No: 427****Topic: 2 – Movement Disorders****REM sleep behavior disorder in Parkinson disease: Association with abnormal ocular motor findings**B.S. Jeon^a, Y.E. Kim^b, H.-J. Kim^a. ^aSeoul National University Hospital, Seoul, Republic of Korea; ^bSeoul National University Bundang Hospital, Seoul, Republic of Korea

REM sleep behavior disorder (RBD) is related with brainstem pathology. We examined whether patients with RBD have abnormal ocular movements suggesting brainstem or cerebellar dysfunction in Parkinson's disease (PD). A total 202 patients were included in this study. Ocular movements were examined by video-oculography (VOG). 116 (57.4%) of the 202 patients have clinically probable RBD, and 32 (27.6%) of the 116 with clinically probable RBD patients had abnormal VOG findings suggesting brainstem or cerebellar dysfunction; whereas 86 of the 202 patients did not have clinically probable RBD, and only 8 (9.3%) of 86 patients had abnormal VOG findings suggesting brainstem or cerebellar dysfunction. ($P = .001$). This study suggests that the presences of RBD are associated with more severe brainstem pathology in PD.

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Abstract – WCN 2013**No: 440****Topic: 2 – Movement Disorders****TIC disorders (TD) in children: The kindergarten matters**V. Sajin^{a,b}, I. Moldovanu^a. ^aInstitute of Neurology and Neurosurgery, Republic of Moldova; ^bState Medical and Pharmaceutical University 'Nicolae Testemitanu', Chisinau, Republic of Moldova

Background: Tics are sudden, rapid, recurrent, non-rhythmic motor movements or vocalizations (European clinical guidelines for Tourette syndrome and other tic disorders, 2011) with the prevalence in children of 0.77–1% (2.99% for transient tics) (Robertson, 2008; Knight et al., 2012).

The poor knowledge about TD leads to the delayed diagnosis and wrong management of TD. Kindergarten teachers (KTs) could contribute to a better behavioral and social integration of children with tics and their parents.

On our knowledge, no precedent informational campaign on TD was performed in Republic of Moldova (RM).

Aims: to assess the necessity to inform KT's about TD

Materials, subjects and methods:

- Video presentation about the TD
- Short questionnaires
- Twenty two kindergarten teachers (mean age 50.86 \pm 11.07 yrs) from 4 kindergartens from RM (549 children). Each teacher supervised about 20 children (2–3 yrs (84 children), 3–4 yrs (128), 4–5 yrs (29), 5–6 yrs (189) and 6–7 yrs (119) old).

Results: Before the presentation, the majority of KT's mistook tics for muscle fasciculations or hysterical reactions. Only two children with TD from 549 were initially reported.

After the presentation, 15 teachers gave a right description of tics, their evolution and educational particularities of children with TD.

Fifteen children with TD (2.73%), 11 boys and 4 girls (3:1) were reported by KT's.

Conclusion: The presented information improved the kindergarten teachers' knowledge about the tic disorders and their communicative and educational skills, as well as their attitude toward such children. KT's can contribute to the active detection and right management of TD.

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Abstract – WCN 2013**No: 384****Topic: 2 – Movement Disorders****Are branded and generic extended-release ropinirole formulations equally efficacious? A rater-blinded, cross-over, multicenter study**N. Kovács^{a,b}, E. Bosnyák^a, M. Herceg^c, E. Pál^a, Z. Aschermann^a, J. Janszky^b, I. Késmárki^d, K. Karádi^a, T. Dóczi^b, F. Nagy^c, S. Komoly^a. ^aDepartment of Neurology, University of Pécs, Hungary; ^bMTA-PTE Research Group, Hungary; ^cDepartment of Neurology, Kaposi Mór County Hospital, Kaposvár, Hungary; ^dDepartment of Neurology, Health Center of City of Pécs, Pécs, Hungary

Objectives: The aim of this rater-blinded multicenter study was to compare the efficacy of branded (Requip modutab®) and generic (Ralnea®) extended-release ropinirole formulations in the treatment of Parkinson's disease (PD).

Methods: Of 32 enrolled patients 29 completed the study. A rater blinded to treatment evaluated Unified Parkinson's Disease Rating

Scale (UPDRS), Fahn–Tolosa–Marin Tremor Rating Scale (FTMTRS), Non-motor Symptom Assessment Scale (NMSS), Montgomery–Asberg Depression Rating Scale (MADRS), and a structured questionnaire on ropinirole side-effects. Besides, the patients self-administered EQ-5D and Parkinson's Disease Sleep Scale 2nd version (PDSS-2).

Results: Branded and generic ropinirole usage achieved similar scores on tests measuring motor symptoms (UPDRS-III 23.5 ± 8.2 vs. 26.9 ± 9.1 points, UPDRS-total: 46.2 ± 16.0 vs. 50.4 ± 18.7 points and FTMTRS 24.2 ± 9.5 vs. 24.93 ± 10.4 points, respectively, $p > 0.05$). The results of questionnaires on the sleep-quality (PDSS-2 14.6 ± 10.5 vs. 14.5 ± 11.2 points, $p > 0.05$), non-motor symptoms (NMSS, 78.5 ± 49.5 vs. 82.3 ± 48.2 points, $p > 0.05$), quality of life (EQ-5D 0.72 ± 0.22 vs. 0.71 ± 0.21 , $p > 0.05$) and depression (MADRS: 11.1 ± 9.3 vs. 11.9 ± 9.2 , $p > 0.05$) were comparable. Except for gastrointestinal problems, both medications had comparable side-effect profile. After completion of the study, the subjects did not prefer neither of the formulations significantly (19 vs. 10 patients intended to continue with the branded or the generic formulation, $p = 0.095$).

Conclusions: Although this study has to be interpreted with limitations, our findings suggest that both branded and generic ropinirole formulations have similar control on the motor- and non-motor symptoms of PD.

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Abstract – WCN 2013

No: 320

Topic: 2 – Movement Disorders

Parkinson's disease: Molecular-genetic investigations in Uzbekistan

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Today, the study of the molecular nature of Parkinson' disease (PD) is one of the priorities.

Aim: To analyze molecular-genetic aspects of PD among indigenous population of Uzbekistan Republic.

Methods: The number of patients was 180 (mean age: 54.8 ± 11.8 years). Control group consisted of 80 persons not suffering from this pathology at the similar age and gender. We carry out the analysis on the presence of associations with PD: mutations Ala53Thr in gene PARK1, T240M gene PARK2, homozygosis for null alleles (0/0) of GSTT1 and GSTM1 genes.

Results: Mutations Ala53Thr in gene PARK1 and T240M in gene PARK2 in investigated population of patients were absent at all patients both in the group with PD and in control group. 0/0 polymorphism for GSTT1 gene was 54.4% and for gene GSTM1–62.8% identified in PD patients. Analysis of the distribution of combined genotypes GSTT1/GSTM1 showed statistically significant differences between PD and control groups in the form of prevalence of combinations of GSTT1 (0/0)/GSTM1/(0/0) genotypes in the group with PD. Clinical and genotypic comparisons showed more early debut of the disease, and prevalence of expanded-mixed form and rapid progression of PD in patients with “null” genotypes of GSTT1 and GSTM1 genes.

Conclusions: High frequency of bearing of null alleles of genes of detoxication GSTT1 and GSTM1, and also their combinations was revealed that indicates to their certain role in the development of Parkinson's disease.

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Abstract – WCN 2013

No: 321

Topic: 2 – Movement Disorders

Parkinson's disease: Sanitary–hygienic investigations in Uzbekistan

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In the whole world, the increase of new cases of Parkinson's disease (PD), among cities, is observed, especially, among agricultural population. It explains the given tendency partially due to the improvement of quality of diagnostics of the disease. However, there are also other reasons for the increase in PD frequency which need to be studied.

Aim: To analyze sanitary–hygienic aspects of PD among indigenous population of Uzbekistan Republic.

Methods: The number of patients was 180 (88 women and 92 men). Sanitary–hygienic researches were made with the help of a specially developed questionnaire, which covered all major factors of biological or sanitary–hygienic character.

Results: Results of questioning of patients with PD among indigenous population of Uzbekistan have shown that the most significant risk factors of PD development were work with pesticides, drinking water from open reservoirs and work in the chemical industry. Influence of harmful factors was not only the risk factor for the disease development, but it also modified clinical picture of disease thus, the median age of patients–countrymen was 53.08 ± 9.14 , patients–city inhabitants– 58.5 ± 10.7 , and the tendency to earlier debut of disease in the agricultural population in comparison with city inhabitants (48.8 ± 9.9 and 53.6 ± 9.9 accordingly) was also observed.

Conclusions: The analysis of the sanitary–hygienic factors of PD has allowed one to reveal most significant of them with high degree of reliability that is important at a substantiation of the concept of PD development, and gives the chance to predict the probability of PD development in persons of Uzbek nationality.

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Abstract – WCN 2013

No: 245

Topic: 2 – Movement Disorders

Postural abnormality as a risk marker for leg deep venous thrombosis (DVT) in Parkinson's disease

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Purpose: Pulmonary thromboembolism is a common cause of death in patients with autopsy-confirmed Parkinsonism. This study investigated the frequency of leg DVT in Parkinson's disease and relationships between DVT and clinical/laboratory findings, including postural abnormalities as assessed by photographic measurements.

Methods: This cross-sectional study assessed the presence of DVT using bilateral leg Doppler ultrasonography in 114 asymptomatic outpatients with Parkinson's disease.

Results: DVT was detected in 23 patients (20%) with Parkinson's disease. DVT was located in the distal portion in 18 patients and in the proximal portion in 5 patients. No significant differences in age, sex, body mass index, disease duration, Hoehn–Yahr stage, anti-Parkinson's drugs, or daily levodopa-equivalent dose were seen between DVT-positive and DVT-negative groups. Univariate analysis for developing DVT in patients with Parkinson's disease identified the following factors: long-term wheelchair use, bent knee, bent spine, and D-dimer elevation. Bending angles were significantly higher in the DVT-positive group at the knee (19.8° , $p < 0.001$) and spine (14.9° , $p = 0.002$) than in the DVT-negative group. In diabetes mellitus cases, long-term wheelchair use, bent knee over 15° , camptocormia, D-dimer elevation, and

two or more risk markers were associated with a significantly increased incidence of DVT. The presence of risk markers contributed to the development of DVT. On multivariate logistic regression analysis, a bent knee posture was strongly associated with an increased risk of DVT.

Conclusion: Presence of leg DVT correlated with postural abnormalities in Parkinson's disease. We recommend non-invasive ultrasonographic screening for DVT in these high-risk patients with Parkinson's disease.

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Abstract — WCN 2013

No: 294

Topic: 2 — Movement Disorders

Gender differences in non-motor symptoms in early, drug naïve Parkinson's disease

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Background: Gender differences in brain structure and function may lead to differences in the clinical expression of neurological diseases, including Parkinson's disease (PD). Few studies reported gender-related differences in the burden of non-motor symptoms (NMS) in treated PD patients, but this matter has not been previously explored in drug-naïve PD patients.

Objective: To assess gender differences in the frequency of NMS in a large sample of early, drug-naïve PD patients compared with age-matched healthy controls.

Methods: Two hundred early, drug-naïve PD patients and sixty age-matched healthy controls were included in the study. Frequency of NMS was evaluated by means of Non-Motor Symptoms Questionnaire. The difference in gender distribution of NMS was evaluated with the χ^2 exact test.

Results: Male PD patients complained of problems having sex and taste/smelling difficulties significantly more frequently than female PD patients. Furthermore, men with PD complained more frequently of dribbling, sadness/blues, loss of interest, anxiety, acting during dreams, and taste/smelling difficulties as compared to healthy control men, while female PD patients reported more frequently loss of interest and anxiety as compared with healthy control women.

Conclusion: This study shows specific sex-related patterns of NMS in drug-naïve PD. In contrast with previous data, female PD patients did not present higher prevalence of mood symptoms as compared to male PD patients. Comparison with healthy controls showed that some NMS classically present in premotor and early stage of disease (i.e. acting out during dreams, taste/smell difficulties) are more frequent in male than in female patients.

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Abstract — WCN 2013

No: 303

Topic: 2 — Movement Disorders

Enhanced susceptibility of GSH-depleted drosophila larvae to rotenone-induced oxidative dysfunctions: Attenuation by *Selaginella delicatula* (A pteridophyte) flavonoids

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Background: Oxidative stress/mitochondrial dysfunctions play a vital role in Parkinson's disease (PD) pathology. Early biochemical

perturbations occurring under compromised redox status consequent to neurotoxin exposure/s may be relevant.

Objective: We studied the biochemical implications of Rotenone (ROT, a Complex-I inhibitor) under GSH-depleted conditions in *Drosophila* larvae and the modulatory effect of *Selaginella delicatula* (Sanjeevani, a pteridophyte) extracts (aqueous-SDAE; methanol-SDME).

Method: Third instar larvae of *Drosophila melanogaster* (Oregon K) were exposed (24 h) in the medium either to ROT (250 μ M) or BSO (6 mM) and their combination. In a satellite study, the protective effects of *Selaginella* extracts were also examined. The biochemical implications were determined in whole body homogenates in terms of oxidative impairments in cytosol and mitochondrial dysfunctions.

Results: ROT caused significant induction of oxidative stress (elevated levels of ROS, protein carbonyls and diminished (31%) activity of complex I). BSO (6 mM) significantly depleted the levels of reduced GSH (50%) and thiols in cytosol. In combination, further depletion of GSH levels occurred with concomitant elevation of oxidative stress. Interestingly, both extracts significantly offset the ROT mediated effects as evidenced by attenuation of oxidative markers, GSH levels and activity of Complex-I.

Conclusion: These data suggest the enhanced susceptibility of GSH-depleted *Drosophila* larvae to Rotenone and provide us an experimental paradigm to examine the relevance of compromised GSH levels. Since larval locomotion is a sensitive readout of locomotor deficits in *Drosophila*, further studies aim to utilize this phenotype and paradigm as a platform to assess the therapeutic propensity of phytochemicals relevant to PD.

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Abstract — WCN 2013

No: 249

Topic: 2 — Movement Disorders

Novel preclinical therapeutic strategies of neurodegeneration with kynurenes: Clinical perspectives (Danube Neurology Session)

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There is appreciable evidence that the neurodegeneration in several neurological disorders (like Alzheimer's disease (AD)) is mediated, at least partly, by neurotoxic products of the kynurenine (KYN) pathway. l-kynurenine can be metabolized through three distinct pathways to form kynurenic acid (KYNA), 3-hydroxy-l-kynurenine and anthranilic acid by the action of kynurenine aminotransferases (KATs), kynurenine-3-monooxygenase (KMO) and kynureninase enzymes, respectively. KYNA is a broad-spectrum, non-selective glutamate receptor antagonist. Correspondingly, KYNA has proved to be a neuroprotective molecule in various experimental models of neurodegenerative disorders, including neurotoxicity induced by kainate, ibotenate, QA and NMDA. Because KYNA has a very limited ability to penetrate the blood brain barrier (BBB), indirect pharmacological approaches are necessary to exploit its therapeutic potential. Possible therapeutic approaches could be to reduce the expression of the neurotoxic agents (like quinolinic acid: QA) or to increase the production of putative neuroprotectant KYNA or make use of its analogues. Among other common mechanisms the shift in the KYN pathway seems to be general over different neurodegenerative diseases and such, neuroprotective therapies influencing the KYN pathway may be beneficial in several neurological pathologies. The most effective preclinical drug candidates are discussed, and attention is paid to currently underinvestigated aspects where modulation of the kynurenine metabolism might be of therapeutic value. Furthermore, the complex anti-inflammatory and neuroprotective properties of KYNA and its analogues provide the

rationale for the screening of these compounds in experimental autoimmune encephalomyelitis (EAE) and multiple sclerosis.

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Abstract – WCN 2013

No: 250

Topic: 2 – Movement Disorders

Violation of metabolism of nitric oxide in patients with Parkinson's disease and vascular parkinsonism

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Nitrogen oxide plays a big role in regulation of a number of physiological and pathological processes in an organism now exposed to intensive studying.

Purpose: To estimate functional activity of NO system and their interrelation with level of dopamine in patients with various forms of parkinsonism.

Methods: A total of 47 patients with Parkinson's disease—PD (62.3 ± 4.51 years) and 45 patients with vascular parkinsonism—VP (63.1 ± 5.5). The control group was composed of 20 patients without symptoms of parkinsonism. The maintenance of stable metabolites of NO—NO₂ and NO₃ were also assessed was estimated in the membranes of erythrocytes. Activities of eNOS, NADFH-NR, to the contents ONOO. Simultaneously in daily urine was determined by the method of dopamine Matlin.

Results: At PD the NO level exceeded control data for 216.9%, and at VP, 150.6%. Increase in the level of NO was associated with significant inhibition in the groups studied eNOS—by 72.3% and 56.8%, respectively, the activation of NADFH-NR—by 75.5 and 43.6%. Overexpression level ONOO at 344.5 and 287.6%, respectively, compared with the control. Installed together, an excretion of dopamine in daily urine was below the control data in patients with PD—for 36.4% and VP—on 21.3 %.

Conclusion: The identified violations of NO-system in red blood cells in patients with various forms of PD and VP, on the one hand reveal some of the fundamental basis of the pathogenesis, the other—open new perspectives suggest means of correction of violations in erythrocytes.

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Abstract – WCN 2013

No: 215

Topic: 2 – Movement Disorders

Hemiballismus: A case series in UYO Southern Nigeria

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Background and aim: Hemiballismus is a dramatic movement disorder that is typically acute in onset. Few cases have been reported in Africa. We describe six cases seen in our hospital between January 2010 and August 2012 with their presentation, management and eventual outcome. This is the first case series to the best of our knowledge in sub-Saharan Africa.

Methods: The participants were six patients seen from January 2010 to August 2012 whose folders were reviewed.

Results: There were two males and four females. The age range was 55–68 years. All the patients were from the Ibibio ethnic group in the South-south Nigeria. The uncontrollable flinging movements all started suddenly. The patients presented within the first week of the movements. All the patients had at least one modifiable risk factor for stroke (hypertension, Diabetes mellitus, obesity and dyslipidaemias).

Each of the patients presented with markedly elevated blood pressure. Two of the patients had morbid obesity with a BMI of >45. Management was mainly supportive: rehydration, sedation and prevention of injuries. The risk factors were controlled. The movements reduced and patients were discharged home well.

Conclusion: Hemiballismus is not rare as previously thought. This sudden surge may be part of the transition to non-communicable diseases noted in sub-Saharan Africa. There may also be an associated environmental factor given that all the patients reside in the same environment. Patients do well on supportive care and Haloperidol.

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Abstract – WCN 2013

No: 189

Topic: 2 – Movement Disorders

Estimation of the caused skin vegetative potentials in patients with Parkinson's disease

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Aim: To study the status of vegetative nervous system in patients with Parkinson's disease (PD) with caused skin vegetative potentials (CSVPs).

Materials and methods: 50 patients were examined, average age 73 ± 10.3 years with duration of disease from 1 till 15 years. The control group concluded 24 persons. CSVP recording was made on a "VNS-SPECTRUM" device, firm Neurosoft (Ivanovo). The comparative estimation of results was made in patients depending on sex, age, form, stage, rate of progression of disease.

Results: The duration of latent periods in patients with PD from feet was more than that in control group ($p = 0.01$), and amplitude indicators in hands ($p = 0.027$) and feet ($p = 0.006$) in group of patients with PD were lower. In patients with duration of disease of more than 1 year amplitude indicators were much lower in hands ($p = 0.017$) and feet ($p = 0.029$), than in patients with duration of disease of less than 1 year. In patients with high quantity of points in section trembling in the UPDRS (>3) amplitude parameters were lower in hands ($p = 0.006$) and feet ($p = 0.008$), than those in patients with less quantity of points. Patients with total of points of more than 30 amplitude parameters in the UPDRS were lower in hands ($p = 0.039$) and feet ($p = 0.008$), than in patients with total of points less or equal to 30. Amplitude parameters in hands ($p = 0.033$) and feet ($p = 0.016$) in group of patients with PD, were lower than in control group.

Conclusions: For authentic diagnostics of progressing vegetative insufficiency it is necessary to use also caused skin vegetative potentials.

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Abstract – WCN 2013

No: 187

Topic: 2 – Movement Disorders

Substantia nigra hypoechoogenicity is not related to Friedreich ataxia

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Background: Previous works have reported that both substantia nigra (SN) hypoechoogenicity and cerebellar dentate nucleus (CDN) hyperechoogenicity coexist in Friedreich's ataxia (FRDA). However, while pathology in CDN is a constant finding in autopsies, no pathological changes have been reported in the SN of FRDA patients.

SN hypoechogenicity has also been observed in patients with rest-less syndrome (RLS), a condition reported occasionally in FRDA patients.

Objective: To investigate the presence of SN hypoechogenicity in a cohort of FRDA patients and its possible association with RLS.

Methods: Fourteen genetically confirmed FRDA patients (mean age: 53.15 ± 17.29 years; mean disease duration: 27.86 ± 14.94 years) and fourteen age and sex-matched healthy controls (mean age: 53.71 ± 15.23 years) underwent transcranial sonography examination (SIEMENS “Acuson X300 PE” 5.0 machine) to evaluate the area of echogenicity of the SN. Ataxia and RLS were also assessed in all participants.

Statistical comparisons of the area of the SN between groups were made using the non-parametric Mann–Whitney *U* test.

Results: The cut-off value for defining SN hypoechogenicity in our controls was 0.21 cm^2 (sum area of both sides). We did not find significant differences between the mean sum area of SN echogenicity in FRDA patients ($0.27 \text{ cm}^2 \pm 0.41$) and in controls ($0.28 \text{ cm}^2 \pm 0.72$) ($p = 0.72$). The areas of SN echogenicity in the two FRDA patients with RLS were 0.21 cm^2 and 0.22 cm^2 , respectively, while the mean area of SN echogenicity of the FRDA patients without RLS was 0.27 ± 0.04 .

Conclusions: Our data do not support the notion that SN hypoechogenicity is related to FRDA itself, although it might be associated with RLS.

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Abstract – WCN 2013

No: 146

Topic: 2 – Movement Disorders

Orthostatic and supine blood pressures are associated with white matter hyperintensities in Parkinson disease

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Background: Several reports on the elderly population have suggested that orthostatic hypotension is associated with white matter hyperintensities (WMHs); however, little information is available on patients with Parkinson's disease (PD).

Objective: To verify the association of orthostatic and supine blood pressures with white matter hyperintensities in Parkinson disease.

Methods: We analyzed the association blood pressure profiles during orthostatic testing with WMH scores in 117 patients with PD. WMHs were rated using the semiquantitative visual rating system proposed by Scheltens et al.

Results: The presence of orthostatic hypotension was associated with increasing tendency of WMH score and the blood pressure changes during orthostasis and supine blood pressure was positively correlated with increasing WMH score.

Conclusion: This finding indicates that hemodynamic changes associated with orthostatic hypotension may be associated with white matter changes in patients with PD.

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Abstract – WCN 2013

No: 194

Topic: 2 – Movement Disorders

Cigarette smoking, coffee intake and alcohol consumption preceding Parkinson's disease: A case-control study

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Background: The lack of a clear genetic underpinning has led to the investigation of the role of environmental factors in the etiology of Parkinson's disease (PD).

Objective: A case-control study was performed in Belgrade in order to investigate the association between PD and smoking, coffee and alcohol consumption.

Methods: 110 new PD cases and 220 hospital controls were interviewed. Cases and controls were matched by sex, age and place of residence. Conditional univariate and multivariate logistic regression methods were used.

Results: With PD, current smoking [odds ratio (OR) = 0.44; 95% confidence interval (CI) = 0.23–0.82], alcohol consumption (OR = 4.78; 95% CI = 2.67–8.55) and coffee consumption (OR = 2.54; 95% CI = 1.36–4.75) were associated independently from each other. In every smoker the risk for PD significantly decreased with the increasing number of cigarettes smoked and with increasing duration of smoking. The risk for PD significantly increased with the increasing quantity of alcohol consumption. PD risk was significantly higher in subjects whose average daily consumption of coffee was 1 and 2–3 cups, and it was lower (but not significantly) in those whose daily coffee consumption was 4+ cups. The results of multivariate analyses did not substantially change after adjustment on family history positive of PD.

Conclusion: The findings of this study support the hypotheses of inverse association of smoking with PD, but an inverse association with coffee was not confirmed. PD was found to be positively associated with increased alcohol consumption.

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Abstract – WCN 2013

No: 197

Topic: 2 – Movement Disorders

Spatial memory and cognition in patients with Parkinson's disease: Evaluating the dopaminergic effect

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Background: Dementia in patients with Parkinson's disease (PDD) has attracted recent attention. However, the cognitive aspects affected are not fully understood.

Objective: We hypothesized that spatial cognition and memory might be deficient in patients with Parkinson's disease (PD) compared with healthy volunteers (HVs). Further, we hypothesized that such functions in PD should not be affected by dopaminergic intervention.

Patients and methods: 14 patients with PD and 14 HVs were recruited. We used a virtual reality (VR) task including turns in virtual corridors. The subjects were first requested to memorize the route, and then to follow it using a joystick afterwards. We prepared two versions including three (run 1) or four (run 2) turns. The success rate and the time were assessed. All the PD patients were tested both in off and on-med conditions. All the subjects completed the Montreal Cognitive Assessment (MOCA). The performances were compared between PD patients and HVs, and between off and on-meds in PD patients.

Results: In both runs, PD patients showed significantly lower success rate and took longer time compared with HVs. There was no significant difference either in the success rate or in the time between off and on-

meds in PD patients. The correlation analysis in run 2 revealed a significant correlation between the MOCA scores and the time taken.

Conclusions: Our results show that the patients with PD have dysfunction in spatial cognition and memory compared to HVs. Additionally, it appears that such functions are not affected by the dopaminergic pathway.

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Abstract – WCN 2013

No: 153

Topic: 2 – Movement Disorders

Exposure of GSM-900 MHz on rat's brain does not support for gene expression involved in DNA damage and repair pathways

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Objective: We investigated whether an acute exposure to GSM-900 MHz irradiation from a mobile phone (3 h/day up to 28 days) could lead to the modulation of a gene expression pattern in the rat brain.

Methodology: All rats were habituated prior to the experiment in a well designed circular cellular phone exposure (CCPE) cage. In the cage, a mobile phone was placed in the center and rats were allowed to move around the device within a 10 cm radius. After 28 days, mRNA from the brains of control, Sham and GSM-900 MHz exposed rats were isolated and subjected to quantitative real time polymerase chain reaction (qRT-PCR) array analysis. Gene expression was measured and expressed in fold change as described by Bandhyopadhyay et.al. (2007). The qRT-PCR was performed by using SYBR green chemistry on an ABI 7900HT platform. The real time quantification of amplification was done by measuring change in fluorescence by SDS 2.3 software and further analyze by RQ manager.

Results: Thus the results obtained from this study have shown that there is no significant change in expressions of genes involved in DNA damage and repair pathways such as chromatin assembly (CHAF1A), DNA damage checkpoints (Chk1), DNA synthesis (POLD1), post-transcriptional processes (RBM4), translation synthesis (POLI) and stress signaling (Hsp90) as compared with controls.

Conclusion: The study concludes that acute exposure to mobile phones (GSM-900 MHz) does not support for gene expression involved in DNA damage and repair pathways.

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Abstract – WCN 2013

No: 83

Topic: 2 – Movement Disorders

Clinical spectrum of cerebral palsy in South Jordan: Analysis of 122 cases

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Background: The clinical spectrum and associated factors of cerebral palsy (CP) may differ between developing and developed countries.

Aim: To evaluate some predisposing factors, clinical spectrum, and some associated problems of cerebral palsy in children.

Setting and design: It is a retrospective study wherein data were extracted from patient file records which contain extensive historical and clinical data in a center for early diagnosis of childhood disabilities.

Patients and methods: One hundred and twenty two children with ages ranging from 7 months to 17 years were reviewed in a 32-month period between September 2007 and April 2010. A simple statistical analysis was used for a percentage calculation.

Results and conclusions: A spastic type was predominant (82.7%), with a quadriplegic subtype being the most common (34.4%). The other types were choreoathetoid (8.2%), mixed type (6.6%) and ataxic (2.5%) being the least. Speech delay was the most common associated problem (71.3%) followed by mental retardation (61.5%), seizures (35.2%), hearing problems (26.2), and autism (4.9%) being the least. The clinical spectrum of CP in Jordan may differ from that reported in Western countries. Prospective studies are needed to evaluate the clinical spectrum and predisposing factors in Jordan.

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Abstract – WCN 2013

No: 78

Topic: 2 – Movement Disorders

Mutation analysis of ATP7B gene in Vietnamese patients with Wilson disease

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Wilson disease (WD) is an autosomal recessive disorder of copper metabolism, which is caused by mutation in copper-transporting P-type ATPase (ATP7B). The principle of this disease is the failure of hepatic excretion of copper through the bile and leads to the deposit of copper in the liver and other organs. The ATP7B gene is located on the long arm of chromosome 13 (13q14.3).

Objective: This study aimed to identify the gene mutation of the Vietnamese patients with WD and to report a novel mutation which is prevalent in Vietnam.

Material and method: 12 unrelated Wilson disease patients were studied. Genomic DNA was extracted from peripheral blood samples. The most common mutation, p.Arg778Leu, was firstly screened by PCR-RFLP then all 21 exons and exon-intron boundaries of the ATP7B gene were analyzed by direct sequencing.

Result: We identified 5 different mutations, accounting for 79.17%, among them was a novel mutation (p.Thr850Ileu). Mutation p.Ser105* was the most prevalent (45.83%). Other mutations are listed as the following: p.Arg778Leu (8.33%), p.Pro992Leu (4.17%), p.Asp1270Ser (4.17%), and p.Leu1371Pro (8.33%). 3 patients were homozygous for a single mutation, and 5 patients were compound heterozygous.

Conclusion: The most prevalent mutation in Vietnam is p.Ser105*. p.Arg778Leu might be not common in Vietnam, and p.Ser105* in exon 2 should be screened firstly on those Vietnamese patients who have a higher risk of WD before sequence analyzing the entire gene.

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Abstract – WCN 2013

No: 97

Topic: 2 – Movement Disorders

Molecular cross-talk between Lrrk2 and NF-κB in neuroinflammation in an animal model of Parkinson's disease (PD)

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Background: Alteration or mutation of LRRK2, highly expressed in microglia and neurons has been implicated to PD. It was found that LRRK2 deficiency attenuated LPS-induced mRNA and/or protein

expression of cytokines and NF- κ B was decreased in LRRK2-knock down cells. LRRK2-induced NF- κ B activation was dependent on the IKK complex, because it was inhibited by a dominant negative form of IKK. The main PD-associated mutant LRRK2-G2019S did not activate NF- κ B differently from wild-type protein. It is not clear whether PD-LRRK2 mutations operate through a gain or loss of function that might act in a dominant negative fashion, recruiting and neutralizing wild-type proteins.

Methods: In our preliminary study we have also found differential expression patterns of p65, TNF- α , p52 and LRRK2 at different time intervals. However the definite correlations between them have not been defined. Therefore, how LRRK2 links with which pathway of NF- κ B that possibly promotes neuroinflammation is far from clear.

Results: In our preliminary study we have also found differential expression patterns of p65, TNF- α , p52 and LRRK2 at different time intervals. However, the definite correlations in between them have not been defined.

Conclusion: Our study demonstrates the future therapeutical point of inhibition/attenuation and/or induction of particular signaling molecules in NF- κ B and LRRK2.

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Abstract – WCN 2013

No: 99

Topic: 2 – Movement Disorders Chorea-acanthocytosis

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Background: Chorea-acanthocytosis is an autosomal recessive neurodegeneration caused by mutations in the *VPS13A* gene, encoding chorein, located on chromosome 9q21. It manifests with acanthocytes in the peripheral blood and a progressive movement disorder, typically including chorea, although dystonia, facial tics and Parkinsonism may occur. Other features include seizures, neuropathy, myopathy, autonomic dysfunction, psychiatric symptoms and dementia.

The case: A 34 year old woman, with consanguineous parents, presented with a 3-year history of facial grimacing, dysphagia, stutter, memory difficulties, depressive symptoms, anorexia, intermittent chest pain and shortness of breath. From the ages of 2 to 4 years she had 'fits', when she would momentarily develop a staring expression.

Examination revealed facial tics, excessive blinking, mild dysarthria, a stammer and intermittent clucking noises and occasional tongue protrusions but no other signs. Formal neuropsychological assessment was unremarkable. Initial creatine kinase (CK) was mildly elevated, blood films were normal and a MR brain scan showed mild non-specific white matter changes.

Four years later she had chorea in her limbs, impaired manual function and an unsteady gait. Dysarthria and dysphagia were apparent and tongue protrusions could extrude food. She excoriated her forehead and wore a 'bite-guard' to prevent injury to her tongue. Peripheral blood smears identified acanthocytes, a MRI showed caudate atrophy and protein function tests showed reduced chorein levels, indicating chorea-acanthocytosis.

Conclusion: Useful clues suggesting chorea-acanthocytosis are an elevated CK and oral-lingual dystonia, often causing tongue protrusions and lacerations. The diagnosis is confirmed by finding acanthocytes in blood, caudate atrophy and a decreased chorein level.

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Abstract – WCN 2013

No: 100

Topic: 2 – Movement Disorders Tremor dominant parkinsonism – Lesion or deep brain stimulation?

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Background: The optimal stereotactic target for the control of medication resistant tremor dominant Parkinson's disease (TDPD) is debatable, as is the issue of lesion versus DBS in cases with predominantly unilateral signs.

The case: A 56 year old lady presented with medication resistant tremor of the right hand secondary to TDPD. There was right sided cogwheel-rigidity, bradykinesia and 'off-phase' dystonic clawing of her toes but no left sided signs. At sixty, a left Vim-ZI-STN lesion was performed which abolished her right sided signs.

Aged 76, her neuropsychological profile was satisfactory. The benefit to her right side was sustained except for occasional mild dyskinesia and dystonia of her right hand and minimal leg tremor. However, severe left sided rest and postural tremor developed and interfered with her activities of daily living when medication (Sinemet – 110 \times 9 tablets/day and Sinemet 250 CR at night) wore off. This was accompanied by mild left sided bradykinesia and rigidity. Consequently, a right VIM-ZI DBS surgery was performed, which initially produced complete left sided tremor suppression but subsequently there was some escape despite reprogramming.

Conclusion: This case illustrates the long term stability and effectiveness of a correctly placed Vim-ZI-STN lesion for controlling TDPD. This benefit was sustained two decades later, without any of the maintenance requirements or cost associated with DBS. Furthermore, over the course of 20 years the only signs apparent in her right sided limbs were minimal dystonia and dyskinesia. The patient was delighted with these results and greatly preferred the lesion to DBS.

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Abstract – WCN 2013

No: 61

Topic: 2 – Movement Disorders Neuroprotective effect of sesamol and quercetin against QA induced neurotoxicity: An experimental paradigm of Huntington's disease

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Quinolinic acid (QA) is an excitatory amino acid (a NMDA-selective glutamate agonist and an endogenous metabolite of tryptophan), whose properties (a preferential degeneration of GABA-ergic neurons and a relative sparing of NADPH diaphorase- and cholinergic interneurons) and simple application account for its wide use in creating a practicable (though not ideal) animal model of HD. The present study was designed to evaluate the neuroprotective effect of sesamol and quercetin against QA-induced neurotoxicity: an experimental paradigm of Huntington's disease by investigating various behavioral and biochemical alterations in rats. Rats were intrastrially administered quinolinic acid and were treated with sesamol (4, 8 and 16 mg/kg, i.p.) and quercetin (25, 50 and 100 mg/kg, i.p.) for 14 days before and 14 days after quinolinic acid administration. Intrastriatal injection of QA leads to increased escape latency, impaired locomotor activity, increased immobility period, raised lipid peroxidation and nitrite concentration and depletion of endogenous antioxidants. Besides, intrastriatal QA significantly increased TNF- α levels suggesting QA mediated

oxidative and neuroinflammatory damage. Intrastriatal administration of QA resulted in significant decrease in the levels of dopamine, serotonin and norepinephrine in the rat forebrain. Chronic treatment with sesamol (4, 8 and 16 mg/kg, i.p.) and quercetin (25, 50 and 100 mg/kg, i.p.) attenuated these behavioral, biochemical and neurochemical alterations in the rat brain and these effects were attributed to their strong antioxidant and anti-inflammatory potential. The present study suggests that sesamol and quercetin could be used as effective agents in the management of Huntington's disease and should be explored further.

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Abstract – WCN 2013

No: 52

Topic: 2 – Movement Disorders

Systemic blood pressure profile correlates with cardiac ¹²³I-MIBG uptake in patients with Parkinson's disease

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Purpose: To examine the correlation between the systemic blood pressure profile and cardiac ¹²³I-metaiodobenzylguanidine (MIBG) uptake in patients with Parkinson's disease (PD).

Methods: We monitored circadian blood pressure patterns of 37 PD patients of 49 to 85 years of age (mean, 71.8 ± 8.4 years) using a portable blood pressure monitoring device. There were 37 age- and sex-matched control subjects. Cardiac MIBG scintigraphy was performed on the 37 PD patients. Based on the nocturnal fall in mean arterial blood pressure (MABP), we classified patients into extreme dippers (nocturnal reduction of MABP >20%), dippers (>10% but <20%), nondippers (<10% but >0%), and inverted dippers (<0%). Average 24-h MABP values revealed reduced BP variability in PD patients.

Results: The percentage nocturnal fall in MABP was significantly different between PD patients and control subjects (p < 0.05). Significant correlations were found between %MABP reduction and the heart-to-mediastinum (H/M) ratio on early and delayed images (p < 0.01). The UPDR motor score, and early and delay H/M ratios were also significantly different between patients who were and were not dippers (p < 0.05).

Conclusion: The present results reported for the first time a significant correlation between the systemic blood pressure profile and cardiac ¹²³I-MIBG uptake in patients with PD. The degeneration between the brainstem and the postganglionic neurons of myocardial sympathetic nerves may progress in parallel in patients with PD.

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Abstract – WCN 2013

No: 51

Topic: 2 – Movement Disorders

Synergetic effect of intrathecal baclofen and deep brain stimulation in treating dystonia

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Dystonia is a syndrome of sustained muscular contractions of opposing muscles with various etiologies. The currently available

symptomatic treatment strategies are quite effective for some of the various types of dystonia. They help in decreasing involuntary movements, correcting abnormal posture, preventing contractures, reducing pain, and improving function and quality of life. Intrathecal baclofen and deep brain stimulation were proven to be fairly effective in controlling dystonia when used separately. We are reporting a synergetic effect of intrathecal baclofen and deep brain stimulation when used simultaneously in two cases of primary generalized dystonia with excellent control of dystonia.

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Abstract – WCN 2013

No: 50

Topic: 2 – Movement Disorders

Botulinum-A toxin in pediatric stiff hips

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Purpose: We aim to determine the functional and orthopedic contributions of botulinum toxin-A in the treatment of pediatric stiff hips.

Methods: Three patients with complicated developmental dysplasia of the hip after surgery and a case of idiopathic chondrolysis were given botulinum toxin injections in selected muscles as an adjunctive therapy to the standard orthopedic management.

Results: All patients experienced significant reduction in pain with significant improvement in posture, range of movement and mobility.

Conclusion: Botulinum toxin-A injection when given to selected muscles appears effective in relieving pain and improving range of mobility in patients with complicated developmental dysplasia of the hip and idiopathic chondrolysis.

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Abstract – WCN 2013

No: 23

Topic: 2 – Movement Disorders

The new hypothalamic proline-rich polypeptide-1 protects motor dysfunction and memory deficits after ischemic stroke in rats

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Background: The proline-rich peptide (PRP-1), which was isolated from the neurosecretory granules of the bovine neurohypophysis (produced by n. supraopticus and n. paraventricularis). It was observed that the PRP-1 produced several beneficial biological effects which included immunoregulatory, hematopoietic, antimicrobial and anti-neurodegenerative properties. The aim of the present study was to investigate the effect of PRP-1 on focal cerebral ischemia induced memory deficits and motor dysfunctions in rats.

Materials and methods: Ischemic stroke was induced in rats via the middle cerebral artery occlusion (MCAO). Assessment of memory was done using the passive avoidance test. The rota-rod test was employed to assess motor coordination. Then after MCAO, the rats were divided into 2 groups: control and PRP-1 treated. Assessment of memory and motor coordination was carried out before MCAO and also after the 3rd, 6th and 12th post-insult days.

Results: Following the MCAO, the memory and motor coordination of the rats in the control group were markedly impaired, in comparison with values achieved before MCAO and after the 3rd and 6th post-insult days; but on the 12th post-insult day there was a noticeable loss of memory and motor coordination. In the case of PRP-1 treatment, after 3rd, 6th and 12th post-insult days there was a noticeable enhancement of basal (before MCAO) values up to normal value.

Conclusion: Thus systematic administration of PRP-1 has been proven to significantly decrease ischemia-induced memory deficits and motor dysfunction. Hence it may be concluded that PRP-1 could be useful clinically in the prevention of stroke.

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Abstract – WCN 2013

No: 47

Topic: 2 – Movement Disorders

Quantitative assessment of hypokinesia using sensor gloves in Parkinson's disease

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Introduction: Instruments that contribute to quantitative (objective) mensuration of cardinal signs in Parkinson's disease (PD) like the bradykinesia, would assist more effective therapeutic intervention.

Objectives: To evaluate the utility of sensor gloves on quantitative evaluation of bradykinesia on Parkinson's disease.

Methods: The authors carried out a study of type cases and controls, organized in two groups: 13 patients with clinical diagnosis of idiopathic PD in "on" state and 24 controls, to which were carried out a registration of the "finger tapping" movement using sensor gloves (5DT-14 Ultra). The analysis of the amplitude was calculated through the power spectra derived from a Fourier transform (FFT). Also we examine the relationship between glove data obtained, the clinical scale employed (UPDRS) and the early and late components of the premotor potential.

Results: The authors observed that patients presented a smaller speed in the execution of movements, demonstrated to reach their maximum amplitude at smaller movement frequency (1.5 Hz) than controls (3.5–4 Hz), as well as, a significant decrease of the amplitude of movement

toward the frequencies of movement >3 Hz. The most favorable values in the clinical scale employed were associated to broader movements toward the high frequencies (>3.5 Hz). Our study demonstrated a relationship between amplitude of movement and the area of the earlier and later components of the premotor potential. A bigger area associated to a broader movement.

Conclusion: The authors conclude that sensor gloves are useful as a quantitative complement of the clinical evaluation in the patients with PD.

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Abstract – WCN 2013

No: 28

Topic: 2 – Movement Disorders

Spheroid bodies: From Seitelberger's disease to neuroaxonal dystrophy and MSA-C

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Background: Since the descriptions of spheroid bodies in grey matter (GM) conditions: Seitelberger's disease and brain iron accumulation (NBIA), new perspectives have emerged considering spheroid bodies' association to autonomic nerve endings and enolase, neurofilaments and chromogranin findings, similar to MSA type C and Niemann–Pick type II specimen.

Objective: To compare the recent definition of Wszolek Z.K. et al. (2009), considering adult onset leukoencephalopathy with axonal spheroid and pigmented glia (ALSP) with current and non-habitual forms of neuroaxonal dystrophy.

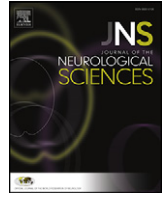
Methods: Comparatives of Seitelberger's disease specimen with Ceuterick C. and Martin J.J. (1984) descriptions of axonal cytoplasmatic bodies.

Sampling of glial cytoplasmatic inclusions (GCIs) in MSA (Wenning) and neuroaxonal dystrophy similarities.

Results: Comparatives may be obtained between molecular layer axonal spheroids in MSA-C, and peripheral nerve ending findings (Yamanaka, 2005).

Conclusions: Suggested correlations would be profiled considering leukoencephalopathy with spheroids (HDLS) and pigmentary leuko-dystrophy (POLD) similarities with peripheral specimen in atypical metachromatic leukodystrophy (Coulter Mackie M.B., 2002).

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Topic: 3 - Stroke

Abstract – WCN 2013

No: 33

Topic: 3 – Stroke

Prevalence of significant carotid artery stenosis in Iranian patients with peripheral arterial disease

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Objectives: The aim of the present study was to determine the prevalence of significant internal carotid artery (ICA) stenosis in a group of Iranian patients with peripheral arterial disease.

Methods: We prospectively screened 120 patients with a known diagnosis of peripheral vascular disease for carotid artery stenosis. Based on the angiographic assessment of abdominal aorta and arteries of the lower extremities, patients with stenosis greater than 70% in the lower extremity arteries were included. A group of healthy individuals aged 50 years was recruited as a control. Risk factors for atherosclerosis, and cerebrovascular disease were recorded. Common carotid arteries (CCAs) and the origins of the internal and external arteries were scanned with B-mode ultrasonography. Significant ICA stenosis, .70% ICA stenosis but less than near occlusion of the ICA, was diagnosed when the ICA/CCA peak systolic velocity ratio was 3.5.

Results: Ninety-five patients, with a mean age of 58.52 ± 11.04 years, were studied. Twenty-five patients had a history of smoking, six patients had a history of coronary artery disease, six patients had hypertension, and ten patients had diabetes mellitus. Significant ICA stenosis was present in four patients (4.2%) with peripheral arterial disease and in one healthy individual (1%) of the control group ($P = 0.05$). In terms of the risk factors for atherosclerosis, no statistically significant relationship was found between individual atherosclerotic risk factors and significant ICA stenosis ($P = 0.05$).

Conclusion: The prevalence of significant ICA stenosis in Iranian patients with peripheral arterial disease is low. In addition, there is no relationship between individual atherosclerotic risk factors and significant ICA stenosis.

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Abstract – WCN 2013

No: 43

Topic: 3 – Stroke

Granulocyte colony-stimulating factor (G-CSF) for acute and subacute ischemic stroke: A meta-analysis

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0022-510X/\$ – see front matter

Introduction: G-CSF is known to mobilize hematopoietic stem cells from the bone marrow into the peripheral blood.⁽⁴⁾ Meta-analysis from the animal studies suggested that G-CSF both reduces infarct size and enhances functional recovery.⁽⁵⁾

Objective: To assess the evidence from randomized controlled trials the effects of granulocyte colony-stimulating factor (G-CSF) compared with placebo in the treatment of patients with ischemic stroke.

Design: Meta-analysis of 5 randomized trials for acute ischemic stroke and subacute ischemic stroke was identified through Medline/Pubmed and Cochrane Library. Summary of the outcome variables was computed using difference of two means of the mean score and their corresponding standard error under fixed effects models. The chi-square test was done to test heterogeneity. Statistical analysis was done using Revman version 5.

Results: The sample size of the studies ranged from 10 to 60 patients. The treatment duration was from 72 h to 5 days. The main outcome measures were changes in the NIHSS, Barthel Index (BI) and Modified Rankin Scale (MRS) from baseline. The mean difference in leukocyte count at the end of treatment showed that the overall effect was statistically significant ($z = 21.98$; $p < 0.0001$) in favor of the control using the fixed effects model. The frequency of adverse events observed in the G-CSF and placebo treated patients in all the studies included did not show any statistically significant difference.

Conclusion: The results of the present meta-analysis do not lend statistically significant support to the use of granulocyte colony-stimulating factor (G-CSF) in patients with acute and subacute ischemic stroke.

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Abstract – WCN 2013

No: 44

Topic: 3 – Stroke

Effect of erythropoietin therapy on vasospasm, cerebral infarction and Glasgow outcome scale in patients with aneurysmal subarachnoid hemorrhage: A meta-analysis

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Introduction: Experimental models of aneurysmal subarachnoid hemorrhage (SAH) have demonstrated that acute systemic erythropoietin (EPO) therapy reverses impaired autoregulation, reduces vasospasm and ischemic deficits, and improves neurological outcome.⁽³⁾

Objective: To assess the evidence from randomized controlled trials the effects of erythropoietin therapy (EPO) compared with placebo on transcranial doppler (TCD) vasospasm, cerebral infarction and Glasgow outcome scale (GOS) in patients with aneurysmal SAH.

Design: Meta-analysis of 2 randomized trials was identified through Medline/Pubmed and Cochrane Library. Data was analyzed using Review Manager (RevMan) 5.1 software. Data on TCD vasospasm, cerebral infarction and GOS score at six months were abstracted from the two studies. Chi-square test of heterogeneity of effect measure was used to determine whether a fixed or random effects model should be used. The Z-test for overall effect was used to test the significance.

Results: The sample size of the studies ranged from 73 to 80 patients. All studies were randomized and placebo controlled. A fixed effects model was used for GOS score at 6 months and TCD vasospasm because tests of heterogeneity were not significant with $p = 0.48$ and $p = 0.54$, respectively. On the other hand, a random effects model was used for cerebral infarction with $p = 0.03$ for the test of heterogeneity. No statistically significant effect was observed on TCD vasospasm (RR = 0.78, $p = 0.23$), cerebral infarction (RR = 0.41, $p = 0.22$) and favorable GOS score at 6 months (RR = 1.04, $p = 0.72$).

Conclusion: The results of the present meta-analysis do not lend statistically significant support to the use of erythropoietin therapy in patients with aneurysmal SAH.

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Abstract -WCN 2013

No: 11

Topic: 3 -Stroke

Walking recovery in the first-year after stroke: Results from the LEAPS RCT

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Background: After acute first-time stroke, almost 50% of people discharged from the acute care hospital to home are at high fall risks and cannot participate in home or community-life due to walking limitations.

Objective: Primary subgroup analysis of the LEAPS RCT reporting the frequency, intensity, and duration of neurorehabilitation to reduce walking disability in the first year post-stroke.

Patients/methods: RCT of 359 adults (age 62.0 ± 12.7 years; 54.9% male) that completed 30–36 physical therapy sessions with 2-, 6-, and 12-month outcome assessments. At 2-mos post-stroke, participants were stratified by walking severity (severe, <0.40 m/s; moderate, 0.40 – 0.79 m/s) and randomization to one of 2 parallel interventions (progressive exercise, EX; task-specific locomotor training, LT), provided early at 2-months (EX, $n = 113$; LT-early; $n = 121$) or later at 6-months (LT-late; $n = 125$). Disability threshold was gait speed change ≥ 0.16 ms (1-level disability change on Modified Rankin Score).

Results: Disability threshold was exceeded and retained at 1-yr with 24 sessions of LT provided at 2 mos for moderate disability group; the severe group needed 36 sessions of LT or EX (Type, 3-way interaction, $p = 0.05$). When LT was delayed to 6-mos, both the severe and moderately impaired groups were more disabled than the early groups (timing, 3-way interaction, $p = 0.03$).

Conclusions: For community-dwelling adults who ambulate at speeds <0.80 m/s at 2-mos post-stroke, a minimum of 24–36 sessions of an intensive, specialized walking program by physical therapists resulted in higher levels of ability, sooner in the recovery year than conventional neurorehabilitation approaches.

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Abstract – WCN 2013

No: 18

Topic: 3 – Stroke

Relationship between neural apoptosis and cell death inducing Dff45-like effector-B gene expression after cerebral ischemic reperfusion injury in rats

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Objective: To explore the relationship of cell death inducing DFF45-like effector-B (CIDE-B) gene activation and neural apoptosis after cerebral ischemic reperfusion injury.

Methods: Total of 144 Wistar rats were randomly divided into sham operation group ($n = 36$) and cerebral ischemic reperfusion group ($n = 108$) consisting of ischemic 30 min, 90 min and 120 min groups with 36 rats respectively. Cerebral ischemia reperfusion models were established with the four artery blocking method according to cerebral ischemia Pusinelli reference. TUNEL method was used to observe the apoptosis of neurons, while immunohistochemical detection was for CIDE-B positive neurons in hippocampus. The expressions of CIDE-B mRNA and protein were detected by RT-PCR technique and Western blot method respectively.

Results: Compared with sham operation group, the number of apoptotic neurons increased significantly after ischemia 30 min, 90 min and 120 min reperfusion 6 h, 1 d, 3 d, 7 d and 14 d ($t = 2.47$ – 3.94 , $P < 0.01$ – 0.05) and CIDE-B positive neurons increased significantly ($t = 2.28$ – 3.94 , $P < 0.01$ – 0.05); CIDE-B mRNA positive neurons in the subgroups of ischemia 30 min, 90 min and 120 min reperfusion 7 d and 14 d which was significantly more than that of ischemia 30 min, 90 min and 120 min reperfusion 6 h, 1 d, 3 d and 28 d ($P < 0.05$).

Conclusion: The neural apoptosis was time-dependent on the expressions of CIDE-B mRNA and protein after cerebral ischemic reperfusion injury in rats.

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Abstract – WCN 2013

No: 19

Topic: 3 – Stroke

Effect of Xing-naojing and Shengmai injection on PAR1 and AQP4 expressions in rats after intracerebral hemorrhage

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Objective: To study the effect of Xingnaojing and Shengmai injection on PAR1 and AQP4 expressions in rats brain tissue after intracerebral hemorrhage (ICH).

Methods: The ICH model was induced by injection of auto-blood. Forty male Sprague–Dawley (SD) rats were randomly divided into ICH group ($n = 10$), saline group (NS, $n = 10$), Xing-naojing and Shengmai injection group (XNJSM, $n = 10$), hirudin group (HIR, $n = 10$). After 72 h, the rats were decapitated for hematoxylin and eosin staining which was performed to examine perihematoma neurocyte morphological changes and the expression of PAR1 and AQP4 in perihematoma tissue by immunohistochemistry and Western blot techniques respectively.

Results: The expression of PAR1 and AQP4 in perihematoma tissue were increased after ICH, however, compared with ICH group, perihematoma tissue morphological changes were improved obviously, and the expressions of PAR1 and AQP4 were also decreased significantly between XNJSM group and HIR group ($P < 0.05$).

Conclusions: The increased expressions of PAR1 and AQP4 may induce some brain injury together following ICH, and Xing-naojing

and Shengmai injection can alleviate neural injuries after ICH by inhibiting the expressions of PAR1 and AQP4.

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Abstract – WCN 2013

No: 91

Topic: 3 – Stroke

Parinaud's syndrome due to a unilateral vascular ischemic lesion

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Background: Parinaud's syndrome includes as core feature paralysis of conjugate vertical eye movements (especially upgaze) with convergence–retraction nystagmus on attempted upgaze. Since the lesions accounting for the paralysis of voluntary vertical eye movements are located adjacent to the pupillomotor pathways, there can be varying degrees of paralysis of convergence, pupillary constriction, and accommodation. Parinaud's syndrome may be caused by extrinsic or intrinsic lesions, with pineal tumors and hydrocephalus being common etiologies.

Clinical case: A 59-year-old man complained of binocular diplopia of sudden onset noted after an exploratory laparotomy complicated by cardiorespiratory arrest during anesthetic induction. On inspection he had Collier's sign. He had complete paralysis of upward vertical gaze associated with convergence–retraction nystagmus on attempted upgaze. Downward vertical gaze and horizontal gaze were intact. We noted skew deviation with hypertropia in the left eye, confirmed by Hess screen with negative Bielschowsky. His pupils were round and unequal (right 2 mm, left 3 mm), both reacting to light. Brain MRI revealed a small lesion in the left paramedian midbrain which was hyperintense in T2 and FLAIR, compatible with vascular ischemic sequelae.

Conclusion: Our patient exhibited Parinaud's syndrome from a unilateral vascular ischemic paramedian midbrain lesion that may result from involvement of the interstitial nucleus of Cajal and/or fibers to and from posterior commissure. We demonstrate similarly to other cases described in the literature, that a single millimetric unilateral lesion, near the midline, may interrupt the pathways involved in the vertical gaze just before and after they decussate, inducing a functionally bilateral lesion.

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Abstract – WCN 2013

No: 104

Topic: 3 – Stroke

Activation of renin–angiotensin system (RAS) in acute stroke

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Background: The RAS plays an important role in the development of arterial hypertension and angiotensin converting enzyme (ACE) is a key enzyme which converts angiotensin I to angiotensin II – the main active product of RAS. But the role of RAS activation in ischemic stroke (IS) is not clear.

Objective: To clarify the association of RAS activation and clinical course of acute IS.

Patients and methods: The examination included 191 hypertensive patients with acute IS. All patients were genotyped for the ACE

insertion/deletion (I/D) polymorphism by polymerase chain reaction. ACE activity and angiotensin II level in serum were determined by enzymatic assay.

Results: In IS patients most frequent variant of genotype was DD – in 44.5%, less frequent is II – in 15.2% and heterozygote ID – in 40.3%. There were strong differences between DD and II variants which are associated with clinical course of stroke and RAS activity. Genotype DD was most unfavorable and associated with more severity of stroke to NIHSS – 16.6 ± 0.9 , with II – 8.8 ± 0.6 and higher blood pressure at admission. This accompanied the increase of ACE activity to 59.7 ± 5.8 U/L with DD genotype and 47.36 ± 8.61 U/L ($p = 0.034$) with II, and elevation of serum angiotensin II level to 50.8 ± 3.3 pg/mL comparable to 36.6 ± 6.9 pg/mL in II genotype ($p < 0.01$). Monozygotic genotype II was more favorable and associated with best restored of neurological function and lower of blood pressure.

Conclusion: The ACE polymorphism is clinically relevant and influences the course of IS. Patients who had D-allele observed higher RAS activity and severity of stroke.

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Abstract – WCN 2013

No: 137

Topic: 3 – Stroke

Ultrasonographic characteristics of ocular ischemic syndrome

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Introduction: The internal carotid artery (ICA) is the main route by which the blood is supplied from the heart to the brain and eye. The ocular and orbital circulation is assured by the ophthalmic artery, which is the main collateral branch of the ICA. Occlusion or severe stenosis of the ICA (which is more than 70% of the arterial lumen's diameter) may lead to transient or permanent symptoms of retinal ischemia and to an increased risk of ischemic stroke.

Purpose: To define orbital circulation abnormalities identified by color Doppler imaging (CDI) of retrobulbar vessels in patients with ICA occlusive/severe stenosis disease.

Methods: We used a Loqic 500 sonographer with 9 MHz linear probe for Doppler investigation of retrobulbar vessels, and an ultrasound equipment (MyLab 50 Esaote) with a 7.5–10 MHz linear array transducer for extracranial Duplex sonography.

Results: We presented 10 patients with severe ICA stenosis/occlusion that developed or not an ocular ischemic syndrome. We discussed the hemodynamic status (orbital and cerebral) in order to elucidate the contribution to the ischemic symptoms. Cerebral and retinal perfusion is dependent not only on the degree of stenosis, and embolic risk, but also on the presence of unilateral or bilateral lesions and on the patency of collateral pathways.

Conclusion: The presentation of ocular ischemic symptoms may be the initial sign of carotid artery stenosis/occlusion and can also be used to predict the severity of ICA's disease.

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Abstract – WCN 2013**No: 138****Topic: 3 – Stroke****An evaluation of 42 cases of cerebral vein and dural sinus thrombosis**

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Background and objectives: Cerebral vein and dural sinus thrombosis (CVT) present a variety of non-specific clinical signs. The aim of our study was to identify CVT causes and risk factors, to describe the demographic, clinical, laboratory and neuroimaging data, and to evaluate the treatment and outcome.

Patients and methods: We included 42 consecutive pts with CVT, which were examined at admission and after three months, using the mRS scores.

Results: Mean age was 36.2 years (SD 7.9), sex ratio: male/female was 1/2. 85.7% of women were fertile. The most frequent neurological syndrome was intracranial hypertension. CT showed direct signs of dural sinuses thrombosis: (dense triangle sign in 5 pts, etc.). In 14 pts, we observed a venous cerebral infarct. MRI identified thrombosis of SSS in 31 pts, transverse sinus in 12 cases, cavernous sinus in 3 pts, cerebral edema in 21 pts. 9 out of 42 MRI had a normal CT. DSA revealed isolated cortical veins occlusion, without sinus occlusion in 3 cases. Risk factors were identified in 32 pts (76.2%); congenital thrombophilia being the most common (11 pts). All pts received anticoagulant therapy. After 90 days from admission, complete resolution of symptoms was seen in 22 cases, minimal neurological deficits in 14 pts, and the death rate was 14.3% (6 pts).

Conclusions: CVT appears to be under diagnosed in our region, due to low percentage of admissions for benign intracranial hypertension. CVT was common in women of fertile age. The outcome was favorable with adequate therapy.

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Abstract – WCN 2013**No: 139****Topic: 3 – Stroke****Diagnosis of carotid body paragangliomas by various imaging techniques**

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Introduction: Carotid body paragangliomas are rare hypervascular lesions arising from neural crest paraganglia cells, with a tendency of slow, but progressive growth, giving rise to external compression, or/and involvement of the carotid arteries, craniofacial nerves and other neighbouring structures.

Patients and methods: We analysed demographics, mode of presentation, imaging features, Shamblin classification, treatment modalities, and neurological complications (stroke, cranial nerve

injuries), of four patients (pts) with carotid body paragangliomas, in order to find specific signs and to elaborate a strategy for diagnosis and treatment.

Results: One pt had two localisations (the second was a glomus tumor of the left prelacrima sac) and another one had a family history for carotid body tumor. All lesions were paragangliomas of the carotid bifurcation, represented by painless lateral neck mass.

There was no evidence of functional tumor. The tumors were confirmed during ultrasonography (all four pts), MRI with MRA (all four pts) and DSA (2 pts). No preoperative embolisation was performed before complete resection of paragangliomas. Postoperatively, 2 pts had a transient twelve cranial nerve deficit. No stroke occurred.

Conclusions: Early diagnosis of carotid body paragangliomas is possible now with color Doppler sonography and MRI + MRA. Early surgery for paragangliomas minimized the risk of complications associated with large tumours.

doi:10.1016/j.jns.2013.07.654

Abstract – WCN 2013**No: 134****Topic: 3 – Stroke****Acute Korsakoff syndrome following bilateral parahippocampal gyrus infarction**

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Damage to the medial brain structures in the area of cognitive-behavioral disorders causes primarily memory disorders, drive (motivation) and affective behaviour. Ischemic stroke within the posterior cerebral artery may result in amnesic disorder resulting from ischemic hippocampal structures and adjacent neighborhoods.

Since Korsakoff syndrome was first described in 1887, the syndrome has been associated with thiamine deficiency. Almost all of the cases described in the literature were related to alcoholism, malnutrition or malabsorption, including persistent emesis.

There are limited case reports of structural lesions causing Korsakoff syndrome.

This report describes acute Korsakoff syndrome following bilateral infarction gyrus parahippocampalis.

63-year-old right-handed man, ex-white-collar worker was admitted to our hospital for an acute anterograde and retrograde amnesia with confabulations. He had a previous history of diabetes, hypertension, chronic heart disease NYHA II/III and paroxysmal atrial fibrillation but not of alcohol abuse.

CT imaging revealed old ischemic stroke in the area of right gyrus parahippocampalis and new ischemic stroke in the area of left gyrus parahippocampalis and occipito-temporal. Conservative treatment of stroke was enabled; patient got the thiamina from first day of hospitalization.

After one week of hospitalization patient developed advanced Korsakoff syndrome with apathy and periodically episodes of aggression. After a month, the patient was discharged home without improvement.

The patient had stayed for six months in the special ward of care and treatment, then his condition improved greatly and the patient returned home.

Mild memory disorders without confabulation remain and the patient needs to be taken care of by other people.

doi:10.1016/j.jns.2013.07.655

Abstract – WCN 2013**No: 156****Topic: 3 – Stroke****Plasma HDL as a predictor of aspirin resistance in stroke patients: A cross-sectional single centre study**

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Background: Aspirin use is known to reduce the recurrence of stroke. However, the clinical response to aspirin has been mixed. A plausible explanation for this is may be the resistance to the effects of aspirin. The causes of aspirin resistance are manifold and multi-factorial. We conducted a study to investigate inherent factors that may predispose towards aspirin resistance in a cohort of aspirin naïve stroke patients.

Methodology: This was a cross-sectional, observational study conducted on patients admitted to our centre with an acute stroke. Fifty consecutive patients were tested for biochemical aspirin resistance using Multiplate platelet analyser after 5 doses of aspirin, corresponding to a total dose of 900 mg.

Results: Aspirin resistance was present in 14% of our patients. There was an inverse relationship between the presence of aspirin resistance and plasma HDL levels ($r = -0.394$; $p = 0.005$). No relationship was observed between aspirin resistance and total cholesterol, triglycerides, LDL, HbA1c, ALT, ALP, urea and creatinine levels. There were no significant differences in the demographic profiles or smoking status between the aspirin-resistant and non-aspirin-resistant groups.

Conclusion: Our results indicate that a lower HDL level is associated with biochemical aspirin resistance. This may increase platelet aggregation and consequently increase the risk of a recurrent stroke. The clinical implications for aspirin resistance are far reaching. Any evidence that correctable factors may negatively influence the action of aspirin warrants further investigation.

doi:10.1016/j.jns.2013.07.656

Abstract -WCN 2013**No: 160****Topic: 3 -Stroke****Anti-beta 2-glycoprotein I autoantibody expression as a potential biomarker for strokes in patients with antiphospholipid syndrome**

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Backgrounds: Stroke patients and control subjects were recruited Kurdistan-Iraq between March 2004 and March 2005. All cases were under 50 years-of-age and had no recognizable risk factors. This study sought to determine the frequency rates of anti-cardiolipin (aCL) dependent on the presence of β_2 -GPI, anti- β_2 -glycoprotein I ($\alpha\beta_2$ -GPI), and anti-phosphatidyl serine (aPS) IgG autoantibodies among selected patients.

Methods: Using ELISA to evaluate the presence of IgG isotype of aCL, $\alpha\beta_2$ -GPI, and aPS autoantibodies in their blood.

Results: The results indicated that the frequency of $\alpha\beta_2$ -GPI was 14/50 (28%), aCL was 11/50 (22%), and aPS was 9/50 (18%) among stroke patients. In contrast, aCL was detected in 2/30 (6.7%) of the control subjects; each of the other anti-phospholipid antibodies (APLA) was never observed. Of all the $\alpha\beta_2$ -GPI⁺ cases, the incidence of stroke patients having the combined profile of $\alpha\beta_2$ -GPI + aCL was 11/14 (78.6%) and of $\alpha\beta_2$ -GPI + aPS was 9/14 (64.3%). Only 2/14 (14.3%) of these $\alpha\beta_2$ -GPI⁺

patients also expressed aCL in the absence of aPS. The frequency of patients expressing all three markers was only 9/14 (64.3 %). In none of the APS/stroke patients were aCL or aPS expressed in the absence of the $\alpha\beta_2$ -GPI. Conversely, IgG $\alpha\beta_2$ -GPI as a sole marker was seen in 3/14 (21.4%) of these patients (i.e. in the absence of either other marker).

Conclusions: It can be concluded that among the three major forms of APLA examined, the presence of IgG $\alpha\beta_2$ -GPI autoantibodies appeared to correlate best with stroke in patients who were concurrently suffering APS.

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Abstract – WCN 2013**No: 157****Topic: 3 – Stroke****Correlation between anticardiolipin antibodies level and stroke severity in antiphospholipid syndrome patients**

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Background: Serum antiphospholipid antibodies (aPLs) are useful to monitor the thrombotic risk in patients with antiphospholipid syndrome (APS). Our goal is to analyze the association of aPL level with stroke severity and outcome in AS patients.

Methods: Observational study that included consecutive young patients with ischemic stroke (IS) up to 55 years from 2006 to 2011. We analysed serum aPL levels including anticardiolipin antibodies (aCL), anti- β_2 -glycoprotein I (aGPI) and antiprothrombin (aPT) in the first 24 h after admission and, in the case of being positive, at least 12 weeks after. Stroke severity was measured by the NIHSS and 3-months stroke outcome by the modified Rankin Scale (mRS). Spearman's rho correlation was used to determine correlations between aPLs and stroke severity, as well as aPLs and outcomes in the APS group, in the whole sample and adjusted by age (≤ 45 and 46–55 years).

Results: A total of 222 patients were included; 63.5% male. In APS patients (9%), a positive correlation were found between IgM aCL and NIHSS ($\rho = 0.480$; $p = 0.032$). Adjusted by age, a positive correlation between IgM aCL and NIHSS ($\rho = 0.789$, $p = 0.011$) and a negative between IgG aCL and NIHSS ($\rho = -0.839$, $p = 0.005$) were observed only in patients >45 years old. There were not statistically significant correlations between aCL/other antibodies and mRS.

Conclusions: In APS patients with brain infarction, IgM aCL serum levels are positively correlated with stroke severity whereas IgG aCL serum levels are negatively correlated, overall in the >45 year old group.

doi:10.1016/j.jns.2013.07.658

Abstract – WCN 2013**No: 171****Topic: 3 – Stroke****Predictors of long-term survival among first-ever ischemic and hemorrhagic stroke in a Brazilian stroke cohort**

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Background: Although pathologic mechanisms for ischemic and hemorrhagic stroke are clearly distinct, most previous studies have failed to compare long-term prognosis including both ischemic and

hemorrhagic stroke. Despite the importance of investigating stroke survival, particularly in developing countries as Brazil that has one of the highest rates of hemorrhagic stroke in Latin America, publications in this field are sparse and come from developed countries.

Objective: Thus, we sought to investigate long-term survival and predictors that could influence adversely ischemic and hemorrhagic first-ever stroke prognosis along 4 years of follow-up.

Patients/material and methods: We prospectively ascertained 665 consecutive first-ever ischemic and hemorrhagic stroke cases from “The Study of Stroke Mortality and Morbidity” (The EMMA Study) in a community hospital in São Paulo, Brazil. The influence of cardiovascular risk factors and sociodemographic characteristics on stroke survival were analysed using life table survival and Cox proportional hazards survival analysis.

Results: We found a lower survival rate among hemorrhagic cases compared to ischemic stroke cases at the end of 4 years of follow-up (52% vs. 44%, $p = 0.04$). The risk of death is two times higher among people with ischemic stroke without formal education. Also, we found consistently higher risk of death for diabetics with ischemic stroke (HR = 1.45; 95% CI = 1.07–1.97) compared to no diabetics. As expected, age equally influenced on the high risk of poor survival, regardless of stroke subtype.

Conclusions: For ischemic stroke, the lack of formal education and diabetes were significant independent predictors of poor long-term survival.

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Abstract -WCN 2013

No: 172

Topic: 3 -Stroke

Comparison of aspirin response measured by urinary 11-dehydrothromboxane B2 and VerifyNow Aspirin assay in patients with ischemic stroke

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Background: There is little data about aspirin resistance in patients with ischemic stroke as compared with those with coronary artery diseases. The purpose of this research is to look for the prevalence of aspirin non-responders and to compare the results of the tests assessing aspirin responses between urinary 11-dehydrothromboxane B2 (dTXB2) measurement and VerifyNow® Aspirin assay in patients with ischemic stroke.

Methods: Patients with ischemic stroke who were treated with aspirin during April 2011–August 2011 were prospectively included. Aspirin response was assessed by urinary dTXB2 measurement and VerifyNow® Aspirin assay. The Spearman's correlation coefficients and kappa statistics were calculated to assess correlation and agreement between the 2 tests.

Results: 101 patients were studied. Prevalence of aspirin non-responders was 40% and 6%, if they were measured by urinary dTXB2 and VerifyNow® Aspirin assay, respectively. Poor correlation in the results between the 2 tests was found ($r = 0.135$, p -value 0.190). The degree of agreement between the 2 tests in relation to resistance status was weak ($\kappa = 0.032$, p -value = 0.590).

Conclusion: Our research confirmed the poor correlation and agreement between the 2 tests. Further research is still needed to identify the best method of diagnosis of aspirin resistance.

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Abstract – WCN 2013

No: 200

Topic: 3 – Stroke

Relevance of genetic polymorphisms in familial intracranial aneurysm in Brazilian population

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Background: Intracranial aneurysm (IA) risk factors can be environmental (smoking/alcoholism) and genetic like variants of endothelial nitric oxide sintase (eNOS), elastin (ELN) and endoglin (ENG). Objectives – analyze eNOS, ELN and ENG polymorphisms, environmental risk factors and association with IA.

Methods: 836 individuals in 6 groups: G1 – 40 (familial IA); G2 – 176 (G1 family); G3 – 113 (sporadic IA); G4 – 277 (G3 family); G5 – 104 (controls); and G6 – 126 (G5 family). Polymorphism analysis eNOS, ELN and ENG was done by PCR (polymerase chain reaction). Significance level $P < 0.05$.

Results: eNOS: allele A more prevalent in G1 (0.93), G2 (0.83), G3 (0.79), G4 (0.89) than G5 (0.61) and G6 (0.75; $P < 0.0001$). A/A genotype more frequent in G1 (86%); G2 (77%) G3 (79%) and G4 (78%), than G5 (26%) and G6 (50%; $P < 0.0001$). ELN: similarity between groups ($P > 0.05$). ENG: allele Wt more prevalent in G5 (0.81) than in G1 (0.61; $P = 0.01$); allele I more prevalent in G2 (0.30) than G6 (0.19; $P = 0.003$) and in G3 (0.34) than G5 (0.24; $P = 0.027$). Genotype –/Wt more frequent in G5 (89%) than G1 (69%; $P = 0.009$) and in G2 (77%) than G4 (88%; $P = 0.003$). Genotype I/I prevailed in G2 (22%), rather than G6 (8% $P = 0.001$). Smokers and alcoholics prevalent in G1 (79%; 40%, respectively) and G3 (61%; 36%), than G5 (29%; 20%; $P < 0.05$).

Conclusions: eNOS and ENG genetic polymorphisms are associated with IA, differentiating familial or sporadic IA and controls, as well as smoking and alcoholism.

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Abstract – WCN 2013

No: 242

Topic: 3 – Stroke

Stroke in patients with diabetes mellitus: A study from Northwestern Nigeria

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Background: In resource poor setting, identification of determinants of death is of overriding significance for clinicians in other that specific therapies and management strategies can be applied to patients at high risk of dying among diabetic mellitus (DM) with stroke.

The study was undertaken to describe the clinical characteristics, outcome pattern and predictors of mortality in a cohort of diabetic patients presenting with stroke in two tertiary hospitals in Northwestern Nigeria.

Materials and method: Between 2007 and 2011, persons with DM and stroke seen in emergency unit of the hospitals were recruited for the study. Each diagnosed patient was followed up for thirty days until either death at the hospital or discharge.

Result: Out of the 536 stroke patients, 85 (15.9%), comprising 48 (56.5%) males and 37 (43.5%) females with mean age of 56.1 ± 10.1 , had diabetes and 64 (75.3%) had infarctive stroke. One-month case

fatality rate was 30.6%. Out of the 26 death recorded 18 (69.2%) occurred in the first 72 h. Factors associated with death included male gender, TIA, abnormal respiratory pattern, abnormal pupillary size and reaction, elevated serum urea, hemorrhagic stroke, aspiration pneumonitis, and worsening GCS. Aspiration pneumonitis and worsening GCS were independent predictors of one month mortality of stroke in the patients.

Conclusion: Factors associated with mortality were male gender, TIA, abnormal respiratory pattern, abnormal pupillary size and reaction, elevated serum urea, hemorrhagic stroke, aspiration pneumonitis, and worsening GCS. Aspiration pneumonitis and worsening GCS were independent predictors of one month mortality of stroke in diabetic patients.

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Abstract – WCN 2013

No: 276

Topic: 3 – Stroke

Brain edema in the course of stroke, ways of correction

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Acute stroke is one of the most common pathologies in neurological practice and clinical medicine in general. As shows many studies, one of important links in pathogenesis of secondary neuronal damage is brain edema as a result of cerebral ischemia.

Purpose: Study effect of complex therapy with anti-edematous drugs on clinical course of disease with studying of secondary hemodynamic effects of acute stroke, comparative clinical dopplerography research of efficiency of using drug Lysine (L-lysine aescinate).

Materials and methods: Open, longitudinal study. 1st control group (20 patients) received basis therapy of stroke. 2nd comparison group (also 20 patients) received Lysine in 10.0 ml dose twice a day intravenously during 5 days and basis therapy of stroke.

Results: Patients age: 34 to 68 years (mean 62.2 ± 1.1). Clinical results of carotid lesion characterized by predominance of focal symptoms, impaired consciousness, secondary dislocation stem syndrome were found.

For objectification condition of patients regarding their neurological status were assessed using two complementary scales – Scandinavian and NIHSS. On NIHSS mean score was 19.3 ± 1.4 , and on Scandinavian scale – 30.0 ± 2.2 . Neurological deficits in the 2nd group on the tenth day on NIHSS decreased by 29.51%, Scandinavian scale increased by 24% ($p < 0.05$) compared to the 1st group.

Cerebral hemodynamics showed statistically significant improvement in arterial and venous components of blood flow.

Conclusions: Research shows that anti-edematous therapy is accompanied by significant improvement in the clinical course of the disease, rapidly decreases the perifocal edema zone, which is confirmed by reduced neurological deficit according to clinical scales and CT.

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Abstract – WCN 2013

No: 297

Topic: 3 – Stroke

Changes in serum interleukin-33 levels in patients with acute cerebral infarction

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Background: Inflammation is widely considered to be involved in the pathogenesis of cerebral ischemic injury. Interleukin-33 (IL-33) has been found to play very important roles in the inflammation of several human diseases such as asthma, inflammatory bowel disease and central nervous system inflammation. Its role in the pathology of acute cerebral infarction, however, has not been reported yet.

Objective: The aims of this study were to measure the levels of IL-33 in patients with cerebral infarction (CI), and examine the correlation of serum IL-33 level and the infarction volume.

Material and methods: Serum IL-33 level was assessed by sandwich enzyme-linked immunosorbent assay (ELISA) in 62 CI samples and compared with 15 healthy controls.

Results: Serum IL-33 levels in large and medium infarct volume patient groups were significantly higher than the control group ($p < 0.01$), but not between the small infarct volume group and the control group ($p > 0.05$). It was further found that the large infarct volume patient group had the highest serum IL-33 levels, which was significantly higher than medium and small infarct volume groups ($p < 0.05$). In addition, the medium infarct volume patient group had higher IL-33 levels than the small infarct volume group ($p < 0.05$).

Conclusion: Serum levels of IL-33 might be involved in the inflammatory responses in acute cerebral infarction patients, especially for severe cases and indicate that IL-33 might play a beneficial role in CI patients.

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Abstract – WCN 2013

No: 295

Topic: 3 – Stroke

Angioplasty and stenting for intracranial arterial stenosis

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Background and purpose: The stenosis of the major intracranial arteries is a major cause of cerebral stroke. We report our experience of endovascular treatment (percutaneous transluminal angioplasty and stenting; PTAS) for symptomatic stenosis of major intracranial arteries.

Material and methods: Totally 134 cases of symptomatic intracranial arterial stenosis were treated by PTAS at Kyoto Medical Center from 1998. Indication of PTAS was 1) symptomatic stenosis, which was not successfully treated by medication, and 2) more than 60% stenotic lesion shorter than 15 mm in length. Balloon angioplasty (BA) was performed first. Stenting was, then, performed in cases of 1) unsuccessful dilatation or arterial dissection after BA and 2) restenosis after BA.

Results: Successful dilatation by BA was obtained in 21/26 internal carotid artery (ICA) lesions, in 5/6 middle cerebral artery (MCA) lesions and in 24/29 vertebral artery (VA) lesions (50/61; 82%). Restenosis occurred in 7/26 ICA lesions, 4/6 MCA lesions and 8/29 VA lesions (totally 19/61 lesions; 31%). Successful dilatation by stenting was obtained in 59/60 ICA lesions, 1/1 MCA lesion and 13/15 VA lesions (totally 73/76 lesions; 96%). Symptomatic complications induced by PTAS occurred in two cases; cerebral stroke and intra cerebral hemorrhage. Stroke recurrence rate after PTAS was 1.1% /year of intracranial ICA lesion and 0.25% /year of intracranial VA lesion.

Conclusions:

- 1) Symptomatic intracranial arterial stenosis can be treated by PTAS with a low rate of stroke recurrence and complications.
- 2) Therapeutic merit of PTAS for intracranial lesion needs to be examined comparing to the recent aggressive medical therapy.

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Abstract – WCN 2013**No: 300****Topic: 3 – Stroke****Thyroid hormone contributes to hippocampal neural progenitor cell proliferation in adult 2vo rats**L. Ting^a, B. Chisulo^{a,b}, Q. Wang^a. ^aNeurology, Nanfang Hospital, Guangzhou, China; ^bMedicine, Kitwe Central Hospital, Kitwe, Zambia

Background: Thyroid hormone (TH) plays a critical role in developing the nervous system and regulating proliferation, survival and differentiation of neural progenitor cells. Hypothyroidism decreases the survival and differentiation of adult dentate granule cell progenitors.

Objective: To investigate the effects of TH on proliferation of stem cells in the hippocampal subgranular zone (SGZ).

Materials and methods: We achieved vascular cognitive impairment in animal model by permanent bilateral occlusion of common carotid arteries, set interventions of thyroid hormone treatment, and observe the effect on proliferation of adult dentate granule cell progenitors after chronic cerebral ischemia by immunofluorescence staining method.

Rats were administered with T₃ (10 mg/30 g·d) for 7 d or a single T₃ (10 mg/30 g) after ischemia, and same dose of saline in control animals. Neurogenesis was studied by Bromodeoxyuridine (BrdU) immunohistochemistry in the SGZ.

Results: T₃ treatment for 7 d after ischemia increased the number of BrdU cells in the SGZ compared to saline (p = 0.005, p < 0.001, respectively). A single T₃ injection 10 h before death also enhanced proliferation in SGZ 7d after chronic cerebral ischemia compared to saline (p = 0.015, p < 0.001, respectively).

Conclusion: The results suggest that T₃ enhance ischemia-induced proliferation of hippocampal in adult rats, and a short T₃ influence on mitotic activity suggested T₃ may act directly on progenitor cell populations.

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Abstract – WCN 2013**No: 301****Topic: 3 – Stroke****The CHA₂DS₂-VASC score reflects clinical outcomes in NVAf patients with an initial cardioembolic stroke**

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Background: Whether the CHA₂DS₂-VASC score reflects severity or clinical outcomes in patients with an initial cardioembolic stroke associated with non-valvular atrial fibrillation (NAVF) was investigated.

Patients and methods: This study included 327 patients hospitalized between April 2007 and March 2012 for an initial cardioembolic stroke associated with NVAf with no prior history of stroke. The NIHSS score on admission and clinical outcome (mRS score after 90 days) was retrospectively evaluated according to the CHA₂DS₂-VASC score.

Results: CHA₂DS₂-VASC scores were: 0, 3.1%; 1, 9.1%; 2, 24.5%; 3, 26%; 4, 20.8%; 5, 14.4%; and 6, 2.1%. The median NIHSS scores for CHA₂DS₂-VASC scores of 0–6 were 4.5, 8, 8, 10, 11, 17, and 23, respectively. Severity differed according to CHA₂DS₂-VASC score. The clinical outcomes according to the CHA₂DS₂-VASC scores were: score 0, mRS score 0–2 (80%) and 3–6 (20%); score 1, mRS score 0–2 (80%) and 3–6 (20%); score 2, mRS score 0–2 (64%) and 3–6 (36%); score 3, mRS score 0–2 (48%) and 3–6 (52%); score 4, mRS score 0–2 (28%) and 3–6 (72%); score 5, mRS score 0–2 (26%) and 3–6 (74%); and score 6, mRS score 0–2 (29%) and 3–6 (71%). The clinical outcome worsened as the CHA₂DS₂-VASC score increased.

Conclusion: The severity of NVAf-induced initial cardioembolic stroke increased with higher CHA₂DS₂-VASC scores, and outcomes were poor. The present study suggests that the CHA₂DS₂-VASC score may be useful not only for the evaluation of stroke risk, but also for the prediction of clinical outcomes after stroke.

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Abstract – WCN 2013**No: 334****Topic: 3 – Stroke****Functional recovery after ischemic stroke – A matter of age. Results from the Austrian stroke unit registry**M. Knoflach^a, B. Matosevic^a, M. Rucker^a, M. Furtner^a, A. Mair^a, G. Wille^a, A. Zangerle^a, P. Werner^b, J. Ferrari^c, C. Schmidauer^a, L. Seyfang^d, S. Kiechl^a, J. Willeit^a. Austrian Stroke Unit Collaboration. ^aNeurology, Medical University Innsbruck, Innsbruck, Vienna, Austria; ^bNeurology, Hospital Rankweil, Rankweil, Vienna, Austria; ^cNeurology, Hospital Barmherzige Brueder, Vienna, Austria; ^dDanube University Krems, Gesundheit Österreich GmbH/BIQG, Vienna, Austria

Objective: To analyze the association between patient age and good functional outcome after ischemic stroke with special focus on the young that were numerically underrepresented in previous evaluations.

Methods: Of 43,163 ischemic stroke patients prospectively enrolled in the Austrian Stroke Unit Registry, 6084 (14.1%) were ≤55 years old. Functional outcome was available in a representative subsample of 14,256 patients free of pre-stroke disability, 2223 were 55 years or younger. Herein we analyzed the effects of age on good functional outcome 3 months after stroke (modified Rankin Scale ≤ 2).

Results: Good outcome was achieved in 88.2% (unadjusted probability) of young stroke patients (≤55 years). In multivariable analysis, age emerged as a significant predictor of outcome independent of stroke severity, etiology, performance of thrombolysis, sex, risk factors and stroke complications. When using the age stratum 56–65 as a reference, odds ratios [95%CI] of good outcome were 3.4 [1.9–6.4], 2.2 [1.6–3.2] and 1.5 [1.2–1.9] for patients aged 18–35, 36–45 and 46–55 and 0.70 [0.60–0.81], 0.32 [0.28–0.37] and 0.18 [0.14–0.22] for those 66–75, 76–85 and >85 years old (P < 0.001). In absolute terms, the regression-adjusted probability of good outcome was highest in the age group 18–35 and gradually declined by 3.1%–4.2% per decade until age 75 with a steep drop thereafter. Findings equally applied to sexes and patients with and without intravenous thrombolysis or diabetes.

Conclusions: Age emerged as a highly significant inverse predictor of good functional outcome after ischemic stroke independent of stroke severity, characteristics and complications with the age-outcome association exhibiting a non-linear scale and extending to young stroke patients.

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Abstract – WCN 2013**No: 353****Topic: 3 – Stroke****Flow-mediated dilatation in ischemic leukoaraiosis**

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Background: Although the exact pathophysiology of ischemic leukoaraiosis (LA) remains to be elucidated it is possible that endothelial dysfunction may play the essential role. Therefore we explored systemic endothelial dysfunction in ischemic leukoaraiosis.

Methods: In order to evaluate systemic endothelial function in subjects with LA (LA1) we examined flow-mediated dilatation (FMD) and compared it to subjects with comparable risk factors without LA (LA0). FMD was investigated in 30 subjects with LA (57.6 ± 6.8 years) and 24 subjects with comparable risk factors without LA (55.7 ± 5.4 years).

Results: FMD was significantly lower in group LA1 compared to LA0 (4.7 ± 3.2 % vs. 7.3 ± 3.7 %; $p = 0.009$). There were no statistically significant differences in the FMD response when subjects from the group LA1 were divided into the subgroup with arterial hypertension and the one without arterial hypertension (4.1 ± 3.2 % vs. 5.4 ± 3.3 %; $p = 0.307$). When the group LA1 were divided according to the leukoaraiosis into Fazekas 1, 2 or 3 groups. When the Fazekas 2 and 3 subgroups were compared to each other, we found that FMD response was significantly different ($p = 0.013$) and even so when the Fazekas 1 subgroup was compared to the Fazekas 3 subgroup ($p = 0.019$). Multiple regression analysis showed only an important influence of LA on FMD ($p < 0.05$).

Conclusions: It seems that systemic endothelial function is diminished in ischemic leukoaraiosis. According to our study it is possible that systemic endothelial dysfunction is one of important aspects of pathophysiology of LA.

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Abstract – WCN 2013

No: 289

Topic: 3 – Stroke

Clinico-epidemiological features of cerebral venous thrombosis in Algarve

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Background: Cerebral vein thrombosis (CVT) is a very uncommon disorder with a wide variety of clinical manifestations. Little information exists about CVT, particularly from peripheral regions.

Objective: To describe the epidemiology and clinical characteristics of CVT in a relatively small region.

Material and methods: Hospital based retrospective study of CVT occurred between January 2001 and December 2012, in the public hospitals serving a population of 395,218–451,005 inhabitants.

Results: Thirty-five cases of CVT were identified. Children (4) and non-residents (3) were excluded from the analysis. Gross incidence was approximately 0.56–0.64/100,000 cases/year. The mean and median ages were 40.4 and 35 years respectively [18–71 years]. The majority (21/75%) were woman. The mean interval between the onset of symptoms and the diagnosis was 8 days [8–41 days]. Patients presented with isolated intracranial hypertension (17/61%), focal syndrome (4/14%), and encephalopathy (7/25%). Brain TC was diagnostic in 10 (42%) cases. Brain MRI and MR venography was performed in all. Use of oral contraceptives was found in 14/66 % of females, with/without other risk factors. In 6 (21%), no risk factor was found. All cases received heparin or LMWH followed by warfarin. Complete recovery occurred in 24 (85 %); death in 1 (3.6%); important sequels in 3 (11%): epilepsy, severe visual loss and motor deficit.

Conclusion: Our findings are comparable to those described in similar case series. However, in comparison to what has been found in our country (0.22/100,000 cases/year), our incidence is high. Diagnosis improvement, availability of MRI, and use of oral contraceptives may explain this.

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Abstract – WCN 2013

No: 325

Topic: 3 – Stroke

The value of apparent diffusion coefficient maps in subcortical infarct lesion

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Background: Prediction of the early neurologic deterioration (END) in patients with acute ischemic stroke is one of great clinical interests.

Objective: This study was designed to analyze whether apparent diffusion coefficient (ADC) values provide reliable quantitative information for the prediction of the neurologic deficits in stroke patients.

Patient and methods: We reviewed 36 patients not treated by thrombolytics who had a diffusion restricted lesion in deep subcortical area within 24 h after stroke onset. Serial National Institutes of Health Stroke Scale (NIHSS) scores were checked from admission. We compared the ADC values in the initial diffusion weighted image (DWI). We tested the value of the ADC to predict tissue outcome and development of END. All images were processed on an independent workstation by using custom software (MIDAS 1.11).

Results: Among 36 patients, 14 patients developed END and 22 patients did not develop END. Univariate analysis showed that the ADC values in diffusion restricted lesion were significantly decreased in patient that developed to END compared with those that remained without END. (p value = 0.037).

Conclusions: Initial ADC values can predict the development of END in patient with deep subcortical infarct.

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Abstract – WCN 2013

No: 266

Topic: 3 – Stroke

The association of infarct pattern on coronal diffusion-weighted MR image and early neurologic deterioration in deep subcortical infarction

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Background: Early neurological deterioration (END) is frequently observed in patients with small deep infarction of subcortical area.

Objective: In this study, we investigated the association between the END and the patterns of small deep infarction in coronal diffusion-weighted imaging (DWI).

Patients and methods: Consecutive 95 patients diagnosed with acute ischemic stroke in subcortical area, who were admitted to stroke center between March 2008 and June 2009, were included. Brain coronal DWI and MR angiography (MRA) were carried out. The patterns of infarct lesions are divided into 3 categories: (1) lacunar pattern; isolated lesion in the subcortical area, (2) skipped pattern: lesions that presented with disconnected shape, separated by unaffected white matter of internal capsule, on coronal DWI, (3) linear pattern; linear lesion extending to the basal surface on coronal DWI. END was defined as an increase of 2 or more points of the National Institutes of Health Stroke Scale (NIHSS) within 7 days of stroke onset.

Results: 23 patients were presented with lacunar pattern, 20 with skipped patterns, and 52 patients with linear pattern. END was observed in 36 (45.8%). Patients with skipped and lacunar patterns on coronal DWI experienced END less frequently than those with linear patterns (25% vs. 55.8% and 8.7% vs. 55.8%, $p = 0.034$ and 0.000, respectively).

Conclusions: More frequent ENDS have been reported in linear patterns than skipped and lacunar patterns in small deep infarction. We found the usefulness of coronal DWI in predicting early clinical course and in depicting small deep infarction.

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Abstract – WCN 2013

No: 328

Topic: 3 – Stroke

Mortality-related factors in ischemic stroke patients 80 years of age and older

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Background: To date, many variables that affect the early and late prognosis in stroke patients have been reported. Many of these factors cannot be changed, such as oldest age. It is important to determine the changeable and non-changeable factors related to disability and death in the oldest age groups.

Objective: The aim was to investigate mortality-related factors in ischemic stroke patients 80 years of age and older.

Methods: We reviewed all ischemic stroke patients admitted to our clinic between January 2010 and January 2012. The patients' database information was retrospectively analyzed. One hundred and ten patients aged older than 80 years with ischemic stroke were included in the study. The patients were divided into two groups based on survival. Age, gender, recurrent stroke, risk factors, clinical syndrome, etiology, radiographic localization, duration of hospitalization, and presence of systemic complications were accepted as mortality-related prognostic factors. The groups were compared according to these prognostic factors.

Results: In the clinical follow-up, 58 (52.7%) patients died; 65.5% died of neurological causes, and 31% died of systemic complications. No significant differences existed between the two groups in age, gender, risk factors, recurrent stroke, or etiology. The frequency of total anterior circulation infarct syndrome was much higher in deceased than living patients (50% and 36.5% respectively; $p < 0.05$). Deceased patients had a statistically significantly higher incidence of total MCA infarct and systemic complications than did living patients ($p < 0.05$).

Conclusion: Starting appropriate treatment and care initiatives as soon as possible is also very necessary in the oldest stroke patients.

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Abstract – WCN 2013

No: 337

Topic: 3 – Stroke

Prevalence of cardioembolic stroke increased significantly in Chinese population in the past 10 years

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Background: Atrial fibrillation-related cardioembolic stroke (AF-CE) is a major cause of cardioembolism, which may be potentially preventable with anti-coagulation.

Objective: The aim of the study was to evaluate the changes in the prevalence of AF-CE over the past ten years in Chinese population.

Patients and methods: We evaluated the prevalence of AF-CE and risk factor profile from data collected prospectively from Prince of Wales Hospital Stroke Registry in 1999, 2004 and 2009.

Results: Total 2744 patients were admitted for ischaemic stroke or transient ischaemic attack in these three years – 946 patients in 1999, 887 in 2004 and 911 in 2009. There was no significant difference in the mean age of patients (71.2 ± 11.3 years, 70.7 ± 12.8 years, and 70.8 ± 12.5 years respectively, $p = 0.644$). The number of AF-CE increased significantly – 92 patients (9.7%) in 1999, 104 patients (11.7%) in 2004 and 216 patients (23.7%) in 2009 ($p < 0.001$).

Among patients with atrial fibrillation, the mean CHADS score prior to the index stroke was 2.7 ± 1.3 in 1999, 3.0 ± 1.3 in 2004 and 2.2 ± 1.3 in 2009. The percentage of patients who were on warfarin before admission was 21.7% in 1999, 23.1% in 2004 and 13.0% in 2009.

Conclusion: Over the past 10 years, the prevalence of AF-CE has increased significantly in Chinese population. Although most of the patients with atrial fibrillation had CHADS score ≥ 2 , only a minority of them were anticoagulated before admission. Raising the awareness of this potentially preventable stroke subtype is warranted in Chinese population.

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Abstract – WCN 2013

No: 369

Topic: 3 – Stroke

The safety and efficacy of the intravenous recombinant tissue plasminogen activator for the ischemic stroke patients in community based hospital

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Background: The results of the NINDS r-tPA Stroke Trial generated considerable hope, but also concerns about whether the results of the NINDS r-tPA Stroke Trial can be replicated in routine clinical practice. We studied whether r-tPA infusion could be applied in a community based hospital with safety and efficacy.

Methods: We analyzed retrospectively the data of thirty-three patients with acute ischemic stroke treated with intravenous r-tPA, from Feb. 2003 to Dec. 2006. Safety was evaluated by intracranial hemorrhage, symptomatic intracranial hemorrhage and mortality. Clinical neurologic status was measured by NIHSS at baseline, 24 h, and 7 days after r-tPA treatment. Efficacy was assessed by the response rate of r-tPA using improvement of NIHSS by 4 or more points at 24 h after treatment and the long term outcome with the modified Rankin scale at 3 months after stroke.

Results: The Median NIHSS was 18. Mean onset to needle time was 140 ± 30 min. Among the 33 patients, 10 patients had intracranial hemorrhage. Two patients had symptomatic intracranial hemorrhage but no death occurred. 15 patients showed improvement of NIHSS by 4 or more points at 24 h after r-tPA. 12 patients showed good outcome at 3 months. 9 patients had no or minimal symptoms, 7 patients had mild to moderate disability, 10 patients had severe disability and 7 patients were died.

Conclusions: The efficacy and safety of intravenous r-tPA for acute ischemic stroke in the community based hospital mirror the results of the NINDS stroke trial.

doi:10.1016/j.jns.2013.07.675

Abstract – WCN 2013**No: 361****Topic: 3 – Stroke****Demographics and risk factors for recurrent ischemic stroke in Aegean part of Turkey***Y. Seçil, Y. Çiftçi, Y. Beckmann. Neurology, Izmir Katip Çelebi University, Atatürk Education and Research Hospital, Izmir, Turkey*

Background: Recurrent stroke is defined as a new cerebrovascular event which occurs after the stabilization of the previous stroke. Recurrence of stroke increases likelihood of disability–mortality associated with stroke. Systematic evaluation of stroke cases can help to reduce the risk of recurrence.

Objective: In order to predict ischemic strokes which carry the risk of recurrence, our study aimed to compare data related to risk factors, stroke type, etiology and disability–mortality rates associated with stroke.

Material and method: Our study including patients with recurrent ischemic stroke admitted to our hospital Neurology department between February 2010 and June 2011. Information about previous strokes and demographics were recorded. Risk factors were classified. Disability–mortality rates and secondary prevention treatment at recurrent stroke onset were evaluated.

Results: 151 patients with recurrent ischemic stroke were included in our study. The mean age was 71.35 ± 12.06 . The most frequent risk factors were hypertension, hyperlipidemia, coronary artery disease and atrial fibrillation. The most frequent recurrence etiology was cardiac embolic events. 53.2% of patients were receiving antiplatelet, 10.4% of patients were receiving anticoagulation therapy. 26.5% of patients died. The mean of modified Rankin score of patients was significantly higher at the recurrent ischemic stroke than the previous ischemic stroke.

Conclusion: Knowing the etiological factor can help to predict recurrence of stroke and prevent death. Effective treatment of modifiable risk factors in recurrent stroke groups is very important in primary and secondary stroke prevention. Recurrent stroke prevention is very important in prognosis, disability and financial basis all around the world.

doi:10.1016/j.jns.2013.07.676

Abstract – WCN 2013**No: 292****Topic: 3 – Stroke****Clinicoradiological localization of language dysfunctions in acute ischemic brain attack***M.H. Ibrahim. Neurology, GMC & GMU University, Ajman, United Arab Emirates*

Objective: The aim of the study was to study the effect of cerebral ischemic vascular insult on the localization of language areas as regard automatic speech, initiation and mutism.

Materials and methods: Seventy ischemic stroke patients selected within the first 48 h of the onset of stroke with left hemispheric stroke with aphasia. Inclusion criteria: At any age above seven years, both sexes, first ever clinical stroke, admitted within the first 7 days of stroke insult. Patients underwent careful history taking, neurological examination, routine laboratory investigations, echocardiography, carotid duplex and language assessments.

Results: Automatic speech disorders were seen in (76%) patients and their MRI lesion distribution showed lesions within the deep periventricular white matter and anterior Perisylvian regions and other areas. Initial difficulty was with lesions in deep periventricular white matter, and other areas. Mutism significantly correlated with lesions in temporal mid-area, external capsule and other areas.

Conclusions: Deep periventricular white matter and insular area in the dominant hemisphere play an important role in many language tasks. Thus aphasia is not a mere cortical function.

doi:10.1016/j.jns.2013.07.677

Abstract – WCN 2013**No: 388****Topic: 3 – Stroke****How accurate are clinicians when estimating stroke outcomes? Results from the Jurassic randomized study***G. Saposnik^a, R. Cote^b, R. Raptis^c, M. Mamdani^a, K. Thorpe^a, J. Fang^d, D. Redelmeier^a, L.B. Goldstein^e JURASSIC Study Group**^aUniversity of Toronto, Toronto, Canada; ^bMcGill University, Montreal, Canada; ^cSt Michael's Hospital, Toronto ON, Canada; ^dInstitute for Clinical Evaluative Sciences (ICES), Toronto ON, Canada; ^eDuke University, Durham NC, USA*

Objective: We compared the accuracy of clinicians and a risk score (iScore) to predict observed outcomes following an acute ischemic stroke.

Methods: The JURASSIC (Clinician **J**udgment vs. **R**isk **S**core to predict **S**troke **o**ut**C**omes) study assigned 111 clinicians with expertise in acute stroke care to predict the probability of outcomes of 5 ischemic stroke case scenarios. The primary outcome was prediction of death or disability (mRS ≥ 3) at discharge within the 95% confidence interval (CI) of observed outcomes. Secondary outcomes included 30-day mortality and death or institutionalization at discharge.

Results: Clinicians made 1661 predictions with overall accuracy of 16.9% for death or disability at discharge, 46.9% for 30-day mortality, and 33.1% for death or institutionalization at discharge. In contrast, 90% of the iScore-based estimates were within the 95% CI of observed outcomes. Nearly 50% of participants were unable to accurately predict the probability of the primary outcome in any of the 5 rated cases. In multivariable analyses, the presence of patient characteristics associated with poor outcomes (mRS ≥ 3 or death) in previous studies (older age, high NIHSS score and non-lacunar subtype) were associated with more accurate clinician predictions of death at 30-days (OR 2.40, 95%CI 1.57–3.67) and with a trend for more accurate predictions of death or disability at discharge (OR 1.85, 95%CI 0.99–3.46).

Conclusions: Clinicians with expertise in stroke performed poorly compared to a validated tool in predicting stroke outcomes. Use of the risk stroke outcome tool may be superior for decision-making following an acute ischemic stroke.

doi:10.1016/j.jns.2013.07.678

Abstract – WCN 2013**No: 437****Topic: 3 – Stroke****Vascular endothelial growth factor: Genetic polymorphisms in patients with intracranial aneurysm and its relation to hypertension and diabetes mellitus***T.D.B. Maluf^a, M.L. Gregorio^b, M.A.D.S. Pinhel^b, M.L.T. Santos^c, M.A. Nakazone^d, J.R.L. Ferraz-Filho^e, W.A. Tognola^f, D.R.S. Souza^b.**^aBiochemistry and Molecular Biology, Sao Jose do Rio Preto Medical School, Sao Jose do Rio Preto, Brazil; ^bMolecular Biology, Sao Jose do Rio Preto Medical School, Sao Jose do Rio Preto, Brazil; ^cNeurosurgery, Hospital School/Sao Jose do Rio Preto Medical School, Sao Jose do Rio Preto, Brazil; ^dCardiology, Sao Jose do Rio Preto Medical School, Sao Jose do Rio Preto, Brazil; ^eNeuroradiology, Hospital School/Sao Jose do Rio Preto Medical School, Sao Jose do Rio Preto, Brazil; ^fNeurology, Sao Jose do Rio Preto Medical School, Sao Jose do Rio Preto, Brazil*

Background: Vascular Endothelial Growth Factor (VEGF) is an important regulator and genetic marker of vasculogenesis and angiogenesis. VEGF polymorphisms are considered as risk factors for intracranial aneurysm (IA). Objective: Analyze VEGF polymorphism, lifestyle and comorbidities in the Brazilian casuistic with IA.

Methods: 856 individuals distributed into groups: G1 – 43 (familial IA); G2 – 177 (G1 family); G3 – 120 (sporadic IA); G4 – 285 (G3 family); G5 – 104 (controls); G6 – 127 (G5 family). VEGF-C936T was analyzed by PCR. $P < 0.05$ was considered statistically significant.

Results: Prevalence of genotype wild C/C in G5 (10.5%) in comparison with G1 (2.3%) and G3 (0.0%) ($P = 0.03$ for both) and risk genotype (T/T) was significant between G4 (75.3%) and G6 (78.0%; $P < 0.0001$). The distribution of allele T was similar between groups ($G1 = 0.86$; $G2 = 0.88$; $G3 = 0.87$; $G4 = 0.87$; $G5 = 0.81$; $G6 = 0.89$; $P > 0.05$). We observed a high frequency of smoking and drinking habits in G1 (79% and 40%, respectively) compared to G5 (30.0% and 22%, respectively; $P < 0.0001$ for all). An association between hypertension and IA was observed in G1 (49%) and G3 (49%) compared to G5 (21%; $P = 0.003$; $P < 0.0001$). Differences between patients and controls in relation to diabetes mellitus were not observed.

Conclusion: The polymorphism for VEGF was more prevalent only in the family of patients with IA. On the other hand, smoking, drinking habits and hypertension were observed in Brazilian patients with IA. Studies in families with a history of IA may allow greater attention to preventive care with respect to the disease.

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Abstract – WCN 2013

No: 452

Topic: 3 – Stroke

Nanowired cerebrolysin attenuated exacerbation of blood–brain barrier breakdown, edema formation and brain pathology after heatstroke in diabetic and hypertensive rats

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Nitric oxide synthase (NOS) upregulation in CNS is common in hypertension or diabetes. Our previous works showed neuronal and inducible NOS upregulation in rat brain after 4-h heat stress (HS) at 38 °C that correlated well with blood–brain barrier (BBB) breakdown, edema formation and neuronal damage.

Possible exacerbation of brain pathology and edema formation in diabetic-hypertensive (DBHY) animals after heat stroke (HS) was examined in a rat model. The role of nitric oxide (NO) in exacerbation of HS-induced brain pathology was also evaluated using nitric oxide synthase (NOS) immunoreactivity. Hypertensive rats (produced by two-kidney one clip (2K1C) method) were made diabetic with streptozotocine (50 mg/kg, i.p./day for 3 days) treatment. After 6 weeks, DBHY rats show 20–30 mM/L Blood Glucose and hypertension (180–200 mmHg). Subjection of these rats to 4-h HS resulted in 6- to 8-fold higher BBB breakdown, brain edema formation and brain pathology. At this time, neuronal or inducible NOS expression was 4 to 6-fold higher in DBHY rats compared to controls. Interestingly, iNOS expression was higher than nNOS in DBHY rats. Cerebrolysin in high doses (10 ml/kg, i.v. instead of 5 ml/kg) or nanowired cerebrolysin (5 ml/kg) induced significant neuroprotection and downregulation of nNOS and iNOS in DBHY animals whereas normal animals need only 5 ml/kg doses for this purpose. Our observations demonstrate that co-morbidly factors exacerbate brain damage in HS through NOS expression and require either double dose of

cerebrolysin or nanowired cerebrolysin for neuroprotection as compared to normal rats.

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Abstract – WCN 2013

No: 482

Topic: 3 – Stroke

Attenuation of glutamate mediated neuronal insult by piroxicam in animal model of focal cerebral ischemia: Possible involvement of GABA agonism

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To explore the importance of brain neurotransmitters in the rodent model of focal cerebral ischemia, we evaluated the effects of Piroxicam, a nonsteroidal anti-inflammatory drug (NSAID) on extracellular brain glutamate and γ -aminobutyric acid (GABA) release, survival time and neuronal cell death. Focal cerebral ischemia led to neuronal infarction and also compromised intrinsic antioxidant status. Thirty minutes pre-administration of Piroxicam (10 mg/kg b.w) showed a significant ($P < 0.01$) reduction in cerebral infarct volume and potentiation of the intrinsic antioxidant status as well. Further, high performance liquid chromatography assay was performed to measure changes in extracellular concentrations of neurotransmitters which were found to be 0.519 ± 0.44 pmol/mg; 1.18 ± 0.28 pmol/mg; 0.63 ± 0.21 pmol/mg respectively while hydroxyl radical (.OH) adduct of salicylate in the frontal cortex and striatum in normal, stroke and drug treated rat models were found to be 0.261 ± 0.06 pmol/mg; 0.68 ± 0.52 pmol/mg; 0.401 ± 0.68 pmol/mg respectively. After the onset of cerebral ischemia, the extracellular level of glutamate in rat brain increases continuously as compared to that of control group. However, by administration of Piroxicam in stroke rat, the elevated extracellular cerebral glutamate was found to be significantly ($P < 0.05$) reduced. The data indicates for the first time that Piroxicam, a NSAID attenuates extracellular glutamate release and also reduces neuronal cell death due to reduction in oxidative stress in cerebral ischemia.

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Abstract – WCN 2013

No: 462

Topic: 3 – Stroke

Hemichorea development after stroke

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Hemichorea is uncommon in cerebrovascular events. Its appearance is in relation to the involvement contralateral of the basal ganglia and thalamus. The first patient we presented was a 71-year-old female and she developed an acute onset right hemichorea. Her cranial MR imaging (MRI) showed a right putaminal infarction. The second patient was a 70-year-old female presented with left hemichorea that developed 15 days ago before admission. Her cranial MRI revealed a right basal ganglia infarction. The third patient was a 57-year-old male with acute onset left hemichorea. His cranial MRI revealed infarctions of right thalamus and caudate nucleus. The fourth patient was a 67-year-old female with right hemichorea as a result of a thalamic haematoma. The pathophysiology of the vascular chorea was discussed by the clinical and imaging findings in these four patients.

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Abstract – WCN 2013**No: 463****Topic: 3 – Stroke****Aetiopathogenesis of cerebral ischemia in young Turkish patients**

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This study evaluates the risk factors and etiological spectrum in young stroke patients (aged 45 or younger).

Methods: Risk factors and aetiology of cerebral ischemia were retrospectively investigated in 230 patients (age ranges 9–45), admitted to our Neurology Unit with cerebral transient ischemic attacks (TIA) or infarction. 230 consecutive patients were divided into two groups according to their ages. Group I consists of patients whose age ranges 9–30 and in Group II, age range was 30–45.

Results: Hypertension, hyperlipidemia, smoking were the most common risk factors. Atherosclerosis was the leading etiology occurring in group II (35%) and vasculopathy was markedly high in group I (23%). In addition cardiac embolism was determined in 20% of all cases. Among these cases cardiac valve disease (58%), atrial fibrillation (16%) and ischemic heart disease (27%) were determined by the investigation methods.

Conclusion: The most striking finding of our study was the high frequency of cardiac valve disease in young adult ischemic strokes in our country when compared with the previous reports in the literature.

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Abstract – WCN 2013**No: 465****Topic: 3 – Stroke****Predictors of mortality in patients with craniocervical artery dissection**

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Background: Dissection of the carotid (CAD) or vertebral (VAD) artery is a major cause of ischemic stroke in young adults. The prognosis varies and the dissection can range from asymptomatic to causing a profound neurological deficit and death. Little is known of the predictors of mortality in these patients.

Objectives: The aim of the study was to investigate the predictors related to mortality in patients with craniocervical artery dissection.

Material and methods: We reviewed retrospectively all patients diagnosed with CAD or VAD admitted to our clinic between January 2000 and January 2013 and included 67 such patients in the study. Age, gender, modified Rankin Scale (mRS) prestroke and at time of admission, clinical presentation of dissection, location of dissection, vascular risk factors, and length of hospital stay were considered mortality-related prognostic factors. Of the 67 patients, 12 (17.9%) patients died: 5 (7.46%) patients with CAD and 7 (10.44%) with VAD. The patients were divided into four subgroups: surviving patients with CAD, dying patients with CAD, surviving patients with VAD, and dying patients with VAD. Then, surviving-CAD versus dying-CAD and surviving-VAD versus dying-VAD compared according to prognostic factors.

Results: Disability at admission (mRS 3–5) was significantly higher in dying-CAD than surviving-CAD. Comparing dying-VAD with surviving-VAD, the mean age, disability at admission (mRS 3–5), presentation with stroke, and dissection location in intracranial vertebral artery were significantly higher in dying-VAD.

Conclusion: Identifying predictors of mortality is important for starting appropriate treatment and setting targets in the management of patients with dissection.

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Abstract – WCN 2013**No: 456****Topic: 3 – Stroke****Heat stress induces selective cell damage in the cerebellum. An experimental study using immunohistochemistry and ultrastructural investigations in rats**

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The possibility that thermal stress caused by systemic heat exposure may lead to a specific cellular damage in the cerebellum was examined in young rats (age 8–9 weeks). The cell changes in the cerebellum were examined using immunohistochemistry and light and electron microscopy.

Subjection of *conscious* young animals to a 4-h HS resulted in marked hyperthermia (41.6 ± 0.45 °C). In these animals the cerebellum showed pronounced and selective cellular damage in the Purkinje cell layers and granule cells. This damage was most marked in specific regions of the vermis and in the lateral cerebellar cortices. Deeper cerebellar nuclei also showed marked cell damage. Immunohistochemistry showed profound upregulation of GFAP, S-100 and Vimentin indicating abnormal glial cell reactions in the cerebellum. Marked decrease in MBP suggests profound degradation of myelinated nerve fibers. Electron microscopy showed marked damage to the cerebral endothelium. Lanthanum an electron dense particle was found to be present within the endothelial cells, basal lamina and in vesicular profiles. Nerve cells showed abnormal reactions, edema and distortion in many parts of the cerebellum. Loss of Nissl substance was much pronounced in several regions of the cerebellum. These nerve, glial and myelin cell changes were less evident in animals subjected to similar heat stress under anesthesia. These observations for the first time show that cerebellum is vulnerable in heat stress and this selective vulnerability is somehow associated with a specific breakdown of the BBB permeability.

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Abstract – WCN 2013**No: 458****Topic: 3 – Stroke****Cardiac arrest alters regional ubiquitin levels in the porcine brain. Neuroprotective effects of methylene blue**

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Ubiquitin is recently implicated the central nervous system (CNS) diseases. Overexpression of ubiquitin occurs in stroke, trauma, Alzheimer's disease, Parkinson's disease and Huntington's disease in the areas associated with neuronal damages. However, the role of ubiquitin in brain pathology or neuroprotection is still not well known. Cardiac arrest (CA) and subsequent cardiopulmonary resuscitation induces widespread brain damages. In a porcine model of CA our laboratory showed marked neuronal, glial and myelin damages in

the cortex, hippocampus, cerebellum, thalamus, hypothalamus and the brain stem following 30 to 180 min after return of spontaneous circulation (ROSC) in pig model. Interestingly these neuropathological changes in CA were considerably reduced by pretreatment with methylene blue (MB), an antioxidant dye following 60 min to 180 min after ROSC. Thus, it would be interesting to see whether ubiquitin levels are also altered after CA and/or ROSC in identical brain areas and this effect could be further influenced by MB treatment.

Our observations using ELISA showed selective and specific increase in ubiquitin levels in most of the above brain areas immediately after untreated CA and progressively from 30 min to 180 min after ROSC. Cerebellum and cortex showed most pronounced increase in ubiquitin levels. Pretreatment with MB during cardiopulmonary resuscitation significantly attenuated ubiquitin levels from 60 min on to 180 min after ROSC. These observations suggest that increased levels of ubiquitin are related with brain ischemic pathology and a reduction in this level is associated with neuroprotection in CA, not reported earlier.

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Abstract – WCN 2013

No: 472

Topic: 3 – Stroke

Indian Aphasia battery: Tool for specific diagnosis of language disorder post stroke

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Background & aims: Aphasia is a language disorder which may disrupt an individual's functioning. To plan a mode of therapeutic/rehabilitative work, it is important to assess problems from neuropsychological perspective focussed on remediation of the impaired processes or compensation via the intact processes or both (Hillis AE, 2001). It targets sequence set of representations (stored visual, orthographic, semantic, phonological information) for solving particular task (e.g. naming). Since no test available to assess north Indian population with specific colloquial expression thus, need was felt to assess these patients. Hence, the aims are to:

1. Develop standardized neuropsychological test to diagnose the presence, degree, specificity & type of aphasic disorders.
2. Assess all language areas, from perceptual modalities (auditory, visual, & gestural), processing functions (comprehension, analysis, problem-solving) to response modalities (writing, articulation, manipulation) for specific prognosis.

Method: The study had 2 phases. 1st phase: development of test material & 2nd phase: administration on aphasics (A), normals (N) & other than aphasics (OA). The test consisted of 5 sections, viz., Acoustic Problems (5), Speech & Language Problems (10), Simple Mathematical Problems (3), Perceptuomotor & Writing Problems (5) & Visual & Verbal Problems (3). All sub-sections were in auditory & orthographic mode. The test was administered on 122 (A), 74 (N) and 64 (OA) aged 18–81 years.

Results: The test was 75% sensitive to (A) while 72% sensitivity to (OA) on the entire task in all modalities.

Conclusion: It can be used for assessment of specific aphasia for planning better prognosis along with other pharmacological treatment, thereby improving quality of patient's life.

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Abstract – WCN 2013

No: 491

Topic: 3 – Stroke

Superficial siderosis of the central nervous system: A post-mortem 7.0 tesla magnetic resonance imaging study with neuropathological correlates

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Background: This study aims to elucidate the pathological substrate of superficial siderosis (SS), regarded as a radiological marker of cerebral amyloid angiopathy (CAA). The present study investigates the prevalence of SS and underlying lesions with 7.0 T MRI in post-mortem brains of patients with various neurodegenerative and cerebrovascular diseases.

Materials and methods: Hundred-twenty post-mortem brains were examined with 7.0 T MRI and their neuropathological correlates. The MRI examination consisted of a positioning, a T2 and a T2*-weighted sequence of coronal sections of a cerebral hemisphere and horizontal sections through pons and cerebellum.

Results: Forty-five brains (37.5%) with 83 separate zones of SS were detected, including 25 zones of disseminated siderosis (macro-SS) and of 58 areas of focal siderosis, restricted to less than 3 sulci (micro-SS). Macro-SS was associated with 14 lobar hematomas and 11 cerebral infarcts ($p = 0.64$), while micro-SS was associated with 19 micro-bleeds and 39 micro-infarcts ($p < 0.001$). Comparison of the 15 CAA to the 30 non-CAA brains showed that macro-SS was due to a lobar hematoma in 53% in the former compared to 3% in the latter group ($p = 0.003$). Micro-SS was due to a micro-bleed in 7% in the CAA brains and to 40% in the non-CAA brains ($p = 0.03$).

Conclusions: SS is due to cerebral bleeds as well as to cerebral infarcts. It should not to be considered as an additional Boston criterion for CAA. Macro-SS is mainly related to lobar hematomas while micro-SS is mainly due to cortical micro-infarcts.

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Abstract – WCN 2013

No: 517

Topic: 3 – Stroke

Factors associated with remote seizure in cerebral vein and dural sinus thrombosis

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Introduction: Whether initiating anticonvulsants treatment for all cerebral vein and sinus thrombosis patients or to wait for a seizure before starting the treatment is a vital question in management of cerebral vein and sinus thrombosis. Here in this study we wanted to define the possible risk factors of presenting, early and remote seizures in cerebral vein and sinus thrombosis patients.

Materials and methods: We have designed a prospective cohort study from April 2007 to April 2012, in 94 patients in al-Zahra Hospital, Isfahan, Iran. On admission, complete neurologic examination and further brain imaging were performed to evaluate probable risk factors for seizure. To recognize seizure predictors, we compared demographic, clinical and imaging factors among patients with or without presenting early and remote seizures.

Results: Among 94 patients, 32 ones (34%) have experienced at least one seizure on the basis of cerebral vein and sinus thrombosis. Bivariate analysis has shown significant association with remote seizure in patients with low consciousness, supratentorial lesion, lesion in occipital, temporal and parietal lobe, thrombophilia, seizure in acute phase and sigmoid sinus thrombosis. In logistic multiple regression analysis, there was no factor who can predict remote seizure.

Conclusions: Our results demonstrate that seizure in acute phase is the most significant factor in relation with remote seizure. Parenchymal lesion in supratentorial area was also found to be considerably associated with both early and remote seizures. But there was no factor predicting seizure in long-term. Larger studies should be performed to study predicting factors for early and remote seizures.

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Abstract – WCN 2013

No: 526

Topic: 3 – Stroke

Nocturnal blood pressure regulation in stroke patients with sleep apnea – Impact of etiology, comorbidity, medication and cerebral lesion

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Background: In clinical routine, stroke patients with sleep apnea (SA) show a different blood pressure (BP) response to respiratory events. The aim of the study was to evaluate the impact of age, comorbidity, medication, etiology and location of cerebral lesions on nocturnal (BP) in stroke patients with SA.

Methods: Respirographic sleep studies were performed in all stroke patients that underwent neurorehabilitation. Their systolic BP was determined by means of a non-linear algorithm and an individual one-point calibration of the pulse transit time obtained with a cuff-based BP measuring (SOMNOmedics GmbH, Germany). The number of systolic rises (defined as >15 mmHg) was scored. Risk factor evaluation, Oxfordshire Community Stroke Project (OCSF) and TOAST classification were performed.

Results: Out of the 203 stroke patients (age 58 ± 12) enrolled in the study, 38% were suffering from SA (AHI > 15). They showed a median of 47 ± 38 BP rises per hour, with a median rise by 19 ± 3 mmHg and a median nocturnal systolic BP of 138 ± 26 mmHg. In 7 patients, no apnea-related BP rises were observed (non-responders). Etiologically, they showed an equal distribution of small-artery occlusion (3), large-artery atherosclerosis (1), stroke of other determined (1) or undetermined cause (1), cardioembolism (1) and cerebral hemorrhage (1). There were neither differences in OCSF vessel lesions nor other influence factors for the non-responders.

Conclusion: 9.1% of stroke patients with SA show an atypical blood pressure response to apneas (non-responders). The missing blood pressure response doesn't seem to be correlated with age, comorbidity, clinical symptoms, medication, etiology, or cerebral lesion.

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Abstract – WCN 2013

No: 523

Topic: 3 – Stroke

Osteoporosis may be a predictor of silent cerebral infarct and white matter change in community-dwelling healthy adults: Present project

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Background: Loss of bone and muscle mass, fall and fractures are common conditions after stroke. Possible association between reduced bone density and cognitive impairment in postmenopausal women are well recognized. However, the relationship between bone mass loss and silent infarcts and cerebral white matter changes remains unknown.

Methods: Bone densitometry measured by Dual-energy X-ray absorptiometry and brain CT scan were performed for 650 subjects (458 female, mean age 63 ± 7.8 years; 192 male, 61.5 ± 8.5 years) among 650 stroke- and dementia-free adults older than 50 years between January 2009 and December 2010. Assessment of vascular risk factors and physical examination by in-person interview and were also taken.

Results: We did analyze 646 subjects who have taken brain CT scan and BMD study. In unadjusted analysis, the odds ratio (OR) for silent cerebral infarcts and/or cerebral white matter changes in men with osteoporosis was 3.8 (95% CI, 1.62–8.86; $p = 0.002$) and 2.2 (95% CI, 1.35–3.53; $p = 0.002$) in women as compared to subjects with normal bone density. Even after adjustment for age, education, hypertension, DM, hypercholesterolemia, and current smoking, the OR was 3.8 (95% CI, 1.43–10.3; $p = 0.008$) for men and 1.9 (95% CI, 1.10–3.18; $p = 0.02$), for men.

Conclusion: These findings suggest that bone mass loss may be a predictor of atherosclerotic vascular lesions in community-dwelling, apparently healthy men as well as women. Severe bone mass loss among men may be more harmful than among women. Further exploration of relationship between bone mass loss and atherosclerotic cerebral lesions is needed.

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Abstract – WCN 2013

No: 238

Topic: 3 – Stroke

The effect of lithium in enhancing post-stroke motor recovery: A double-blind, placebo-controlled, randomized clinical trial

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Background: Evidences from cultured cells and animal models of ischemia suggested that lithium has neuroprotective and neurotrophic effects and may play a desirable role in reducing infarct volume and improving the brain insults from stroke.

Objective: The aim of the study was to evaluate the efficacy of lithium in motor recovery after ischemic stroke with focus on distal upper limb motor recovery.

Material and methods: This was a placebo-controlled, double-blind, and randomized clinical trial on 80 patients with first ever stroke, allocated randomly in lithium 300 mg/twice daily or placebo. Treatment was initiated 48 h after stroke and continued for 30 days. Modified National Institute of Health Stroke Scale (mNIHSS) and hand subsection of Fugl-Meyer Assessment (hFMA) were used to evaluate impairment on the 5th and 30th of treatment.

Results: 66 subjects completed the study, including 32 subjects in lithium and 34 in placebo group. In general, there were no statistically significant difference in the improvement in mNIHSS ($p = 0.40$) and hFMA ($p = 0.07$) after 30 days. However, a subgroup analysis showed that patients with cortical strokes in lithium group had significantly better improvement in both mNIHSS and hFMA in comparison to placebo group ($p = 0.003$). About 44% ($n = 14$) of patients in lithium group, mainly from cortical stroke subgroup, regained more than 25% of full function based on hFMA, while this rate in placebo group was 14.7% ($n = 5$) ($p = 0.009$).

Conclusion: We observed some promising effect from lithium in enhancing recovery in cortical sub-group of ischemic stroke. Larger trial with more cortical stroke patients is needed to better investigate this effect.

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Abstract – WCN 2013**No: 507****Topic: 3 – Stroke****Shoulder hand syndrome in hemispheric stroke**

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Introduction: Shoulder-hand syndrome (SHS) is the post-stroke pain in the paretic limb characterized by regional pain, sensory changes (allodynia), abnormalities of temperature and sudomotor activity, oedema formation, abnormal skin color changes that usually occur between two weeks to five months after stroke.

Incidence: 1.5%–70%.

Clinical criteria: Pain-spontaneous, allodynia, hyperalgesia.

Motor disturbances: tremor, weakness, lack of muscular coordination, decreased range of motion, myoclonus, dystonia.

Skin changes: Color changes, sudomotor abnormalities, temperature dysregulation, oedema, nail growth, anxiety, depression.

Possible causes: Aetiology of stroke, side affected, sensory impairment, spasticity, shoulder subluxation, the severity and recovery of motor deficit, Phenobarbital use, etc.

Method/objective: The aim of the study was to review the clinical features of SHS.

Study type: Cross-sectional study.

Study Place: Neurology department, Bangabandhu Sheikh Mujib Medical University, Dhaka.

Study Period: July 2009–June 2011 (2 years).

Subjects: Stroke with SHS.

Sample size: thirty cases. Sampling technique: Convenient sampling.

Data collection: By interviewed questionnaire, SHS was diagnosed by clinical criteria.

Result: Age range of the cases was 45–75 years. SHS was more common in 56–65 years of age. Clinical features were Weakness (96.7%), Spontaneous Pain (93.3%), Oedema (86.7%), Raised temperature (63.3%), Tremor (36.7%), Hyperalgesia (33.3%), Color change (30%), Dystonia (30%), Normal temperature (30%), Allodynia (26.7%), Sudomotor change (23.3%), Myoclonus (10%), Decreased temperature (6.7%).

Conclusion: SHS is not very uncommon in our clinical setting. This study aimed at reviewing the clinical features of SHS after hemispheric stroke. It is not related to any type of stroke or any side of stroke affected.

doi:10.1016/j.jns.2013.07.693

Abstract – WCN 2013**No: 538****Topic: 3 – Stroke****Predictors and causes of hospital readmissions after acute ischaemic stroke: Local data from a hospital-based registry**

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Background: Acute ischaemic stroke patients have significant neurological deficits, which may lead to increased hospital readmission rates during the first year after index stroke.

Objective: This study looks for the predictors of hospital readmission within the first year of discharge for index ischemic stroke, and investigates for the common causes of hospital readmission.

Methods: 1797 patients were admitted to our hospital from January 1, 2009 to December 31, 2009 for acute ischaemic stroke. Four hundred patients were sampled by simple randomization and they

were dichotomized into those with hospital readmission and those without. Their demographics were analyzed with SPSS 16.0, and their causes and times to next hospital readmission were also investigated.

Results: A total of 400 patients were studied in detail, but after exclusion, 334 patients were recruited for this study. 168 patients were found to have hospital readmission within 1 year of discharge for index acute ischaemic stroke. The predictors of hospital readmission include elderly patients with age <72 years, cardioembolic stroke, diabetes mellitus, complications occurring during index admission, and NIHSS > 5. Commonest causes of hospital readmission include infection and recurrent stroke.

Conclusion: Certain patients who are admitted to hospital for acute ischaemic stroke are at increased risk of readmission after the index event. Risk factors for early readmission, i.e. after the first 30 days, and for late readmission, i.e. from the 31st day onwards, can be identified early and readmissions can be prevented. This may minimize the cost brought about to the health care burden.

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Abstract – WCN 2013**No: 397****Topic: 3 – Stroke****Electrocardiographic maximal P-wave duration in patients with acute ischemic stroke: A novel yardstick in diagnosing ischemic-stroke subtypes?**

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Background: P-wave parameters of electrocardiogram (ECG) have associations with atrial fibrillation (AF), myocardial ischemia, or systemic embolism.

Objective: We aimed to evaluate whether maximal P-wave duration (P-max), one of the P-wave parameters, provides additional diagnostic information in determining ischemic-stroke subtypes.

Patients and methods: We enrolled 160 acute stroke patients who showed sinus rhythm on admission. The P-max was defined as the greatest P-wave duration among all 12-leads of ECG measured by calipers manually. The patients were classified into 3 groups by the P-max value; group 1: <80 ms (n = 33), group 2: 90–110 ms (n = 73), and group 3: >120 ms (n = 54). In these 3 groups, clinical background, ECG findings during admission, and ischemic-stroke subtypes were compared. The ischemic-stroke subtypes were classified into large-artery atherosclerosis (LAA), cardioembolism (CE), small-vessel occlusion (lacunar), and others with determined etiology.

Results: In post hoc analysis, chronic kidney disease, development of premature atrial contraction and paroxysmal AF, and dilation of left atrial dimension on echocardiogram were significantly more frequent in group 3, compared with the other groups. In chi-square test, group 3 had CE more frequently than other ischemic-stroke subtypes ($P < 0.001$). Meanwhile, groups 1 and 2 had CE less frequently than the other ischemic-stroke subtypes ($P = 0.04$ and 0.006 , respectively). In contrast, there was a tendency that the lacunar was more frequently in groups 1 ($P = 0.07$).

Conclusion: The P-max in acute stroke patients who show sinus rhythm on admission may be a simple, convenient and practical yardstick in diagnosing ischemic-stroke subtypes.

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Abstract – WCN 2013**No: 578****Topic: 3 – Stroke****A case of relapse and remitting cerebral infarction associated with polycythemia vera**

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Background: Polycythemia vera (PV) is a chronic myeloproliferative disorder characterized by increased production of leukocytes, erythrocytes and platelet. We present a case of relapse and remitting ischemic stroke associated with PV, which resulted in aggravation of infarction by hematologic abnormalities.

Objective: The aim of the study was to present a case of relapse and remitting cerebral infarction with PV and discuss the relationship between aggravation of ischemic stroke and hematologic abnormalities.

Patients and methods: A fifty-two-year-old woman visited our department of emergency with transcortical sensory aphasia. Her brain MRI and MR angiography (MRA) showed the acute ischemic stroke in left parietotemporal lobe and mild stenosis of ipsilateral middle cerebral artery (MCA). She was diagnosed with PV according to WHO criteria at the first hospitalization. She was hospitalized five times for recurrent ischemic stroke during about 3 month. The brain DWI, MRA and laboratory test were evaluated at each hospitalization.

Results: The patient's neurological symptoms got worse and MRA revealed the progression of stenosis and occlusion of proximal left MCA finally. While the level of hemoglobin and hematocrit has been improved in her laboratory test, but the white blood cell (WBC) count and platelet count have been increased despite continued treatment of PV.

Conclusion: In this case, increased WBC and platelet count may play an important role in formation of local thrombus and contribute to the progression of large artery stenosis. Unlike the other cases, we demonstrate the relationship between aggravation of intracranial artery stenosis and hematologic deterioration by serial MRA and laboratory test.

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Abstract – WCN 2013**No: 577****Topic: 3 – Stroke****Effects of uric acid levels on outcome in severe ischemic stroke patients treated with intravenous Rtpa**

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Background: Uric acid (UA) has been known to be a neuroprotective antioxidant because of its free radical scavenger activity. However, many observational studies indicate that elevated serum levels of UA increase the risk of cardiovascular events. We studied the influences of UA in patients with acute ischemic stroke after thrombolytic therapy.

Objective: To confirm the association of admission UA level with clinical outcome in intravenously thrombolized patients stratified by stroke severity.

Patients and methods: Two hundred eighteen consecutive patients treated with intravenous thrombolysis were included for this analysis in our prospective stroke registry. Early improvement was defined as an improvement from baseline in the National Institutes of Health Stroke Scale (NIHSS) score by ≥ 4 points 24 h after stroke onset and excellent functional outcome was defined as 3-month modified Rankin Scale score by using responder analysis.

Results: There was no significant relationship between serum UA levels and early improvement or excellent functional outcome in total group. However, in the patients with severe baseline stroke deficits (NIHSS score ≥ 15), patients with higher tertile of UA levels had a significant association with excellent functional outcome ($p = 0.003$). Excellent functional outcome in patients with severe baseline stroke deficits might be associated with serum UA levels particularly in men but not in women.

Conclusions: Increased serum UA levels are significantly associated with better outcome in severe ischemic stroke patients treated with intravenous thrombolysis, but the effectiveness of UA can differ by initial stroke severity and gender.

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Abstract – WCN 2013**No: 533****Topic: 3 – Stroke****Characterization of cerebral venous thrombosis in a stroke unit – Cross-sectional study**

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Background: Cerebral venous thrombosis (CVT) is the least common form of acute cerebrovascular disease, 0.5% of all strokes. Diagnosis may be delayed due to large spectrum of clinical symptoms and nonspecific signs on neuroimaging.

Objective: The aim of the study was the characterization of CVT cases diagnosed in our stroke unit.

Patients and methods: 13 patients with CVT admitted in our department from January 2008 to December 2012 were assessed through retrospective, epidemiological characteristics, clinical features, risk factors and prognosis.

Results: Of the 13 patients with confirmed CVT, 10 were female, aged between 21 to 57 years (average 38.25). We found thrombotic risk factors in 10 patients: oral contraceptives use ($n = 4$), infection ($n = 2$) and thrombophilia ($n = 2$). The diagnosis was made by MRI in 11 cases, 1 by brain Venography and another by Angiography. Median time from onset of symptoms to diagnosis was 2 days. In 5 patients, multiple sinuses were affected; the most frequent was lateral sinus ($n = 7$), followed by superior sagittal sinus ($n = 6$). Headache, with or without associated deficits, was the most common symptom, occurring in 12 patients. Six patients had a good outcome with complete recovery at early stage of treatment and the rest remained with mild disability.

Conclusion: The data of our patients were similar to literature. However, in our series no deaths occurred and the prognosis was good. This can be explained by rapid diagnosis (only 2 days, less than other studies) and treatment initiation. CVT should be considered in diagnosis of secondary headache even in the absence of other symptoms.

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Abstract – WCN 2013**No: 600****Topic: 3 – Stroke****Hematoma size but not Gcs score on admission predicts in-hospital case fatality in the very elderly. A case control study**

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Background: Despite extensive investigations in the stroke medicine, still there is little known about the course of spontaneous intracerebral haemorrhage (SICH) in the very elderly.

Material and methods: A retrospective study of 231 consecutive cases diagnosed with SICH was carried out in the Department of Neurology at the Medical University of Warsaw. A case control study included patients with SICH aged ≥ 80 years. Data on risk factors, clinical presentation, neuroimaging and outcomes were collected and statistically analysed.

Results: The whole studied population consisted of 231 patients (52% male): 66 aged ≥ 80 years and 165 < 80 years of age. There were more female cases among the very elderly (65% vs 46%, $p = 0.009$). Hypertension was the most common risk factor in both age groups ($p = 0.21$). Coronary artery disease and atrial fibrillation were statistically more frequent in the elderly (CAD: $p < 0.001$; AF: $p < 0.001$). No significant differences were observed in GCS score or the SICH site. Larger hematoma size (3.6 cm vs 3.2 cm, $p = 0.04$) and more frequent occurrence of intraventricular extension (IVH) (41% vs 24%, $p = 0.05$) were found in patients aged ≥ 80 years. The mortality rate was higher in the older patients (in-hospital mortality: 40.9% vs 15.2%, 30-day mortality: 33.3% and 13.3%; both $p = 0.02$). Hematoma size predicted in-hospital case fatality in the very elderly ($p = 0.005$), whereas hematoma size and GCS score had predictive value in patients aged < 80 years ($p = 0.001$ and $p = 0.017$, respectively).

Conclusion: The very elderly patients with SICH are a high risk group as far as clinical outcomes and prognosis is concerned. Hematoma size seems to be the most powerful parameter in predicting early mortality. Further prospective studies are needed to identify prognostic factors in this subpopulation.

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Abstract – WCN 2013

No: 605

Topic: 3 – Stroke

Prevalence and radiological peculiarities of metabolic syndrome and hypertension in stroke patients

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Background and purpose: Hypertension, obesity and diabetes (metabolic syndrome) are significant risk factors of stroke. A better understanding of relation between metabolic syndrome and hypertension may lead to the reduction of cerebrovascular risks and improve the stroke treatment and outcome as well.

Methods: A total of 345 ($F = 198$, 69.7 ± 5.3) patients with acute stroke were investigated. Diagnosis of Metabolic syndrome (MetS) was ascertained according to the revised National Cholesterol Education Program Adult Treatment Panel III (ATP III-r). Stroke was diagnosed using NIHSS criteria. Type, side and site of stroke were assessed by MRI. Hypertension (HYP) was defined according to cardiovascular criteria, based on target organ damage. In addition to hypertension, other modifiable and non-modifiable risk factors were recorded. Neuropsychological battery and MMSE tests were performed in the target population. The data statistically evaluated by SPSS-11.0.

Results: Among stroke patients 124/35.9% found to have MetS, 106/85.4% diagnosed as ischemic stroke with lacunar or multiple brain lesions. Most of the patients with female preponderance (76) had moderate to severe leucoaraiosis (71%), mild-to moderate cognitive disturbances (54%) and dementia (14%). The HYP and obesity alone-146/42.3% patients had more haemorrhagic stroke (79/54.1%) with male dominance and psychological disturbances, such as depression (46%) or depression-anxiety disorders (31%).

Conclusions: Mets may be considered as independent risk factor for ischemia and dementia ($P > 0.001$), while the Hypertension and obesity alone may carry the risk of bleeding and neuropsychological disturbances ($P > 0.001$).

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Abstract – WCN 2013

No: 607

Topic: 3 – Stroke

Comparison of comorbidity of hypertension and mild cognitive impairment in atrial fibrillation's patients after stroke and myocardial infarction

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Background: Mild Cognitive Impairment is the frequent sequela of patients with neurovascular and cardiovascular disorders, especially patients with Atrial Fibrillation and may lead to vascular dementia.

Aim: The aim of the study was to establish the role Hypertension on development of MCI after stroke and Myocardial Infarction.

Methods: 192(93/99) poststroke and post-MI patients with AF were investigated and underwent MRI scan. Cognition and daily activity evaluated by MMSE, ADL Index, Cognitive function investigated by neuropsychological battery test and acute phase and after 3 months. Depression was evaluated by Hamilton Depression Rating Scale.

According to the test data both neurological (after Stroke) and cardiovascular (after Myocardial Infarction) patients with atrial fibrillation were separately divided in III groups:

I – MCI alone, II – Hypertension alone, III – MCI + Hypertension.

Groups did not differ due to vascular risk-factors and demographic variables.

Results: From patients with MI-MCI and Hypertension alone was diagnosed in 56 (56.5%) and 38 (38.3%), but after Stroke in 42 (45.1%) and 76 (81.7%) cases respectively. Hypertension + MCI – were revealed after MI in 42 (42.2%) and postStroke in 68 (71.1%) cases, – Comparison of both groups revealed that Hypertension + MCI and Hypertension alone have more frequently were in poststroke group as well as ADL Index and MMSE, but depression alone 58 (58.5%) and Executive Dysfunction Syndrome alone 63 (63.6%) more frequently as well their comorbidity (34/34.3%) in cardiac patients vs poststroke. ($p < 0.05$), which were correlated with multiple lacunar silent damage radiologically.

Conclusion: In AF-poststroke patients more demented and hypertensive (OR 9.90, $P = 0.031$), but the AF-MI patients have more frequently mild cognitive decline and depression (OR 8.9, $P > 0.001$).

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Abstract – WCN 2013

No: 619

Topic: 3 – Stroke

Tracking of mesenchymal stem cells labeled with Gd-Dtpa by Mr imaging in cerebral ischemia model

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Objective: Recent developments in stem cell and gene therapy will require methods to monitor stem cell survival and integration repeatedly and non-invasively with a high temporal and spatial

resolution in vivo. The aim of the study was to explore the possibility of the rat mesenchymal stem cells (MSCs) labeled with standard contrast agents (Gd-DTPA) for stem cell tracking.

Methods: MSCs from bilateral femur of rats were cultured and propagated. Intracellular uptake of Gd-DTPA was achieved by using a non-liposomal lipid transfection reagent (Effectene) as the transfection agent. Electron microscopy was performed to detect the distribution of Gd-DTPA particles in MSCs, and labeling efficiency of Gd-DTPA particles on MSCs was evaluated using spectrophotometric. Viability and proliferation of labeled MSCs were evaluated using MTT assay. Labeled MSCs were detected with T1-weighted MR imaging in vitro and in rat brain.

Results: The presence of Gd-DTPA particles inside the MSCs was definitely detected by transmission electron microscopy. Labeling efficiency was highly. There was no difference in viability and proliferation between the labeled and unlabeled confirmed by MTT values of light absorption. The labeled MSCs demonstrated the high signal intensity on T1-weighted MRI in vitro and in rat brain.

Conclusion: Rat MSCs can be labeled with Gd-DTPA particles without changing the cell viability and proliferation. Obviously labeled MSCs can be imaged in vitro and vivo. Gd-DTPA shows no evident adverse effect on the function of labeled MSCs. Gd-DTPA can be used for the MR imaging tracking of labeled MSCs.

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Abstract – WCN 2013

No: 637

Topic: 3 – Stroke

Epidemiology of stroke risk factors in Belarus

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Background: Stroke is one of the leading causes of death. Death from stroke in people of working age is particularly alarming. Studying the prevalence of stroke risk factors will help develop an optimal system for prophylaxis thus reducing morbidity and mortality from stroke.

Objective: The aim of the study was to study prevalence of stroke risk factors among people of working age.

Patients and methods: Using a specially developed standardized questionnaire card we carried out the screening of open population. We examined 189 people aged between 40 and 62 years (mean age 51.68 years) of one of the district polyclinics in Minsk, including 135 women (71.43%) and 54 men (28.57%).

Results: The following risk factors were revealed:

- Overweight – 128 people (67.72%), including 66 people with a BMI of 26–29 (34.92%) and 62 people with a BMI \geq 30 (32.8%);
- Arterial hypertension – 87 people (46.03%), including 33 people with hypertensive crisis in medical history;
- Family history of hypertension before the age of 60 years – 98 people (51.85%);
- Hypodynamia – 71 people (37.57%);
- Smoking – 46 people (24.34%);
- Coronary heart disease – 35 people (18.52%);
- Family history of stroke/heart attack before the age of 60 years – 31 people (16.4%);
- Diabetes mellitus – 11 people (5.82%);
- Alcohol abuse – 7 people (3.7%).

Conclusion: The most common stroke risk factors in the studied population are overweight, hypertension and hypodynamia. Particularly disturbing is the fact that only 44 people (50.57%) with hypertension regularly take antihypertensive medication.

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Abstract – WCN 2013

No: 470

Topic: 3 – Stroke

The prevalence and association with cerebrovascular risk factors of cerebral microbleeds in Korean ischemic stroke patients

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Background and Purpose: Advanced age, presence of intracerebral hemorrhage or ischemic stroke, dementia, hypertension, and use of antithrombotic therapy have been reported to be associated with a higher risk of cerebral microbleeds. In addition, the severity of white matter hyperintensity (WMH) was known to be associated with cerebral microbleeds. We tried to investigate the frequency and the number of microbleeds in Korean ischemic stroke patients and risk factors as well as their relationship to microbleeds.

Methods: Four hundred subjects (mean age \pm SD: 66.5 \pm 13.5) visited for acute cerebral infarctions or transient ischemic attacks were enrolled in this study. For each subject, we researched the baseline demographic characteristics, cerebrovascular risk factors, the number and localization of the microbleeds or WMHs.

Results: The prevalence of microbleeds in this study was 31%. We revealed that male gender, history of heart disease and WMH acted as burdens of microbleeds. After regional subdivision, hypertension was a risk factor for microbleeds especially located in basal ganglia of human brain. Regardless of the location of microbleeds, we understood that WMH was considerably correlated with microbleeds.

Conclusion: A higher prevalence of microbleeds was proved in the Korean ischemic stroke patients than Caucasians. Also, we found that male or WMHs might be the risk factors for microbleeds. Furthermore, hypertension was a risk factor of the microbleeds existed in basal ganglia. WMHs were one of the most important risk factors for microbleeds in spite of bleeding site.

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Abstract – WCN 2013

No: 621

Topic: 3 – Stroke

Clinical characteristics of stroke patients in the Northern Thailand

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Background: Acute stroke required emergency management.

Objective: This study aimed to analyze the clinical characteristics and management of stroke in the Northern Thailand.

Material and methods: We interviewed patient/relative and analyzed the data of the acute stroke patients admitted to twelve government hospitals in the Northern Thailand during October 2010 to September 2012.

Results: Total of 802 acute stroke patients were recruited, mean age was 65 \pm 13.5 years old, 54.1% were female. Most patients had regularly exercise and rarely consumed alcohol. Co-morbidities such as hypertension, diabetes, dyslipidemia were 57.2%, 15.6%, 18.4% respectively. Ischemic and hemorrhagic strokes were 79.7% and 20.3%, respectively. Clinical characteristics of the patients including BP, pulse, GCS, NIHSS, and blood tests were not different between ischemic and hemorrhagic strokes. Patients admitted to general ward were 73% and only 13.1% were admitted to stroke unit. Thrombolytic agent was given in 10.4% of the ischemic stroke patient. The reasons that this agent was not used were unavailable in the hospital for 24.5%, and 75.5% had contraindications including delayed onset

(37.9%). Bleeding complication from thrombolytic agent was 15.4%. Antithrombotic agents for secondary stroke prevention were subsequently used in 94.7% of ischemic stroke patients.

Conclusion: Hypertension was the most important risk factor in the Northern of Thailand. Majority type was ischemic stroke. Due to resource limitation, most of the patients were admitted to general ward. Thrombolytic therapy was given in minority of ischemic stroke patients, reasons apart from contraindication were unavailable and delayed onset. Improvement system of care is needed.

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Abstract – WCN 2013

No: 623

Topic: 3 – Stroke

Goal achievement of risk factor control for patient with ischemic stroke in the Northern Thailand

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Background: Stroke causes morbidity and mortality.

Objective: This study aimed to evaluate the goal achievement of risk factor control of ischemic stroke in the Northern of Thailand.

Material and methods: We analyzed the data of the ischemic stroke patients with more than 6 months of onset who regularly followed up during October 2010 to September 2012. The defined goal of the risk factor control was according to ASA 2006, and ADA 2008 in patient with diabetes.

Results: Total of 498 patients were enrolled, aged 66 ± 12.4 years old, and 52.1% were male. The proportion of the patients living in the city is 51.9%. Healthcare scheme has a universal coverage of 53.4%, and civil servant coverage 34.1%. Proportion of patient with BMI ≤ 25 kg/m² was 72%, regular exercise was 61.2%, and quit smoking or never smoking was 87.2%. Control of HT (SBP/DBP in DM vs. Non DM) was 77.6/91.2%, and 61.6/76.8%, respectively. Dyslipidemia; LDL ≤ 100 mg/dl, DM vs. non-DM was 48.1% vs. 44.7% and diabetes; FBS ≤ 120 mg% and HbA1C $< 7.0\%$ was 50.0%, and 31.3%, respectively. Antithrombotic agents were prescribed in 91.8% of patient.

Conclusions: Goal achievement of risk factor control for patients with ischemic stroke in the Northern of Thailand is relatively compatible with data from international studies but there is still a quality gap. These findings will be used to encourage participating hospitals to improvement stroke care.

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Abstract – WCN 2013

No: 650

Topic: 3 – Stroke

Transient ischaemic attacks in young adults

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Background: Transient ischemic attacks (TIAs) are risk factors for the future development of ischemic stroke.

Objective: To analyze predisposing factors of TIAs in young adults.

Materials and methods: 50 patients with TIA were examined, the 1st group consisted of patients under 30 years old (10%), the 2nd group consisted of 30–40 years old patients (18%), the 3rd group – 40–50 years old (62%). Laboratory and neuroimaging studies were conducted.

Results: The risk factors of TIAs revealed in group 1 were: 35% – malformation of cerebral vessels, such as open Willis circle (anterior and

posterior trifurcation, posterior communicating artery's aplasia); 20% – sugar diabetes with angiopathy; 20% – non-ischemic cardiac arrhythmias, 10% combined oral contraceptives intake, and 5% hyperhomocysteinemia. In group 2, 46% cerebral vessels malformation, 18% – non-ischemic cardiac arrhythmias, 9% – arterial hypertension (AH) and 9% – cerebral atherosclerosis. In group 3 – 42% AH, 30% – cerebral atherosclerosis, 22% – malformation of cerebral vessels and 6% – non-ischemic cardiac arrhythmias. 25% of patients had both atherosclerosis and other risk factors. Patients with TIA in anamnesis developed ischemic stroke in 60% (group 1), 67% (group 2) and 74% (group 3) of cases.

Conclusion: The appropriate screening for cerebral vessels' malformations (as the major risk factor of TIA at the age under 40 years old), atherosclerosis, metabolic disorders and other risk factors is necessary for the TIA's prophylaxis.

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Abstract – WCN 2013

No: 648

Topic: 3 – Stroke

Plasma neutrophil gelatinase-associated lipocalin and functional outcome in ischemic stroke

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Background: A range of circulating inflammatory reactants have been evaluated as biomarker candidates in acute ischemic stroke. However, no study has shown an additional value of circulating inflammation related proteins for the prediction of the patients' functional outcome.

Patients and methods: At one week from ischemic stroke onset, we measured plasma levels of neutrophil gelatinase-associated lipocalin (NGAL) in 46 consecutive ischemic stroke patients. Protein levels were related to the occurrence of post-stroke infections and the patients' modified Rankin Scale (mRS) at day 90 after stroke.

Results: Plasma levels of NGAL correlated significantly with the mRS at day 90. In predictive models for unfavourable (mRS 3–6) vs. favourable (mRS 0–2) outcomes which included the National Institutes of Health Stroke Scale on admission, the patients' age, and thrombolytic therapy, the addition of NGAL significantly increased the area under receiver operating characteristics curves. Plasma NGAL levels did not add prognostic information when the occurrence of post-stroke infections was also included into the predictive models.

Conclusions: Elevated circulating NGAL levels one week following ischemic stroke are associated with worse functional outcomes at three months and reflect the occurrence of post-stroke infections.

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Abstract – WCN 2013

No: 651

Topic: 3 – Stroke

Plasma midregional pro-adrenomedullin improves prediction of functional outcome in ischemic stroke

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Background: To evaluate if plasma levels of midregional proadrenomedullin (MR-proADM) improve the prediction of functional outcome in ischemic stroke.

Patients and methods: In 168 consecutive ischemic stroke patients, plasma levels of MR-proADM were measured within 24 h from symptom onset. Functional outcome was assessed by the mRS at 90 days following stroke. Logistic regression, receiver operating characteristics (ROC) curve analysis, net reclassification improvement (NRI), and Kaplan–Meier survival analysis were applied.

Results: Plasma MR-proADM levels were found significantly higher in patients with unfavourable (mRS 3–6) compared to favourable (mRS 0–2) outcomes. MR-proADM levels were entered into a predictive model including the patients' age, NIHSS, and the use of recanalization therapy. The area under the ROC curve did not increase significantly. However, category-free NRI of 0.577 ($p < 0.001$) indicated a significant improvement in reclassification of patients. Furthermore, MR-proADM levels significantly improved reclassification of patients in the prediction of outcome by the Stroke Prognostication using Age and NIHSS-100 (SPAN-100; NRI = 0.175; $p = 0.04$). Kaplan–Meier survival analysis showed a rising risk of death with increasing MR-proADM quintiles.

Conclusions: Plasma MR-proADM levels improve the prediction of functional outcome in ischemic stroke when added to the patients' age, NIHSS on admission, and the use of recanalization therapy. Levels of MR-proADM in peripheral blood improve the reclassification of patients when the SPAN-100 is used to predict the patients' functional outcome.

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Abstract – WCN 2013

No: 602

Topic: 3 – Stroke

Cardio-embolic causes of stroke in young adults

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Background: Numerous aetiologies are responsible for stroke in young adults. A fundamental challenge in approaching a young stroke patient is that the cause of the stroke can be one of the many disease processes.

Objective: To determine the frequency of cardioembolic causes in young adults with acute stroke and compare this with other causes.

Methods: Prospective study of young adults presenting with acute first-ever stroke. The cases included people aged between 17 and 55 years, who had acute stroke, during thirty month period. The stroke mechanism was determined on patient's characteristics, presenting clinical features, risk factors for stroke and the results of the investigations performed.

Results: One hundred and thirty five patients with first-ever stroke aged 17–55 years were identified. The mean age was 45.4 ± 9.0 (SD) for men and $39.8 \pm (11.5)$ SD for women. The presumed stroke aetiology for men and women was established in 72 (53.5%) patients. Of these, Cardio-embolic causes were presumed in 16/72 (22%). Atherosclerosis accounts for 16 (22%) of cases. Haematological causes were presumed in 10/72 (14%) patients. Other disorders including non-atherosclerotic vasculopathies explained 30/72 (42%) cases. The causes of stroke were undetermined in 63 (46.5%) of the patients.

Conclusions: In this population of young patients, cardiac embolism is a very important cause of stroke. Even after taking an exhaustive

history, examining the patient in great details and undertaking numerous investigations, there may still be patients in whom no reasonable explanation for their stroke can be found.

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Abstract – WCN 2013

No: 732

Topic: 3 – Stroke

Where should we direct our efforts among in-hospital steps from arrival to achieve faster recanalization in acute ischemic stroke?

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Background and purpose: Faster recanalization is related to better clinical outcome in acute ischemic stroke. The purpose of our retrospective study is to analyze the time taken in various aspects of in-hospital care from ER arrival to arterial puncture in patients undergoing endovascular therapy and to find which step the longest is.

Method: Stroke Center Database was investigated retrospectively and included in our analysis were patients:

- (1) who were admitted to our institution from January 2004 to June 2012,
- (2) who presented some neurological symptoms,
- (3) who underwent MRI following CT, which showed acute ischemic stroke caused by major vessel occlusion and no intracerebral hemorrhage,
- (4) who underwent endovascular therapy including local intra-arterial fibrinolysis, mechanical thrombectomy using balloon, Merci retrieval system and Penumbra system and stenting.

We examined the times from ER arrival to arterial puncture (ER–AP), from ER arrival to CT (ER–CT), from CT to MRI (CT–MR), and from MRI to arterial puncture (MR–AP).

Result: During the study period, 212 patients were included for retrospective analysis. The average ER–AP time was 2.73 ± 1.39 h, the average ER–CT, CT–MR and MR–AP times were 0.47 ± 0.38 h, 0.81 ± 0.37 h and 1.45 ± 0.68 h, respectively. The MR to AP step spent the longest time and the CT to MR step was the second.

Conclusion: CT to MR and MR to arterial puncture times show wide variability and the times must be reduced to achieve faster recanalization by endovascular therapy.

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Abstract – WCN 2013

No: 746

Topic: 3 – Stroke

Adipokines level and insulin resistance in abdominally obese patients with primary acute ischemic non-lacunar hemispheric stroke

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Background: During the last decade there are numerous reports regarding obesity as a factor that could influence stroke severity and outcome.

Objective: The aim of this study was to determine humoral and endocrine changes among patients with abdominal obesity II class in the acute phase of ischemic non-lacunar hemispheric strokes.

Design and methods: This study evaluated 18 non-diabetic abdominally obese patients (class II) aged 61.3 ± 0.7 years (39% women) with

primary acute ischemic non-lacunar stroke. Patients were matched for infarct volume and co-morbidities. Serum concentrations of adiponectin and leptin, as well as HOMA-IR index were assessed at 1st and at 10th day after stroke onset in fasting state. For comparison, a control group of 9 abdominally obese conditionally healthy humans with chronic lumbalgia has been formed.

Results: Abdominally obese patients have significant and sustained reduction of adiponectin level at 1st ($23.4 \pm 1.5 \mu\text{g/ml}$, $p < 0.05$) and at 10th post-stroke day ($27.9 \pm 4.2 \mu\text{g/ml}$, $p < 0.05$) in comparison with control group of obese healthy humans ($42.0 \pm 2.7 \mu\text{g/ml}$). Moreover, abdominally obese patients have significant transient elevation of leptin serum level ($22.3 \pm 3.8 \text{ ng/ml}$, $p < 0.05$) and HOMA-IR (21.26 ± 5.94 , $p < 0.05$) at 1st day after stroke with subsequent return to basal values on the 10th post-stroke day ($11.4 \pm 3.4 \text{ ng/ml}$ and 2.25 ± 0.49 , respectively). Maybe, these peculiarities are interdependent and constitute the syndrome of adipose tissue reaction in stroke.

Conclusion: Under the conditions of acute ischemic non-lacunar hemispheric stroke, patients with abdominal obesity II class have sustained reduction of adiponectin serum level and transient elevation of leptin serum level as well as HOMA-IR.

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Abstract – WCN 2013

No: 751

Topic: 3 – Stroke

Stroke mechanisms and prognosis of monoparetic stroke

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Background: Although isolated monoparesis following stroke has been discussed in the literature, there are only a few studies about this topic. In this study, we attempted to clarify the lesion topography, mechanisms and prognosis for this rare entity of stroke.

Methods: We prospectively studied 295 consecutive acute stroke patients. Monoparetic stroke was defined as follows:

- 1) isolated motor deficit in either a leg or an arm without coordination deficit, language deficit and significant involvement of speech or face,
- 2) the motor deficit was caused by an acute ischemic or hemorrhagic stroke.

Results: Among the 295 patients, 11 had a monoparetic stroke (6 male and 5 female, mean age 63.2 years) involving an upper limb (7 hand palsies and 1 arm paresis) or a lower limb (3 patients). They had an acute ischemic stroke except one who manifested a right hand palsy related to the bleeding of metastatic brain tumor. The lesions were mainly located in frontoparietal cortex. Only two cases had a subcortical lesion. Cardioembolism was the cause of stroke in 4 patients, and large artery atherosclerosis (artery to artery embolism) in 4 patients, small vessel disease in 1 patient, and intratumoral hemorrhage in 1 patient. Besides, one case had two potential causes of stroke (cardioembolism and atherosclerosis). All patients except one case with metastatic tumor showed a good prognosis at discharge (modified Rankin scale ≤ 2).

Conclusion: Monoparetic stroke was mostly related to embolism, and had a good prognosis of neurological recovery if it was not a tumorous condition in the brain.

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Abstract – WCN 2013

No: 705

Topic: 3 – Stroke

The influence of VEGF-A in brain edema on acute ischemic stroke

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Background: Hypoxia causes transcription of genes that are controlled by HIF-1, which activates VEGF-A and influence blood brain barrier leakage, causing brain edema. Brain edema can lead to progressive neurological deterioration. The purpose of this study is to prove that the decreased levels of VEGF-A contribute to brain edema in acute ischemic stroke.

Material and methods: Design of this study was a case-control in acute ischemic stroke patients treated in Stroke Unit Dr. Sardjito Hospital Yogyakarta Indonesia. To find significant independent factors causing brain edema, logistic regression analysis was used. The significance level used was 0.05 with 95% level of confidence ($p < 0.05$).

Results: There were 37 subjects as a brain edema, and 34 subjects as non-brain edema. The mean levels of VEGF-A on brain edema group is 436 pg/ml, and in non-brain edema group is 746 pg/ml, this difference is significant (95% CI: 5.5–615, $p = 0.046$). The proportion of VEGF-A levels $< 638.3 \text{ pg/ml}$ in brain edema group is greater than non-brain edema (83.78% vs. 58.82%), this difference is significant. (OR = 3.6; 95% CI = 1.06–13.26). Results of multivariable analysis were lipid profile (OR = 4.26; 95% CI: 1.28–14.15), MMP-9 (OR = 5.59; 95% CI: 1.58–19.78), and score SSGM (OR = 5.29; 95% CI: 1.47–19.08).

Conclusion: The decreased levels of VEGF-A significantly contribute to the brain edema in acute ischemic stroke, and the influence of decreased levels of VEGF-A on the brain edema will increase if considered with unification of lipid profile, levels of MMP-9 and score SSGM.

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Abstract – WCN 2013

No: 713

Topic: 3 – Stroke

Intravenous thrombolysis for acute ischaemic stroke after recent transient ischaemic attack

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Background: Intravenous thrombolysis (iv-thrombolysis) in patients after recent transient ischaemic attack (TIA) is an important clinical problem.

Objective: The objective was to assess the impact of TIA preceding acute ischaemic stroke (AIS) on the safety and efficacy of iv-thrombolysis.

Methods: We retrospectively evaluated the data of Caucasian patients with AIS who were consecutively treated with iv-thrombolysis from September 2006 to May 2011 in three experienced stroke centres.

Results: There were 400 patients in the analysed group (54.50% male; mean age 69.56 ± 11.77). At three-month follow-up, 58.00% of patients were independent (mRS 0–2), 17.75% had died, 17.00% suffered HT and 4.25% experienced SICH.

There were 29 patients (7.25%) who had a previous ipsilateral TIA within 24 h before established stroke. Most of the patients (75.86%) had a single episode of TIA, while seven patients (24.14%) had more

than one episode. The duration between the time of the first TIA episode and the time of onset ranged from 60 to 1320 min with a median duration 369 min (range 160–720 min). The presence of SICH was higher in patients with prior TIA ($p = 0.153$); however, mortality was lower in this subgroup ($p = 0.563$).

Multivariate analysis showed the impact of prior TIA within 24 h before stroke onset on the presence of SICH. There was no impact of TIA on unfavourable outcomes after three months or on the mortality rate within three months.

Conclusion: We found no impact of recent TIA on the safety and efficacy of iv-thrombolysis in patients with AIS.

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Abstract – WCN 2013

No: 714

Topic: 3 – Stroke

Delirium in acute stroke: Frequency and risk factors

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Introduction: Delirium is common in acute stroke and worsens overall prognosis. The aim of this study was to identify the frequency and risk factors for the development of delirium in acute stroke.

Methods: We recruited patients consecutively admitted to neurovascular unit of city hospital between 11/05/12 and 30/06/12, within 3 days after stroke onset. Exclusion criteria were subarachnoid haemorrhage, comatose state at admission, TIA and history of psychiatric disease. The criteria of DSM-IV were used to make the diagnosis of delirium. Severity of neurological deficit was assessed by NIHSS, the functional state – modified by Rankin scale (mRS). We assessed frontal lobe reflexes: snout, nasolabial, palmo-mental and grasp reflexes. Stroke lesion location, cortical and central cerebral atrophy and anterior and posterior white matter lesions (WML) were assessed on admission head CT.

Results: Ninety six of 271 admitted patients were included in the study. M/F ratio was 50/46, mean age was 68.02 STD 10.52; median NIHSS was 8 with interquartile range (IQR) 5–12.5; mRS – 3, IQR 2–4.

Twenty two of 96 patients revealed symptoms of delirium. Patients of older age, those with severe stroke, with prominent atrophy and WML, those with fever, with urine catheter and with positive snout reflex were more likely to develop delirium. Severity of posterior WML was the only independent predictor of delirium according to multivariate analysis.

Conclusion: Delirium affected 23% of stroke patients in our cohort. Chronic brain changes and stroke complications were the main risk factors for the development of delirium.

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Abstract – WCN 2013

No: 728

Topic: 3 – Stroke

Could pro-Bnp, uric acid, bilirubin, albumin and transferrin be used in making the distinction between stroke subtypes?

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The importance of establishing the pathogenesis of ischemic strokes in determining an adequate therapy.

To observe if certain plasma parameters could be used as biomarkers in distinguishing between stroke subtypes.

Plasma pro-BNP (chemiluminescence), serum uric acid, bilirubin (colorimetric assay), albumin and transferrin (nephelometric assay) levels were performed in 168 admitted patients (mean age 68.7 ± 11.6 years, 52 men and 116 women) with different subtypes of acute ischemic strokes within 24 h and at 7 days after stroke onset as TOAST and OCSF criteria, NIHSS and Glasgow Coma Score at baseline and at 7 days were used.

The mean value of pro-BNP level was significantly higher in the cardioembolic stroke (CE), in patients, within 24 h ($p < 0.001$) and at 7 days ($p < 0.001$) after stroke onset. A negative correlation between pro-BNP levels and GCS ($r = 0.05$, $p < 0.0002$) and a significant difference between pro-BNP levels of NIHSS groups were observed ($p < 0.08$, respectively $p < 0.01$). We observed significantly higher values within 24 h of uric acid ($p < 0.05$), significantly lower values within 24 h of transferrin ($p < 0.05$), significantly lower values at 7 days of albumin and transferrin ($p < 0.001$), and significantly higher values at 7 days of uric acid and bilirubin ($p < 0.001$). No significant statistical differences between the values of oxidative stress parameters and stroke subtypes, GCS and NIHSS score were observed.

The level of plasma pro-BNP may be useful in distinguishing CE stroke from other stroke subtypes. Oxidative stress parameters could not be used to differentiate stroke subtypes.

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Abstract – WCN 2013

No: 820

Topic: 3 – Stroke

Col4a2 mutation causing recurrent intracerebral hemorrhage – Importance of screening both Col4a1 and Col4a2 in ICH of unknown origin

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Background: Type IV collagen $\alpha 1$ and $\alpha 2$ chains form heterotrimers that constitute an essential component of basement membranes including those of the cerebral vasculature. Mutations in COL4A1, encoding the $\alpha 1$ chain, cause a large spectrum of clinical manifestations, including porencephaly, bleeding prone cerebral small vessel disease, intracranial aneurysms and variable systemic abnormalities. Mutations in COL4A2 have recently been reported in a few porencephaly families.

Objective: Herein, we report on a young adult patient with recurrent intracerebral hemorrhage, leukoencephalopathy, intracranial aneurysms, nephropathy and myopathy associated with a novel COL4A2 mutation.

Methods: We examined an otherwise healthy 29-year-old male patient with recurrent deep intracerebral hemorrhages occurring after physical exertion. General physical and neurological examination, brain MRI, abdominal ultrasound, ophthalmologic examination, laboratory tests and genetic testing of COL4A1 and COL4A2 were performed.

Results: MRI showed deep intracerebral hemorrhages of different ages, diffuse leukoencephalopathy, multiple cerebral microbleeds and small aneurysms of the carotid siphon bilaterally. Laboratory workup revealed significant microscopic hematuria and elevation of creatine-kinase. We found a glycine mutation within the COL4A2 triple helical domain.

Conclusions: This study reports the first COL4A2 mutated adult patient with a phenotype very close to the spectrum of manifestations in COL4A1 mutations, and emphasizes the importance of screening both

COL4A1 and COL4A2 in patients showing recurrent ICH of unknown etiology particularly if associated with leukoencephalopathy.

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Abstract – WCN 2013

No: 801

Topic: 3 – Stroke

Patent foramen ovale prevalence in patients with acute ischemic stroke and the risk of subsequent events

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Background: Association between patent foramen ovale (PFO) and stroke is controversial.

Objective: To determine the prevalence of PFO in patients with acute ischemic event (AIE) and recurrence rate.

Patients and methods: Observational retrospective study in patients admitted to a neurology ward with AIE in 5 years. In those with transesophageal echocardiogram (TEE), clinical data, PFO presence, lesion pattern and recurrent events were collected. We present descriptive and inferential statistics with *t*-test, chi-square and recurrence risk by Kaplan–Meier curves with LogRank, using 95% confidence intervals (95%CI).

Results: Of 1300 subjects, 337 underwent TEE (25.9%), mean age 55.7 years, and 62.9% men. PFO was identified in 69 (20.5%; 95% CI 16.2–24.8%). Prevalence was similar in cryptogenic events (21.1%; 95% CI 17.0–25.3%). There was a negative association with diabetes ($p = 0.003$), hyperlipidemia ($p = 0.03$) and smoking ($p = 0.047$). No association was found with other risk factors or lesion pattern. Paradoxical embolism sources were evaluated in half and found in 23.2%. Six PFO (8.7%) underwent percutaneous closure. In a mean follow-up of 604.7 days, recurrence was low in patients with PFO, indistinguishable from those without PFO (4.8% vs 11.8%, LogRank $p = 0.157$). Patients with PFO under anticoagulation had fewer recurrent events than those non-anticoagulated (0.0% vs 7.5%) and those without PFO with/without anticoagulation (8.5%/12.8%), although not significant (LogRank $p = 0.174$).

Conclusion: The prevalence of PFO was similar to the general population, even in cryptogenic events. The negative association with some risk factors and the presence of paradoxical embolism source suggest an implication of the PFO in the event etiology, although not associated with recurrence.

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Abstract – WCN 2013

No: 815

Topic: 3 – Stroke

Rotational vertebral artery occlusion: Mechanisms and long-term outcome

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Background and objective: To elucidate the mechanisms and prognosis of rotational vertebral artery occlusion (RVAO).

Materials and methods: We analyzed clinical and radiological characteristics, patterns of induced nystagmus, and outcome in 21

patients (13 men, aged 29–77 years) with RVAO documented by dynamic cerebral angiography over an 8-year period at three University Hospitals in Korea. The follow-up periods ranged from 5 to 91 months (median = 37.5 months). Most patients ($n = 19$, 90.5%) received conservative treatments.

Results: All the patients developed vertigo accompanied by tinnitus (38%), fainting (24%), or blurred vision (19%). Only 12 (57.1%) patients showed the typical pattern of RVAO during dynamic cerebral angiography, a compression of the dominant VA at the C1-2 level during contralateral head rotation. The induced nystagmus was mostly downbeat with horizontal and torsional components beating toward the compressed VA side. None of the patients with conservative treatments developed posterior circulation stroke, and four of them (21.1%) showed resolution of symptoms during the follow-ups.

Conclusions: RVAO has various patterns of VA compression, and favorable long-term outcome with conservative treatments. In most patients with RVAO, the symptoms may be ascribed to asymmetrical excitation of the bilateral labyrinth induced by transient ischemia and/or by disinhibition from inferior cerebellar hypoperfusion. Conservative management might be considered as the first-line treatment of RVAO.

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Abstract – WCN 2013

No: 795

Topic: 3 – Stroke

Metabolic syndrome is strongly associated with ischemic stroke and carotid stenosis in non-diabetics

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Background: Insulin resistance (IR) with compensatory hyperinsulinemia plays a crucial role in the pathogenesis of atherosclerosis, but their role in ischemic stroke (IST) has not been elucidated. Study was aimed to analyze IR and plasma insulin (PI) levels, dyslipidemia pattern, obesity and plasminogen activator inhibitor-1 in 100 patients with atherothrombotic IST (group A), 100 patients with asymptomatic carotid stenosis (ACAS) $\geq 50\%$ (group B), 100 patients with lacunar stroke (group C) and 65 healthy controls (group D). Patients with diabetes mellitus and ischemic heart disease were excluded.

Methods: IR was determined by Homeostasis Assessment Model, PI levels by radioimmunoassay. Total-, LDL- and HDL-cholesterol and triglyceride levels were measured in all groups. Central obesity was determined by waist circumference and hypercoagulable state by plasminogen activator inhibitor (PAI-1) levels.

Results: IR was significantly higher in group A compared to groups B, C and D (4.82 ± 0.27 vs. 3.69 ± 0.22 , $p < 0.05$; 4.82 ± 0.27 vs. 2.71 ± 0.21 , $p < 0.01$, 4.82 ± 0.27 vs. 1.50 ± 0.19 , $p < 0.01$). PI levels were significantly higher in group A in comparison to groups B, C and D (19.00 ± 1.2 vs. 15.95 ± 0.88 , $p < 0.05$; 19.00 ± 1.2 vs. 11.12 ± 0.19 , $p < 0.01$, 19.00 ± 1.2 vs. 7.12 ± 1.08). Different patterns of dyslipidemia were observed in ACAS and IST. PAI-1 levels, and waist circumference were significantly higher in groups A, B, and C in comparison to controls ($p < 0.01$, respectively).

Conclusion: Our results indicate that all subtypes of ischemic stroke as well as ACAS are strongly associated with IR and increased PI and PAI-1 levels. Specific patterns of dyslipidemia between ACAS and IST were observed.

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Abstract – WCN 2013**No: 804****Topic: 3 – Stroke****Dual antiplatelet therapy clopidogrel with low dose cilostazol intensified platelet inhibition in patients with ischemic stroke**

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Objective: We previously reported that antiplatelet action is intensified with combined use of clopidogrel and cilostazol in ischemic stroke patients using the VerifyNow P2Y12 Assay. In this study, the relationship between cilostazol dose and platelet function with combination therapy was investigated.

Methods: The subjects were 231 patients with non-cardiogenic ischemic stroke patients treated at our hospital (18 patients with a combination of clopidogrel 75 mg and cilostazol 100 mg, 52 patients with a combination of clopidogrel 75 mg and cilostazol 200 mg, 126 patients with clopidogrel 75 mg alone, and 35 patients with cilostazol 200 mg alone). Platelet function with 20 μ M adenosine diphosphate was measured using the VerifyNow P2Y12 Assay. Clopidogrel resistance was defined as P2Y12 Reaction Units (PRU) >230 and/or % inhibition <20%.

Results: PRU was >230 in 32 patients (25.4%) receiving clopidogrel alone, 1 patient (5.6%) receiving combination therapy with cilostazol 100 mg, and 1 patient (1.9%) receiving combination therapy with cilostazol 200 mg. The percent inhibition was <20% in 41 patients (32.5%) receiving clopidogrel alone, 1 patient (5.6%) receiving a combination with cilostazol 100 mg, and 1 patient (1.9%) receiving a combination with cilostazol 200 mg. The rate of PRU >230 and/or % inhibition <20% was significantly lower in both cilostazol combination groups than in the clopidogrel alone group.

Conclusion: Clopidogrel resistance was clearly decreased with combination clopidogrel 75 mg and low-dose (100 mg) cilostazol therapy. The combination therapy with clopidogrel and low-dose cilostazol may be one means to overcome clopidogrel resistance.

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Abstract – WCN 2013**No: 708****Topic: 3 – Stroke****A novel pathway for atrial fibrillation: Concise, practical, easy-to-use**

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Background: Atrial fibrillation (AF) is the most common cardiac arrhythmia and increases the annual risk of stroke by five-fold. A recent study at our center demonstrated that a significant proportion of patients (43.6%) with known AF presenting with acute ischemic stroke were not on any antithrombotic therapy, with only 1 of 58 patients at high risk of stroke on therapeutic anticoagulation. While there are guidelines for the management of AF, a care pathway to optimize AF management is not widely practiced.

Objective: To develop a novel one-page AF pathway that is practical, relevant and easily adopted by all physicians, be it in hospital or in primary practice.

Material and methods: The latest available guidelines on AF management in USA, Europe, UK, Canada and Singapore were reviewed, with added emphasis on pathways and algorithms in these countries.

Results: A single-page pathway consolidating key components of diagnosis and management of AF was developed. A table juxtaposing the CHADS₂ and HAS-BLED score was incorporated for direct comparison and documentation of risk-versus-benefit of anti-coagulation. The modified Rankin Scale (mRS) was included, as pre-morbid functional status is an important consideration prior to the commencement of antithrombotic therapy. The pathway was created for use by any general physician, with sections specially color-coded for cardiologist input if needed.

Conclusion: A well-established AF care pathway is currently lacking in clinical practice. Our one-page pathway serves as a ready-to-use guideline-cum-management reference, as well as a possible research and audit tool in all patients presenting with AF.

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Abstract – WCN 2013**No: 783****Topic: 3 – Stroke****Early clinical manifestations in patients with cerebral venous thrombosis**

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Introduction: Cerebral venous thrombosis (CVT) has a diverse etiology, sometimes very hard to establish, representing a major neurological emergency.

Objectives: Early recognizing onset symptoms and fast diagnosis in patients with CVT.

Method: This is a retrospective study which included 18 cases with CVT admitted in the Neurology clinic of the Mureş Clinical County Emergency Hospital during 01.01.2012–31.12.2012, noticing the onset, early clinical manifestations, age, sex, demographics and affected venous area. For establishing the diagnosis, the patients have been investigated using paraclinical (electroencephalogram, eye examination, blood test results) and imagistic (computed tomography, magnetic resonance, angio magnetic resonance) investigations.

Results: From the total of 1196 patients with stroke, CVT represented 1.50%. Out of the studied cases, the onset was acute in 50%, 27.77% were subacute and 22.22% were chronic. The most frequent early symptom was headache (44.44%), followed by cranial nerve syndrome (27.77%), seizures (11.11%), aphasia (11.11%), among loss of consciousness, focal neurologic deficit, nystagmus and papilledema (5.55%). Most often, the lesions were located in the superior transverse sinus (33.33%), sigmoid sinus (27.77%), superior sagittal sinus (22.22%), cavernous sinus (11.11%) and deep venous system (5.55%).

Conclusions: The most common form of debut in CVT is the atypical form. Recognizing this pathology is essential for prompt treatment in particular clinical circumstances.

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Abstract – WCN 2013**No: 785****Topic: 3 – Stroke****Influence of serum glucose on the risk of symptomatic haemorrhagic transformation in non-diabetic patients treated by thrombolysis for supratentorial stroke**

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Background: Intravenous (i.v.) recombinant tissue plasminogen activator (rt-PA) improves outcome in ischaemic strokes, despite an increased risk of symptomatic haemorrhagic transformation (sHT). As most studies also included diabetic patients, part of the effect may be due to micro-angiopathy.

Objective: To test the hypothesis that baseline serum glucose concentration is associated with sHT in non-diabetic patients treated by i.v. rt-PA.

Methods: We studied the influence of baseline serum glucose concentrations on sHT (ECASS2 definition) in non-diabetic patients. Secondary end-points were death <7 days, between 8 days and 3 months, and <3 months, and unfavourable outcome at 3 months (modified Rankin scale 2 to 6).

Results: 505 consecutive patients met inclusion criteria (242 men [47.9%]; median age 71 years; median baseline national institutes of health stroke scale score 12; 37 with sHT [7.3%]). Baseline serum glucose concentrations were higher in patients with sHT, death between 8 days and 3 months, death and unfavourable outcome, and tended to be in patients with death <7 days. After adjustment, baseline serum glucose concentrations were independently associated with sHT (adjOR: 1.176 for 1 mmol/l increase; 95% CI: 1.020–1.357; $p = 0.025$), but not with death (<7 days, <3 months, 8 days to 3 months), and unfavourable outcome.

Conclusion: Increased serum glucose concentrations in non-diabetic patients treated by i.v. rtPA for cerebral ischaemia are associated with an increased risk of sHT, but not with death or unfavourable outcome after adjustment on confounders.

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Abstract – WCN 2013**No: 793****Topic: 3 – Stroke****Sexual life of post-stroke women with mild or no disability: A qualitative study**

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Objective: To investigate the perspectives and experiences of post-stroke women regarding sexual issues, along with interactions in social life.

Material and methods: A qualitative design was adopted for the study. According to the Rankin score with mild or no disability, a convenience sample of women ($n = 16$, married and ≥ 18) with the history of at least 6-month stroke and active sexual life prior to stroke were admitted. Individual and audio-taped interviews were performed. Transcribed interviews were qualitatively and thematically analysed using content data analysis to code and categorize emerging themes.

Results: Four principal themes were identified as follows: physical, emotional and spiritual changes experienced after stroke; negative effects of changes on the roles of post-stroke women as housewives and mothers; changes of sexual life experienced by women in post-

stroke period; and, support from healthcare providers in coping with emotional and sexual challenges.

Conclusion: In light of our findings, it is definite that physical, social and emotional challenges commonly influence sexual lives of post-stroke women, and that such women are lack of medical counselling and assistance. Therefore, in the management of post-stroke women, the following recommendations may be suggested:

- To provide stroke women and their spouses with counselling and assistance, healthcare professionals, during in-service training, should concentrate on physical and psychosocial sequelae of stroke related to sexuality.
- To help women cope with their physical, social and emotional challenges in post-stroke period, easily accessible health facilities and opportunities should be formed under the supervision of the ministry of health for stroke rehabilitation.

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Abstract – WCN 2013**No: 857****Topic: 3 – Stroke****A prospective study of vascular cognitive impairment in patients with stroke**

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Background: Cognitive impairment occurs in around 30% patients following stroke. It has been extensively studied in the developed countries recently, especially in the last decade. However, there is few data regarding post stroke vascular cognitive impairment (VCI) from developing countries like India.

Objective: The purpose of this study was to determine various factors including demographic, clinical, neuro-imaging, laboratory parameters which contribute to the development of vascular cognitive impairment (VCI) in patients with stroke.

Material and methods: In the interim analysis of this prospective study, 56 patients fulfilling inclusion criteria were evaluated. All of them underwent clinical examination including cognitive assessment, neuro-imaging in the form of MRI. Repeat assessment was done at 3 and 6 months. Patients with VCI included those with vascular dementia (VaD, as diagnosed by NINDS-AIREN criteria) and VCI-ND (VCI–no dementia). **Results:** Of the 56 patients presented in this interim analysis, 22 (39%; VaD = 8, VCI-ND = 14) patients developed VCI. Factors associated with the development of VCI included hypertension ($p = 0.012$), prior stroke ($p = 0.013$), strategic site infarction ($p = < 0.001$), high blood sugar level on admission ($p = 0.011$), high LDL level levels ($p = 0.006$) and NIHSS score ($p = < 0.001$).

Conclusion: Post-stroke cognitive impairment is commonly seen. Our study revealed that post stroke cognitive impairment occurs as a result of stroke itself as well as its risk factors such as hypertension and high LDL levels. Prior history of stroke, high blood sugar level and stroke severity on admission also contribute significantly to post stroke cognitive decline.

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Abstract – WCN 2013**No: 859****Topic: 3 – Stroke****Comparison of stroke severity and comorbidity between patients with acute stroke in basilar (Ba) or medial cerebral arteries (Mca) areas**

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Background: Neurologic deficit severity and co-morbidities impact prognosis in patients with stroke.

Objective: Comparison of survival, co-morbidity frequencies, complications and length of hospitalization between patients with an acute stroke in either BA or MCA area.

Methods: We performed prospective, observational, single-center study on consecutive 69 patients allocated according to stroke area in group 1: BA (30 patients, 23 males); group 2: MCA (39 patients, 24 males). Age, Glasgow coma scale (GCS), The National Institute of Health Stroke Scale (NIHSS), complications as mechanical ventilation (MV), infections, acute myocardial infarction (AMI), hospital length of stay and survival were compared among groups, as well as co-morbidity frequency: atrial fibrillation (AF), diabetes mellitus (DM), hypertension (HTN), and hyperlipidemia.

Results: Age was similar in both groups. Women were older than men in group 1 (71.1 vs. 57.2, $p = 0.0046$). At the admission into ICU, GCS was lower in group 1 than group 2 (8.16 ± 3.92 vs. 10.85 ± 5.37 , $p = 0.025$) and NIHSS was greater (28.79 ± 11.38 vs. 19.17 ± 9.88 , $p = 0.0006$). Complications occurred similarly in both groups: MV, infections, AMI (all $p > 0.05$). Group 1 had longer ICU length of stay (12.6 ± 20.4 vs. 6.9 ± 5.8 , $p = 0.17$), hospitalization duration (18.9 ± 24.4 vs. 12.8 ± 9.2 , $p = 0.13$) and greater NIHSS at dismissal (14.12 ± 13.55 vs. 10.46 ± 7.07 , $p = 0.38$). Survival was similar in both groups (53% vs. 46%, $p = 0.73$). Atrial fibrillation was more frequent in group 2 ($\chi^2 = 3.859$, $P = 0.044$).

Conclusion: Acute stroke in region of BA was more severe. There were no differences comparing to MCA stroke for complication rate, ICU length of stay and survival. In MCA stroke AF was more frequent.

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Abstract – WCN 2013

No: 861

Topic: 3 – Stroke

Gamma-glutamyl transferase levels and cerebrovascular risk factors in patients with acute ischemic stroke

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Objective: The aim of this study was to investigate the relationship between gamma-glutamyl transferase (GGT) levels and cerebrovascular risk factors in patients with acute ischemic stroke (AIS).

Patients and methods: Sixty patients with AIS and 44 controls who had no cerebrovascular disease were included to the study. The patients were divided into four groups as those with total and partial anterior circulation infarcts (TACI and PACI, respectively), lacunar infarcts (LACI) and posterior circulation infarcts (POCI). All patients and controls were evaluated for GGT levels and the presence of diabetes mellitus (DM), hypertension (HT), and hyperlipidemia (HL). The patients with AIS were classified as normal or those with nonstenotic carotid plaques, internal carotid artery stenosis, and vertebrobasilar insufficiency as evaluated by carotid, and vertebral artery Doppler ultrasound examinations.

Results: The frequency of DM, HT, HL, and gender distributions was similar in all groups. The mean GGT levels were significantly higher in patients with AIS. There was no relationship with GGT levels and the largeness and distribution of infarct areas. The mean GGT levels were at their nadir in LACI, while at their maximal levels in the PACI group. The lowest GGT levels were detected in patients with nonstenotic plaques.

Conclusion: Higher GGT levels in AIS patients support the relationship of GGT with inflammation and oxidative stress. However absence of any

correlation among GGT levels, infarct areas and Doppler findings suggests that GGT is not a marker of degree of ischemia.

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Abstract – WCN 2013

No: 863

Topic: 3 – Stroke

Cerebral venous thrombosis: Patients with and without parenchymal lesions

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Background: The prognosis of cerebral venous thrombosis (CVT) may range from complete recovery to death. If CVT patients show parenchymal involvement, such as venous infarction with or without hemorrhage, their clinical pictures may be more severe.

Objective: To analyze clinical features, radiological findings, risk factors and prognostic outcomes in CVT patients with (CVTP) and without (CVTW) parenchymal lesions.

Patients and methods: We recruited 37 patients with CVT who were admitted to our clinic between 2005 and 2011. CVTW group included 22 patients (11 women, 11 men with ages ranging from 29 to 46 years), remaining 15 patients (8 women, 7 men within 23 to 55 age range) belonged to CVTP group.

Results: CVTW cases had longer diagnostic delay than the CVTP cases: 18.7 ± 19.6 days and 5.2 ± 5.4 days, respectively ($p < 0.01$); CVTW cases more often presented isolated intracranial hypertension than CVTP cases: 21/22 vs 2/15 ($p < 0.00$). CVTP cases admitted the hospital more often than CVTW cases with decreased alertness (4/15 vs 0/22 $p < 0.02$), seizures (9/15 vs 2/22, $p < 0.00$) and motor deficits (9/15 vs 1/22, $p < 0.00$). CVTP patients had more often deep venous thrombosis and genetic risk factors, the difference being only marginally significant ($p = 0.06$). CVTW group had good outcome. In CVTP group, bad outcome was correlated with involvement of parenchymal area ($p < 0.001$).

Conclusions:

- (i) CVTW may frequently be related to isolated intracranial hypertension and have longer diagnostic delay with benign course;
- (ii) CVTP may have more serious clinical pictures.

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Abstract – WCN 2013

No: 848

Topic: 3 – Stroke

Ataxic disorders after acute isolated cerebellar infarction

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Background: Considering somatotopic organization of cerebellar functions, investigations of disease progression ataxic disorders in patients taking into account the localization of infarction lesion remain a permanent challenge.

Objective: To investigate disease progression in ataxic disorders in patients with acute isolated cerebellar infarction depending on affected vascular territory.

Material and methods: 20 patients (13 men and 7 women, mean age 57.9 ± 14.7) after acute isolated cerebellar infarction were examined by neurological scales (ICARS), transcranial Doppler, duplex and triplex scanning, brain MRI.

Results: The MRI showed infarctions of SCA in 9 patients, infarctions of PICA in 8, and infarctions of AICA in 3. Ataxic disorders depended

on the localization of infarction lesion and were different according to Fisher's test ($p < 0.05$): patients with SCA infarctions presented with dysarthria, limb ataxia due to a lesion of the paravermis and lateral parts of the anterior lobe from lobules II to V; typical signs of PICA infarction included dizziness, gait ataxia, nystagmus associated with a lesion of posterior inferior lobules IX–X of the posterior lobe of the hemisphere; AICA infarctions were manifested with dizziness, vestibular disturbances, nystagmus and associated with affected anterior inferior and medial cortex of the cerebellar hemisphere.

Conclusions: The study demonstrated that clinical assessment of the early signs of cerebellar ataxia after isolated infarction, consideration of guiding symptoms of ataxia syndrome and the use of visualization enables to determine vascular territory lesions in the cerebellum.

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Abstract – WCN 2013

No: 824

Topic: 3 – Stroke

Long-term risk of stroke and myocardial infarction in women with low social support in Russia: Monica-psychosocial epidemiological study

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Objective: To study the effect of social support (SS) on relative risk of stroke and myocardial infarction (MI) in female population of 25–64 years in Russia over 16 years of follow-up.

Material and methods: Under the third screening of the WHO “MONICA-psychosocial” program random representative sample of women aged 25–64 years ($n = 870$) were surveyed in Novosibirsk. Berkman-Sym test was used to measure indices of close contacts (ICC) and social network (SNI). From 1995 to 2010, women were followed for the incidence of stroke and MI by using Myocardial Infarction Registry data. Cox regression model was used for relative risk assessment (HR) of stroke, MI.

Results: Low levels of ICC and SNI were revealed in 57.1% and 77.7% of women, respectively. Stroke was developed in 5.1% of women, MI – in 2.2%.

HR of MI over 16 years of study was in 4.9-fold (95.0% CI: 1.108–21.762; $p < 0.05$) and 2.9-fold (95.0% CI: 1.040–8.208; $p < 0.05$) higher for low ICC and SNI, respectively, compared to higher SS levels. HR of stroke was 4.1 (95.0% CI: 1.193–14.055; $p < 0.05$) and 2.7 (95.0% CI: 1.094–6.763; $p < 0.05$) for low ICC and SNI, respectively. Married and unmarried women with low ICC and hard physical laborers with low ICC and SNI had higher rate of stroke development (p for all < 0.05).

Conclusions: The prevalence of low SS in women aged 25–64 in Russia is high. Risk of stroke and MI over 16 years of study was significantly higher in women with low ICC and SNI, especially in married ones in manual labor class.

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Abstract – WCN 2013

No: 887

Topic: 3 – Stroke

Predictors of short-term outcome in posterior circulation strokes

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Prediction of acute ischemic stroke outcome is essential for treatment planning, guidance of patient and relatives, and in the search for new

therapeutic strategies. Previous examinations have focused mostly on anterior circulation infarcts with few studies examining diffusion-weighted MRI (DWI) data and functional outcome in posterior circulation (PC) infarcts.

Objective: To investigate predictors of short-term mortality and major disability in patients with PC ischemic strokes.

Patients and methods: We evaluated prospectively 145 consecutive patients (85 men and 60 women) aged 32 to 85 years in acute period of ischemic PC strokes. Comprehensive examination included analysis of the baseline characteristics, risk factors; attentive clinical study; assessment of neurological status with the use of scales NIHSS, B. Hoffenberth et al., modified Rankin scale (mRS), Barthel index; laboratory data. Localization and size of the ischemic lesion were verified with the DWI.

Results: All patients at 21-days of follow-up according to the mRS were classified into the favorable (F) group (mRS 0–2) – 92 patients (63.4%) and unfavorable (U) group (mRS 3–6) – 53 patients (36.6%). Carried out statistic analysis revealed NIHSS score ($p < 0.001$), B. Hoffenberth et al. (1990) score ($p < 0.001$), Glasgow coma scale score ($p < 0.001$), atrial fibrillation ($p = 0.03$), cardioembolic subtype of stroke ($p = 0.004$), transient ischemic attacks (TIA) in anamnesis ($p = 0.02$) and multiple PC intracranial territories involvement ($p < 0.001$) correlated with poor outcome.

Conclusions: Stroke severity, its mechanism, TIA and atrial fibrillation, lesion location rather than lesion volume in PC strokes are critical for short-term functional outcome.

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Abstract – WCN 2013

No: 870

Topic: 3 – Stroke

Brain MRI findings in Fabry Disease

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Background: Stroke is a serious complication of Fabry Disease (FD) affecting 15% of males and 11% of females. Moreover FD has been identified in 4% of young patients with unexplained strokes. The evaluation of MRI in FD patients without a prior history of stroke may lead to earlier treatment.

Objective: To evaluate the presence and distribution of ischemic and hemorrhagic lesions in brain MRI of children and adults with FD lacking history of CVA.

Patients and methods: Brain MRI studies in 73 consecutive patients without history of stroke or TIA were evaluated using classic sequences as well as GRE-weighted images, for ischemic lesions and chronic microbleed detection. Of the 62 adult patients, 5 were excluded due to dialysis antecedent. Among the remaining group, 25 were males and 32 females (mean age 34 and 40.3 years old, respectively).

Results: Two patients (18.18%) under 18 years of age presented abnormal MRI. Nineteen adult patients (33.33%) had brain MRI evidence of small vessel disease in the basal ganglia, corona radiata, thalamus, brainstem, and periventricular white matter. Patients with MRI abnormalities were older (51.3 vs. 30.6 years old, $p = 0.0001$). Four patients (mean age 61 years old) presented chronic microbleeds identified by GRE, in the pallidum and thalamus. Moreover, FLAIR and T2-weighted images showed evidence of white matter and deep grey matter involvement.

Conclusion: In our group, 33.33% of adult patients with FD lacking history of CVA or prior dialysis had evidence of small vessel disease on MRI and 21.05% of those presented cerebral microbleeds.

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Abstract – WCN 2013

No: 950

Topic: 3 – Stroke

Internuclear ophthalmoplegia caused by an isolated ischemic lesion affecting the medial longitudinal fascicle

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Background: Internuclear ophthalmoplegia (INO) frequently appears in multiple sclerosis, but is rare as an isolated sign in stroke.

Objective: To present clinical and imaging data of 3 cases of ischemic stroke with uni- or bilateral INO.

Patients and methods: Two patients with unilateral and another one with bilateral INO were investigated by a 3 Tesla Philips Achieva scanner.

Results: A 66-year-old female and a 62-year-old male were admitted with acute right sided and an 80-year-old male with bilateral INO. Diffusion weighted MRI scans (DWI) confirmed restricted water diffusion in the medial longitudinal fascicle (MLF) in all cases. Unilateral INO was associated with unilateral MLF lesion whereas in the patient with bilateral signs the MLF was also affected on both sides. Hypertension was present in all three patients. Two patients had hyperlipidemia, one had diabetes, and one was a heavy smoker. The patient with bilateral INO had malignant melanoma which could also have caused prothrombotic state. Antiplatelets (aspirin or clopidogrel) were administered in all patients. The clinical signs were resolved in the patient with bilateral INO after two months. Patients with unilateral INO also improved during the first month after their stroke and their follow-up is ongoing.

Conclusion: INO due to a small infarct affecting the MLF may cause uni- or bilateral signs. As Fisher proved, signs can be bilateral if the terminal branch of a paramedian artery divides and these vessels supply the MLF on both sides. Such small arteries are not visible however even for a 3 Tesla MRI.

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Abstract – WCN 2013

No: 949

Topic: 3 – Stroke

Assessment on preadmission treatment of cardioembolic stroke patient in Malaysian hospital

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Background: Atrial fibrillation (AF) increases the degree of stroke risk up to five times and doubled the overall mortality rate. Hence, anticoagulant as stroke primary prevention is vital.

Objectives: We aim to assess the preadmission treatment received by the cardioembolic stroke patients, and review the mortality rate according to pre-treatment received.

Method: This prospective study has been carried out from 1st Jan 2010 to 31st December 2012 by 11 Malaysian hospitals. Data have been collected by using Malaysian National Stroke Registry (NSR) case report form which then will be transferred to web application. For analysis, data was extracted and analyzed by using PASW 18.

Results: There were 126 cardioembolic stroke patients registered to NSR with mean age of 65.97 ± 12.72 years old.

Of all, only 45 patients were on Warfarin with 13 (10.3%) of them were within therapeutic and 32 patients (25.4%) received sub-therapeutic Warfarin. 3.2% of patients received dual antiplatelet therapy, 26.2% on single antiplatelet therapy and 34.9% of patient did not received any treatment.

The mortality rate was significantly high in no treatment group (34.1%) as compared to Warfarin (13.3%) and antiplatelet group (16.2%) with p value of 0.038. However, the mortality rate between Warfarin therapeutic (7.7%) with sub-therapeutic group (15.6%) and between single antiplatelet (18.7%) with dual antiplatelet were not significantly different.

Conclusion: Our findings warrant greater effort to prevent stroke in AF patients, thus high mortality outcome which was significantly associated with treatment received prior to stroke event.

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Abstract – WCN 2013

No: 937

Topic: 3 – Stroke

Establishing the aetiology of stroke in a young cohort

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Background: In 2010 in Ireland, 34.3% of the almost 23,000 strokes diagnosed occurred in patients under 65 years of age. This figure may underestimate the prevalence of stroke in young patients as some may have not been recognised or misdiagnosed.

Materials and methods: A retrospective audit of all patients under age 65 presenting with ischaemic and haemorrhagic strokes to a University Hospital between 1st January 2010 and 31st December 2011. Aetiology was determined using the TOAST criteria.

Results: A total of 80 patients under 65 were treated for stroke (13.6% of the total 586 strokes in 2010/2011). 26% [n = 21] were under 45 years. Average age was 51.1 years (range 20–64 years). 49 were males (61%), and 31 females (39%). 63 were admitted via the emergency department (ED); 10 transfers from other hospitals; 7 existing inpatients. Of 63 ED admissions, 24 (38%) patients received CT Brain within 1 h. Of 80 patients, 56 (70%) had MRI Brain during their inpatient stay. Using TOAST classification of stroke aetiology, 9 (11.3%) strokes were caused by large artery atherosclerosis, 19 (23.7%) cardioembolism, 10 (12.5%) lacunar, 13 (16.2%) other determined aetiology e.g. dissection, 16 (19.9%) undetermined, 10 (12.5%) haemorrhagic and 3 (3.7%) were iatrogenic. 57.1% of under 45 year strokes were due to other determined aetiology. 12.5% of under 65 years received thrombolysis, compared to 5.8% of all strokes.

Conclusion: The under 65 population accounts for 13.6% of strokes presenting to a tertiary referral hospital. They often require more extensive investigation to determine aetiology.

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Abstract – WCN 2013**No: 926****Topic: 3 – Stroke****Alice in Wonderland syndrome: A case report**

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Objective: Alice in Wonderland syndrome (AIWS), named after a novel that was written by Lewis Carroll, was first reported in 1955 by J. Todd. We herein report a case of AIWS following a cerebral infarction.

Patients: A 61-year-old right-handed male developed a cerebral infarction and was admitted to the hospital. He was transferred to our hospital for rehabilitation on the 19th day after the stroke. A neurological examination performed on admission revealed no abnormalities, except for mild impairment of the two-point discrimination on the right side and a mild motor aphasia. On admission he complained of a bizarre sensation in his right extremity, which he described as feeling that his right extremity length was half that of the left extremity. He was distressed by this bizarre feeling throughout the day, particularly due to a fear of falling down the stairs. The sense of a short extremity disappeared when he looked at his right limb visually. He felt that the weight of his right extremity was lighter than that of the left extremity and stated that the objects grasped tactually in his right hand were lighter in weight and smaller in size than those grasped in the left hand. No prosopagnosia or autotopagnosia were observed. MRI and SPECT images revealed broad hypoperfusion in the left hemisphere.

Discussion and conclusions: The bizarre sensation in the patient's extremity was considered to be consistent with a microsomatognosia, as part of a cluster of symptoms called AIWS. The occurrence of AIWS in stroke patient is extremely rare.

doi:10.1016/j.jns.2013.07.738

Abstract – WCN 2013**No: 657****Topic: 3 – Stroke****Effect of functional SNPs in UCP2 gene on cerebral infarction among the Korean older male subjects**

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Stroke is a serious disease which can cause death. Many genetic polymorphisms in many genes are known to be related with stroke, cerebral infarction and cardiovascular disease. In this study, we investigated the relationship between UCP2 polymorphisms and stroke among the Korean population. A total of 586 normal subjects and 1857 stroke patients (184 ischemic hemorrhage, 1659 cerebral infarction) were enrolled. Three SNPs of UCP2, G-1957A, G-866A and A55V were genotyped by the TaqMan method. Association of the UCP2 SNPs with stroke and blood parameters was statistically analyzed by logistic regression or general linear model using SAS 9.0. The G-866A and A55V UCP2 SNPs were related with stroke ($p = 0.029$, $p = 0.039$ in dominant model). These SNPs were significantly associated with cerebral infarction among stroke types. The frequency of subjects with A allele at the -866 position and the V allele at amino acid 55 was, respectively, 71.25% and 70.23% which was significantly lower than the frequencies observed for frequencies observed for G-866A (76.62%) and A55V (75.64%) in the normal group ($p = 0.023$ and $p = 0.028$, respectively). This association varied according to gender and age. The G-866A and A55V SNPs were significantly associated with cerebral infarction in male and

older subjects. These two SNPs were also linked with decrease of serum triglyceride levels in the normal male subjects. This study showed that the G-866A altering of promoter activity and A55V changing of amino acid were significantly related with stroke, especially cerebral infarction in the Korean male population.

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Abstract – WCN 2013**No: 922****Topic: 3 – Stroke****A clinical study of the patients with “ataxia optique”**

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Objective: So-called “ataxia optique” is a disorder of hand movement in which the patient is unable to grasp objects located in the peripheral visual field. This study was performed to evaluate the neuropsychological and neuroradiological findings of this disorder.

Patients: We studied six patients with ataxia optique, 56–79 years of age. All subjects were able to grasp objects located in the central visual field. Four patients bumped into the contralateral objects due to the presence of lesions.

“Ataxia optique”: Two patients had lesions in the left hemisphere. In one of these patients, reaching with both hands was affected in the contralateral visual field and the right hand in the ipsilateral field. The other patient demonstrated impairment in the ability to reach with the right hand in the contralateral field. Two patients with right hemisphere lesions exhibited reaching impairments with both hands in the right and left visual fields. Two patients had bilateral hemisphere lesions, and their ability to reach with either hand was affected in both the right and left visual fields. The SPECT analysis (vbSEE) revealed the presence of a converging region, including the inferior parietal lobule, angular gyrus, and superior parietal lobule.

Discussion and conclusions: The pattern of the reaching impairments may differ according to the side of the hemisphere lesion. The results of the SPECT analysis indicated that the localization leading to ataxia optique occurs in the above-mentioned converging region, thus leading to an impairment of the visual dorsal stream.

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Abstract – WCN 2013**No: 924****Topic: 3 – Stroke****A pure form of amorphognosia: A case report**

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Background: Two types of tactile agnosia have been reported, apperceptive and associative. Recently we encountered a patient who showed an amorphognosia, but hylognosia and tactile object recognition were not impaired.

Case report: A 54-year-old, right-handed female suddenly developed a left hand numbness in Jan. 2011. She was admitted to a hospital and pointed out a cerebral infarction in the right frontoparietal region due to Moyamoya disease. On June 22, she was admitted to our hospital for rehabilitation. The neurological examinations revealed no abnormal findings. Neuropsychological examinations such as MMSE, WMS-R, WAIS III, Kohs block design test, BADS, BIT, and CAT were all within normal ranges. The somatosensory functions such as response to light

touch, tactile localization, double simultaneous stimulation and two point discrimination were normal. No tactile extinction was observed. Weight and texture discrimination, and objects recognition presented tactually were all correct in both hands. She only missed to recognize the two dimensional shapes such as triangle, square, rectangular, and so on, and three dimensional shapes such as cube, parallelepiped, sphere, and so on, in her left hand, but not in the right hand. MRI and SPECT images revealed a broad hypoperfusion in the right frontoparietal region.

Discussion and conclusion: The tactile agnosia arises from a parietal lesion damaging the high level processing of somatosensory information that culminate in the structured description of the object. A pure form of amorphognosia is extremely rare. However, we could not specify the causative lesion because of the broad cerebral infarction.

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Abstract – WCN 2013

No: 687

Topic: 3 – Stroke

Seasonal variation in stroke occurrences in Japan

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Background: Seasonal variations in stroke were examined from the data in the Kyoto Stroke Registry (KSR).

Methods: We analyzed data on stroke patients identified from January 1999 to December 2009 in the Kyoto prefecture, and registered in the KSR which is administered by the local government. The study cohort had 13,788 patients divided into 9011 cerebral infarction (Celn), 3549 cerebral hemorrhage (CH) and 1197 subarachnoid hemorrhage (SAH) cases. OR and 95% confidence intervals (CI) for stroke occurrence in each season and the effects of risk factor histories (RFH) were calculated for each stroke subtype.

Results: ORs (CI) for stroke occurrence were highest in winter for Celn (1.06; 1.00–1.12) and CH (1.37; 1.25–1.51) and in spring for SAH (1.51; 1.28–1.79), with summer serving as a reference. After adjustments for age, sex and RFH (hypertension, arrhythmia, diabetes mellitus and hyperlipemia), ORs for stroke occurrence were 1.06 (0.93–1.21), 0.90 (0.78–1.03) and 1.05 (0.91–1.20) in spring, autumn and winter for Celn; 1.53 (1.22–1.93), 1.36 (1.07–1.72) and 1.61 (1.28–2.02) for CH; and 1.53 (1.06–2.23), 1.08 (0.73–1.61) and 1.36 (0.94–1.99) for SAH, respectively. Hypertension history had a significant effect on the onset of CH (0.77; 0.62–0.96, $p = 0.018$) in spring. Arrhythmia history affected the onset of Celn in autumn (1.18; 1.02–1.37) and onset of Celn (1.29; 1.12–1.48) in winter. Effects of other RFH on the seasonal occurrence of stroke did not reach statistical significance.

Conclusions: Seasonal variation remained significant after adjustments for RFH. However, RFH had significant effects on the seasonal occurrence of some stroke subtypes.

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Abstract – WCN 2013

No: 958

Topic: 3 – Stroke

Childhood strokes in SCD drop sharply since stop study/protocol adoption in the US

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Background and aims: In 1998 Adams et al. (NEJM 1998:339(1) 5–11) reported the results of the first primary prevention clinical trial in children, called the STOP study which was halted early when it was clear that Transcranial Doppler Ultrasound (TCD) had identified a high risk group (10%/year untreated risk of stroke with a TCD velocity of 200 cm/s or greater) and that regular transfusions in this high risk group had reduced first stroke dramatically (by >90%). Did these dramatic results affect childhood stroke in the US?

Methods: Review of published literature since STOP.

Results: In the last 10 years, reductions in stroke post STOP compared to before 1998 have been reported using data from:

- 1) the National Inpatient Sample (NIS);
- 2) California Hospitalization data base; and
- 3) two large SCD clinics in the US and one in France.

Recently data from the National Center for Health Statistics showed that, since STOP, the black white childhood stroke death rate disparity has been cut by 74% for ischemic stroke but not for hemorrhages, which are not prevented by the STOP protocol.

Conclusions: Phase IV clinic and national data document a major reduction in stroke in SCD since STOP. As STOP was the only evidence based change in treatment during this era, these large reductions in stroke in SCD are most likely due to wide adoption of the STOP trial results into clinical practice in the United States. Further adoption should drop rates further still.

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Abstract – WCN 2013

No: 977

Topic: 3 – Stroke

Patients with atrial fibrillation and CHa₂DS₂-Vasc ≤ 1 still develop stroke – Why so?

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Objective: To determine the influence of AF and of additional thromboembolic risk factors on stroke development and evolution with comparison of several schemas' stroke predictive value.

Methods: Retrospective study of all ischemic stroke patients admitted during one year in a municipal hospital.

Results: The study included 735 patients with ischemic stroke of whom 519 had primary stroke (70.6%). AF was determined in 206 cases (28.4%), these patients being older (70.1 ± 0.65 vs. 64.3 ± 0.46 years, $p < 0.001$), mainly females – 57.8% (119/206) and having more vascular risk factors. AF patients have had more severe signs of cerebral lesion on admission (79% vs. 37%, $p < 0.01$), also with a higher hospital mortality rate (30.6% vs. 13.2, $p < 0.001$). According to CHADS₂ score, about 37.4% of patients with AF and stroke were included in the low and medium stroke risk groups, but still have had stroke. Upon analysis 46% of them were women, 38% were aged between 65 and 75 years, and 36.7% were having coronary heart disease, including 16% old myocardial infarction and 7.7 – intermittent claudication. By including these factors in the CHA₂DS₂-VASc score, 0% had a low risk, and 4.1% had a medium one and 95.9% had a high thromboembolic risk.

Conclusions: Patients with AF without previous thromboembolic events have a high risk of stroke with more severe evolution and consequences. Female gender, age between 65 and 75 years and vascular disorders should be taken into consideration when calculating the thromboembolic risk, CHA₂DS₂-VASc being the most sensible score in determining thromboembolic risk in patients with non-valvular AF.

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Abstract – WCN 2013**No: 1020****Topic: 3 – Stroke****Clinical feasibility and usefulness of three depression scales in acute stroke patients**

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Objectives: To investigate clinical feasibility and usefulness of three depression scales, which are Beck depression inventory (BDI), aphasic depression rating scale (ADRS), and emotional behavior index (EBI) in acute stroke patients.

Methods: Twenty stroke patients, 2 weeks to 4 weeks after onset, were prospectively recruited. BDI was recorded by self or caregiver with yes/no question. ADRS and EBI were checked by observing patient's behavior for three days. EBI which recorded behaviors most frequently observed, had 7 domains such as sadness, aggressiveness, disinhibition, adaptation, passivity, indifference, and denial. Descriptively we analyzed two rating scales and one behavior index.

Results: 13 patients had left hemisphere lesions, and 7 had right lesions. For BDI, only 9 patients could be evaluated because 11 patients had global aphasia or cognitive dysfunction (MMSE < 20). In 9 patients, three patients had severe depression (BDI score > 16) and 6 patients did not have depression. For ADRS, all patients could be evaluated. Twelve patients had depression by ADRS score (>10). Among them, 10 were from BDI unfeasible 11 patients and 2 were from feasible 9 patients. For EBI, 6 patients showed sadness, 5 aggressiveness, 2 disinhibition and 1 indifference. Six patients were not observed specific emotional behavior.

Conclusion: It is difficult to assess depression by BDI in patients with communication deficiencies or severe cognitive disorders. ADRS and EBI could help access to emotional changes in these patients. Further research of large sample size and good design is needed for appropriate depression rating scale in acute stroke patients including ADRS and EBI.

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Abstract – WCN 2013**No: 986****Topic: 3 – Stroke****Hospitalized patients in the mortality rate from stroke**

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Hospital registry of the Department of Neurology, Clinical Center Nis, contains information about the treatment of more than 30,000 patients. The aim of our study was to compare the mortality rates of different types of stroke.

Statistical processing was carried SPSS V19.

In the period from 01 January 2000 to 31 December 2010 in-hospital registry of the Department of Neurology, registered 10,739 hospital admissions of patients with a diagnosis of stroke. Since there were 5476 (51%) men and 5263 (49%) were women. The average age of patients was 68.12 ± 11.36 years.

Of the total number of recorded hospital admissions, 487 (4.5%) were diagnosed with SAH, 1804 (16.8%) with a diagnosis of ICH, 7553 (70.3%) with a diagnosis of acute ischemic stroke, and 895 (8.3%) with unspecified type of stroke. In the hospital 3285 (30.6%) deaths were registered, while 7454 (69.4%) patients were discharged from the hospital stay. The mortality rate was 19.7% in patients with SAH, 47% of patients with ICH, 23.1% of patients with acute ischemic stroke and 66.5% in patients with an unspecified type of stroke.

High mortality rate was registered in the case of patients with unspecified type of stroke and bearing in mind that these are the

patients who did not set a definitive diagnosis because they died shortly after admission. Hospital mortality rate patients with ICH is somewhat higher than in the literature, because in our hospital surgery is not indicated in severe illness.

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Abstract – WCN 2013**No: 985****Topic: 3 – Stroke****Cerebrovascular reactivity and systemic haemodynamic parameters in response to acute hyperoxia in stroke and diabetic patients with stroke**

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Background: The aim was to determine the effect of acute hyperoxia on cerebrovascular reactivity (CVR), thromboxane B₂ (TXB₂) levels and systemic haemodynamic parameters in acute stroke patients (SP) and diabetic patients with stroke (DPwS).

Methods: In 26 SP (age 68 ± 6 years) and 26 DPwS (69 ± 6 years) mean blood flow velocity (MBFV) in middle cerebral artery (MCA), index of pulsatility (IP) and index of resistance (IR) were determined by transcranial Doppler before (basal) and during 15 min of hyperoxia (100% O₂ via facial mask). Systolic and diastolic blood pressure (SBP, DBP), heart rate (HR), pO₂ and TXB₂ blood levels were assessed at the same time points. Data are expressed as mean ± SD; p < 0.05 was significant.

Results: During hyperoxia MBFV significantly increased in both groups; SP (from 56 ± 11 to 60 ± 12), DPwS (from 53 ± 13 to 58 ± 16), while IP and IR or HR were not affected in both group compared with basal values. Hyperoxia only caused mild HR increase by 3% in SP and 4% in DPwS (p < 0.05). There was a significant increase in SBP, DBP and pO₂ in both groups and 2% decrease in TXB₂ levels in SP and 12% decrease in TXB₂ in DPwS during hyperoxia (p < 0.001) without significant difference between groups. TXB₂ showed weak negative correlation with MBFV in both groups (p < 0.05).

Conclusion: This study demonstrated impairment of CVR accompanied with decrease in TXB₂ levels in both groups, suggesting the role of TXB₂ in CVR. Haemodynamic parameters were not affected, except for improved oxygenation.

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Abstract – WCN 2013**No: 989****Topic: 3 – Stroke****ICH evacuation by MIN techniques**

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Objective: STICH I and II trials examined a situation that actually is not representing today's reality regarding MIN strategies and techniques. EndoSTICH trial and MISTIE trial are studying two minimally invasive techniques (endoscopic evacuation and catheter-lysis) which however do not compete the needs of the majority of hemorrhages. We elaborated a MIN technique with high effectiveness and applied it up to now in 150 cases. We present a retrospectively analysis of this first series.

Material and method: This MIN concept combined several techniques to assist microsurgery: High-end neurosonography with small probes ("burr-hole-probe/ALOKA") and mouth tracking of the microscope, both mandatory. Additionally we added endoscopy (Wolf, Aesculap, Storz) and LASER (Th-YAG Revolix).

150 patients underwent this application within 10 years by the presenting author. The approaches varied from burr-hole to 1€ or 2€ in size depending from the imaging findings and expected difficulties.

Results: In nearly all cases it was possible to evacuate the hematoma within 1 h and the hematoma evacuation decreased the ICP to normal levels. Clinical results were excellent in lobar bleedings with isochoric before surgery. Large and deep-seated hemorrhages needed longer recovery time but in all cases postop CT showed fast reduction of perifocal edema.

Conclusion: Combination of ultrasound, mouth tracking, endoscopy and LASER enabled evacuation of all types of hematoma minimal invasively and very effectively in less than one hour.

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Abstract – WCN 2013

No: 996

Topic: 3 – Stroke

The significance of serum gamma-glutamyltransferase in acute cerebral infarction

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Background: Serum gamma-glutamyltransferase (GGT) is known as a possible marker for metabolic syndrome and cardiovascular disease. The relationship between serum GGT and acute cerebral infarction is not well known.

Objective: We evaluated the significance of serum GGT in acute ischemic stroke and ischemic stroke subtype.

Patients and methods: We included 178 patients with acute cerebral infarction within three days of onset prospectively. Age- and sex-matched 180 controls were enrolled. All patients were performed brain image including MRI/MRA or CT/CTA and cardiac workup. Serum GGT in first admission day, C-reactive protein (CRP), uric acid, total cholesterol, high density lipoprotein (HDL), low density lipoprotein (LDL), leukocyte count, fasting glucose, systolic and diastolic blood pressure were measured. Stroke subtype was classified into five groups by the Trial of Org 10172 in Acute Stroke Treatment (TOAST) classification.

Results: The mean level of GGT in acute stroke patients was significantly high compared with the control group ($p < 0.001$). In subgroup analysis, the GGT was high in groups of large artery atherosclerosis, small vessel occlusion and cardiac embolism (all $p < 0.001$).

Conclusion: Serum GGT was elevated in patients with acute cerebral infarction in small vessel disease, atherosclerotic and cardiogenic stroke. There was no association between serum GGT and site of large artery atherosclerosis. Serum GGT may play roles of serum biomarker for acute cerebral infarction.

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Abstract – WCN 2013

No: 1032

Topic: 3 – Stroke

Hospital mortality associated with stroke in Southern Iran

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Background: Unlike the western hemisphere, information about stroke epidemiology in Southern Iran is scarce.

Methods: A retrospective, single-center, hospital-based longitudinal study was performed at Namazi Hospital in Shiraz, Southern Iran. Patients with a diagnosis of hemorrhagic and ischemic strokes were identified based on the International Classification of Diseases, 9th

and 10th editions for the period between 2001 and 2010. Demographics including age, gender, area of residence, socioeconomic status, length of hospital stay and discharge destinations were analyzed in association with mortality.

Results: A total of 16,351 patients were included in this analysis with the mean age 63.4 (95% CI: 63.1, 63.6). Males were slightly predominant (53.6% versus 46.4%). Fifty six percent of the total sample were older than 65, 14% were younger than 45, and 0.026% were children younger than 18. The mean hospital stay was 6.3 days (95% CI: 6.2, 6.4). Among all types of strokes, the overall hospital mortality was 20.5%. A higher stroke mortality was associated with females, children, and low socioeconomic status ($P < 0.001$).

Conclusions: In comparison to western countries, a larger proportion of patients are young adults, and the mortality rate is higher.

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Abstract – WCN 2013

No: 1033

Topic: 3 – Stroke

Allelic polymorphism of the LMP2 and PSMA6 gene in acute ischemic stroke patients in Ukrainian population

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The proteasome activity significantly changes in ischemia. Polymorphisms in genes of proteasome subunits have been studied as risk factors of cardiovascular disease but we don't find any articles concerning role of allelic polymorphism of proteasome genes in the susceptibility to acute ischemic stroke (IS). Different allelic variants of proteasome subunits PSMA6 and LMP2 can have different effects on the susceptibility to acute IS in Ukrainian population.

Methods: Fast Real Time PCR was used for determination of LMP2 (rs 4331349) and PSMA6 (rs 1048990) and detection of the various variants of the LMP2 and PSMA6 genes in 102 patients with IS and in 92 control subjects. We tested some risk factors of IS in patients with different variants of these genes.

Results: It was shown that the distribution of the major homozygotes, heterozygotes and minor homozygotes for the LMP2 gene (60Arg/His) was 55.9%, 34.3% and 9.8%, respectively (in control group: 53.3%, 43.5% and 6.7%; $p = 0.046$); for the PSMA6 (–8C/G) polymorphism the percentages were 75.5%, 21.4% and 3.1%, respectively (in control group: 80.2% homozygotes and 19.8% heterozygotes; $p = 0.22$). We identified that polymorphism of LMP2 gene (60Arg/His) associated with diabetes mellitus (DM) ($p = 0.03$). The PSMA6 (–8G/G) minor homozygotes was associated with repeated IS ($p = 0.06$).

Conclusion: We found relationship between LMP2 gene polymorphism and we didn't found relationship between PSMA6 gene polymorphism and risk of IS. We suggest that G/G promoter variant of PSMA6 can be considered as a risk factor for repeated IS and polymorphism of LMP2 gene was associated with previous DM.

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Abstract – WCN 2013

No: 1043

Topic: 3 – Stroke

Cerebral venous thrombosis – Clinical presentations amongst Sudanese patients

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Cerebral venous thrombosis (CVT) is an under diagnosed cause for acute or slowly progressive neurological deficit. It has a wide spectrum of symptoms and signs that may be clinically challenging and great mimicker.

We studied 45 patients with CVT at our centre. 41 were females (91%). The mean age was not different between the sexes (18.55 years in females and 18.35 years in males).

Of the 41 females 15 were postpartum (36.5%); five were pregnant while the other five were on oral contraceptive pills. Two were positive for antiphospholipid antibodies (APLA). Only one patient had protein C deficiency. The associations included, septic abortion (1), non-specific febrile illness (4), SLE (1), neoplasia (2), rheumatoid arthritis on methotrexate therapy (1), and indeterminate cause (2). Amongst the four males one had periorbital cellulitis, one had severe dehydration and the last two were of indeterminate cause.

We will present illustrative cases. One patient had a haemorrhagic infarct in the left temporal lobe, misdiagnosed as brain tumour. A second female patient was diagnosed as multiple sclerosis and was planned for disease modifying therapy, the third patient had marked psychiatric disturbance, while a fourth had periorbital cellulitis and developed a squint.

We identified 2 patients with venous sub-arachnoid haemorrhage – a unique presentation and 3 with suspected associated viral encephalitis – not previously reported.

MRI/MRV was the examination of choice for confirmation of cerebral venous thrombosis.

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Abstract – WCN 2013

No: 1053

Topic: 3 – Stroke

Ibudilast inhibits Th17 differentiation

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Background: T helper cells secreting interleukin (IL)-17 (Th17 cells) play a crucial role in the pathogenesis of autoimmune diseases, such as multiple sclerosis. In addition, Th17 cells are associated with secondary inflammation induced by cranial infarction. Ibudilast, a nonselective phosphodiesterase inhibitor that influences inflammation and neurodegeneration in patients with ischemic stroke and multiple sclerosis, is used in Japan to treat bronchial asthma and cerebrovascular disorders. Several reports have shown that ibudilast decreases the production of tumor necrosis factor-alpha, IL-1-beta, IL-6, and interferon-gamma. It also suppresses the differentiation of T helper 1 cells. However, it remains unknown whether ibudilast suppresses Th17 differentiation and IL-17 production.

Methods: For the in vitro study, naïve T cells were isolated from 5 healthy volunteers using a magnetic bead kit. These cells were stimulated for Th17 differentiation, with/without ibudilast for 1 week. Population of Th17 was measured by flow cytometry using anti-IL-17 antibodies. For the in vivo study, mRNA were isolated from 5 patients with a history of ischemic stroke before and after ibudilast consumption for 6 weeks. We compared the IL-17 mRNA before ibudilast consumption with those after ibudilast consumption. This study was approved by institutional ethics board.

Results: In vitro, Th17 differentiation was significantly inhibited by ibudilast. In vivo, the IL-17 mRNA were significantly lower in patients treated with ibudilast than in patients not treated with ibudilast.

Conclusion: Ibudilast inhibits Th17 differentiation and may regulate harmful secondary inflammation induced by ischemic changes in the brain.

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Abstract – WCN 2013

No: 970

Topic: 3 – Stroke

“Silent” strokes in hydrocephalus

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Background: According to the data of WHO, cardio-vascular diseases (CVD) are nowadays one of the major problems of the population and tend to be growing. Therefore, timely diagnostics of impairments, caused by CVD has a great importance for treatment and predicting complication leading to increased patient morbidity and mortality.

Objective: In this article, we report cases of right hemisphere strokes that had atypical or rare symptomatology and did not get intensive care, due to the absence of severe focal symptoms and some ambiguity in anamnesis. Comparative analysis was performed for detachment of the similarities in patients. We also attempted to form some theory of pathogenesis of such “silent” strokes and performed some tips for practitioners.

Patients and methods: Three cases of patients (two males, aged 45 and 73, and female, aged 53) with clinically silent stroke were investigated. Patients got treatment in neurological department and underwent standard clinical analyses, blood coagulation tests, lipoprotein phenotyping tests, neuroimaging scannings and MMSE-2 test. All data were standardized and compared to normal test results and to each other.

Results: All patients had MR-signs of hydrocephalus, cortex degeneration and had some insignificant deviations in analyses, pointing on the predisposition to atherosclerosis. All patients had high MMSE-2 scores. Males also had burdened labor anamnesis.

Conclusion: We pointed a correlation between the presence of the hydrocephalus and inconsistency of the severity of symptoms in relation to neuroimaging data. We also suggested theory of pathogenesis of such discrepancies.

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Abstract – WCN 2013

No: 1116

Topic: 3 – Stroke

The temporal evolutions of Lac, Cr and NAA concentrations in whole brain hyperacute ischemia rat model detected quantitatively by HPLC

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Objective: To explore the dynamic changes of brain metabolites in hyperacute ischemia brain injury of rat model.

Method: 96 rats were randomly divided into 12 groups. Each group had 8 rats and all rats were sacrificed by cutting the head. Concerning the

time course, the rat head was dropped into liquid nitrogen at time points of 0 s, 20 s, 40 s, 2 min, 3 min, 4 min, 5 min, 6 min, 10 min, 14 min and 20 min. The N-acetylaspartate (NAA), lactate (Lac) and creatine (Cr) concentrations in rat brain were measured by high performance liquid chromatography method.

Results: In the normal control group, the average concentration of Lac was $4.13 \pm 0.05 \mu\text{mol/g}$, N-acetylaspartate was $7.34 \pm 1.80 \mu\text{mol/g}$, Cr was $10.11 \pm 1.60 \mu\text{mol/g}$. In the ischemic groups, the average concentrations of Lac were significantly higher in each ischemia group than in the normal control group. While the ischemia time delayed from 0 s to 20 s, brain lactate concentration increased rapidly while NAA and Cr concentrations decreased. During 3 to 5 min, the lactate concentration kept on increasing, then maintained at a high level after 5 min. The concentration of NAA temporarily increased between 3 min to 14 min, then decreased progressively after 14 min. The Cr concentration fluctuated up and down from 1 min to 3 min, decreased significantly from 3 min to 4 min, then maintained at a low level after 4 min.

Conclusion: Lac, NAA and Cr concentrations dynamically change with different ischemic time in hyperacute ischemic brain injury.

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Abstract – WCN 2013

No: 1111

Topic: 3 – Stroke

Minimally invasive removal of intracerebral haemorrhage

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Objective: The main purpose of surgery of intracerebral hemorrhage is minimally invasive removal of hematoma with maximal volume evacuation. For the purpose of increasing the efficiency of surgery of intracerebral hemorrhage, we developed and applied an endoscopy assisted manipulator for minimally invasive evacuation of intracerebral hemorrhage and analyzed the results of treatment of patients using that technique.

Material and methods: The manipulator is consisted from rigid endoscope of Aesculap AG, including irrigation and suction channels, transparent sheath and its holder. The main advantages of the surgical evacuation of intracerebral hemorrhage using of the endoscopy assisted manipulator are:

- 1) Burr hole approach under local anesthesia.
- 2) The transparent sheath improves visualization of the border between brain parenchyma and hemorrhage.
- 3) Free-hand surgery without fixing the endoscope.
- 4) The capability of manipulation in deep area of the brain through narrow surgical approach.

We performed surgery using the manipulator in 78 patients with intracerebral hemorrhage during 24 h after onset. The range of hematoma volume was 18–106 cm³, the mean hematoma volume – $59.2 \pm 7.4 \text{ cm}^3$.

Results: The main duration time of surgery was $65.4 \pm 5.4 \text{ min}$, the mean hematoma removal rate was $95.2 \pm 2.6\%$. The lethal outcome rate was 10.8%. No postoperative recurrence of hemorrhage with deterioration of symptoms occurred.

Conclusion: We consider that evacuation of intracerebral hemorrhage using the endoscopy assisted manipulator allows improving the treatment outcome of patients with intracerebral hemorrhage.

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Abstract – WCN 2013

No: 1089

Topic: 3 – Stroke

Possibilities of neuroprotection during arterial hypertension and cerebro-vascular disorders

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Aim: Study of comparative effect of two variants of neuroprotection in patients with arterial hypertension (AH) and cerebrovascular disorders (CD).

Materials and methods: 60 I–III level AH patients of both sex were observed (age $55.3 \pm 2.8 \text{ years}$) with CD.

All patients received basic therapy (BT) (valsartan + ACK + indapamide + atorvastatin). By the character of neuroprotection patients were randomized into 2 groups: 1st (n = 30) received cerebrolysine of 10 ml i/v 10, 2nd (n = 30) – citicoline 1000 mg i/v N_o 10. Before, during and after 3 month therapy all patients' cardiological, neurological and psychological statuses were observed with the use of AH daily monitoring, different psychodiagnostic tests, electroencephalography, CT and MRT of brain, and Doppler of cerebral vessels.

Results: Analysis showed that groups were equally effective for dynamics of cardiological status and AH. But in group of BT + cerebrolysine there was a significant regress of overall (n = 24) and microneurosymptoms (n = 22), anxiety-depressive (n = 27) and cognitive (n = 28) disorders (P < 0.01). During BT + citicoline regressed overall (n = 29) and microneurosymptoms (n = 24) (P < 0.01), but less effective during psychological (n = 18) and cognitive (n = 24) disorders. Cerebrolysine was better for dynamics of neuro-physiological parameters, citicoline – of symptoms liquor hypertension (P < 0.01). This data verifies by variables the neurovisualization methods. Acute cerebrovascular events were not registered during neuroprotection in both groups (P < 0.05).

Conclusion: Cerebrolysine and citicoline demonstrated positive neuroprotective effect in patients with AH and CCVD. Thus, effect of cerebrolysine was significant during neuropsychological and cognitive disorders, and citicoline – local neurological symptoms and liquor hypertension. Combination of citicoline and cerebrolysine is increasing the possibility of neuroprotection during AH.

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Abstract – WCN 2013

No: 807

Topic: 3 – Stroke

Surgical therapy of internal carotid artery occlusion (ICAO) in nine patients

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Background: ICAO is a common clinical event. Although the incidence of stroke caused by the occlusion is not high, it leads to the serious change of the cerebral hemodynamics. In particular, when operation of contralateral carotid stenosis is needed, the risk is increasing.

Objective: To explore the indications for operation and recanalized techniques of ICAO.

Patients and method: DSA showed ICAO in 9 male patients, aged 57 to 77 years old. Among them, four patients showed recurrent TIA episodes, three showed dizziness with unilateral weakness, one showed unilateral blindness caused by paroxysmal amaurosis, and one showed paresis of left lower limb. Tail-like gradual occlusion occurred in 6 patients and neatly blunt occlusion in 3 patients. Distal carotid varying degrees of progress occurred in 6 patients.

Results: Six cases showed recanalization, while another three were not. After 2-year follow-up, seven cases did not show further cardiovascular and cerebrovascular events. Two patients died of heart disease.

Conclusion: Hemodynamic analysis is an important part for the operation. Tail-like gradual occlusion and forward or retrograde blood flow developing of the distal carotid artery should serve as strong evidence to choose operation. With regard to ICAO techniques, the length of the hard tough thrombosis exceeds the carotid incision by 3 cm, Fogarty catheter was applied to insert into the distal embolization, slowly retraced after filling the balloon, and repeated until blood flows out of the thrombosis. According to the intraoperative angiography, we can decide whether the stent is placed in the distal artery occlusion or not.

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Abstract – WCN 2013

No: 1143

Topic: 3 – Stroke

Intra-arterial mechanical procedures in treatment of acute stroke

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Some of the methods of treatment for acute stroke after therapeutic window for intravenous thrombolysis (3–4.5 h) are neuro-interventional procedures: intra-arterial thrombolysis and endovascular potentiating of thrombolytic effect.

Case report: Patient (64), admitted at 13 h, because of stroke, with severe right-sided limbs pyramidal deficit, motor dysphasia (NIHSS 14). First symptoms have appeared abruptly at 7 h.

The patient underwent digital subtraction angiography (DSA) due to MSCT malfunction. The DSA finding 7 h after onset was occlusion of temporal branch of left MCA.

Following DSA, aspiration thrombectomy was performed, using aspiration catheter size 4Fr, with thromb aspiration with negative pressure using syringe of 60 ml, and procedure is repeated until complete thromb extraction is achieved. Control DSA showed complete recanalization. Patient showed signs of fast recovery, initially in lower limb, immediately followed by recovery of upper limb, maintaining motor dysphasia (NIHSS 5).

MR endocranium on the next day showed hyper density in zone of left MCA, 1 × 1.5 cm.

Discussion: Based on experience in MMA in treatment with intra-arterial thrombolysis and aspiration techniques, of 5 treated patients, full recovery has been achieved in 4, while one resulted in death. It is a fact that patients with time expired on the therapeutic window for intravenous fibrinolysis, but immediately treated with intra-arterial fibrinolysis or mechanical thromb extraction as possible therapeutic option, have much better prognosis.

Conclusion: Blood vessel recanalization in presented patient is achieved by effect of mechanical thromb extraction and it does not represent standard therapeutic procedure. Presented data indicate possibility of widening the therapeutic window.

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Abstract – WCN 2013

No: 1135

Topic: 3 – Stroke

Low prevalence etiology in a patient with hemispheric stroke

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Objectives: Atherosclerotic intracranial arterial stenosis is an important cause of stroke and it is increasingly being treated with percutaneous transluminal angioplasty and stenting to prevent recurrent stroke. In patients with intracranial arterial stenosis, aggressive medical management was superior to percutaneous transluminal angioplasty and stenting with the use of the Wingspan stent system, but there are no studies that compare transluminal balloon angioplasty to medical treatment.

Methods: We present the case of a 76-year-old patient who presented to our clinic with recurrent syncope for neurologic assessment. The patient has a history of permanent atrial fibrillation, central retinal artery occlusion, and hypertension and has no history of prior neurological deficits. The ultrasonographic examination of cervical vessels was unremarkable for atherosclerotic disease but the CT examination of the head revealed a stroke in the cortical border zone between MCA and PCA. The cervico-cerebral artery digital subtraction angiography revealed an intracranial stenosis of middle cerebral artery in M1 segment.

Results: The patient underwent percutaneous transluminal balloon angioplasty of the stenosis of MCA in M1 segment without neurologic incidents or haemodynamic instability and was discharged with Clopidogrel and oral anticoagulant-cumarin derivate.

Conclusion: We choose to perform percutaneous transluminal balloon angioplasty in a selected case of a patient with middle cerebral artery stenosis and multiple risk factors for stroke with good results and without complications.

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Abstract – WCN 2013

No: 1200

Topic: 3 – Stroke

Clinico-immunological assessment of carotid atherosclerosis

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Background: Inflammation of the vessel wall is one of the pathogenic mechanisms of initiation and progression of atherosclerotic lesions, that are the most common cause of ischemic stroke.

Objective: A comparison of the level of interleukin-6 (IL-6) and C-reactive protein (CRP) in blood serum in patients with different clinical manifestations of atherosclerosis of carotid arteries.

Materials and methods: Atherosclerotic lesion of carotid arteries was detected by duplex scanning in 61 patients. 23 of them had clinical manifestations of carotid atherosclerosis (stroke or transient ischemic attack) in history. The level of IL-6 and CRP in serum was determined in 61 patients with carotid artery disease by immunosorbent assay.

Results: Patients with stroke in history had the increased concentration of IL-6 (8.0; 4.0–14.0 pg/ml), the level of IL-6 in patients with asymptomatic carotid atherosclerosis was lower (4.6; 2.1–7.7 pg/ml, $p = 0.042$). Concentration of CRP was higher (6.8; 4.9–14.2 mg/l) in patients with clinical manifestations of atherosclerosis of precerebral arteries, than in patients with asymptomatic atherosclerosis (2.1; 1.3–6.3 mg/l, $p = 0.002$). The level of CRP in patients with atherothrombotic

stroke in history was higher (9.3; 5.2–19.2 mg l) than in the other patients (2.8, 1.8–6.9 mg l, $p = 0.011$).

Conclusion: The increased concentration of IL-6 and CRP in serum is associated with complications of carotid atherosclerosis. CRP is associated with stroke, formed by the mechanism of atherothrombosis.

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Abstract – WCN 2013

No: 1222

Topic: 3 – Stroke

Stroke in Kenya and its co-morbidities and risk factors in Kenya hospitals

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Background: Stroke is the most common causes of morbidity and mortality the world over. Established risk factors such as arterial hypertension, diabetes mellitus, cigarette smoking, hyper-lipidaemia, micro-vascular rupture, gender, age/co-morbidities like sickle cell disease, HIV/AIDS infection and cerebral malaria are encountered in the tropics.

Objectives: To determine pathological, risk factors, in-hospital period prevalence and in-hospital outcome of stroke.

Results: A total of 2629 patients were admitted at the hospital during study period. 80 patients had diagnosis of stroke; giving an in-hospital period prevalence of 3042/100,000. Mean age was 61.3 years, mode; 63 years, range 34–95 years. M to F ratio 1.2:1 stroke sub-types: ischaemic stroke 68 (85%) and haemorrhagic stroke seven (8.8%). Five patients (6.3%) no evidence of stroke sub-type was on-record. Established risk factors are hypertension/diabetes mellitus. Hypertension was found in 64 patients (80%) and diabetes-mellitus in 27 (33.7%). 23 patients (28.8%) had both hypertension/diabetes-mellitus. Co-morbidities were observed and included mitral-stenosis, cardiac-arrhythmias cardio-myopathy, HIV/AIDS, Left Ventricular Hypertrophy (LVH), infective endocarditis, atrial septal aneurysm, carotid plaques with or without stenosis and hyper-homocystenemia. 75 patients (93.8%) were discharged and four (5%) died in hospital. All patients who died had anterior circulation ischaemic stroke as per Trial of Org 10172 in Acute Stroke Treatment (TOAST) classification.

Conclusion: Ischaemic stroke is the most common pathological sub-type observed in this study. Hypertension is the leading observed risk factor for stroke. Hospital period prevalence for stroke of 3042/100,000 was found. 75 patients (93.8%) were discharged and four (5%) died in hospital.

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Abstract – WCN 2013

No: 460

Topic: 3 – Stroke

Anterograde amnesia due to fornix infarction

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Background: The fornix serves an important role in human memory. Amnesia resulting from fornix infarction has been rarely reported.

Objective: To describe the clinical and radiological features of a patient who developed anterograde amnesia following fornix infarction and review the published literature.

Patients and methods: A patient who was admitted to the Department of Neurology, National Neuroscience Institute, Singapore.

Results: A 39-year-old lady with poorly controlled diabetes mellitus and dyslipidaemia presented with acute confusion and anterograde amnesia. Neurological examination was normal except for MMSE score of 27 upon 30, with deficits in orientation and recall. MRI brain showed an acute infarction of the columns of the fornices bilaterally. The right septal nuclei and anterior commissure were also involved. CSF studies, trans-thoracic echocardiogram, ultrasound of the carotid arteries and thrombophilia workup were unremarkable. She was started on clopidogrel with optimisation of her cardiovascular risk factors. 2 months after admission, she reported persistent episodic memory loss and cognitive tests showed residual impairment in delayed recall. MMSE score was 28 upon 30 (minus 2 points for recall) and MOCA score was 26 upon 30 (minus 4 points for recall). MRI brain done 4 months after admission showed a small focal area of high T2, low T1 signal and volume loss at the inferior aspect of the column of the right fornix, consistent with previous infarction.

Conclusion: This rare case illustrates persistent memory impairment following fornix infarction and emphasises the importance of excluding a strategic infarct in a patient presenting with acute amnesia.

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Abstract – WCN 2013

No: 1248

Topic: 3 – Stroke

A national survey to establish current methods of venous thromboembolism prophylaxis in stroke patients practiced by Polish neurologists

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Background: Venous thromboembolism (VTE) is associated with considerable morbidity and mortality in stroke patients.

Objective: The purpose of our survey was to establish the methods of VTE prophylaxis currently practiced by Polish neurologists. We also aimed to determine whether there is enough variation in practice to justify the development of evidence based upon detailed Polish guidelines for VTE prevention.

Methods: Postal self-administered questionnaires about VTE prophylactic methods practiced by Polish neurologists were sent to 218 (all) neurological wards where stroke is treated. If there was no response the questionnaire was faxed and finally we attempted to obtain information by telephone.

Results: Data from 176 (80.7%) of 218 centers was finally collected. Majority 137/176 (77.8%) had a stroke unit. The median admission rate of surveyed wards was estimated to be 320 patients/year (range 20 to 1000 patients/year). The most common method VTE prophylaxis reported was low molecular weight heparins (LMWH) (98.9%), the least common was intermittent pneumatic compression (IPC) (6.8%). Large centers (admitting ≥ 200 patients/year) (70.7% vs 45%; $p = 0.039$), and those with stroke units (95 vs. 20; $p = 0.031$) were more willing to join a randomized trial evaluating any of the methods. All the centers participating in the survey reported a need for detailed Polish VTE prophylaxis guidelines.

Conclusions: LMWH is the predominant VTE prophylaxis strategy for stroke patients practiced by Polish neurologists. Due to the variation of methods practiced in VTE prophylaxis it might be

reasonable to further specify this section of stroke treatment guidelines.

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Abstract – WCN 2013

No: 1260

Topic: 3 – Stroke

Neuroradiological reporting from abroad – Is it possible?

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Objective: To evaluate whether reporting of emergency examinations is possible over a distance of 12,000 km.

Methods: Evaluation of time duration for the sending process for MR and CT scans under emergency conditions.

Results: 3472 MRI and 1118 CT reports have been prepared between 01.07.2010 to 30.06.2012, median time for sending process for CT was 47.1 s and for MR 252.4 s.

Conclusion: In case of broadband cable capacity and dedicated IT-network specialized reporting of neuroradiological cross sectional imaging examinations is feasible in acceptable length of time even over long distance and in an emergency setting. In any case the requirements of Germany for teleradiological projects are fulfilled. This allows neurological treatment even of intensive care patients according to quality level of European centers.

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Abstract – WCN 2013

No: 1139

Topic: 3 – Stroke

Myocardial Infarction as an early complication in acute stroke patients: Results from the Austrian Stroke Unit Registry

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Background: Patients with transient ischemic attack (TIA) and stroke have an increased risk for subsequent cardiac events including myocardial infarction (MI). Information regarding MI as a complication of stroke in the very acute phase is limited.

Objective: To assess the frequency of MI, the characteristics and short-term outcome of such patients in the stroke unit setting.

Methods: 41,619 patients with ischemic stroke or TIA, and 4984 with primary intracerebral haemorrhage (ICH) prospectively enrolled in the Austrian Stroke Unit Registry in the last 6 years were analysed.

Results: 421 patients with ischemic stroke or TIA (1%) and 17 patients with ICH (0.3%) suffered from MI during stroke unit treatment. Patients with ischemic cerebrovascular events were significantly older, had more severe strokes and generally more vascular risk factors, atrial fibrillation and previous MI. Total anterior circulation and left hemispheric stroke syndromes were more often observed in MI patients. Patients with MI not only suffered from a worse short-term outcome at stroke unit discharge (mortality 14.5 versus 2%, $p < 0.001$), but also acquired more other well known stroke complications. In

multivariate analyses factors that remained independently associated with MI were previous MI and stroke severity at admission. Hypercholesterolemia was inversely associated with MI.

Conclusions: Although MI in the acute phase after stroke seems to be rare, it is associated with a poor short-term outcome. Furthermore our results identify patients with previous MI and more severe strokes to be at an increased risk for the early complication of MI.

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Abstract – WCN 2013

No: 1244

Topic: 3 – Stroke

The cultural acceptance of robotic tele-stroke medicine among patients and healthcare providers in Saudi Arabia: Results of a pilot study

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Background: The technical feasibility and cultural acceptance toward the use of telestroke technology in Saudi Arabia has not been investigated.

Objectives: To determine the degree of satisfaction toward the use of this technology among stroke patients, their relatives and healthcare providers in Saudi Arabia.

Materials and methods: The remote presence robot (RPR), the RP-7i® (FDA-cleared) provided by InTouch Health was used between October and December, 2012 at King Abdulaziz Medical City in Riyadh, Saudi Arabia. Patients and their relatives were informed that the physician would appear through a screen on the top of a robotic device, as part of their clinical care. A convenient sample of consecutive stroke patients (and relatives) admitted to the stroke unit through ED and healthcare providers completed a self-administered satisfaction questionnaire following consultation sessions.

Results: A total of fifty participants completed the questionnaire. All agreed or strongly agreed that the consultant interview through robotic tele-stroke was useful and that the audiovisual component of the intervention was of high quality; 98% agreed or strongly agreed that they did not feel shy or embarrassed during the remote interview, were able to understand the instruction of the remote consultant and recommended its use in stroke management. In addition, 92% agreed or strongly agreed that the use of this technology can efficiently replace the physical presence of a neurologist.

Conclusion: Results suggest that the use of tele-stroke medicine is culturally acceptable among stroke patients and their families in Saudi Arabia and is favorably received by healthcare providers.

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Abstract – WCN 2013

No: 1271

Topic: 3 – Stroke

Retinal vasoreactivity in patients with ischemic stroke

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Background: As the eye and brain share embryological, anatomic and physiological similarities, studies of retinal blood vessels may

prove useful as a surrogate marker for cerebrovascular disease and help to assess stroke risk. Retinal arteriovenous ratios (AVRs) have been shown to indicate risk of stroke, but the association between retinal vasoreactivity, ischemic stroke, and cerebrovascular function remains unknown. Retinal vasoreactivity assessments may detect early stages of endothelial dysfunction and allow direct assessment of blood vessel physiology.

Objective: To examine

- 1) retinal microvessel behavior in patients with ischemic stroke
- 2) the relationship between retinal and cerebral vascular reactivity.

Methods: Cohort study of 12 patients with ischemic stroke presenting 3+ months after the stroke, and 8 healthy controls (44–85 years). Retinal vasoreactivity was measured with the dynamic vessel analyzer following flicker light stimulation. Middle cerebral artery (MCA) vasoreactivity following hyperventilation/breathhold was measured using transcranial Doppler ultrasound. AVRs were obtained using funduscopic photographs.

Results: Patients with stroke had significantly attenuated retinal venous ($p = <0.0002$, CI 95%) and arterial ($p = 0.001$, CI 95%) vasodilation responses compared to healthy controls, and decreased cerebral vasoreactivity following hyperventilation/breath hold ($p = 0.0068$, CI 95%). Across all groups an attenuated venous flicker response was associated with an increase in MCA RI ($r = -0.5$, $p = 0.03$, CI 95%) and PI ($r = -0.5$, $p = 0.04$, CI 95%) and a decrease of MCA vasoreactivity ($r = 0.46$, $p = 0.05$, CI 95%).

Conclusion: In this study impairment of retinal microvascular function is associated with ischemic stroke and measures of cerebrovascular function. Microvascular dysfunction of retinal blood vessels may predict ischemic stroke risk.

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Abstract – WCN 2013

No: 1072

Topic: 3 – Stroke

“Mobile Stroke Unit” for stroke treatment at the emergency site

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Background: Currently only 2–5% of all acute stroke patients receive thrombolytic therapy due to delay in hospital arrival or diagnostic work-up. In this feasibility study, we tested the efficacy of a new approach of stroke diagnosis and treatment starting at the emergency site, rather than after hospital arrival, in reducing delay in stroke therapy.

Methods: We constructed a “Mobile Stroke Unit”, an ambulance that delivers imaging, point-of-care-laboratory analysis, and neurological expertise directly at the emergency site. In a prospective, randomised single-centre trial, we compared the time from alarm (emergency call) to therapy decision, between Mobile Stroke Unit (MSU) and standard stroke hospital intervention.

Results: As prespecified the trial was stopped after a planned interim analysis at 100 of 200 patients because the primary endpoint showed a clear superiority of the MSU treated group.

Prehospital stroke treatment reduced the median time from alarm to therapy decision substantially to 35 min (IQR 31–39) in the MSU group versus 76 min (63–94) in the hospital control group ($p < 0.0001$). We also detected similar gains regarding times from alarm to end of the different diagnostic steps, although there was no substantial difference in the number of patients who received intravenous thrombolysis or in neurological outcome. Safety endpoints seemed similar across the groups.

Conclusions: This first trial demonstrates feasibility of prehospital diagnostic stroke work-up and treatment, with substantially reduced time until therapy decision.

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Abstract – WCN 2013

No: 1283

Topic: 3 – Stroke

Ipidacrine for post-stroke cognitive impairment

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Objective: To determine cognitive status and to assess efficacy of ipidacrine in post-stroke patients.

Methods: Ipidacrine (neiomidin) is a nonselective inhibitor of acetylcholinesterase and butyrylcholinesterase. The drug was prescribed to 43 patients (aged 33–75, 26 males, 17 females) one month after the first hemispheric ischemic stroke. Patients received ipidacrine in increasing dosages from 10 to 60 mg during 2 months. Clinical and neuropsychological investigation with the use of NIHSS, Barthel index (BI), MMSE, FAB, attention test (AT), semantic verbal fluency test (SVFT), phonetic verbal fluency test (PVFT) was carried out twice: at the baseline and 2 months after. Control group consisted of 35 persons without stroke.

Results: The data differences in patients and control group were fixed in MMSE (26.51 ± 3.06 and 28.71 ± 1.07 , $p = 0.0001$), FAB (12.74 ± 2.97 and 16.40 ± 1.74 , $p = 0.0000$), SVFT (15.79 ± 5.32 and 21.03 ± 5.79 , $p = 0.0002$), PVFT (7.16 ± 3.60 and 12.76 ± 4.52 , $p = 0.0000$), and AT (99.53 ± 42.06 and 64.80 ± 18.95 s, $p = 0.0000$) one month after stroke. A statistically significant improvement of NIHSS (4.42 ± 2.69 and 2.70 ± 2.01 , $p = 0.0001$) and BI (95.35 ± 10.60 and 98.84 ± 4.06 , $p = 0.0074$) was observed during treatment. Neuropsychological reinvestigation demonstrated that MMSE (28.16 ± 2.33 , $p = 0.0000$), FAB (15.05 ± 2.61 , $p = 0.0000$), SVFT (19.40 ± 5.76 , $p = 0.0000$), PVFT (8.35 ± 3.62 , $p = 0.0063$) and AT (83.79 ± 30.11 , $p = 0.0017$) significantly increased and differences with control group in MMSE and SVFT disappeared.

Conclusion: Post-stroke cognitive impairment reflecting decrease of frontal functions, verbal fluency and attention was identified. A statistically significant regress of neurological and cognitive deficiency was revealed during ipidacrine treatment. The distinct positive dynamics of executive function, attention, verbal fluency and general mental activity were observed. The drug was well tolerable.

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Abstract – WCN 2013

No: 1291

Topic: 3 – Stroke

How to use verbal autopsy to study stroke incidence in developing countries

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It is well recognized that good quality population-based studies are the most reliable source of information about stroke incidence on a population level but identifying all new stroke in a population is particularly challenging. However, those criteria may be not practical for stroke studies undertaken in developing countries, where most strokes occur and resources are limited.

In a door-to-door survey in Rabat–Casablanca regions, in Morocco, involving a total of 60,031 individuals (36,756 urban and 3275 rural), investigators identify through the use a specific questionnaire a possible stroke patient and also households where one person died of a possible stroke. In a second step neurologists confirmed the stroke in survivors and stroke death by using a specific verbal autopsy.

We identified 51 first-ever stroke during one year. The incidence rate standardized to Segi's world population was 97.9 per 100,000 (95% CI 73–123) and the incidence rate in people aged over 55 years was 550.6 per 100,000 (95% CI 381–721) with 467.4 (95% CI 269–666) in urban and 691.4 (95% CI 381–1002) in rural.

Our results show that incidence rate of stroke is significantly high in people aged over 55 years specially in rural area, which are similar to those reported in community studies both in developed countries (Feigin et al., 2003) or in emerging countries (Lavados et al., 2005). We believe that the use of specific verbal autopsy in door-to-door survey of stroke provides good approximation of stroke incidence in developing countries.

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Abstract – WCN 2013

No: 1137

Topic: 3 – Stroke

Serial hemodynamic and autonomic function studies in acute ischemic stroke: Relation to outcome, stroke type and location

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Background: Hypertension and autonomic changes commonly accompany acute ischemic stroke (AIS); however, their pathophysiology is poorly understood.

Aim: To study serial hemodynamic and cardiovascular autonomic functions among patients with AIS and to explore their relation to stroke outcome, location, type and severity.

Patients and methods: Patients with AIS of <96 h duration underwent serial hemodynamic and cardiovascular autonomic function studies using thoracic electrical bioimpedance technology over the first 4 days of admission. Patients with arrhythmias or pulmonary edema were excluded. Correlations between NIHSS scores, outcome at follow-up (mean 10 months), stroke type, location and hemodynamic/autonomic parameters were explored using Mann–Whitney U-test and logistic regression analysis.

Results: 55 patients (M:F:27:28; age:62 ± 12 years) with AIS were studied. 33 had large artery stroke, 18–lacunar and 4–cardioembolic. 6 patients (11%) died. Mean BP decreased by 8 mm Hg by day 4 (D4); while cardiac output remained stable, systemic vascular resistance (SVR) changes reciprocated BP. Lower NIHSS at admission, SVR (D2), systolic BP (D4), and cardiac index (CI, D1) correlated significantly ($p < 0.01$) with good outcome, as did following autonomic indices: sympathetic activity (LFNU; LFNUBPV); parasympathetic activity (HFNU); sympathovagal balance (Lf/HF); baroreflex sensitivity (BRS) and ventricular work (LVWI). Systolic BP (D4) predicted outcome independently. Stroke location in the basal-ganglia was related to left ventricular ejection time and sympathovagal balance (Lf/HF) ($p < 0.05$).

Conclusion: Hypertension in AIS may be mediated predominantly by changes in SVR. Hemodynamic and autonomic parameters may predict outcome and possibly stroke location. Further studies among patients with homogenous stroke types may help understand the interaction between focal brain injury and autonomic changes.

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Abstract – WCN 2013

No: 1336

Topic: 3 – Stroke

Thrombolysis in acute ischaemic stroke: Initial experience in a general hospital in a developing country

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Background: The aim of the study was to present our initial experience with the use of intravenous thrombolytic therapy in patients with acute ischaemic stroke. We present results related to demography, time, and data on the clinical outcome of patients and compare the results with other studies (Serbian Experience with Thrombolysis in Ischemic Stroke (SETIS) and Safe Implementation of Treatments in Stroke (SITS)).

Objective: Define risk factors in our community, characteristics of clinical course and outcome in patient with acute ischemic stroke proven neuroradiologically (CT) after IV application rt-PA (0.9 mg/kg).

Patients and methods: Study involved 15 patients treated with rt-PA in our hospital between the years 2011–2013.

Results: The research included 15 patients (F:M = 4:11), median age 65 (53–77). The most frequent risk factors were: hypertension 86.6%, atrial fibrillation (AF) 40%, diabetes mellitus 33.3%, hyperlipidemia 53.3%, and smoking 26.6%, 6.6% had no risk factors. Ischemic lesion proven CT in 8 patients was on left side hemisphere 8/15 (53.3%), right side hemisphere 5/15 (33.3%), cerebellum 1/15 (6.6%) and brainstem 1/15 (6.6%). Due the hospitalization the improvements were registered in 11/15 (73.3%), unchanged at 3/15 (20%) and one lethal result. Initial NIHSS score was 8.45 (4–17), modified Rankin scale (mRS) after 3 month 0–2 (1.53), and onset-to-needle time was 168.5 (125–250) min.

Conclusion: Of course, we are aware that a good outcome (above the average monitoring studies) is the result of an engaged approach. We want to show the results in order to encourage colleagues working in hospitals of similar size and equipment in countries with similar social, demographic and economic characteristics.

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Abstract – WCN 2013

No: 1312

Topic: 3 – Stroke

Systemic allostatic overload in acute stroke

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The aim of our study is to characterise cerebrovascular patients concerning cardiovascular functions.

Haematological analyses were performed within 24–48 h on 476 Acute Stroke (AS), 1142 Chronic Cerebro-Vascular Disease (CCVD) and 216 Other Neurological Disease (OND) patients, among which 161 AS, 504 CCVD, 83 OND underwent echocardiography.

Troponin Ths (TnT_{hs}, pg/ml) and Pro Brain Natriuretic Peptide (PBNP, pg/ml) were higher in AS (74.61 sd 217.88, $p = 0.007$; 3809.68 sd 6798.65, $p = 0.0004$) and CCVD (30.15 sd 49.29, $p = 0.0001$; 1608.78 sd 3982.99, $p = 0.0007$), compared to OND (9.66 sd 7.63; 100.76 sd 145.86). A tendency to lower PBNP was found in AS without CCVD (A) compared to AS with CCVD (B) patients.

Echocardiography showed decreased ejection fraction (EF %) and increased pulmonary arterial pressure (PAP, mmHg) in AS (EF 59.52 sd 9.96, $p = 0.0001$; PAP 43.85 sd 11.47, $p = 0.0001$) and CCVD (EF 60.02 sd 10.55, $p = 0.0001$; PAP 40.53 sd 10.89, $p = 0.006$), compared to OND (EF 64.96 sd 9.66; PAP 34.32 sd 9.45). Significant differences were observed concerning such parameters and left atrial dilatation (LAD, cm) in subgroup B (LAD 4.63 sd 0.72 $p = 0.0003$; EF 58.52 sd 9.97, $p = 0.01$; PAP 44.58 sd 11.59, $p = 0.05$) compared to A (LAD 4 sd 0.41; EF 64.4 sd 8.65; PAP 37.2 sd 6.11). Significant correlations were found between haematological and echocardiographic parameters.

Our data highlight the key role of early identification of cerebrovascular patients with a wider due to restricted therapeutical window because of cardiovascular dysfunctions.

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Abstract – WCN 2013

No: 1350

Topic: 3 – Stroke

Spinal cord infarction presenting as unilateral diaphragm paralysis

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Background: Cervical cord ischemia leads to tetraplegia and pulmonary insufficiency in the case of transverse extension of ischemia. Paralysis of the diaphragm can be experimentally modeled by lateralized, high cervical lesions that interrupt descending inspiratory drive to the corresponding phrenic nucleus. Most reported cases of unilateral diaphragm paralysis were caused by trauma. Paralysis of unilateral diaphragm by spinal cord infarction is rare.

Case report: A 79-year-old woman with a history of hypertension visited our hospital with a 2-day history of suddenly developed quadriparesis and dyspnea. On admission her vital signs were normal except for dyspnea. Neurologic examination revealed normal mental state and cranial nerves. Motor evaluation showed weakness of both upper and lower extremities, upper extremities more than lower extremities, right more than left. Sensory evaluation revealed a sensory level at C4. The patient showed no pathologic reflex and deep tendon reflex was hypoactive in all extremities. We ordered cervical spine MRI. It showed an elongated hyperintensity on T2WI at the ventral portion of cervical spinal cord from C2 to C6 level without enhancement. The patient was treated with anticoagulation and corticosteroid. Over the next two days, we checked chest fluoroscopy and it showed upward displacement of the right diaphragm with relatively fixation of movement suggestive the right diaphragm paralysis. 2 weeks later we checked phrenic nerve conduction study (MacLean's method). It showed prolonged latency and lowered amplitude of compound muscle action potentials (CMAPs) of the right phrenic nerve. 2 month later, she discharged with only mild right hemiparesis without dyspnea.

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Abstract – WCN 2013

No: 1389

Topic: 3 – Stroke

External carotid artery angioplasty and stenting – A good choice for patients with symptomatic ipsilateral internal carotid artery occlusion

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Background: In patients with internal carotid artery occlusion (ICA), the ipsilateral external carotid artery (ECA) may supply the cerebral circulation through collateral vessels. Although endarterectomy for ECA stenosis has been described in literature, angioplasty with stent-graft has rarely been reported.

Objectives: The aim of our small case series is to present few cases with ECA stenosis and ipsilateral ICA occlusion in which stenting of ECA was performed.

Patients and methods: We present four patients with uni- or bilateral ICA occlusion (one of the cases with occlusion secondary to thrombosis after endarterectomy), aged 48 to 69 years old, who presented to our service for neurologic symptoms suggestive for an ischemic event, confirmed by cerebral CT scan or MRI. The ultrasonographic exam of cervico-cerebral arteries showed the presence of ICA occlusion with ipsilateral hemodynamic significant ECA stenosis, so we performed an angiogram which confirmed these findings. The digital subtraction angiography also revealed that the cerebral blood flow was supplied from the ECA collaterals. All the ECA stenoses were ipsilateral to the ischemic stroke or transient ischemic attack.

Results: We proceeded to angioplasty with stenting of the ECA with optimal postangioplasty results and improved intracranial cerebral blood flow, without periprocedural incidents. The patients were discharged few days later with double antiplatelet therapy.

Conclusions: We chose to present these cases to underline the importance of preventing further ischemic events in patients with occluded ICA and ipsilateral ECA stenoses using angioplasty and stenting of ECA.

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Abstract – WCN 2013

No: 1392

Topic: 3 – Stroke

Ability reading comprehension in patients with speech and language impairments after stroke

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Background: Impairments of speaking caused by stroke decreases ability to express and comprehend speaking, reading and writing. Dyslexia is loss or impairment of ability to read brought on by brain damage, in part with the aphasia syndrome or other cognitive neurological impairments.

Objective: The aim is to determine age, sex, education level, stroke etiology comorbidity, relation between locations of brain impairment after the stroke with reading comprehension of sentences and paragraphs in patients with impaired speaking communication.

Patients and methods: During six months of speaking, 135 patients from neurological ward of the Institute of Rehabilitation "Dr Miroslav Zotović" in Banja Luka have been analyzed. Subtests of Boston Diagnostic Aphasia Examination were used.

Results: The average age of the patients was 66.77 years in samples where there were more males than females. Brain lesions were

distributed on 35 different locations. The most numerous were multifocal lesions, than left sided lesions of parietal, frontoparietal, temporal, parietal temporal, frontoparieto temporal and other brain regions. Low results were found on word–picture matching (average 45.40% of tests) and comprehension of sentences and paragraphs (average 27.55% of tests). Understanding of read materials was lower at left sided compared to right sided brain impairments. There is a high correlation between comprehension of read materials with auditory understanding of speech and language like the ability of nomination tested patients.

Conclusion: Reading comprehension of patients with speech and language impairments after stroke on examined samples is low. The degree of these functions depends on location and size of brain damage and other factors.

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Abstract – WCN 2013

No: 1393

Topic: 3 – Stroke

Frequency of writing and reading disorders in Bosnia and Herzegovina population of acute stroke patients

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Objective: Writing and reading disorders usually followed aphasias but the frequency of acalculia, agraphia and alexia are not often studied.

The aim of the study was to determine the frequency of alexia, agraphia and acalculia in acute stroke patients, and determine their relation to the type of stroke (ischemic and hemorrhagic) and lesion site (left or right hemisphere) in Bosnian and Herzegovinan population of acute stroke patients.

Patients and methods: It was analyzed in 194 consecutive hospitalized patients with acute first-ever stroke at the Department of Neurology, University Clinical Center Tuzla. For clinical assessment of alexia, agraphia and acalculia, Minnesota test for differential diagnosis of aphasia's was used. The patients were evaluated in the first week of stroke onset.

Results: Out of 194 analyzed patients, 59 (30.40%) of them had alexia, agraphia and acalculia or different combinations of these disorders. The frequency of alexia, agraphia and acalculia among patients with stroke in the left (dominant) hemisphere was significantly higher (33; 55.9%) than in the right one (23; 37.7%) ($\chi^2 = 4.003$; $p = 0.0459$). In 170 subjects with ischemic stroke, there were 53 (31.1%) cases of alexia, agraphia and acalculia, and 6 (42.8%) cases among 14 subjects with hemorrhagic stroke then hemorrhagic and ischemic stroke ($\chi^2 = 0.363$; $p = 0.394$).

Conclusion: Post-stroke reading and writing disorders are present in one third of stroke cases, and more common in patients with stroke in the left hemisphere, than in the right. There were no significant differences in the frequency of alexia, agraphia and acalculia between hemorrhagic stroke and ischemic stroke.

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Abstract – WCN 2013

No: 1398

Topic: 3 – Stroke

Ischemic stroke subtype and clinical characteristics of patients with locked-in syndrome

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Background: Locked-in syndrome (LIS) is a state characterized by paralysis of all extremities and the facial musculature, but preserved consciousness and cognition. The most frequent cause is a bilateral ischemic lesion of ventral brainstem.

Objective: To determine the clinical characteristics, neurovascular findings, and ischemic stroke subtyping based on the TOAST classification system in case series of patients with ischemic LIS.

Patients and methods: Cases with LIS were identified from a prospectively acquired database of patients with acute ischemic stroke admitted or transferred to specialized hospital for cerebrovascular diseases over 3-year period (between 2009 and 2011).

Results: Our case series consists of 19 patients (mean age 62, 2 ± 10 years; range 46–82). Periodic episodes of rotatory vertigo or dizziness, double vision, and slurred speech were the most prevalent premonitory neurological symptoms. The mean time from symptom onset to clinical diagnosis of LIS was 3 days (range 1–15). In the majority of subjects the most extensive injury was localized in the middle part of the basis pontis. Based on the TOAST criteria seventeen cases were classified as large artery occlusive disease (90%), and the rest as a cardioembolic stroke subtype of undetermined etiology. The sites of most extensive atherosclerosis were the intracranial segment of vertebral arteries, and the proximal half of the basilar artery.

Conclusions: Ischemic LIS is commonly caused by large artery disease in the proximal intracranial vertebrobasilar territory. Transitory dysfunction of vestibulocerebellar or motor/oculomotor brainstem's structures was the most often reported premonitory symptoms.

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Abstract – WCN 2013

No: 1400

Topic: 3 – Stroke

The clinical syndromes after acute isolated thalamic stroke

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The clinical syndromes and specific symptoms occurring after the acute thalamic stroke on different anatomic and vascular territories are poorly highlighted in the literature.

Objective: To describe the clinical syndromes and the specific symptoms in patients with the isolated thalamic stroke.

Methods: Complex assessment of neurological cognitive status of 22 patients (12 men, 10 women, average age – 61.9 + – 10.2 year old) with confirmed MRI diagnosis of thalamic stroke on different vascular territories has been performed.

Results: Thalamic stroke was more frequently localized in the classic territories – paramedial (27.3%) and lower lateral (49.9%) ones; less frequently in marginal vascular zones – lateral (22.7%), and central (9.1%).

Stroke clinical syndromes of various territories differed: the unilateral stroke of paramedial zone was manifested by posteromedial syndrome – suppressed consciousness, cognitive and psychosensory disorders, up-gaze paresis; the bilateral one – by the thalamic pyramidal stroke syndrome; the syndrome of central territory impairment was accompanied by faints, cognitive disorders, contralateral hemihypesthesia, hemiataxia, hemianopsia, central mimetic paralysis during the emotional reactions; pure full sensor syndrome or in unusual combinations and moderate cognitive disorders are typical manifestations of lacune stroke of the lateral territory; and stroke of the lower lateral territory was manifested by the thalamic syndrome of Degerin–Russi – heterolateral hemihypesthesia, hemiataxia combined with painful feeling, autonomic disturbances with hyperpathia and cognitive deficiency.

Conclusion: The isolated thalamic strokes represent the heterogenic clinical group; they are manifested by different neurological syndromes

and specific symptoms that have to be taken into account during the therapy selection.

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Abstract – WCN 2013

No: 1177

Topic: 3 – Stroke

Cilostazol plus aspirin versus aspirin alone in patients with symptomatic intracranial arterial stenosis: Results of catharsis

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Background: The Cilostazol–Aspirin Against Recurrent Stroke with Intracranial Artery Stenosis (CATHARSIS) (Clinicaltrials.gov identifier: NCT00333164) was an investigator-driven nationwide multicenter cooperative randomized controlled trial to compare cilostazol plus aspirin with aspirin alone in patients with symptomatic intracranial artery stenosis (IAS).

Methods: Subjects were patients at age of 45–85 years with ischemic stroke after two weeks to six months from onset and >50% stenosis in responsible intracranial arteries on MRA. They were randomly allocated to either group of cilostazol 200 mg/day plus aspirin 100 mg/day (CA group) or aspirin 100 mg/day alone (A group), who were followed up for two years.

Results: A total of 165 patients (109 males, average 68 years) were randomized. There was no difference in the progression of IAS between both groups (9.6% in CA group and 7.6% in A group, $p = 0.5326$). Stroke recurrence occurred in 2.4%/year in CA group (ischemic 4, hemorrhagic 0) and 5.5%/year in A group (ischemic 6, hemorrhagic 2). After correction with background characteristics, rates of stroke plus silent brain infarct (5.8% vs 12.5%, HR 0.34, 95% CI 0.12–0.96, $p = 0.04$) and all vascular events plus silent brain infarct (10.7% vs 25.0%, HR 0.37, 95% CI 0.14–0.97, $p = 0.04$) were significantly lower in CA than A group.

Conclusion: There was no difference in the progression of IAS between two groups. Rates of stroke plus silent brain infarct and all vascular events plus silent brain infarct were lower in CA than A group.

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Abstract – WCN 2013

No: 1368

Topic: 3 – Stroke

Nonhemiplegic hand strength is weaker in those with deglutition problems at initial one month after stroke

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Background: Dysphasic stroke patients with nasogastric feeding tubes (NGT) are at high risk of malnutrition, which may lead to

prolonged hospital stay and poor functional recovery. Hand grip strength (HGS) is a useful marker of patients' nutritional status at an early stage.

Objective: The objective is to determine if HGS of the nonhemiplegic side at one month post-stroke is significantly different in patients with NGT compared to those without NGT and to assess if HGS measurements correlated to serum markers and functional status.

Patients and method: We retrospectively reviewed medical record of 218 patients admitted to our department from September 2010 to April 2012.

Material and method: We recorded patients' serum markers (albumin), modified Barthel index scores (MBI), mini mental status examination (MMSE), and Berg balance scale (BBS). Correlations analysis of hand strength to biochemical markers was performed using independent t-test.

Result: Mean \pm SD values of MBI, BBS, MMSE albumin, with NGT ($n = 151$)/without NGT ($n = 67$) were $12.5 \pm 20.0/51.3 \pm 31.3$, $6.26 \pm 12.2/27.6 \pm 20.9$, $10.0 \pm 10.1/19.1 \pm 8.0$, and $6.7 \pm 0.7/7.4 \pm 6.7$. The mean lateral pinch and grip strength (lb) of these two groups were $6.5 \pm 7.0/11.7 \pm 5.3$ and $20.1 \pm 23.5/42.3 \pm 20.6$, respectively and all these values showed statistically significant differences ($P < 0.05$).

Conclusion: NGT patients showed weaker grip and lateral pinch strength in nonhemiplegic hand and this correlated to functional status and serum markers. Initial assessment of grip and lateral pinch strength of the contralateral hand may be useful to evaluate the functional nutritional status in those with prolonged NGT from an early stage.

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Abstract – WCN 2013

No: 1373

Topic: 3 – Stroke

On the question of the application of silver nanoparticles in antithrombotic therapy

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Platelets play a key role in the development of thrombotic lesions. Urgent action to prevent platelet activation affecting the process of fibrin polymerization and slowing clot formation.

Objective: To study the effect of silver nanoparticles with 10, 20, 70 nm on the system of cellular and plasma hemostasis in vivo. For the study of platelet aggregation, we used the method of scanning electron microscopy. Silver nanoparticles were pre-incubated with platelet-rich plasma of human blood in the presence of ADP in the control units. The particles do not exceed the diameter of 2.9–3.2 μm . Addition of 60 mM and $d = 70$ nm nanoparticles in platelet rich plasma, caused the formation of aggregates a diameter of 3.5–4.2 μm . It caused the decrease in prothrombin time, which indicates additional aggregation and thrombus formation. By chemiluminescence, registered superweak glow and intense bursts are accompanied by activation of blood neutrophils in the presence of luminol by adding nanoparticles of $d70$ nm at a concentration of 0.064×10^{-11} M, indicating an inflammatory action. When added to the plasma particles 10 and 20 nm, it manifested an anti agregational effect – formed units 1.8–2.5 μm , prothrombin time did not change. Adding 10–20 nm silver nanoparticles in a concentration of 10 to 100 mM prevented fibrin polymerization.

Conclusion: Nanosilver, $d = 20$ nm or less, reduces platelet aggregation, practically does not activate neutrophils and prevents fibrin polymerization. The data suggest that nanosilver $d = 20$ nm or less is

not cytotoxic and may be used in clinical practice as an anti-thrombotic agent. A further study of the effect of nanosilver on the hemostatic system is required.

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Abstract – WCN 2013

No: 1423

Topic: 3 – Stroke

α-Lipoic acid treatment is neurorestorative and promotes functional recovery after stroke in rats

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Background: A considerable body of evidence suggests that oxidative stress is a fundamental mechanism of brain damage in stroke. A recent study suggests that the antioxidant properties of alpha-lipoic acid (aLA) correlate with its ability to promote neuroproliferation. However, there have been no reports of comprehensive studies examining the neurorestorative effects of aLA administered after the onset of ischemia.

Purpose: Here we report the role of aLA as neurorestorative agent in focal cerebral ischemia.

Methods: The middle cerebral artery (MCA) of adult rats was occluded for 2 h and then reperfused. aLA (20 mg/kg) was administered in 71 animals (aLA group) through the left external jugular vein immediately after reperfusion. An equivalent volume of vehicle was administered to 71 animals (control group). A neurological deficit score (NDS) was obtained, and motor impairment was assessed by the accelerating rotarod test. Levels of endogenous neural precursor and glial cell were analyzed by immunohistochemistry.

Results: Immediate aLA administration group after reperfusion significantly reduced the mortality, infarct size, and NDS than control group ($n = 71$, $P = 0.005$, $P = 0.002$, and $P = 0.001$, respectively). Long-term functional outcomes by rotarod test were also markedly improved by aLA treatment ($P = 0.013$). And aLA treatment enhanced the earlier proliferation of endogenous neural precursor and glial cell using nestin- and GFAP-antibody along the infarct and infarct core regions.

Conclusions: These results indicate that urgent treatment with aLA after ischemic injury may have significant neurorestorative effects through enhanced neuroproliferation. Thus, aLA may be a useful intervention for the treatment of acute ischemic stroke.

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Abstract – WCN 2013

No: 1421

Topic: 3 – Stroke

Degree of recovery in motor functions of patients with CVI in relation to the degree of damage on speech comprehension

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Background: Patients with brain damage brought on by cerebrovascular insult (CVI) with damage to the motor function system, often have difficulty in comprehension of speech and language.

Objective: To research the degree of functional impairment and recovery of patients following CVI in relation to the degree of speech and language comprehension by patients with present speech and language disability.

Method: The functional ability was estimated with Barthel index for 102 patients with present speech and language impairments after

CVI, also an estimate on speech and language status with the subtest auditory comprehension BDAE. A correlation was observed between speech comprehension and functional ability.

Results: Average age of the patients was 64.94 years in samples where there were more males than females. The most common type of stroke was ischemic. The most numerous were multifocal lesions in the area of left ACM.

At the beginning stages of rehabilitation the average Barthel index of the patients was 47.95 and after rehabilitation the index was 65.09.

Patients with average comprehension complex ideational material (KIM) under 50% of the test had a starting Barthel index of an average of 39.02, and after rehabilitation it was 55.91.

Patients with average comprehension KIM above 50% of the test had a starting Barthel index of 55.45 and after rehabilitation 72.07. A low correlation was found between the results of auditory comprehension KIM and functional ability of patients. The results are statistically significant.

Conclusion: Patients with larger impairments of comprehension KIM had a lower degree of functional repair in relation to patients with better comprehension of speech and language.

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Abstract – WCN 2013

No: 126

Topic: 3 – Stroke

Recurrent stroke from free-floating carotid artery thrombus: 14-year experience from a tertiary institute in Singapore

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Background: Free-floating carotid artery thrombus is a rare cause of cerebral infarcts. Current understanding is limited to small case-series. Recurrent stroke in free-floating thrombus is believed to be due to delayed embolism from the tail of the carotid thrombus to the intracranial circulation.

Objective: To review recurrent stroke mechanism in free-floating carotid artery thrombus.

Patients and methods: All patients admitted from 1999 to 2012 for cerebral infarcts were included. Cases were retrospectively identified from carotid ultrasound reports showing free-floating carotid artery thrombus. Case notes, neuroimaging and electronic records were retrieved and patients with recurrent stroke were studied.

Results: A total of 28,126 carotid ultrasound reports were screened and 61 patients with free-floating thrombus were identified. Of these, recurrent stroke occurred in 4 patients. Age range: 60 to 83 yrs. Clinical syndrome: pure motor stroke ($n = 2$), sensorimotor stroke ($n = 1$), hemispheric stroke ($n = 1$). BP on admission ranged from 145/75 to 210/80 mm Hg. In 3 patients, neurological deterioration occurred in the first week of admission coinciding with a drop in BP. Etiology of hypotension included dehydration, anemia and inadvertent administration of anti-hypertensive. Brain scan showed watershed infarcts, which collaborated with hypotension as the underlying cause. Recurrent stroke due to embolism from the tail of the carotid thrombus occurred in only one patient with hemispheric syndrome. All patients were treated with heparin and warfarin.

Conclusion: Hypotension is the cause of recurrent stroke in 75% of patients with free-floating carotid artery thrombus. Clinicians treating such patients must be vigilant to this potentially preventable complication.

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Abstract – WCN 2013**No: 1431****Topic: 3 – Stroke****Stroke and leucoencephalopathy**

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Background: Encephalopathy means brain disease, damage and malfunction. It presents broad spectrum of symptoms such as memory loss and subtle personality changes to severe such as dementia, seizures, coma, and death.

Posterior leucoencephalopathy syndrome was first described in 1996. It is a condition where diagnosis depends on clinical and radiological features. Leukoencephalopathy seen in association with severe hypertension, disease leading to failed out regulation, hyperperfusion, and endothelial injury vasogenic edema. Vasoconstriction and hyperperfusion lead to brain ischemia and vasogenic edema. MRI scan is useful in giving prognosis.

Material: We have in stroke unit for 225 consecutive patients with ischemic stroke investigated systematically with MRI.

Imaging 156 patients (69,333%), MRI shows edema involving the white matter in the posterior portions of the cerebral hemispheres, especially in the parieto occipital regions. Diffusion weighed sequences can be differentiated between edema from leaking capillaries and cell.

Conclusion: In 78% (123 patients) of patients, the acute infarct was deep. And leucoencephalopathy was more frequent in patients with a deep infarction than in patients in whom the cortex was involved. Hypertension was the most common risk factors 87% and was more frequent in study than in control group. This study suggests that hypertension may be more strongly associated with leucoencephalopathy than with deep infarction.

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Abstract – WCN 2013**No: 1430****Topic: 3 – Stroke****Effect of short-term changes of air pollution on the development of acute ischemic stroke**

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Background: Thrombolytic treatment (TT) of acute ischemic stroke (AIS) requires a highly organized hospital service due to the strict therapeutic protocol. All data that can be used to predict the demand on the healthcare system have practical benefit. According to literature data, air pollution (AP) plays a role in the development of stroke.

Objective: We examined the role of short-term changes of AP in the development of AIS. Our previous results did not confirm the effect of AP averaged by season on the development of stroke. Analysis of TT-s — due to their narrow therapeutic window — allowed us to determine the onset of stroke events, which is important for the examination of short-term meteorological effects. We had to reduce the sample size in order to assess highly localized environmental effects.

Patients and method: Stroke onset dates and times of 80 patients who received TT at our department between 2009 and 2012 were compared with measurement data (PM₁₀, NO, NO₂, NO_x, SO₂, O₃, CO) from nearby meteorological stations. Correlation was analyzed with SPSS software, Pearson's, Kendall's tau-b and Spearman's tests, and Student's t-test.

Results: There was a significant correlation between the increase of concentration of most polluting materials (NO: 0.1050, NO₂: 0.0490, NO_x: 0.1060, CO: 0.1140) and the development of stroke. A negative correlation was found with sulfur-dioxide and ozone-levels (SO₂: -0.0670, O₃: -0.0900) and no correlation with PM₁₀-levels. The t-test did not show significance due to the low number of cases.

Conclusion: Higher air pollution only slightly increases the short term risk of stroke.

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Abstract – WCN 2013**No: 1436****Topic: 3 – Stroke****Kinetics of inflammatory markers in acute ischemic stroke and their relevance in stroke-induced immunosuppression**

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Background: Stroke-induced immunosuppression syndrome (SIIS) leads to severe complications in stroke patients. Infection is the most common of these complications and the chief cause of morbidity and mortality in stroke survivors.

Objective: We aimed to identify an inflammatory marker for the distinction between the inflammatory response evoked by stroke and inflammation later caused by infection due to SIIS.

Patients and method: We investigated the kinetics of CRP, WBC, neutrophil count, suPAR, CD4 + CD25high Tregs, CD64 + and CD177 + neutrophils and monocytes in 12 acute ischemic stroke (AIS) patients within 6 h (Stroke 1) and one week after the insult (Stroke 2). Patients with infection after stroke were excluded. As controls, 14 age-matched healthy individuals with similar cardiometabolic risk factors, but with a negative history of stroke or other neurological disorders were included.

Results: CRP, WBC and neutrophil count values were comparable in Stroke 1 and controls; however, they were elevated in Stroke 2. suPAR levels were higher in both stroke groups compared to controls. The prevalence of CD64 + neutrophils was higher in Stroke 1 than in controls and it decreased in Stroke 2 below the level in controls (5.95 [5.41–8.75]% vs. 32.38 [9.21–43.93]% vs. 4.06 [1.73–6.77]%, p < 0.05). The prevalence of CD177 + neutrophils was higher in both stroke groups compared to controls.

Conclusion: Our pilot study identified the prevalence of CD64 + neutrophils as an inflammatory marker that best represents immunological alterations in AIS. Its level is a suitable candidate for the indication of the developing inflammatory response due to infection.

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Abstract – WCN 2013**No: 1428****Topic: 3 – Stroke****Dysphagia tests and intensive special neurorehabilitation should be regular part of diagnostic and neurorehabilitation algorithmus in stroke unit**

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Background: Dysphagia is a frequent symptom of ischaemic or haemorrhagic cerebral stroke (iCI, ICH). The incidence of dysphagia is ranging from 25% to 81%. The most often complication of dysphagia is aspiration which often leads to pneumonia. The reason why the determination and therapy of dysphagia and aspiration is so important is that the aspiration (especially silent aspiration) contributes to higher mortality as well as to the increase of costs.

Aim:

1. To analyse the occurrence of dysphagia and its complication: aspiration “silent” aspiration.
2. To evaluate the results of intensive special neurorehabilitation.

Material and methods: Material consists of 113 subjects. Whole group was divided into two subgroups: the 1st iCI patients with neurorehabilitation by certified dysphagiologist (iCI-d, n = 66), mean age 68.08 ± 11.41 years, male 59.09%, the 2nd subgroup with different neurological disorders (n = 47), mean age 65.09 ± 15.52 years, male 62.62%. iCI with various degree of neurological deficit (NIHS) was confirmed by CT/MRI. For confirmation of dysphagia clinical swallowing examination and instrumental evaluation—videofluoroscopy was used. For evaluation of aspiration Rosenbeck P/A scale was used.

Results: The occurrence of dysphagia was documented in 57 from 66 iCI patients (86.36%) and 29 (43.93%) of them had aspiration, and “silent” aspiration was found in 10 (15.15%) patients. Significant improvement following neurorehabilitation was documented in all stroke patients with dysphagia. From the 47 patients 2nd subgroup (non-stroke patients) we have found out 20 patients (42.55%) with dysphagia and 6 (12.76%) of them had aspiration. 1 (2.12%) patient had “silent” aspiration.

Conclusions:

1. Special tests for dysphagia should be the obligatory part of diagnostic algorithm.
 2. Special neurorehabilitation by certified dysphagiologist should be part of therapeutic algorithm in stroke patients.
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Abstract – WCN 2013

No: 1406

Topic: 3 – Stroke

Predictors of early efficacy of thrombolytic therapy in patients with ischemic stroke

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The aim of the study was to evaluate association of age, smoking, atrial fibrillation and time from symptoms onset with early efficacy of thrombolytic therapy in patients with ischemic stroke.

Materials and methods: We conducted retrospective analysis of 44 cases of systemic and intraarterial thrombolytic therapy in patients with ischemic stroke that are admitted at Sverdlovsk Regional Clinical Hospital and Demidovskaya Hospital in Nizhnii Tagil in 2011. Neurologic deficit was evaluated according to NIHSS on admission and at the 7th day of hospitalization. Early effectiveness of thrombolytic therapy was defined when NIHSS at the 7th day decreased in ≥ 4 points.

Results: Median age was 59.5 (IQR 53.0–70.0) years. 19 (43.2%) patients smoked. Atrial fibrillation was noted in 34.1% (15) of patients. Median time from symptoms onset to beginning of rtPA infusion was 187.5 (IQR 152.5–217.5) min. Median NIHSS on admission was 11.5 (IQR 9–16.5), NIHSS at the 7th day of hospitalization was 6.0 (IQR 4–12). Patients with cardioembolic stroke were noted to have more severe neurologic deficit on admission, median NIHSS was 14.0 (IQR 12.0–18.0). Improvement at

the 7th day of hospitalization was achieved in 29 (66%) patients. Statistical analysis revealed significant association of early neurological improvement and time from symptoms onset. Spearman's coefficient of rank correlation was -0.424 (95% CI, -0.640 to -0.145 , $P = 0.0042$).

Conclusion: Early efficacy of rtPA infusion depended on time from symptoms onset to rtPA infusion. Statistical analysis didn't showed significant correlation of other factors with early improvement after rtPA infusion due to insufficient sample size.

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Abstract – WCN 2013

No: 1405

Topic: 3 – Stroke

Severe stress and post-stroke depression

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Purpose: To identify the main factors assisting to the development of post-stroke depression.

Patients and methods: Totally 75 ischemic stroke patients are investigated. Stroke severity is assessed by NIHSS. Stroke risk factors are studied retrospectively: age, ethnicity, previous stroke/TIA, atherosclerosis, hypertension, atrial fibrillation, diabetes mellitus, recent infection, smoking, alcohol abuse, severe social and psychological stress. Patients are divided as right hemispheric stroke (1st group—18 patients), left hemispheric stroke (2nd group—26 patients), brainstem stroke (3rd group—15 patients), and cerebellar stroke (4th group—16 patients). Ischemic lesion is evaluated on conventional CT at 24 h after onset. Stroke outcome is evaluated by mRS at 1 month. Depression is assessed by Zung Depression Scale at 1 month. Statistics are performed by SPSS-11.0.

Results: In the 1st group initial NIHSS was 21.5 ± 4.5 . Severe depression was found in (46.15%), moderate depression in (7.69%), and mild depression in (15.38%) of patients. In the 2nd group the initial NIHSS was 14.0 ± 3.5 . Severe depression was found in (33.3%), moderate depression in (13.4%), and mild depression in (6.7%) of patients. In the 3rd group initial NIHSS was 16 ± 4.5 . Severe depression was found in (44.5%), moderate in (11.2%), and mild in (16.7%) of patients. In the 4th group the initial NIHSS was 12 ± 2.5 , and severe depression was found in (12.5%), and mild depression in (6.25%) of patients. The initial NIHSS positively correlated with depression ($r = +0.25$; $p < 0.05$). Correlation was not found between lesion location and depression. Multivariate logistic regression revealed the significance of initial NIHSS in conjunction with severe stress for depression at 1 month post-stroke.

Conclusion: Probably, the pre-stroke psychological instability and stroke initial severity predispose toward post-stroke depression.

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Abstract – WCN 2013

No: 1347

Topic: 3 – Stroke

Estimated glomerular filtration rate and risk of survival in acute stroke

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Objective: To assess the risk of survival in acute stroke using the Modification of Diet in Renal Disease (MDRD) equation derived from estimated glomerular filtration rate (eGFR).

Method: 83 acute stroke patients had GFR calculated within 48 h of admission after basic data were captured. Stroke outcome was defined as either discharged or still-in-care (survived) or all cause in-hospital death.

GFR was estimated by the MDRD equation and stroke severity was assessed by the Canadian Neurological Scale (CNS). Data were compared between the GFR groups of <60 ml/min and ≥60 ml/min. Relative risks (RR) and odds ratios (OR) for stroke outcomes (survival and death) were estimated between the GFR groups and the homogeneity of the odds ratios among the different layers of stroke severity (CNS <6.5 and ≥6.5) was determined by Breslow–Day and Tarone's test. Matanel Hazensel and Cochran's tests were used to determine conditional independence and the common odds ratio with stroke severity as a layering variable.

Results: RR of survival and death of the GFR groups—less than 60 ml/min and above or equal to 60 ml/min were 0.425 and 1.204 and 2.360 and 0.830, respectively. The OR of survival of GFR below 60 ml/min compared to GFR above or equal to 60 ml/min was 0.353. There was homogeneity across the two layers of stroke severity (CNS score less than 6.5 and above or equal to 6.5), $p = 0.612$ and 0.612 .

Conclusion: Independent of stroke severity, GFR is a surrogate in the assessment of the risk of survival in acute stroke.

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Abstract – WCN 2013

No: 1414

Topic: 3 – Stroke

Venous infarction in young female patients

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Purpose: To investigate the risk of venous infarctions in young females with ischemic stroke.

Patients and methods: Totally 57 female stroke patients aged from 35 to 55 years investigated. Stroke Etiology determined according to TOAST. Stroke severity assessed by NIHSS. Stroke risk factors investigated retrospectively. Brain visualized by MRT (1.5 T) at 24 h. Dopplerography conducted in all patients. Statistics performed by SPSS-11.0.

Results: Cardioembolic stroke found in 19 patients (33.3%), lacunar stroke – in 12 patients (21%), other etiology – in 26 patients (45.7%), where 17 cases (65.4%) were venous infarction. Main risk factors in cardioembolic stroke were congenital cardiac valve deficiency – 9 patients (47.3%), cardiac post-operational period – 5 patients (26.3%), and acute myocarditis – 5 patients (26.3%); In lacunar stroke-hypertension due to chronic kidney pathology in 6 patients (50%), heredity hypertension – in 3 patients (25%), chronic vasculities – in 2 patients (16.6%), and diabetes mellitus – in 1 patient (8.4%). Other etiology stroke risk factors included long term (≥10 years) oral contraceptives usage – in 21 patients (80.7%), smoking – in 20 (76.9%) patients, systemic lupus – in 4 patients (15.3%), Fabry disease – in 2 patients (7.6%), and anti-phospholipids – in 3 patients (11.7%). Multivariate logistic regression revealed the significance of long term oral contraceptives application in conjunction with smoking and chronic venous insufficiency (blood flow velocity in cavernous and transverse sinuses >30 cm/s) for brain venous infarctions.

Conclusion: Presumably, the long-term application of oral contraceptives in conjunction with chronic venous insufficiency crates the risk for venous infarctions in females.

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Abstract – WCN 2013

No: 1333

Topic: 3 – Stroke

Decompressive hemicraniectomy worth in patients with cerebral infarction in the territory of the middle cerebral artery and treated with rt-PA

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Background: The aim of this paper is presentation of patients with malignant MCA infarction treated concomitant therapy rt-PA and decompressive hemicraniectomy.

Method: In 2009–2012, 215 patients were treated with rt-PA. Malignant brain edema diagnosed clinically and CT occurred in 6 of them (2.3%), aged up to 61, treated with decompressive hemicraniectomy. Analysis included: the time of the beginning malignant edema after the beginning of the stroke, mortality (at 30 and 90 days), quality of recovery evaluated with the mRs.

Results: Among 6 patients with malignant MCA infarction with space-occupying edema there were two women and four men aged 40–61. On the average, the patients scored 15.8 point in NIHSS on admission. On day one of brain edema was present in all the patients. At the moment of qualifying a patient for hemicraniectomy the NIHSS score deteriorated by 9 points, on the average. CT showed brain swelling, and in 2 secondary hemorrhagic cerebral infarction. One patient died on day 11 following hemicraniectomy (the surgery was performed when one pupil was dilated and nonreactive), one – died on day 40. The other four patients survived and are still alive.

Conclusions:

1. Decompressive hemicraniectomy in malignant MCA patients treated with rt-PA is a safe procedure, which improves their chances for survival.
2. Secondary hemorrhagic cerebral infarction should not be a contraindication for decompressive hemicraniectomy.
3. Decompressive hemicraniectomy should be performed as promptly as possible.
4. Good cooperation between a neurologist and a neurosurgeon is necessary to successfully perform hemicraniectomy.

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Abstract – WCN 2013

No: 1472

Topic: 3 – Stroke

The dissemination and conditions for endovascular treatments in acute ischaemic stroke in Europe

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Introduction: Endovascular approaches are new emerging methods in the management of acute stroke. The aim of this study is to establish the organization of endovascular treatments for stroke in European countries.

Material and methods: A structured questionnaire regarding stroke unit (SU) organization and usage of endovascular treatments for acute stroke in the years 2009–2011 was acquired from representatives of the European countries, mostly being members of the EFNS Stroke Scientist Panel or local opinion leaders.

Results: Data from 19/47 (40%) European countries was received. The number of comprehensive SU overall was 181 (mostly – 100 in Germany). Intraarterial thrombolysis was reimbursed in 13/19 and

mechanical thrombectomy in 11/19 countries. 6301 procedures were performed (mostly in Germany – 4428 and none in Iceland and Cyprus). An estimate of 0.19% of stroke patients were treated and (highest in Slovenia – 0.92%) .91% of the procedures were done by neuroradiologists and only 0.8% by neurologists.

Conclusions: There is a lot of diversity in the use of endovascular procedures for acute stroke within Europe. The method is underused, but lack of evidence does not support the notion for widespread use. Neurologists' involvement is insufficient, but current UEMS guidelines support the training of neurologists in interventional neuroradiology.

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Abstract – WCN 2013

No: 983

Topic: 3 – Stroke

Cerebrovascular risk factors and depression: Report from National Ambulatory Medical Care Survey 2002–2009

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Background: Depression is a common psychiatric disorder that can occur as a result of many factors. Recently, a link between cerebrovascular risk factors and depression has been suggested. We performed this study to identify the association between depression and cerebrovascular risk factors.

Method: We analyzed nationally representative data derived from patients enrolled in the National Ambulatory Medical Care Survey (NAMCS) between 2000 and 2009. Data analyzed included patients' age, gender, race/ethnicity, presence and control of cerebrovascular risk factors and self-reported depression.

Results: During the 9-year study period, an estimated 148,857 patients enrolled in this study. There were 14,849 (7.4%) of 148,857 patients who reported depression. Of the patients with depression, 2.8% had a history of cerebrovascular diseases. Among patients with depression (compared with those without), frequency of body-mass index >30 (41.1% versus 31.9%, $P < 0.05$), cigarette smoking (33.8% versus 25.2%, $P < 0.05$), hypertension (28.4% versus 23.6%, $P < 0.05$), diabetes mellitus (12.3% versus 10.1%, $P < 0.05$), and hyperlipidemia (18.1% versus 13%, $P < 0.05$), was significantly higher.

Conclusion: High frequencies of cerebrovascular risk factors were identified among persons with depression despite absence of any overt cerebrovascular disease. Further studies need to identify whether cerebrovascular risk factors have a causative role in depression or are a consequence of depression.

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Abstract – WCN 2013

No: 1432

Topic: 3 – Stroke

Epidemiological and prognosis aspects of stroke of the elderly in the neurological clinic of Fann Teaching Hospital, Dakar – Senegal

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Background: Strokes are a vital emergency and functional. Their frequency increases with the aging population so that the senior pays a heavy price for this condition.

Objective: The objective of this study was to describe the epidemiological and prognosis of stroke in the elderly at the Fann Teaching Hospital, in Dakar.

Patients and methods: This is a retrospective study from 1st January 2001 to 1st November 2003 on patients aged 55 and older with stroke and had a brain CT scan. Sociodemographic data, medical history and surgical signs of severity associated with neurological symptoms and the prognosis were collected.

Results: Records of 314 patients with stroke collected, 228 were aged 55 and over for a prevalence of 72.6%. The patient population consisted of 54.8% of women with a mean age of 67.9 years (± 8.5). Risk factors were dominated by high blood pressure (74.1%), diabetes (11.8%), the previous stroke (10.1%) and heart disease (3.1%). Ischemic stroke accounted for 65.8%. Coma (22.4%) and high blood pressure (21.9%) were associated with neurological symptoms. Lethality of 26.3% was observed with the main predictor coma (OR = 33.3, confidence interval 95% of 13 to 83.3).

Conclusion: It should develop and implement a health education program to reduce morbidity and mortality from stroke in the elderly.

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Abstract – WCN 2013

No: 1443

Topic: 3 – Stroke

Neuroplasticity in neurodegenerative diseases and stroke

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Neurodegenerative diseases and stroke are foremost causes of medical, as well as socioeconomic problems in modern society leading to quality of life impairment of patients and caregivers.

Disorders like stroke, dementia, Alzheimer's and Parkinson's disease, Huntington disease, multiple sclerosis and acquired brain trauma contribute to the decline of cognitive, motor, and sensory abilities.

Neuroplasticity, also known as cortical remapping, challenges the idea that brain functions are fixed in certain time. It refers to changes in neural pathways and synapses which are due to continuous stimulation and practice enhanced by previous experience.

Mirror-neurons system is activated both in action and observation. Practicing mental stimulation improves memory and attention.

Neuroplasticity can act through two possible mechanisms on motor deficit and cognitive impairment, by forming new, or improving existing pathways. The changes in the cortex organization include an increase in the number and density of dendrites, synapses and neurotrophic factors. After damage has been afflicted to the motor cortex, changes of activation in other motor areas are observed. These changes occur in homologue areas of the non-affected hemisphere which can substitute for the lost functions or in the intact cortex adjacent to the damage. Thanks to these cortical reorganizations the patients can recover, at least in part, their lost abilities.

Brain health is maintained through control of conventional vascular risk factors, practice of physical activity, healthy nutrition, stress management and mental stimulation. Challenging the brain with different tasks creates new neural connections and intensive exercise leads to improvement in neuroplasticity.

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Abstract – WCN 2013**No: 1440****Topic: 3 – Stroke****Neuroimmune correlations in acute ischemic stroke**I. Solodovnikova, A. Son. *Odessa National Medical University, Odessa, Ukraine***Background:** Neuroinflammation and neuronal apoptosis play important role during early brain injury in acute ischemic stroke (AIS).**Objective:** The aim of this study was to analyze the correlations between CD4+/8+, CD7+, CD25+, CD95+, CD25+/95+, CD38+, and CD54+ cell counts and clinical outcome in patients with AIS.**Patients and methods:** 56 males (63.2 ± 4.7 years) with AIS were examined. CD4+/8+, CD7+, CD25+, CD95+, CD25+/95+, CD38+, and CD54+ cell counts were measured using immunoenzymometric PAP method. The clinical outcomes were assessed by the modified Rankin Scale (mRS).**Results:** Patients with CD4+/8+ 1.70–4.50 and CD7+ 151.00–591.00 cells/Mcl or CD4+/8+ 1.70–4.63 and CD38+ 122.00–726.00 cells/Mcl cell counts during first 24 h after AIS showed mild disability (mRS ≤ 1) (p < 0.01). Patients with CD25+/95+ 0.29–1.15 and CD7+ 238.00–330.00 cells/Mcl cell counts by 8–10th day after AIS showed severe disability (mRS ≥ 4) (p < 0.01). Patients with CD4+/8+ 1.90–2.71 or CD25+ 382.00–652.00 cells/Mcl cell counts by 21st day after AIS showed mild disability (mRS ≤ 2) (p < 0.01). Patients with good outcome (mRS ≤ 1) three weeks after AIS had CD54+ cell counts 473.00–748.00 cells/Mcl during first 24 h after AIS (p < 0.01).**Conclusion:** Neurological deficit and clinical outcome in patients with AIS correlate with CD4+/8+, CD7+, CD25+, CD95+, CD25+/95+, CD38+, and CD54+ cell counts.

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Abstract – WCN 2013**No: 1445****Topic: 3 – Stroke****Effects of afobazole on brain tissue under the conditions of new model of local ischemia**K. Alikhanyan^a, M. Balasanyan^b, A. Mkhitarian^c. ^aDepartment of Clinical Pharmacology, Muratsan Hospital of Yerevan State Medical University, Yerevan, Armenia; ^bDepartment of Pharmacology, Yerevan State Medical University after M. Heratsi, Yerevan, Armenia; ^cDepartment of Clinical Pathology, Muratsan Hospital of Yerevan State Medical University, Yerevan, Armenia

Investigations with neuroprotective drugs have shown great promise in preclinical testing but disappointment in clinical trials. So, the aim of our investigation was to develop new local ischemia model more precisely reflecting the clinical picture of stroke, when the acute ischemic disorders develop on the background of chronic ischemization of cerebral tissue and to study neuroprotective effects of afobazole in this model.

White inbred rats were used. Chronic ischemization of brain tissue was caused by hypokinesia (rats were individually housed in narrow cages for 15 days). Local ischemia was caused by the ligation of middle cerebral artery (MCA).

Data obtained have shown local ischemia caused in hypokinetic rats was conducted by big areas of laminar neuronal necrosis in cortex, accumulation of neutrophils, massive glial scars, focal hemorrhages, which were more significant after 12 days of MCA occlusion compare with 6th day of MCA ligation. Histopathological analyses of brain of rats treated with afobazole (twice daily, 5 mg/kg, i.p.) during 6 and 12 days have shown diminished subarachnoidal edema, macrofagal reaction was manifested without infiltration of brain tissue, the short areas of laminar necrosis of brain cortex and single areas of pancortical necrosis

were mentioned. Afobazole has shown also ability to limit the size of ischemic zone after 6 and 12 days of MCA occlusion.

Thus, afobazole evidently displays neuroprotective effect in the new more relevant to clinical stroke experimental model of local ischemia. It opens new possibilities for afobazole as a potent neuroprotective agent for the treatment of stroke.

doi:10.1016/j.jns.2013.07.801

Abstract – WCN 2013**No: 633****Topic: 3 – Stroke****The recent trends of the change of risk factors with stroke patients at the hospital in Central Tokyo**T. Takemi, T. Kimura, A. Unaki, H. Okayasu. *Neurology, St. Luke's International Hospital, Tokyo, Japan***Background:** In recent years, stroke patients with dyslipidemia as their risk factor have been increasing at our hospital. We assumed the changes in lifestyle and environment of modern times to explain this trend.**Objective:** Regarding the above clinical question, we consider it is important to examine the change of risk factors prevalence in the younger patients compared with the elder patients, for the prevention of stroke.**Patients and methods:** This study involved consecutive 1853 patients admitted to our hospital with the diagnosis of ischemic stroke from April 2001 to March 2013, and was conducted by comparative review on patients of two groups about the common risk factors (hypertension, diabetes mellitus, dyslipidemia, and atrial fibrillation). Group A includes patients aged less than 60 years old. Group B includes patients aged 60 and over.**Result:** Group A has 415 patients and Group B has 1438. The percentage of patients with hypertension, diabetes mellitus, dyslipidemia, and atrial fibrillation as their risk factor was 48.7%, 27.5%, 45.5%, and 9.4% in the Group A, and in the Group B, 63.9%, 29.8%, 34.4%, and 23.7%, respectively.**Conclusion:** In the Group A, the percentage of patients with dyslipidemia is higher than in the Group B. However, in recent several years, it has been increasing also in the elder patients. Our result indicates the importance of vigorous control of dyslipidemia for the prevention of stroke. To get national trends, it is requested to expand further study throughout all of Japan.

doi:10.1016/j.jns.2013.07.802

Abstract – WCN 2013**No: 1494****Topic: 3 – Stroke****Quality of life in aphasia after stroke in Cotonou, Benin**D.D. Gnonlonfoun^a, C. Adjien^a, P.M. Ossou-Nguiet^b, T. Adoukonou^c, D. Houinato^a, D.G. Avodé^a. ^aNeurology, CNHU-HKM, FSS/UAC, Cotonou, Benin; ^bNeurology, Faculté de Médecine de Brazzaville, Brazzaville, Congo; ^cNeurology, Université de Parakou, Parakou, Benin**Background:** Aphasia, a consequence of stroke is an inability to understand, formulate language or combination of both.**Objective:** The aim is to report the perceived quality of life among aphasic patients in Cotonou.**Methods:** It is transversal, prospective study of descriptive and analytic aim conducted in extern neurological department of CNHU-HKM and orthophonie clinic in liberal consultation, from October to December 2012, including all patients with aphasia after stroke by dating six months at least. For each patient, the Sickness Impact Profile or SIP-65 was administered. The demographic characteristics

and the clinic data were the variables studied. Statistical analysis was performed by Epi info 3.5.

Results: Forty one patients were enrolled including 27 males and 14 females. The age varied from 36 to 72 years with of a mean of 55.3 ± 2.5 years. The mean duration of disease was 10 months. The Broca's aphasia was predominant (68.3%) and also the ischemic stroke (58.5%). Most of our patients answered "false" to more than half of the questions, except for communication, resumption of work and distraction. Bivariate analysis showed that: the age, the sex, the mode of home, the support, the profession, the type of stroke and of the aphasia and the orthophonic treatment were associated to the quality of life.

Conclusion: The aphasia is a common condition after stroke, affecting the quality of life, especially the resumption of work and the speech. Those items must be considered in order to get a good reeducation.

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Abstract — WCN 2013

No: 1485

Topic: 3 — Stroke

Clinical significance of vertebral artery hypoplasia in lateral medullary infarction

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Background: It is controversial whether vertebral artery hypoplasia (VAH) is a risk factor for stroke in the posterior cerebral circulation or not. The aim of our study was to investigate the frequency and clinical relevance of VAH in lateral medullary infarction (LMI).

Methods: In total of 40 patients with LMI, the frequency of VAH, defined as a diameter of ≤ 2.2 mm by contrast-enhanced magnetic resonance angiography, was measured. We assessed the risk factors for stroke in each patient. In all LMI patients, transcranial Doppler was performed.

Results: Among them, 13 patients (32.5%) had VAH. It was detected in 6 patients (28.6%) of the 21 patients with age range of 15–64 years and in 7 patients (36.8%) of the 19 patients with age range of 65–99 years. In 3 young patients with age range of 15–40 years, all patients had VAH (100%) but had no additional risk factors for stroke. In very elderly patients (5 with age range of 80–99 years), 3 patients (60%) had VAH and had only hypertension as an atherosclerotic risk factor. 13 patients with VAH demonstrated low mean flow velocity (20–35 cm/s) in distal symptomatic vertebral artery.

Conclusion: The clinical significance of VAH is not yet clearly defined. But VAH, which shows decreased flow velocity, might be more susceptible to pro-thrombotic or atherosclerotic processes than normal or dominant vertebral artery in young or very elderly patients with few risk factors.

doi:10.1016/j.jns.2013.07.804

Abstract — WCN 2013

No: 1377

Topic: 3 — Stroke

Extensive cerebral sinus thrombosis involving bilateral internal jugular, sagittal, sigmoid and transverse sinuses with clinical presentation of subarachnoid hemorrhage

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Introduction: Cerebral sinus thrombosis is less common than most other types of strokes and is thousand times less prevalent than

arterial stroke. Causative factors are different from arterial stroke that includes infection, dehydration, local trauma, rheumatologic diseases, hypercoagulable states, cancer and use of oral contraceptives. We are presenting an unusual case with extensive cerebral sinus thrombosis.

Case description: This case is about a 36 year-old Indian male who was working in Dubai as a laborer for the last 9 years. He presented with sudden onset severe thunderclap headache with some vomiting but without any other neurological or medical complaints. On clinical examination he was a little confused but no other neurological signs except neck stiffness and bilateral papilloedema. Plain CT brain showed evidence of subarachnoid hemorrhage in bilateral frontal and parietal areas. CT venogram showed extensive thrombosis involving bilateral internal jugular veins, posterior two third of superior sagittal sinus, sigmoid sinus and bilateral transverse sinus thrombosis. Further workup including thrombophilia profile, vasculitic profile, thyroid function, coagulation profile, paraneoplastic workup was negative. There was no evidence of any underlying neoplastic process and any hypercoagulable state.

Discussion: This case is interesting and unusual as his clinical presentation resembles subarachnoid hemorrhage and secondly patient is very stable except headache in spite of extensive involvement of the sinus thrombosis. And finally all workup for sinus thrombosis was negative except mild relation to dehydration.

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Abstract — WCN 2013

No: 1495

Topic: 3 — Stroke

Formation of the register for patients with intracranial arterial aneurysms

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Introduction: Aneurysm is a chief cause of nontraumatic subarachnoid hemorrhage (SAH). The risk of SAH development accounts for about 1% a year, and lethality reaches 40–50%.

Objective: Formation of a register for patients with intracranial arterial aneurysms (IA) with SAH and incidental IA and assessment of their functional outcome.

Materials and methods: The register involves 45 patients with treated in the Neurosurgical Department in 2012. Analysis of the functional outcomes has assessed according to the Modified Rankin Scale and Rivermead Mobility Index at the time of discharge from the hospital and a year later, just now 15 patients.

Results: 12 of 15 patients (6 — SAH) had undergone surgery (4 SAH, and 8 — 6–12 months after SAH). In 3 — lethal outcome (3 SAH). In 4 — improvement of the functional condition in a year after surgery as compared to the moment of discharge from the hospital. In 4 — aggravation of the functional condition (1 — SAH). In 1 — none of alterations. 3 patients had not undergone surgery (2 — lethal outcome, Hunt and Hess Grade >4), 1 — functional condition didn't change over a year.

Conclusion: As the register of patients with arterial aneurysms is a single objective method to determine the rate of subarachnoid hemorrhage in patients with arterial intracranial aneurysms, efficiency of neurosurgical interventions, and assessment of functional outcome, it is necessary to continue its formation.

doi:10.1016/j.jns.2013.07.806

Abstract – WCN 2013**No: 1506****Topic: 3 – Stroke****Bedside dysphagia screening after acute stroke: Incidence, screening, and clinical outcomes**J.A.S. Vatanagul, C.P.V. Gallemmit. *Internal Medicine, Perpetual Succour Hospital, Cebu City, Philippines*

Background: Dysphagia is a common finding in the clinical setting of a patient with acute stroke, which can give rise to a risk of aspiration, pulmonary infections, fluid depletion, and malnutrition. Therefore, early identification of dysphagia is crucial to avoid these adverse health consequences.

Objectives: To determine the incidence and clinical outcomes of acute stroke patients with dysphagia admitted at a tertiary hospital in Cebu City from December 2011 to July 2012.

Study design: Prospective, single center, descriptive study.

Materials and methods: The study population included all patients aged 18 years old and above admitted during the study period due to an acute stroke, whether an ischemic infarct or hemorrhage who underwent dysphagia screening test within 24 h of admission and with no previous documented dysphagia. Patients who passed the screening test were fed orally and those who failed were inserted with nasogastric tube for feeding and oral medications.

Results: A total of 74 patients with acute stroke admitted at a tertiary hospital in Cebu City from December 2011 to July 2012 were enrolled in this study. Out of the total 74 respondents, 38 patients passed the dysphagia screening test and 36 patients failed. Eight (22%) of the 36 patients developed pneumonia, and only one patient died of pneumonia.

Conclusion: Early identification of dysphagia with a simple and inexpensive bedside screening procedure can certainly decrease the risk of pneumonia to develop among patients who have failed the dysphagia screening test, as well as reduce mortality and overall healthcare expenditure.

doi:10.1016/j.jns.2013.07.807

Abstract – WCN 2013**No: 1510****Topic: 3 – Stroke****Cerebral venous sinus thrombosis (CVST): Study of four Filipino patients and literature review**J.A.S. Vatanagul, I.A. Rulona. *Internal Medicine, Perpetual Succour Hospital, Cebu City, Philippines*

Research design: Case series study.

Background: Cerebral venous sinus thrombosis (CVST) is rare, with estimated 3–4 cases per million annual with the introduction of venography. Most cases of cerebral venous sinus thrombosis are due to hypercoagulability. Other important risk factors include pregnancy, primary antiphospholipid syndrome, and hereditary thrombophilias.

Objective: To present four Filipino patients with radiologically-proven cerebral venous sinus thrombosis (CVST) and discuss their demographic, etiologic and clinical characteristics and clinical outcomes.

Methods: A retrospective and descriptive analysis of the medical records of four patients with CVST, who were admitted to a single tertiary hospital.

Results: Four patients were identified with a mean age of 36.25 years old. The youngest patient was 22 years old. All of the patients were women. Two of the cases had previous use of oral contraceptives. The most frequent clinical manifestations were headache as seen in all patients (100%), dizziness (50%), vomiting (25%), motor weakness (50%) and seizures (25%). Three patients (75%) presented with thrombophilia – protein C deficiency.

Cranial CT scan revealed subarachnoid hemorrhage in 50% of the patients while parenchymal hemorrhage was present in the other 2 cases. Superior sagittal sinus (50%) and transverse sinus (50%) were the most common locations. All cases received oral anticoagulation with warfarin. All patients were discharged stable and without any neurologic deficits and adverse outcomes.

Conclusions: Venous sinus thrombosis may present to the physician in a number of clinical presentations. Early recognition of the condition and investigation of appropriate therapy probably reduce mortality and morbidity.

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Abstract – WCN 2013**No: 1515****Topic: 3 – Stroke****Expression profile of microRNAs in the peripheral lymphocyte of acute stroke patients**H. Zhao^a, P. Liu^a, R. Wang^a, X. Liu^a, X. Wu^b, C. Xu^c, X. Ji^a, L. Gao^c, Y. Luo^a. ^aCerebrovascular Diseases Research Institute, Xuanwu Hospital of Capital Medical University, Beijing, China; ^bDepartment of Neurology, The First Affiliated Hospital of Liaoning Medical University, Jinzhou, China; ^cDepartment of Neurology, Xuanwu Hospital of Capital Medical University, Beijing, China

MicroRNAs (miRs) profile of whole blood in acute stroke patients has been reported, but the profiling of peripheral lymphocyte has not been characterized. Since the key role of immunity and inflammation in stroke pathology, we currently profiled peripheral lymphocyte miRs of acute stroke patients and healthy persons with Agilent human microarray. We identified 70 down-regulated and 35 up-regulated miRs (>2-fold change) in lymphocyte by differential analysis. Next, 7 deregulated miRs (miR-99a, -99b, -181c, -181d, -212, -424, and -532-5p) were selected randomly and detected in patients' plasma by qRT-PCR, and we found their trends were just the same as their counterparts in peripheral lymphocyte. Pathway analysis indicated 58 significantly changed KEGG pathways targeted by the deregulated miRNAs including MAPK pathway, ubiquitin mediated proteolysis and focal adhesion pathways concerning molecular mechanisms of cell apoptosis and inflammation response. Moreover, a miRs-mRNAs network was created based on bioinformatics analysis of pathways related with immune cell proliferation, chemotaxis and adhesion, presenting 22 critical miRs and their 139 targets to visualize hypothetical interactions. Taken together, except for profiling miRs of lymphocyte in acute stroke patients, we also showed that some miRs in lymphocyte might be related with plasma miRs, and further provide important miRs as therapeutic targets of regulation of lymphocyte function in acute stroke patients.

doi:10.1016/j.jns.2013.07.809

Abstract – WCN 2013**No: 1537****Topic: 3 – Stroke****Circulating beta-antithrombin glycoform increases during the acute ischemic cerebrovascular event**A.M. Garcia^a, M. de la Morena-Barrio^b, J. Corral^b, J. Iniesta^a, A. Miñano^b. ^aNeurology, Reina Sofia Hospital, Spain; ^bCentro Regional de Hemodonación, Servicio de Hematología y Oncología Médica, Hospital Morales Meseguer, Universidad de Murcia, Murcia, Spain

Despite control of thrombin is crucial in the development of atherothrombosis, antithrombin, the main endogenous inhibitor of

thrombin, has been classically considered to play a role only in venous thrombosis. Available methods to study antithrombin do not discriminate between the two main glycoforms present in plasma. We have developed a method able to specifically distinguish b-antithrombin (higher heparin affinity, predominant vascular localization and less abundant in plasma, 10%) from a-antithrombin. We used this novel method and the classical one that determines the whole antithrombin anticoagulant activity (a + b) in plasma of patients with ischemic cerebrovascular disease during the acute event and one year later. We recruited 117 consecutive patients (mean age 73 years, 52% males). Total levels of antithrombin did not significantly differ from that of a cohort of 97 healthy control subjects ($97.6 \pm 12.7\%$ vs. $99.9 \pm 6.4\%$, respectively $p = 0.105$) and did not significantly change one year after the event ($100 \pm 13.1\%$). Plasma b-antithrombin of patients during the acute event was significantly higher than the control cohort ($105.0 \pm 17.7\%$ vs. $97.1 \pm 12.2\%$, respectively; $p < 0.001$). We observed a significant decrease of b-antithrombin over time ($p = 0.04$), one year after the thrombotic event, levels reached values observed in controls. Results might be explained by the release of b-antithrombin from the vascular compartment to the circulation during the acute event.

Conclusions: Data sustain a role of antithrombin in stroke and show new knowledge on b-antithrombin, one potentially relevant glycoform that might be studied in thromboembolic diseases using the method that specifically detect this glycoform.

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Abstract – WCN 2013

No: 1529

Topic: 3 – Stroke

Risk factors for the progression of cognitive impairment after ischemic stroke

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Background: Vascular cognitive decline appears in 20–30% of ischemic stroke patients, being in the same time a risk factor for recurrent stroke and mortality. The arterial changes in the ageing brain are associated with hypoperfusion in periventricular regions, basal ganglia and hippocampus. The age, systemic hypotension episodes, low diastolic blood pressure and previous cognitive impairment are main risk factors for neurodegenerative and vascular dementia after ischemic stroke.

Objective: The objective of this study is to assess the risk factors associated with the progression of cognitive impairment in patients after ischemic stroke.

Patients and methods: We studied 56 patients (65 to 75 years old) with ischemic stroke, with and without previous cognitive impairment, admitted into the Neurological Clinic Timisoara between October 2012 and March 2013. All patients were clinically, metabolically (glycemic and lipidic profile), IRM and MMSE assessed.

The patients were followed up on 3 and 6 months after stroke onset. IRM performed showed small infarcts, cerebral atrophy, medial temporal atrophy and leukoaraiosis. Cognitive impairment was diagnosed according to the score of MMS test (mild >21 , moderate 10–20, severe <9).

Results: After 3 months: no cognitive decline 28 patients (50%); mild decline 15 patients (26.7%); moderate impairment, with previous cognitive decline, 10 patients (17.8%); and severe impairment–dementia, with previous cognitive decline, 3 patients (5.3%).

After 6 months: no cognitive decline 19 patients (33.9%); mild decline 18 patients (32.1%); moderate impairment 14 patients (25%); and severe impairment 5 patients (8.9%).

Conclusions: The risk factors of progression of cognitive impairment after 3 and 6 months from stroke onset are older age, previous cognitive decline, genetically predisposed to brain vessel damage,

insufficient control of vascular diseases and low systemic blood pressure (hypotension episodes).

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Abstract – WCN 2013

No: 1300

Topic: 3 – Stroke

Frequency of cerebral venous sinus thrombosis and predisposing factors in Sanandaj (west of Iran) 2010–2011

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Background and objectives: Venous sinus thrombosis is a rare cerebral vascular disorder with different geographical and age distributions. The prevalence of disease is 5 cases per million per year. Diversity among different regions, has been reported. This study aimed to evaluate the prevalence of cerebral venous thrombosis, symptoms and risk factors in Sanandaj (west of Iran).

Patients and methods: This study is a descriptive study including all patients with cerebral venous thrombosis during the years 2011 until 2012 that have been admitted to Tohid Hospital in Sanandaj. The data were gathered from the medical files and it was analyzed using SPSS software and descriptive statistics.

Results: Of all 30 patients, 24 patients (80%) were female. The mean of age was 36.2 ± 13.7 years. Headache was a common symptom that it was seen in 26 patients (86.7%). Common sign was left hemiparesis in 10 patients (33.3%) and papilledema in 9 (30%). The most common risk factor was OCP using. Upper sagittal and right transverse sinus were involved in 25 patients (83.3%) and 9 patients (30%), respectively. The most common involved brain hemisphere was the right side. One patient was died.

Conclusion: Cerebral venous sinus thrombosis with high prevalence in women than men can be a sign of a strong association with disease risk factors such as OCP, pregnancy and the puerperium. Also involvement in the right brain hemisphere was common but it did not had a logical reason and it needs more studies.

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Abstract – WCN 2013

No: 1524

Topic: 3 – Stroke

Free radical formation in patients with cerebrovascular accidents and comorbid cardiovascular pathology

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Background: Cerebrovascular accidents (CA) are associated with an increase of free radical production, which leads to oxidative stress and contributes to brain damage. Comorbid cardiovascular diseases (CVD) aggravate the condition.

Objective: To evaluate the disturbances in free radical balance and their prognostic value in patients with CA and comorbid CVD.

Patients and methods: 141 patients with CA (Male – 72 (51.1%), mean age 65.48 ± 13.44 years) and history of CVD (coronary artery disease (CAD), myocardial infarction (MI), atrial fibrillation (AF)). Free radical formation was assessed in plasma based on oxidative (chemiluminescence intensity index – basal (CIb) and zymosane-stimulated (CIIs)) and lipid peroxidation markers (anti-peroxide plasma activity (APA), malondialdehyde (MDA)).

Results: In patients with CA and CAD CIIB was 1.97-fold lower (64.24 ± 7.18 vs. 126.5 ± 17.19 mV/s * 106 leu) and CIIs 1.55-fold higher (1106.71 ± 107.34 vs. 716.21 ± 93.37 mV/s * 106 leu) compared to patients with no CAD ($p < 0.05$). In MI patients APA is 1.41-fold (2.92 ± 0.24 vs. 3.54 ± 0.13) higher than in patients with no MI ($p < 0.05$). We observed a marked increase in free radical formation on admission as demonstrated by CIIB 1.23-fold (64.75 ± 11.94 vs. 94.31 ± 10.00 mV/s * 106 leu) in patients with AF compared to patients with CA only ($p < 0.05$).

Conclusion: In patients with CA and CVD there is an increased free radical formation even in the first 24 h, which justifies adjunctive antioxidant therapy.

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Abstract – WCN 2013

No: 1538

Topic: 3 – Stroke

Stroke awareness in Munich and Moscow

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Background: Different data of public stroke awareness in several researches may result from different methods of investigations.

Objectives: We aimed to compare the stroke awareness among patients in Moscow and Munich.

Material and methods: 180 patients in Moscow (48% female, mean age 63.09 ± 13.83) and 180 patients in Munich (47.8% female, mean age 61.12 ± 14) hospitals, divided into three groups (after stroke, high risk of stroke and without risk of stroke) answered 33 questions, including 4 open-ended questions about stroke symptoms (SS) and risk factors (RF).

Results: In group of patients without risk of stroke the difference in number of SS named was not significant. In group of patients with high risk of stroke the awareness of SS (1.96 ± 1.28 in Munich and 1.03 ± 1.07 in Moscow, $p = 0.000$) and the awareness of stroke RF (2.25 ± 1.29 in Munich and 1.34 ± 1.2 in Moscow, $p = 0.000$) were better among patients in Munich. In group of patients with stroke in anamnesis the awareness of SS (2.08 ± 0.99 in Munich and 1.25 ± 0.89 in Moscow, $p = 0.000$) was also better among patients in Munich. The difference in awareness of stroke RF in this group was not significant. The direct dialogue with the doctor and specialized brochure was named as most preferable forms of increasing the knowledge of risk factors and stroke symptoms.

Conclusion: The educational activities in Moscow should be focused on patients with high risk of stroke and should increase the recognition of stroke onset and highlight the importance of early hospitalization.

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Abstract – WCN 2013

No: 1550

Topic: 3 – Stroke

Arterial CNS involvement in Behcet's disease

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Background and objective: Although Behcet disease (BD) tends to involve vascular structures, the incidence of arterial involvement is

far behind that of venous involvement. In this study, we evaluated all the cases with BD and arterial CNS involvement.

Methods: All the files of Neuro-Behcet outpatient clinic of Istanbul Medical Faculty were retrospectively evaluated. Demographical findings, clinical characteristics and radiological findings were evaluated.

Results: Between 1984 and 2011 there were 18 cases with BD and cranial arterial involvement out of 400 patients with neurological involvement. 4 were female and 14 were male (M:F = 3.5). Their age at arterial CNS involvement ranged between 25 and 64 years (median: 42; 42.5 ± 11.7). 15 had presented with acute hemiparesis/hemiplegia, accompanied by aphasia in two, ataxia in one, crossed brainstem syndrome in two patients. One patient had an asymptomatic intracranial aneurysm. One patient presented with seizures due to ACA infarct, and one patient presented with multiple cranial neuropathies due to external carotid aneurysm. One of the female patients had an underlying rheumatic mitral stenosis, and 3 patients were over age 50 which may suggest that stroke may not be directly associated to BD in those 4 cases. In 2 patients vasculitic involvement could be shown on angiography. In the remaining patients no other etiologies were found. **Conclusions:** Arterial CNS involvement is rarely seen in BD. A minority of those cases could be shown to have vasculitis. Other etiological factors unrelated to BD should be sought exclusively in such cases before attributing the stroke to BD.

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Abstract – WCN 2013

No: 1572

Topic: 3 – Stroke

Cancer patients are at increased risk of recurrent stroke and cardiovascular mortality

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Background: Cancer patients are at increased risk of cardiovascular and cerebrovascular events. However, the risk of recurrent stroke or cardiovascular mortality amongst cancer patients with stroke is currently unknown.

Objective: To determine the risk of recurrent stroke and cardiovascular mortality in cancer patients with ischemic stroke.

Patients and methods: This is a single center, observational study comparing the clinical characteristics and outcome of 58 ischemic stroke patients with cancer to 1047 ischemic stroke patients without cancer recruited from 2004 to 2008. Mean follow-up period was 76 ± 18 months. Primary endpoint was recurrent stroke and secondary endpoint was cardiovascular mortality.

Results: The three most common malignancies were breast (17%), colorectal (14%) and nasopharynx (13%). 26% of cancer subjects had active malignancy and 7% had metastatic disease. 22 patients with cancer (38%, 13.94 per 100 patient-years) and 219 patients without cancer (21%, 4.65 per 100 patient-years) developed a recurrent stroke ($p < 0.01$). 13 patients with cancer (22%, 4.3 per 100 patient-years) and 143 patients without cancer (14%, 2.35 per 100 patient-years, $p = 0.08$) died due to cardiovascular causes. After adjusting for age, gender, cardiovascular risk factors and co-morbidities, cancer was an independent predictor for recurrent stroke (HR 2.68; 95% CI: 1.58 to 4.54, $p < 0.01$) and cardiovascular mortality (HR 2.17; 95% CI: 1.12 to 4.22, $p = 0.02$).

Conclusion: Stroke patients with underlying cancer are at increased risk of developing recurrent stroke and cardiovascular mortality.

doi:10.1016/j.jns.2013.07.816

Abstract – WCN 2013**No: 1574****Topic: 3 – Stroke****Application of chemical exchange saturation transfer (CEST) MRI in acute stroke to visualize tissue acidosis and infarction risk**

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Background: In stroke patients with uncertain symptom onset or patients beyond the current time window for ivTPA there is a need for better imaging modalities to guide clinical decision making. Currently, MRI (perfusion weighted imaging) can be used to assess acute cerebral hypoperfusion. However, the technique is invasive, limited to some patients, and distinguishing penumbra from benign oligemia remains difficult. Amide Proton Transfer (APT) MRI is a non-invasive technique sensitive to the exchange of protons between amide-groups and water. The exchange rate is base-catalyzed and thus pH-sensitive.

Objective: To investigate the potential for APT imaging, which is sensitive to lactate formation and reduced pH in tissue at risk for infarction, in human acute stroke patients.

Patients and methods: 10 patients with acute stroke were scanned on a Phillips 3T system using our standard acute MRI protocol (including DWI and PWI) as well as a 3-minute APT-sequence. Patients were offered a follow-up scan and a clinical evaluation one month post-stroke.

Results: We found a large variation in APT effect between patients. However, in patients with penumbra, based on DWI/PWI mismatch, we found a lower APT effect compared to the ischemic core pointing to early lowering of pH in the penumbra, and possibly changes in protein and water content in the ischemic core.

Conclusions: These first results of APT in human acute stroke patients show that the technique could serve as a tool to show early tissue acidosis, and thus guide clinical decision making in stroke care.

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Abstract – WCN 2013**No: 1575****Topic: 3 – Stroke****Clinical factors related to severity of post stroke dementia (A retrospective review from 50 geriatric hospitals in Korea)**

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Background and purpose: The results of clinical studies about the severity of post-stroke dementia patients are limited in Korea. We investigate clinical factors related to severity of dementia and to inspect the clinical factors related to the progression of dementia severity.

Methods: The patients who visited the hospital by first time between March 2010 and February 2012, among the patients with post-stroke dementia admitted to 50 geriatric hospitals spread all over Korea, formed the analysis cohorts. Retrospective review of medical records was performed.

Results: A total of 2965 patients were included. The average duration of illness was 24.61 ± 28.18 months. By the severity of illness, mild cases were 1032 patients (34.81%), moderate 1278 (43.10%), severe 655 (22.09%), and mean score of MMSE was 14.82 ± 6.24 . The severity of dementia is higher in patients with overweight by 3.10 times

($p = 0.017$) existence of inmate by 5.92 times ($p = 0.0002$), past history of aphasia symptom by 0.18 times ($p = 0.0004$). Among the clinical factors related to the progression of dementia severity, female patients showed longer duration of illness by 2.89 times compared with average, by the results of univariate analysis of 120 severe dementia patients.

Conclusions: Among the clinical factors related to severity of post-stroke dementia in inpatients of 50 geriatric hospitals in Korea, severity of dementia is higher in patients with overweight, existence of inmate, and past history of aphasia symptom. The progression speed of dementia is suggested to be slow in female, regarding longer duration of illness in severe dementia patients.

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Abstract – WCN 2013**No: 1557****Topic: 3 – Stroke****Clinical and radiologic characteristics of RCVS (Reversible Cerebral Vasoconstriction Syndromes) in 23 patients**

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Background: Reversible cerebral vasoconstriction syndrome (RCVS) is a clinicoradiologic syndrome believed to be benign conventionally, however it is pointed out that cerebrovascular events including infarction and hemorrhage are not rare, suspecting that this syndrome is not necessarily in good prognosis.

Subject and method: We have experienced 23 patients of RCVS corresponding to the diagnostic criteria from April, 1988 to December, 2012. The background disease included 17 obstetrical disease (eclampsia/preeclampsia/PCA), posttransfusion 1 case, idiopathic 4 cases and nephrotic syndrome 1 case. We examined cranial CT/MRI and MRA in all 23 cases, cerebral angiography in 8 cases in the acute phase and followed up clinical and radiological findings up to 3 months.

Results: Headaches was recognized in 18 cases convulsion in 16 cases. Radiological examination showed posterior reversible encephalopathy syndrome (PRES in 20 cases, cerebral infarction in 5, subcortical subarachnoid hemorrhage in 2, cerebral hemorrhage and subdural hematomas in 1 case). mRS at 3 months later was 0–1 in 22 cases, but one case was 3. We recognized asymptomatic irreversible change in MRI in six cases at 3 months later.

Discussion: RCVS has various symptoms and background disease in addition to a headache. Our patients had larger portions of gynecological related RCVS patients. This may reflect higher incidence of PRES in acute phase. Hemorrhagic complications were not rare. Although outcome was favorable in most of our cases, some cases showed irreversible lesion in MRI images, suggesting that reversibility was not always true in radiologic images.

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Abstract – WCN 2013**No: 1601****Topic: 3 – Stroke****Is anger a precipitating risk for stroke?**

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Background: Emotional well-being is closely related to health. A lot of medical illnesses are closely linked to disrupted mental health. Men with introvert personality are particularly at high risk of ischaemic stroke.

Hypothesis/objective: Suppressed anger within the past 2 weeks is a risk for stroke. Patients with introvert personality are more likely to develop ischaemic stroke, whereas those with bad temper and recent rage outburst tend to present with intracranial haemorrhage.

Patients and methods: Patients admitted to the ward with stroke (both ischaemic and haemorrhagic) were interviewed about whether they have suppressed anger over the past 2 weeks.

Results: More than 50% of strokes admitted that they have suppressed anger or upset within the past 2 weeks. The suppressed anger intensified to maximum just 1–2 weeks prior to onset of the disease. In cryptogenic young stroke group who has no other medical risks, suppressed anger was clearly identified.

Conclusion: Our findings suggest that suppressed anger is a clear risk for stroke. We hope that the public will become aware these finding. In order to prevent recurrent stroke, it is advisable not to suppress the anger and instead try to channel out the suppressed anger.

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Abstract – WCN 2013

No: 1559

Topic: 3 – Stroke

Hyperhomocysteinemia and hyperlactatemia in lacunar ischemic stroke in young adults

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Background: The recent studies suggested a possible effect of high levels of homocysteine (Hcy) on the redox potential of the cells, which may be important in the formation of ischemic penumbra.

Objective: To determine the possible correlation relationship between Hcy and indicators of the lactate-pyruvate metabolism in acute lacunar ischemic stroke (IS) in young adults.

Materials and methods: The study included 30 patients aged from 32 to 45 years with lacunar IS in the acute phase. The risk factors had been presented by hypertension, hypercholesterolemia, and metabolic syndrome. The diagnosis of lacunar stroke had been confirmed by typical clinical symptoms, MR imaging, as well as the lack of data in favor of cardiac or another source embolism. In addition to conventional methods, the survey included an analysis of Hcy, lactate and pyruvate.

Results: 21 patients (70%) with lacunar IS had elevated Hcy up to 14.12 ± 1.18 mkmol/L, lactate up to 5.2 ± 1.34 mmol/L, and a slight increase in pyruvate up to 0.16 ± 0.05 mmol/L. There was a statistically significant correlation relationship between Hcy and lactate levels ($r = 0.37$, $p < 0.05$). There was no correlation between Hcy and pyruvate.

Conclusions: This research revealed statistically significant correlation between homocysteine and lactate levels in the lacunar stroke in young adults. Despite the controversies in the literature, hyperhomocysteinemia may have its role in the developing of the lacunar stroke, especially in young adults.

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Abstract – WCN 2013

No: 1586

Topic: 3 – Stroke

Trends in oral anticoagulation for acute ischemic stroke prevention in patients with preexisting atrial fibrillation

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Background: Effective anticoagulation with vitamin K antagonists (VKA) is the standard treatment for stroke prevention in the majority of patients with atrial fibrillation (AF). The proper anticoagulation in AF patients is still problematic and many of them receive suboptimal treatment. Hence, further efforts are necessary in this field.

Objective: Our aim was to evaluate the changes in the use of VKA for ischemic stroke prevention in patients with preexisting AF over the past 15 years.

Methods: We analyzed consecutive patients admitted to our center due to acute ischemic stroke between 1995 and 2011. Data regarding the use of VKA were analyzed. We distinguished between periods: 1995–2000, 2001–2005 and 2006–2011.

Results: In the analyzed time periods prestroke history of AF was reported with similar frequency (years 1995–2000: 25.2%; years 2001–2005: 24.3%; years 2006–2011: 24.7%). The proportion of patients with AF using VKA before stroke was significantly ($p < 0.001$) increasing (8.5%, 14.6%, 27.4%), as well as the proportion of AF patients using antiplatelets (37.6%, 47.3%, 47.2%). However, the proportion of AF patients using VKA with INR 2–3 has decreased (29.4%, 11.9%, 9.6%). It explains why the number of strokes due to AF did not change during the observation period. During hospital stay AF was diagnosed in additional 2.9%, 4.9% and 7.0% of patients.

Conclusions: The number of acute stroke patients with AF treated with VKA before the stroke onset is systematically growing. However, high ratio of patients receiving suboptimal doses suggests the constantly low guideline adherence among physicians and/or patient noncompliance.

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Abstract – WCN 2013

No: 1579

Topic: 3 – Stroke

A case of infant with ischemic stroke after varicella infection

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Stroke due to central nervous system vasculopathy is a recognized as rare complication of varicella-zoster virus infections. We describe a case of an 18-month-old previously healthy infant presented with partial seizures and left faciobrachial paresis 2 months after varicella infection. Magnetic resonance imaging of the brain showed acute and subacute ischemic lesions in the right cerebral hemisphere and in the basal ganglia. VZV-DNA was detected by PCR analysis in the CSF and blood, the patient was treated with intravenous acyclovir followed by oral treatment for 4 weeks and low doses of corticosteroids.

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Abstract – WCN 2013**No: 1602****Topic: 3 – Stroke****Stroke campaign: An experience report**

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Background: Cerebrovascular accident (CVA) is the second cause of mortality in Brazil and third in the world. Data from the World Health Organization (WHO) predict a 1.4% increase in stroke mortality in men and 0.3% in females by 2030. It is possible to reduce stroke mortality and disability through decreasing incidence and lethality of the disease and that is why stroke campaigns are indispensable.

Objectives: Experience report on campaign to promote health and prevent stroke.

Material and methods: In the campaign about stroke, medical students teach about the FAST test to recognize earlier the symptoms. The FAST test consists of “F”, Face (to smile), “A”, arm (any arm drifts downward?), “S”, Speech (speech sound slurred or strange?) and “T”, Time (if any of signs are observed) to call emergency immediately. Blood pressure measurements were also made to identify the main risk factor for hemorrhagic stroke – hypertension.

Results: People were instructed to the prevention of stroke, following stroke prevention, guidelines make aware of blood pressure, smoking, control alcohol abuse, check cholesterol levels, control diabetes, manage exercise and diet, and act FAST test at the first warning sign of stroke. The population empowerment from campaigns like this can reduce the interval between symptoms and diagnosis, reducing stroke morbidity and mortality.

Conclusion: Objectives were achieved and from primary attention to harm reduction, health promotion was entirely contemplated. It is a health promotion experience to be perpetuated.

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Abstract – WCN 2013**No: 1701****Topic: 3 – Stroke****Adult on set moyama like atherosclerotic disease with spontaneous revascularization**

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Background: Moya moya disease is characterized by chronic occlusion of internal carotid arteries with development of fragile new vessels at the base of the brain with unknown etiology. It is often presented with ischemic vascular events in children and intraventricular or intraparenchymal hemorrhages in adults. Adult onset moya moya disease must be differentiated from atherosclerotic disease. We presented an adult onset moya moya like atherosclerotic disease with spontaneous external revascularization.

Case presentation: 57 year old female patient was admitted in emergency clinic with sudden loss of consciousness and vomiting. Her systemic examination revealed mild hypertension. Neurological examination revealed a drowsy patient with stiff neck, and no cranial nerve dysfunction was found. She had no motor and sensory lateralizing deficit. Bilateral extensor planter reflexes and brisk tendon reflexes were found. She had normal blood count and screening tests. Computerized tomography revealed intraventricular hemorrhage

filling all ventricles. She gained her consciousness in the next day and informed that she had had another intracerebral eight years ago with full recovery. Cranial angiography revealed bilateral occlusion of carotid arteries at the level of supraclinoid segment and stenosis of right internal carotid artery after bifurcation from common carotid artery and moya moya vessels at the base of the skull. There was also anastomosis of right external carotid and right middle cerebral artery at the dural level convexity of the right hemisphere.

Conclusion: This case demonstrates that spontaneous revascularization of dural anastomosis may be a part of the moya moya like syndrome due to atherosclerosis.

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Abstract – WCN 2013**No: 991****Topic: 3 – Stroke****Isolated oculomotor nerve palsy: A rare manifestation of spontaneous internal carotid artery dissection**

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Introduction: Internal carotid artery (ICA) dissection commonly presents with headache, neck pain, Horner syndrome or with focal cerebral ischemic symptoms. Cranial nerve palsies can also be found, accounting for 12% of spontaneous ICA dissection cases and usually presenting as a syndrome of lower cranial nerve palsies. Therefore, an isolated oculomotor nerve involvement is extremely rare.

Case report: A 58 year-old man complains of a sudden-onset double vision for 3 days. He denies recent inflammation/infection, headache, cervical pain and trauma. He had well-controlled diabetes, hypertension and dyslipidemia. He was afebrile, without ocular erythema, proptosis or palpable orbital mass. Neurologic examination: left-sided ptosis and binocular horizontal diplopia in dextroversion without apparent extraocular-muscle paralysis or pupillary involvement. Other cranial pairs, motor, sensory and coordination systems were spared. The laboratory screening was negative for infections and immunologic diseases. The brain-MRI revealed a crescent-shaped mural hyperintensity in left ICA at the skull base extending to intra-petrous segment, implying reduced caliber/flow on MRI-angiography – left ICA dissection. The patient started antiaggregation therapy and progressively recovered. A year later he was asymptomatic and CT-angiography confirmed ICA recanalization.

Discussion: An ICA dissection presented only by an incomplete oculomotor nerve palsy is extremely rare. According to the literature, this can be explained by a blood supply impairment to *vasa nervorum* (embolic/hemodynamic) as the distal III is supplied by the inferior cavernous sinus and meningo-hypophyseal arteries, both derived from the intracavernous ICA. This case report enhances the importance of considering a cervicocerebral dissection in less typical clinical presentations.

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Abstract – WCN 2013**No: 1694****Topic: 3 – Stroke****Medullary infarcts: Analysis of 67 patients**

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Objective: To investigate the localization, etiology, symptomatology and prognosis of medullar infarcts.

Background: Vascular supply of medulla arises from anterior spinal artery, posterior inferior cerebellar artery and from direct penetrant arteries of the vertebral artery. Occlusion of these arteries either singular or in combination leads to different clinical symptoms and syndromes. Analyzing the relation of the involved arteries to the clinical symptoms may lead to better understanding of ischemia of medulla oblongata.

Patients and methods: 67 patients were identified with medullary infarct according to their clinical symptoms and definite ischemic lesions due to diffusion-weighted magnetic resonance imaging (DWI). Location of the infarct was also classified as according to upper middle or lower part of medulla oblongata besides lateral medullar, medial medullar or combination of lateral and medial territories. All patients underwent routine blood tests, etiologic investigations for arteriosclerosis and vasculitis if necessary. Doppler ultrasonography of vertebral and carotid arteries, echocardiography, MR angiography, and if necessary digital subtraction angiography were done.

Results: There were 41 male, 26 female patients of whom 32 had hypertension, 22 had diabetes mellitus, 20 with atrial fibrillation, 45 with smoking history and 15 had hypercholesterolemia. Some patients have more than one risk factor. 55 patients had lateral medullary syndrome, 10 patients had medial medullary syndrome and 2 patients had combined syndrome. All patients had a clear correlation between topographic localization and clinical signs. There was no death among the patients.

Conclusion: Anatomical localization is the leading factor in medullary infarcts and detailed examination and investigation is needed.

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Abstract – WCN 2013

No: 1612

Topic: 3 – Stroke

Epidemiological analysis of stroke in Goiás and in Brazil from 2008 to 2012

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Introduction: Cerebral Vascular Accident (CVA) is a leading cause of physical disability, hospitalization and death, creating a high impact on quality of patients' lives and their families.

Objective: To compare epidemiological data of individuals affected by stroke in Goiás and Brazil, considering gender and annual fluctuations between 2008 and 2012.

Methods: We obtained epidemiological data of Goiás and Brazil in the Unified Health System Database (DATASUS) and literature data for comparison criteria.

Results: We observed an increase in Brazil in cases of stroke in all analyzed years, being the highest around 20%, from 2008 to 2009. In Goiás, there was a reduction of approximately 16% in the number of cases from 2008 to 2009, followed by an increase of approximately 19% and 17% in the next two years, with a further decline of approximately 8% from 2011 to 2012. In males, the number of cases was always superior to females in each year, both in Brazil and Goiás.

Conclusion: With the increase in incidence of risk factors such as hypertension and diabetes, an increase in the number of strokes was expected, as seen in Brazil. But we did not observe such pattern in the state of Goiás. Literature predicts that in males the incidence of stroke is about 40% higher. However, the major increase observed in Brazil was about 7%, in the year 2012. In Goiás, there was a maximum raise of approximately 17% in 2008.

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Abstract – WCN 2013

No: 1687

Topic: 3 – Stroke

Stroke in the very elderly patients in Albania

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Aim: To evaluate vascular risk factors, stroke subtypes and clinical outcomes in very old patients admitted at our hospital with acute stroke.

Methods: We included in this study 252 stroke patients admitted to 4th Ward in the Clinic of Neurology in University Hospital Centre "Mother Teresa", Tirana, Albania from February 2011 to December 2012. All of them were male. We assess if there were any significant differences between patients under 80 years old compared with older patients (80 years or older), with regard to vascular risk factors, stroke type and clinical outcome.

Results: Of 252 patients, 189 patients were less than 80 years, 63 were 80 years old and older.

Younger patients were more likely to have diabetes (30, 6% versus 16, 2%, $p < 0.001$).

Older patients were more likely to have ischemic heart disease (38% versus 30, 1% $p = 0.02$), or atrial fibrillation (34, 5% versus 15, 2%, $p < 0.001$).

Older patients were more likely to have Total Anterior Circulation Infarcts (TACI) strokes (17, 6% versus 11, 1%, $p < 0.009$) or Partial Anterior Circulation Infarct strokes (PACI) (30, 1% versus 23, 5%, $p = 0.04$).

Outcome data, which was available for 91% patients, showed that older patients stayed longer in the hospital (median length of stay 23 days versus 18 days ($p = 0.008$)) and had a higher inpatient mortality (14% versus 3, 7% ($p < 0.001$)).

Conclusion: Very elderly patients have a different risk factor profile, have more anterior circulation infarcts and have a worse prognosis with increased mortality and increased length of stay in the hospital.

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Abstract – WCN 2013

No: 1675

Topic: 3 – Stroke

Variation of some oxidative stress and inflammatory markers in acute ischemic stroke

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Background: The role of inflammation in atherogenesis and appearance of the tissue lesions in ischemic stroke are demonstrated and more recently there are strong indirect evidences that oxidative stress, by free radical production, detains a major contribution in ischemic brain injury.

Objective: To assess the variation of some oxidative stress and inflammatory markers in the acute phase of ischemic stroke and to find if there is any correlation between them or with the clinical status evaluated by NIHSS scale.

Patients and methods: In a prospective study, we evaluated levels of oxidative stress markers – uric acid, albumin, copper and Total Antioxidant Status (TAS) and of two inflammatory markers – CRP (C Reactive Protein) and fibrinogen – in 55 patients with acute ischemic stroke compared with 19 controls.

Results: We obtained significant ($p < 0.001$) lower values for albumin, copper, TAS, and significant ($p < 0.001$) higher levels for

CRP in the patients with ischemic stroke compared with controls. We didn't find a direct correlation between neurological status and oxidative stress markers, instead NIHSS score correlated significantly with CRP levels at the onset of stroke.

Conclusion: These results prove once more the implication of oxidative stress and inflammation in the pathogenesis of ischemic stroke.

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Abstract – WCN 2013

No: 1665

Topic: 3 – Stroke

Management of multiple intracerebral hemorrhages due to myxomatous aneurysms – A case report and literature review

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Background: The natural history of cerebral aneurysms which derived from metastatic spread of cardiac myxomas is not well known and their management may prove very difficult.

Objective: Description of a patient's history with multiple intracerebral hemorrhages due to myxomatous aneurysms and progression who underwent whole brain radiotherapy.

Patients and methods: Case report and literature review.

Results: A 35-year-old woman presented with headache due to multiple intraparenchymal hemorrhages five months after resection of an atrial myxoma. Angiography showed several small fusiform aneurysms. During the following four months the patient developed a worsening of her neurological deficits correlating with a progression of the amount as well as the size of the hemorrhages. Since undergoing a whole brain radiotherapy with 30 Gy/à 2 Gy the patient has been clinically and radiologically stable over a 15 month period up to now.

Conclusions: Myxomatous cerebral aneurysms are important entities for neurologists to recognize which can present before, but also months to years after diagnosis of a cardiac myxoma. So far there are only a few case reports about the management of intraparenchymal hemorrhages of this etiology published: most of them recommend an observational procedure. In one case radiotherapy with 50 Gy and chemotherapy has been described.

To our knowledge this is the first report about a whole brain radiotherapy with 30 Gy/à 2 Gy in a patient with multiple intracerebral hemorrhages due to myxomatous aneurysms.

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Abstract – WCN 2013

No: 1681

Topic: 3 – Stroke

Efficacy of carotid endarterectomy performed by urgent indications

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Background and purpose: Carotid endarterectomy (CEAE) is a standard method of primary or secondary ischemic stroke (IS) prevention. In our days there is no common consensus concerning the period when to perform CEAE. Majority of authors consider that CEAE should be performed after acute period of IS. But some authors' opinion is to perform CEAE to patients who need it by urgent indications. We estimate efficacy of CEAE by urgent indication in acute period of stroke.

Methods: 54 patients with acute IS developed due to occlusive and stenotic lesion of carotid arteries were investigated. Males were 45 (83.3%), females were 9 (16.8%). Average age was 60.7 ± 2.6 years. The period from the IS sign appearance to CEAE operation performance ranged from 2 h to 18 days. CEAE was executed when hemodynamically significant stenosis of carotid arteries with embologically dangerous atherosclerotic plaque occurred. Indications for CEAE were minor stroke – 36 patients (66.6%), transient ischemic attacks (TIA) – 4 patients (7.4%), stroke progression – 11 patients (20.4%) and floating embolus – 3 patients (5.6%). In 37 patients (68.5%) classic CEAE and in 17 patients (31.5%) eversion CEAE were performed.

Results: Regression of neurological deficiency in 2–3 days was observed in 48 (88.9%) patients in post operative period. Among these 48 patients full neurological recovery occurred in 36 (66.7%) patients. Obtained data testified about efficacy of CEAE in acute period of IS when urgent indications presented.

Conclusions: CEAE performed at the stage of reversible ischemic and neurological lesions (minor stroke and TIA) had better results.

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Abstract – WCN 2013

No: 1658

Topic: 3 – Stroke

Efficacy of intra-arterial thrombolysis with tissue plasminogen activator in acute ischemic stroke. Uzbekistan results

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Background and purpose: Intravenous tissue plasminogen activator (tPA) application within 3 h after onset of acute ischemic stroke (AIS) is established therapy. The alternative for thrombolysis in patient admitted within 3–6 h after onset is intra-arterial thrombolysis (IAT). The purpose was to evaluate the efficacy of IAT with tPA in AIS due to middle cerebral artery (MCA) occlusion.

Methods: Cerebral angiography (CA) and standard Seldinger technique were applied in IAT. After angiographic detection of MCA occlusion IAT with tPA in dose 15–25 mg was performed. 10 patients (7 male, 3 female, aged 48–75 years, average age 64.4 ± 8.6) were investigated. All patients were selected by the National Institute of Neurological Disorders and Stroke recommendations. National Institutes of Health Stroke Scale (NIHSS) and modified Rankin scale (mRs) were used.

Results: MCA occlusion was detected by CA in all patients. NIHSS average score at admission was 14.7 ± 3.9, 272 ± 36.5 min elapsed from symptom onset. We obtained the following clinical results: 5 patients (50%) had excellent outcome (mRs score 0 to 1), 3 patients (30%) a good outcome (mRs score 2), and 2 patients (20%) had no changes (mRs score 3 to 4). In this patient brain ischemic zone was detected on control CT. In the group with excellent and good outcomes 3 patients had asymptomatic hemorrhagic transformation (HT) with small petechiae along the margins of infarct zone.

Conclusions: IATT with tPA performed under CA control can be considered as an effective method in patients with AIS. Thrombolysis-related HT is a marker of successful recanalization.

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Abstract – WCN 2013

No: 1664

Topic: 3 – Stroke

Mortality trends by stroke in Uruguay

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Background: Stroke is one of the major death causes in Uruguay and in the world.

Objective: For this motive, we have designed this study to describe and to analyze stroke mortality tendency in Uruguay between 1950 and 2008. Its relationship with gender, imaging techniques, intensive treatments, gross domestic product (GDP), and mortality in other countries was analyzed.

Methods: An ecologic study of mortality trends, standardized gender and age rates, and Poisson regression models and autocorrelation was performed.

Results: The mortality trend has decreased throughout the studied time, with two periods, an increasing one until 1971, and then a decreasing period until 2008. The female gender presented higher mortality. The decreasing period coincides with the introduction of intensive care units (ICU) and the computerized tomography (CAT) scan. The GDP showed a negative correlation with the decrease of stroke mortality.

Conclusions: Uruguay has had a decline in stroke mortality placing itself in an intermediate position in comparison to other countries. CAT scan techniques, ICU centers and an increase in the GDP seem to have had a positive effect in the decline of stroke mortality.

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Abstract – WCN 2013

No: 1651

Topic: 3 – Stroke

Multidisciplinary stroke early supported discharge program in a local hospital: Outcome and safety

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Background: Recent meta-analysis had demonstrated that early supported discharge is equally effective as compared with prolonged inpatient rehabilitation. Stroke Early Supported Discharge (SESD) Program was then implemented since Dec. 2011. Timely home based rehabilitation and careers' training would be provided to those patients eligible for early discharge.

Objective: This study aims to show whether our SESD program can be effective and safe outside clinical trial setting.

Method: This study reviewed the outcome and safety parameters after the introduction of the program. Data were prospectively collected and compared before and after the introduction of the program.

Results: From Dec. 2011 till Sept. 2012, 283 patients were recruited into the SESD program. Their mean age was 70. Half of the patients had Modified Barthel Index (MBI) less than 85. After the program was implemented, the proportion of acute stroke patients directly discharged without transfer to rehabilitation unit, was significantly increased from 43.5% to 49.2% ($p = 0.005$). The average length of stay of those patients was 5.08 days. For the functional recovery, the MBI and Berg Balance Scale (BSS) were significantly improved after home based rehabilitation as compared with their baseline upon discharge (mean MBI from 81.2 to 91.9, $p < 0.001$, mean BSS from 40.4 to 46, $p < 0.001$). Their unplanned readmission rate, within 4 weeks from discharge, was 10.2%, which was lower than that of all the stroke patients (12.3%) in the same period ($p = 0.055$).

Conclusions: The SESD program was effective and safe to facilitate direct discharge of acute stroke patients from acute hospital.

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Abstract – WCN 2013

No: 1636

Topic: 3 – Stroke

Predictive value of imbalance grade and F-score to identify stroke in dizziness patients

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Background: In patients presenting dizziness, there is no definite criterion to distinguish between the central and peripheral etiology based on their symptom quality. Although dizziness is usually considered to be benign etiologies, not infrequently acute dizziness cases could herald a cerebrovascular disease. But lack of objective risk stratification schemes of cerebrovascular disease makes clinicians perplexed. At this point, we aimed to assess the effectiveness about the cerebrovascular disease risk stratification systems such as hard coronary heart disease 10 year risk score in Framingham heart study (F-score) and specialized examinations such as imbalance grade.

Methods: We performed retrospective study presenting symptom with whirling/non-whirling type dizziness, vertigo or disequilibrium. Peripheral type vertigo was excluded. MRI was taken in all included patients to define ischemic lesions. ABCD2, CHADS2, F-Score, and imbalance grade (0 to III) were calculated using clinical information from the medical records. Statistical analysis was done with chi square and logistic regression.

Results: Among the 145 patients (mean age, 59.8 ± 10.3), ischemic stroke was diagnosed in 27 (18.6%). Univariate analysis revealed that F-score (13.6 ± 9.1 vs 7.4 ± 7.5 , $P = 0.001$), imbalance grade ($P = 0.014$), male (70.4% vs. 39.8%, $P = 0.004$), smoking (40.7% vs. 10.2%, $P = 0.000$) and low high density lipoprotein (HDL) cholesterol (47.7 ± 16.3 vs. 52 ± 13.8 , $p = 0.05$) were associated with stroke. In multivariable analysis, imbalance grade only ($p = 0.037$) was associated with stroke. Most common lesion in stroke patients with isolated dizziness was PICA territory.

Conclusion: Those patients with isolated dizziness, imbalance grade could be helpful to predict central origin.

So prompt evaluations were needed to define stroke to those patients with high imbalance grade score presenting with dizziness.

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Abstract – WCN 2013

No: 1451

Topic: 3 – Stroke

Isolated right hand palsy due to cortical ischaemia

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Case: 75 year old male patient with no known previous illness, noted a weakness on his right hand one day before his presentation to our polyclinic. He consulted the physical therapy and rehabilitation polyclinic and an ENMG study was ordered with a preliminary diagnosis of entrapment neuropathy. The ENMG was found to be normal, and a control ENMG was ordered 10 days later. The patient consulted medical advice from the neurology polyclinic 1 day after the beginning of his complaints. His neurological examination at the time of his application was as follows: Right hand fingers were in flexion posture, the motor strength of the opposition and flexion of the right hand first finger, abduction and adduction of the other fingers were 0/5. The strength of the right hand extension was 4/5. Hoffmann sign was (+) on the right side. The other neurological examination signs were normal. Cranial MR imaging revealed widespread bilateral periventricular chronic lacunar ischaemia as well as an acute ischaemic

infarct area in the sol cortical area corresponding to the motor area of the right hand. Ultrasonographic investigation showed 75% stenosis in the left carotid artery and 50% stenosis in right carotid artery. No ischaemic cardiac pathology was detected on cardiac examination.

Discussion: Isolated hand palsy due to cerebral ischaemic pathology is rarely seen and is usually confused with acute entrapment neuropathies. Usually cortical ischaemic lesion is seen and cerebral atherosclerosis is present in the etiology.

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Abstract – WCN 2013

No: 1570

Topic: 3 – Stroke

Hemorrhage into a gastrointestinal stromatumor (GIST) under Dabigatran at a patient with stroke caused by a paroxysmal atrial fibrillation

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Background: An 87 year old patient had a stroke with a hemiparesis left under aspirin. In their anamnesis she had an arterial hypertension and a benign gastrointestinal stromatumor. Should we treat the patient with new oral anticoagulation therapy or Warfarin? After information the patient agreed to therapy with Dabigatran.

Methods: The classical diagnostics in neurology and internal medicine.

Objectives: Sensomotoric hemiparesis left.

Results:

MRI brain: Several ischemic lesions in the region of right middle cerebral artery.

Holter EKG: Paroxysmal atrial fibrillation.

24 h blood pressure measure: Arterial hypertension I.

CT abdomen: Great hemorrhage into the GIST.

Gastroscopy and colonoscopy: No hemorrhage.

Laboratory 1: Regular finding.

Lab II: Hb 6.7 mg/dl, following leukocytosis of 12000 and CRP 30 mg/dl.

Conclusions: At patients with a stroke caused by atrial fibrillation and a benign tumor in the history the indication for therapy with Dabigatran should be done carefully or under monitoring.

3 days after starting the therapy with Dabigatran (2×110 mg/daily) the patient had a decrease of hemoglobin to 6.7 g/dl and a hemorrhage into the GIST, but not a cerebral hemorrhage. After finishing the therapy with Dabigatran the hemorrhage stopped slowly without a special coagulation management. A therapy with Imatinib, which should melt down the tumor, needed to be stopped because of side effects. A urosepsis, the next complication, faded away under a special antibiotics. After 6 weeks the patient left the hospital with a Barthel index of 60 points; the same as to the admission.

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Abstract – WCN 2013

No: 1565

Topic: 3 – Stroke

The prevalence of modified risk factors for stroke in working-age people, according to the screening of an open population

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Background: WHO stresses that up to 80% of all deaths due to heart diseases, stroke and diabetes can be prevented (Rosamond W. et al., 2008).

Aim: To evaluate the prevalence of the most studied modified risk factors for ischemic stroke in people of working age.

Materials and methods: Ulyanovsk population in the age of 40–59 years underwent screening. The “modifiable risk factors for stroke” (AHA, ASA) were determined during clinical examination of the patient.

Results: We surveyed 300 people: 86 (15.93%) men, 214(25.78%) women. Hypertension was diagnosed in 162 subjects (54.0%). In 24 cases (8.0%) the blood pressure above 140/90 mmHg was first found out at a doctor's office. Smoking in the present – 40 people (13.33%), in the past (gave up smoking more than 1 year ago) – 28 (9.33%). Men predominated over women in smoking: 40 (46.51%) males in comparison with 28 females (13.08%), $p < 0.05$. 4.3% of patients had diabetes II type. Cardiovascular diseases (angina pectoris, acute myocardial infarction in past history) are diagnosed in 10.33–22.67% (“definite” cases and “certain” + “possible” cases). 45.33% of patients suffered from hypodynamia. There were 86 (28.67%) obese patients in the study. Diagnosis of ciliary arrhythmia was pronounced in 4 cases. 9 observed women take oral contraceptives. According to the results of incremental discriminant analysis of the given data, the most significant risk factor for stroke was hypertension.

Conclusions: Among the identified modified risk factors the most prevalent in the working-age group are hypertension, cardiovascular diseases, hypodynamia and smoking in men.

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Abstract – WCN 2013

No: 1619

Topic: 3 – Stroke

Atherosclerotic carotid artery diseases and atrial fibrillation in ischemic stroke patients

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Introduction: Atrial fibrillation (AF) is the most common sustained cardiac arrhythmia, affecting 1–2% of the general population. The most common causes of AF are ischemic heart disease and atherosclerosis. The aim of this study is to determine the frequency of atherosclerotic cervical carotid disease with Doppler ultrasonography (USG) in patients with AF.

Material and methods: Ninety-three patients who had AF, treated with warfarin and being followed-up in the Cerebrovascular Diseases outpatient clinic were included to this prospective study. Seventy-one subjects were chosen as control group among warfarin treated patients with indications other than AF. Patients and controls were evaluated with carotid Doppler USG.

Results: The mean age of patients and controls were 71.5 and 64.2 years, respectively. Fifty of the 93 patients (54%) had atherosclerotic changes in carotid Doppler USG compared with 40 of 71 (56%) control patients ($p = 0.74$). Among the 93 patients who had atherosclerotic changes 27 had an increased intima-media thickness, 15 had atherosclerotic plaques causing <50% of lumen stenosis, 2 had 50–69% of lumen stenosis, 5 had >70% of lumen stenosis and 3 had internal carotid artery (ICA) occlusion. Proportion of patients who had greater than 50% stenosis or ICA occlusion was 11%.

Discussion: Carotid artery stenosis of 50% or more occurs in about 11% of ischemic stroke patients with AF, due to other risk factors like hypertension, diabetes mellitus and hyperlipidemia. This patient group must be followed-up carefully for the progression of carotid artery disease for the prevention of new stroke due to large artery disease.

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Abstract – WCN 2013**No: 1609****Topic: 3 – Stroke****Cognitive functioning in ischemic stroke patients with physical disability**

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Background: Stroke is a major cause of disability in the elderly. It is also related to a wide spectrum of cognitive deficits. Post-stroke cognitive impairment is particularly important and frequent. Whether there is relationship between cognitive functioning and physical disability in ischemic stroke patients is unclear.

Objective: To access the relationship between cognitive functioning and physical disability in ischemic stroke patients.

Patients and methods: We prospectively evaluated consecutive patients with acute ischemic stroke. 145 patients were administered by tests of Modified Rankin Scale (MRS), Mini-Mental State Examination (MMSE) and National Institute of Health Stroke Scale (NIHSS) and the Barthel Index (BI). We accessed the relationship between MRS and MMSE and compared the cognitive functioning with mild to moderate physical disability ($MRS \leq 3$) and severe physical disability ($MRS > 3$) in ischemic stroke patients across 4 weeks while observing the changes both in MMSE and NIHSS/BI.

Results: MMSE scores were significantly different among different degree of MRS over time with $p < 0.05$. The change of MMSE in $MRS \leq 3$ group was different from that in $MRS > 3$ group after 2 and 4 weeks ($p < 0.01$), the improvement of MMSE scores was 0.75 ± 3.78 in $MRS \leq 3$ group and 3.10 ± 5.68 in $MRS > 3$ group after 2 weeks ($p < 0.001$), and 0.58 ± 4.10 in $MRS \leq 3$ group and 3.71 ± 5.59 in $MRS > 3$ group after 4 weeks. The change of MMSE was linear for NIHSS or BI scores after 2 and 4 weeks.

Conclusion: Physical disability correlated with cognitive functioning in ischemic stroke patients. The improvement in cognitive functioning correlated with the degree of physical disability.

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Abstract – WCN 2013**No: 1721****Topic: 3 – Stroke****Ischemic stroke and migraine**

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Background: Migraine is recognised as a cardiovascular risk factor and connection with stroke especially migraine with aura.

Objectives: Relationship between migraine and stroke and compared to other types of headache.

Methods: The study included 135 patients with stroke and headache. After statistical adjustment for age, gender and affected vascular brain system, we formed two groups: 15 patients with stroke and migraine and second 17 patients with stroke and other types of headache. All of them had MRI. Welch criteria were applied for the relationship between migraine and stroke.

Results: I group: 15 pts, mean age 45 ± 11 years, 9 pts had migraine with aura and 6 pts without aura. The II group: 16 pts, mean age 53 ± 8 years, 12 pts tension type headache, 3 headache

attributed to HTA and one unclassified. There were no differences between groups on main vascular risk factors (blood pressure, cholesterol level and ischemic cardiopathy). According to Welch criteria 2 pts had stroke which occurs in patients with migraine but not during an attack, 9 pts had migraine induced stroke with risk factors. Migraine with aura produced stroke in the presence of another risk factor and 5 pts had uncertain history of migraine without aura and stroke during a migraine attack. The Rankin disability scale was significantly worse compared to stroke and other types of headache.

Conclusion: A patient with migraine had high probability to have severe stroke than other types of headache. Relationship between migraine and stroke is complex and difficult.

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Abstract – WCN 2013**No: 1733****Topic: 3 – Stroke****Insular and caudate lesions release abnormal yawning in stroke patients**

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Background: Abnormal yawning (chasm) is a rare phenomenon in patients in anterior circulation stroke.

Objective: We aimed at identifying core regions involved in abnormal yawning in an attempt to assign abnormal yawning to lesions in the anterior circulation.

Patients and methods: We analyzed the extent of brain areas encompassing restricted diffusion after ischemic stroke in ten patients presenting with at least three yawns per 15 min associated with stroke symptoms. All patients were classified according to the NIH stroke scale (NIHSS), daytime of symptom onset, yawning duration, Glasgow Coma Scale (GCS), partial oxygen pressure, body temperature, blood pressure, pulse, and modified Rankin scale. Lesion maps were segmented on diffusion weighted images in native space using MRICroN and spatially normalized using the unified segmentation algorithm in SPM5. The extent of overlap between the different stroke patterns was calculated.

Results: Abnormal yawning persisted for 2.8 ± 1.3 days. Average GCS and NIHSS scores were 12.8 ± 2.6 and 10.5 ± 6.5 , respectively. Oxygen saturation, body temperature, blood pressure, and heart rate were within normal limits. Ischemic brain lesions overlapped in eight out of ten patients: in six patients in the insula and in six in the caudate nucleus. The decrease of the apparent diffusion coefficient within the lesions correlated moderately with the duration of abnormal yawning (insula; $r^2 = 0.58$; caudate nucleus; $r^2 = 0.32$).

Conclusion: The intensity of targeted ischemic lesions in the insula and the caudate nucleus correlates with the duration of abnormal yawning in anterior circulation stroke.

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Abstract – WCN 2013**No: 1736****Topic: 3 – Stroke****Poststroke depression**

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Comorbidity of depression and stroke significantly reduces quality of patient's life after stroke. Squeal after stroke also determines quality of life and impact on the occurrence of depression after stroke. In our study we investigated occurrence of depression in patients after different types and subtypes of stroke measured by Hamilton scale compared to the level of disability on NIHSS scale.

The goal was to make comparative analysis of depression after stroke, according to gender and age, side of lesion and severity of neurological deficit.

Material for our work is 210 patients with stroke treated at the Neurology Clinic, Clinical Center of Sarajevo University in 2012, 105 male and 105 female. The mean age of patients was 67.12 with SD of 9.5 years. Ischemic stroke was present in 65% cases. There was no statistical significant difference between ischemic and hemorrhagic stroke among genders (chi-square = 6.563, $p = 0.082$). Depression was more prevalent among younger patients (52–60 g) with 39.2% then in the group of older patients (61–70 g) with 32% of depressed. In relation to gender there was significantly more patients with depression among women compared to men (63.8:27.2%) (chi-square = 14.38, $p = 0.00019$). Depression was more frequent in patients with stroke in the left hemisphere medial localization (63%). NIHSS scale average was 16.07 with the minimum of 11 and maximum of 22, $F = 52.56$, $p = 0.001$.

We can conclude that depression after stroke is more frequent in younger patients, female patients, patients with localized stroke in the medial left hemisphere and with higher disability score.

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Abstract – WCN 2013

No: 1695

Topic: 3 – Stroke

Is treatment with alteplase beneficial in elderly patients or patients with mild deficit?

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Background: Treatment with iv thrombolysis is not approved for patients with ischaemic stroke (IS) over 80 years or patients with mild deficit despite subgroup analyses from randomised studies and observational data pointing to a benefit for these patient groups.

Objective: To analyse efficacy of iv thrombolysis in matched-pairs of patients (with and without iv thrombolysis) over all age groups and the whole range of stroke severity in a large cohort of prospectively enrolled patients with ischemic stroke (Austrian Stroke Unit Registry).

Methods: Patients with and without therapy with iv tPA were matched for age, sex, the pre-stroke modified Rankin scale (mRS), stroke severity (according to the NIH stroke scale (NIHSS)), hypertension, diabetes, hypercholesterolemia, etiology and clinical stroke syndrome. The outcome was assessed by the mRS at 3 months. Means of outcome differences in matched pairs were used for interpretation. For data-visualization weighted averages of outcome differences were computed for all age-severity combinations along a grid and mapped to a colour.

Results: 60230 patients with IS 7615 were treated with iv tPA. Of those 25.3% were >80 years and 18.1% had a mild deficit (NIHSS ≤ 4). 2124 matched pairs were included into the analysis. Colour mapping showed a beneficial effect on outcome in thrombolized patients as compared to matched cases over all ages and the whole range of stroke severity. In patients with mild stroke effect sizes were smaller but still present.

Conclusion: Treatment with iv tPA might be beneficial in elderly patients and patients with mild deficit.

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Abstract – WCN 2013

No: 1732

Topic: 3 – Stroke

Clinical presentation of internal carotid artery dissection in series of 28 patients

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Background: Cervical artery dissection is defined by the existence of a hematoma in the arterial wall. Internal artery dissection (ICAD) is an important cause of stroke among young and middle-aged patients.

Objective: The aim was to analyze the spectrum of clinical presentation in 28 ICAD patients.

Patients and methods: 28 patients with ICAD, 26 with unilateral and 2 with bilateral, aged 35–59 (mean age 47.6) were evaluated in the last 12 year period. The ICAD diagnosis was established in all cases using MRI, MRA and duplex sonography.

Results: Facial and neck pain and Horner's syndrome were the only presenting symptoms in 5 patients; headache and visual disturbances in 2; facial pain, Horner's syndrome and contralateral sensorimotor deficit in 7; headache and contralateral sensorimotor deficit in 5; and contralateral sensorimotor deficit in 9. ICAD was triggered by mild trauma in 7 patients (1 while unloading sacks of corn, 2 following sudden head turning, 1 during sports activity, 1 during sexual intercourse, 1 during roller-coaster ride, and 1 in car accident), and spontaneous in 21. MRI revealed infarction in 21 patients, while in the 5 patients presenting with facial and neck pain and Horner's syndrome and in 2 patients with headache and visual disturbances MRI did not show evidence of infarction. Good outcome (defined as modified Rankin score of 0–2) was seen in 26 patients (92.85%).

Conclusion: The clinical presentation of ICAD is variable and can be similar to other stroke etiologies. ICAD is not always associated with brain infarction.

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Abstract – WCN 2013

No: 1707

Topic: 3 – Stroke

Cerebral venous thrombosis: Clinical, genetic and neuroradiological study

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Background: Cerebral Venous Thrombosis (CVT) is a rather uncommon cerebrovascular disease with a wide spectrum of clinical presentations and a variable prognosis.

Objective: To study the CVT clinical presentation and outcome.

Patients and methods: We investigated clinical and imaging features of twenty-three consecutive patients with CVT (17 F, 6 M; mean age 39.8 yrs) admitted to our department from August 2005 to February 2013. Diagnosis of CVT was confirmed by brain MRI and MR-venography. All patients underwent an extensive systematic etiological and genetic work-up at admission. Treatment was based on intravenous unfractionated heparin, followed by oral anticoagulation for six months or longer. Outcome was evaluated by modified Rankin Scale (mRS) at three months.

Results: Genetic risk factors were identified in twelve out of twenty patients. Patients presented with the following clinical syndromes: focal syndrome (12/23, 52.1%), isolated headache (8/23, 34.7%) and isolated intracranial hypertension (3/23, 13.0%). Headache was the most

frequent symptom, present in twenty out of twenty-three patients (86.9%). Other often occurring symptoms were seizures (9/23, 39.1%), motor deficits (7/23, 30.4%) and aphasia (6/23, 26.0%). The sinus most frequently involved was the lateral sinus (22/23, 95.6%), either isolated or in association with other sinuses. A complete recovery (mRS = 0–1) was observed in 16/23 (69.5%) cases. One patient died of heart failure.

Conclusion: Our results confirm that headache is the most frequent symptom and may be the only clinical presentation of the CVT in a high percentage of cases. CVT seems to have a more benign prognosis than previously considered.

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Abstract – WCN 2013

No: 1742

Topic: 3 – Stroke

Prediction of tissue outcome in acute stroke with perfusion CT

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Background: Perfusion CT allows assessment of tissue viability in acute cerebral ischemia.

Objective: To evaluate the outcome of ischemic tissue depending on the initial perfusion deficit.

Patients and methods: Thirty patients (21 men, median age 59 years) with acute hemispheric stroke in the first 24 h of symptoms onset underwent perfusion CT on admission and non-contrast CT on Day 10. Relative (intact/affected side) values of perfusion parameters within hypoperfused area on initial perfusion CT scan in the regions corresponding to hypodense and normal tissue zones on non-contrast CT were studied with ROC analysis. No patients were eligible for recanalization therapies, and received standard antiplatelet treatment and supportive care.

Results: Median initial NIHSS score was 8.1. Thresholds for relative values of perfusion parameters indicating tissue survival (i.e., no infarction on final CT scan) were 5.06 for rCBF, 1.13 for rCBV, and 0.41 for rMTT. Despite no difference was found between areas under ROC curves ($p > 0.05$), AUROC value was numerically higher for rCBV (0.84 vs 0.80 vs 0.59 compared with values for rCBF and rMTT, respectively). The rCBV threshold had sensitivity of 88.9% and specificity of 75.0%.

Conclusion: In patients with acute stroke who did not receive thrombolysis the viability of tissue can be best predicted with the change of CBV parameter relative to intact tissue on initial perfusion CT, with decrease of CBV > 13% indicating worse tissue outcome.

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Abstract – WCN 2013

No: 1792

Topic: 3 – Stroke

Specification of amino acid neurotransmitters in the cerebrospinal fluid in intracerebral hemorrhage

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Introduction: Current knowledge of the mechanisms of neuronal death in stroke shows that the damaged nerve cells play an important role in imbalanced neurotransmitter amino acids, set in motion a cascade of calcium glutamate.

The purpose of the study is to evaluate the content of excitatory (glutamate and aspartate) and inhibitory (GABA, glycine) neurotransmitter amino acids (NTA) in the cerebrospinal fluid (CSF) of patients with intracerebral hemorrhage (ICH), and give a comparative description with ischemic hemispheric stroke (IHS).

Materials and methods: The study included 75 patients: 40 with ICH, 20 with IHS and 15 dyscirculatory encephalopathy (DE) under sub-compensation (control group).

All patients with ischemic acute stroke had severe disease severity (according to clinical scales). Determining the level of excitatory and inhibitory NTA in the CSF was performed by liquid chromatography (Cohen's method).

Results: These studies have shown that in patients with ICH, the average content of glutamate was 402.0 ± 11.60 nmol/ml, aspartate – 345.0 ± 13.83 nmol/ml, glycine – 316.5 ± 10.4 nmol/ml and GABA – 100.3 ± 4.8 nmol/ml. In the early days of ICH, average content of excitatory amino acids increased dramatically, significantly surpassing both the control group (4.14 times), and patients with IHS (2.65 times).

Conclusions: Thus, the ICH and IHS accompanied by significant changes in the content of both excitatory and inhibitory amino acids in the acute phase of the disease.

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Abstract – WCN 2013

No: 1774

Topic: 3 – Stroke

The role of hemodynamic disturbance on extracranial carotid artery on the occurrence of brain aneurysm

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Introduction and purpose: An aneurysm occurs as a result of weakening of the wall. They may be congenital or acquired. The most common causes are atherosclerosis, hemodynamically induced damage of the vessel wall and vasculopathy.

The purpose of this study was to find a relationship between hemodynamic disturbance on extracranial carotid artery and occurrence of intracranial aneurysm in our patients.

Material and methods: We tested 52 patients with the diagnosis – Aneurysm cerebri non ruptum (confirmed by CTA, MRA or DSA), 44 (84.6%) women and 8 (15.4%) men, age 24–78 years (average 58.2 years). We used findings of Doppler sonography to evaluate blood flow in the cervical carotid arteries and compare with the presence of an aneurysm at the side with hemodynamic disturbance (HD) with high flow velocity. For statistical analysis we used Chi-square test.

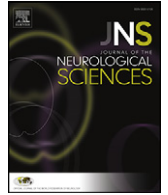
Results: We have found HD in 28 (53.8%) ICA right, and 25 (48.1%) ICA left; significant arterial stenosis on carotid bulb was observed only in three patients.

There was statistical significant difference between the hemodynamic disturbance in the left cervical ICA as a consequence of presence of Kinking and the occurrence of intracranial aneurysms in carotid branch ($p < 0.02$).

We observed the following co-morbidities in our patients: 43 patients (82.7%) had arterial hypertension, 8 (15.4%) had diabetes, 9 (17.3%) had hypercholesterolemia, 22 (42.3%) had smoking and 6 (11.5%) had aneurysm in family.

Conclusion: Hemodynamic disturbance due to kinking may play a role in the development of an aneurysm in the smoker's brain vessel.

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Topic: 3 - Stroke

Abstract – WCN 2013

No: 1095

Topic: 3 – Stroke

A pooled analysis identifies the potential and limitations to improve upon intravenous rt-PA

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Background: Intravenous thrombolysis (IVT) remains the most effective therapy for ischemic stroke. While non-randomized series suggested better outcomes for add-on and other approaches, larger randomized clinical trials (RCTs) were mostly negative. Because imbalances in important baseline factors between treatment and control arms affect all but the largest trials and may have produced misleading results, we developed techniques to identify potentially efficacious therapies by providing a pooled comparison sample to test against.

Objective: To accommodate imbalances by testing outcomes of trials intended to improve upon IVT against pooled models at each trial's own baseline NIH Stroke Scale and age.

Patients and methods: We generated outcome models (modified Rankin Scale 0–1 & 0–2) with the novel feature of multi-dimensional statistical intervals pooled from the thrombolytic arms of all published IVT RCTs. A function was fitted and individual trial results were tested against this model to determine whether outcomes surpassed the $p < .05$ surface. We analyzed IVT add-on and endovascular/thrombectomy trials.

Results: Functions derived from 14 IVT RCT thrombolytic arms representing 1785 subjects were generated ($r^2 \geq .67$). Nineteen trials were analyzed representing 1748 subjects. Only TUCSON (3 hour IVT/ultrasound with microspheres), CLOTBUST (3 hour IVT/ultrasound), and SYNTHESIS (3 hour IA rt-PA) achieved significant improvement, but notably SYNTHESIS-EXPANSION (4.5 hour IA rt-PA) did not. No extended window endovascular/thrombectomy trial showed benefit.

Conclusion: Three hour treatment with adjunctive ultrasound or IA rt-PA appears to be the most promising approach, indicating that there remains little evidence that the therapeutic window can be extended other than by IV rt-PA.

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Abstract – WCN 2013

No: 1793

Topic: 3 – Stroke

The importance of ultrasound in the early diagnosis of vertebral artery dissection

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Introduction: Arterial dissection occurs as a result of penetration of the blood vessel wall, which leads to the separation of the layers of the wall and formation of intramural hematoma, or dissection aneurysm.

Objective: To determine the correlation of ultrasound findings with MR angiographic diagnosis of unilateral vertebral artery (VA) dissection.

Materials and methods: We retrospectively examined 12 patients, 4 men and 7 women aged 33 to 60 years (mean age 48 years) with unilateral vertebral artery dissection who were hospitalized at our institution during the period from 2010 to 2012. All patients had undergone echosonographic examination of neck blood vessels, followed by MRI with MR angiography of the head and neck.

Result: Echosonography of neck blood vessels showed a significant asymmetry with reduced flow in 9 patients, 4 of whom had stenosis of a longer segment of VA1, VA2 and/or VA3 segment. In two patients occlusion of VA1 segment was registered. In one patient normal findings were registered. MR angiography of blood vessels of the head and neck showed dissection of the extracranial segment in 8 patients, craniocervical transition in two patients and VA4 segments of three patients. Ten patients had stenosis of blood vessels (50–90%) in the field of dissection and occlusion in two patients.

Conclusion: Ultrasonography is also highly sensitive to the disturbance of hemodynamic flow characteristics in the vertebral arteries, which further indicates the MR angiographic diagnosis.

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Abstract – WCN 2013

No: 1797

Topic: 3 – Stroke

Intracranial carotid artery dissection in puerperium

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Introduction: Intracranial artery dissection is an important cause of stroke in young adults, but it has rarely been reported as a cause of stroke in puerperium.

Case report: We report a case of a 27-year-old female with a past history of migraine, who presented with unilateral left headache and transient episode of dysphasia and right hemiparesis, 30 days after vaginal delivery. The first symptom started six days after the uncomplicated childbirth.

She was admitted to our department, with strong headache, confusion, and right hemiparesis. CT and laboratory tests were all unremarkable. Cervical ultrasonography revealed a different flow in the distal part of the internal carotid artery, with low flow velocity in the left ICA. MRI revealed some high signal abnormalities, with one predominant lesion in the left parahippocampal region suggestive of ischemia,

MRA with 3D TOF confirmed supraclinoid carotid artery subocclusion and suggested intracranial carotid artery dissection.

After 5 days she was impaired and we did DSA that showed a left intracranial ICA occlusion.

The patient was placed under oral anticoagulation and one month after she was discharged, control ultrasonography showed good flow in the left internal carotid artery that suggests recanalization.

Conclusion: The pathophysiology of intracranial artery dissection appears multifactorial, with evidence suggesting traumatic and genetic contributions. Vessel wall injury related to the Valsalva maneuver during labor, with pressure vessel wall on skull bones as well as hemodynamic and hormonal changes related to pregnancy may be the cause of peripartum spontaneous intracranial artery dissection.

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Abstract – WCN 2013

No: 1808

Topic: 3 – Stroke

The importance of localization of dissection of the internal carotid artery on clinical outcome

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Introduction: Carotid artery dissection occurs as a result of the split of the vessel wall with the formation of intramural hematoma that leads to narrowing of the lumen and is usually localized in the extracranial internal carotid artery (ICA).

Objective: To determine the importance of localization of the internal carotid artery dissection on clinical outcomes.

Materials and methods: We retrospectively studied 30 patients, 20 men and 10 women aged 27–66 years (mean age 48.8 years) hospitalized in our institution in the period of 2009–2012. A total of 29 patients had unilateral and 1 patient had bilateral dissection of ICA. Before the hospital discharge all patients had undergone assessment through the National Institute of Health Stroke Scale (NIHSS) and magnetic resonance imaging (MRI) with MR angiography of the head and neck.

Results: MR angiography in 23/30 patients showed dissection of the extracranial segment of 11 (48%) with occlusion of a blood vessel, and 12 (52%) in the long segment stenosis. 7/30 patients had dissection of the intracranial segment of the ICA, of which 6 (86%) had occlusion, and 1 (15%) had stenosis of the vessel. The average NIHSS at discharge was 8 in patients with extracranial ICA dissection, and 4 in patients with intracranial dissection.

Conclusion: Dissection of the intracranial segment of the ICA often goes with occlusion of a blood vessel, but with better clinical recovery compared to dissection of the extracranial segment of the ICA.

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Abstract – WCN 2013

No: 1801

Topic: 3 – Stroke

Rheumatic stroke

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Introduction: Cerebrovascular pathology in rheumatic fever has a variety of clinical manifestations. Stroke can be caused by rheumatoid vasculitis of cerebral vessels.

Purpose: To study the clinical, immunological, and neurophysiological changes in the diagnosis of stroke in rheumatic fever.

Materials and methods: Rheumatic probes, ECG, echo-cardiography, immunological methods.

The study included 44 patients with stroke in rheumatic fever. The age of patients range from 20 to 68 years (mean age 46.6 ± 1.3 years), 9 of them (20.5%) men and 35 (79.5%) women.

Results: Stroke was more frequent between the ages of 30 and 60–37 (84%).

All cases revealed marked changes in the heart rate by type of atrial fibrillation, arrhythmia.

Stroke more often (91%) occurred on the background of an existing visceral rheumatic fever. The following forms of rheumatic stroke were observed: transient ischemic (5 patients), ischemic (37) and hemorrhagic (2) strokes.

The majority of patients had changes in the peripheral blood and immunobiochemical indicators: increasing titer of Asl-O, positive C-reactive protein, and increased haptoglobin.

The study of the immune status in patients with RI showed a significant deficit of T-lymphocytes in the 1.6 and 1.9-fold (relative and absolute content, respectively), T-suppressor cells in 1.9 and 1.8 times the level of B-cells – 1.2 and 1.3 times compared with the control.

Conclusions: Thus, acute rheumatic stroke tends to occur in young and middle-aged individuals, 3.9 times more often in women, against rheumatic fever activity and gross lesions of cardiac rhythm disturbances, with immunodeficiency. These methods have diagnostic and prognostic values.

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Abstract – WCN 2013

No: 1028

Topic: 3 – Stroke

Public awareness of the main risk factors of stroke and its primary prevention

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Background: In the effectiveness of prevention of stroke the primary role played by the awareness of the population about the problem and ways to prevent it.

Objective: To study the state of public awareness of risk factors of a stroke and measures for its primary prevention.

Methods of research: This continuous study interviewed the population of Toy-tepa town using a questionnaire whose question list is specially formulated by us, consisting of 28 questions, covering main risk factors of stroke.

Results and discussion: 1346 people were interviewed, the average age was 42 ± 3.2 years. Women comprised 67.6%, while men comprised 32.4% of the respondents. 49.71% of the respondents didn't know the level of their blood pressure. 87 people were suffering from arterial hypertension. 20.68% of them regularly take hypotensive drugs, 22.98% take only at an increased level of blood pressure, and 56.32% didn't take antihypertensive drugs. The most common group of hypotensive drugs taken by the population were ACE inhibitors, raunatinum, and dibasolum. 2% of respondents know their level of cholesterol in the blood. 17.16% of the patients knew their blood glucose level, with increased levels in 7.2% of them. 17.75% of respondents smoke, with smokers accounting for 48.5% of all men, and 3.07% of all women. 29.92% of patients responded that they suffer from being overweight.

Conclusions:

- 1) Analysis points to the lack of knowledge of the population about risk factors of stroke, such as arterial hypertension, cerebral atherosclerosis, and diabetes mellitus.

2) The population is not sufficiently actively engaged in non-drug prevention of the diseases, which consists of smoking refusal, the struggle with physical inactivity, and overweight.

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Abstract – WCN 2013

No: 1030

Topic: 3 – Stroke

Peculiarities of the etiology of ischemic stroke (IS) in young patients

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Background: Stroke is becoming increasingly medico-socially and economically significant. Stroke in young patients is becoming an everyday reality.

Objective: Explore and compare the etiology of ischemic stroke in young people and the elderly.

Materials and methods: The study included 40 patients (20 young people; 20 elderly) with IS. We used the descriptive method and entered data on a special form.

Results: All patients were divided into two groups: the main group of young patients (50% men and 50% women) with ages ranging from 23 to 44 years, the average age was 40.5 years; and the control group of elderly patients (70% men, 30% women) with ages ranging from 61 to 78 years, the average age was 67.35 years.

Etiological factors of IS in young patients were: essential arterial hypertension (AH) – 65%; atrial fibrillation – 15%; cerebral atherosclerosis + AH – 10%; symptomatic AH – 5%; neuro-rheumatism – 5%; AH + the use of combined oral contraceptives – 5%; AH + the use of psychoactive substances – 5%; AH + alcohol abuse – 5%. Etiological factors of IS in the elderly were: the essential AH – 55%; cerebral atherosclerosis + AH – 25%; diabetes mellitus type II + AH – 15%; and atrial fibrillation – 5%.

Conclusions: 1. Young patients met the following causes of IS, which were not found in elderly patients: neuro-rheumatism, symptomatic arterial hypertension, a combination of arterial hypertension with the use of psychoactive substances, the use of combined oral contraceptives, and abuse of alcohol.

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Abstract – WCN 2013

No: 1819

Topic: 3 – Stroke

The effect of opium addiction on carotid intima media thickness

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Background: The hazardous effects of smoking on the progression of atherosclerosis have been well studied, but little is known about the effect of opium addiction on the progression of atherosclerosis.

Methods: A total of 202 subjects who were screened for brain disease (at annual medical checkups) between April 2010 and March 2012 were studied to clarify the effects of smoking and opium addiction on maximum carotid intima media thickness (IMT). Univariate analyses were performed to investigate the relationships between maximum IMT and independent variables, such as smoking, opium addiction, age, gender, hypertension, and diabetes.

Results: Although maximum IMT was significantly higher in the opium addiction group (7.9%) in comparison with the non-addicted group

($p = 0.02$), univariate analysis revealed that age ($p = 0.000$), diabetes ($p = 0.004$), hypertension ($p = 0.006$), and smoking ($p = 0.02$) had a significant relationship with maximum IMT. Opium addiction ($p = 0.22$) and sex ($p = 0.43$) had no significant relationship with maximum IMT.

Conclusions: Opium addiction has no hazardous effects on the progression of atherosclerosis. Prospective studies with a larger sample size are needed to confirm this hypothesis.

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Abstract – WCN 2013

No: 1645

Topic: 3 – Stroke

A reversible cause of bilateral thalamic infarct and cognitive impairment

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Background: Bilateral thalamic infarct is rare and associated with cognitive impairment, gaze palsy and altered consciousness. We report a patient who had a reversible cause of bilateral thalamic infarct and his cognitive function improved significantly following an intervention.

Result: A 61 year old right handed man presented with one-day history of acute confusional state to the emergency department. He then developed intermittent agitation, anterograde amnesia and increased drowsiness three days after the hospital admission. His routine blood test, ECG, EEG and CSF study were unremarkable. His CT head showed bilateral thalamic haemorrhagic infarct. His CT venogram raised a suspicion of venous hypertension and possible recanalisation as there was irregularity in the right internal cerebral vein and hypoplastic straight sinus although no thrombus was seen. He was given intravenous heparin initially even though CT venogram was not diagnostic of cerebral venous sinus thrombosis. Subsequently a digital subtraction angiography was performed which revealed a vein of Galen dural arteriovenous fistula. It was treated successfully with embolisation. His consciousness level stabilized and cognitive function steadily improved. He was discharged home with mild short-term memory deficit, impaired executive function and visual-spatial awareness two months later.

Conclusion: Investigating bilateral thalamic infarct requires a careful workup. When standard imaging is inconclusive, digital subtraction angiography should be considered as highlighted in this case which showed that the detection of vein of Galen dural arteriovenous led to definitive treatment and improvement of the cognitive function.

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Abstract – WCN 2013

No: 1850

Topic: 3 – Stroke

Poststroke pain: A prospective study of the association and risk factors in patients presenting with stroke

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Introduction: Pain is a significant part of many stroke patients' experience. Although pain is defined as an individualised experience, specific types of pain which commonly occur following stroke are well described.

Aims of this study were: To determine prevalence; intensity and risk factors of pain after stroke.

Methods: It is about a prospective study in a Tunisian population containing 121 patients with stroke who were selected from Sahloul Hospital in Sousse going from January 2012 till December 2012.

The criteria of inclusion: Diagnosed stroke by the regular doctor.

The criteria of exclusion: Cognitive disorders (confusions), aphasia.

Measuring Instrument: Neuropathic pain was evaluated by DN4. A score upper or equal to four “4/10” defines a positive test, which confirms the positive diagnosis of a neuropathic pain.

Worst pain intensity during the previous 48 h was assessed on a visual analogue scale (VAS), ranging from 0 to 100: a score of 0 to 30 was defined as no or mild pain; and 40 to 100 as moderate to severe pain. Screening for depression was done using HAD-S. SF36 was used to evaluate life quality.

Results: Moderate to severe pain was reported by 63% of patients (VAS median = 70). The DN4 was positive in 52%. Predictors of pain were younger age, male sex, worse HAD-S score and short disease duration. Pain impaired quality of life in 2/3 of cases.

Conclusion: Pain following stroke is common and may have a considerable effect on the everyday life of the patient.

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Abstract – WCN 2013

No: 1844

Topic: 3 – Stroke

Juvenile stroke as presenting manifestation of essential thrombocythemia with JAK2^{V617F} mutation

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Thromboses of cerebral veins or arteries may herald a latent/subclinical essential thrombocythemia (ET), which is a myeloproliferative disorder (MPD) with sustained elevated platelets. Juvenile stroke as presenting manifestation of ET has rarely been reported.

Case report: A 31 year-old man, without cardiovascular risk factors, presented with acute onset of gait ataxia and transient mesic disturbances. Brain MRI showed multiple acute ischemic lesions in the cerebellum and in the right occipital lobe.

Blood tests were normal except for an elevated platelet blood count (600,000/mm³).

The acquired Janus kinase 2 V617F mutation (JAK2^{V617F}) resulted positive. Bone marrow examination showed clustered hyperlobulated megakaryocytes. He was diagnosed with ET. The patient had complete recovery from stroke and underwent therapy with antiplatelet agents and hydroxyurea.

Discussion: MPD are hematologic malignant conditions that include: ET, polycythemia vera (PV), primary myelofibrosis (PMF) and chronic myeloid leukemia.

Detection of the JAK2^{V617F} mutation is a useful tool for early MPD diagnosis: it is present in over 90% of patients with PV, 50–70% with ET and 30–58% with PMF, while it is absent in healthy subjects.

An early diagnosis of MPD, especially in patients with thrombotic events in the latent MPD phase, would be beneficial for preventing further morbidity.

Special attention must be given to patients with acute stroke at juvenile onset, especially in the absence of cardiovascular risk factors. Therapeutic decision can be based on recent literature data where administration of antiplatelet agents, in combination of hydroxyurea, has been shown to be beneficial for MPD patients with thrombotic events carrying the JAK2^{V617F} mutation.

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Abstract – WCN 2013

No: 1836

Topic: 3 – Stroke

Aspergillus fumigatus ethmoidal sinusitis invading cavernous sinus responsible for cerebral infarction: A case report

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A 56-year-old immunocompetent man presented to the emergency department with a right hemiplegia of sudden onset. He reported a history of recent protracted fever after a surgery for ethmoidal chronic sinusitis. MRI showed an ischemia of the left lenticular-caudate nucleus and a tight stenosis affecting the supracavernous segment of the left internal carotid artery and the first segment of the left middle cerebral artery. Stenosis was due to a surrounding T2-hypersignal structure, enhanced by gadolinium injection, wrapping and compressing both arteries and infiltrating the left cavernous sinus. Blood cultures, transesophageal echocardiogram and PET scan were normal. Aspergillus antibodies were positive with high titer. CSF analysis revealed lymphocytic meningitis with elevated eosinophils. Beta-d-glucans were positive in CSF and negative in serum consistent with a fungal infection limited to CNS. *Aspergillus fumigatus* was identified after cultures on surgical ethmoidectomy samples.

The diagnosis is an external compression of the left internal carotid secondary to an aspergilloma coming from a remaining post ethmoidectomy skull-base breach leading to cerebral infarction. An antifungal treatment by 800 mg per day of oral voriconazole has been started for 6 months. A slight lesion regression was noted after 2 months of treatment with stable blood flow in the carotid and serum anti-aspergillus IgG drop. CSF analysis improved with a decrease in beta-d-glucans level and white cell count.

This case emphasizes the possibility of a local arterial damage, in an immunocompetent patient, due to *A. fumigatus* without systemic infection or cerebral vasculitis commonly described.

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Abstract – WCN 2013

No: 1891

Topic: 3 – Stroke

Number of patients and current use of antithrombotic agents before the onset of intracerebral hemorrhage

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Background: Antithrombotics are widely used to treat some vascular diseases. Intracerebral hemorrhage is the major risk associated with antithrombotics.

Objective: To assess how many patients had taken antithrombotic agents and the type of agent used before onset of hemorrhagic stroke transferred to a suburban emergency hospital in Japan. The antithrombotic agent most used annually was also assessed.

Patients and methods: This retrospective analysis focused on patients who were transferred to our institution due to intracerebral hemorrhage (ICH) from January 2009 through December 2012, inclusive. Antithrombotic agents were classified into three groups: antiplatelets (P), anticoagulants (C) and both agents (PC).

Results: There were 492 consecutive patients with ICH. Mean age was 74 ± 12.67 years (males 270, female 220), including 128 (26.02%)

patients previously treated with antithrombotic agents. Groups consisted of the following: P (69.5%; 89/128), C (20.3%; 26/128) and PC (10.16%; 13/128). In group P, aspirin was used in 64 of 128 patients (50%), and thienopyridine agents in 19 patients (14.8%). In group C, warfarin was only used in 26 patients (20%) and no new oral anticoagulants. In the PC group, every patient took warfarin (13 patients; 10%) and many patients also used aspirin. Aspirin was the most widely used agent (~60% annually) followed by warfarin (~30% annually).

Conclusion: Over one-quarter of ICH patients had taken antithrombotic agents before hospitalization annually and these agents included aspirin and/or warfarin. Our results suggest that new antithrombotics, with fewer bleeding risks, may decrease the incidence of ICH.

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Abstract – WCN 2013

No: 1898

Topic: 3 – Stroke

Relationship of brachial-ankle pulse wave velocity and cerebral microbleeds, lacunar infarcts and white matter lesions in acute ischemic stroke

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Background: Brachial-ankle pulse wave velocity (BaPWV) is a representative and simple method for evaluating arterial stiffness. Increased arterial stiffness may lead to vessel damage and atherosclerosis. Vessel wall damage in relation to arterial stiffness develops more frequently in the microvessels of end organs.

Objective: We investigated the relationship between arterial stiffness, as measured by BaPWV and cerebral small vessel disease, visualized on MRI as white matter lesions (WMLs), lacunar infarcts (LIs), and cerebral microbleeds (CMBs).

Patients and methods: A total of 88 patients with acute ischemic stroke who had undergone BaPWV measurement and brain MRI were enrolled. The number of CMBs and LIs were rated. The locations of CMBs and LIs were divided into infratentorial, lobar and deep regions including the basal ganglia, thalamus and internal capsule. The WMLs were separated into deep and periventricular regions and the severity of WMLs was assessed.

Results: Patients with higher BaPWV tended to have more LIs and more severe WML in both deep and periventricular regions ($p < 0.05$). There was no significant association between the number of CMBs and BaPWV. Patients with higher BaPWV had more number of CMBs and LIs in the deep regions than in the lobar or infratentorial regions ($p < 0.05$).

Conclusion: These findings suggest that increased BaPWV is associated with the severity of cerebral small vessel diseases. Moreover, deep regions, such as the basal ganglia and thalamus, are more related with systemic arterial stiffness.

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Abstract – WCN 2013

No: 1198

Topic: 3 – Stroke

Comparison between multiphasic perfusion CT guided selection and perfusion MRI selection of patients with acute ischemic stroke for endovascular treatments

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Background: The objective of this study was to compare between multiphasic perfusion computed tomography (CTP) selection and perfusion MRI (MRP) selection of patients with acute ischemic stroke to identify those patients who stand to benefit most from endovascular recanalization.

Methods: We reviewed acute stroke patients who received intra-arterial thrombolysis initiated within 6 h of stroke between October 2009 and August 2011. In acute ischemic stroke patients within 4.5 h from symptom onset, tPA of 0.45 mg/kg was infused intravenously after exclusion of hemorrhage by CT screening, and subsequently a further decision on intra-arterial recanalization was made after CTP or MRP screening.

Results: A total of 98 patients received intra-arterial thrombolytic therapy within 6 h of stroke onset during the study period, 54 patients with the CTP guided group and 44 patients with the MRP guided group. The CTP guided group had a shorter door-to-imaging time ($P = 0.07$) and door-to-procedure time ($P = 0.05$), compared with the MRP guided treatment group. The post thrombolysis outcome of the CTP guided group (difference of NIHSS from initial to post thrombolysis; CTP guided -2.59 , MRP guided -2.39) is better than that of the MRP guided patients ($P = 0.03$) but favorable outcome at 7 days after onset was similar in 47 (79.7%) of the CTP guided versus 45 (70.3%) of the MRP guided patients ($P = 0.737$).

Conclusion: These results suggest that CTP guided endovascular treatment could provide useful information when deciding upon intervention in hyper-acute ischemic stroke patients. If MRI equipment is not available or patients are contraindicated for MRI, the perfusion CT can be used as an alternative tool.

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Abstract – WCN 2013

No: 1872

Topic: 3 – Stroke

CYP2C19 genotype and risk of in-stent restenosis among vertebral artery stent patients treated with clopidogrel: A retrospective study

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Preventing stroke through endovascular treatment with vertebral artery stenting (VAS) remains a great challenge due to the occurrence of an in-stent restenosis (ISR) and a lack of randomized controlled trials. In this study, a retrospective analysis was conducted involving 163 patients who had been treated with VAS between January 2004 and December 2011 in the Drum Tower Hospital of Nanjing University Medical School. Patients were followed-up at 3, 6, and 12 months after treatment and annually thereafter. The cumulative long-term incidence of ISR was evaluated and risk factors for ISR were identified. DNA was extracted from blood for genotyping CYP2C19 (636G>A, 681G>A), CYP3A4 (894C>T), and P2Y12 (34C>T, 52G>T). The results demonstrated that in-stent restenosis occurred at a higher rate, mainly within the first year after VAS treatment (4.05%) during long-term follow-up. A multiple binomial regression analysis showed that patients with smoked, or hypertension or hyperlipidemia or CYP2C19 (*2 or *3), were more likely to develop ISR. The strongest statistically significant predictors of subsequent ISR was hyperlipidemia (OR 4.305, 95% CI 2.988–18.761, $p = 0.042$), and CYP2C19 (OR 0.521, 95% CI 0.17–0.699, $p = 0.035$) by multivariate logistic regression analysis. 21.4% patients had stroke with an ISR, 8.3% patients had stroke without ISR during a median of 54 months follow-up time (OR: 0.331, 95% CI: 0.112–0.973). CYP2C19 with *2 or *3 alleles had a higher incidence of restenosis ($P = 0.004$) and stroke (Kaplan–Meier analysis) than that with *1 allele.

Conclusion: Hyperlipidemia and CYP2C19 impotency are risk factors for the development of ISR in VAS treated ischemic patients.

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Abstract – WCN 2013

No: 806

Topic: 3 – Stroke

Isolated infarction of medullary pyramid: Its clinical features and pathology

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Background: Medial medullary infarction (MMI) is caused by hypoperfusion of the anteromedial territory in which blood flow is supplied by bilateral vertebral arteries and the anterior spinal artery. MMI usually causes hemiparesis, deep sensation disturbance, and hypoglossal nerve palsy. Isolated infarction of medullary pyramid (IIMP) is considered an incomplete form of MMI. Its semiology and etiology are unknown due to its rarity. We report the clinical characteristics, prognosis, and one autopsy finding of IIMP.

Patients and methods: We checked MRI reports of all the patients with ischemic stroke (n = 3185) hospitalized between January 2005 and October 2012. We reviewed the medical records of patients diagnosed with IIMP by brain MRI-DWI taken 48 h after onset to obtain information about the distribution of muscle weakness, concomitant symptoms, TOAST classification, and 3-month prognosis.

Results: Seventeen (14 males/3 females) patients (0.53%) had IIMP, and all presented with contralateral hemiparesis with the weakness more prominent in the lower extremities (n = 15). Six patients (35%) had ipsilateral hypoglossal palsy. When applying TOAST classification, twelve cases (71%) were confirmed as large-artery atherosclerosis and four (24%) as small-artery occlusion. Thirteen patients (76%) have become independent with stable walking and had a modified Rankin Scale score of 2.3 ± 1.4 three months after stroke onset. An autopsy case of a 77-year-old man exhibited necrotic cells restricted to the left medullary pyramid and severe arteriosclerosis of vertebral arteries.

Conclusion: IIMP more often causes leg dominant hemiparesis and generally has a good 3-month prognosis. Its etiology closely associates with atherosclerosis of the vertebral arteries.

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Abstract – WCN 2013

No: 1864

Topic: 3 – Stroke

The predisposing factors to hematoma enlargement in spontaneous intracerebral hemorrhage

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Hematoma enlargement occurs commonly in patients with spontaneous intracerebral hemorrhage and worsens their clinical outcome. This study analyzed the main predisposing factors of hematoma enlargement in spontaneous intracerebral hemorrhage through a retrospective study data of 268 patients. All patients underwent their first CT scan within 24 h of arrival and a second CT scan within 72 h of admission. These factors that were assumed to affect the growth of hematoma, such as consciousness level, blood pressure, clinical outcome, hematological

parameters, and CT findings (hematoma shape, location, midline shift and broken into the ventricles), were analyzed by means of univariate and multivariate logistic regression analyses. Univariate analyses indicated that Glasgow coma scale (GCS), hematoma shape, midline shift, breaking into the ventricle, thrombin clotting time (TT), white blood cells, neutrophil percentage, serum creatinine, and fasting plasma glucose (FPG) were significantly different between the patients with or without hematoma enlargement ($P < 0.05$). Multivariate analyses revealed that the following four factors were independently associated with hematoma enlargement: the midline shift, the GCS score, the FPG, and the breaking into the ventricles. Therefore, timely review of CT findings and dynamic monitoring of peripheral blood biochemical indicators should be taken in patients who have a risk of developing hematoma enlargement, which is very important for further therapy.

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Abstract – WCN 2013

No: 1862

Topic: 3 – Stroke

Ambient air pollution and emergency department visits for stroke

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Background: Many epidemiological studies have shown the short-term effects of air pollution on cardiovascular diseases. Several studies have investigated the relationship between air pollutant exposure and stroke but the findings have been inconsistent.

Objective: We conducted a time-series study to assess the association between ambient air pollution and emergency department (ED) visits for stroke in Seoul, Republic of Korea.

Materials and methods: The ED data from 2005 to 2009 were obtained from the Health Insurance Review and Assessment Service, and the number of ED visits for stroke was 118,171. We used a generalized additive model with Poisson distribution, adjusting for temperature (cubic spline, $df = 6$), relative humidity (cubic spline, $df = 3$), day of the week, and national holidays. The risk was expressed as a RR with one standard deviation increase and its 95% CI. We explored the lag effects with cumulative lag models (lag0–1 to lag0–7).

Results: In the same day exposure, the RRs of ED visits for stroke were 1.016 (95% CI, 1.011–1.023) per 12.04 ppb increment of NO₂ and 1.014 (1.007–1.020) per 0.24 ppm increment of CO. In lag0–7, the RRs for stroke were 1.009 (1.002–1.017) per 2.33 ppb increment of SO₂, 1.009 (1.004–1.015) per 36.70 µg/m³ increment of PM₁₀, 1.018 (1.010–1.026) per 10.04 ppb increment of O₃, 1.012 (1.006–1.018) for NO₂, and 1.010 (1.003–1.017) for CO. Positive correlations were observed in PM₁₀ ($p = 0.029$) and O₃ ($p < 0.001$) for hemorrhagic stroke and all pollutants for ischemic stroke in lag0–7.

Conclusion: Ambient air pollutants are significantly correlated with ED visits for stroke.

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Abstract – WCN 2013

No: 1861

Topic: 3 – Stroke

Psychosis after thalamic infarcts: Is it just post-stroke delirium or does the thalamus have a pivotal role to play?

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Background: As the ‘gatekeeper’ of information and stimuli from the outside world reaching the brain, the thalamus plays a key role in the processing and conveyance of this information. The prevalence of post-stroke psychotic disorders is rare, and apart from an association with frontoparietal lesions, they have also been described after infarcts in the thalamus.

Objective: We describe the case of a 51 year-old gentleman who suffered acute bi-thalamic infarcts. By describing the sequence of events from presentation to the subsequent development of cognitive sequelae, as well as the changing nature of his psychotic episodes, we are able to make a distinction between post-stroke delirium and psychosis.

Patients and methods: We undertook a retrospective analysis of the medical notes (including entries from the medical, psychiatric and therapy teams) and investigations performed in the London Hyper-Acute and Acute Stroke Units.

Results and conclusion: The patient's post-stroke delirium developed shortly after the bi-thalamic infarcts, had a strong diurnal component to it and was also associated with disorientation in time and place, whereas the psychotic disorder evolved over a period of weeks and included worsening paranoid ideas and delusions. A possible link between thalamic dysfunction and psychosis is explored after considering the neuronal networking of thalamic relays with the reticular brainstem formation as well as layers 6 and 5 of the cerebral cortices.

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Abstract — WCN 2013

No: 1856

Topic: 3 — Stroke

Cerebral vasoreactivity with breath holding test in Transcranial Doppler and functional MRI in patients with carotid stenosis

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Background: Impaired cerebral vasoreactivity (CVR) on Transcranial Doppler (TCD) with breath holding test (BHT) is predictive of stroke in patients with carotid stenosis. Functional MRI (fMRI) through BOLD-signal change can also evaluate the hemodynamic response, with the advantage of imaging resolution. Few studies have shown a good correlation between these methods, but none of them used BHT.

Objective: To apply the BHT on TCD and fMRI for CVR evaluation in patients with carotid stenosis and controls in order to verify if fMRI can show any difference on BOLD-signal between these groups and detect those subjects with impaired CVR on TCD.

Patients and methods: Fifteen patients with carotid stenosis (11 men; 52–90 yo; mean: 67.7 yo) and 7 matched-controls (5 men; 51–85 yo; mean: 64.6 yo) performed BHT during TCD. (Impaired CVR = mean percentage increase of MCA velocities $\leq 31\%$ on 3-consecutive intervals of 30 s of apnea intercalated by 4-minute-intervals of normal breathing.) During fMRI, BHT was performed through a traffic light-based code for instruction on when to stop breathing (3-consecutive intervals of 24 s of apnea intercalated by 4-minute-intervals of normal breathing). BOLD-signal intensity was measured in the lentiform nucleus ipsilateral to the carotid stenosis. In controls, the mean signal intensity of both lentiform nuclei was considered.

Results: Patients and controls showed significantly different BOLD-signal intensities ($p = 0.0053$). Based on CVR status by TCD, there was an agreement between TCD and fMRI of 6/7 (85.7%) in controls, 11/15 (73.3%) in patients, and 17/22 (77.2%) overall.

Conclusion: Through this inexpensive technique for hypercapnia induction, BHT, fMRI showed different BOLD intensity signals between

patients and controls and, additionally, presented a good correlation with TCD.

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Abstract — WCN 2013

No: 1936

Topic: 3 — Stroke

Do cervical spine spondylosis and osteophytes influence dysphagia in stroke patients? Case-control study

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Introduction: Although there are many risk factors that influence dysphagia outcome, the role of cervical spine abnormalities in stroke patients has not been reported, yet. This study was carried out to determine whether the presence of cervical spine abnormalities can influence the severity and outcome of dysphagia in stroke patients.

Method: We retrospectively reviewed 800 patients' medical records and video fluoroscopic swallowing study (vfss) reports performed at our department. We identified 120 stroke patients with severe cervical spondylosis. Among these, 19 patients were identified who had single brain lesion. We case matched these 19 patients according to their lesion site, gender and age, to a control stroke group with dysphagia, with normal cervical spine findings (total $n = 38$, mean age = 71.6 ± 10.9). We compared the two groups' dysphagia severity scale scores, the parameters recorded at their initial vfss and swallowing outcomes at follow-up.

Result: Compared to controls, the case group showed a more severe degree of dysphagia ($P < 0.005$) than the control group from their dysphagia severity scale. The case group also had more cases who showed residues at the vallecular (68%, $P < 0.008$) and pyriformis sinuses (100%, $P < 0.000$) and were more likely to have poorer dysphagia outcome ($P = 0.004$) at follow-up than the control group.

Conclusion: Cervical spine abnormalities can influence the initial severity and outcome of dysphagia in stroke patients. Although cervical spine abnormalities do not cause more severe aspiration, it seems to have an adverse effect on swallowing, by causing greater amounts of residues with reduced clearance of bolus.

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Abstract — WCN 2013

No: 1943

Topic: 3 — Stroke

Comparative study of combination therapy of Korean-Western medical treatment versus Western medical treatment only in acute ischemic stroke patients

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Background: Stroke is one of the 3 major causes of death worldwide and can leave irreversible neurological deficits, which is why appropriate treatment is important.

Objective: The objective of this study is to report the effectiveness of Korean and Western medicine combination therapy on cerebral infarction.

Patients and methods: Subjects were patients diagnosed with acute cerebral infarction within 10 days of onset, admitted to Kyung-Hee University Medical Center (KMC) and Kyung-Hee University Korean Medical Center (KKMC) from 2010.3 to 2011.10. Patients admitted to

KMC were set as the Western Medical Treatment Only (W-Tx) group. Patients admitted to KMC were set as the Combination Treatment (C-Tx) group. Motricity Index (MI) score and Scandinavian Stroke Scale (SSS) were checked at the start of treatment, and 3 weeks after.

Results: 15 patients were included in the W-Tx group, and 30 patients were included in the C-Tx group. After 3 weeks, the MI arm score of the W-Tx group increased from 87.0 ± 21.2 to 88.3 ± 20.9 ($p = 0.480$), and the leg score increased from 86.8 ± 23.1 to 89.5 ± 20.5 ($p = 0.210$). The MI arm score of the C-Tx group increased from 81.4 ± 12.6 to 85.9 ± 12.1 ($p < 0.001$), and the leg score increased from 78.2 ± 18.6 to 86.7 ± 17.7 ($p < 0.001$). Only the C-Tx group showed significant MI score increase. The SSS of the W-Tx group increased from 51.6 ± 9.1 to 53.1 ± 7.9 ($p = 0.034$), and the SSS of the C-Tx group increased from 44.8 ± 8.9 to 50.0 ± 5.8 ($p < 0.001$). Both groups showed significant SSS improvement.

Conclusion: This study implies that receiving Korean medical treatment with Western medical treatment may have a positive effect on improving the motor function of stroke patients.

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Abstract – WCN 2013

No: 1934

Topic: 3 – Stroke

Abdominal obesity and body mass index as a risk factor for stroke in transient ischemic attacks

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Background: Transient ischemic attack (TIA) is a proven risk factor for stroke, but on the liaison with a body mass index (BMI) is little known.

Aim: To study abdominal obesity and BMI as risk factor for stroke in TIA.

Methods: The study conducted on a “case–control”. Individuals with diagnosis of TIA (study group–“cases”) were compared with patients with arterial hypertension (AH) and control group.

Results: In total, 87 patients of the main group were examined, 84 with arterial hypertension (AH), and 94 individuals from the control group. There was a general relationship between TIA and large waist circumference (independent of BMI) compared with controls (OR = 2.23, 94% CI: 1.18–3.98, waist circumference > 80 cm vs < 80 cm). There could be a “plateau of risk” with an increased risk for circumference > 80 cm, but in the absence of a reliable trend tended to regard as compared to patients with hypertension (p value for trend = 0.03). There was no connection between TIA and BMI. Waist circumference was associated with the hypertension severity of symptoms (OR = 1.78, 94% CI: 1.04–3.37), the risk of heavy weekly high blood pressure – 10 cm circumference) correction by AH partially reduced the liaison between TIA and circle waist.

Conclusions: Waist circumference, not BMI, has moderate independent association with the risk of TIA. These data partially favor the hypothesis that abdominal obesity promotes hypertension, which in turn may increase the risk of TIA.

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Abstract – WCN 2013

No: 1923

Topic: 3 – Stroke

Endovascular treatment for acute ischemic stroke: Is immediate post-procedural extubation a new prognostic factor?

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Background: Endovascular therapy may benefit selected patients with acute ischemic stroke. Yet, it remains uncertain which factors directly influence functional outcome. Following general anaesthesia (GA), stable patients are extubated before transfer to Neurointensive Care Unit.

Objective: We aim to study if immediate post-procedural extubation correlate with functional outcome at 3 months after stroke.

Material and methods: We retrospectively reviewed and analyzed the medical records of 38 consecutive patients treated between December 2006 and June 2011 in our institution, including their GA records.

Results: 22 and 16 patients suffered acute ischemic stroke in the anterior and posterior circulations respectively. Atrial fibrillation was present in 36.8%. Mean time from symptom onset to therapy was 370 min. 7 patients were not successfully recanalized due to unfavourable anatomy. 16 patients were extubated immediately after endovascular procedure, 9 of whom enjoyed favourable functional outcome (mRS ≤ 2) at 3 months, compared to 3 of 22 who had remained intubated ($p = 0.0116$). Time to therapy, location of stroke and presence of AF were not significantly correlated with favourable functional outcome at 3 months poststroke. Mortality was 36.8%, the odds being 8.4 times higher in those who were not extubated immediately after endovascular procedure.

Conclusion: Acute ischemic stroke patients under GA for endovascular therapy are immediately extubated when they are assessed to be breathing well, able to maintain their own airway and haemodynamically stable. These patients are observed to have significantly better functional outcome at 3 months after stroke. Immediate post-procedural extubation may be an important prognostic factor.

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Abstract – WCN 2013

No: 1918

Topic: 3 – Stroke

An evaluation of neuroendocrine dysfunction following acute aneurysmal subarachnoid hemorrhage: A prospective study

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Background: Evidence that aneurysmal subarachnoid hemorrhage (SAH) is associated with neuroendocrine dysfunction comes mainly from case reports or small series of patients.

Objective: To investigate the incidence and pattern of neuroendocrine changes in cases of acute aneurysmal subarachnoid hemorrhage (SAH).

Methods: Endocrine assessment was performed in 100 consecutive cases of acute aneurysmal SAH presenting within 7 days of ictus. The gonadotropic, somatotropic, thyrotropic, and corticotropic axes and prolactin were evaluated for their possible dysfunction.

Results: A total of 100 cases (38 males, 62 females; age range–17–76 years; mean age–43.6 years) of acute SAH were studied. The aneurysms were located in the anterior circulation ($n = 95$) and posterior circulation ($n = 5$). The commonest hormone deficiency was of growth hormone ($n = 67$) followed by gonadotropin ($n = 50$), corticotropin ($n = 49$) and thyrotropin ($n = 35$). Hyperprolactinemia was noted in 10 cases. One pituitary hormone axis deficiency was noted in 26 cases while 67 cases had two or more pituitary hormone axes

deficiency. A total of 93 cases had hormonal deficiency in one or more pituitary hormone axes and 7 cases had no hormonal deficiency.

Conclusions: Endocrine dysfunction occurs in 93% of cases of acute SAH and multiple pituitary hormone axes deficiency occurs in 67% of cases. It is suggested that hormonal evaluation should be considered as a part of management of acute SAH.

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Abstract – WCN 2013

No: 1909

Topic: 3 – Stroke

Factors involved in the outcome of ischemic stroke among the Asian population: A Chinese study

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Background: Ischemic stroke (IS) is an issue in many countries and the outcome depends on the events, causation and the medical care facilities present in the health centers. The aim of this study was to evaluate the outcome of IS among patients presenting in the rural area of Eastern China.

Methods: This study was conducted in Hubei province, over a period of 1 year. A total of 335 cases of IS and suffering for the first time were recruited. The mean age was 55 ± 8 years. 60% were males. Information was obtained using a standardized questionnaire including demographic details, symptoms and signs, risk factors, Glasgow Coma Score, stroke severity, admission baseline investigations and presence of complications. The end point was mortality or survival at 1 month from the day of stroke.

Results: Strong factors predicting mortality were ruled out to be stroke severity on admission (74%), hypertension (67%), admission hyperglycemia (54%), and presence of complications (94%) especially concomitant renal or cardiac failure and chest infections during the hospitalization period. Of these factors, the presence of complications had the strongest correlation with fatality ($r = 0.52$; $p = 0.001$). Overall, 1 month fatality was 31%.

Conclusion: Our study clearly reflected that the overlapping complications seem to be the strongest factor for mortality among patients of ischemic stroke in the hospital. Early detection and prompt treatment can easily prevent these conditions and responsibility lies primarily on the physician and paramedical staff to monitor these complications in every case of ischemic stroke while taking care of the general stroke management.

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Abstract – WCN 2013

No: 1908

Topic: 3 – Stroke

State of coronary bed in patients with acute ischemic stroke with reversible and irreversible neurological impairment

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Purpose: To examine the state of coronary bed in patients with acute ischemic stroke with reversible and irreversible neurological deficits.

Material and methods: 48 patients with reversible (group 1) and 44 patients (group 2) with an irreversible neurologic deficit (57 males and 35 females) were involved in the study.

Results: Vessel disease of coronary arteries was revealed in all the patients. Group 1 patients showed lesion of main branches, and in group 2 the frequency of lesions was less. This fact requires a detailed

study of the degree of coronary lesions. Identified: in group 1 – the prevalence of stenotic coronary lesions more than 50%, stenosis of the anterior interventricular branch, particularly, in 77% cases, stenosis of circumflex branch – at 75% cases, stenosis of the right coronary artery – in 53% cases. In group 2 there is dysfunction-occlusion. A greater percentage of hemodynamically significant lesions were found in females of group 1 – 41.8%, in comparison with males – 13%.

Distal localization of lesions and large length of lesions were found in group 2, more often in women – 67%. In group 2 a sharp deterioration in hemorheology, hemostasis and fibrinolysis in acute period of ischemic stroke was detected.

Conclusions: We hope that the research results will attract the attention of physicians in choosing new ways of prevention strategies of cardio-cerebrovascular diseases. Timely and qualified early diagnosis with earlier pharmacologic therapy will reduce mortality in young people.

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Abstract – WCN 2013

No: 1962

Topic: 3 – Stroke

Management of tandem occlusion stroke with endovascular therapy eventually preceded by systemic intravenous thrombolysis

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Background: Approximately 50% of patients with internal carotid artery (ICA) occlusion have a concomitant proximal middle carotid artery (MCA) occlusion. Tandem occlusion of ICA and MCA (TIM) has a poor outcome when treated by intravenous thrombolysis (IVT).

Objective: Tandem occlusion stroke can be caused by occlusion/critical stenosis/occlusive dissection of ICA with thrombosis of MCA. The endovascular treatment can be performed in all these types of stroke, alternatively in the combination with IVT.

Patients and methods: We retrospectively studied 25 patients with TIM who underwent acute endovascular treatment. 14 patients had occlusion of ICA, 6 had critical stenosis of ICA and 5 patients suffered from occlusive dissection of ICA. Either angioplasty with stent implantation of proximal ICA followed by thrombectomy of MCA or single thrombectomy of MCA was performed. 9 patients underwent treatment with IVT before revascularisation, 16 patients were treated only with endovascular intervention.

Results: Median National Institute of Health Stroke Scale (NIHSS) score at admission was 14 (range 5–20), median NIHSS score after 24 h was 10 (range 0–35). 16 patients (64%) had good clinical outcome (mRS scale 0–2) and 3 patients (12%) died. In the group with IVT intracerebral bleeding was present in 6 patients (66%), in two cases with subsequent death. In the group without IVT 3 intracerebral hemorrhage (6%) occurred.

Conclusion: Acute thrombectomy with/without stenting ACI is a promising method in tandem occlusive disease with good clinical outcome. IVT before acute endovascular intervention in our group showed higher incidence of intracerebral hemorrhage and mortality.

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Abstract – WCN 2013

No: 1984

Topic: 3 – Stroke

Metamorphopsia restricted to the nose and mouth with right medial temporooccipital lobe infarction

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Background: Metamorphopsia includes a broad spectrum of visual perceptual distortions, such as alteration of perceived object size or, rarely, altered perception of faces, termed prosopometamorphopsia. The fusiform face area is known to play a key role in face perception.

Case: A 75-year-old right-handed woman was admitted for a sudden onset of nausea, dizziness, and blurred vision. She complained of dimmed vision, and the central part of faces, particularly the nose and mouth, appearing out of shape. She claimed, “The nose looks very narrow as well as lengthened toward the mouth, which looks small and round in shape”. Her description of how she saw faces seemed as if viewed through a convex lens. She had no prosopagnosia. She had no impairment in her visuoperceptual performances (describing a complex scene, drawing figures, reading and writing) or in color perception. Left homonymous quadrantanosia was seen. Diffusion weighted MRI and T2-weighted brain MRI revealed an infarction in the right medial temporooccipital lobe, including the parahippocampal gyrus.

Conclusions: In light of recent empirical evidence, face perception is thought to be mediated by a distributed neural system including all regions of the core and extended systems of which the major entry node is the lateral fusiform gyrus. We speculated that any injury on this pathway could bring about prosopometamorphopsia involving whole or unilateral face perception, or very rarely, such as in our case, distortion restricted to the central area of the face.

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Abstract – WCN 2013

No: 1973

Topic: 3 – Stroke

Anaemia as a predictor of the severity and functional outcome of ischaemic stroke

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Background: Korea Veterans Hospital lower haemoglobin levels may impair cerebral oxygen delivery and predispose to infarct expansion and impaired neuronal survival in the ischaemic penumbra following stroke. Whether anaemia is associated with greater stroke severity and worse outcome remains unestablished.

Objective: To identify the association of anaemia with stroke severity and functional outcome among patients with ischaemic stroke.

Methods: A cross-sectional study was conducted among 190 prospectively-recruited patients with newly-diagnosed ischaemic stroke at the National Hospital of Sri Lanka. Their socio-demography, co-morbidities, stroke type (Oxford Community Stroke Project classification), stroke severity by NIHSS, and functional outcome by Modified-Rankin-Score (MRS) and Barthel index (BI) were assessed within 48 h of stroke onset. Anaemia was defined as Hb < 13 g/dl in males and < 12 g/dl in females.

Results: Mean age was 63.1 years (SD = 12.0); 75.8% were males. Mean NIHSS was 11.07 (SD = 7.04); 27.4% had moderate-severe and severe stroke (NIHSS ≥ 15). Mean MRS was 3.63 (SD = 1.19); 68.3% had dependent mobility (MRS > 3). Mean BI was 38.26 (SD = 30.6); 85.8% had moderate-severe dependent mobility (BI < 75).

58.4% had anaemia (59.0% males; 56.5% females). Mean Hb was 11.7 (SD = 2.2) [males = 11.7 (SD = 2.3); females = 11.65 (SD = 1.9)]. Compared to non-anaemic patients, anaemic patients had significantly higher functional disability based on MRS (77.3% vs. 55.7%) and BI (90.1% vs. 79.7%) but showed similar stroke severity (27.8% vs. 27.0%). When adjusted by gender, co-morbidities and stroke subtypes, functional disability by MRS was significantly associated with anaemia [adjusted odds ratio = 2.17 (95% CI: 1.13–4.17)].

Conclusions: Anaemia is independently associated with increased functional disability, as measured by MRS.

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Abstract – WCN 2013

No: 1654

Topic: 3 – Stroke

Results of Russian prospective controlled multicentric clinical study “SOKOL” (comparable efficacy of Cavinton in standard acute stroke treatment)

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Background: The protective effect of vinpocetine (Cavinton) was demonstrated using in vitro models of oxidative stress. In vitro data supports additional clinical trials of vinpocetine to be carried out.

Objectives: The aim of the study was to evaluate the efficacy of Cavinton with standard acute stroke therapy as compared to standard therapy.

Patients and methods: 661 patients with acute stroke (7–14 day) were enrolled in the study. One group (334 patients) received standard therapy and intravenous infusions of Cavinton (4 days – 25 mg, 3 days – 50 mg) and then Cavinton 30 mg/day in tablets for 90 days. The other group (327 patients) received only standard therapy. Patient disability was the primary endpoint (the Oxford handicap scale) accessed at baseline (visit 1), after infusions (visit 2) and after 1 (visit 3), 2 (visit 4) and 3 (visit 5) months of treatment.

Results: In the Cavinton group Oxford handicap scale score was 2.9 points at the 1st visit, 2.4 points at the 2nd, 1.8 points at the 3rd, 1.6 points at the 4th and 1.3 points at the 5th visit. In the control group this rate was 2.9 points at the 1st visit, 2.5 points at the 2nd, 2.2 points at the 3rd, 2.0 points at the 4th and 1.8 points at the 5th visit. The significant difference between groups (p < 0.05) was at the 3rd, the 4th and the 5th visit.

Conclusions: Addition of Cavinton to standard treatments lowers patients' disability and may be considered as an option for long term treatment.

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Abstract – WCN 2013

No: 1999

Topic: 3 – Stroke

Transcranial colour Doppler in adult patients with sickle cell disease

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Background: The risk of stroke in patients with Sickle Cell Disease (SCD) is much higher in the paediatric population. Actually there is little knowledge about adult patients.

Aim: Try to standardise Doppler parameters in adult patients with SCD, using Transcranial Colour Doppler (TCCD) with the correction of the angle of insonation.

Material and methods: We enrolled 52 outpatients followed in the Congenital Anaemias Center in Milan (15 with Sickle Cell Anaemia, SCA;

26 with Sickle β -Thalassemia, HbS-Thal and 11 with haemoglobinopathy SC disease), over the age of 16, and 25 control subjects, matched for sex, ethnicity and age. We evaluated haemoglobin (Hb) values, systolic peak and pulsatility index in the principal intracranial and extracranial vessels using TCCD.

Results: Mean Hb values of SCD and control subjects were 9.9 ± 1.5 and 14.8 ± 1 g/dl. Adults with SCD had a higher peak-systolic velocity (120.1 ± 19.16 cm/s) compared with healthy controls (109.2 ± 14.7 cm/s); in particular SCA patients had much higher velocities than the others (126.32 ± 15.18 cm/s), according with lower Hb values and worse clinical features. We found a significant difference comparing the velocities of MCA, ACA, ICA and the pulsatility index in this vessels between the patient group and the control group ($p < 0.05$), and in particular a statistical difference only between the SCA group and the control group ($p < 0.01$).

Conclusion: Velocities in adults are lower than those provided by the STOP trial in children, confirming that the speeds disclose an age-related decline, however are higher than in healthy controls, in particular in SCA patients.

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Abstract – WCN 2013

No: 1202

Topic: 3 – Stroke

Long-term natural history of intracranial arterial stenosis:

An MRA follow-up study

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Objective: Intracranial arterial stenosis (ICAS) is a major cause of ischemic stroke in Asian populations. In the present study, we demonstrated the results of a long-term follow-up of ICAS and comparisons between symptomatic and asymptomatic stenosis.

Methods: A series of 102 subjects underwent follow-up MR angiography (MRA) five years after initial MRA. Because the sample size is too small to adjust for multiple confounders, we applied propensity score. We used a logistic regression analysis with adjustment for the propensity score. In addition, we matched the patients by propensity scores and built the second logistic regression model.

Results: The median time interval between initial and follow-up MRA was 5.7 years (range 3.6–8.5 years). The progression rate of ICAS differed significantly between symptomatic and asymptomatic patients (22% vs. 8%), indicating that symptomatic stenosis had a 3-fold risk of progression compared with asymptomatic stenosis [OR 3.27, 95% confidence interval (CI) 1.08–9.95]. After adjustment for propensity score, the OR was 4.84 (95% CI, 1.40–16.7). In the propensity score matched cohort, the relative risk of stenosis progression was 5.20 for symptomatic stenosis (95% CI 1.00–27.23) compared with asymptomatic stenosis. Furthermore, the progression rate of concomitant asymptomatic stenosis in subjects with symptomatic ICAS was largely comparable to that of asymptomatic stenosis; 3% progressed, 65% were stationary, and 32% regressed.

Conclusion: We found that symptomatic stenosis had a greater risk of progression compared with asymptomatic stenosis. In addition,

our results suggested that concomitant asymptomatic stenosis in symptomatic patients also had a benign course.

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Abstract – WCN 2013

No: 2027

Topic: 3 – Stroke

Cervical medullar infarction in multileveled discopathy

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Spinal cord infarction is a rare but devastating pathology causing acute neurological deficits. The incidence has been estimated to 1% of all strokes. In that case report, our patient has presented anterior spinal artery infarction in C5 and C6.

The only risk factor found was a multileveled discopathy which is known to be an uncommon cause of anterior spinal artery syndrome.

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Abstract – WCN 2013

No: 1076

Topic: 3 – Stroke

Evaluation of left ventricular diastolic dysfunction of acute ischemic stroke patients by transthoracic echocardiography

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Background: Left ventricular diastolic dysfunction (LVDD) and atrial fibrillation (AF) have similar risk factors. Some cohort studies showed the relationship between LVDD and the incidence of AF, though the mechanism was not revealed well. Also, the relationship between LVDD and acute ischemic stroke (AIS) has not been studied well.

Objective: We examined LVDD of AIS patients by transthoracic echocardiography (TTE).

Patients and methods: Among 920 serial AIS patients hospitalized between January 2011 and December 2012, this study included 208 cardioembolic stroke patients and 455 non-cardioembolic stroke (lacunar infarction and atherothrombotic infarction) patients who were evaluated with TTE. Hemodialysis patients, AIS patients with other mechanisms, and cryptogenic stroke patients were excluded. Left atrial diameter (LAD), transmitral diastolic peak velocity (E-wave) and ratio E-wave to the lateral mitral annular diastolic peak velocity (E/E') were evaluated as an index of LVDD.

Results: The mean LAD, E-wave and E/E' in cardioembolic stroke patients were significantly higher than those in non-cardioembolic stroke patients (40.7 ± 7.0 mm vs. 35.3 ± 5.7 mm, 82.4 ± 25.0 cm/s vs. 60.0 ± 18.2 cm/s, 12.9 ± 6.4 vs. 10.2 ± 4.4 , respectively. $p < 0.001$ for all). The areas under the receiver operating characteristic curve for diagnosing cardioembolic stroke were 0.736, 0.768, and 0.643, for LAD, E-wave, and E/E', respectively. Only E/E' showed a positive correlation with D-dimer, NIHSS on admission, and modified Rankin Scale at discharge.

Conclusion: LVDD is useful for the diagnosis of cardioembolic stroke. Cryptogenic stroke patients with LVDD require further investigation for detecting their cardioembolic sources. Only E/E' correlates with stroke severity in cardioembolic stroke patients.

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Abstract – WCN 2013**No: 2014****Topic: 3 – Stroke****Necrosectomy by MIN techniques versus craniectomy in stroke**K.D.M. Resch. *Neurosurgery, Klinikum Darmstadt, Darmstadt, Germany*

Objective: In the series of 150 hematoma evacuations some cases were hemorrhagic infarctions. In these cases additional to the hematoma the center of the necrosis was evacuated also. As these patients made a much better recovery than craniectomy cases we developed a novel concept.

Methods: Hemorrhagic infarctions were operated through key-holes under ultrasound control (burr-hole probe, Alpha 7, ALOKA) and by mouth-switch tracked microscope. Before surgery CTA or TCD was made to see if the infarction showed reperfusion, because in the latter condition the operation may be more difficult. Perfusion CT determined the center of the necrosis.

Results: In all cases we saw a much better course of recovery than in craniectomy cases. The patients were not disabled additionally by stigmata like craniectomy defect, big scar, loss of hair and psychological trauma. The beginning of rehabilitation therapy could start within one week only, the wound of 3 cm was not visible and during rehabilitation there was no fear by the rehabilitation clinics to soon start the full training program. We did not see any complications due to the MIN strategy.

Conclusion: The first promising results of MIN concept application in stroke decompression seem logical regarding pathophysiology. It is worthwhile to study this novel concept by trials. The operative technique, however is much more sophisticated and also much faster and economic than a simple craniectomy.

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Abstract – WCN 2013**No: 1925****Topic: 3 – Stroke****Neural coupling of cooperative hand movements in stroke patients**M. Schrafl-Altermatt, V. Dietz. *Balgrist University Hospital, Spinal Cord Injury Center, Zurich, Switzerland*

The neural control of cooperative hand movements reflecting “opening a bottle” has been studied in healthy subjects. It revealed a task-specific neural coupling of the upper limbs resulting in bilateral EMG responses following unilateral ulnar nerve stimulation. In contrast, during control conditions (e.g. pro-/supination) only ipsilateral reflex responses were detected.

The objective of this study was to examine this neural coupling in chronic stroke patients.

Mildly affected post-stroke patients were included 6.4 years (± 2.5) after stroke. Patients had a mean age of 53.3 years (± 7.9), were right-handed and had a left hemispheric insult resulting in a right hemiparesis. The average Fugl-Meyer score for the upper limb was 49.1 (± 7.7) out of 66. Unilateral ulnar nerve stimulations were randomly delivered during three conditions, a dynamic cooperative movement, an isometric and a dynamic pro-/supination control condition. EMG of forearm extensors and flexors was analyzed.

Stimulation of the right (affected) arm led in both conditions in ipsilateral muscles to an early response followed by segmented EMG responses. Contralateral no reflex response could be recorded in neither one of the conditions. Stimulation of the left (unaffected) arm resulted in bilateral EMG response patterns similar to the ones found in healthy subjects during the dynamic cooperative movement. Only ipsilateral responses appeared during the control conditions.

The results indicate that the task-specific neural coupling during cooperative hand movements is partially impaired after stroke

corresponding to movement performance. This has consequences for rehabilitation of upper limb function of these patients.

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Abstract – WCN 2013**No: 882****Topic: 3 – Stroke****Impact of prior IV thrombolysis on early recanalization and clinical outcome after endovascular treatment in acute ischemic stroke**D. Sanak^a, D. Skoloudik^b, T. Veverka^a, M. Kuliha^b, M. Kral^a, M. Kocher^c, M. Roubec^b, K. Langova^d, V. Prochazka^e, M. Nevrlý^a, D. Franc^a, K. Mensikova^a, R. Marak^a, P. Otruba^a, P. Kanovsky^a.

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Background: Although different endovascular approaches are now largely used for acute intracranial arterial occlusion (IAO) in ischemic stroke (IS), intravenous thrombolysis (IVT) is standard option within first 4.5 h after stroke onset. The aim was to assess the impact of prior IVT before endovascular treatment (EVT) on early recanalization and clinical outcome.

Methods: In retrospective study, 372 consecutive acute IS patients with IAO were treated with EVT and divided into 2 groups according to the presence/absence of prior IVT. Stroke severity was assessed using National Institutes of Health Stroke Scale (NIHSS), 90-day clinical outcome using modified Rankin Scale (mRS) with good outcome defined as 0–2. Early recanalization was quantified using Thrombolysis in Cerebral Ischemia scale and symptomatic intracerebral hemorrhage (SICH) using SITS-MOST criteria.

Results: Group 1 consisted of 193 patients (51% males, mean age 62.9 ± 13.8 years) treated with IVT and Group 2 of 179 patients (60% males, mean age 65.5 ± 12.4 years) without IVT. Patients treated with IVT had higher baseline NIHSS (18 vs. 15 points, $p < 0.0001$). No difference was found in recanalization rate (80.9 vs. 87.6%, $p=0.089$) including complete (41.6 vs. 46.4%, $p=0.403$), occurrence of SICH (4.1 vs. 4.0%, $p=1.000$), good clinical outcome (43.8 vs. 51.7%, $p=0.146$) and 3-month mortality (22.7 vs. 23.0%, $p=1.000$).

Conclusion: Prior IVT probably does not increase the chance for early recanalization and good clinical outcome after EVT of IAO in acute IS patients.

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Abstract – WCN 2013**No: 2034****Topic: 3 – Stroke****Fibrocartilaginous embolism myelopathy in a young healthy woman**M. Kucharik, D. Petřelnicova, M. Saling, L. Prochazkova. *2nd Dept. of Neurology, Comenius University, University Hospital Bratislava, Bratislava, Slovak Republic*

Objective: Fibrocartilaginous embolism (FCE) is a rare cause of spinal cord and cerebral ischemia, characterized by progressive paraplegia following a back pain and also symptoms from the posterior circulation. First case was reported in 1961 by a 15-year-old boy, since then, over 40 cases have been reported. MRI shows T2 hyperintensity; CSF studies are normal. There are no specific diagnostic criteria for FCE. The autopsy, which makes the diagnosis definitive, shows diffuse embolic material of the nucleus pulposus in the anterior spinal cord artery and basilar arterial system. Fibrocartilaginous disk material may travel to the spinal bone marrow through communicating sinusoids and venules. High internal pressure such as minor trauma or Valsalva maneuver may permit retrograde flow of emboli through anastomoses within spinal arterial system as a result of valveless peripheral venous system. There is no specific treatment for FCE.

Patients and methods: We present a case of a young previously healthy 25-year old female who experienced sudden severe neck pain immediately after intensive physical exercise (performing crunch) and became rapidly quadriplegic with respiratory paralysis. MRI showed spinal cord infarction in the anterior spinal artery territory from Th3 level up to the medulla oblongata.

Results: Lack of cerebrovascular risk factors, clinical findings and neuroimaging findings made FCE the most likely cause.

Conclusion: In case of sudden progressive quadriplegia and severe neurologic deterioration, one has to take rare syndromes into consideration. Urgent MRI scan including angiography may help to distinguish this syndrome and start treatment. Prognosis for recovery is uncertain.

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Abstract – WCN 2013

No: 1903

Topic: 3 – Stroke

Correlation of electrophysiologic abnormality with motor recovery in stroke

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Objectives: The objective was to determine whether the abnormal findings of somatosensory evoked potentials (SEPs) and motor nerve conduction study correlate with Motor Recovery in Stroke.

Methods: Eighty-two stroke patients were enrolled in this study from January, 2011 to December, 2011. The initial clinical data were reviewed including age, gender, main type of stroke (ischemic or hemorrhagic), lateralization of the lesion (right, left, or bilateral), location, motoricity index, MBI and hand function test. SEP tests (N20 in median nerve, P37 in posterior tibial nerve) and motor nerve conduction study (median, lateral, common peroneal, and tibial nerve) were performed. The amplitude of hemiside motor amplitude was compared with that of sound side and confirmed the relationship among other parameters.

Results: Relative value of median and tibial motor amplitude correlated significantly with the hemiplegic median SEP, posterior tibial SEP, and initial MBI. And the absent median SEP was correlated significantly with MFT ($p < 0.05$). Binary logistic regression analysis of factors showing significant correlations with final MBI indicated with relative value of motor amplitude.

Conclusion: In conclusion, relative value of motor amplitude as well as SEP was a primary predictor of motor recovery in the sub-acute phase of stroke.

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Abstract – WCN 2013

No: 2123

Topic: 3 – Stroke

Septic hydatid cerebral emboli with a subarachnoid hemorrhage: A case report

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Introduction: Hydatid disease is a parasitic disease caused by the larval stages of *Echinococcus granulosus* and the most common sites of involvement are the liver and lungs. hydatidosis is a rare disease even in the endemic areas and though most of the patients may remain asymptomatic, a number of serious and lethal complications have been described.

Case report: We present a patient with cardiac hydatidosis leading to a septic cerebral embolus accompanied by a subarachnoid hemorrhage, probably due to the rupture of a mycotic (embolic-septic) aneurysm.

Discussion: Cerebral hydatid cysts are relatively rare and account for up to 2% of cases. Cerebral involvement generally occurs secondary to dissemination from a cardiac source. Mycotic aneurysms also known as infective or microbial aneurysms are rare inflammatory neurovascular lesions that count for 0.7–6.5% of all intracranial aneurysms. Because their spontaneous rupture results in subarachnoid and intracerebral hemorrhage, they are associated with significance and morbidity. The possibility of hydatid disease should be kept in mind, especially in endemic zones.

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Abstract – WCN 2013

No: 2110

Topic: 3 – Stroke

Caveolin-1 phosphorylation plays an important role in inhibition of oxygen-glucose-deprivation-induced endothelial cell apoptosis via regulating STAT3 pathway

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Background: Caveolin-1 is abundantly expressed in endothelial cells and serves as a “master regulator” of signal molecules. Our previous study suggests that caveolin-1 could protect blood brain barrier (BBB) from ischemia-reperfusion injury through regulation of MMP activation. However, whether caveolin-1 could protect brain endothelial cells under hypoxic/ischemic conditions is unknown yet.

Objectives: In order to explore the potential mechanisms of caveolin-1 in protection of BBB, we investigated the roles of caveolin-1 in protection of brain endothelial cells from oxygen and glucose deprivation (OGD)-induced endothelial apoptosis, and related mechanisms.

Material and methods: Brain microvascular endothelial b.End3 cells were subjected to OGD for 6 h. siRNA was performed to partially knock down caveolin-1. Western blot analysis was used to determine the expression of P-CAV-1, CAV-1, Stat3, p-Stat3, Bcl-2, Bax, caspase-3, active caspase 3. Hoechst staining and flow cytometry were used to detect cell apoptotic cell death.

Results: OGD treatment down-regulated phosphorylation of caveolin-1 and p-Stat3, up-regulated the ratio of Bax/Bcl-2 and activated caspase-3, and subsequently inducing apoptotic cell death. Interestingly, partial silence of caveolin-1 remarkably down regulated p-Stat3 and aggravated the rates of apoptosis in the OGD treated cells.

Conclusion: Phosphorylation of Caveolin-1 could attenuate OGD-induced apoptotic cell death in brain microvascular endothelial cells and its mechanisms could be associated with p-Stat3 pathway.

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Abstract – WCN 2013

No: 1749

Topic: 3 – Stroke

Acute stenting in treatment of acute ischemic stroke

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Background: Acute stenting alone or in combination with other modalities is one possibility for acute stroke treatment in certain indications.

Objective: The purpose of our study is to demonstrate the results of acute stenting in 44 patients with acute ischemic stroke during January 2006 and December 2012 in our hospital.

Patients and methods: We retrospectively reviewed 44 patients (36 males, 8 females, median 58 years) with acute stroke caused by occlusion or severe stenosis of large artery of anterior (35 patients) or posterior circulation (9 patients). Patients underwent acute stenting alone or in combination with intravenous thrombolysis (IVT), percutaneous angioplasty (PTA), intraarterial thrombolysis (IAT) or mechanical thrombectomy (MT). The procedure was performed within 8 h after stroke onset. We determined median score of National Institute of Health Stroke Scale (NIHSS) at baseline and after 24 h and modified Rankin score (mRS) in 3 months. The Thrombolysis in Myocardial Infarction (TIMI) reperfusion score was assessed by angiography performed immediately after each procedure.

Results: The median NIHSS score at baseline was 14 (range 4–25), after 24 h 8 (range 0–35). Good clinical outcome in 3 months (mRS 0–2) had 29 patients (65.9%). The mortality rate was 22.7% (mRS 6). Complete recanalization (TIMI 3) was achieved in 23 patients (52.3%). Symptomatic intracranial hemorrhage occurred in 8 cases (18.0%).

Conclusion: Acute stenting has a high recanalization rate. In our study late stent implantation did not show higher risk of complications.

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Abstract – WCN 2013

No: 2132

Topic: 3 – Stroke

Peroxyntirite could regulate proliferation and neuronal differentiation of neural stem/progenitor cells through activating WNT/ β -catenin signaling pathway

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Background: Hypoxia/ischemia could mediate the differentiation of neural stem/progenitor cells (NSCs) into mature neurons. In hypoxic/ischemic brain, nitric oxide and superoxide are simultaneously produced and they rapidly react to form peroxyntirite. Whether peroxyntirite can regulate neurogenesis is unknown yet.

Objectives: Present study aims to understand the roles of peroxyntirite in regulating self-renewal, proliferation and differentiation of NSCs.

Materials and methods: Primary cultured NSCs were subjected to different stimulations including peroxyntirite donor SIN-1, synthesized peroxyntirite and hypoxia treatment. For visualizing the formation of peroxyntirite, we developed a highly sensitive and specific fluorescent

probe and detected the formation of peroxyntirite in the NSCs. We applied different biomarkers including Ki67, BrdU, Tuj1 and DCX, etc. to identify the self-renewal proliferation, and neuronal differentiation of NSCs respectively.

Results: Low concentrations of extraneous peroxyntirite (<1 μ M) promoted NSC proliferation, self-renewal and neuronal differentiation but high level of peroxyntirite (>5 μ M) induced cytotoxicity. Hypoxic treatment induced the production of peroxyntirite and promoted NSC proliferation and self-renewal, and neuronal differentiation. Treatments of peroxyntirite decomposition catalysts (PDCs, FeTMPyP and FeTPPS) reduced hypoxia-induced peroxyntirite formation, NSC proliferation, self-renewal and neuronal differentiation. The neurogenesis promoting effects were partly mediated through activating Wnt/ β -catenin signaling pathway.

Conclusion: Low concentration of peroxyntirite may serve as a cellular signaling for promoting NSC proliferation, self-renewal and neuronal differentiation.

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Abstract – WCN 2013

No: 2125

Topic: 3 – Stroke

Perfusion defect and initial clinical status might predict the poor prognosis of DWI-negative ischemic stroke

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Purpose: Clinically acute stroke is not always accompanied by positive lesion on the diffusion-weighted image (DWI). In this diffusion negative ischemic stroke (DNIS), the clinical prognosis would be difficult to predict. We investigated which factors are associated with the clinical course in the DNIS.

Methods: We retrospectively reviewed data from prospectively collected Inha University Hospital Stroke Registry from January 2007 to December, 2010. We identified and divided the DNIS into two groups, according to the persistence of symptoms after seven days, and analyzed the differences in clinical and imaging factors between two groups.

Results: A total of 1398 subjects were enrolled and 142 DNIS patients were identified. Compared to DWI(+) group, the patients with DNIS were younger, and had the low frequency of atrial fibrillation and hypertension. Among DNIS, the patients with fully recovery within seven days, had the lower ABCD2 score, the lower NIHSS on admission, and the lower frequency of perfusion delay on perfusion MRI, compared to the patients with persistent symptoms. In the multivariable analysis, initial NIHSS(≥ 1) and perfusion delay were independently associated with persistent symptoms (NIHSS, $p < 0.001$, OR 17.2, 95% CI 5.5–54.0; perfusion delay, $p = 0.03$, OR 36.7, 95% CI 1.4–977.0).

Conclusions: Our results suggested that the perfusion defect and focal neurological deficit might be related to the symptom persistence in the DNIS.

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Abstract – WCN 2013

No: 2108

Topic: 3 – Stroke

The sex hormone ratios of acute cerebral infarction

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Background: Sex hormones may be associated with higher incidence of clinically significant stroke or stroke related events. The purpose of the present study was to assess the role of sex hormone ratio in acute ischemic stroke.

Methods: Between January 2011 and March 2013, a total of 114 patients with acute cerebral infarction or transient ischemic attack, and 96 control subjects were included in this study. Sex hormones including estradiol, estrogen, testosterone, free testosterone and progesterone of all patients were investigated. We analyzed sex hormone ratio of these patients.

Results: In men, compared with control group, estradiol/testosterone (E/T) ratio and estradiol/free testosterone (E/T free) ratio were significantly elevated in the stroke patient group. ($P = 0.017$ and $P = 0.014$). On the contrary, there were no evidence for an association between ischemic stroke and E/T, E/T free ratio in women.

Conclusion: In men, higher E/T or E/T free ratio was associated with ischemic stroke. Other sex hormone ratios were not related with acute ischemic stroke. These findings support the hypothesis that increased estradiol and reduced testosterone were associated with ischemic stroke, particularly in men.

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Abstract – WCN 2013

No: 2142

Topic: 3 – Stroke

Unusual central nervous system hemorrhagic complications associated with dengue fever

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Background: Dengue viral infection is emerging as significant public health problem, particularly in developing world. The dengue infection mainly presented with classical dengue fever, dengue hemorrhagic fever, and dengue shock syndrome.

Objectives: We intended to report spontaneous intracranial hemorrhage, associated with dengue virus infection without other bleeding manifestations.

Patient and methods/material and methods: The clinical details and outcome were described in two patients of CNS hemorrhagic manifestations with dengue fever. Both the patients were serum positive for IgM capture ELISA for dengue fever and showed fourfold increase in titer subjected one week apart. There was no history of trauma.

Results: In Case-1, a 28 years old male was admitted with complaint of high grade fever, malaise and severe bone pain for 15 days. Later on, he developed acute headache, vomiting and altered sensorium with the presence of thrombocytopenia. The computerized tomography head demonstrated acute subdural hematoma with associated subarachnoid hemorrhage. Bilateral fronto-parietal craniotomy with right sided evacuation of acute SDH was done. The patient recovered completely at follow-up.

In Case-2, a 19 years old male reported with complaints of fever, retroorbital pain, arthralgia and thrombocytopenia. He subsequently developed headache, seizures and unconsciousness. Magnetic Resonance Imaging of the brain revealed acute extradural hematoma. The patient was managed conservatively and was completely improved at follow-up after 3 months.

Conclusion: Our case series illustrate that spontaneous CNS hemorrhage associated with dengue fever can occur without other bleeding

manifestation and it required a high index of suspicion to have good clinical outcome.

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Abstract – WCN 2013

No: 1882

Topic: 3 – Stroke

Cerebral vascular accident caused by intracranial aneurysm: Evaluation of fifteen cases

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Background: Hemorrhagic stroke due to intracranial aneurysm (HS-IA) is caused by the rupture of aneurysms predominantly formed in the anterior circulation (90%), which leads to subarachnoid hemorrhage. HS-IA has a higher incidence around the age of 50, and it is more common in women than in men. The clinical features are sudden headache, dizziness, hemiplegia, and nausea, among others. Complications are frequent: vasospasm, seizures, rebleeding, and hydrocephalus. **Objectives:** Investigate the characteristics of patients with HS-IA, comparing with literature.

Methods: A retrospective review of 15 patients admitted with the diagnosis of HS-IA in Santa Monica Hospital, Goiânia – Brazil, from July 2008 to January 2009 was performed.

Results: Among the 15 cases of HS-IA analyzed, a peak occurrence between 51 and 60 years was observed. There were 9 women and 6 men. The most common affected arteries were: middle cerebral (23%), ophthalmic (23%), anterior communicating (18%), posterior communicating (18%), internal carotid (12%), and posterior inferior cerebellar (6%). The most common symptoms were: 86.7% headache, 33.3% vomiting, 33.3% change in the consciousness, 33.3% visual impairment, and 20% nuchal pain. Of the 14 patients undergoing surgery, 8 patients had no complications, while the other 6 had different frames: headache, facial nerve paresis, neck hematoma and hypertensive peak; spasm, motor impairment and atrophy of the left temporal muscle, and paresis of the lower limbs.

Conclusion: In this study, the data obtained are consistent with the epidemiological literature, except in the category of postoperative complications, proving that the profile of the patient is important to recognize a HS-IA.

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Abstract – WCN 2013

No: 2137

Topic: 3 – Stroke

Cranial nerve syndromes and vision loss in cerebral venous thrombosis: A rarity

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Background: Cerebral venous thrombosis (CVT) can present as raised intracranial tension, focal deficits, cavernous sinus thrombosis, sub-acute encephalopathy and cranial nerve syndrome. Cranial palsy may be seen in 10–12%. CVT presenting as cranial nerve palsy and vision loss is rare.

Objective: To analyse patients of CVT presenting with cranial nerve palsies and vision loss due to raised intracranial tension.

Patients and methods: Retrospective analysis of CVT patients admitted in stroke unit from 2004 to 2012 was performed from medical records of our university hospital.

Results: Among 405 patients, 8 patients of CVT had combination of visual loss and cranial nerve palsies. Mean age at presentation was 27.6 years. Male:Female: 5:3. The most common risk factor was elevated homocysteine levels ($n = 4$). Duration of presentation ranged from 4 days to 1 year. Patients presented with headache ($n = 7$), neck pain ($n = 7$), seizures ($n = 4$), papilledema ($n = 8$), visual impairment ($n = 8$), visual field defects ($n = 5$), bilateral 3,4 and 6th cranial nerve palsy ($n = 5$), LMN facial palsy ($n = 4$), bilateral 6th nerve palsy ($n = 3$) and radiculopathy ($n = 4$). None had focal deficits.

CSF pressure ranged from 235 to 400 mm of Hg. Superior sagittal sinus and transverse sinus thrombosis were the commonest sites of thrombosis by MR venography. Treatment included anti-oedema measures, methyl prednisolone ($n = 4$) and 5000 IU subcutaneous heparin 6th hourly followed by acenocoumarol. Optic nerve fenestration ($n = 3$) followed by thecoperitoneal shunt ($n = 4$) was undertaken in those with significant vision loss and headache, though the outcome remained poor.

Conclusion: Cerebral venous thrombosis can have diverse presentations. CVT with vision loss and cranial nerve palsy may be misdiagnosed as chronic meningitis, pachymeningitis or vasculitis.

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Abstract – WCN 2013

No: 2039

Topic: 3 – Stroke

Recovery of bimanual coordination is delayed after left hemispheric and/or purely cortical stroke lesions

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Background: One-fit-all post-stroke rehabilitation has limited plausibility. Better understanding the effects of lesion location on the recovery process may help to develop rehabilitation strategies adapted to individual patients' needs.

Objective: The purpose of this study was to assess the natural evolution of bimanual coordination over standard rehabilitation aiming to identify a potential responder-profile for bimanual-oriented therapy.

Patients and methods: 12 hemiparetic, moderately impaired patients were included within 30 days after a first unilateral stroke. Kinematic and clinical assessments were performed once a week for 6 weeks and at 3 months after inclusion. Patients performed a synchronous bimanual reach-to-grasp task. Clinical evaluation included Fugl-Meyer Assessment, Box and Block test, 9-Hole Peg test and Barthel Index.

Results: Although no clinical difference was found between groups, left-hemisphere damaged (LHD) patients tended to reach less smooth than right-hemisphere damaged (RHD) patients ($p = .07$). Moreover, it was found that patients with purely cortical damage moved slower than patients with subcortical lesions ($p < .01$). These kinematic inter-group differences disappeared, respectively, after 4 and 6 weeks of standard therapy, likely indicating a shift in the recovery phase.

Conclusion: Because patients with a subcortical lesion/and or right hemispheric lesion were naturally more coordinated during bimanual movements over recovery, they may benefit more from bimanual rehabilitation.

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Abstract –WCN 2013

No: 1471

Topic: 3 – Stroke

Troponin-T in acute ischemic stroke patients – Results of the prospective history study

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Background: Multiple interactions are considered to occur between the various forms of cardiovascular and cerebrovascular diseases. The aim of the study was to assess the serum level profile of cardiac troponin T (cTnT) in acute ischemic stroke (AIS) patients to evaluate factors associated with increased serum level of cTnT.

Methods: AIS patients admitted within 12 h from stroke onset were enrolled in this prospective study. Neurological examination and brain CT or MRI at admission, standard laboratory tests, including cTnT and other cardiac markers, at admission and 4 h later, and repeated electrocardiograms (ECGs) were performed. Correlations between cTnT and several baseline parameters were tested and multivariate regression analysis was used to assess the predictors of cTnT elevation.

Results: In total, 107 consecutive AIS patients (65 males, mean age 67.2 ± 14.2 years) were enrolled. 39 (36.4%) patients presented with elevated cTnT above the upper limit. The cTnT levels correlated significantly with age ($r = 0.448$) and the levels of NT-proBNP ($r = 0.528$), cystatin C ($r = 0.457$), CK-MB mass ($r = 0.253$), urea ($r = 0.281$), and albumin ($r = -0.219$). Multiple logistic regression analysis found creatinine $>90 \mu\text{mol/L}$ (OR = 3.45; 95% CI: 1.09–10.85), NT-proBNP (OR = 1.09 per 100 $\mu\text{g/L}$ increase; 95% CI: 1.03–1.16) and CK-MB mass (OR = 1.45 per 1 $\mu\text{g/L}$ increase; 95% CI: 1.04–2.04) to be associated with cTnT elevation in AIS patients.

Conclusion: Elevated cTnT can be relatively frequently detected in AIS patients. To reliably identify the patients with current acute myocardial impairment, more in-depth clinical investigation is needed.

Trial registered at <http://www.clinicaltrials.gov> (no. NCT01541163). Supported by the IGA MH CR grant number NT/11046-6/2010.

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Abstract – WCN 2013

No: 2139

Topic: 3 – Stroke

Stroke outcome after endovascular treatment: Influence of the site of occlusion

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Background: After IV thrombolysis, patients with occlusion of both M1 and ICA (tandem occlusion) have a worse outcome than patients with isolated M1 occlusion (iM1 occlusion).

Objective: We aimed to evaluate the influence of the site of occlusion on stroke outcome after endovascular treatment.

Patients and methods: We included consecutive patients treated by endovascular procedure because of a stroke related to an occlusion of M1, isolated or associated with an ICA occlusion. Mechanical

thrombectomy was performed using a stentriever (Solitaire FR). We evaluated clinical severity of stroke (NIHSS), lesion volume (pre-treatment diffusion MRI), haemorrhagic complications (T2*MRI at 24 h (ECASS II)), recanalisation at the end of the procedure (TICI criteria), and 3-month outcome (mRS). Favourable outcome was defined as a mRS ≤ 2 or identical to pre-stroke mRS.

Results: Among 28 consecutive patients (median age 58.5 years, 8 men, median NIHSS 18, 21 treated by IV rtPA), 14 had iM1 occlusion, 14 tandem occlusion. Demographics, NIHSS, stroke volume, delay and duration of endovascular procedure, and number of patients with IV rtPA did not differ between groups. The rates of recanalisation of M1, of hemorrhagic complications and of favourable outcome at month-3 (57% in iM1 occlusion and 64% in tandem occlusion) were similar in the 2 groups of patients. Multivariate analysis found infarct volume to be the only independent predictor of 3-month outcome.

Conclusion: After endovascular treatment, 3-month outcome was similar in patients with tandem occlusion compared to patients with iM1 occlusion, without clear influence of the site of occlusion.

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Abstract – WCN 2013

No: 2194

Topic: 3 – Stroke

Alcohol attributable fraction of stroke mortality in Russia

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Background: Stroke is an international health problem with high associated human and economic costs. The mortality rate from stroke in Russia is one of the highest in the world. Risk factor identification is therefore a high priority. Epidemiological evidence suggests that binge drinking increases the risk for ischemic and hemorrhagic stroke.

Objectives: The aim of the present study was to estimate the premature stroke mortality attributable to alcohol abuse in Russia on the basis of aggregate-level data of stroke mortality and alcohol consumption.

Methods: Age-standardized sex-specific male and female stroke mortality data for the period 1980–2005 and data on overall alcohol consumption were analyzed by means ARIMA time series analysis.

Results: The results of the analysis suggest that 26.8% of all male stroke deaths and 18.4% female stroke deaths in Russia could be attributed to alcohol. The estimated alcohol-attributable fraction for men ranged from 16.2% (75+ age group) to 57.5% (30–44 age group) and for women from 10.3% (75+ age group) to 43.5% (30–44 age group).

Conclusions: The outcomes of this study provide support for the hypothesis that alcohol is a major contributor to the high stroke mortality rate in Russian Federation. Therefore prevention of alcohol-attributable harm should be a major public health priority in Russia. Given the distribution of alcohol-related stroke deaths, interventions should be focused on the young and middle-aged men and women.

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Abstract – WCN 2013

No: 2106

Topic: 3 – Stroke

Perilesional and subcortical plasticity after focal ischemic stroke is associated with motor recovery: A tensor-based morphometry study

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Background: In preclinical models of cortical sensorimotor stroke, extensive remodeling of perilesional and subcortical gray matter (GM) has been described. It is unknown whether these morphological changes can be detected in clinical populations as well.

Objective: To measure the longitudinal GM plasticity in stroke survivors using MRI, and to identify its relationship with motor recovery.

Patients and methods: We tracked motor recovery in 28 patients suffering from hand paresis after cortical sensorimotor stroke with a detailed battery of hand function tests. We used a longitudinal principal component analysis to quantify and classify recovery subgroups. To assess GM volume change, we performed tensor-based morphometry and statistical parametric mapping on high-resolution T1-weighted MRI scans acquired between 3 and 9 months post-stroke. Significance was estimated using threshold-free cluster enhancement. Cytoarchitectonic regions-of-interest were used for structure–behavior correlations and subgroup comparisons.

Results: Multivariate classification yielded three recovery subgroups. We found significant GM expansion in perilesional premotor cortex, ipsilesional caudate and ipsilesional mediodorsal thalamus, and GM atrophy in the contralateral cerebellum. A 3×2 -ANOVA with recovery subgroup (fast, slow, impaired) and site of plasticity (premotor, thalamic) as between-subject factors revealed a significant interaction effect ($F(2,106) = 3.837$, $p = .025$): fast recoverers showed more perilesional, impaired recoverers more thalamic GM expansion. Thalamic GM change was significantly correlated with chronic motor impairment ($r = .829$, $p = .011$).

Conclusions: Subcortical GM plasticity can be detected non-invasively during subacute phases of recovery, and correlates with post-stroke motor outcome. This might reflect an adaptive, but insufficient, shift of motor control from pyramidal to extrapyramidal circuits.

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Abstract – WCN 2013

No: 2203

Topic: 3 – Stroke

Acute ischemic stroke with medial cerebral artery hyperdense sign – Comparison of intravenous thrombolysis and catheter-based therapy: A pilot study

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Introduction: Acute ischemic stroke due to occlusion of medial cerebral artery (MCA) has an unfavourable prognosis in spite of intravenous rtPA administration. Alternative treatment options are still a matter of debate. In this study, we compared the results of rtPA and mechanical thrombectomy in the matched study groups.

Material and methods: Ten patients underwent a direct mechanical thrombectomy (catheter-based therapy, CBT group) of MCA occlusion and were compared to 10 matched individuals, who were treated for acute ischemic stroke by intravenous thrombolysis (IVT group). All 20 patients had hyperdense sign on initial non-contrast CT scan. IVT group (8 females, average 58 yrs, range 38–79) had an initial NIHSS of 15 points. CBT group (8 females, average 59 yrs, range 32–79) had the initial NIHSS of 14 points. Mean onset to needle time was 124 min in IVT group and 140 min in CBT group.

Results: Early MCA recanalization was reached in nine subjects in the CBT group and in three patients in IVT group as assessed by ultrasound. Symptomatic intracerebral haemorrhage occurred in one subject in CBT group and in two patients in IVT group. Favourable outcome (mRS 0–2) on acute hospital discharge was achieved in five individuals treated by CBT and in two persons, who received IVT.

Conclusion: This pilot study showed promising outcomes (angiographic and clinical) after direct CBT when it is initiated with minimal delay. Further research comparing direct CBT versus IVT is warranted.

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Abstract – WCN 2013

No: 2165

Topic: 3 – Stroke

Functional condition of central and cerebral hemodynamic in patients with coma caused by acute ischemic stroke

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The aim of study was to evaluate the functional condition of central hemodynamic and cerebral blood flow in acute ischemic stroke comatose patients.

Objective: The continuous simultaneous assessment of invasive central hemodynamic and intracranial pressure (ICP), transcranial Doppler measurement of cerebral hemodynamic was performed in 35 comatose ischemic stroke patients on 1, 3, 5 and 7th day of acute period.

Material and methods: The cardiac index (CI) was significantly higher on 3rd and 7th day of acute period. The mean arterial pressure (MAP) was increased.

Results: The systemic vascular resistance index (SVRI) was higher than normative range. ICP was significantly higher on 3rd and 5th day in comparison with 1st day ($p < 0.05$ – 0.001) and was higher than normative range value. Cerebral perfusion pressure (CPP) was assessed in normative range value and on 7th day it was 96.6 ± 2.12 mm Hg. Time average maximum velocity (TAMX) in middle cerebral artery on stroke side increased significantly on 7th day ($p < 0.05$ – 0.001). Pulsatility index (PI) on 3rd day was 1.2 time higher than 1st day value and was 1.16 ± 0.07 and it was a result of increased ICP. The maximal value (0.64 ± 0.02) of the resistive index (RI) was on 3rd day ($p < 0.05$ – 0.001).

Conclusions: The reaction of central hemodynamic was aimed to provide an adequate cerebral perfusion. The elevation of cerebral blood flow and mean arterial pressure on 7th day of acute period of ischemic stroke was a result of cerebral autoregulation impairment.

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Abstract – WCN 2013

No: 2219

Topic: 3 – Stroke

Quality of care registry of cerebrovascular disease in Veneto region: One year of experience in monitoring and implementing stroke care

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Objective: To pilot and promote the use of a regional “quality of care register” for cerebrovascular disease to document procedures, treatments, compliance with evidence-based indications, and to guide improvements in ischemic stroke patient management.

Materials and methods: The multicentric, observational prospective study, funded by the Regional Government of Veneto with the participation of Pfizer Italia, was designed to collect data on patients admitted to Ist and IInd level SUs for cerebrovascular diseases in the Veneto Region from 1/09/2011 to 31/8/2012. Patients' complete care pathway was assessed from the pre-hospital phase, to the emergency department, to admission to SU, discharge and follow-up.

Results: During the collection period 3357 admissions for cerebrovascular diseases were recorded. Of the ischemic stroke patients, 75% reached the ER and underwent a brain CT within 4.5 h of symptoms onset and were thus in time to undergo reperfusion therapy, but thrombolysis involved only 18% of all ischemic stroke. In terms of compliance with the main guidelines, the study revealed poor use of the leading standardized neurological assessment scales (Rankin and NIHSS), and that a high percentage of patients were not taking adequate preventive treatment, a significant percentage of patients were not recommended to take statins after ischemic stroke (46%), and patients with cardioembolic stroke were not discharged with anticoagulant therapy (46%).

Conclusions: The system for monitoring of patients with cerebrovascular pathology is designed both to identify structural weaknesses in the stroke pathway requiring organizational/legislative intervention, and to evaluate the quality and standardization of the care pathway.

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Abstract – WCN 2013

No: 2211

Topic: 3 – Stroke

Predictors of early post-stroke seizures in patients with spontaneous intracerebral hemorrhage

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Background: Seizures are common in patients with spontaneous intracerebral hemorrhage (ICH). Understanding of the risk factors of seizures following ICH is needed to predict which patient will require treatment.

Objective: To determine the factors predicting the occurrence of early post-stroke seizures in patients with spontaneous ICH.

Methods: Seventy patients (43 males and 27 females) with first time spontaneous ICH were included in the study. Their age ranged from 27 to 90 years (mean 58.5 ± 11.1). All the patients were subjected to clinical examination, laboratory investigations (including early morning serum cortisol level), computed tomography brain scan at the time of admission and after 48 h, and electroencephalography. Patients were followed up for 7 days for the occurrence of early seizures.

Results: On the follow-up period, 13 patients (18.6%) developed early post-stroke seizures. Of them, 4 patients (30.7%) had focal seizures and 9 patients (69.3%) had generalized seizures. Patients with early post-stroke seizures were significantly more likely to have Glasgow Coma Scale (GCS) <12, high systolic and diastolic blood pressure, high body temperature, high random blood glucose level, lobar cortical hematoma, hematoma volume >30 cc, peri-hematoma edema, midline shift, hematoma expansion after 48 h and periodic lateralized epileptiform discharges (PLEDs) than patients without seizures.

Conclusions: GCS <12, high systolic and diastolic blood pressure, high body temperature, high random blood glucose level, lobar cortical hematoma, hematoma volume >30 cc, peri-hematoma edema, midline shift, hematoma expansion after 48 h and PLEDs can be considered as predictors of early post-stroke seizures.

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Abstract – WCN 2013

No: 2214

Topic: 3 – Stroke

A case of anterior spinal artery syndrome (ASAS) demonstrated by angiography

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A 27-year-old woman experienced pain, paresthesia and weakness after her mother made massage to her neck. Nearly 1 h after, her upper extremities became plegic and her lower extremities became weaker and 5 h later she became totally quadriplegic. Neurological examination revealed extreme neck pain, quadriplegia, decreased deep tendon reflexes, sensory disturbance of pain and temperature below the neck and urinary incontinence. MRI T2 weighted images showed a high-intensity lesions which did not enhance after contrast material at C4–7 segments. Vasculitic markers were negative. Echocardiography was normal. Serum viral markers were negative. In CSF, cell count and biochemistry was normal, viral markers were also negative. ASAS was diagnosed according to the owl's eye appearance which is large, round, single, central lesion locating in the anterior side of the spinal cord supporting an ischemic disease in cervical diffusion MRI. Angiography was performed and it showed that vertebral arterial flow was intact but there was an interruption in the blood flow of anterior spinal arteries at the 4–7th cervical segments. ASAS is a very rare entity and its MRI findings such as the "owl's eye" appearance is specific and demonstrative and angiographic demonstration of occlusion of anterior spinal arteries is relatively rare. We report this case because it has typical clinical and MRI findings and could also be demonstrated by angiography.

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Abstract – WCN 2013

No: 1246

Topic: 3 – Stroke

The association of sleep quality in ischemic stroke patients

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Background: Sleep-disordered breathing (SDB) represents both a risk factor and a consequence of stroke.

Objective: To determine the diversity of sleep quality in patients with different TOAST classification.

Method: With cross-sectional study, 61 patients came from the Chang-Bing Show-Chwan Memorial Hospital. Utilize the structural formula questionnaire to collect demography and polysomnography, describes the materials centralized and dispersing the situation in terms of average, standard deviation, percentage of number of times. Use one-way ANOVA pays inference statistics.

Results: In this study, our male patients are about 55.7% and mean age is 63 ± 12.7 , which has $26.1 \pm 4.5 \text{ kg/m}^2$ in Body mass index (BMI), and Apnea-hypopnea Index (AHI) is 15.2 ± 17.8 . With the analysis of related risk factor in AHI and Athens Insomnia Scale (AIS), it has been showed that BMI in sleep disorder patients is higher than that in normal people (all p values < 0.05). It also shows that patients with sleep disorder have the problem with overweight (BMI $\geq 24 \text{ kg/m}^2$). Furthermore, the potential risk factors showed that cardioembolism is higher in BMI and AHI (all p values > 0.05). However, large artery atherosclerosis is higher in AIS and Epworth Sleepiness Scale (all p values > 0.05). These data showed significance with sleepiness in daytime, but no significance with age, smoking behavior and alcohol intake.

Conclusion: The sleep quality would be affected in overweight people. Moreover, sleepiness in daytime in different TOAST classifications of stroke had been confirmed.

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Abstract – WCN 2013

No: 2238

Topic: 3 – Stroke

Quality control of carotid stenting with digital subtraction angiography and computed tomography angiography: Similarities and differences

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Background: The effect of carotid stenting must be continuously monitored to ensure good patient outcomes. Mostly this is done by means of digital subtraction angiography DSA. Increasingly, computed tomography angiography CTA is used as an alternative. To assess differences between both methods, we retrospectively compared DSA and CTA after carotid stenting.

Patients and methods: Retrospective analysis of pre- and post-interventional CTA and DSA in carotid stenting patients. We found 59 patients with complete records. Two readers independently assessed the images on PACS workstations and measured carotid diameters according to NASCET criteria. Stenosis as well as technical success rates (improvement over 20% and less than 50% residual stenosis) and interrater reliability and correlations were computed for both methods.

Results: Mean preinterventional stenosis rates were 69.9% for CTA vs 59.3% for DSA, postinterventional stenosis was 20.5 vs. 17.4% respectively. Technical success rates were calculated for DSA to be 97.5% and for CTA 85.6%. The correlation of both methods was higher preinterventionally (.69, $p < .0001$) than postinterventionally (.48, $p = .0001$). Interrater reliability was mostly moderate to good (lambda .46–.79).

Conclusion: Due to the volumetric dataset with the possibility to perform measurements orthogonal to the vessel and to assess the minimal diameter independent from projection angles of DSA, the measurement of preinterventional stenosis was deemed more

accurate with CTA, thus giving higher stenosis rates. Postinterventional assessment with DSA is prone to a bias since the intervention is performed under control in one projection. CTA however demonstrated regularly less optimal results in other projections.

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Abstract – WCN 2013

No: 617

Topic: 3 – Stroke

Optimized amide proton transfer imaging of ischemic stroke

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Objective: The identification of ischemic penumbra is very important to guide the treatment in ischemic stroke. Amide proton transfer (APT) imaging, a new MRI technology, is expected to detect the ischemic penumbra more precise than PWI–DWI mismatch in hyperacute cerebral infarction. But APT effect is typically small and sensitive to pre-saturation power, which needs to be optimized in the first 3 h after stroke.

Material and methods: Twelve adult male Sprague Dawley rats underwent permanent middle cerebral artery occlusion by thread embolism. MRI experiments were conducted under an Agilent 7T animal MRI system in the first 3 h after ischemic stroke. APT imaging was obtained by the home-made APT sequence. Parameters were used: Offset at 3.5 ppm and –3.5 ppm, TR/TE = 26/2.5 ms, FA = 5°, slice thickness = 2 mm, FOV = 34 × 34 mm², NEX = 64, matrix = 64 × 64 and bandwidth = 50 kHz. APT MRI was optimized as functions of pre-saturation power. The pulse power (Gauss pulse, 15 ms) was serially set from 0.4 μT to 1.6 μT with intervals of 0.2 μT. Amide proton transfer imaging and contrast to noise rate (CNR) were processed in Matlab.

Results: The optimized pre-saturation power in the first 3 h was 0.6 μT, which had better spatiotemporal resolution and contrast than others. The best CNR was about 4. Moreover, the APT effect almost disappeared when the pre-saturation power was varied more than 0.4 μT.

Conclusion: Our study demonstrates the optimized APT imaging in the first 3 h of ischemic stroke, which showed the ischemic lesion precisely and be promising for an accurate gauge of ischemic penumbra.

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Abstract – WCN 2013

No: 2248

Topic: 3 – Stroke

Endovascular treatment of acute basilar artery occlusion

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Background: Acute ischemic stroke (AIS) caused by basilar artery occlusion (BAO) is often associated with a severe and persistent neurological deficit and a high mortality rate. Nevertheless, the most effective therapeutic approach has not been established yet.

Objective: To evaluate safety and efficacy of multimodal endovascular treatment (ET) of acute BAO, including bridging therapy (intravenous thrombolysis [IVT] with subsequent ET).

Patients and methods: In the retrospective, bicenter study, the set consisted of 37 AIS patients (29 males; mean age 60.8 ± 9.2 years) with radiologically confirmed BAO. Following data was collected: baseline characteristics, risk factors, pre-event antithrombotic treatment, neurological deficit at time of treatment, time to therapy, recanalization rate (with successful recanalization defined as Thrombolysis in Cerebral Infarction score 2–3), post-treatment imaging findings. 90-Day outcome was assessed using modified Rankin scale (mRS) with good clinical outcome defined as 0–3 points.

Results: The mean National Institutes of Health Stroke Scale score at presentation was 21.3 ± 8.3 points. The mean time to treatment was 5.31 ± 0.13 h. In 17 bridging group patients, mean time from IVT to ET 1.39 ± 0.05 h. Successful recanalization was achieved in 89.2% patients. In patients with BAO recanalization versus those without recanalization, good 90-day clinical outcome was observed in 36.4% versus 0% (p > 0.05), 90-day mortality was 30.3% versus 75.0% (p > 0.05) and mean 90-day mRS was 3.80 versus 5.75 points (p > 0.05).

Conclusion: Data in this series showed that multimodal ET was an effective recanalization method of acute BAO. BAO recanalization was associated with lower mortality, but not necessarily with good clinical outcome.

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Abstract – WCN 2013

No: 2234

Topic: 3 – Stroke

Influence of acute aneurysmal subarachnoid hemorrhage on subpopulations of mononuclear cells

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Background: Aneurysmal subarachnoid hemorrhage (aSAH) causes specific changes in transcription profiles of peripheral blood cells with a downregulation of transcripts related to T lymphocytes and an upregulation of transcripts related to monocytes and neutrophils. However, it is uncertain whether those changes reflect changes in cell count or in their activities.

Objective: To analyze changes in subpopulations of mononuclear cells in the acute phase of aSAH.

Patients and methods: Nine patients with acute nontraumatic aSAH (7 females) and 10 control subjects (8 females) were recruited into the study. Flow cytometry method was used to analyze following subpopulations of leukocytes: T lymphocytes (CD3+, CD4+, CD8+, invariant natural killer T cells [iNKT]) and monocytes (classical CD14++CD16-, intermediate CD14++CD16+, nonclassical CD14+CD16++).

Results: Significant changes in leukocyte composition with an increase of granulocyte percentage (85.3% vs 63.3%) and a decrease of monocyte (4.9% vs 6.2%) and T lymphocyte (10.0% vs 26.5%) percentages were noted in aSAH patients comparing with controls (p < 0.05). Among monocytes a significant decrease only of nonclassical monocyte count was observed in aSAH patients (p < 0.05). Analysis of subsets of T lymphocytes revealed significantly lower count of CD3+, CD4+, and iNKT in aSAH patients comparing with controls.

Conclusion: Acute aSAH strongly influences the composition of leukocytes in the peripheral blood. In particular, we found a decrease of nonclassical monocyte count concomitantly with decreased numbers of CD3+, CD4+ and iNKT cells.

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Abstract – WCN 2013

No: 2225

Topic: 3 – Stroke

Is thrombolysis realizable at the Department of Neurology, Fann Teaching Hospital, Dakar-Senegal?

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Background and purpose: Stroke is a real health problem with a high morbidity and mortality in developing countries. This figure is increasing and worsening because of a problem of management. Nevertheless, the organization of care in the acute phase significantly reduces the cost, morbidity and mortality of stroke. Among these treatments, thrombolysis is the most effective therapeutic means to prevent lasting disability. However neurological clinics exist in Africa, thrombolysis in stroke patients is not updated. The study focused on the feasibility of thrombolysis in our neurological department.

Methods: This is a prospective study from September 2010 to April 2012 in a population of patients attending the Department of Neurology, Fann Teaching Hospital of Dakar-Senegal for stroke. Data on sociodemographic, lifestyle, delay to attend the clinic and to realize the CT scan of the brain were collected. We performed univariate and multivariate comparing the time of consultation and achievements of brain scan with socioeconomic data.

Results: Data of 285 patients was collected. They had a mean age of 56.5 years, male (50%), and ischemic stroke (73%). The main risk factors for stroke were: hypertension and diabetes.

The consultation period was within 3 h for 19.8% and within 6 h for 15.1%. 23.3% came by ambulance and 66.7% by public transport. 9% had the CT scan performed within 3 h and 18% within 6 h. No statistically significant difference between time compared to variables of interest before 6 h was observed.

Conclusions: Thrombolysis appears feasible in our clinic.

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Abstract – WCN 2013

No: 2278

Topic: 3 – Stroke

Time is brain: Stroke thrombolysis in Brunei Darussalam

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Background: Stroke is emerging as a major public health problem for Brunei Darussalam. The Neuroscience, Stroke and Rehabilitation Centre (NSRC) has established a dedicated stroke unit since 2010.

Objective: To assess the demographic of patients admitted with ischaemic stroke into NSRC who were within the 4.5 hours thrombolytic window.

Methods: Medical case-notes of all patients admitted with stroke (ischemic or haemorrhagic) from July 2010 to October 2012 were reviewed. Data collected included age, gender, referral centres, time of symptoms, time of arrival in NSRC, time of thrombolysis and length of stay.

Results: Out of 456 charts reviewed, 21 patients (5%) received intravenous thrombolysis with Alteplase. There were 13 (62%) males and 8 (38%) females. 86% of patients were referred from Brunei Darussalam main hospital (RIPAS) while 14% were referred from Jerudong Park Medical Centre. The mean age of patients was 56 ± 12 years (range 23–88).

The mean time from onset of symptoms to iv needle thrombolysis was 179 ± 62 min. The mean door to iv needle thrombolysis was 48 ± 22 min. Thus the mean time from onset of symptoms to reaching our unit (door) was 131 min.

Conclusion: In our preliminary study (with small number of patients), there is a quick and efficient door to needle thrombolysis time of 48 min. However there is a need to improve the time delay (131 min) from the onset of symptoms to reaching our stroke unit.

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Abstract – WCN 2013

No: 2153

Topic: 3 – Stroke

Demographic of stroke: A comparative study between Brunei Darussalam Neuroscience, Stroke and Rehabilitation Centre and Krankenhaus Nordwest, Frankfurt, Germany

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Background: The Neuroscience, Stroke and Rehabilitation Centre (NSRC) of Brunei Darussalam has been working in collaboration with Krankenhaus Nordwest Hospital, Frankfurt since 2010.

Objective: To compare the demographic of patients between the two centres.

Methods: All patients admitted with stroke (ischemic or haemorrhagic) from July 2010 to November 2011 to both centres were reviewed. Data collected included age and gender.

Results: In the Brunei data, 254 patients (59% male, 41% female) were studied (mean age of 60 ± 14 years, range 22–95). The male to female ratio in age group 21–30 is 1:0, age group 31–40 is 3:1, age group 41–50 is 2:1, age group 51–60 is 3:2, age group 61–70 is 1:1, age group 71–80 is 1:1, and age group > 80 is 1:1.

In the Frankfurt Krankenhaus Nordwest data, 3498 patients (50% male, 50% female) were studied (mean age 72.45 ± 20 years, range 19–103). The male to female ratio in age group 21–30 is 1:3, age group 31–40 is 1:2, age group 41–50 is 1:1, age group 51–60 is 2:1, age group 61–70 is 2:1, age group 71–80 is 1:1, and age group > 80 is 1:2.

Conclusion: Comparing Brunei NSRC to Frankfurt Krankenhaus Nordwest the mean age of stroke disease is 12 years younger in the Brunei patients and higher male to female ratio in the age group of 31–40 and 41–50. This warrants further study to identify the possible reasons for the demographic difference between an Asian and Western population.

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Abstract – WCN 2013

No: 2200

Topic: 3 – Stroke

Cortical deafness as a sole manifestation of ischemic cerebrovascular disease

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Background: Cortical deafness is a very rare condition characterized by the complete disruption of central auditory processing with intact peripheral auditory function.

Objective: To describe a case of cortical deafness associated with ischemic cerebrovascular disease.

Patients and methods: A 61 year-old female with a history of arterial hypertension presented to our department complaining of a sudden complete hearing loss. On examination, she was alert but slightly confused and agitated. Her spontaneous speech was fluent with occasional paraphasic errors. She was unable to follow verbal commands, but could understand simple written information. She appeared totally deaf, with no startle response to loud sounds. The rest of the neurological examination was unremarkable. Brain CT scan demonstrated a chronic right temporoparietal ischemic infarct. Otologic examination revealed no evidence of external or middle ear disease. Audiometry showed normal middle ear reflexes. Brainstem auditory evoked potentials were normal, consistent with normal auditory processing up to and including the inferior colliculi. An MRI scan demonstrated the old right temporoparietal infarct and an additional hyperacute ischemic infarct in the left posterior parietal area, extending into the Heschl's gyrus, the superior temporal gyrus and the Sylvian fissure. In three months' follow-up, the patient showed no improvement and she was able to communicate only through reading and writing.

Conclusion: Our case emphasizes on cortical deafness as a rare sole manifestation of ischemic cerebrovascular disease associated with bilateral damage of the primary auditory cortex.

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Abstract – WCN 2013

No: 1900

Topic: 3 – Stroke

Central obesity in the poorest region in Chile

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Background: Stroke is the first most common specific cause of death in Chile, since 2008. The increased risk in our region is explained by the prevalence of poverty, diabetes, a sedentary lifestyle and overweight.

Object: Is to assess the waist circumference (WC) in Clínica Alemana Temuco, during January 2011 to March 2013, in patients with a stroke.

Methods: We evaluated patients with a stroke diagnosis, in our primary Stroke Unit, who were hospitalized for ≥ 72 h. Waist circumference (WC) was measured according to WHO's recommendations, by two nutritionists. The study patients were divided into 3 groups according to WC for cardiovascular risk, low risk (LR): < 94 cm (men); < 80 cm (women); moderate risk (MR): 94 – 102 cm (men); 80 – 88 cm (women); and high risk (HR): > 102 cm (men); > 88 cm (women). We obtained a diagnosis of central obesity (CO) using NCEP-ATP-III criteria.

Results: 81 patients were included. 48 were males (59.3%). Men mean aged: 62 years (32–89), mean WC: 103.9 (SD: ± 9.9). BMI: 29. LR: 14.6%, MR: 37.5%, HR: 47.9% (CO). Women mean age: 57.4 years (35–81). Mean WC: 102.3 (SD: ± 13.6), BMI: 29.9. LR: 3%; MR: 15.2%; HR: 81.8% (CO). Total sample corresponds to LR: 9.9%, MR: 28.4%; and HR: 61.7% (CO).

Conclusion: Central obesity is a major risk factor for cardiovascular disease. This series of cases only shows that the problem is very important in Chile. The obesity and central obesity are epidemic. We

should perform behaviors to lower these indicators, with much education and government guidelines.

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Abstract – WCN 2013

No: 2198

Topic: 3 – Stroke

Preliminary evaluation of diffusion kurtosis imaging in acute stroke patients

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Aims: To translate and evaluate diffusion kurtosis imaging (DKI) in stroke patients.

Methods: Four patients underwent DKI and conventional MRI (including T1WI, T2WI, FLAIR and DWI) using a GE 1.5T HDx echo speed plus an MRI scanner. Mean kurtosis (MK), radial kurtosis (Kr) and axial kurtosis (Ka) were derived using DKI software in GE ADW4.3 workstation. Fourteen regions of interest (ROI) were outlined in infarction regions of stroke patients, including 2 ROIs in super-acute, 3 ROIs in acute, 6 ROIs in subacute and 3 ROIs in chronic lesions. The stroke duration was determined from onset time and DWI signal intensity. Stroke prognosis was evaluated by follow-up MRI and clinical manifestations.

Results: During super-acute stroke, regions of hyperintense MK and Kr progressed to infarction with mild bleeding without noticeable DWI and ADC abnormalities. For the case of sub-acute stroke, DKI (MK and Kr) appeared hypointense without significant change in DWI and ADC maps. In addition, 10 ROIs showed abnormality in both DKI and DWI/ADC maps.

Conclusions: DKI displayed contrast different from the standard DWI and ADC maps. DKI remains promising to address false negative signals in the standard DWI and ADC maps, and complement existing stroke MRI for improved stroke diagnosis.

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Abstract – WCN 2013

No: 2301

Topic: 3 – Stroke

Predictors of pneumonia in acute stroke inpatients in emergency unit

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Background: Pneumonia is a serious complication in acute stroke patients. Several mechanisms might be implicated including impairment of the immune system and aspiration. The analyses of clinical predictors are useful to guide the clinical management and to adopt preventive measures in order to improve the patients' outcome.

Objective: To determine the independent predictors of pneumonia in patients with acute stroke.

Methods: This is a retrospective study from July to December 2011. All patients admitted with a diagnosis of stroke in an academic medical center were included. We review all the medical charts and

extracted information using a structured questionnaire, including the demographics, NIH and Glasgow Coma scales; risk factors (hypertension, diabetes, dyslipidemia, smoking, alcohol, previous vascular disease); type (ischemic vs. hemorrhagic) and localization of stroke (anterior vs. posterior circulation); motor deficit; time use of mechanical ventilation. Pneumonia was defined clinically and treated with antibiotics.

Results: One hundred and fifty nine patients aged 18 to 90 (mean 63 ± 13.5) years were admitted. Incidence of pneumonia was 32.1%. Pneumonia incidence was higher in patients with ischemic stroke (OR: 4.36, 95% CI: 1.9–10.0). Pneumonia was most common in those with higher NIH and lower Glasgow coma scales ($p < 0.0001$). Patients with pneumonia had longer hospitalization ($p < 0.0001$). Logistic regression analyses identified only the NIH stroke scale as an independent predictor of pneumonia ($p = 0.001$).

Conclusion: The severity of the deficit as evaluated by the NIHSS scale was shown to be the only independent risk factor for pneumonia in acute stroke patients.

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Abstract – WCN 2013

No: 2318

Topic: 3 – Stroke

New prognostic score for the prediction of 30-day outcome in spontaneous supratentorial cerebral hemorrhage

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Objectives: The purpose of the present study was to evaluate predictors of outcome in primary supratentorial cerebral hemorrhage. Furthermore, we aimed to develop a prognostic model to predict 30-day fatality.

Methods: We retrospectively analyzed a database of 156 patients with spontaneous supratentorial hemorrhage to explore the relationship between anamnestic, clinical and CT characteristics, and fatal outcome within 30 days using multiple logistic regression analysis. The analyzed factors included volumetric data assessed by neuropathological and CT volumetry. A second CT scan in survivors, or neuropathological ABC/2 volumetry in non-survivors was used along with the baseline CT to assess the growth index of hematoma.

Results: Systolic blood pressure, serum potassium and glucose levels, platelet count, absolute and relative hematoma volumes, presence and size of intraventricular hemorrhage statistically significantly predicted the fatal outcome within 30 days. Based on our results we formulated a six-factor scoring algorithm named SUSPEKT to predict the outcome.

Conclusions: After validation the SUSPEKT Score may be applicable in general clinical practice for early patient selection to optimize individual management or for assessment of eligibility for treatment trials.

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Abstract – WCN 2013

No: 2315

Topic: 3 – Stroke

Association between metabolic syndrome and homocysteinemia in ischemic stroke

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Background: Stroke is one of the leading causes of morbidity and mortality worldwide. The role of metabolic syndrome and homocysteinemia as risk factors for ischemic stroke is not completely clear.

Aim: To determine the frequency of metabolic syndrome and homocysteinemia, as well as their association in patients with ischemic stroke.

Method: The research included 53 subjects being on rehabilitation after stroke and 40 clinical controls without vascular disease that were on rehabilitation due to the back pain problems.

Results: The frequency of metabolic syndrome was much higher in patients with stroke compared to control group (88.7% vs. 70.0%, $p < 0.05$). The level of homocysteinemia and the frequency of hyperhomocysteinemia were increased in the patients with stroke ($15.0 \pm 5.50 \mu\text{mol/L}$ vs. $11.2 \pm 2.51 \mu\text{mol/L}$, $p < 0.01$ and 39.2% vs. 11.4%, $p < 0.01$, respectively). Among patients with stroke, those with metabolic syndrome had higher frequency of hyperhomocysteinemia (42.2% vs. 16.7%, $p < 0.05$) and the serum level of homocysteine was significantly higher in patients with more individual components of metabolic syndrome (11.1% in patients with 3 components, 36.8% in patients with 4 components and 64.7% in patients with 5 components, $p < 0.05$).

Conclusion: Our results suggest that metabolic syndrome and homocysteinemia represent significant risk factors for ischemic stroke. It seems that there is an association between these two factors in pathogenesis of the ischemic stroke, but further analyses are needed to confirm this hypothesis.

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Abstract – WCN 2013

No: 2347

Topic: 3 – Stroke

Thrombolysis in ischemic stroke in a familial amyloidotic polyneuropathy patient – Is there an increased risk of hemorrhagic complication?

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Background: Patients with familial amyloidotic polyneuropathy (FAP) type I have typically a severe somatic and autonomic peripheral neuropathy. Amyloid deposits of the mutated transthyretin (frequently TTR Val30-Met, almost all produced by the liver) are said to mostly spare the brain in this type of FAP. Nevertheless, with longer survival induced by liver transplantation the choroid plexus TTR Val30-Met production might become relevant.

Patients and methods: A 46-year-old woman with TTR Val30-Met FAP, with liver transplant and pacemaker since age 37, and on immunosuppression with tacrolimus, was admitted by acute motor aphasia (NIHSS 4). Brain CT and analytical study were normal. Thrombolysis was started 3.5 h after symptom onset. At the stroke unit she worsened and repeated CT scan disclosing left temporal and right fronto-basal hematoma. Due to pacemaker MRI was not performed. Cervical and transcranial duplex scan was normal. She was discharged a week later asymptomatic, on triflusal 600 mg/d.

Discussion: There is no data on thrombolysis safety in FAP patients. It is known that in some mutations there is an important amyloid

deposition in the leptomeningeal and cortical vessels, with frequent intracerebral hemorrhagic stroke (IHS).

Conclusion: Although with TTR Val30-Met mutation IHS is not common, in our liver transplanted patient the local and remote parenchymal hemorrhage after thrombolysis raises the hypothesis that cerebral amyloid angiopathy might develop in the long-term in these patients, due to maintained abnormal transthyretin production in the choroid plexus. Brain pathological studies are necessary to confirm this hypothesis in the TTR Val30-Met liver transplanted patients.

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Abstract – WCN 2013

No: 2319

Topic: 3 – Stroke

Setting up a neuroscience stroke and rehabilitation centre 12,000 km away with the help of telemedicine

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Background: Due to world-wide aging population neurologists are urgently needed for stroke/non-stroke.

In Brunei Darussalam we set up a national stroke unit, intensive care unit, neurorehabilitation on-site including tele-neurology and tele-teaching.

Objective: The aim of this project “to teach to treat-to treat to teach” was to set up a world class centre of neurology.

Methods: In 7/2010 set-up of the Bruneian Neuroscience Stroke and Rehabilitation Centre (BNSRC) has started. To overcome the distance of 12,000 km a telemedical network between the Department of Neurology (KHNW) and BNSRC started. This international cooperation includes a “specialist neurology” training program, accredited in Brunei Darussalam by an international advisory board. Daily tele-teaching, 24/7 tele-neurology service, as well as all neurological laboratories have been set up on site by tele-cytology, and tele-electrophysiology including EEG and ultrasound. Awareness campaigns and telescience have been successfully started.

Results: The BNSRC already has been successfully audited according to the STU requirements of the German Stroke Society. Over 2000 patients have been treated so far. Several Bruneian doctors are enrolled in the specialist curriculum.

Conclusion: The program to teach to treat-to treat to teach resulted in a local set up of a BNSRC, being built up by KHNW. This outstanding project of a cooperation between the KHNW and BNSRC succeeded in building up a centre of excellence of high end acute neurological department including a stroke unit and neurorehabilitation.

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Abstract – WCN 2013

No: 2345

Topic: 3 – Stroke

Stroke in patients with active and non-active malignancy

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Background: Global burden of stroke and cancer is steadily increasing over the years. According to the previous studies, the pattern of stroke in cancer patients may differ from the conventional one.

Objectives: To compare vascular risk factors, stroke etiology and outcome among patients with positive or negative history of cancer, with emphasis on the activity of the malignancy.

Methods: All consecutive acute stroke patients admitted to our department between September 2006 and September 2011 were included. We distinguished between patients:

- 1) with active malignancy (the diagnosis of cancer within 12 months before stroke, AM);
- 2) with non-active malignancy (non-AM);
- 3) cancer-free patients (CF), used as a reference.

Results: Of 1558 acute stroke admissions, 90 (5.8%) had an additional diagnosis of cancer, including 41 cases of AM (2.6%) and 49 cases of non-AM (3.1%). Compared to CF group, non-AM patients did not differ in terms of vascular risk factors; they more frequently had stroke due to small vessel occlusion (18.4% vs 7.0%, $p = 0.003$). AM patients less frequently had atrial fibrillation (7.3% vs 23.5%, $p = 0.016$) and history of previous stroke (2.4% vs 24.9%, $p = 0.018$). The most frequent stroke etiology in the AM group was undetermined (61.0%) and they had higher levels of serum inflammatory parameters. Stroke severity and outcome were similar across all groups.

Conclusions: Compared to the CF patients, the pattern of stroke in non-AM patients appears similar. However, stroke pattern in patients with AM may be different from the conventional one, but it does not affect the short term prognosis.

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Abstract – WCN 2013

No: 2355

Topic: 3 – Stroke

Late cerebrovascular complications of radiation therapy for pediatric primary central nervous system tumors

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Background: Radiation therapy (RT) plays an important role in the management of primary central nervous system tumors (PCNST) in children. Despite field and dose optimization, late cerebrovascular complications (LCCs) still occur. However, the number of studies assessing the burden of these complications is limited.

Objective: To describe the occurrence of LCCs in adult survivors who underwent RT for PCNST during childhood.

Methods: Retrospective consecutive case series description of LCCs from a cohort of adult survivors of pediatric PCNST. Diagnosis of hemorrhagic LCCs was confirmed by gradient-echo MRI sequence.

Results: We identified LCCs of RT in 35 of a total of 103 patients (34.0%); median age at RT was 8.4 years; and median follow-up was 18.7 years. Histological diagnoses were: medulloblastomas/primitive neuroectodermal tumors (16/45.7%), gliomas (15/42.9%) and germ cell tumors (4/11.4%). The most frequent PCNST site was the posterior fossa (21/60%). Mean total radiation dose was 48 Gy (18–54 Gy); most patients received focal plus entire neuraxis RT (23/65.7%). Thirty-three patients had microbleeds, all asymptomatic; cavernomas occurred in eleven patients, causing epilepsy in 2; two patients had a symptomatic ischemic stroke (6 and 25 years after RT); two patients presented with symptomatic superficial siderosis (15 and 26 years after RT) and one patient developed symptomatic moyamoya syndrome 4 years after RT. In some patients, microbleeds and cavernomas increased in number during follow-up.

Conclusions: LCCs are very common after RT for pediatric PCNST, with microbleeds and cavernomas being more frequent in a continuous and dynamic pathophysiological process.

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Abstract – WCN 2013

No: 1779

Topic: 3 – Stroke

Long-term imaging follow-up in patients with adequately coiled basilar tip aneurysms is essential to timely detect late reopening

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Background and objective: Reopening after coiling at mid-term follow-up is more frequent in basilar tip aneurysms than in aneurysms at other locations. To quantify this phenomenon of late reopening, we performed MRA at 3 T in a cohort of patients with basilar tip aneurysms coiled between 1994 and 2005, that were adequately occluded at short- or mid-term and had no imaging follow-up in the last 7 years.

Patients and methods: Between 1994 and 2005, 148 patients with basilar tip aneurysms were coiled. In 2012, 29 were deceased and 56 had prolonged imaging follow-up after previous reopening and/or retreatment. Of the remaining 63 patients, 11 were untraceable, 11 were not eligible because of contra-indications for MRI or poor clinical condition, and 9 refused follow-up MRA. Finally, 32 patients with adequately coiled basilar tip aneurysms and without imaging follow-up in the last 7 years were scheduled for MRA follow-up after a mean of 12.8 years (median 12.7, range 7–19 years).

Results: Of 32 patients, 31 had MRA and 1 patient had angiography after a rebleed 5 days before scheduled MRA. Four of 32 patients (12.5%) had late reopening of the coiled basilar tip aneurysm requiring additional coiling.

Conclusions: Basilar tip aneurysms that are adequately occluded at short- or mid-term after treatment may reopen (and rebleed) many years later with a frequency of 1 in 8 in this cohort. In patients with coiled basilar tip aneurysms, prolonged imaging follow-up at regular intervals is recommended to timely detect late reopening.

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Abstract – WCN 2013

No: 2320

Topic: 3 – Stroke

The association between routine serum C-reactive protein and outcome in ischemic stroke patients treated with intravenous alteplase

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Background: The clinical usefulness of blood biomarkers in acute stroke is not yet fully established, especially after intravenous thrombolysis.

Objective: Our aim was to investigate the association between routine serum C-reactive protein (CRP) measured very early after stroke and outcome in patients treated with intravenous thrombolysis, adjusting for a history of recent infection.

Patients and methods: We analyzed the data of consecutive patients treated with intravenous alteplase in our center between October

2003 and December 2011. The information was prospectively collected in a detailed registry. Patients or their proxies were additionally interviewed for a history of an infectious event during 7 days before stroke onset. Routine serum CRP was measured within 24 h from admission, concentration >5 ng/ml was considered elevated.

Results: Serum CRP was measured in 341/406 patients treated with intravenous rt-PA. Patients with elevated CRP (135/341, 42.5%) compared to those with normal CRP values, were significantly older, more frequently presented with a pre-existing disability, comorbidities and suffered more severe strokes. They had a higher proportion of symptomatic intracranial hemorrhage according to ECASS II definition (7.2% vs 1.6%, $p = 0.010$), higher 3-month mortality (25.6% vs 11.3%, $p = 0.001$), and were less frequently alive and independent after 3 months (45.9% vs 63.7%, $p = 0.002$). However, those associations were not confirmed in a multivariate analysis.

Conclusions: Routine serum CRP determined very early after stroke may be helpful in establishing long-term prognosis for patients treated with intravenous thrombolysis. Elevated CRP seems to coexist with other factors associated with poor outcome.

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Abstract – WCN 2013

No: 2422

Topic: 3 – Stroke

Cerebral perfusion changes in symptomatic and asymptomatic patients with carotid artery stenosis

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Background: Optimal strategy of ischemic stroke prevention in symptomatic and asymptomatic patients with severe internal carotid artery (ICA) stenosis may depend on the impairment of cerebral hemodynamics.

Objective: To evaluate changes in cerebral perfusion, measured with CT perfusion imaging, in symptomatic and asymptomatic patients with ICA stenosis >70%.

Patients and methods: We studied 39 patients with unilateral carotid artery stenosis >70% (19 symptomatic and 20 asymptomatic patients, median age 67 years; study group) and 40 otherwise healthy subjects with mild hypertension (median age 60 years; control group). Patients underwent CT perfusion at the level of the basal ganglia and semioval center. Mean transit time (MTT), cerebral blood volume (CBV), and cerebral blood flow (CBF) were calculated in the middle cerebral artery area.

Results: In symptomatic patients MTT and CBV values were significantly different in study vs. control group (median MTT: 7.1 s vs 4.4 s, $p < 0.01$; median CBV: 3.4 ml/100 g vs. 3.7 ml/100 g, $p < 0.05$, respectively). In asymptomatic patients only MTT increased significantly in study group (5.7 s vs 4.4 s, $p < 0.01$, respectively).

Conclusion: Both symptomatic and asymptomatic patients with carotid stenosis >70% demonstrated deterioration of cerebral perfusion. The pattern of changes may indicate that cerebral blood flow autoregulation is more severely impaired in symptomatic patients than in asymptomatic ones. This may lead to the more weighted approach to the stroke prevention.

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Abstract – WCN 2013**No: 2381****Topic: 3 – Stroke****Post-partum angiopathy: A challenging clinical-case**

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Background: Postpartum angiopathy is one of the reversible cerebral vasoconstriction syndromes. It usually presents as a combination of symptoms such as severe headache, seizure, encephalopathy, motor deficit or ataxia. Intraparenchymal or subarachnoid hemorrhage, ischemic stroke and vasogenic brain edema have been reported.

Clinical case: Three days after an uncomplicated delivery, a 36 yo woman developed progressively intense holocranial headache. Within 1 h of headache onset, she was aphasic and had right motor deficit. Her past medical history included migraine and aplastic anemia. Her pregnancy was complicated by anemia, so she needed transfusion support. Blood pressure was normal and she did not develop albuminuria or preeclampsia throughout pregnancy.

Cerebral CT scan showed left basal ganglia hemorrhage. Angiography showed multiple bilateral stenosis involving large and medium size cerebral arteries. Transcranial Doppler showed bilateral flow accelerations in all intracranial cerebral arteries, more severe on the right anterior circulation. She was treated with oral nimodipine, endovenous magnesium sulphate and mild hypervolemia. On day 16 she had a focal seizure, with right temporal paroxysmic activity on interictal EEG. A cerebral MRI scan showed acute cerebral infarction involving right anterior cerebral artery. Due to the lack of clinical recovery, a presumptive diagnosis of non-convulsive status epilepticus was made and levetiracetam perfusion was started, with clinical and electroencephalographic response. The patient neurological status and cerebral vasospasm gradually improved.

Conclusion: We illustrate a challenging clinical case of postpartum angiopathy that presented with several of the possible complications of this syndrome: intraparenchymal cerebral hemorrhage, seizures and ischemic stroke.

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Abstract – WCN 2013**No: 2285****Topic: 3 – Stroke****Retrospective evaluation of patients with oral anticoagulant treatment introduced for secondary prevention**

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Background and objective: Warfarin reserves an important role for secondary prevention. The purpose of the study was to determine the efficacy of anticoagulation for secondary prevention in our clinical practice in strokes of different etiology.

Material and methods: We enrolled 181 ischemic stroke patients categorized according to TOAST classification. These patients started as either only with warfarin or warfarin and antiagregan combination were examined as retrospectively (1 month–16 years) for recurrent vascular incident and bleeding complication.

Results: Patients that are grouped according to their warfarin use period (first 1 year, 1–5 years, more than 5 years), minor bleeding complication was found statistically significant ($p = 0.023$) in more than 5 years warfarin using group and there was no other difference between the groups. During the follow, recurrent vascular incidence was determined in 17 patients, 13 of these patients (7.2%) had

recurrent ischemic strokes. 6 (3.3%) major and 23 (12.7%) minor bleeding complications were observed. Average INR value was 1.69 ± 0.31 during the recurrent ischemic stroke, on the other hand it was 3.15 ± 1.30 during major bleeding complication. There was no significant difference between only warfarin using and warfarin with antiagregan using patients in terms of recurrent ischemic stroke and bleeding complications. During the diagnosis, ECG showed that patients had normal sinus rhythm, 48.8% of these patients' Holter results showed PAF.

Conclusion: This study is important as it reflects clinical practice and we can say that our department is successful with INR follow on the strength of the similarities from our study with the literature.

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Abstract – WCN 2013**No: 2399****Topic: 3 – Stroke****Thrombolysis or anticoagulation for cerebral venous thrombosis (TO-ACT trial)**

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Background: Endovascular thrombolysis (ET), with or without mechanical clot removal, may be beneficial for a subgroup of patients with cerebral venous sinus thrombosis (CVT), who have a poor prognosis despite treatment with heparin. Published experience with ET is promising, but only based on case series.

Objective: The objective of the TO-ACT trial is to determine if ET improves the functional outcome of patients with a severe form of CVT.

Methods: The TO-ACT trial is a multi-centre, prospective, randomized, open-label, blinded endpoint (PROBE) trial. Patients are eligible if they have a radiologically proven CVT, a high risk of poor outcome (defined by presence of one or more of the following: mental status disorder, coma, intracranial hemorrhagic lesion, or thrombosis of the deep cerebral venous system) and if the responsible physician is uncertain whether ET or standard treatment is better. 164 patients will be included.

Intervention: Patients are randomized to receive either ET or standard treatment (therapeutic doses of heparin). ET consists of local application of rt-PA or urokinase within the thrombosed sinuses. Mechanical clot removal, such as thrombosuction, is allowed, but not mandatory.

Outcomes: The primary endpoint is the modified Rankin score (mRS) at 12 months, with a score ≥ 2 defined as poor outcome. Secondary outcomes are 6 months mRS, mortality and recanalization rate. Principal safety outcomes are major intra- and extracranial hemorrhagic complications. Results will be analyzed according to the "intention-to-treat" principle. Blinded assessors not involved in the treatment of the patient will assess endpoints with standardized questionnaires.

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Abstract – WCN 2013**No: 2432****Topic: 3 – Stroke****P-wave dispersion & transthoracic echocardiography to predict paroxysmal atrial fibrillation as a cause of stroke or transient ischemic attack**

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Background: One-third of stroke and transient ischemic attack (TIA) are cryptogenic requiring additional investigation and intervention. Occult paroxysmal atrial fibrillation (PAF) has been suggested as a possible cause for these strokes.

Objective: The aim of our study is to evaluate simple ECG & bed side echocardiographic parameters for the prediction of PAF in patients presenting with stroke or TIAs.

Patients and methods: Sixty patients with non-hemorrhagic stroke were included in the study, during 1 week of continuous ICU monitoring 30% of patients were discovered PAF (group 1) and the remaining 42 patients didn't develop PAF (group 2). Both groups were subjected to detailed comparative analysis regarding demographic data, risk factors, clinical examination including NIHSS, serial ECGs for calculation of Pmax & Pdis, echocardiography with evaluation of LAV, LAVI, CT Brain & Duplex on carotid arteries.

Results: It was found that Pmax & Pdis were significantly higher in group 1 (PAF) than in group 2 (NSR) (147.7 ± 9.6 & 54.1 ± 7.5 in group 1 VS 114.3 ± 9 & 30.2 ± 7 in group 2) with P value < 0.001. Also LAV & LAVI were significantly higher in group 1 in comparison to group 2, (57.1 ± 10 & 28.9 ± 3 respectively) in group 1 & (40.1 ± 12 & 20.1 ± 8 respectively) in group 2 with P value < 0.001. On multivariate logistic regression analysis Pmax, Pdis, and LAVI were the most significant independent predictors of PAF.

Conclusion: Paroxysmal atrial fibrillation is common in cryptogenic stroke (30%), Pmax, Pdis in single surface ECG & LAVI in echocardiography are highly significant predictors of PAF.

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Abstract — WCN 2013

No: 2437

Topic: 3 — Stroke

Clinical characteristics and short term mortality ischemic stroke patients with atrial fibrillation

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Background: Ischemic stroke (IS) accounts for 85% of all strokes. Atrial fibrillation (AF) is a strong independent risk factor for stroke.

Objective: AF is responsible for about 25% of all IS and the risk of stroke attributed to AF increases with age. This study investigated the clinical characteristics and case fatality for the first-ever IS patients with AF.

Patients and methods: 102 patients were included in the study. A stroke was classified as IS when intracranial hemorrhage was excluded by a CT. AF was defined as a history of persistent AF or paroxysmal AF, supported by past ECG or diagnosed based on ECG and/or 24-hour ECG monitoring during admission. Short-term mortality was assessed as 30-day case-fatality rate.

Results: Of the 102 patients included in this study 21 (20.6%) had AF. Compared to patients without AF, patients with AF were older (71.2 vs. 68.3) and had higher frequency of coronary heart disease (15.1 vs. 12.3%) and myocardial infarction (4.1 vs. 1.2%), but a lower incidence of hypertension, diabetes mellitus, and hyperlipidemia. These patients had a higher NIHSS score on admission (median 12 vs. 7) and hemorrhagic transformation (16.2 vs. 7.8). Within AF patients, only 3 (14.3%) received oral anticoagulants before stroke onset. 9 subjects with AF died within 30 days with an overall mortality of 42.9%.

Conclusion: Our results indicate that patients with AF have a higher frequency of cardiovascular comorbidities, stroke-related complications and a higher early death than patients without AF. The oral anticoagulants are rarely used in stroke prevention.

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Abstract — WCN 2013

No: 2340

Topic: 3 — Stroke

Carotid baroreceptors. Their role in ischemic cerebral stroke, coronary artery disease, and arterial hypertension.

Are the changes in baroreceptor sensitivity age-related?

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Background: The changes in baroreceptor sensitivity in ischemic cerebral stroke (iCI), coronary artery disease (CAD), arterial hypertension (AH), carotid atherosclerosis (CA) and their age-related changes are poorly documented.

Objective: Hypothesis that the changes in baroreceptor sensitivity (BRS) may correlate with arterial wall, stiffness, IMT, plaque morphology and also age was assessed. The changes in BRS can influence compensatory mechanisms in different clinical situations. Prospective, multidisciplinary, multicentric study.

Material and methods: Material consists of 1034 subjects, divided in four groups: 1. iCI (n = 296), mean age 65.4 ± 9.6 , men 50.6%, BMI = 29.7 ± 3.8 , 2. AH, (n = 261), mean age 59.2 ± 9.36 , men 47.8%, BMI = 28.9 ± 5.1 , 3. CAD (n = 220), mean age 69.2 ± 10.4 , men 37.5%, BMI = 29.6 ± 3.8 , and 4. healthy controls (C, n = 237), mean age 50.9 ± 11.3 , men 47.1%, BMI = 23.8 ± 4.2 , three age-related subgroups were created:

1st group <40 yrs,
2nd group: 41–60 yrs,
3rd group >60 yrs.

Neurological, cardiological examinations by certified stroke-neurologist, certified cardiologist, Diagnoses clearly documented (CT/MRI). Battery of biochemical/hematological investigations. BRS measured by Finometer (Finapres Medical Comp. Amsterdam), stiffness (SphygmoCor, AtCor, Sydney), IMT, plaque morphology (USG).

Results: BRS is significantly lower in all three groups (AH, iCI, CAD) with lowest values in iCI, comparing to controls (92 ± 1.76 vs 4.41 ± 1.70 , $p < 0.001$). Stiffness (PWV $11.06 \pm 4.12 \pm 4.8$ vs 7.48 ± 1.96 , Aix 26.5 ± 11.5 , 28.3 ± 9.0 , 24.9 ± 9.8 vs 14.2 ± 6.7), IMT (0.798 ± 0.195 vs 0.648 ± 0.123 mm) and plaque morphology showed significant higher values in all groups of pts comparing to controls ($p < .0001$). The changes are in inverse relationship to arterial wall flexibility (-0.52 , $p < .001$). BRS changes are age related (from 40 yrs to more than 60 yrs ($p < 0.001$)).

Conclusions: BRS changes have shown an inverse close relationship to all parameters of arterial wall (Stiffness, PWV, Aix) in all groups of pts comparing to controls. They are age-related. The results provoke opinion regarding possible unfavorable influence of BRS changes on compensatory mechanisms related to the onset and outcome of cerebrovascular and cardiovascular events.

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Abstract – WCN 2013

No: 2445

Topic: 3 – Stroke

Multiple lacunar state and post-stroke dementia

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Aim: Epidemiologic assessment of post-stroke dementia and its correlation with imaging data.

Methods: We studied the charts of 241 stroke patients followed up at the neurology unit, Speciality Polyclinic 2, Tirana, during the last three years period. They are presented at least three months after the first or recurrent stroke. All patients underwent a detailed neurological examination. HIS score was applied. Diagnosis of dementia was made according to the NINDS-AIREN criteria. The imaging (CT and/or MRI) of the brain is requested. Patients who had previous history of psychotic and delusional disorders, impairment of speech and communication after stroke and patients who died were excluded from the study. SPSS 17.0 program was applied for data analyzed.

Results: The mean age is 68.6 (± 9.9) years old. There are 72 (29.8%) females and 169 (70.2%) males. We analyzed the data of 69 (28, 63%) patients of them who met criteria for dementia. 23 (33.3%) patients of them had 4–7 in HIS score. Mix dementia. 46 (66.7%) patients had >7 in HIS score (or vascular dementia). There is a statistically significant correlation between post-stroke dementia and sub-cortical lacunar infarction, bilateral thalamic lacuna and multiple white matter lesions (p value < 0.01). No significant correlation between post-stroke dementia and bulbar lacuna is found.

Conclusion: This study suggests that post-stroke dementia is most frequent in multiple lacunar state and bilateral ischemic stroke patients.

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Abstract – WCN 2013

No: 1788

Topic: 3 – Stroke

Cerebral thrombophlebitis and β -thalassemia intermedia

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Introduction: The presence of a high incidence of thromboembolic events, mainly in β -thalassemia intermedia patients, has led to the identification of a hypercoagulable state in thalassemia. The purpose of our case report is to highlight the thrombotic risk and the pathophysiological characteristics of hypercoagulability in thalassemia patients.

Case report: A 23-year-old man with medical history of β -thalassemia intermedia and splenectomy was hospitalized for an acute headache beginning two weeks before consultation associated to bilateral blurred vision and vomiting. Clinical examination found bilateral papilledema on optic fundus. He had no meningeal irritation. Biological data found a mild anemia on blood count. Cerebral Magnetic resonance imaging revealed cerebral venous thrombosis of the superior sagittal sinus. Anticoagulants were indicated at life.

Discussion: Diverse factors contributing to the hypercoagulable state in patients with thalassemia have been identified: platelet activation, increased platelet aggregation, alteration in RBCs, and elevated levels of endothelial adhesion proteins.

Clinical observations have suggested that splenectomy can contribute to an increased susceptibility to thrombosis attributed to the presence of high platelet counts following splenectomy and to increased number of abnormal RBCs.

The higher incidence of thrombotic events in thalassemia intermedia compared to thalassemia major patients is mainly attributed to transfusion naivety and splenectomy, both of which promote an underlying procoagulant activity.

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Abstract – WCN 2013

No: 1826

Topic: 3 – Stroke

Endothelial dysfunction in acute ischemic stroke

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Aims: To evaluate indicators of intima-media thickness (IMT), brachial flow-mediated dilatation (FMD) and GTN-mediated dilatation in patients with different subtypes of ischemic stroke.

Methods: The study included 72 patients with acute ischemic stroke (age, 58.24 ± 5.64 years) and 32 control patients (56.29 ± 4.75 years). Endothelial function was assessed using the method of D. Celermajer. An endothelium-dependent vasodilation (FMD) was caused by a cuff of a sphygmomanometer. To assess an endothelium-independent vasodilation (GTN-mediated) we performed pharmacological tests with glyceryltrinitrate. FMD and GTN-mediated responses were calculated as the difference between the maximum diameter and initial. Coefficient of endothelial dysfunction was calculated as to divide GTN-mediated dilatation by FMD.

Results: According to the criteria TOAST the subtypes of stroke were identified as large-artery atherosclerosis (ATS) – 36.1%, cardioembolism (CES) – 27.8%, small-artery occlusion – 19.4%, other determined cause – 8.3%, and undetermined cause – 5.5%.

Significantly detected a difference IMT in patients with stroke (0.72 ± 0.19) and in control group (0.6 ± 0.13 , $p < 0.05$). IMT in patients with CES (0.69 ± 0.01) was lower than in patients with ATS (0.73 ± 0.08 , $p < 0.05$). FMD test was significantly higher in patients with CES (0.62 ± 0.03) than in patients with ATS (0.57 ± 0.02 , $p < 0.05$). GTN-mediated test showed no significant differences in stroke subtypes. Coefficient of endothelial dysfunction was higher in patients with stroke than in control group (1.91 versus 1.44, $p < 0.05$), and in patients with ATS than CES (1.40 versus 1.28, $p < 0.05$).

Conclusions: Indicators of endothelial function was deteriorated to a greater extent in patients with ATS, compared to patients with CES.

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Abstract – WCN 2013

No: 2472

Topic: 3 – Stroke

Vascular risk factors, etiology and outcome comparison between cancer and non-cancer patients with acute ischemic stroke: Nested case-control study

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Background: Concerning common vascular risk factors, recent data comparing cancer and non-cancer patients shows no significant

differences. However, in cancer patients, several clinical conditions appear to increase the risk of stroke.

Objective: Compare ischemic stroke in cancer and non-cancer patients according to common modifiable vascular risk factors, etiology and outcome at discharge and three months later.

Material and methods: Retrospective case–control study conducted in a stroke unit, between January 2007 and December 2012. Cases: diagnosis of cancer and acute ischemic stroke; controls: only stroke. Groups were compared based on: vascular risk factors, etiology (TOAST classification) and outcome at discharge (NIH Stroke Scale, modified Rankin Scale scores).

Results: 56 cases were identified; 64.3% men, mean age of 71 years (31–92 years); 37.5% had evidence of active cancer. Gastrointestinal cancer (25.9%) was the most common; 151 controls, matched for gender and age. Vascular risk factors were not significantly different, except diabetes mellitus, more frequent in the control group [16.07% vs 33.77%, $p = 0.02$]. Cancer cohort had more previous thrombotic events [8.93% vs 0.66%, $p = 0.007$]. Another determined etiology subtype was more frequent in the cancer patients [17.86% vs 0.66%, $p < 0.001$], with no significant differences on the other TOAST subtypes. Functional outcome, both at admission and discharge were not significantly different. Mortality rate was higher in cancer patients at discharge and three months later [16.07% vs 3.31%, $p < 0.01$].

Conclusion: Results reveal similar vascular profile, although ischemic stroke in cancer patients seems to be more frequently associated with a hypercoagulable state and poorer outcome.

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Abstract – WCN 2013

No: 2498

Topic: 3 – Stroke

Treatment possibilities of basal ganglia hemorrhage: Retrospective trial of fifty-four patients

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Background: Intracerebral hemorrhage presents a challenge for both neurologists and neurosurgeons and constitutes up to 15% of the total number of cerebrovascular incidents.

Objective: The aim of this trial is to present an outcome of conservative and surgical treatment of intracerebral hemorrhage in our hospital.

Patients and methods: All patients underwent CT on admission. The treatment depended on CT confirming intracerebral hemorrhage and regarding progressive neurological deterioration of consciousness, brain edema or shift effect. These patients underwent a prompt surgical treatment with decompressive craniotomy and clot removal or external ventricular drainage as standard approach. The other patients underwent conservative treatment which consisted of antiedematous therapy and blood pressure regulation.

Results: In a group of fifty-four patients, fifty-one patients were treated conservatively and three patients were treated surgically. Conservatively treated patients and three patients treated surgically showed clinical improvement with satisfying additional postoperative CT scan within 48–72 h.

Conclusion: The trial showed that there is no difference between conservative and surgical treatment, although surgical treatment was superior in cases of progressive neurological deterioration with GCS 5–11. According to our retrospective trial, we can conclude that there was no significant difference between conservative and surgical treatment during the follow-up period of three months.

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Abstract – WCN 2013

No: 2477

Topic: 3 – Stroke

Impact of heat shock protein on ischemic stroke risk

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Heat shock protein 72 (HSP72) has been found to reduce ischemic injury. HSP72 itself may be a marker for neuroprotection in ischemic stroke.

The aim of the study was to investigate the association between HSP plasma concentration and risk of ischemic stroke. We also investigated MTHFR and CBS genotypes.

Methods: We have measured the level of antibodies against HSPs in vitro, in serum of ischemic stroke patients and control group using ELISA. We study the prevalence of MTHFR (C677T, A1298T) and CBS (T833C, insertion 68 bp in position 844) genotypes.

Results: Heterozygotic genotype C677T of MTHFR was detected in 45% of patients, and A1298T in 43%. 5% of homozygotic C677C and 8% of T1298T were detected. In 87.5% of subjects at least one heterozygotic mutation was detected. Hyperhcy was correlated with mutations in either gene ($p = 0.04$). The level of antibodies against one of the bacterial HSPs, GroEL, correlates with the level of serum hcy. It points at the possible significance of autoimmune reactions contributing to atherosclerosis. We have also purified MTHFR and tested the influence of HSPs of the KJEB system in vitro, using spectrophotometric test for MTHFR activity. We found that MTHFR is partially protected from the effect of heat shock by KJEB system and that KJEB proteins can restore that activity of the enzyme when it is lost due to thermal denaturation.

Conclusions: Our study confirms diagnostic significance of antibodies to HSP in stroke. HSPs can be involved in the development of atherosclerosis, as well as protection against stroke.

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Abstract – WCN 2013

No: 2463

Topic: 3 – Stroke

Tympanic temperature matches the brain temperature, fact or fiction?

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This study is a case series, which was conducted on 10 CVA patients who were admitted in the ICU ward. These subjects consist of unilateral or bilateral ICH.

GCS, vital sign and tympanic temperature were controlled for the subjects within one week. Tympanic temperature measured from the left and the right ear. After this time we observed the meaningful difference between the left and the right tympanic temperature in patients with a unilateral lesion, but this difference was not seen in patients with bilateral lesions.

Based on these results, it seems that we used tympanic temperature in early diagnosis of hemorrhagic lesion in the affected side of the brain, just as the case in unilateral hemorrhagic lesion in patients with different etiologies.

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Abstract – WCN 2013**No: 1435****Topic: 3 – Stroke****Isolated right carotidynia: CT, MRI, FDG-PET and contrast-enhanced ultrasound carotid examination findings**N. Villain^a, C. Nifle^a, J.-M. Baud^b, M. de Malherbe^c, F. Pico^a.^aDepartment of Neurology and Stroke Center, Versailles, France;^bAngiology and Ultrasound Unit, Versailles, France; ^cDepartment of Radiology, Versailles Saint Quentin en Yvelines University, Versailles Hospital, Versailles, France

Introduction: Carotidynia as a distinct nosological entity is a controversial issue. Some authors have described consistent MRI findings in patients with carotidynia. We report a patient with this MRI pattern and present findings of contrast-enhanced ultrasound carotid examination.

Case description: A 52 year-old woman was admitted in the Stroke Unit for a suspected carotid artery dissection, after a 3-day history of intense, sudden and isolated right-neck pain. Clinical exam was strictly normal except for the tenderness at the right-carotid bifurcation. Ultrasound carotid examination depicted a hypoechogenic tissue thickening around the right-carotid bifurcation, with an enhancement after injection of a microbubble contrast agent, without evidence for dissection or plaque. CT and MR findings evidenced a T1 isosignal, T2 hypersignal, and CT isodense infiltration around the right-carotid bifurcation, together with a gadolinium enhancement, without argument for artery dissection. The MR-angiography was normal except for a mass effect on the homolateral intern jugular vein, also observed on CT-angiography. A full biological testing for vasculitis (including biopsy of the temporal artery), auto-immune, infectious or metabolic disorder remained negative, including for a biological inflammatory syndrome. The FDG-PET scan, and the aorta-renal arteries CT-angiography were also normal. Symptoms spontaneously disappeared after day 9. Repeated MR confirmed the disappearance of initial abnormalities.

Discussion: We provide a full clinical/imaging/biological report of an isolated and remittent right carotidynia, arguing for idiopathic carotidynia as a distinct radio-clinical syndrome. MRI findings in this case are consistent with previous descriptions. To our knowledge, this the first report of contrast-enhanced ultrasound aspects of idiopathic carotidynia.

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Abstract – WCN 2013**No: 2444****Topic: 3 – Stroke****Profile impact of depressive symptoms after stroke**Z. Adwan. *Privat Clinic, SMS HOSP, Swaida, Syria*

Objective: The impact of the profile of depressive symptoms after stroke using the Stroke Impact Scale (SIS).

Background: Stroke effects have impacts on memory and thinking, communication, and social functioning. The effect of depression on functional outcomes.

Methods: A total of 287 stroke patients recruited from acute and sub-acute facilities in a community were followed prospectively from stroke onset till 3 months post-stroke. All subjects were assessed with standardized stroke outcome measures. Subjects were also assessed with SIS about 90–120 days after stroke.

Results: An average age 73 ± 10.1 years and 47% men. 94% were ischemic stroke patients and the median baseline NIHSS was 6. On average, this stroke cohort has 4.2 depressive symptoms. Mean scores of SIS domains were 62, 78, 74, 81, 60, 67, 56 and 59 for strength, memory/thinking, emotion, communication, mobility, ADL/IADL, upper extremity and participation, respectively, with 0 being the worst and

100 being the best. The baseline GDS scores (number of depressive symptoms) significantly correlated with strength; memory and thinking; communication; ADL/IADL; mobility; hand function; and social participation. Mean SIS domain scores were significantly different between subjects who were considered depressive ($GDS \geq 6$) and subjects who were not depressive ($GDS < 5$) (all $p < 0.0005$). Mean scores of communication domain were significantly different between the two groups at the 0.0119 level.

Conclusions: Depressive symptoms affected more than just the physical aspects of functioning. They also restricted memory and thinking, communication, emotion and social participation.

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Abstract – WCN 2013**No: 2501****Topic: 3 – Stroke****Arterial stiffness and small vessel disease in patients with obstructive sleep apnea**J. Pretnar-Oblak^a, A. Resman-Gasparsic^a, L. Dolenc-Goselj^b, B. Žvan^a, M. Zaletel^a. ^aDepartment for Vascular Neurology, Ljubljana Medical Center, Ljubljana, Slovenia; ^bDepartment for Neurophysiology, Ljubljana Medical Center, Ljubljana, Slovenia

Background: Obstructive sleep apnea (OSA) is a common sleep disorder associated with increased body mass index, metabolic syndrome, hypertension etc. Stroke and in particular small vessel disease (SVD) is common in OSA patients. Pathophysiology of SVD is not well known. Increased arterial stiffness seems to play an important role. The aim of this study was to show that OSA is an independent risk factor for increased arterial stiffness. We hypothesized that patients with OSA have increased arterial stiffness compared to controls with similar cerebrovascular risk factors.

Methods: 32 subjects with OSA (average AHI 53.9) and 28 age and gender matched controls with similar risk factors were included. Common carotid artery stiffness was determined using β index. Magnetic resonance tomography of the head was performed in all patients and Fazekas scale was used to assess the stage of SVD.

Results: The two groups did not differ in cerebrovascular risk factors except for BMI (OSA: 35.0 ± 4.0 vs. control: 31.2 ± 4.1) ($p < 0.01$). Less OSA patients had a normal MRI exam but the difference was not significant (OSA: 43% vs. controls 54%). β index was similar in both groups (OSA: 8.0 ± 2.4 vs. controls: 7.0 ± 2.3) ($p = 0.11$). Logistic regression model has shown a significant relationship between BMI and OSA but not between AS and OSA.

Conclusions: Arterial stiffness in OSA patients does not differ significantly from controls with similar risk factors. OSA patients frequently have small vessel disease but there is no significant difference compared to controls.

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Abstract – WCN 2013**No: 2543****Topic: 3 – Stroke****Cerebral amyloid angiopathy (CAA), an often missed cause of intracranial bleeding**Y. Duodu^a, S. Mathews^b, S. El Tawil^a. ^aMilton Keynes General Hospital, Milton Keynes, UK; ^bHistopathology, Milton Keynes General Hospital, Milton Keynes, UK

Background: CAA is caused by deposition of amyloid in the walls of small cortical and meningeal vessels. Clinical presentations include

cognitive decline, cerebral haemorrhages and TIA like episodes. Early diagnosis is important to avoid catastrophic bleeding with antiplatelet or anticoagulant medication.

Objective: To demonstrate practical difficulties in diagnosis and management of CAA in patients with vascular risk factors.

Patients and methods: We present a case of pathologically proved CAA. Clinical presentation and investigation results are discussed.

Results: This gentleman initially presented to psychiatric services with memory problems at the age of 76. Brain MRI showed evidence of small vessel disease. Other vascular risk factors included hypertension, previous smoking and IHD. His medication included aspirin, clopidogrel, citalopram and ramipril. At the age of 79 he had a brief episode of language difficulties lasting for a few hours. One month later he developed a similar but longer episode. CT of the brain showed a right frontal subarachnoid haemorrhage. MRI of the brain 2 days later showed a further left superficial parietotemporal bleeding. Antiplatelets were stopped and the patient made a good recovery. Further fatal intracranial bleeding occurred 2 months later. Pathological examination confirmed CAA.

Conclusion: CAA is common in older patients who may have other vascular risk factors. Cognitive impairment and changes on CT or MRI are often attributed to ischemic small vessel disease. This case demonstrates the importance of obtaining specialised MRI sequences in older patients before decisions are made on the use of several antiplatelet agents or oral anticoagulation.

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Abstract – WCN 2013

No: 2524

Topic: 3 – Stroke

Rescue thrombectomy after failure of intravenous thrombolysis in acute ischemic stroke: Preliminary results of a multicenter prospective observational study

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Background: Intravenous (iv) thrombolysis has a poor rate (less than 30%) of recanalisation in proximally occluded mean cerebral artery (MCA). Thrombectomy has been shown to achieve a higher rate of recanalisation, up to 80%. The outcome of stroke patients is closely related to the recanalisation rate. A protocol of thrombectomy is prospectively performed in stroke patients with failure of iv thrombolysis in case of proximal MCA occlusion.

Material and methods: All patients admitted within the time window (4.5 h) for iv thrombolysis had an angioscanner to assess the level of occlusion of MCA. In case of no clinical recovery after iv thrombolysis, a thrombectomy was performed under general anaesthesia in case of persisting proximal occlusion. The device used was the Solitaire FR. The mRS and the NIHSS were performed at discharge, 3 and 6 months. A CT scan was performed at 24 h, and an angioMRI at one month.

Results: 16 patients underwent the procedure. There were 2 cases of procedural failure. The mean age is 64 +/- 10.5 years. The mean NIHSS at admission is 16 +/- 2. From stroke onset, the mean time of iv thrombolysis is 110 +/- 48 min, the mean time of stent deployment is 297 +/- 60 min. Good outcome (mRS 0 to 2) is 78.5% (11/14). Intracranial haemorrhage occurred in 35.7%. The post-procedure angiography showed 12/14 complete recanalisation (TIMI = 3).

Conclusions: Our first results show a favourable outcome of stroke patients with failure of iv thrombolysis successfully recanalised by thrombectomy.

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Abstract – WCN 2013

No: 2528

Topic: 3 – Stroke

Acute spinal cord infarction: Outcomes of a Portuguese center

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Introduction: Acute spinal cord infarction syndrome (ASCIS) is responsible for 5–8% of all acute myelopathies. Main causes include aortic pathology, atherosclerosis and infection. Current knowledge of long-term outcome is limited, but seems to be worst for patients with severe deficits and/or no initial improvement.

Objective: To examine clinical features and assess motor and functional outcome of patients with ASCIS.

Patients and methods: Retrospective analysis of 104 consecutive patients with spinal cord lesions (from 1989 to 2013). Ten patients presenting with ASCIS were included. Data concerning demographic and clinical variables were analyzed. Neurological syndrome was defined and initial and long term outcomes were assessed using the American Spinal Injury Association (ASIA) motor score and the Modified Rankin Scale (mRS).

Results: Five women and five men were included (mean age 56.3 years). In 60% of patients the first symptom was motor deficit. Possible causes were atherosclerosis (n = 4), hypoperfusion (n = 1), degenerative spine disease (n = 3) and cryptogenic (n = 2). Mean ASIA motor score was 71.0 ± 15.23 at onset and 75.3 ± 17.4 24 h after admission. Median mRS was significantly worse at discharge (median 4, range 1–4) when compared to admission (median 0, range 0–2) (p = 0.010), but there was no change at one year follow-up (median 3, range 1–4) when compared to discharge (p = 0.18). Worst ASIA scores 24 h after admission correlated with worst mRS at discharge (p < 0.05).

Conclusion: In this series motor outcome was fundamentally related to the severity of the neurological deficits at presentation. Nevertheless, the majority of patients regained ambulatory capacity.

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Abstract – WCN 2013

No: 2550

Topic: 3 – Stroke

Beyond DWI – Emerging candidate MRI biomarkers associated with risk of early stroke after TIA

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Background: Early recurrent stroke is a major cause of disability after TIA. The presence of acute DWI hyperintensity after TIA

symptoms is an independent predictor of early stroke risk and provides prognostic information when included in the ABCD³-I score. Little information exists on other MRI biomarkers for stroke risk.

Objective: We aimed to investigate the association between candidate new MRI biomarkers and early stroke risk in the Dublin BIO-TIA study.

Patients and methods: The BIO-TIA Study is a multi-hospital registry of TIAs over a 5-year period. Patients with stroke-physician confirmed TIA and MRI within 72 h of TIA onset were included and followed prospectively for early recurrent stroke, confirmed by a stroke physician. Standardized blinded MRI assessment was performed by a neuroradiologist.

Results: 230 patients met inclusion criteria, of whom 14 had stroke recurrence by 90 days. Early recurrence was associated with any acute DWI lesion (71.4% vs. 35.6%, $p = 0.007$, c -statistic = 0.68, OR = 4.53, CI = 1.27–14.87). Among candidate MRI biomarkers, 90-day recurrent stroke was associated with FLAIR Vascular Hyperintensity (16.7% vs. 3.4% in non-stroke patients, $p = 0.03$, c -statistic = 0.57, OR = 5.7, CI = 1.04–30.9), focal stenosis on intracranial MRA (25% vs. 3.7%, $p = 0.01$, OR = 8.8, CI = 1.22–63.1) and FLAIR chronic ischaemic lesions (28.6% vs 11.6%, $p = 0.065$).

Conclusion: In addition to acute DWI lesions, other MRI biomarkers were associated with stroke risk after TIA, including FLAIR Vascular Hyperintensity, focal intracranial stenosis, and FLAIR chronic infarcts. If validated, these non-DWI biomarkers may improve identification of patients at high early risk for intensive stroke prevention therapy in clinical practice or randomized trials.

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Abstract – WCN 2013

No: 841

Topic: 3 – Stroke

Perfusion computed tomography in the diagnosis of acute focal neurological symptoms: Ischaemic stroke vs. seizures

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Background: Misdiagnosing stroke may lead to unnecessary thrombolytic therapy. There are contradictory reports about the role of perfusion computed tomography (PCT) in differentiating stroke and seizures as causes of acute focal neurological symptoms.

Objective: To define the role of PCT in differentiating between those two conditions in acute setting.

Patients and methods: The study covered 35–85 year old patients with acute focal neurological symptoms, who underwent PCT and EEG within 12 h after symptoms onset. Presumptive ischaemic focus would be revealed in non-contrast CT in the 3rd day after symptoms onset or, when no CT changes appeared, in MRI. Neurological status was evaluated according to the National Institutes of Health Stroke Scale (NIHSS). Perfusion parameters were set up as asymmetry indices for corresponding regions of brain hemispheres. EEG was described in a 5-stage scale.

Results: Statistical analysis covered 17 patients with ischaemic stroke and 12 patients with focal neurological symptoms in the course of seizures. Those groups, taking potential effect of sex and age into account, were significantly statistically different for two parameters of PCT: MTT (mean transit time) and TTP (time to peak) in lateral middle cerebral artery territory (linear regression, p value < 0.05). TTP revealed to be the most significant parameter (logistic regression, $p < 0.05$). Analyzed groups were not significantly statistically different in NIHSS evaluation (T -test, p -value > 0.05) and EEG (Mann-Whitney test, p -value > 0.05).

Conclusion: PCT may differentiate between ischaemic stroke and seizures as causes of acute focal neurological symptoms.

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Abstract – WCN 2013

No: 2626

Topic: 3 – Stroke

The association of Bcl gene polymorphism and – 148C/T promoter β fibrinogen with plasma fibrinogen level in acute ischemic stroke

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Background: Stroke is a leading cause of disability and death. High fibrinogen levels are a risk factor for ischemic stroke and are associated with interaction of genetic and environmental factors.

Objective: To analyze the relationship of *gene polymorphism Bcl* and – 148C/T β fibrinogen with elevated levels of fibrinogen in acute ischemic stroke.

Method: This is a case control study with acute ischemic stroke patients and non-stroke in the Neurology Department of Dr. Wahidin Sudirohusodo Hospital, Makassar, Indonesia. The sample was determined based on their sexes, age group, risk factor, and taken by consecutive sampling technique. All samples of stroke were conducted, fibrinogen level, DNA isolation, PCR and RFLP, using the *HindIII* and *BclI* restriction enzymes. Furthermore, these were analyzed by electrophoresis and sequencing.

Result: There were a total of 216 samples consisting of 106 stroke and 110 non-stroke patients. The average fibrinogen level of the sample group is 423.48 (SD \pm 131.07) mg/dl, while in the control group was normal. There was significant correlation of high fibrinogen level and ischemic stroke with RO 16:50 (95% CI 7.72–35.26). The Chi-square analysis result proves that fibrinogen (cut off 375 mg/dl) plays a significant role in causing ischemic stroke (OR 16:50 $p = 0.000$). The polymorphisms of *BclI* gene and of – 148C/T have a correlation with stroke incidence (*BclI* gene: $p = 0.000$ and OR 2.16, as for – 148C/T: $p = 0.000$ and OR 6.84).

Conclusion: *BclI* gene and – 148C/T were significantly associated with an elevated plasma fibrinogen level, both genes are also risk factors for stroke ischemic events.

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Abstract – WCN 2013

No: 2100

Topic: 3 – Stroke

Capability of navigated repeated transcranial magnetic stimulation in stroke rehabilitation (Randomized blind sham-controlled study) (Clinicaltrials.gov identifier: NCT01652677)

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Background: Among several noninvasive brain stimulation techniques repetitive transcranial magnetic stimulation (rTMS) demonstrates beneficial effect in motor recovery after stroke. The transcranial magnetic stimulation with MRI navigation (nTMS) permits to take into account individual brain anatomy and to repeat stimulation focally.

Objective: Examine effectiveness, safety and tolerability of different regimens of nTMS for stroke rehabilitation.

Patients and methods: The randomized blind sham-controlled study of repetitive nTMS of primary motor cortex for motor stroke rehabilitation was started. 22 patients have been recruited (mean age = 55.15 ± 8.77 years). Patients were randomly assigned to one of the four groups:

- 1) sham group (n = 4);
- 2) low-frequency stimulation group (n = 6);
- 3) high frequency stimulation group (n = 9);
- 4) both hemispheric stimulation: low-frequency to unaffected hemisphere than high-frequency to affected (n = 3).

For nTMS we used NBS eXimia Nexstim and Magstim Rapid2. Clinical condition of patients was assessed with a set of scales before and after stimulation.

Results: Patients in the fourth group have significant better clinical output: Fugl-Meyer scale distinction = 27.00 ± 12.50 (p = 0.003, U-test). Preliminary results show the relief of the post-stroke spasticity and pain after high-frequency stimulation of the affected M1. Motor improvement was demonstrated for low-frequency stimulation of the unaffected M1. Headache appeared at 6 patients (27%) after stimulation which held itself. Four patients had an increase of epileptiform discharges on EEG (45%). Generalized epileptic seizure occurred in 2 patients (9%) during TMS.

Conclusion: Repeated nTMS is a safe and effective add-method in post-stroke rehabilitation, but continuous study and forming protocols are necessary to validate this method.

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Abstract – WCN 2013

No: 2613

Topic: 3 – Stroke

Clinical and radiologic imaging study of patients with ruptured and unruptured cerebral aneurysms submitted to the endovascular or microsurgical treatment

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Introduction: Endovascular therapy is an effective method to treat cerebral aneurysms due to it being a low invasive procedure and causes less complication than microsurgical technique, the standard approach.

Objective: Perform a clinical and radiologic imaging study of patients with ruptured and unruptured cerebral aneurysms submitted to the endovascular or microsurgical treatment.

Material and methods: Patients treated between 2001 January and 2009 December were analyzed through protocol including sex, age, subarachnoid hemorrhage presence or absence, aneurysm variety, size and more frequently localization, multiplicity, clinical complications regarding hemorrhage and embolization, associated diseases, dome-to-neck ratio, sort of treatment, retreatment, angiographic control and Fischer, Hunt–Hess and Glasgow Outcome scales. We studied 105 patients and discovered 146 aneurysms using transversal observational study. Statistic study has p < 0.05.

Results: Aneurysms were most common in the posterior communicating artery of women in the sixth decade. Patients submitted to the endovascular treatment had satisfactory evolution when analyzed by Glasgow Outcome Scale in 3 months and by angiographic control in 1, 3 and 5 years.

Conclusion: Endovascular therapy is an effective method comparing with microsurgery to therapeutic approach of cerebral aneurysms and its symptoms.

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Abstract – WCN 2013

No: 2608

Topic: 3 – Stroke

Tissular and motor analysis of neuroprotective and anti-inflammatory effects of copaiba oil (*Copaifera reticulata* Ducke) after focal cortical ischemia

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Background: Cerebrovascular diseases are a common cause of mortality worldwide and brain ischemia is its main manifestation, inducing a large variety of injuries and neurological processes.

Objective: Evaluate the antiinflammatory and neuroprotective effects of Copaiba oil, in tissue and motor preservation after focal ischemia in the motor cortex, with Endothelin-1 (ET-1).

Material and methods: 16 Wistar adult rats were distributed in groups of ischemia + tween (GIT), ischemia + Copaiba (GIC) and nonischemic (GNI), examined microscopically and by behavioral tests, with emphasis on locomotor activity, motor coordination and balance of the animals.

Results: Microinjection of ET-1 induced tissue loss, with necrotic features and tissue edema, accompanied by intense inflammatory response and ischemic penumbra area in the GIT animals. In GIC the reduction of inflammatory response, lesional edema and ischemic penumbra with tissue preservation were observed. Evaluated locomotor activity in the GIT noticed a significant reduction when compared to GNI (p < 0.0001). Comparing the GIC with the GIT, there was a statistically significant difference between the two groups (p < 0.0001). For the evaluation of GIC compared with GNI, a significant difference (p < 0.05) was found. In the evaluation of coordination and balance, the result reveals that the groups showed no statistical difference.

Conclusion: It was found that copaiba reduced the inflammatory response, with repercussions on the improvement of behavioral performance of the animals in locomotor aspect, setting it as a neuroprotective factor.

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Abstract – WCN 2013

No: 2605

Topic: 3 – Stroke

An ominous disease mimicking multiple sclerosis

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Background: Multiple sclerosis is a CNS demyelinating inflammatory disease defined by clinical demonstration of disease dissemination in space and time, after excluding other etiologies that could explain the neurologic manifestations (2010 McDonald criteria). Paraclinical evidence (MRI, visual evoked potentials, CSF analysis) also support the MS diagnosis. The attempts to fulfill the criteria could mislead clinical investigation, forcing an equivocal MS diagnosis while disregarding the correct etiology.

Objective: To present a case of severe cerebrovascular disease first diagnosed as multiple sclerosis.

Case report: A 50-yr-old, hypertensive, smoker, Caucasian male was referred to our reference center for treatment of CNS demyelinating diseases for consultation. He had received the diagnosis of multiple sclerosis relapsing–remitting in the month before, and was sent to us for the purpose of human immunoglobulin therapy – since a 5 day course of methylprednisolone was ineffective. The neurological examination revealed severe ataxia of limbs and trunk, slighted decreased muscle strength, brisk deep tendon reflexes in all limbs and bilateral Babinski sign, left internuclear ophthalmoplegia, right hemifacial

hypesthesia. He also exhibited frequent presyncope when standing in upright position. The symptoms started around 50 days earlier, and were evolving. Five years before, he noticed episodes of sudden weakness of his left leg, with instantaneously improvement.

Gadolinium-enhanced brain MRI showed T2-hyperintense lesions in the inferior thalamus, cerebral peduncles, pons, cerebellum and a small periventricular lesion. Brain angiography pointed complete occlusion of basilar artery and irregular pericallosal arteries.

Conclusion: We reported a case of total basilar occlusion confounded with multiple sclerosis presentation.

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Abstract – WCN 2013

No: 2475

Topic: 3 – Stroke

Ischemic stroke revealing a celiac disease: A case report and review of the literature

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Objective: To report a case of celiac disease 'CD' revealed by ischemic stroke.

Case report: A 38 year old woman is investigated for ischemic stroke associated with generalized convulsive crisis which are stabilized by medical antiepileptic drugs. The examination does not show any sign of clinical call suggesting a disease thrombosis (systemic, neoplastic...). We noted an iron deficiency anemia (hemoglobin at 9 g/100 ml microcytic hypochromic anemia). The levels of calcemia, cholesterol, glycemia, prothrombin index, and albumin are normal. The cardiovascular morphological assessment (Holter ECG, echocardiography, Doppler of the supra-aortic trunks 'SAT'...) do not objectify any abnormalities (patent foramen ovale, stenosis, thrombosis...). In addition the homocysteinemia, C, S, and antithrombin protein levels are normal. Leiden factor's mutation, antiphospholipid antibodies, anti nuclear antibodies, the flux cytometry etc. are without abnormalities. The anti-gliadin and antitransglutaminase of the type (IgA) antibodies are strongly positive and of this fact the diagnosis of the CD is established.

Discussion: The absence of cardiovascular diseases or the metabolic risk factor of young's stroke and the negativity of the immunological assessment plead in favor of the existence of a causal link between the CD and the ischemic stroke. The most widely incriminated factor is autoimmune central nervous system vasculitis, in which tissue transglutaminase, the main auto-antigen contributing to maintaining the integrity of endothelium tissue, plays a major role like reporting in our observation.

Conclusion: Being a potentially treatable cause of stroke, CD disease must appear among the etiologies to seek in front of stroke of unknown cause especially in young patients.

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Abstract – WCN 2013

No: 2525

Topic: 3 – Stroke

Epidemiology study of stroke about 1256 cases

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Background: Stroke is the frequent cause of hospitalization in the neurology department.

Objective: We sought to clarify epidemiology, clinical and etiology aspects of stroke on a Moroccan study.

Material and methods: The authors present a retrospective study of 1256 cases of stroke selected among the hospitalized patients at the Department of Neurology of Military Hospital Mohammed V during a period from January 1st 1997 until December 31, 2012. The patient may fulfill the criteria of ad hoc committee of cerebrovascular disease and have imagery which confirms stroke, and minimum of laboratory data.

Results: This analysis showed male predominance (77.5%). The patients are aged 24 to 104 years. The most frequency localization was territory of the middle cerebral artery (75.6%). The etiology was multiple, but dominated by angiitis and cardiomyopathy before 45 years and by atherosclerosis over this age.

Conclusions: Our study showed that the characteristics of stroke in our country were not different of other studies in the world.

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Abstract – WCN 2013

No: 2380

Topic: 3 – Stroke

Sports-related ischemic stroke

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Background: Ischemic stroke symptoms installation during usual sports activity (Sports-related stroke) has been rarely reported, with only small case series in the literature.

Objective: Describe clinical cases of patients with acute stroke with installation during usual sports. We analyzed the clinical features, imaging and prognosis.

Patients and methods: Review of patient database of our Neurology Service, searching for patients with acute stroke during sporting activity (2010–2013).

Results: We identified 3 patients (2 men) with a mean age of 54 years (47–59). First patient was 47 years old, with a history of hypertension, stroke during gym manifested by paresis of the left upper limb, in the ER with NHISS 4, OSCP-PACI, TOAST indeterminate; outcome: mRS NHISS 3 and 2. Second patient was 57 years old with a history of dyslipidemia, stroke during football practice manifested by imbalance and right hemiparesis, in the ER with NHISS 13, OSCP-POCI, TOAST indeterminate; outcome: mRS NHISS 6 and 2. Third patient was 59 years old, with a history of hypertension and dyslipidemia, stroke during running manifested by imbalance and right hemiparesis and global aphasia in the ER with NHISS 17, OSCP-PACI, TOAST cardioembolic; outcome: mRS NHISS 5 and 1.

Conclusion: Although our case series is too small, we can observe that patients had good functional recovery. It will also be taken into account that patients had low risk factors. We cannot correlate etiology with incidence or prognosis.

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Abstract – WCN 2013

No: 2334

Topic: 3 – Stroke

Prevalence of Fabry disease in young patients with ischemic stroke in Tokyo Japan

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Background: Fabry disease is an X-linked lysosomal storage disorder caused by mutations in the alfa-galactosidase (alfa-GAL) gene. Recently, the multicenter European study Stroke in Young Fabry Patients (sifap) reported that definite Fabry disease was diagnosed in 0.5% of all patients. This European study suggested an increased prevalence of unrecognized Fabry disease in stroke patients. In Asia, the frequency of Fabry disease with stroke remains unclear. We investigated the prevalence of Fabry disease in young patients with first ischemic stroke or transient ischemic attack (TIA) in Japan.

Objective: To reveal the prevalence of Fabry disease in young Japanese ischemic stroke patients.

Patients and methods: We recruited young Japanese (aged 18 to 55 years), which admitted with first ischemic stroke or TIA to the neurology department of Nippon Medical School or Fuchu Keijinkai Hospital in 2010 to 2012. Those hospitals are serving the greater metropolitan area of Tokyo. The leukocyte alfa-GAL activity was assessed in all patients, followed by genetic testing for alfa-GAL gene in those with low enzyme activity.

Results: Among a total of 86 patients (69 males and 17 females), definite diagnosis of Fabry disease was established in one (1.1%) male patient, in whom a complete enzyme deficiency was found. The mutation R342Q was identified in this patient.

Conclusion: Our study suggests that the prevalence of Fabry disease in young Japanese ischemic stroke patients is probably below 1.4% of male patient. Further large sample size study is needed.

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Abstract — WCN 2013

No: 2584

Topic: 3 — Stroke

Fabry disease northern Sardinia: Screening in young adults with cerebrovascular disorders

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Background: The etiologic determination of stroke in young adults remains a diagnostic challenge in up to one-fourth of cases. Increasing evidences, since 2005, lead to consider Fabry disease (FD) as a cause to check on juvenile stroke.

Objective: We aimed to evaluate frequency of unrecognized FD in a cohort of patients with cerebrovascular disease.

Methods: We assessed prevalence of FD in 107 patients consecutively admitted to our Neurological Department for ischemic stroke, transient ischemic attack, intracerebral hemorrhage, neuroradiological evidence of white matter lesion even if in the absence of clinical manifestation and cerebral venous thrombosis. The qualifying events have to occur between 18 and 55 years of age.

Results: We found 2 patients affected by FD, with a prevalence of 1.9. The first patient included for silent stroke, the second one for ischemic stroke.

Conclusions: Our study is in a middle position between studies that found prevalence up to 4% and other researches that retrieve no FD diagnosis.

We are prone to think that FD should be always considered in the differential diagnosis and in the case of patients with personal or familial positive anamnesis for cardiologic and/or renal concomitant involvement or recurrence of cerebrovascular events. FD should be suspected not only in cryptogenic stroke, but also in the other TOAST classes and cerebrovascular disorders, paying particular attention to silent strokes.

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Abstract — WCN 2013

No: 554

Topic: 3 — Stroke

Survival of intracerebral hemorrhage in Chile. A population based register

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Background: The average incidence of intracerebral hemorrhage (ICH) is 25 per 100,000 inhabitants in the world; in Chile is 20 per 100,000. The mortality of hemorrhagic stroke is between 30 and 45%.

Objective: To stud intrahospitalary mortality and survival of ICH through the country.

Method: Is a descriptive and ecological study of national discharge database from 2003 to 2007 from ICD-10 category I 61.0 (0 to 9) and I 62.9 of the National Health Ministry (MINSAL) and National Socioeconomic Survey (CASEN). A survival analysis estimated long term survival. Kaplan–Meier survival curves and Cox proportional hazard models were used to demonstrate predictors.

Result: 13,256 registered and 10,267 persons were identified, 93.6% had only one event. 37% died during the hospitalization, 44% the first day and 70% before five days. The survival is higher in female than in male ($p = 0.02$). The higher case-fatality rates are in IV, VI, VIII and Metropolitan regions (≥ 40 per 100,000). The lethality has been decreasing from 42.6% to 36.4%. The correlation of lethality rates was only with poverty ($r = 41\%$, $p = 0.0001$).

Conclusions: The lethality was decreasing during the study period. Most of the patients died during the first five days, survival is higher in women.

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Abstract — WCN 2013

No: 2273

Topic: 3 — Stroke

Carney complex in a young patient with ischemic stroke

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Introduction: Carney complex is a rare neoplasia syndrome characterized by atrial and cutaneous myxomas, endocrine abnormalities and testicular tumors.

Case report: Male, 29 years old, presented with sudden onset of horizontal diplopia, dysarthria, paresthesia of the left hemibody and gait imbalance; with a previous history of unilateral orchiectomy at the age of 14 for testicular cancer. He denied family history of skin changes or cardiac or endocrine problems. Observation disclosed the presence of multiple cutaneous pigmented lentiginous lesions (including lips, conjunctiva, oral and genital mucosa) and a small exophytic papule in the dorsal region. The neurological examination revealed dysarthria, bilateral appendicular ataxia and ataxic gait. MRI showed multiple recent ischemic lesions (cerebellum, pons and mesencephalic bilaterally and left temporo-occipital). Transthoracic and transesophageal echocardiogram revealed a large and friable mass located in the left atrium, attached to the atrial septum. The patient underwent cardiac surgery with successful and total extraction of the tumor, whose pathological diagnosis was myxoma. The endocrinological evaluation revealed increased IGF-1 and urinary cortisol (24 h) and testicular microcalcifications in ultrasound examination. The histopathological diagnosis of the papule

was cutaneous myxoma. Though the patient met the diagnostic criteria for Carney complex, PRKAR1A sequencing was negative for the presence of mutations in the coding region and flanking intronic regions.

Conclusion: We report a case of cardioembolic ischemic stroke in a young patient leading to Carney complex diagnosis. Recognizing this rare situation is important since specific patient follow-up must be planned for early detection of potentially fatal tumors, including atrial myxoma.

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Abstract – WCN 2013

No: 2401

Topic: 3 – Stroke

Red blood cell distribution width (RDW) association with ischemic stroke among adults younger than 55 years

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Background: Red blood cell distribution width (RDW) measures the size variability of these cells. It has been recently reported as a novel independent marker of macrovascular disease, including stroke, even at normal range values.

Objective: Evaluation of RDW values in adults younger than 55 years presenting with ischemic stroke.

Patients and methods: All patients aged under 55 years and presenting with ischemic stroke during 2012 were enrolled. Socio-demographic information was registered before clinical examination in addition to RDW, cholesterol, triglycerides, smoking, alcohol use, presence of diabetes and arterial hypertension. Cases with anemia were excluded.

Results: 158 patients (133 males, 25 females), aged 16–55 years (mean 48.4 years, SD 7.22) presented with ischemic stroke. RDW values were 14.27% (SD 1.1%, normal range being 10–15%). No case presented with RDW under 12%. Cases were divided in quartiles: 9.6% had 12–13% RDW value (1st quartile), 30.8% had 13–14% RDW value (2nd quartile), 34.6% had 14–15% RDW value (3rd quartile), and 25% had >15% RDW value. Hypertension was present in 90 patients (57%). Diabetes was present in 42 patients (26.6%). 58 patients (36.7%) were smokers. 48 patients (30.9%) were alcohol users. Mean cholesterol level was 211.4 mg/dl (SD 66.71 mg/dl). Mean triglyceride level was 170.3 mg/dl (SD 95.32 mg/dl).

Conclusions: Adults under 55 years of age presenting with ischemic stroke have upper normal or over the range RDW values, therefore RDW may be of great help as a future risk marker for ischemic stroke, being a cheap and readily available clinical laboratory examination.

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Abstract – WCN 2013

No: 2659

Topic: 3 – Stroke

Influence of antiplatelet treatment on the NIHSS score after intravenous thrombolysis

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Background and aims: The influence of antiplatelet treatment on the NIHSS scores and the risk of cerebral hemorrhage after intravenous rt-PA are not well known. We assessed the effect and the safety of rt-PA under antiplatelet agents (AP).

Methods: We studied data from 78 patients with ischemic stroke undergoing intravenous rt-PA within 3 h after onset, between April 1, 2006 and November 30, 2011. The patients with anticoagulation were excluded. Neurological improvement is defined by reduction of 2 points in the NIHSS score within the first 24 h after intravenous rt-PA. We assessed all 6 h and 24–36 h follow-up CT and MRIs for cerebral hemorrhage.

Results: 70 patients were included; 50 men, mean age of 71 ± 11 , 20 patients used AP prior to rt-PA, 14 with aspirin, 1 with ticlopidine, 1 with clopidogrel, 1 with cilostazol, 1 with cilostazol and clopidogrel, and 1 with aspirin and ticlopidine. The proportion of patients with recovery on the NIHSS scores was 18 patients in the AP group, 28 patients in the non-AP. Recovery on the NIHSS scores was higher in the AP group compared to the non-AP group. Any type of cerebral hemorrhage was observed: 4 patients in the AP group, 19 patients in the non-AP group. There is no significant difference between the AP group and non-AP group. None of the patients between two groups experienced symptomatic cerebral hemorrhage.

Conclusions: Our study disclosed that intravenous rt-PA with patients using AP is effective and safe.

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Abstract – WCN 2013

No: 2655

Topic: 3 – Stroke

Risk factors and etiology of ischemic stroke in young adults admitted to the stroke unit of Policlinico “Gemelli” of Rome

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Background: Approximately 10–14% of ischemic strokes occur in young adults.

Objective: To investigate risk factors and etiologies of young adults admitted to the “Stroke Unit” of Policlinico “Gemelli” of Rome from December 2005 to January 2013.

Patients and methods: 150 consecutive patients aged ≤ 50 years diagnosed with ischemic stroke. Clinical evaluation consisted of a complete neurologic examination and the National Institutes of Health Stroke Scale. Diagnostic work-up consisted of anamnesis, extensive laboratory, radiologic, and cardiologic examination. Stroke etiologies were classified according to the Trial of Org 10172 in Acute Stroke Treatment (TOAST).

Results: Patients' mean age was 41 ± 8 years. The most common risk factors were dyslipidemia (52.7%), smoking (47.3%), hypertension (39.3%), and patent foramen ovale (32.8%). Large-artery atherosclerosis (TOAST 1) was diagnosed in 17 patients (11.3%). Cardioembolism (TOAST 2) was presumed in 36 patients (24%), most of them presented a PFO at transesophageal echocardiography. Small-vessel occlusion (TOAST 3) was diagnosed in 12 patients (8%); all of them were hypertensive and most of them presented additional risk factors. 41 patients (27.3%) presented a stroke of other determined etiology and 44 (29.3%) presented a stroke of undetermined etiology. 3-year survival was 96.8%; recurrent strokes occurred in only 3 cases.

Conclusion: Traditional vascular risk factors are very common also in young adults with ischemic stroke, but probably they increase the susceptibility to stroke being dependent on other causes, as TOAST 1 and 2 represent less than 20% of cases. Prognosis quoad vitam is good, being characterized by low mortality and recurrence rate.

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Abstract – WCN 2013**No: 1340****Topic: 3 – Stroke****Optimum site for cTBS in stroke-patients:****A quantitative evaluation**

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Background and objective: Continuous Theta Burst Stimulation (cTBS) is a new trans-cranial magnetic stimulation protocol that transiently alters cortical excitability in the human brain. We aimed to test the proper stimulation for the recovery of the target site.

Subjects and methods: Two stroke patients at the restorative stage (A. 68-year-old female; B. 74-year-old male) were treated with informed consent (Uekusa Gakuen University and Nissan Tamagawa Hospital Ethics Committee approved the study). cTBS was applied to the non-affected side of M1 (hand, shoulder area) for 200 times consecutively for 40 s. Motor evoked potential (MEP) before and after cTBS was compared. The motion of the paretic arm was analyzed by motion capture method with Frame-DIASIV (DKH) software. The changes in angle and velocity of the movement of several U/E joints were measured.

Results: Both patients showed decreased MEP after cTBS. Patient A showed moderate improvement at the fingers but poor at the shoulder. cTBS targeting the fingers showed more effects than when targeting the shoulder. Of the fingers, the thumb had better outcome. Patient B showed moderate improvement at the shoulder but poor at the fingers by both stimulations targeting the fingers or shoulder. The targeting site of cTBS is not corresponding to the recovery site. The mechanism of the improvement is not only from regaining the affected M1 but also from the change of the spasticity, associated reaction and the other factors.

Conclusion: cTBS is a potential tool for the treatment to the stroke patients. Our results showed cTBS can regain the motor function through several mechanisms.

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Abstract – WCN 2013**No: 1396****Topic: 3 – Stroke****Kinesiological evaluation after cTBS to contralesional motor cortex in restorative stage stroke patients**

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Background and objective: Continuous theta burst stimulation (cTBS), is a repetitive trans-cranial stimulation technique that causes significant inhibition of synaptic transmission for over 1 h when applied to the primary motor cortex (M1). cTBS studies for the stroke related symptoms such as motor paralysis and neuropsychological dysfunctions at the chronic phase were reported. We aimed to investigate whether cTBS over the unaffected site of the M1 can induce an improvement of the upper arm at the restorative phase. For the evaluation of the improvement, the shoulder joint angle and

velocity of the movement were measured by the motion capture analysis.

Subjects and methods: Five stroke patients (mean age: 74.9 yrs) were treated under the informed consent (Uekusa Gakuen University and Nissan Tamagawa Hospital Ethics Committee approved the study). cTBS was applied to the unaffected side of M1. Paretic shoulder motion during flexion and abduction was analyzed with Frame-DIAS VI software. Baseline maximal angle and velocity prior to cTBS (600 stimuli, at 80% active motor threshold) or sham stimulation were set at 100 and post-stimulation change was calculated. Pre- and post-scores were compared by Mann–Whitney test.

Results: cTBS improved shoulder movements. In particular, cTBS significantly improved shoulder joint abduction angle and flexion velocity ($p < 0.05$).

Conclusion: The findings demonstrated the feasibility and efficacy of cTBS on the restorative stage recovery. Even at the restorative stage the inhibitory effect of cTBS resulted in improved paretic arm movement with faster time and wider range. The results have important implications for the use of cTBS for stroke rehabilitation.

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Abstract – WCN 2013**No: 2652****Topic: 3 – Stroke****Balance and gait measures as predictors of cognitive function in post-stroke patients**

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Background: Stroke patients are at risk for developing cognitive impairment. While motor function changes and decline in physical performance may precede the onset of cognitive impairment, it is not easy to identify which stroke patients are likely to develop cognitive impairment.

Objectives: To test whether quantitative balance and gait parameters can enhance the prediction of post-stroke long-term cognitive outcome.

Methods: The TABASCO is a prospective study of first-ever mild-moderate ischemic stroke patients who were cognitively intact at baseline. Quantitative movement, balance and cognitive tests were obtained at admission, 6, 12 and 24 months later.

Results: Data obtained from 211 patients. During 2 years of follow-up, 15.6% were found to have clinically significant cognitive decline (CD). The CD group and cognitively intact group did not differ in their neurological deficits 6 months after the index event. Nonetheless, at 6 months, timed up and go (TUG) test times were longer and Berg Balance Scale (BBS) scores were lower in the CD group, compared to the cognitively intact group ($p < 0.001$). Multivariate regression showed that the TUG and BBS below 45 were significant independent predictors of cognitive decline ($p = 0.002$ and $p = 0.021$, respectively).

Conclusions: Our results show that balance and mobility are significant predictors for cognitive status after stroke. Relatively simple, performance-based measures of balance and mobility may enhance the identification of patients who have a heightened risk of developing cognitive decline. Using these tests may enable early adaptation of rehabilitation strategies to delay the onset of cognitive decline and dementia in post-stroke patients.

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Abstract – WCN 2013**No: 921****Topic: 3 – Stroke****The volume DWI method increases detectability of small ischemic lesions in patients with transient global amnesia**

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Background: Transient global amnesia (TGA) is a clinical syndrome of reversible anterograde amnesia that occurs acutely. One of the possible causes of TGA is an ischemic deficit in the hippocampus.

Objective: We studied the acute ischemic changes in the hippocampus following TGA onset using magnetic resonance imaging (MRI).

Patients and methods: A total of 13 patients who visited our hospital within three days after the onset of acute amnesia were clinically diagnosed with TGA. The patients were evaluated using the conventional diffusion-weighted imaging (DWI) with a 6-mm slice thickness and the volume DWI method to generate three-dimensional images with 1.6-mm isotropic voxels.

Results: The initial MRI study of each patient was performed within three days after TGA onset. High signal-intensity lesions in the hippocampus were detected in six of the 13 patients (46.2%) on conventional DWI, and seven of 11 patients (63.6%) using the volume DWI method. Subsequent MRI studies were performed in six patients within two days after the initial MRI examination. In three patients, the volume DWI method revealed high signal-intensity lesions in the hippocampus that were not detected on the initial MRI examinations. The conventional DWI was unable to detect these lesions at the subsequent MRI examinations. Ultimately, we detected hippocampal lesions in 11 out of the 13 patients (84.6%).

Conclusion: The volume DWI method and repeated MRI examinations are useful for detecting small ischemic lesions in the hippocampus after TGA onset. The results suggest the presence of a relationship between TGA and hippocampal ischemic deficits.

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Abstract – WCN 2013**No: 2650****Topic: 3 – Stroke****Is there an evidence to perform thrombectomy in patients over 80 years old?**

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Background: Endovascular mechanical recanalization (EMR) is the most effective therapeutic strategy regarding large artery occlusion in stroke. Advanced age has been identified as one of the strongest predictors of poor outcomes.

Objective: The aim of this study is to analyze the clinical outcomes after thrombectomy in elderly patients in comparison to younger ones.

Methods: Patients treated with EMR were registered prospectively. Patients eligible for treatment received either intravenous thrombolysis (IVT) as a bridging concept and/or intra-arterial thrombolysis (IAT) in addition to EMR. Outcome parameters and recanalization of the occluded vessels were documented using the National Institutes of Health Scale (NIHSS), the modified Ranking Scale (mRS) and the Thrombolysis in Cerebral Infarction (TICI) score, respectively. Follow-up evaluations were performed up to 90 days. Patients were divided into three groups: age <60 (n = 53); age 60–79 (n = 92); age ≥80 (n = 33).

Results: The baseline NIHSS score of all patients was 18. TICI 2b and TICI3 were documented in 70% of patients <60, in 74% of patients 60–79 and in 70% of patients ≥80. At 90 days follow up we documented 59% for patients who aged <60, 39% for patients who aged 60–79 and 18% for patients who aged ≥80 with mRS ≤2. The highest mortality rate was documented in patients aged ≥80.

Conclusion: Recanalization results were equal in all patient age groups. Despite a higher mortality rate, successful revascularization is strongly associated with improved outcome in elderly stroke patients treated with thrombectomy.

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Abstract – WCN 2013**No: 162****Topic: 3 – Stroke****Anxiolytic effects of 5-hydroxiadamantane-2-on under the conditions of local ischemia of brain in rats**

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Stroke is the most common life-threatening neurological disease, one of the major causes of death and disability worldwide. Prevalence of anxiety remains a common problem several years after the stroke. 5-hydroxiadamantane-2-on an adamantane derivative has cerebrovascular activity and improves cerebral blood flow by activating GABA-ergic system.

The aim of this study was to reveal and evaluate anxiolytic effect of 5-hydroxiadamantane-2-on after local ischemia.

White inbred male rats were used. Anxiety was evaluated by two main parameters of elevated plus-maze (EPM) test: percentage of entries to open arms (OA) and time spent on OA. As a model of acute local ischemia, the middle cerebral artery (MCA) occlusion was chosen. The control group didn't receive any drug, while the experimental group has received 5-hydroxiadamantane-2-on immediately after MCA occlusion and on the following 6 days (100 mg/kg, i.p., once daily). In both groups the neurobehavioral changes have been tested through EPM test a day before and on the 6th day after MCA occlusion.

The percentage of entries to OA and time spent on OA of control group before occlusion were 27.25 ± 9.69 and 33.52 ± 3.86 and on 6th day – 19.38 ± 14.2 and 17.25 ± 11.1, relatively. For experimental group the percentage of entries to OA and time spent on OA were 12.55 ± 7.1 and 21.0 ± 5.8 before occlusion and 36.66 ± 9.12 and 69.0 ± 22.33 on the 6th day.

Thus, the results have shown that 5-hydroxiadamantane-2-on displays anxiolytic-like effect in animals with anxiety caused by local ischemia and can be suggested for further investigation of its anxiolytic activity.

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Abstract – WCN 2013**No: 1516****Topic: 3 – Stroke****Correlation between anti-thrombotic drugs and hematoma expansion in acute intracerebral hemorrhage under strict blood pressure-lowering management: SAMURAI-ICH study**

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Background: Few reports have documented the effect of pretreatment of anti-thrombotic agents on hematoma expansion in patients with intracerebral hemorrhage (ICH) under strict blood pressure-lowering management in the acute phase.

Methods: Patients (n = 211) with hematoma in the supra-tentorial region were enrolled. Continuous intravenous administration of the anti-hypertensive drug nicardipine was commenced within 3 h of ICH onset at 10 hospitals in Japan, and systolic blood pressure was controlled between 120 and 160 mm Hg. The correlation between pretreatment of anti-thrombotic agents and hematoma expansion (growth volume and rate 24 h after admission) was analyzed.

Results: Subjects (n = 211; 81 females, mean age, 65.6 ± 12.0 years) had a median NIHSS score on admission of 13. Twenty-four patients were taking anti-thrombotic agents on admission, 14 aspirin alone, 1 aspirin + dipyridamole, 2 aspirin + cilostazol, 1 aspirin + clopidogrel, 2 ticlopidine alone, 2 cilostazol alone, and 2 warfarin. When divided into 3 groups according to hematoma volume on admission (≤3.0 ml, 3.1–11.9 ml, >11.9 ml), a significant difference (p = 0.008) was observed between patients with and without pretreatment of anti-thrombotic agents (mean growth volume 24 h after admission: not taking anti-thrombotic agents, 4.73 ml; taking anti-thrombotic agents, 12.15 ml) only in the 3rd tertile group (volume > 11.9 ml). By three months after discharge, four deaths unrelated to anti-thrombotic agents had occurred.

Conclusions: Pretreatment of anti-thrombotic agents is likely significantly related to hematoma expansion in ICH patients with higher hematoma volume (>11.9 ml) on admission under strict blood pressure management in the acute phase.

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Abstract – WCN 2013

No: 2752

Topic: 3 – Stroke

Hemorheological study in a group of patients with clinically silent multifocal vascular cerebral lesions

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Clinically silent vascular cerebral lesions/CSVCL have been a problem of great importance in neurology. It involves patients with risk factors for stroke such as hypertension, diabetes mellitus, and cardiac diseases with arrhythmia but our knowledge about the pathomechanism of these changes has not been clear enough. We analysed hemorheological profile in a group of 35 patients with CSVCL diagnosed by magnetic resonance imaging or computed tomography in relation to the control group/10 subjects without such changes. The following hemorheological parameters were estimated: relative blood viscosity at various shear rates, plasma viscosity, haematocrit and parameters of Quemada's rheological model (RCD as red cell deformability and RCA as red cells aggregation). We also estimated biochemical factors: fibrinogen, IgM, IgG, IgA, CRP, cholesterol and

albumin/globulin ratio [A/G]. We found a significant increase of red cell elasticity (p < 0.04) in a group of patients as well as a significant decrease of IgM level (p < 0.018), cholesterol (p < 0.04) and A/G (p < 0.036). We detected also the following significant correlations: between A/G and RCA/negative in patients, p < 0.05, cholesterol and RCA/positive in controls, p < 0.007, IgM and fibrinogen/negative in both groups, p < 0.04 and p < 0.01 respectively, IgM/fibrinogen ratio and RCA/negative in patients, p < 0.02, cholesterol and fibrinogen/positive in patients, p < 0.035. In conclusion we may suggest that a better red cell elasticity in patients results from a self-regulatory compensation. IgM molecules seem to play an opposite role to fibrinogen molecules in red cells aggregation phenomenon.

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Abstract – WCN 2013

No: 2696

Topic: 3 – Stroke

A case of acute cerebral infarction in young adult associated with the polycythemia vera

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Background: Several primary hematologic disorders have been associated with ischemic stroke. The main cause of stroke in the hematologic disorders is coagulopathy or thrombosis.

Purpose: We report one patient of acute cerebral infarction associated with polycythemia vera.

Patients and methods: A 44-year-old man admitted to our hospital was complaining of left extremity weakness. His symptoms occurred suddenly 2 days ago. He had been well before admission and his past medical history was unremarkable. He had no history of any medical or neurological illness. Vital sign was stable at admission. In the neurological examination during admission, he was alert and well oriented. In motor examination, left hemiparesis was Medical Research Council (MRC) grade 3. He had no sensory abnormality and no ataxia. Diffusion weighted imaging showed hyperintense signal on right MCA territory. Brain CT angiography showed no evidence of definitive intra and extra cranial aneurysm or stenosis. Laboratory studies revealed normal serum electrolytes and renal function except for elevated hemoglobin, which shows decreased erythropoietin. An ultrasound scan of the abdomen revealed splenomegaly and mild fatty liver. PBS revealed moderate polycythemia. Phlebotomy and BM biopsy were performed. Then he was diagnosed with polycythemia vera. He took hydroxyurea 500 mg bid daily.

Results: Base upon clinical manifestation and radiologic findings, we diagnosed right middle cerebral artery infarction. Laboratory studies and bone marrow biopsy revealed polycythemia vera.

Conclusions: The case presented herein is characterized by an association between acute cerebral infarction and hematologic disorder. We should be curious of other causes except for cerebral atherosclerosis when young adult patient without other risk factors was diagnosed with ischemic infarction.

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Abstract – WCN 2013

No: 2727

Topic: 3 – Stroke

Swallow therapy for dysphagia

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Swallowing disorder is a difficulty in moving food from mouth to stomach. After any cerebrovascular accident, 90% of patients encounter some form of swallowing disorder. Swallow therapy techniques reduce any delay in triggering the oral, pharyngeal swallow & improve pharyngeal transit time. Therapy procedures were designed to improve five areas of swallowing.

- Position of food in mouth
- Manipulating food in the mouth with tongue
- Chewing the bolus of varying consistencies
- Recollecting bolus in oral cavity
- Organizing lingual action to propel the bolus posteriorly.

Case: A 57 years old male patient came with a history of pontine infarct 15 days back, complaining of slurring of speech and dysphagia. He was fed by NG tube.

MRI findings: *Acute pontine infarct*

Barium swallow: Right pyriform fossa pooling.

Dysphagia assessment: A detailed assessment of swallowing revealed:

- Delayed bolus transit time.
- No anterior and posterior propulsion
- Food residue after eating
- Gurgly and wet voice quality
- The patient had pharyngeal dysphagia with all diet and liquid levels.
- Frequency of therapy was decided four times a week.

Treatment plan: Therapy plan consists of three important techniques oral motor stimulation, postural maneuvers and diet modifications.

Result: With the help of these techniques he was able to take partial oral feed within one week. After 2 weeks he was able to take regular food but felt a little difficulty in thin liquid diet. This clearly shows that swallow therapy works well for patients who have difficulty in swallowing.

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Abstract – WCN 2013

No: 2743

Topic: 3 – Stroke

Silent cerebral infarctions and MR perfusion imaging in children with sickle cell anemia

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Objectives: Children with sickle cell anemia (SCA) and clinically “silent” cerebral infarcts (SCI) have poorer academic performance and lower intelligence quotients compared to healthy peers, and are at higher risk of stroke. Mechanisms that lead to SCI, however, remain poorly understood. Here we determined whether occurrence and volume of SCI in a neurologically intact children with SCA who did not have intracranial stenosis were related to alterations in cerebral blood flow (CBF).

Methods: We studied prospectively a cohort of 62 children with SCA without prior clinically overt stroke or TIA (median age, 8.8 years; range limits, 2.3–14.4 years; 33 females) and mean velocity <170 cm/s on transcranial Doppler ultrasound. They underwent MR-angiography of intracranial arteries, anatomical MR-imaging and continuous arterial spin labeling (CASL) MR-perfusion imaging of the brain.

Results: We found silent infarcts in 20 children (32.3%, 95% CI: 21.9–44.7%). The median volume of SCI in relation to total brain volume was 0.020% (range limits, 0.001–0.101%). Total CBF mean value was 81 ± 18 ml/100 g/min. CBF in the right hemisphere (79 ± 19 ml/100 g/min) was significantly lower than in the left hemisphere (82 ± 18 ml/100 g/min, $p = 0.025$). Low CBF was associated with high volume of SCI ($p = 0.042$; $F = 4.33$; $R^2 = 0.07$). Low hemispheric CBF was associated with high occurrence ($p = 0.021$; $\rho^2 = 0.036$), number and volume of SCI (Spearman $R = -0.199$, $p = 0.027$ and $R = -0.210$, $p = 0.021$ respectively). Also, systolic blood pressure was an independent predictor of high occurrence of SCI and their bigger volume.

Conclusion: High occurrence of SCI and their bigger volume are associated with low CBF and high systolic blood pressure in children with SCA.

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Abstract – WCN 2013

No: 2744

Topic: 3 – Stroke

The effect of nasogastric tube in stroke patient with swallowing disorder

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Background: Nasogastric (NG) tube is common route of feeding in stroke patient with swallowing problem. The size of tube has an effect on both patient and food passage.

Objective: To determine the effect of NG tube size on feeding in stroke patient.

Material and methods: A randomised cross-over study was conducted among 10 stroke patients (eight male, median age 66 years) in rehabilitation ward, Srinagarind hospital. All participants were fed via small-bore tube (10 F) and large-bore tube (14 F) for separate session (7 days for each session). Total feeding time, the event of tube obstruction, and patient satisfaction were recorded.

Results: Despite reports of patients being more comfortable in using small-bore tube, there was no significant difference between tube sizes in satisfaction ($P = 0.13$). There were 4 patients who experienced tube obstruction during placing of small-bore tube, whereas no report of obstruction in those placing large-bore tube. Median feeding time was longer while feeding via small-bore tube (49 min VS 29 min, $P = 0.03$).

Conclusion: Small-bore NG tube (10 F) may be used to improve patient comfort. Modified diet and well drug preparation should be used to reduce tube obstruction.

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Abstract – WCN 2013

No: 2701

Topic: 3 – Stroke

Vascular risk factors predisposing to white matter hyperintensities: Cross-sectional study

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Objective: To analyze risk factors predisposing to white matter hyperintensities (WMH) in patients.

Methods: 284 participants (mean age 50 +/– 5) were enrolled from local community and stratified into two groups: group 1 (without

WMH) and group 2 (with WMH). MRI studies were done on scan 1.0 Tesla (Philips panorama) with a slice thickness of 0.5 cm; T1 and T2, T2d–fluid attenuated inversion recovery (FLAIR) scans were reviewed.

Results:

Group 1 was consisted of 193 participants (age 48 ± 5 ; 124 women and 69 men).

Group 2 included 91 participants (age 53 ± 5 ; 55 women and 36 men).

Participants from group 2 were significantly older ($p < 0.02$).

The analysis for risk factors showed the prevalence of these factors in group 2 vs group 1, respectively: coronary artery disease (CAD) (11% vs 4%, $p = 0.03$); Diabetes Mellitus (DM) (6% vs 1%, $p = 0.005$); arterial hypertension (AH) (60% vs 33%, $p = 0.007$); and family history of vascular diseases (CVD) (25% vs 14%, $p = 0.03$) were associated negatively without WMH participants.

Thus, CAD, AH, DM and family history of CVD were significantly more common in participants with WMH.

Participants in group 2 were noticed to have AH grade 1 (140–159/90–99 mmHg) more often than were participants in group 1 (30% vs 19%, $p < 0.05$).

There was found no significant difference in prevalence of smokers and cardiac arrhythmia between the two groups (30% vs 29%, $p > 0.5$) and (1% vs 2%, $p > 0.5$).

Conclusion: We found that in our patient cohort WMH were associated with cardiac ischemia, diabetes, arterial hypertension, and family history of vascular diseases.

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Abstract – WCN 2013

No: 2719

Topic: 3 – Stroke

A Brain-Computer Interface for rehabilitation after stroke

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Brain-Computer Interface (BCI) technology has not only been used widely for communication but also for control purposes in a closed loop setup [1]. Recently, it has been shown that exercising motor imagery (MI), i.e. the mental rehearsal of movement execution, could be used as an effective therapy in stroke rehabilitation [2] even if no feedback about the performance is given to the user. However, when providing additionally a real-time feedback, the Hebbian plasticity can be elicited that is likely to result in an increased cortical plasticity, and has the potential to improve the functional recovery. In this manuscript a MI based Brain-Computer Interface (BCI) is connected either to an upper limb rehabilitation robot or a Virtual Reality (VR) system. Both, the VR system and the rehabilitation robot provide online feedback about the performed MI to the user. A total of eleven post-stroke patients and a control group of eleven healthy people took part in the VR based experiment and the first results on 4 healthy users performing the experiment with sensory feedback from the rehabilitation robot are available. Additionally, five of the stroke patients agreed to participate to further sessions, allowing to observe the possible improvements in accuracy due to training effects.

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Abstract – WCN 2013

No: 2721

Topic: 3 – Stroke

The association between urinary 6-sulfatoxymelatonin level, cognitive and emotional status and sleep quality in acute stroke

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Aim: To determine the association between night urinary 6-sulfatoxymelatonin (aMT6s) level, cognitive, emotional status and sleep quality in acute stroke.

Methods: 53 patients with ischemic stroke and 4 patients with subarachnoid hemorrhage (SAH) were examined with clinical (NIHSS) and neuropsychological methods: MMSE, FAB, MoCA-test, Semantic Verbal Fluency Test (SVFT), State-Trait Anxiety Inventory (STAI) and Pittsburgh Sleep Quality Index (PSQI). The assessment of aMT6s level in night urine was performed. For the comparison we used published data on aMT6s level in 49 healthy subjects.

Results: The stroke patients had lower aMT6s level in comparison with the control: 8.6 ± 6.92 vs. 27.2 ± 13.3 ng/mL ($p < 0.001$). Patients with ischemic stroke had lower aMT6s level than patients with SAH ($p = 0.036$). We found correlation between aMT6s level and NIHSS ($r = -0.29$; $p = 0.027$). The aMT6s concentration was interacted with MMSE subtests, especially «recall» ($r = 0.43$; $p = 0.001$); FAB ($r = 0.42$; $p = 0.01$), especially «mental flexibility» ($r = 0.29$; $p = 0.029$), «sensitivity to interference» ($r = 0.30$; $p = 0.025$) and «inhibitory control» ($r = 0.42$; $p = 0.001$); efficiency of categorical cue in delayed recall MoCA subtest ($r = 0.52$; $p = 0.0004$) and SVFT ($r = 0.29$; $p = 0.037$). Also aMT6s level was associated with state anxiety level ($r = -0.42$; $p = 0.041$) and PSQI ($r = -0.47$; $p = 0.024$). In general patients with low memory function and sleep quality had decreased aMT6s level ($p = 0.022$).

Conclusion: The aMT6s level was decreased in stroke patients. The reduction of aMT6s level was more pronounced in ischemic stroke than in SAH and depended on stroke severity. The association between low aMT6s level and domain-specific cognitive impairment, high level of trait anxiety and low sleep quality was found.

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Abstract – WCN 2013

No: 2723

Topic: 3 – Stroke

Vestibular evoked myogenic potentials in cerebellar and brainstem strokes

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Objective: Cervical vestibular evoked myogenic potential (cVEMP) is a short-latency myogenic response recorded over the sternocleidomastoid muscle (SCM) in response to saccular stimulation. The aim of this study was to make recordings in patients with brainstem and cerebellar strokes to see how lesions involving vestibular nuclei or the vestibulo-cerebellar connections affect the potentials.

Method: 20 patients with brainstem and cerebellar strokes were included in the study. Patients were seated on an armchair and the reference electrode was placed on the upper third of sternum and the ground electrode on the middle of the forehead. To record the surface EMG activity, an active electrode was placed on the upper half of the SCM ipsilateral to the stimulated ear. The latencies of peaks p13 and n23 and peak-to-peak amplitude of p13–n23 were measured and compared with the potentials recorded from 50 healthy controls.

Results: cVEMPs were recorded in all healthy controls and in 17 patients from both SCM muscles. p13, n23 latencies and p13–n23

amplitude recordings of these patients were not different from the healthy controls. In a patient with cerebellar stroke the potential from the ipsilateral SCM was absent. In two other patients (one with cerebellar and the other with lateral medullary infarction) contralesional responses showed latency and amplitude abnormalities.

Discussion: Normal responses recorded in most patients with vestibular nuclear or vestibulo-cerebellar lesions or contralesional abnormal responses cast doubt on the known pathways involved in the generation of the cVEMPs and necessitate further studies to understand the anatomical connections.

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Abstract – WCN 2013

No: 2254

Topic: 3 – Stroke

Early recurrent ischaemic stroke (ERIS) – under recognised cause of early neurological deterioration (END) in stroke patients undergoing thrombolysis

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Introduction: ERIS is a rare cause of END associated with intravenous thrombolysis. We present a case with ischaemic stroke with sudden neurological deterioration resulting in a new vascular territorial infarction during thrombolysis.

Case: A 67 year old gentleman with left-sided hemiplegia and dysarthria. Risk factors included atrial fibrillation and hypertension. NIHSS score was 16. CT scan revealed a right Middle Cerebral Artery (MCA) hyperdense sign. Just after the completion of thrombolysis, GCS dropped from 13 to 3 and NIHSS rose from 16 to 25. An urgent CT scan revealed a new left MCA hyperdense sign. MRI head showed extensive infarction encompassing both MCA, the left anterior and posterior cerebral artery territories and left pons. ECHO did not show thrombus in the heart. Intensive care treatment was withdrawn and patient subsequently died.

Discussion: The pathophysiological mechanism of ERIS is thought to be related to destabilisation of thrombus by tPA and a consequent release of emboli from a more proximal vascular location towards the brain. The incidence and prognosis of ERIS documented in the literature is poor. The incidence ranges from 0.2% to 1.0%. An evaluation of risk is impossible in the absence of clear characterisation, implying a need for larger scale multicentre data. In future, if we evaluate the risk, endovascular treatment can be utilised in treating ERIS in high risk cases.

Conclusion: Our case suggests AF is a risk factor for developing ERIS. Increased awareness and risk evaluation of ERIS would allow clinicians to consider endovascular treatment in future.

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Abstract – WCN 2013

No: 2729

Topic: 3 – Stroke

Presenting symptoms for the identification of acute stroke and transient ischemic attack in the emergency department

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Background: Formalized assessment tools, based upon easily collected clinical variables, may help emergency department staff diagnose acute stroke more efficiently. However, a substantial number

of patients detected by acute stroke response system (ASRS) turn out to be non-stroke patients.

Objective: To compare the difference of initial presenting symptoms between the patients with definite stroke and those without.

Methods: We prospectively recruited consecutive patients with suspected acute stroke during 6 months. An emergency department staff activated ASRS if they found:

- (1) patients were presented to emergency room within 6 h after symptom onset;
- (2) patients had at least one of 8 symptoms: unilateral weakness, unilateral numbness, loss of consciousness, difficulty in speaking or understanding, sudden loss of vision, diplopia, loss of equilibrium or dizziness, and headache with vomiting.

The difference of presenting symptoms of ASRS was investigated between the definite stroke and stroke mimics.

Results: We recruited 188 patients with suspected acute stroke by ASRS. The definite stroke was identified in 103 (54.8%). The patients with definite stroke (42/103, 40.8%) had more unilateral weakness than those with stroke mimics (6/85, 7.1%) ($p < 0.001$). On the other hand, the patients with stroke mimics (33/85, 38.8%) had more loss of equilibrium or dizziness than those with definite stroke (8/103, 7.8%) ($p < 0.001$).

Conclusion: Whether the patients were diagnosed as definite stroke or not was significantly associated with presenting symptoms of ASRS. The proper selection of presenting symptom for assessment tool used by emergency department staff may be important to improve its performance in ASRS.

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Abstract – WCN 2013

No: 2730

Topic: 3 – Stroke

Distribution of the corticobulbar tract in the internal capsule

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Background and objectives: It is generally thought that the corticobulbar tract descends through the genu of the internal capsule (IC). There have been several reports that genu lesions cause bulbar symptoms such as facial palsies, dysarthria, and dysphagia. However, the precise location of the corticobulbar tract in the IC remains controversial. The purpose of our study is to assess whether the corticobulbar tract passes through the IC genu.

Methods: We reviewed 26 patients with selective IC infarction and located the sites related to bulbar symptoms. In addition, using diffusion tensor imaging, we reconstructed tracts passing through the IC in ten subjects without cerebral infarction.

Results: Patients with genu infarction, which extended to more than half of the posterior limb of the IC, showed bulbar symptoms. However, patients with genu infarction, which was limited to the genu, did not have bulbar symptoms. In contrast, patients with lesions limited to the posterior limb may show bulbar symptoms. According to statistical maps of the region of interest, the lesions related to bulbar symptoms were localized to areas that were beyond the midpoint of the posterior limb of the IC. In diffusion tensor imaging of subjects without cerebral infarctions, the corticobulbar and corticospinal tracts did not pass through the IC genu.

Conclusion: Our data provide evidence that the corticobulbar tract does not pass through the IC genu. The proposed location of the corticobulbar tract in the level of the IC lies beyond the midpoint of the posterior limb.

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Abstract – WCN 2013**No: 2359****Topic: 3 – Stroke****Acute ischemic stroke, classification in terms of ethiopathogenesis, correlation between neuroradiological, clinical and prognostic findings**

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Background: Determination of 'core' which is defined as centre of ischemia, in other words tissue around the irrecoverable one and with the early diagnosis and emergent therapy that can be saved, causes an evident regression in mortality and morbidity because of the stroke. Besides the diagnosis, determining reperfusion signs at the perfusion imaging is useful to clarify the prognosis of stroke subtypes during the monitoring patients with acute stroke. Causitive Classification of Stroke is a standardized and reliable program which is organized by Harvard University. Everybody can reach this program by internet.

Objective: Our purposes were interpreting the correlations between DWI/PWI MRI mismatch methods and ethiological factors, an effect of this correlation on clinical findings and prognosis.

Material and methods: Totally, 30 patients with acute ischemic stroke were involved in the study. Cranial MRI imaging DWI/PWI MRI mismatch methods and clinical measurements that were determined by NIHSS and Glasgow outcome scale were performed three times at first 24 hours, at 48 hours and at discharge. CCS forms were filled with whole clinical and laboratory findings. rCBV, rCBF, and rTTP mapping were done with perfusion MRI.

Results: End of the study deficit areas on rCBV map was found to be effective on prognosis of patients. rTTP maps were used in the calculation penumbra. Also we detected that more meaningful prognostic results may be obtained with rCBV maps.

Conclusion: Early diagnosis and treatment of the disease affect the patient's quality of life significantly. Perfusion studies can be useable to appropriate patient selection eligible for thrombolytic therapy.

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Abstract – WCN 2013**No: 2732****Topic: 3 – Stroke****ICT-supported CVD prevention through phone-based automated lifestyle coaching**

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The development and maintenance of a healthy lifestyle (smoking cessation, healthy nutrition, moderate physical exercises etc.) is a major objective concerning the primary and secondary prevention of CVD. CAPSYS is a computer-based lifestyle coaching system, which aims at supporting CVD patients in performing appropriate behavior changes in order to minimize their individual risk factors. Patients can access CAPSYS by dialing a local-rate telephone number and answer to a set of previously known questions concerning their current nutrition, physical activity, blood pressure, smoking etc. In an interactive voice response approach, questions are issued by the system in natural language using a text-to-speech module, and the patient can provide the required values using the phone keypad (DTMF touchtone). Based on the gathered values for each patient, the system automatically generates personalized verbal feedback at runtime and presents it to the patient during the phone dialog. Depending on the individual

development of the patient's risk factors, the system feedback can contain advice for improvement, praise for healthy behavior and motivation to pursue a certain goal.

The automated feedback is generated based on rules derived from the guidelines of the Luxembourg Conseil Scientifique for prevention of cerebro-cardiovascular diseases and obesity. CAPSYS has been developed by researchers from the Public Research Centre Henri Tudor in Luxembourg in collaboration with neurologists from the Centre Hospitalier de Luxembourg (CHL). Currently, the user acceptance and efficacy of the system is being evaluated in a six-month randomized controlled study with eligible participants recruited at CHL's neurology department.

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Abstract – WCN 2013**No: 2800****Topic: 3 – Stroke****Epidemiology of stroke in a teaching hospital in subsaharan africa: 3 years prospective study in douala, cameroon**

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Background: Stroke is the leading cause of hospitalization in neurology but epidemiological data for this condition are scarce in Cameroon.

Objective: Describe the epidemiology of stroke in a tertiary hospital in Cameroon.

Patients and methods: We carried out a prospective study from 1st January 2010 to 31st December 2012 at the Douala General Hospital in 2 sites: the neurological unit (NU) and the intensive care unit (ICU). All patients admitted for confirmed stroke were included consecutively. Socio-demographic, clinical and paraclinical data were collected.

Results: 318 patients presented with stroke (252 in the NU and 66 in the ICU) with a mean age of 58.84 ± 13.04 years; 38.1 were female. The major risk factor before stroke were high blood pressure (69.6%), alcohol consumption (28%), diabetes mellitus (20.4%), tobacco (16%), heart disease including atrial fibrillation (11.3%), dyslipidemia (8.5%), HIV infection and sleep apnea constituting each 3.1%. The mean delay for consultation was 27.19 ± 97.52 h. 53.8% had an ischemic and 46.2% a hemorrhagic stroke. The mean duration of hospitalization was 10.56 ± 23 days with a global mortality rate of 26.4% (13% in the NU). Septic conditions were the leading cause of death.

Conclusion: Fighting vascular risk factors by informing and screening the population should be emphasized in our context in order to reduce the burden of stroke.

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Abstract – WCN 2013**No: 2808****Topic: 3 – Stroke****Metabolic burden and intracranial atherosclerotic disease in an asymptomatic population**

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Background: To date, few data are available with respect to intracranial atherosclerotic disease (ICAD) in general population. This study examined whether metabolic abnormalities, metabolic syndrome (MetS) and non-alcoholic fatty liver disease (NAFLD), are surrogate markers for intracranial atherosclerotic disease (ICAD) among apparently healthy population.

Methods: Over 3 years, study data were collected on subjects who underwent a comprehensive health check-up. The presence of ICAD was ascertained based on the findings of brain MRA, and the diagnosis of NAFLD was made by liver ultrasound work-up. Finally, 6146 subjects without a previous stroke history (mean age, 54.7 ± 8.9 years; male, 70.5%) were enrolled in this study.

Results: The prevalences of MetS, NAFLD, and ICAD were 46.4% (n = 2853), 23.8% (n = 1460), and 4.6% (n = 282), respectively. After the adjustment for atherosclerosis risk factors, NAFLD (OR: 1.89, 95% CI: 1.45–2.46) and MetS (OR: 1.50, 95% CI: 1.14–1.96) were independently associated with increased risk of ICAD. In addition, the number of individual components of metabolic syndrome was associated with the risk of ICAD in a dose-response manner.

Conclusions: The present study suggested that the detection of NAFLD and metabolic syndrome in an asymptomatic population was independently associated with the increased risk for ICAD; however, further prospective studies would be needed to confirm our findings.

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Abstract – WCN 2013

No: 2809

Topic: 3 – Stroke

Treat and transfer: Efficacy and safety of the telestroke approach in Salzburg, Austria

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Background: Five hospitals without 24 h neurology coverage in the Austrian state of Salzburg are connected to a stroke hub via videoconferencing. Prompt transfer to the comprehensive stroke center (20–129 km distance) is carried out after administration of recombinant tissue plasminogen activator (rt-PA).

Objective: To assess efficacy and safety of teleconference-assisted thrombolysis for acute ischemic stroke and instant transfer to the stroke center.

Patients and methods: Retrospective chart review of patients treated with acute ischemic stroke from 2006–2009. Inclusion criteria: rt-PA administration within 4.5 h from symptom onset, age ≥ 18 years. Exclusion criteria: arrival at the stroke center beyond 24 h from symptom onset, initial NIHSS > 25, and previous stroke. The measures for efficacy were mortality, NIHSS and mRS at 3-month follow-up.

Results: Forty-seven patients were moved to the stroke center after rt-PA treatment. The control group consisted of 304 patients who received rt-PA directly at the stroke center. Mean time till admission to the stroke unit was 231 and 108 min, respectively (P < 0.001). Patient demographics, NIHSS on admission and door-to-needle time did not differ between the groups. No transfer-related complications were reported. The rate of complications during stroke unit care did differ between the groups. There were no differences in the outcome measures.

Conclusion: This study confirms that the efficacy of telemedicine-assisted systemic thrombolysis is comparable to in-house administration at a comprehensive stroke center. Importantly, rapid patient relocation was safe and provides the added benefit of care at a stroke unit and interdisciplinary management in case of complications.

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Abstract – WCN 2013

No: 2750

Topic: 3 – Stroke

The investigation of neuroprotective activity of 5-hydroxyadamantane-2-on in rats under the conditions of local permanent brain ischemia

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The influence of 5-hydroxyadamantane-2-on, an adamantane derivative, on morphological state of brain tissue of rats with local permanent brain ischemia was studied. As a model of ischemia the occlusion of left middle cerebral artery was chosen. The comparison was done between serial histological preparations of the control group (with occlusion and without drug administration) and two experimental groups (with occlusion and administration of 5-hydroxyadamantane-2-on 100 mg/kg i.p., for 6 and 12 days respectively). Moreover, within the same histological preparations the morphological changes were compared between histological preparations from basins of left and right (not occluded) middle cerebral arteries.

It was determined that the administration of 5-hydroxyadamantane-2-on (dosage 100 mg/kg) was once per day after the occlusion of the middle cerebral artery brings to significant regenerative processes of brain tissue. It was also shown that regenerative processes are more profound in the group of rats that was treated with 5-hydroxyadamantane-2-on for 12 days.

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Abstract – WCN 2013

No: 2811

Topic: 3 – Stroke

Correlations of intima-media thickness with atherosclerosis and obesity markers in first degree relatives of stroke patients

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Background: Stroke is the first cause of mortality and major invalidity. The intima media thickness (IMT) was proved to be a good marker not only for subclinical atherosclerosis but also for predicting future cerebrovascular events.

Methods: We evaluated two groups. Group A included 47 stroke patients and group B 48 first degree relatives of the previous stroke patients. IMT was measured by cervical ultrasound mode B, at the common carotid artery distal wall, on both sides. Other parameters measured were weight, height, body mass index (BMI), smoker status, arterial systolic and diastolic blood pressure.

Results: Group A had 53.2% males and group B 65.4% women. The abdominal circumference per height ratio showed a strong

correlation with the BMI, but without establishing one with the IMT. 34% of the stroke patients were smokers, whereas 27.1% of their relatives smoked; the relative risk of stroke for relatives was over 2 times higher for smokers than for non-smokers. Although in both groups the value of the IMT was high (0.91–0.97 mm), there was no statistical significance ($p > 0.05$). IMT was correlated to almost all the other parameters measured.

Conclusion: In our study the IMT was correlated with atherosclerosis and obesity markers, but had no statistical significant value as an independent risk factor for cerebrovascular disease in first degree relatives of stroke patients.

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Abstract – WCN 2013

No: 2789

Topic: 3 – Stroke

Obstructive sleep apnea and ischemic stroke

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Background: The obstructive sleep apnea syndrome (OSAS) is a chronic respiratory disorder which is at present a real public health problem because of its cardiac and vascular side effects.

Objective: The purpose of the study is to detect OSAS in patients with ischemic stroke to optimize secondary prevention.

Patients and methods: This is a prospective study of 50 patients suffering from ischemic stroke admitted to the Neuro-vascular unit of the University Hospital Hassan II of Fez in whom nocturnal oximetry was performed in search of OSA, polysomnography is supplemented by the following if nocturnal oximetry appears abnormal. The exclusion criteria of the study were: disorders of consciousness, oxygen saturation of less than 95%, and obstructive lung disease.

Results: The average age was 60.6 years, the sex ratio (M/F): 1.38. In 50 patients recruited 13 patients (26%) had abnormal nocturnal oximetry. Polysomnography was applied systematically in 13 patients. The review has been performed in 8 patients and OSAS was confirmed in 6 patients and the syndrome of mixed sleep apnea (obstructive and central) in 2 patients. In 13 patients, snoring was found in 46% of cases, hypertension in 46% and diabetes in 30.7%.

Conclusions: Screening for obstructive sleep apnea syndrome appears an important issue in the treatment of ischemic stroke to reduce the morbidity associated with ischemic stroke.

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Abstract – WCN 2013

No: 2754

Topic: 3 – Stroke

Acute statins treatment for aneurysmal subarachnoid hemorrhage might be limited: A systematic review and meta-analysis

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Background: The role of statins in the management of patients with aSAH remains unclear. Designers considered whether statins delayed the time to CVS and DIND from ictus, and shortened duration of CVS and DIND might solve this problem indirectly. Therefore, the efficiency of statins was evaluated via comparing different outcomes to discuss the role of statins for aSAH.

Objective: The role of statins for aneurysmal subarachnoid hemorrhage remains confusing. The relevance of different end points was evaluated to discuss the role of statins.

Methods: Systematic literature retrieval was carried out to obtain studies of randomized controlled trials researching on statins for aneurysmal subarachnoid hemorrhage before March 2013. Data extraction and quality evaluation of studies were performed by 2 investigators. A meta-analysis was performed by RevMan 5.2.3.

Results: 7 RCTs with 347 patients which met the inclusion criteria were included in the meta-analysis. The results showed statins did not delay the time to vasospasm or delayed ischemic neurological deficit from ictus, or shorten duration of vasospasm in clinical practice, but reduced 22% vasospasm on transcranial Doppler (RR = 0.60; 95% CI 0.42 to 0.86, $P = 0.005$), 26% delayed ischemic neurological deficit (RR = 0.48; 95% CI 0.32 to 0.70, $P = 0.0002$), and 10% mortality in hospital (RR 0.54; 95% CI 0.32 to 0.91) compared to the placebo group.

Conclusion: It might be a choice that acute statins treatment is for aneurysmal subarachnoid hemorrhage, but the role was limited. Further large scale, well-designed RCTs on this topic are still needed.

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Abstract – WCN 2013

No: 2749

Topic: 3 – Stroke

Eicosapentaenoic acid prevents cerebral vasospasm after aneurysmal subarachnoid hemorrhage: A meta-analysis

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Background: In recent years, studies have shown that eicosapentaenoic acid has multiple effects: including reduced platelet aggregation, plaque stabilization, vasodilation and anti-inflammation, could reduce the recurrence of stroke. Studies in vitro have shown that eicosapentaenoic acid inhibited the SPC-Rho-kinase pathway, without affecting the calcium dependent vascular smooth muscle contraction. In vivo model of cerebral vasospasm also found that eicosapentaenoic acid could be used for adjuvant therapy preventing cerebral vasospasm after aneurysmal subarachnoid hemorrhage.

Objective: To systematically evaluate the curative effectiveness and safety of eicosapentaenoic acid preventing cerebral vasospasm after aneurysmal subarachnoid hemorrhage.

Studies and methods: Systematic literature retrieval was carried out to obtain studies of randomized controlled trials preventing cerebral vasospasm after aneurysmal subarachnoid hemorrhage with eicosapentaenoic acid before January 2013. Study selection, data collection and methodological quality assessment were performed by two individual reviewers. A meta-analysis was performed by RevMan 5.2.3 software.

Results: 3 studies and 307 patients were included totally. Meta-analysis showed that eicosapentaenoic acid decreased symptomatic cerebral vasospasm (RR = 0.46, 95% CI (0.29, 0.72), $P = 0.0006$), cerebral infarction was caused by vasospasm (RR = 0.27, 95% CI (0.14, 0.51), $P < 0.0001$), but did not improve the modified Rankin Scale (RR = 1.15, 95% CI (0.98, 1.33), $P = 0.08$). Few gastrointestinal adverse effect has been reported.

Conclusion: Eicosapentaenoic acid is safe and reliable to be used for the prevention of aneurysmal cerebral vasospasm after subarachnoid hemorrhage. However, further studies are needed to confine the long-term clinical outcomes.

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Abstract – WCN 2013**No: 2819****Topic: 3 – Stroke****Level of von willebrand factor in patients with carotid artery stenosis**

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According to the results of numerous studies, von Willebrand factor (vWF) is one of the markers of endothelial dysfunction in stenosis of the carotid arteries.

An objective of the study is to explore vWf patients with stenosis of the carotid artery.

Target groups and techniques: The group included 83 people with carotid stenosis. The average age of 59.07 (± 6.62). In two groups of patients: I – stenosis < 50% (n = 70, 34 men, 36 women), II – stenosis > 50% (N = 13, 6 males, 7 females). Not stable plaques identified in 75.7% (n = 53) in the first and 100% (n = 13) of the second groups.

Results: vWfV first group with unstable plaque was significantly higher than those with stable plaque (P = 0.004). Significant differences were found in men (p = 0.024) and women (p < 0.05). When comparing groups I and II with the unstable plaque, significant differences were detected (p = 0.065).

Conclusions: The comparative study has indicated relationship between the vWF level and the structure of plaques in patients with occlusive lesions of the carotid arteries as well as absence of the relationship between the level of vWF and extent of the carotid arteries stenosis.

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Abstract – WCN 2013**No: 2835****Topic: 3 – Stroke****Nimodipine in treatment of cognitive impairment in patients with subarachnoid hemorrhage**

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Background: Subarachnoid hemorrhage is a neurologic emergency characterized by high case fatality rates and lifelong care of survivors. Neurologic complications include rebleeding, symptomatic vasospasm, ischemic stroke, hydrocephalus and cognitive dysfunction. The acute treatment of subarachnoid hemorrhage is well documented but less is known about the long-term effects of subarachnoid hemorrhage on cognitive and functional outcomes.

Objective: Taking into consideration that nimodipine is a lipophilic calcium channel blocker, which easily crosses the blood–brain barrier, having a marked effect on the cerebral circulation, explore the nimodipine's usefulness, as an adjunct in the management of patients with subarachnoid hemorrhage by improving their neurologic outcome.

Methods: We enrolled 130 patients with subarachnoid hemorrhage. The patients closely followed clinically with repeated neuropsychological assessment. Nimodipine therapy was begun as soon as possible or within 4 days of the diagnosis.

Results: 34 patients died, 12 had cerebral infarction with improvement in neurological outcome, 66 showed stable improvement in cognitive performance measures, 30 patients had no significant improvement after treatment, although half of them improve in memory, executive function, and language with time (1–2 years

later). Deficits in cognition and day-to-day functioning were further compounded by depression, anxiety, fatigue, and sleep disturbances. Adverse events were decreases in blood pressure, headache and extrasystoles. Discontinuation of therapy required in 2 patients.

Conclusion: We found nimodipine decreased the incidence of cerebral infarction and cognitive dysfunction, improved the outcome in patients with subarachnoid hemorrhage. Benefits are attributable to reducing of cerebral arterial spasm, neuroprotective effects and less of side effects.

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Abstract – WCN 2013**No: 2837****Topic: 3 – Stroke****Comparison of risk factors between ischaemic stroke and haemorrhagic stroke**

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Background: Post hoc analysis of SPARCL trial showed that low LDL was associated with haemorrhagic stroke. Different meta-analyses also revealed conflicting results as to whether cigarette smoking favours ischaemic or haemorrhagic stroke.

Objective: To study the demography and risk factors of our stroke patients; to compare any difference in risk factors between ischaemic and haemorrhagic stroke; and to study whether low LDL favours haemorrhagic stroke.

Patients and methods: All patients with a diagnosis of stroke admitted to the Neurology Ward, Penang General Hospital in the past year were included. Data was collected from the stroke registry, and telephone interviews were conducted to fill in missing data.

Results: Of the 335 stroke patients being admitted, 83.3% had ischaemic stroke (mean age 64.5 years, male 60%), and 16.7% had haemorrhagic stroke (mean age 64.7 years, male 57%). The top three risk factors were: hypertension (32%), high LDL (21%), and diabetes mellitus (17%). The presence of diabetes mellitus was shown to favour ischaemic stroke (p < 0.05). Hypertension, high LDL, cigarette smoking, atrial fibrillation, prior stroke/TIA, alcohol intake, ischaemic heart disease, and valvular heart disease were common risk factors for both ischaemic and haemorrhagic stroke.

Conclusion: In our study, diabetes mellitus was shown to favour ischaemic stroke. On the contrary, LDL level and cigarette smoking were not shown to favour either type of stroke. Further large-sample study may be needed to clarify this relationship.

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Abstract – WCN 2013**No: 2815****Topic: 3 – Stroke****Glycated peptides induce endothelial dysfunction in the brain vasculature in normoalbuminuric patients with type 2 diabetes mellitus**

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Introduction: The aim of study was to clarify if advanced glycation end-products (AGEs) may explain the endothelial variability in microvascular territories, (kidney, brain) and exert toxic effects upon the proximal tubule (PT) in patients with type 2 diabetes mellitus (DM).

Methods: A total of 70 patients with type 2 DM and 11 gender- and age-matched healthy subjects were assessed concerning urine albumin: creatinine ratio (UACR), urinary alpha₁-microglobulin, urinary neutrophil gelatinase-associated lipocalin (NGAL), plasma and urinary AGEs, plasma asymmetric dimethyl-arginine (ADMA), and serum cystatin C. Fully automated chip-Nano electrospray ionization in conjunction with high-capacity ion trap multistage mass spectrometry was utilised to characterize the urinary proteomic profile; extra- and transcranial Doppler and the breath-holding test for the assessment of cerebrovascular reactivity were performed.

Results: The cerebral haemodynamic indices were correlated with plasma ADMA, plasma AGEs, inflammation, DM duration, GFR, and cystatin C. Urinary glycosylated peptide fragments were identified, in normo-(m/z ratio 700–900 Da) and microalbuminuric patients (m/z ratio 1000–1200 Da). Urinary alpha₁-microglobulin and urinary NGAL were correlated with urinary AGEs ($R^2 = 0.586$; $P < 0.001$; $R^2 = 0.415$; $P < 0.001$), UACR ($R^2 = 0.292$; $P < 0.001$; $R^2 = 0.116$; $P < 0.002$), GFR ($R^2 = 0.172$; $P < 0.001$; $R^2 = 0.135$; $P < 0.001$), cystatin C ($R^2 = 0.146$; $P < 0.001$; $R^2 = 0.129$; $P < 0.001$), but not with ADMA.

Conclusion: In normoalbuminuric type 2 DM patients, AGE-induced endothelial dysfunction plays a putative role in the brain vasculature. AGEs are toxic for the PT, but spare the glomerular endothelium. The proteomic pattern of urinary glycosylated peptides and PT dysfunction are the key factors which could make the difference between normo- and microalbuminuric patients.

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Abstract – WCN 2013

No: 2639

Topic: 3 – Stroke

Predictors of mortality in stroke subtypes and subdural haematoma 2000–2007: A cox regression analysis

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Background: Stroke is the second most common cause of death worldwide and is the leading cause of acquired disability in adults. However few studies have investigated predictors of outcome in a large dataset.

Objective: We sought to identify comorbidities that predict death in hospital inpatients.

Patients and methods: We compiled an anonymised database of all patients admitted with ischaemic stroke, haemorrhagic stroke, subarachnoid haemorrhage and subdural haematoma to a large

multiethnic teaching hospital in Birmingham, UK during the period 2000–2007. The ICD 10th revision coding was used to identify these cases. Stepwise Cox regression was performed incorporating age, gender and ethnicity, and all comorbidities with a frequency over 1% in each of the four intracranial pathologies.

Results: Shorter duration of hospital stay predicted death in all stroke subtypes ($p < 0.001$ respectively) and in subdural haematoma patients ($p < 0.05$). *Clostridium difficile* infection predicted death in ischaemic (HR: 2.27) and haemorrhagic (HR: 8.47) stroke ($p < 0.01$ respectively). Urinary tract infection was associated with death in subdural haematoma (HR: 5.07, $p < 0.001$). Pneumonia predicted death in ischaemic stroke (HR: 2.28, $p < 0.001$) and subarachnoid haemorrhage (HR: 3.98, $p < 0.001$).

Conclusion: Our findings indicate that patients are dying due to swift hospital discharge following their stroke. This may be related to shortages in hospital beds, and inadequate community care. Further studies are required urgently to investigate this further, incorporating cause of death data. We also demonstrate that hospital-acquired infections are a leading cause of death in stroke patients.

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Abstract – WCN 2013

No: 2740

Topic: 3 – Stroke

Knowledge and attitudes towards stroke among workers in three university hospitals in Egypt

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Background: The rising global burden of stroke is more pronounced in the less developed world, including Egypt.

Objective: To evaluate the level of knowledge and attitudes towards stroke among workers affiliated to hospitals in Egypt.

Material and methods: This is a hospital-based cross-sectional observational study of workers affiliated to three university hospitals in Egypt using convenience sampling over 3 months. They were classified into clinical (Health care staff and pre-clinical medical students) and non-clinical workers (e.g. Administrative & sanitary staff). A structured self-administered questionnaire of knowledge and attitudes towards stroke was developed.

Results: Of 468 questionnaires administered, 396 were completed. Most respondents have heard of stroke before (98.4%). The most commonly identified risk factors of stroke were hypertension, stress, smoking and high cholesterol levels. Age and lack of exercise were among the least recognized. There were no differences between clinical and non-clinical workers in knowledge about the risk factors of stroke expect for unhealthy diet ($p = 0.007$) and obesity ($p < 0.001$). Clinical workers were more likely to identify the symptoms of stroke. Non-clinical workers were more likely to go to hospital ($p < 0.001$).

Conclusion: Knowledge of stroke among workers in the three studied university hospitals is poor. There is a discrepancy in level of knowledge and attitude towards stroke between clinical and non-clinical workers. This addresses the need for public stroke education programs to improve public awareness of stroke in Egypt.

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Abstract – WCN 2013**No: 2825****Topic: 3 – Stroke****Serum levels of YKL-40 in acute arterial ischemic stroke patients, and its relationship to infarct volume**

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Background: The aim of this study was to investigate the levels of YKL-40 in serum from patients with acute arterial ischemic stroke (aIS), and its relationship to infarct volume.

Patients and methods: A total of 90 subjects, including 50 patients with aIS, (28 males, 22 females, at 42–72 years of age) and 40 controls (21 male, 19 female) were enrolled in our study. Trials of Org 10172 in Acute Stroke Treatment (TOAST) was used for etiologic classification. Serum YKL-40 was measured by ELISA. Acute cerebral infarction was defined as an area of high signal intensity on the diffusion weighted images. Infarction volume was calculated by multiplying the manually contoured hyperintense region by the slice thickness plus the intersection gap with the aid of Scion image software.

Results: Serum YKL-40 levels of aIS patients were significantly higher than in healthy controls. Serum YKL-40 levels were 227.28 ± 78.23 ng/ml in aIS patients and 74.78 ± 13.94 ng/ml in controls ($p < 0.001$). There were significantly positive correlations between serum YKL-40 levels and infarct volume in patients with aIS ($r = 0.931$, $p < 0.001$). Serum YKL-40 levels were especially elevated in patients with large artery (carotid) atherosclerosis according to TOAST.

Conclusions: Our study showed that YKL-40 is higher in aIS patients. In our opinion, the relationship between the serum YKL-40 level and cerebral infarction volume may represent a new opportunity for the possible utility of serum YKL-40 as an inflammatory marker for ischemic stroke.

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Abstract – WCN 2013**No: 2738****Topic: 3 – Stroke****Acute cerebrovascular accidents and sudden weather changes: Any association?**

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Introduction: It is suggested that changes of climate, weather or seasons are the risk factors of the acute cerebrovascular accidents (CVA). We hypothesized, that independently of the seasonal atmospheric changes, abrupt weather changes may influence the incidence of CVA. In this study we analyzed the association between sudden changes of atmospheric pressure and air temperature with the incidence of CVA.

Material and methods: In 607 consecutive patients (mean age 69.7 years, 56% women) admitted to our Stroke Unit in the year 2010 we analyzed the association of the time of onset of CVA (ischemic stroke 70.0%, hemorrhagic stroke 7.3%, transient ischemic attack 21.9%, subarachnoid hemorrhage 0.8%) with abrupt changes of atmospheric pressure and air temperature measured in half-hour intervals before admission (17,479 recordings).

Results: With the use of logistic regression we observed significant increase of the incidence of CVA after decrease of air temperature of at least 8 °C during 19 h ($p < 0.001$; OR = 2.72; 95%CI 2.13–3.47), or after decrease of atmospheric pressure of at least 8 hPa during 7 h ($p = 0.012$; OR = 2.95; 95%CI 1.42–6.14). However, respective regression models allowed for interpretation of only 1.0% and 0.1% in

the variability of the incidence of CVA. The incidence of CVA was not associated with the seasons (chi-square $p = 0.439$).

Conclusions: After sudden drops of atmospheric pressure or air temperature the incidence of acute cerebrovascular accidents rises independently of seasonal weather changes. Although the statistical models are significant, this association appears to be very subtle or incidental and therefore probably clinically not important.

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Abstract – WCN 2013**No: 2851****Topic: 3 – Stroke****CD34⁺/CXCR4⁺/CD45⁻ stem cells and cerebrovascular risk factors in acute stroke patients**

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Background: Endogenous non-hematopoietic stem cells seem to play a protective role in acute ischemic stroke.

Objective: We hypothesized that:

- levels and dynamic changes of CD34⁺CD45⁻, CD34⁺CXCR4⁺CD45⁻ and CXCR4⁺CD45⁻ stem cells correlate positively with neurological and functional status of acute stroke patients;
- certain comorbidities may correlate with lower levels of the abovementioned cells and negatively influence their dynamics.

Patients and methods: Venous peripheral blood was sampled and assessed by flow cytometry on days 1, 2, 4, and 6 after acute stroke onset in 34 patients. Parallel to blood sampling and after 3 and 6 months patients were assessed with the National Institute of Health Stroke Scale, Barthel Index, Scandinavian Stroke Scale, and modified Rankin Scale.

Results: Higher initial levels of all 3 subgroups of cells correlated with better neurological or functional status on admission. Higher increase in CD34⁺CD45⁻ and CD34⁺CXCR4⁺CD45⁻ stem cells and lower increase in CXCR4⁺CD45⁻ stem cells during the 6 days following stroke correlated positively with initially more severe stroke.

Diabetes in anamnesis correlated with initially lower levels of CD34⁺CXCR4⁺CD45⁻ and CXCR4⁺CD45⁻ cells. Hypertension correlated with initially lower levels of CD34⁺CD45⁻ cells. Diabetes and hypercholesterolemia correlated with higher increase or smaller decrease of all 3 subgroups of cells.

Conclusions: The lower initial levels and the subsequent increase in the levels of the chosen subgroups of stem cells seem to indicate more severe neurological/functional state of the stroke patients. Diabetes, hypertension and hypercholesterolemia seem to negatively influence the initial levels of the cells and/or augment their dynamics.

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Abstract – WCN 2013**No: 2854****Topic: 3 – Stroke****Pharmacogenomic assessment of clopidogrel – A step toward personalized medicine for patients with ischemic stroke**

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Background: Stroke is a heterogeneous disease. The most common causes of cerebral infarction are: large artery atherosclerosis, small vessel disease and cardioembolism. Furthermore, there are several genetic variations in drug metabolism in regard to the most frequently used antiplatelet agents and anticoagulants that potentially underlie variability in drug efficacy and adverse drug reactions. **Objective:** This study discusses investigation, outcomes and the clinical effectiveness of clopidogrel administration for secondary stroke prevention in Bulgarian patients with cerebral infarction based on an assessment of “clopidogrel resistance”.

Patients and methods: Participants included 214 patients (104 male and 110 females, range 41–89 years) with ischemic stroke. They were assigned to receive 75 mg of clopidogrel daily despite regular medical therapy. Assessment of individual pharmacological response to antiplatelet therapy was performed on 24 cases by platelet function tests, including multiple electrode aggregometry (MEA).

Results: Among the 28 subjects evaluated by laboratory tests, only 3 showed a resistance to clopidogrel. During the clinical follow-up for a period of 5–8 months, the prevalence of consecutive cerebrovascular events was greater in 6 patients who underwent tests, and none of them were resistant to clopidogrel. Our study presents confirmatory results with respect to efficiency and safety of clopidogrel administration for secondary stroke prophylaxis. In fact, only 4% of Bulgarians are not-responding to clopidogrel treatment.

Conclusion: Antiplatelet drug clopidogrel is the main point for secondary stroke prevention with or without assessment of platelet function testing. We recommend an individualized approach for each patient with cerebrovascular disease.

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Abstract – WCN 2013

No: 2832

Topic: 3 – Stroke

Age, sex and stroke type differences in stroke patients at Mukalla, Hadhramout, Republic of Yemen: Analysis of 1072 cases

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Background: Stroke is the third leading cause of death, and a cause of long-term disability among survivors.

Objectives: To identify age, sex and stroke type differences in stroke patients.

Patients and methods: A retrospective study of stroke patients admitted at Mukalla, Hadhramout in 1/2009–12/2011. Data were collected in a questionnaire from patients' files.

Results: There were 1072 stroke cases; ischemic stroke was 78.5% and hemorrhagic 21.5%. Males 56.5% and females 43.5%. Old patients' > 60 years were 72% of cases. Hypertension was more in middle-aged. Diabetes was in 36.3% and 39.5% of middle-aged and old, and 7.4% of young. Old patients had ≥ 2 risk factors (89.9%). Ischemic stroke increased with age, while hemorrhagic type was more in young. Coma was more in young. Non-significant difference between males and females, in hypertension, diabetes previous attacks and family history. 94.2% of males had ≥ 2 risk factors and females (70%). Ischemic stroke occurred in 76.2% of males and 81.3% of females. Hypertension in hemorrhagic stroke was significantly higher than in ischemic type, non-significant difference between diabetes in ischemic and hemorrhagic stroke. Previous attacks were significantly higher in hemorrhagic than ischemic type. Sudden onset of presentation and coma on hospitalization were significantly high in hemorrhagic type.

Conclusions: Stroke increased with age. Ischemic stroke was more in old while hemorrhagic type in young patients. Males were more affected. No sex differences noted in relation to risk factors. Hemorrhagic stroke was more related to hypertension, previous attacks, sudden onset and coma.

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Abstract – WCN 2013

No: 2887

Topic: 3 – Stroke

Recombinant tissue plasminogen activator and stroke – Case report

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A case report: Patient 46 years old, male, smoker, with history of stroke in the immediate family without comorbidities (normal blood pressure with normal values of lipids and glucose). Patient is admitted for examination in emergency service after 2 h of acute weakness in right limbs and speech difficulties. Immediately an internal medicine and neurological examination was performed with CT examination showing normal findings. Afterwards fibrinolytic therapy was administered as per protocol for the treatment of ischemic stroke, with good tolerance of the drug and a good therapeutic response. Nine hours after the administration of the drug almost complete withdrawal of right motor deficit was present, but with persisting speech difficulties. Treatment was continued with low molecular weight heparin. On the second CT an infarction was seen in the vascular area of the left ACM, and on the third CT was showing almost complete regression of the ischemic area. The patient was released from hospital with latent weakness of limbs and right sensorimotor dysphasia.

Conclusion: Our experience of an acute stroke thrombolysis with recombinant tissue plasminogen activator shows that adequate thrombolysis in accordance with established treatment guidelines saves lives.

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Abstract – WCN 2013

No: 2893

Topic: 3 – Stroke

The locked-in plus syndrome

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Background: There has been discussion about the nomenclature and examination protocols of patients with disorders of consciousness (DOC) and related syndromes, as the locked-in syndrome (LIS). As publications increase finding brain responses to external stimuli, using fMRI or EEG, in these patients, there is need for a diagnosis scheme which best suits these patients. Locked-in-plus syndrome (LIPS) is proposed as a category for patients who show typical signs of LIS combined with DOC.

Objective: To collect clinical and instrumental data of these patients to develop a new classification for patients with LIS and LIPS.

Methods and material: Seven patients with ischemic pontine and other brain lesions were examined clinically, with standardized behavioral assessment scales and with MRI and functional MRI.

Results: All patients presented different degrees of arousal, consciousness, and other neurological and behavioral symptoms. The extent of structural brain damage and brain response in fMRI was found to be variable in spread.

Conclusion: The relevant differences between LIS and LIPS lie on the variety of additional pontine lesions in LIPS and in symptoms caused by these lesions: Frequently occurring features in LIPS include hypersomnia, frontal release signs, thalamic posturing of hand and/or feet. Rarely an akinetic mutism may occur.

Extra pontine brain lesions may occur in mesencephalic, thalamic and cerebellar brain structures, as well as occipital, temporal brain regions, depending on varieties of the vertebro-basilar artery blood supplying system. Due to the heterogeneity of the data collected, a new diagnostic category should be implemented in clinical practice.

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Abstract – WCN 2013

No: 2880

Topic: 3 – Stroke

Early stroke predictors in hypertensive obesity men aged 60–69: Pulse feeling and therapy

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Background: Hypertension is the major risk factor for stroke.

Aim: Research of early predictive value of pulse feeling in hypertension, complicated stroke.

Methods: Pulse was palpated in 12 positions in the points Cun, Guan, and Chi. ECG and computed tomography were investigated. Valuation of the pulse was made taking into account the gender, age, season, and given region. Data of rhythm, heart rate frequency, filling, tension, depth, length, hardness, form, width, and smoothness of pulse were determined. Pulse was investigated to the healthy with systolic and diastolic blood pressure less than 140/90 mm Hg respectively, in comparable 27 patients with II (basal Gr1) and in 36 patients also with II (control Gr2) degree of hypertension (WHO classification) survivors of stroke. Patients had BMI 30.2–34.6 kg/m².

Results: The healthy had balanced pulse. The patients a long time before stroke had the pulse disbalance, which was correlated with the level of blood pressure. Frequent and sudden weather changes (magnetic storm, the changeability of barometric pressure, relative humidity, air temperature), and stresses enhanced the disbalance of pulse among non-treated by acupuncture Gr2, that correlated with the increase of cases' frequency and the hardness of stroke. The patients treated by acupuncture and other therapy methods, in Gr1 had the best prognosis.

Conclusion: Pulse feeling early predicts stroke. Correct acupuncture using stress free needles based on the everyday results of PF normalizes the pathological pulse, in complex with the antihypertensive, neuro-protective treatment reduces the risk of stroke, improves prognosis and the life quality.

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Abstract – WCN 2013

No: 2788

Topic: 3 – Stroke

The prevalence of heart problems and risk factors in patient with ischemic stroke and their relationship with severity of stroke

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Background and objective: Stroke is the most common cause of morbidity and mortality. Ischemic stroke is a brain dysfunction due to vascular occlusion. Many studies are discussed about some risk factors for cerebral infarction such as: systolic and diastolic hypertension, diabetes, heart disease, cardiogenic emboli, hyperlipidemia, smoking, and taking OCPs. In this study, we describe the prevalence of heart problem and other important risk factors in ischemic stroke and their relationship with severity of stroke.

Patients and methods: The study population included patients with ischemic stroke admitted in neurology ward in Sanandaj Tohid Hospital between September 2011 and March 2012. Data and information were obtained from Hx, Ph. Ex, E.C.G, Echocardiography, and Biochemistry and it was analyzed using SPSS software and descriptive statistics.

Results: Of the 130 patients, 53.1% were male. The mean age of patients was 68 ± 12.9. The most important causes of stroke include: hypertension 83.1%, heart disease 60%, hyperlipidemia 39.7, diabetes 36.6% and 15% OCPs. 14.6% of patients showed atrial fibrillation based on admission E.C.G. Mitral regurgitation on echocardiography in 55 patients (42%) had the highest prevalence. In patients mean Ejection Fraction (EF) was 50.1 ± 10.7, and the average strength was 2.43 ± 1.7 in organs involved. Right and left extremities of muscle strength were shown to decrease with increasing age ($p = 0/04$ and $p = 0/07$), but ischemic lesion sizes were increased ($p = 0/008$).

Conclusion: The severity of stroke was increased by age. There were no significant difference between the severity of stroke in men and women, in smokers and non-smokers, those with diabetes, hyperlipidemia, hypertension and those who without these risk factors.

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Abstract – WCN 2013

No: 2864

Topic: 3 – Stroke

Sensitivity of the color trails test (CTT) in stroke and lacunar infarction

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Background: The color trails test (CTT) is a culture free neuropsychological test to measure sequencing, visual scanning, and speed of mental processing abilities. No clinical data are available about the utility of this test in the Greek population.

Objective: To provide clinical data on the sensitivity of the CTT in 2 groups of patients (6 month post stroke and lacunar infarction) and 2 groups of pair matched (in age) controls.

Patients and methods: Twenty-four patients exhibiting ischaemic stroke (IS) and 14 patients exhibiting lacunar infarction (LI) were compared to 38 controls. All patients were seen in the Neurology department of the Athens Navy Hospital and the Eginition Hospital of Athens between December 2011 and December 2012.

Results: The mean age for the IS group was 64.75 years (SD = 12.79) and the mean age for the LI group was 66.74 years (SD = 10.06). The Mann–Whitney statistical tests were used to find differences between the groups. Significant differences were found between the IS group and the controls (CTT1 $U = 132.500$, $p = 0.001$, CTT2 $U = 126.000$, $p = 0.001$), and the LI group and the controls (CTT1 $U = 66.500$, $p = 0.150$; CTT2 $U = 48.500$, $p = 0.021$).

Conclusion: The CTT can be a useful tool to measure the speed of mental processing in stroke and lacunar infarction. Further research is needed to prove its clinical utility in larger samples.

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Abstract — WCN 2013**No: 2861****Topic: 3 — Stroke****The clinical significance of the intraoperative ultrasound research during carotid endarterectomy**R. Medvedev^a, G.K. Kuntcevic^b, E. Gemdjan^c, S. Scrylev^d, M. Krotenkova^e, V. Shchipakin^d, A. Koshcheev^d, M. Tanashyan^f.^aDepartment of Reanimation and Intensive Therapy, Research Center of Neurology of the Russian Academy of Medical Sciences, Russia;^bUltrasound Investigations Laboratory, Russia; ^cBiostatistics, National Research Center for Hematology, Russia; ^dVascular Surgery Department, Research Center of Neurology of the Russian Academy of Medical Sciences, Moscow, Russia; ^eRadial Diagnostics Department, Research Center of Neurology of the Russian Academy of Medical Sciences, Moscow, Russia; ^fNeurological Department, Research Center of Neurology of the Russian Academy of Medical Sciences, Moscow, Russia**Objective:** To appraise the possibility of intraoperative blood flow monitoring of artery ophthalmic (AO) and middle cerebral arteries (MCA) during the carotid endarterectomy.**Material and methods:** The research included 67 patients with stenosis of ICA at the age of 43 to 73 (the median of age is 57). The volume of research included clinical data, intraoperative blood flow monitoring in AO and MCA, and results of DWI of brain made 24 h after the operation.**Results:** Monitoring blood flow in the AO was carried out with 40 (60%) patients and with 27 in MCA (40%). With low sensitivity and hence with low prognostic value in registering material emboli during the AO blood flow monitoring, the further analysis was based on the data received during the blood flow monitoring in MCA. The combination of same signs such as material emboli with 12% of patients and gaseous signals during 5 and 16 gaseous showers with 8% of patients made it possible to determine the sensitivity of the method in revealing of acute ischemic lesions in 65% of cases. The prognostic significance of positive results made up 95%.**Conclusions:** The intraoperative ultrasound control of the blood flow made it possible to prognosticate the development of acute ischemic lesions in brain.

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Abstract — WCN 2013**No: 2874****Topic: 3 — Stroke****Hypertension rates and mortality in spontaneous intracerebral hemorrhage and ischemic cerebrovascular disease**H.L. Gul^a, O. Karadaş^b, S.K. Gul^c, S. Firtina^d. ^aDepartment of Neurology, Kartal Education and Research Hospital, Istanbul, Turkey;^bDepartment of Neurology, Erzincan Military Hospital, Erzincan, Turkey;^cDepartment of Radiation Oncology, Kartal Education and Research Hospital, Istanbul, Turkey; ^dDepartment of Cardiology, Erzincan Military Hospital, Erzincan, Turkey**Introduction:** In the prevention of spontaneous intracerebral hemorrhage (ICH) and ischemic cerebrovascular disease (ICVD), treatment against risk factors takes an important role. Hypertension (HT) is one of the most important independent modifiable risk factor in ICH and ICVD development. In this study, it was aimed to underline the importance of hypertension as a risk factor of cerebrovascular disease.**Methods:** 133 patients (69 male, 64 female with a mean age of 65.9) diagnosed as ICH and 137 patients (56 male, 81 female with a mean age of 69.7) diagnosed as ICVD were included to our study. Hypertension rates of both groups were noted and after one year of follow up, mortality rates were recorded.**Results:** 115 patients had a history of hypertension in ICH group (86.4%) and 102 patients had a history of hypertension (74.4%) in ICVD group. There is a relation between hypertension and development of ICH and ICVD. One year of mortality is 49.6% (66 patients) for ICH group and 42.3% (58 patients) for ICVD group.**Conclusion:** As the number of risk factors like hypertension increases, the risk of development of ICH and ICVD increases. Treatment of HT decreases the incidence of both ICH and ICVD. It was shown that maintaining blood pressure within normal limits reduces mortality and morbidity significantly.

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Abstract — WCN 2013**No: 2865****Topic: 3 — Stroke****Are stable patients on warfarin treatment actually stable?**D. Necioglu Orken, L. Gundogdu-Celebi, E. Turgut, S. Mumcu. *Neurology, Sisli Etfal Education and Research Hospital, Istanbul, Turkey*

Anticoagulant agents, especially warfarin, are widely used for the prevention of cardioembolic infarction related to atrial fibrillation (AF) but it is limited by inherent problems. New anticoagulant agents are expected to revolutionize the field of stroke prevention in AF. But the question of which patients should use these new agents is not yet answered.

The aim of this study is to determine the ratio of anticoagulated patients with warfarin who are inside the target therapeutic range at any given time.

Material and methods: Among 204 patients who had ischemic cerebrovascular disease, treated with warfarin and being followed-up in the Cerebrovascular Diseases outpatient clinic, 131 who had AF were included to this study. The INR in their last visit is evaluated whether it was in therapeutic range or not.**Results:** The mean age (\pm SD) of patients was 73.9 ± 9.6 years, and 65 of patients (49.6%) were females. INR of 80 patients (61%) was inside, while INR of 51 patients (39%) was outside of therapeutic range. INR of 25 patients (49%) was below and 26 (51%) patients were above the therapeutic range.**Discussion:** Despite regular monitoring of INR, 39% of our patients on warfarin treatment are outside of therapeutic range at any given time. In spite of careful dose adjustment, the INR is frequently outside the target therapeutic range. This uncontrollable situation increases the risk of thromboembolism and bleeding. New anticoagulant agents might be used in preference to warfarin in most cases.

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Abstract — WCN 2013**No: 2873****Topic: 3 — Stroke****Assessment of functional outcome and quality of life after aneurysmal subarachnoid hemorrhage**I. Gabrielyan^a, A. Zakaryan^b, H. Manvelyan^a. ^aDepartment of Neurology, Yerevan State Medical University, Yerevan, Armenia; ^bDepartment of Neurosurgery, Yerevan State Medical University, Yerevan, Armenia**Background:** Subarachnoid hemorrhage from a ruptured intracranial aneurysm (aSAH) accounts for approximately 5% of all strokes, occurs at relatively young age and has poor prognosis of survival, despite improvements in medical care. Survivals often have disability with reduced quality of life (QoL).**Objective:** The aim of the study is to assess the level of disability and QoL to unveil any association between functional outcome and QoL.

Methods: The study is a cross-sectional survey of 50 patients with surgically treated aSAH. Functional outcome was evaluated by the modified Rankin Scale (mRS), Barthel index (BI) and QoL was measured with SF-36 after discharging from hospital, during one month period after hemorrhage.

Results: Mean age of patients was 46.7 year, 31 male and 19 female. By the mRS 38 (76%) patients were independent (mRS < 4), 3 (6%) were assisted independent with BI > 60 for 40 patients (80%). The mean BI was 84.56 (\pm SD 26.2). Scores of almost all domains of SF-36 were higher than one-half of total score of 100 with close correlation between the subscales. The mean scores for PH and MH were 46.2 \pm 12.0 and 46.3 \pm 10.5 respectively. Statistically significant correlation was established between physical and mental components of SF-36, BI and mRS scores showing the reduced QoL associated with the more severe disability ($p \leq 0.05$).

Conclusion: There are certain correlations between the level of disability and the QoL: patients with neurological deficit are more prone to suffer from low QoL, so the treatment of aSAH must aim to lesser neurological deficit.

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Abstract – WCN 2013

No: 2756

Topic: 3 – Stroke

Cerebral amyloid angiopathy-related leukoencephalopathy: A case of successful steroid treatment for neurological deficit and white matter lesions

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Background: Cerebral amyloid angiopathy (CAA)-related leukoencephalopathy (LE) is a relatively rare cerebrovascular disease in which amyloid β -protein (A β) is deposited on the vessel walls in the cerebrum. However, pathophysiology of A β in the CAA-related LE has not been understood completely.

Objective: We report a case of CAA-related LE whose neurological symptoms were improved by steroid therapy. In this case, the brain biopsy samples showed glial activations around the A β -deposit vessels with absence of acute inflammatory component.

Patients and methods: A 76-year-old woman developed subacute cognitive decline, consciousness impairment and right hemiparesis. Magnetic resonance images (MRI) showed prominent edema in the left-hemispheric dominant white matters and multiple cerebral microbleeds.

Results: Pathological studies of biopsied brain specimen disclosed hyaline-like materials on the vessel walls that were positive for Congo red and were dichroic under the polarized light, indicating amyloid depositions. Immunohistochemical examination revealed A β 40 deposits on the vessel walls. Numerous reactive astrocytes and upregulation of microglia were observed in the brain parenchyma around the affected vessels. There was no apparent inflammatory cell infiltration or granuloma formation. Repeated high-dose methylprednisolone pulse therapy improved consciousness, cognitive function, and resolved MRI lesions.

Conclusion: The CAA-related LE is considered to be produced by microbleeds and microinfarcts induced by A β deposition on the vessel walls. The steroid therapy efficacy in the present case suggests that the activated glial response plays additional role in pathophysiology of the CAA-related LE.

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Abstract – WCN 2013

No: 2176

Topic: 3 – Stroke

Recurrent ischemic stroke revealing Takayasu's arteritis

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Background: Takayasu's arteritis is of an unknown cause. This pathology is under described in Africa.

Objective: To describe Takayasu's arteritis associated to recurrent ischemic stroke in a young woman.

Patients and methods: We described a clinical case.

Results: A 35 year old young woman was admitted in the Neurology department on April 24th, 2011 with left hemiplegia and hypertension history. There was no history of hypertension in her family. She has episodic loss of conscious and headaches in past months. At the physical examination, the height was of 174 cm, the weight 63 kg (BMI = 20.8 kg/m²) and the temperature at 37 °C. The blood pressure was of 170/60 mm Hg on the left arm and no pulse on the right arm. The patient was in normal conscious. There was a left flask hemiplegia. The brain CT scan displayed a right MCA ischemic stroke and a non-calcified thickening of the right CA and subclavian artery. The erythrocyte sedimentation rate (ESR) was of 78 mm in the first hour. The HIV test was negative. The patient was treated with a beta blocker, an anti-platelet agent (dipyridamole), iron, a corticosteroid (prednisone) and functional motor rehabilitation. At six weeks, the ESR was 20 mm. The patient had recurrent ipsilateral ischemic stroke with a worsening of the motor deficit on the third month after the first episode. The CT scan displayed a recent ischemic stroke in the right MCA territory.

Conclusion: Supra-aortic arteries exploration is important in ischemic stroke.

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Abstract – WCN 2013

No: 2916

Topic: 3 – Stroke

Cerebral venous thrombosis: An overlooked complication of lumbar puncture

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Background: Cerebral venous thrombosis (CVT) accounts for 0.5–1% of all strokes. Lumbar puncture is proposed as one of the risk factors. There is increased risk for CVT following a lumbar puncture in patients with other predisposing conditions for thromboembolism, such as hematological factor deficiencies, malignancy, pregnancy or recent surgery. The diversity of clinical symptoms may cause a delay in diagnosis and administration of appropriate treatment. We investigated the prevalence and risk factors for CVT following a lumbar puncture in our stroke cohort. We discuss the possible disease mechanisms and prevention strategies as well as the importance, frequency, risk factors and treatment of CVT after lumbar puncture.

Methods: The medical records of patients diagnosed with cerebrovascular diseases were reviewed retrospectively. Cerebral magnetic resonance imaging and venous magnetic resonance angiography were used in the diagnosis of CVT. A total of 37 patients were enrolled in screening.

Results: Eight (21.6%) out of 37 patients diagnosed with CVT had dural puncture recently. All patients were younger than 45-years of

age and had at least one predisposing condition for thromboembolism other than lumbar puncture. Five were young women in postpartum state.

Conclusion: Lumbar puncture may trigger CVT especially in patients with congenital or acquired thrombophilia such as pregnancy. We draw attention to the importance of neurological reevaluation of patients with persistent and progressive headache following lumbar puncture. Since missed diagnosis could result in mortality and serious morbidity, prevention strategies should be carried out for patients undergoing lumbar puncture, with known prothrombotic conditions.

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Abstract – WCN 2013

No: 2914

Topic: 3 – Stroke

Predictive factors of mannitol induced acute renal insufficiency

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Background and objective: Mannitol is commonly used to treat cerebral edema. Acute renal insufficiency (ARI) is known as one of the mannitol's most common side effects. Although concerns about renal insufficiency are a major limiting factor in mannitol use, it is very little known about mannitol induced ARI. The goal of this study is to determine predicting factors of ARI.

Methods: We retrospectively reviewed the medical records of all ischemic stroke patients treated with mannitol due to brain edema from 2010 to 2012 in Chosun University Hospital. From among 125 patients, 25 patients received mannitol below 3 days were excluded and 100 patients were selected. We identified demographic factor, medical history, NIHSS, baseline renal function, exposure to nephrotoxic or nephrophilic drugs, and laboratory data. An ARI was defined as an increase in the creatine level of >0.5 mg/dl if the base-line value is <2 mg/dl or an increase >1 mg/dl if the base-line value is >2 mg/dl.

Results: The 14 patients (14%) were diagnosed as ARI. From mannitol use to ARI development, median hospital days are 5. Glucose level before use of mannitol and peak osmolality during mannitol treatment were associated with ARI. In logistic regression analysis with suspected factors independent predictive factors of mannitol induced ARI were glucose level before use of mannitol and peak osmolality.

Conclusion: Our study presented in 14% of mannitol induced ARI. Strict glucose control before mannitol treatment may be able to reduce the rate of ARI occurrence.

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Abstract – WCN 2013

No: 2904

Topic: 3 – Stroke

In-hospital fatality in stroke patients in relation to risk factors and clinical presentation at Hadhramout: Analysis of 1072 cases

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Background: Stroke is the third leading cause of death, and a cause of long-term disability among survivors.

Objectives: To identify the frequency of in-hospital deaths among stroke patients admitted in Ibnseena Teaching Hospital, Mukalla, Hadhramout, Republic of Yemen.

Patients and methods: A retrospective cross-sectional study of stroke patients admitted at Mukalla, Hadhramout in 1/2009–12/2011. Data were collected in a questionnaire from patients' files.

Results: There were 1072 stroke cases; ischemic stroke were 78.5% and hemorrhagic 21.5%. Males were 56.5% and females 43.5%. In-hospital deaths were higher in old (36.5%) than in middle-aged and young (23.8% and 25.9% respectively), non-significantly higher in females (35.2) than in males (31.4%), highly significantly higher in hemorrhagic Stroke (**50.6%**) than ischemic type (28.2%). Hypertension, diabetes, smoking, previous attacks and family history of stroke significantly increased in-hospital deaths. Deaths among patients without risk (22.2%) with one risk factor (25%) and with ≥ 2 risk factors (34.6%). Death in late hospitalization (>24 h from onset) was significantly higher (43.4%) than in early hospitalization (within 24 h from onset) (29%). Death in comatose cases was significantly higher (67.5%) than fully (21.8%) or disturbed conscious (31.2%).

Conclusions: In-hospital death was more common in elderly, in females, in hemorrhagic stroke, and in patients with hypertension, with diabetes, with previous attacks, and with family history. Multiple risk factors, late hospitalization, and coma at hospital admission also increased fatality.

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Abstract – WCN 2013

No: 2965

Topic: 3 – Stroke

Immune thrombopenic purpura (ITP) elicited by childhood vaccination and antiphospholipid syndrome (APS) manifested as cerebral venous thrombosis: Coincidence or no?

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APS is defined by: the occurrence of at least one clinical feature: vascular event or pregnancy morbidity and the presence of at least one type of an antiphospholipid antibody (APA) on two separate occasions 12 weeks apart. Cerebral venous thrombosis (CVT) is an uncommon manifestation of APS. ITP is an autoimmune disease defined by low platelet count due to antiplatelet antibodies.

A 21 year old female was admitted due to headache, vomiting and tinnitus, beginning 15 days ago. She had a history of ITP, after MMR vaccination. Neurological examination and brain CT were normal. Fundoscopy showed bilateral papilloedema and CSF pressure was 100 cm H₂O, indicating intracranial hypertension. MRI and MRV demonstrated CVT. Anticoagulation treatment and acetazolamide were initiated. Investigation revealed elevated titers of APAs. Intracranial pressure remained elevated and she was complicated with decreased visual acuity. Consequently, she had a lumboperitoneal shunt with total regression. Based on the evidence of CVT and the elevated APAs after a 3-month interval, we diagnosed APS.

In our case, APS was first manifested as CVT in an apparently healthy young woman. However, she had an episode of severe thrombocytopenia after MMR vaccination. The importance of the presence of APAs in patients with ITP is not clear. Recent studies suggest that the measurement of APAs in patients with ITP may identify a subgroup with high risk of developing APS, within a maximum period of 72 weeks. We are not certain that our patient fits the case, in the absence of prospective studies with a more prolonged follow-up. Yet, we would like to suggest that more attention should be devoted to episodes of ITP in early childhood, as they may be connected with hypercoagulable states, like APS, in young adults.

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Abstract – WCN 2013**No: 2981****Topic: 3 – Stroke****A rare involvement of a rare disease; ischemic stroke due to relapsing polychondritis**

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Relapsing polychondritis (RPC) is a chronic inflammatory disease associated with an autoimmune disorder in cartilaginous tissue, eyes, labyrinth, blood vessels, and central nervous system. A minority of patients with RPC develop neurological involvement (3%).

A 43-year-old right handed woman acutely developed a left hemiparesis. A noncontrast CT scan revealed a right MCA infarct. During the previous three years she had history of peripheral vascular disease (deep vein thrombosis), posterior uveitis, two episodes of auricular chondritis and an episode of polyarthritits of her small peripheral joints. The blood count showed 3,100,000 red cells and 634,000 platelets per cubic millimeter. Serum electrolytes, creatinine, glucose, coagulation tests, liver functional tests, lacticodehydrogenase, and creatine kinase were normal. C-reactive protein (CRP) (119 mg/dL) and the erythrocyte sedimentation rate (ESR) (88 mm/h) were elevated. Laboratory data associated with collagen disease were all within the normal range. Echocardiography showed a mild mitral valve prolapsus and regurgitation suggesting the heart involvement.

Ischemic stroke with RPC is a rare condition which has been related to vasculitis of the CNS. The diagnosis is challenging because of its rarity and the diversity of clinical presentations. Early manifestations often remain unrecognized for prolonged periods. As a result, the diagnosis is frequently obtained only after the emergence of classic features such as auricular inflammation or other features of cartilage destruction.

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Abstract – WCN 2013**No: 2976****Topic: 3 – Stroke****Ischemic stroke in a young man with MTHFR A1298C and ACE I/D polymorphism**

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Background: Ischemic stroke is a leading cause of death and disability worldwide. Mutations in several candidate genes involving angiotensin-converting enzyme (ACE) and methylenetetrahydrofolate reductase (MTHFR) gene have been found to be associated with ischemic stroke.

Patients and methods: This report describes the case of a 29 year-old man who presented with sudden onset left hemiparesis and hemihypoesthesia. Magnetic resonance imaging investigations showed multiple acute ischemic infarct in the right temporo-parieto-frontal region. Traditional risk factors for ischemic stroke hypertension, hyperlipidemia, cardiac pathology, cigarette smoking, obesity, and alcohol consumption were all negative. Serum tests including platelet count, prothrombin time, activated partial thromboplastin time, fibrinogen, protein C, protein S, antithrombin III activity, anticardiolipin antibodies, lupus anticoagulant antibodies, autoimmunity studies, and peripheral blood smear were all within the normal limits except for mild hyperhomocysteinemia. Mutation analysis revealed homozygous MTHFR A1298C and ACE D/D polymorphism.

Conclusion: The etiology of ischemic stroke is complex and several risk factors could be involved. The present case suggests that ACE

D/D genotype together with homozygous MTHFR A1298C polymorphism may be a risk factor in the etiology of cerebral infarction.

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Abstract – WCN 2013**No: 3017****Topic: 3 – Stroke****Not so fast! The public struggles to recognise stroke**

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Introduction: Sensible suspicion of stroke and early administration of indicative physical tests are essential skills that should be possessed by all members of the general public, and which potentiate timely presentation to hospital.

Objectives: This study aims to evaluate the ability of the public in Malta to suspect and quickly test for stroke.

Methodology: Volunteers, recruited according to a probability quota sample, submitted to a structured interview detailing demographics, first aid training and knowledge about stroke. Individual chi-squared tests were performed to relate knowledge to expected influences.

Results: 500 subjects were interviewed, of whom 34% (n = 169) had been trained in first aid. 43% (n = 143) spontaneously identified unilateral weakness/numbness as indicative of stroke, 28% (n = 92) confusion or slurred speech, 20% (n = 65) loss of balance and coordination, 6% (n = 20) visual disturbances, and 5% (n = 17) unprovoked severe headache. Whilst 70% (n = 348) were able to mention at least one test to attempt to elicit warning signs of stroke, only 3% (n = 15) mentioned all three touted in the popular FAST approach. 50% (n = 248) were able to correctly identify abnormalities consistent with stroke that would be demonstrated by performing each of the three tests. Previous first aid training was not associated with significant improvement.

Conclusions: Knowledge of symptomatology indicative of stroke is lacking, as is familiarity with simple physical tests that could aid increase suspicion by first aiders. Current first aid training fails to improve awareness.

doi:10.1016/j.jns.2013.07.1026

Abstract – WCN 2013**No: 3034****Topic: 3 – Stroke****Thrombolysis combined with antiplatelet and anticoagulant therapy in young patient with hypercoagulable state and acute severe ischemic stroke**

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Background: Present guidelines for thrombolysis in stroke do not allow use of any other antithrombotic medications prior and within 24 h of tissue plasminogen activator (t-PA) administration.

Objective: To evaluate use of antithrombotic medications prior and within 24 h of t-PA treatment, followed by intensive platelet function monitoring, in young patient with severe neurological impairment and increased risk for re-thrombosis.

Patient and methods: A 48-year old woman developed acute severe right-sided hemiparesis and sensorimotor aphasia, NIHSS 23, mRS 2,

brain MSCT was normal, while MSCT cranial angiography showed occlusion of left middle cerebral artery. Patient had mastectomy due to breast carcinoma 2.5 month earlier, received chemotherapy 2 days prior the stroke and was on every day anti-estrogen therapy for 2 months. After 3.5 h of stroke onset, patient received 300 mg of aspirin, 30 mg enoxaparin iv, abciximab 10 mg iv, and t-PA 0.9 mg/kg/60 min iv. After 15 min of t-PA initiation, platelet function (multiplate method) showed suboptimal inhibition of platelet aggregation, so she received additional 10 mg of abciximab, with confirmed optimal inhibition of platelets. DSA was performed after thrombolysis, normal blood flow was registered in the left middle cerebral artery. A 24 h control brain MSCT excluded brain hemorrhage and brain MRI 3 days later showed minor infarction. Patient completely recovered within 7 days.

Conclusion: In young stroke patients with severe neurological impairment and known hypercoagulable state, combined antithrombotic medications, with intensive platelet function monitoring, should be considered to facilitate thrombolysis and prevent early re-thrombosis.

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Abstract — WCN 2013

No: 3033

Topic: 3 — Stroke

ApoE polymorphisms and ischemic stroke in a Greek population

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Background: Apolipoprotein E (ApoE) is a plasma protein exhibiting three common isoforms (E2, E3, E4). Its involvement in lipoprotein metabolism may have an impact on stroke occurrence. As results in literature are inconclusive further studies are needed to elucidate its role.

Objective: To study the role of ApoE isoforms and its interplay with environmental risk factors in patients with first ischemic stroke occurrence in Greek population.

Patients and methods: 329 patients with first ever ischemic stroke were included in our study. Strokes of cardioembolic origin and patients with autoimmune or prothrombotic syndromes were excluded. A control group of 361 subjects with no stroke history was also included in our study. Risk factors (hyperlipidemia, hypertension, diabetes mellitus and smoking) were assessed. ApoE alleles were determined in all subjects participating in the study.

Results: Genotype $\epsilon 3/\epsilon 3$ was found to have a protective role against stroke occurrence compared to other genotypes (OR = 0.674, CI: 0.480–0.946) especially in female patient subgroup. In multivariate analysis after adjustment for age, BMI, hypertension, dyslipidemia, diabetes mellitus and smoking, the role of genotype was limited and outweighed by risk factors in both genders. No association between ApoE alleles and BMI, cholesterol, triglycerides or HDL plasma levels was noted.

Conclusions: Our study supported a protective role of $\epsilon 3/\epsilon 3$ genotype, especially in female patients. However, risk factors such as age, BMI, hypertension, dyslipidemia, diabetes mellitus and smoking have strong impact on stroke occurrence and outweigh the protective role of $\epsilon 3/\epsilon 3$ genotype.

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Abstract — WCN 2013

No: 3028

Topic: 3 — Stroke

The underlying problem in stroke; the public waffles in darkness

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Introduction: There exists much uncertainty amongst members of the general public as to the actual site of the problem in a stroke.

Objectives: This study aims to evaluate patient's understanding of which organ has suffered insult in the case of stroke.

Methodology: Volunteers, recruited according to a probability quota sample, underwent a structured interview detailing demographics, first aid training and basic facts about stroke. Individual chi-squared tests were performed to relate knowledge to expected influences.

Results: 500 individuals (47% male, n = 233) were interviewed, of whom 34% (n = 169) had previously attended a formal training in first aid. 59% of subjects correctly indicated that it is the brain that has sustained damage in the case of a stroke; with a significant improvement noted amongst those who had previously attended training (68% vs 54%, p < 0.01). 25% of people questioned identified the problem to be in the limbs or face, and 10% in the heart. 90% of interviewees (n = 452) indicated the need to seek urgent medical attention even in the case transient symptomatology suggestive of stroke; whilst 98% (n = 488) correctly identified hospital as the appropriate first port of call.

Conclusions: A significant proportion of the general public has a poor understanding of the nature of stroke; first aid training results in a significant increase in prevalence of this insight. There is almost universal appreciation of the urgency and seriousness of a “stroke”, rendering the onus on prompt recognition.

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Abstract — WCN 2013

No: 3041

Topic: 3 — Stroke

Unusual cerebral venous thrombosis presenting with peripheral facial palsy: A case report

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Background: Cranial nerve palsy in cerebral venous thrombosis (CVT) is a rare clinical sign resulted from cavernous sinus thrombosis or elevated intracranial pressure. Peripheral facial palsy is extremely rare and individually reported in CVT. Herein, we present a rare case of peripheral facial palsy in transverse sinus thrombosis related to methylenetetrahydrofolate reductase (MTHFR) and factor V Leiden mutations.

Case report: A 17-year-old obese girl admitted with a 2-day history of new onset headaches, horizontal diplopia, and right-sided facial drooping. The examination was notable for right-sided peripheral facial palsy, bilaterally sixth nerve palsy, and bilaterally papilledema. Cerebral MRI and MR venography showed thrombosis of right transverse sinus. Lumbar puncture revealed clear and colorless cerebrospinal fluid with an opening pressure of 320 mm of water. Subcutaneous low-molecular-weight heparin 4000 UI twice in a day was immediately started. Acetazolamide 500 mg and topiramate 50 mg twice in a day were also given. After the prothrombotic study we found the homozygous mutations for MTHFR A1298C and heterozygous mutation for factor V Leiden. MRI performed 30 days

after discharge showed total recanalization of the affected sinus. Complete recovery of facial palsy occurred concomitant with recanalization of the transverse sinus.

Conclusions: Patients with MTHFR and factor V Leiden mutations may rarely lead to transverse venous thrombosis. The pathophysiology of peripheral facial palsy is unknown in CVT and most likely represents a pressure related situation and generally resolves with decreasing of the elevated intracranial pressure.

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Abstract – WCN 2013

No: 3043

Topic: 3 – Stroke

Internal carotid transfer of autologous bone-marrow mononuclear cells for the treatment of severe stroke in patient with thrombotic thrombocytopenic purpura

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Background: Stem cell therapy has potential regeneration capacity in patients with severe stroke.

Objective: To establish safety and potential efficacy of regenerative capacity of autologous bone-marrow derived stem cells in patient with severe stroke and thrombotic thrombocytopenic purpura (TTP).

Patient and methods: A 44-year female with known diagnosis of TTP, during relapse of the disease (thrombocytopenia $75 \times 10^9/L$) developed severe right-sided hemiparesis, and sensorimotor aphasia (NIHSS 18, mRS 5). Brain MSCT confirmed probable ischemia in the left middle cerebral artery area, though major strokes are very rare in TTP. Five daily plasma exchanges were performed and patient entered TTP remission. On the 6th day brain MRI and MRI angiography confirmed the diagnosis of ischemic stroke with open middle left cerebral artery. On the 7th day, 250 ml of autologous bone-marrow was harvested from the posterior iliac crests. After processing of bone-marrow, 8.2×10^8 of mononuclear cells (MNC) were isolated and transfused into the left internal carotid arteries 4 h after harvesting. No side effects of the procedures were noted. Serial brain MRI imaging and clinical examinations have revealed significant improvement (NIHSS 6, mRS 3 at six month follow-up) and reduction of the infarction size.

Conclusion: Internal carotid transfer of autologous bone-marrow MNC in patient with severe acute ischemic stroke and TTP was safe and patient had significant improvement of neurological deficit 6 months after procedure.

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Abstract – WCN 2013

No: 3031

Topic: 3 – Stroke

Vertebral artery dissections an under diagnosed stroke

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Background: Dissections even though theoretically considered being a cause of young stroke practically not included in the differential diagnosis of stroke quite often. Most of the reported data on dissections prove that dissections are one of the commonest causes of stroke in the

young and account for up to 2.5% all strokes and 60–75% of these are said to be a carotid artery. Analyzing our data at EMS Hospital we found that vertebral artery dissections are very common and they account 60–70% all dissections.

Material and method: First thousand cases of cerebral DSA at the Department of Interventional Neurology, EMS Hospital were included in the retrospective study.

Discussion: When analyzed we found that dissections are common at our population also and account up to 3% strokes of this 65% of them were at vertebral artery. Majority of them were without a recognizable injury. Most of them on young people but can occur at any age, most of them improve on supportive treatment and if fluctuating or deteriorating on treatment they need revascularization.

Once suspected a cerebral DSA is a must for most of the patients for confirmation of the diagnosis and planning the treatment.

Conclusions:

- Dissections are not uncommon at our population
- They are one of the commonest causes of stroke in young adults
- In the population studied vertebral artery dissection (70%) is common than the carotid artery dissection
- Most of the dissection occurs without a recognizable trauma.

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Abstract – WCN 2013

No: 3024

Topic: 3 – Stroke

A retrospective study of 100 cases of carotid artery stenting at a non teaching interventional neurology department

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Background: Carotid artery stenting (CAS) has been considered as a revascularization method with high morbidity and mortality and is considered as a second option after carotid endarterectomy (CEA) till recently. Now with refinement in techniques and technological advances made it superior to CEA. In our experience CAS is a very safe procedure and the complications are much less than the reported one and safer than the CEA. In this background we wish to publish our experience in doing last 100 cases of CAS.

Aim: Aim of the study is to compare the results and complications of the first 100 cases of CAS with the reported data on CAS & CEA.

Materials and methods: All the 100 cases of CAS done at EMS Hospital were included and compared with the reported data.

Results: In our study CAS is found to be a very safe procedure with a mortality of 1% (due to reperfusion bleeding), morbidity was also less and includes reperfusion bleeding with clinical worsening of 1%, local puncture site hematoma of 2%, and local pseudoaneurysm of 1%. No clinically significant embolic strokes were detected. Minor fluctuations in pulse and BP were seen during carotid bulb manipulations and would improve with symptomatic treatment.

Conclusions:

1. CAS is a safe procedure with a mortality of 1% and a serious morbidity of 1%
2. CAS can be done much safer than previously reported
3. Clinically significant embolic strokes are very uncommon
4. In stenosis of more than 95% in elderly with long duration of illness reperfusion bleeding is more common than embolic stroke.

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Abstract – WCN 2013**No: 2977****Topic: 3 – Stroke****Sudden-onset of hemichorea with hypomania**

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Introduction: The key role of thalamic and subthalamic nuclei in motor control through the basal ganglia thalamocortical circuit is well recognized. It is known that the thalamus is a central structure in the processing of impulses, mood and emotional behavior. The involvement of the subthalamic nucleus is however less clear. Focal lesions in these areas caused by strokes, tumors or metabolic diseases may disturb these functions.

Case report: We report the case of a 59-year-old hypertensive female, with no previous history of neurological or psychiatric dysfunction, who was admitted with sudden-onset left involuntary movements. Neurological examination was normal except for choreiform movements involving the left face, trunk and arm. There was also an acute-onset of inappropriate behaviors and mild elevation of mood characterized by disinhibition, increased energy and incessant verbal flow, with strong impulses to talk to strangers and ridiculing other patients, fulfilling ICD-10 criteria for hypomania. MRI disclosed an acute circumscribed right thalamic infarct with involvement of the subthalamic region, suggesting disease of the thalamoperforating arteries of the posterior circulation. The mood symptoms improved gradually, but persistent and discrete choreic movements were still seen interfering with voluntary movements.

Conclusion: Hemichorea is a frequent and well established manifestation of infarcts in the basal ganglia circuitry. Nevertheless, the co-occurrence of mood symptoms is rarely seen in this setting. Our case suggests an involvement of the thalamo-prefrontal projections. The role of subthalamic nucleus remains elusive but reports have been made concerning its possible role in mood regulation.

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Abstract – WCN 2013**No: 2869****Topic: 3 – Stroke****Young patients' carotid atherosclerosis: Frequency, lesion pattern, localization and risk factors**

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Purpose of study: Define the prevalence and the intensity of the young patients' extracranial arteries atherosclerosis.

Materials and methods: 1563 patients at a young stage (under 45 years of age) had the duplex ultrasound. All survey patients were broken up into groups depending on their age.

Results summary: 7.1% (n = 112) of survey patients have the carotid atherosclerotic lesion. Among them the patients were selected who had Local Intima-Media Thickening (LIMT) and Atherosclerosis Plaques (AP). Among the patients with AP there were 69% of men and 31% of women; among the patients with LIMT – 58% of men and 42% of women. 90.9% (n = 50) of cases are the patients with predominantly hypochoic AP. 67.2% (n = 37) are the cases with the stenoses up to 30% of the diameter. The stenoses more than 50% of diameter were detected in 7.2% (n = 4) of cases. In 52.5% (n = 40) of cases AP were found out in the common carotid artery (CCA) bifurcation area (31.5% rightward and 21% leftward); in 25% (n = 19) – in the brachiocephalic trunk; in 7.8% (n = 6) – in the vertebral arteries ostial area and in 14.4% (n = 11) of cases AP were located in the external carotid arteries ostial

area. The main risk factors for atherosclerosis in young patients were smoking (45.5%), uncontrolled hypertension (19.1%), and genetic factors (8.7%).

Conclusion: The young patients' carotid atherosclerosis was discovered in 7.1% of cases, among which were LIMT (45.6%) and AP (54.4%) that included hypochoic AP (90.9%) and AP localized in the CCA bifurcation (52.5%), predominantly among men (69%).

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Abstract – WCN 2013**No: 3008****Topic: 3 – Stroke****Left atrial enlargement as an indicator of severe cardioembolic stroke risk in patients with low risk atrial fibrillation**

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Background: Finding a predictive marker for stroke in patients with non-valvular atrial fibrillation (NVAF) who are considered to be low risk, is a concerning issue. We evaluated the usefulness of left atrial diameter (LAD) as a predictor in patients with cardioembolic stroke due to NVAF who had low CHADS₂ scores.

Patients and methods: We recruited eligible cases among cardioembolic stroke patients admitted to our hospital. Inclusion criteria were as follows; CHADS₂ score before stroke onset of <3, no prior anticoagulation therapy, no other cardiac or extra-cardiac complications which may affect stroke diagnosis and cardiac function. Patients diagnosed with other subtype or unclassified etiology were excluded. LAD and fractional shortening (%FS) were measured within one month of stroke onset using transthoracic ultrasonography. We also defined infarctions with a largest final diameter of 30 mm (by MR or CT scan) as "large infarctions" and all others as "small infarctions".

Results: 41 consecutive patients (16 women, mean age 73.7) were registered into our study. We found no statistical difference between the two groups with regards to age, gender, CHADS₂ score distribution or individual components. Whereas no statistical difference in %FS was found between the two groups, LAD in the large infarction group was statistically larger than that of the small infarction group (46.1 ± 9.9 mm and 38.1 ± 8.3 mm, respectively, p = 0.01).

Conclusion: With this, we speculate that enlargement of the left atrium may be a strong predicting factor of severe cardioembolic stroke in patients with NVAF and a low CHADS₂ score.

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Abstract – WCN 2013**No: 3007****Topic: 3 – Stroke****Evaluation of the fluency profiling system (FPS) as a measure of the efficiency of dynamic language networks**

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Background: Recent advances in neuro-imaging confirm the proposition that large-scale neural networks underpin natural language, and, therefore, fluency in spontaneous speaking.

The FPS provides an automatic solution to the measurement of fluency in natural language provided that minimal criteria are met in regard to Signal:Noise ratio and sample duration. The analysis uses a series of algorithms to define and characterize three distributions, for short pauses, long pauses and speech segments. The three distributions have natural log means of ~ 4.2 (~ 70 msec), ~ 6.2 (700 msec) and ~ 7.1 (1300 msec), and it may therefore be assumed that they each reflect interaction among families of variables, the sine qua non of complex dynamic systems.

Objective: To assess the validity and sensitivity of the FPS in the context of an acquired neurogenic communication disorder.

Patients and methods/material and methods: Two analyses based on the AphasiaBank Database are considered. The first analysis demonstrated that fewer than 20% of the speech samples supported effective analysis, indicating that stringent guidelines will be required for databases in this domain.

The second analysis involved a detailed comparison between small samples of aphasics and non-brain damaged controls. The analysis indicated that speech segment duration provided the most consistent distinction between the groups.

Results: Broca's aphasia showed shorter mean speech segment durations than the control group.

Conclusion: The FPS provides inferential statistics that quantify function across cognitive and motor domains beyond those provided by traditional categorical or model based diagnostic tools.

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Abstract – WCN 2013

No: 3016

Topic: 3 – Stroke

Endothelial dysfunction in smoker patients with acute ischemic stroke

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Aim: To assess intima-media thickness (IMT), endothelial function in smokers or nonsmoker patients with ischemic stroke.

Methods: The study included 72 patients with acute ischemic stroke (age, 58, 24 ± 5 , 64 years, mean \pm SD), according to the criteria TOAST (Adams et al, 1993), and 32 control patients (56, 29 ± 4 , 75 years). Of which 68, 5%-smokers, 51, 4%-nonsmokers. Endothelial function was assessed using the method of D.Celermajer (Celermajer et al., 1992). An endothelium-dependent vasodilation (FMD) was caused by a cuff of sphygmomanometer. To assess an endothelium-independent vasodilation (GTN-mediated) we performed pharmacological tests with glyceryltrinitrate. We also assessed coefficient of endothelial dysfunction, as more delicate index than ratio of GTN-mediated dilatation to FMD.

Results: Coefficient of endothelial dysfunction was higher in stroke patient on 24, 5% than in health people. In this time IMT in these patients had no different. But in smoker patient IMT was higher than in nonsmoker (0.77 ± 0.19 – smokers versus 0.59 ± 0.12). Coefficient of endothelial dysfunction is also higher in smoker patients on 6, 8% than in nonsmokers.

Conclusion: IMT and coefficient of endothelial dysfunction is higher in smoker patients than in smokers with ischemic stroke that means having of a factor of poor outcome in this disease.

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Abstract – WCN 2013

No: 3062

Topic: 3 – Stroke

Posterior circulation stroke as the initial manifestation of vertebral artery angiosarcoma

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Background: Angiosarcomas are rare highly malignant tumors of vascular origin that typically manifest with systemic signs or metastatic complications. Embolic stroke as the initial presentation of angiosarcoma of the aortic arch or its branches has been only twice reported in the literature; in both cases emboli involved the anterior circulation.

Objective: To present a unique case of vertebral artery angiosarcoma manifesting as embolic stroke of the posterior circulation.

Results: A 49 year old male was admitted to our department with a 6-day history of vertigo and gait instability. On examination he presented gaze-evoked nystagmus and pronounced body lateropulsion to the left. MRI of the brain revealed acute ischemic infarcts involving the left cerebellar hemisphere and the left occipital cortex. Digital subtraction angiography disclosed a well-circumscribed vascularized lesion around the left vertebral artery. Chest MRI detected an enhancing mass of 5×4 cm in the left thoracic outlet. The patient was subjected to high axillary thoracotomy. Histology revealed a well-differentiated angiosarcoma, composed of multiple anastomosing vessels that were lined by malignant endothelial cells showing little nuclear pleomorphism. The patient did not receive post-operative radiation or chemotherapy; two years later he remains tumor-free, with no neurological complications.

Conclusion: This is the first reported case of posterior circulation stroke complicating an angiosarcoma. Although extremely rare, angiosarcomas should be taken into account in the differential diagnosis of cerebral embolism.

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Abstract – WCN 2013

No: 3098

Topic: 3 – Stroke

Effect of atorvastatin on the CD36+, vWF+ and plasma nitric oxide level in acute lacunar stroke

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Background: The beneficial effects of statins may be related to the reduction of CD36, as anti-angiogenic factor, and vWF+ expression.

Objective: We aimed to determine of CD36+ and vWF+ and plasma NO level in acute lacunar infarctions (LI) resulting from cerebral small vessel disease (CSVD) after atorvastatin treatment.

Patients and methods: The level of CD36+, vWF+ and plasma NO were examined using flow cytometry at 48 h and after 7 days of treatment in control group of patients with LS receiving standard therapy ($n = 22$, a mean age of 62.2 ± 15.4 years), and in main group of patients with LS ($n = 17$, a mean age of 61.2 ± 17.6 years) treated with atorvastatin 20 mg, once in 20 healthy volunteers.

Results: Reduction of CD36+ and vWF+ on the 7th day of atorvastatin treatment compared to the results of control group and healthy volunteers ($p < 0.05$) indirectly reflected the decline tendency to

thrombosis due to endothelial repair. Increased expression of vWF receptor of 1.4 times was also detected by MIF density, after 7 days of atorvastatin treatment it was reduced to 107 ± 30 ($p < 0.05$). Median levels of NO ($11, 5$ mmol/L) increased ($p = 0.05$) after 7 days of atorvastatin treatment ($22, 6$ mmol/L) and in control group of patients with LS ($p = 0.02$).

Conclusion: The results of our study indicate the high potential of atorvastatin in stimulation endothelial repair in acute LS resulting from CSVD by decrease of cells bearing CD36+ and vWF+ markers and increase of plasma NO level.

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Abstract – WCN 2013

No: 3092

Topic: 3 – Stroke

Quantitative sensory testing in stroke

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Background: Sensory testing is an important part of the clinical examination in patients with stroke. Depending on the area of the lesion, different sensory pathways can be affected, which results in different clinical syndromes. Quantitative Sensory Testing (QST) is a technique to determinate thresholds for cold, warmth, heat and cold pain and it is preferably used for the detection of small fiber neuropathies, and it may serve to identify deficits in the spinothalamic tract system.

Methods: A Medoc TSA-II equipment was used to determinate thresholds. The results were compared with the site in MRI imaging.

Case series: Two illustrative cases

Patient 1: Thalamic infarct: the patient described “numbness” of his left arm and leg at presentation. The QST evaluation detected an elevated threshold for cold and warm. In correlation of clinical examination, QST results and MRI findings the lesion can be limited to the ventral posterolateral nuclei of right thalamus.

Patient 2: Brainstem infarct with hemiataxia and lesion of the medial lemniscus: This patient presented with hemiataxia, and sensory symptoms which were described as “numbness”. Vibration perception was absent. The SEPs were pathologic, however QST was normal. In accordance with the MRI image, the sensory findings could be located in the medial lemniscus.

Discussion: QST in stroke can serve as a precise tool to identify lesions affecting the temperature/pain system. Lesions in this pathway can be associated with specific handicaps, as well as neuropathic pain syndromes. It can be an important addition to clinical and imaging findings.

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Abstract – WCN 2013

No: 3095

Topic: 3 – Stroke

Cadasil, masks and mimics

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Introduction: Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is an autosomal dominant inherited vascular disease. Pathologically there is destruction of the smooth muscle cells in the small and medium-sized arteries, with deposition of eosinophilic material and fibrosis leading to progressive thickening of the arterial wall and lumen narrowing.

Case report: Female, 32 years old, complains of frequent migraine attacks since the age of 16. Some of these episodes included sensory aura but she also describes aura without migraine. The neurological examination was unremarkable. She performed a head MRI which revealed multiple focal T2/FLAIR hyperintense white-matter lesions involving the fronto-opercular regions, anterior temporal lobe horns and the semioval center. Laboratory tests were performed to screen for thrombotic risk factors. She was found to be heterozygous for the factor V Leiden mutation and MTHFR A1298C polymorphism with normal homocysteine levels and also had abnormal activated protein C resistance. Autoimmune screening, viral and bacterial serologies were unremarkable. She started on clopidogrel 75 mg and low molecular weight heparin 40 mg per day. Her follow up MRI revealed an increased number of brain lesions. She had a positive testing for the R15C missense mutation on exon 4 in the Notch3 gene.

Discussion and conclusion: This case clearly illustrates the diagnostic challenges underlying vascular disorders in young adults. The first approach is to exclude the most common etiologies; nonetheless some patients require a more complex diagnostic work up. For that purpose, clinicians must rely on the technological advances regarding brain imaging and genetic diagnosis.

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Abstract – WCN 2013

No: 3086

Topic: 3 – Stroke

Clinical applicability of stroke prognostication using age and NIH stroke scale (SPAN) index for thrombolysis in Slovenian population

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Background: Neurologist is constantly facing a dilemma, how successful thrombolysis will be in his/her patient. A possible help may be a recently introduced SPAN index¹, where only NIHSS score before thrombolysis and patient's age are considered.

Objective: With our study we wanted to evaluate clinical applicability of SPAN index in Slovenian population for Modified Rankin Score of ≤ 3 after 3 months.

Patients and methods: One-hundred-twenty-three patients were included (58 women). We evaluated SPAN index and Modified Rankin Score (after 3 months) for each patient. Cut-off was evaluated with Receiver Operating Characteristic (ROC) curve.

Results: The mean age was 65.4 (SD 11.1) years and mean NIHSS was 16.1 (SD 5.6) points. Majority of patients had moderate stroke (48.8%), 33.3% had moderate to severe and 17.9 suffered severe stroke. The mean SPAN was 81.5 (SD 17.8). Only 9 patients scored more than 100 in SPAN index. The mean Modified Rankin Score after 3 months was 2.7 (SD 1.8) points. For Modified Rankin Score of ≤ 3 , area under ROC curve was 0.67. For cut-off 71 on SPAN index, sensitivity was 95%, specificity was 31%, positive predictive value was 38% and negative predictive value was 93%.

Conclusion: SPAN index with cut-off 71 has high sensitivity and negative predictive value for our population. Despite a bit lower specificity, it may be useful with some restriction, for quick prognostication which patients might need assistance in daily living 3 months after thrombolysis.

¹Neurology. 2013 Jan 1;80(1):21-8.

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Abstract – WCN 2013**No: 3110****Topic: 3 – Stroke****Angiographic thrombectomy in venous sinus thrombosis:****Case review and series**

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A 61-year-old man with a weeklong occipital headache and acute onset confusion, dysphasia, and vomiting presented to the Emergency department. Plain CT scan of head was suggestive of venous sinus and cerebral deep venous thrombosis, a finding confirmed on CT Venogram. Following anticoagulation with low molecular weight heparin he experienced a sudden reduction of level of consciousness. Repeat CT scan of head revealed a large right occipital and a smaller left temporoparietal haemorrhage. Subsequent MR imaging revealed significant oedema and venous ischaemia in the left and right occipital, temporal and parietal lobes and additionally the left thalamus causing mass effect. An extensive angiographic thrombectomy was carried out, clearing a channel through the previously occluded saggital, transverse and straight sinuses. Repeat MR imaging and MR venogram revealed that although the sinus had reclosed, a significant resolution in oedema and venous ischaemia had occurred. Three months the patient is mobilising independently with mild cognitive impairment and a mild cortical visual deficit. Two additional cases would be presented where a similar procedure was attempted with partial success.

With reference to these cases, we propose a role of angiographic thrombectomy in venous sinus thrombosis and suggest that it may provide therapeutic benefit, particularly when significant venous congestion and ischaemia are present and prove refractory to systemic anticoagulation in a patient with deteriorating level of consciousness.

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Abstract – WCN 2013**No: 3100****Topic: 3 – Stroke****Risk of myocardial infarction polymorphisms on chromosome 9P21.3 and the risk of ischaemic stroke-replication of the results obtained in GWAS**

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Recent studies revealed that polymorphisms on chromosome 9p21.3 were risk factors for ischaemic heart disease and myocardial infarction. In few studies conducted in different populations, including two meta-analyses, it was found that polymorphisms in this region of chromosome 9 were also associated with the risk of ischaemic stroke due to large vessel. This study focused on 6 single nucleotide polymorphisms (rs496892, rs7044859, rs564398, rs7865618, rs1537378, rs2383207) in a sample of 1300 patients with ischaemic stroke admitted to Department of Neurology, University Hospital in Cracow and 700 controls.

The study did not revealed association between single polymorphisms on chromosome 9p21.3 with the risk of ischaemic stroke without differentiation of its etiologies. The C-allele of the rs1537378 polymorphism increased the risk of ischaemic stroke in logistic regression model after adjustment for age, gender, vascular risk factors, including ischaemic heart disease and myocardial infarction, however. This result was not statistically significant after Bonferroni correction.

The A-allele of the rs7044859 polymorphism on chromosome 9p21.3 increased the risk of ischaemic stroke due to large-vessel disease in a dominant model after adjustment for age, gender, ischaemic heart disease and myocardial infarction.

The A-allele of rs496892 polymorphism on chromosome 9p21.3 was protective factor for ischaemic stroke due to small-vessel disease. Meta-analysis, including own results and the results of the recently studies, revealed that five out of six study polymorphisms on chromosome 9p21.3 (rs496892, rs564398, rs7865618, rs1537378, rs2383207) were associated with the risk of ischaemic stroke due to large-vessel disease and with ischaemic stroke without differentiation of its etiologies.

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Abstract – WCN 2013**No: 3102****Topic: 3 – Stroke****Time trends in the primary and secondary prevention of stroke**

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Background: Management of modifiable risk factors for stroke is important in the primary and secondary prevention. Opinions on the optimal preventive treatment of stroke risk factors are changing.

Objective: The objective of this study was to investigate the time trends in antiplatelet treatment, management hypertension, dyslipidemia and diabetes in stroke patients treated on a stroke unit in the past eight years.

Patients and methods: The study employed a retrospective cohort design using pharmacy retrospective record analysis of medication history in the group of 3290 patients. Time trends of drug relative frequency in the past eight years were modelled by linear regression. Statistical significance of regression coefficients was estimated by Matlab statistical software.

Results: Statistically significant decreasing trend was found for ticlopidine, clopidogrel, dipyridamole and aspirin, inzulin, amiloride, amlodipine, enalapril, hydrochlorothiazide and simvastatin. Significant increase was found for atorvastatin, ramipril and metformin.

Conclusion: Time trends in the treatment of stroke risk factors reflect the growing evidence of the effectiveness and risks of individual drugs.

doi:10.1016/j.jns.2013.07.1046

Abstract – WCN 2013**No: 3118****Topic: 3 – Stroke****Stroke in young Malawian adults is associated with HIV infection and recent antiretroviral drug initiation: A prospective case-control study**

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Background: An increasing incidence of strokes in young adults (≤ 45 years) has been noted in Africa. This is postulated to relate to

HIV. We therefore investigated HIV infection and other risk factors for stroke in Malawi.

Methods: We performed a prospective case–controlled study of 222 patients (61 were ≤ 45 -years-old) meeting the WHO case definition of acute stroke, confirmed by MRI and 503 community-based (otherwise healthy) controls, loosely frequency matched for age, sex and place of residence. Multivariate logistic regression models were used for case–control comparisons.

Results: HIV infection was the predominant risk factor for stroke in under 45-year-old: recent combined antiretroviral treatment (cART) initiation (adjusted OR 15.36 [3.72,63.41] $p < 0.001$; population attributable fraction [PAF] 15%), or untreated HIV infection (3.79 [1.54,9.35] $p = 0.004$; PAF 19%), were the strongest risk factors. Abdominal obesity (PAF 11%) and heating the house with wood (PAF 8%) were also important with this younger group. Hypertension did not have a significant effect (1.36 [0.59,3.10] $p = 0.470$; PAF 6%). Risk factors for stroke in patients aged ≥ 45 years were hypertension (8.72 [4.31,17.65] $p < 0.001$; PAF 68%), smoking (2.67 [1.46,4.87] $p = 0.001$; PAF 8%) and to a lesser extent, untreated HIV infection (2.58 [1.00,6.66] $p = 0.049$; PAF 3%) and recent cART initiation (9.55 [1.45,62.9] $p = 0.019$; PAF 2%).

Conclusion: A better mechanistic understanding of stroke in this young HIV-infected population and why there is such a strong association with cART initiation will be an important step towards the reduction of stroke burden in high HIV prevalent settings.

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Abstract – WCN 2013

No: 3111

Topic: 3 – Stroke

Strokes of Takayasu disease

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Introduction: Takayasu arteritis (TA) is a chronic inflammatory disease of the aorta and its main branches and about 10 to 20% of patients with TA develop ischaemic stroke (or transient ischaemic attacks).

Objectives: To report 2 cases of strokes observed in TA revealing the disease (1) and occurring at 20 years after the first vascular manifestations.

Observations: Two women aged 40 years old are admitted for stroke's investigations. The first one has an antecedent of vascular surgery in the younger (humeral stenosis). Both presented strokes with sequellae (1). None have hypertension, valvulopathy, arrhythmia, endocarditis or intracardiac thrombosis. One patient suffered from ischaemic attacks (in the territory of a high-grade stenosis of the middle cerebral artery) and the second has one MRI of the brain showed small lesions of the white matter that were presumably of vascular origin. Both have loss pulses. The investigations by sonography and MR angiography there were still prominent vascular changes of the subclavian and common carotid arteries (2) and several vessels showed a characteristic concentric thickening of the wall on duplex sonography. The renal and the coronary investigations are normal. Upon immunosuppression, the disease took a stable course and the evolution was favorable (1) but we have observed neurological sequellae referring to the delay on diagnosis and an appropriate treatment.

Conclusion: Strokes are rare but recognized manifestations of TA so the examination of intracranial vessels (occurrence of intracranial

arterial stenosis) in TA may be helpful for primary or secondary prevention strategies.

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Abstract – WCN 2013

No: 2325

Topic: 3 – Stroke

Unilateral asterixis following thalamic stroke

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Background: Movement disorders are a recognized, albeit rare, complication of ischemic and hemorrhagic stroke. Several types of hyperkinetic and hypokinetic disorders have been reported, in the acute stage or in a delayed fashion, after vascular lesions in multiple locations. Thalamic stroke is characterized by prototypical manifestations related to the topographical location of the lesions that often point to the occluded vessels.

Objectives: We describe a case of unilateral asterixis following thalamic stroke, highlighting an unusual sign of structural CNS disorders.

Patients and methods: Case report.

Results: A 75-year old obese woman with a history of hypertension, hyperlipidemia and mild heart failure presented with acute left-sided weakness. Observation revealed left-sided ataxia and hypesthesia, with ipsilateral asterixis of the upper limb and bilateral limitation of ocular abduction. ECG revealed atrial fibrillation, and brain MRI showed acute lesions in the right postero-lateral thalamus, internal temporal and occipital regions, attributed to embolic stroke in the territory of the right PCA and postero-lateral choroidal arteries. Blood work revealed hypercholesterolemia and carotid ultrasound showed diffuse intimal thickening and bilateral plaques. Transthoracic echocardiogram showed no evidence of thrombi.

Conclusions: Asterixis is characterized by involuntary loss of tone during posture maintenance, and can be regarded as a form of negative myoclonus. It is typically bilateral and associated to toxic or metabolic encephalopathies. Unilateral presentation is rare and often indicative of a focal lesion, especially of the thalamus. Although it is an uncommon sign of stroke, vascular etiology is an important consideration, particularly in patients with risk factors.

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Abstract – WCN 2013

No: 2567

Topic: 3 – Stroke

Clinical features and outcome of stroke in the very elderly

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Background: As the population ages, it is expected that a larger proportion of the stroke population would be aged 85 and more. Specific information about this age group is limited.

Objective: To evaluate clinical characteristics, management and outcome in patients over the age of 85 as compared to patients below the age of 85.

Patients and methods/material and methods: The study included all patients discharged from Milton Keynes hospital with a diagnosis of stroke over a period of one year commencing on March 2011. All patients above the age of 85 were included in the study. Clinical data was collected and compared to a similar number of patients below the age of 85 admitted during the same period.

Results: Patients above the age of 85 represented 18.8% of all strokes included. There were no significant differences in stroke risk factors or gross markers of stroke severity between the two groups. Pre-stroke functional level was significantly worse in older patients. Similar brain imaging modalities were used, but the use of carotid Doppler was significantly less in older patients ($p = 0.04$). Inpatient mortality rates were significantly higher for older patients with 26.3% of patients dying during their hospital stay. Of those surviving, a significantly higher proportion was discharged to nursing home ($p = 0.002$). There were no significant differences in medication on discharge between the two groups.

Conclusion: Older age at stroke onset is associated with poor stroke outcome. This is related to worse pre-stroke functional levels rather than more severe stroke.

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Abstract – WCN 2013

No: 3107

Topic: 3 – Stroke

Is the frequency of symptomatic intracranial stenosis in Japanese-descendants living in western country different from the observed in Japan?

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Background and purpose: Symptomatic intracranial stenosis is responsible for 30% to 50% of all ischemic strokes in Asian patients. In North America Caucasians, the prevalence of symptomatic intracranial stenosis is around 8%. The role of risk factors related to environment and lifestyle is not yet clearly understood.

Objective: To describe the frequency of symptomatic intracranial stenosis in a multicenter sample of Japanese-descendants living in Brazil.

Patients and methods: Seventeen patients (11 women; mean age 67 yo; range: 24–89 yo) who were admitted with stroke to any of the three major referral hospitals in a 1 million inhabitants city in Southeastern Brazil (Campinas) were included. Neuroimaging were performed in the etiological workup; intracranial stenosis was diagnosed by MRI/MRA or CTA.

Results: Fourteen patients had ischemic stroke. The mechanism of vascular ischemia was attributed to intracranial stenosis in four patients, extracranial stenosis in one, cardioembolic in two, lacunar infarcts in five, and undetermined in two patients. Three patients had hemorrhagic stroke; two had subarachnoid hemorrhages and one had parenchymal hemorrhage. Hypertension was present in 11 patients, diabetes in six and smoking in three.

Conclusions: The distribution of ischemic stroke subtypes in Japanese living outside Japan shares similarities to residents in that country, such as high frequency of lacunar infarcts. The frequency of intracranial stenosis of Japanese-descendants living abroad in a western country was higher than the Caucasians, but lower than the frequency observed in Japan. This suggests that there might be a genetic predisposition that may be modified by environmental factors.

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Abstract – WCN 2013

No: 3132

Topic: 3 – Stroke

One year outcome of young adults with lateral medullary infarction

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Background: There is little study on the stroke mechanisms and prognosis according to age in patients with lateral medullary infarction (LMI). We investigated the difference of risk factors, etiologies, and prognosis between young age and old age.

Methods: We enrolled 106 consecutive patients with acute lateral medullary infarction who admitted within 7 days from stroke onset between February 2004 and January 2011. Neurological outcomes were measured by National Institutes of Health Stroke Scale (NIHSS), Barthel Index (BI) and Modified Rankin Scale (mRS) at 3 months and 1 year. We performed MR or conventional angiography to evaluate vascular pathology in all patients.

Results: Total of 106 patients was selected, with mean age of 61.5 ± 12.2 years and 76 (71.7%) male patients. There were 37 patients in young (18–59 years) and 69 in old age group (≥ 60 years). Hypertension and diabetes mellitus were frequent significantly in old age group. Most common etiologic mechanism was large artery atherosclerosis in both groups (45.9% in young age and 75.4% in old age), but, arterial dissection and small vessel occlusion were predominant in young age group (29.7% versus 2.9% in dissection and 24.3% versus 8.7% in small vessel occlusion). Pneumonia was significantly higher in unfavorable group ($p = 0.009$). Multivariable logistic regression identified age (OR = 1.06, 95% CI = 1.01 to 1.12) as a significant predictor of favorable outcome ($mRS \leq 1$) at 1 year.

Conclusion: This retrospective study suggests that younger group with LMI have a good clinical outcome and the arterial dissection and small vessel disease are frequent vascular pathologies in young patients.

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Abstract – WCN 2013

No: 2563

Topic: 3 – Stroke

Prospective USG-Doppler evaluation of the condition of CCA/ICA in patients with symptomatic and asymptomatic carotid stenosis

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Aim: The goal of our study was prospective monitoring of changes of condition of CCA/ICA in patients with symptomatic and asymptomatic carotid stenosis.

Material and methods: The study was performed on 48 patients with carotid stenosis, and 10 healthy controls. Evaluation of carotid arteries and FMD tests were done with Doppler Duplex ultrasound, and were repeated along with lab tests every 4 months for a period up to 3 years and 8 months (on average – 20 months) – patients were on antiplatelets and statins. We evaluated stenosis, morphology of plaques, IMC and endothelium functional test – flow-mediated dilation (FMD).

Results: We noticed various changes of the condition of carotid stenosis, but different in different periods of observation. In general there was progression of stenosis in the examined group. At some stages (periods of observation) progression of carotid stenosis was statistically significant ($p < 0.05$). In 2 persons stenosis asymptotically progressed to occlusion while in 2 others we found tiny spontaneous revascularization of an occluded artery. In some cases we observed transformations of arterial plaques. In some stages (periods) we observed correlation ($p < 0.05$) of the observed changes of stenosis of a carotid artery with FMD abnormalities.

Conclusion: Frequent Doppler Duplex Ultrasound monitoring of carotid arteries allows detecting dynamic changes of the condition of carotid arteries like quick progression of stenosis, or even some improvement, transformations of atherosclerotic plaques like breakdowns and ulcerations, even “spontaneous” recanalization of an

occluded artery. In some patients it “shifted” them from pharmacological treatment to endarterectomy.

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Abstract – WCN 2013

No: 2660

Topic: 3 – Stroke

Research in relationship between fibrinogen-beta chain genetics polymorphisms and intracranial atherosclerotic stenosis

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Objective: To investigate classical risk factors of intracranial atherosclerotic stenosis and its relationship with genetics polymorphisms of rs1800787 and rs1800790 which are located in promoter area in FGB gene.

Method: The research was a case-control study. Patients in Department of Neurology of Second People's Hospital of Lianyungang were enrolled consistently from December, 2010 to February, 2012. Patients enrolled were grouped into 4 groups, symptomatic intracranial atherosclerotic stenosis group (SIAS), ischemic stroke caused by small vessel disease group (SVD), asymptomatic intracranial atherosclerotic stenosis group (aSIAS) and health control (HC). Basic characteristics, history and laboratory data including level of plasma fibrinogen were recorded. We used Taqman real-time polymerase chain reaction (PCR) for genotyping.

Results:

1. A total of 164 patients were enrolled, 81 for SIAS group, 43 for SVD group, 15 for aSIAS group and 25 for HC.
2. As for classical stroke risk factors, level of CRP was in relation to IAS independently ($P = 0.012$), while other risk factors were not significantly correlated to IAS ($P > 0.05$).
3. There were no significant differences between groups in distribution of genotypes and alleles of rs1800787 and rs1800790. And significant relations were not found between IAS and alleles of rs1800787 and rs1800790.

Conclusion: Elevated level of CRP was independently related to IAS. We did not find significant correlations between rs1800787, rs1800790 genetics polymorphisms and IAS.

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Abstract – WCN 2013

No: 3144

Topic: 3 – Stroke

Spontaneous intracerebral haemorrhages in the posterior FOSSA: Characteristics, prognostic factors and outcome

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Background: Posterior fossa (PF) haemorrhages account for 11–25% of all intracerebral haemorrhages (ICH). Our knowledge concerning risk factors, pathophysiology and long-term outcome is still limited.

Objective: To evaluate the baseline characteristics, prognostic factors and long term outcome of patients with PF-ICH and to discuss hypothesis on their pathogenesis.

Material and methods: The PITCH cohort is a prospective, hospital-based cohort of 562 consecutive adults with a spontaneous ICH (recruitment 11/2004–03/2009), those who presented a PF-ICH were

eligible for the study. We performed multivariate analyses (logistic regression and survival models).

Results: 64 ICH (11% of the cohort) were located in PF: 48% were in the brainstem and 52% in the cerebellum. The in-hospital mortality was 52% and was highly influenced by the NIHSS at presentation ($p < 0.0001$), associated factors were: brainstem location (OR = 16.0; 95%CI 1.7–149.7) and ICH volume (OR = 1.1 per 1 ml increase; 95%CI 1.01–1.2). Mortality was not affected by the baseline characteristics or pre-ICH treatments. Compared with cerebellar ICH, patients with brainstem ICH were more likely to have an excessive alcohol consumption (OR = 4.2; 95%CI 1.1–16.3) and less likely to be on antihypertensive drugs (OR = 4.9; 95%CI 1.5–15.7). Three years after ICH, 36% of patients were still alive. Half of those who survived had good functional outcome. None suffered ICH recurrence.

Conclusion: The high prevalence of in-hospital mortality of PF-ICH is mainly influenced by the acute clinical presentation, but the functional long term outcome may be better than expected. The pathogenesis of PF-ICH remains unexplored, but indirect markers suggest the involvement of deep perforating vasculopathy in the pathogenesis of brainstem ICH.

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Abstract – WCN 2013

No: 3141

Topic: 3 – Stroke

Effects of MLC601 on early recurrent vascular events in post-stroke patients – The chimes study

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Background: Early vascular events are an important cause of morbidity and mortality particularly in the first three months after a stroke. We aimed to investigate the effects of MLC601 on the occurrence of early vascular events within three months of stroke onset.

Methods: Post-hoc analysis of data from the CHIMES Study, an international, multi-center, randomized, placebo-controlled, double-blind trial that compared MLC601 to placebo in improving functional outcome and reducing neurological deficit in patients with ischemic stroke of intermediate severity in the preceding 72 h (Chen et al, published online before print June 18, 2013, <http://dx.doi.org/10.1161/STROKEAHA.113.002055>) was performed. Early vascular events were defined as a composite of recurrent stroke, acute coronary syndrome and vascular deaths.

Results: In the CHIMES Study, 550 subjects were randomized to MLC601 and 549 to placebo. Risk factors and use of secondary prevention treatments were similar between the MLC601 and placebo-treated groups. The frequency of early vascular events during the 3-month follow-up period was significantly less in the MLC601 group than in the placebo group: 16 (2.9%) versus 31 (5.6%); odds ratio = 0.50 (95% CI 0.27 to 0.93; risk difference = -2.74% (95% CI -5.13% to -0.35%); $p = 0.025$ without an increase in non-vascular deaths. Kaplan-Meier analysis showed a difference in the risk of vascular outcomes between the two groups as early as the first month post-stroke ($p = 0.024$).

Conclusion: Treatment with MLC601 reduced early vascular events among patients with acute ischemic stroke in the CHIMES study. Further studies are needed to confirm this beneficial effect and elucidate its mechanism.

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Abstract – WCN 2013**No: 3210****Topic: 3 – Stroke****Gender and age characteristics of the risk factors of ischemic stroke**

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Introduction: In recent years, increased attention has been paid to the problem of gender and age specifics of the ischemic stroke. According to various studies are conflicting results about the gender and age differences of risk factors of the ischemic stroke.

Objective: To identify gender and age differences of risk factors of the ischemic stroke.

Methods: On the basis of the republican clinical hospital of Kazan the medical records of 112 inpatients aged 27 to 88 years (37 men, 75 women) with acute ischemic stroke were analyzed. Data processing was carried out using the program Biostat.

Results: The average age of women and men with acute ischemic stroke were 63,5 and 60,3 years respectively. Women were more common at the age under 55 years. At the age under 55 most frequently were atherothrombotic (69%) and cardioembolic (25%) subtypes of the ischemic stroke, while atherothrombotic and lacunar subtypes were revealed in older group. Hypertension, diabetes mellitus, and dyslipidemia were common among men and women. Smoking and transient ischemic attack were more frequently in men ($p < 0.05$), while atrial fibrillation and history of myocardial infarction were more frequently in women ($p < 0.05$). Left-hemispheric infarctions occurred with equal frequency in men and women.

Conclusion: According to our study we do not found significant gender differences among such risk factors as hypertension, diabetes and dyslipidemia. Women were more frequent at the age under 55.

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Abstract – WCN 2013**No: 3180****Topic: 3 – Stroke****Transient neurological deficit as a rare presentation of spontaneous ICH**

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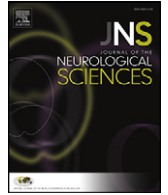
Backgrounds: Clinicians are well aware of the clinical severity and poor prognosis of patients with a spontaneous intracerebral haemorrhage (ICH). However, minor deficits or transient symptoms may be an uncommon presentation, rarely described. We aimed at identifying the clinical and radiological features of patients admitted for an ICH revealed by a transient neurological deficit.

Materials and methods: The Lille and Helsinki cohorts are hospital-based cohorts of consecutive adults admitted with a spontaneous ICH. We performed bivariate analyses on clinical and radiological variables comparing patients with NIHSS 0 at admission versus those with NIHSS ≥ 1 .

Results: Among 1490 consecutive adults, 41 patients (2.7%) had a NIHSS = 0. The median age was 63 years (IQR 54–75) vs. 70 years (IQR 59–79) in patients with NIHSS ≥ 1 ($p = 0.01$). NIHSS = 0 patients had less frequently arterial hypertension ($p = 0.049$) and atrial fibrillation ($p = 0.046$), while other vascular risk factors were similar in the two groups. 71% of the patients ($n = 29$) with NIHSS = 0 had a cortical ICH. The median ICH volume was 4.6 cm³ (IQR 1.03–7.45). After 1 year of follow-up, 39 (95%) patients were alive, while 1 patient died from a recurrent ischemic stroke and 1 was lost to follow-up.

Conclusion: Clinicians should be aware that ICH may be revealed by a transient neurological deficit in about 3% of ICH cohorts. Even if this represents a rare occurrence, a rapid assessment with neuroimaging minimizes potential misdiagnosis and mismanagement of these patients. The frequent cortical location might suggest a link with the underlying vascular disease (cerebral amyloid angiopathy).

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Topic: 4-Neuro-critical care

Abstract – WCN 2013

No: 70

Topic: 4 – Neuro-critical care

The role of the A2A receptor in cell apoptosis caused by MDMA

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Objective: Ecstasy, also known as 3,4-methylenedioxyamphetamine (MDMA), is a psychoactive recreational hallucinogenic substance and a major worldwide recreational drug. There are neurotoxic effects observed in laboratory animals and humans following MDMA use. MDMA causes apoptosis in neurons of the central nervous system (CNS). Withdrawal signs are attenuated by treatment with the adenosine receptor (A2A receptor). This study reports the effects of glutamyl cysteine synthetase (GCS), as an A2A receptor agonist, and succinylcholine (SCH), as an A2A receptor antagonist, on Sprague Dawley rats, both in the presence and absence of MDMA.

Materials and methods: In this experimental study, we used seven groups of Sprague Dawley rats (200–250 g each). Each group was treated with daily intraperitoneal (IP) injections for a period of one week, as follows: i. MDMA (10 mg/kg); ii. GCS (0.3 mg/kg); iii. SCH (0.3 mg/kg); iv. GCS + SCH (0.3 mg/kg each); v. MDMA (10 mg/kg) + GCS (0.3 mg/kg); vi. MDMA (10 mg/kg) + SCH (0.3 mg/kg); and vii. normal saline (1 cc/kg) as the sham group. Bax (apoptotic protein) and Bcl-2 (anti-apoptotic protein) expressions were evaluated by striatum using RT-PCR and Western blot analysis.

Results: There was a significant increase in Bax protein expression in the MDMA + SCH group and a significant decrease in Bcl-2 protein expression in the MDMA + SCH group ($p < 0.05$).

Conclusion: A2A receptors have a role in the apoptotic effects of MDMA via the Bax and Bcl-2 pathways. An agonist of this receptor (GCS) decreases the cytotoxicity of MDMA, while the antagonist of this receptor (SCH) increases its cytotoxicity.

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Abstract – WCN 2013

No: 2388

Topic: 4 – Neuro-critical care

A comparative effectiveness study of recombinant activated factor VII for acute intracerebral hemorrhage: Monotherapy versus combination therapy

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0022-510X/\$ – see front matter

Aim: To evaluate the effectiveness of recombinant activated factor VII (rFVIIa) and vitamin K combination therapy for acute intracerebral hemorrhage (ICH) and to compare it with rFVIIa monotherapy results of other studies.

Methods: We relied on our clinical experience and up-to-date medical literature found in Medline.

Based on pre-established criteria we selected 40 cases with spontaneous intracerebral hemorrhage documented by CT/MRI scanning, admitted to Slatina Neurological Intermediate Care Unit between 2008 and 2012. They received a single dose of 20 µg/kg of rFVIIa within four hours after the symptoms onset followed by vitamin K therapy. Each patient was evaluated by neurological (GSC and NIHSS) and neuroimaging exam (CT/MRI).

The primary endpoints: Poor outcome (defined as severe disability or death), the frequency of adverse events.

The secondary endpoints: Total hospital days.

The families were informed and their written consent obtained.

Results: The growth in volume of intracerebral hemorrhage was reduced by rFVIIa combined with vitamin K therapy comparable to those receiving higher doses of rFVIIa monotherapy. Our study did not revealed significant differences between rFVIIa monotherapy and combination therapy regarding disability but there were no thromboembolic adverse events or deaths in our group.

Conclusions: Our findings raised the question of whether lower doses of rFVIIa combined with vitamin K may be as effective as higher doses in the treatment of ICH.

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Abstract – WCN 2013

No: 2995

Topic: 4 – Neuro-critical care

Secretoneurin as an auxiliary novel biomarker for hypoxic brain injury after cardiac arrest

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Background: Accurate prediction of functional outcome in survivors of cardiac arrest (CA) is a major challenge. Up to now, data on the diagnostic accuracy of serum biomarkers are conflicting.

Objective: This study investigated serial serum concentrations of the neuropeptide secretoneurin (SN) in survivors of CA, and tested whether SN might prove as an auxiliary biomarker of post-cardiopulmonary resuscitation (CPR) prognosis.

Methods: Adult patients admitted to an academic intensive care unit after successful CPR were prospectively enrolled from September 2008 to October 2012 into this observational study. Serum SN and neuron-specific enolase (NSE) concentrations were determined daily after CPR over a period of 7 days. Patients were followed for 6 months or until death after CPR. Neurological outcome was assessed using the Cerebral Performance Categories (CPC) scale.

Results: 137 patients were included with 43.8% surviving to good neurological outcome (CPC 1–2). SN levels were significantly higher in patients with poor outcome compared to patients with favorable outcome (0–24 h: 68.6 [42.8–124.6] versus 35.7 [24.9–58.1] fmol/ml, $p = 0.009$; 24–48 h: 48.1 [27.2–63.1] fmol/ml versus 28.3 [16.9–35.9] fmol/ml, $p = 0.006$). The area under the curve for prediction of unfavorable neurological outcome was 0.706 for SN compared to 0.629 for NSE. SN levels were not influenced by MTH.

Conclusions: SN is released early after resuscitation following CA, and is associated with neurological outcome. Nevertheless, for clinical decision making concerning the continuation of treatment in comatose survivors of CA a “multimodality” approach integrating findings of clinical, neurophysiological and neuroimaging studies together with serum biomarkers is essential.

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Abstract – WCN 2013

No: 2885

Topic: 4 – Neuro-critical care

Acute motor neuropathy associated with ‘bone marrow necrosis’

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Introduction: Bone marrow necrosis (BMN) is a rare antemortem diagnosis where severe destruction of haemopoietic tissue with preservation of bone was observed. We present a patient who presented with acute motor neuropathy associated with BMN.

Case history: A previously healthy 65 year old man presented with a week history of ascending weakness. There was associated fever, cough, bone pain and night sweats. Examination demonstrated weakness, absent reflexes, and flexor plantars. CNs normal. Sensation intact.

A pancytopenia developed. Calcium and ALP were high. CT chest, abdomen and pelvis were normal. Anti-AChR Ab, VGCC, ANA, ANCA, paraneoplastic Ab, *Campylobacter*, tumour markers normal.

Repetitive stimulation, SNAPs, NCV normal. CMAP attenuated. No evidence of LEMS. Consistent with motor neuropathy (AMAN).

Bone marrow: BMN with a few blast cells. No malignant infiltration.

Progress: Condition rapidly worsened and he was admitted to the ICU. He had profound generalised weakness, severe pain even to passive movements requiring morphine analgesia. He developed type 2 respiratory failure. Further bone marrow studies confirmed BMN. Despite excellent care condition deteriorated and the patient died.

Discussion: LEMS is the only reported association with BMN. As BMN is a severe, usually fatal condition the opportunity to investigate and report neurological associations could be missed. The pathophysiology of BMN is not established but may be associated with myeloid leukaemia; hence paraneoplastic or auto-immune neurological syndromes may be the likely association. The awareness of such association may help focus investigations as presentation of BMN could be very complicated.

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Abstract – WCN 2013

No: 2897

Topic: 4 – Neuro-critical care

Severe unresponsive cerebral vasospasm treated with mild hypothermia: Case report and literature review

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Background: Hypothermia was used in selected patients with refractory vasospasm following SAH, in which no other intervention proved itself useful. Early moderate hypothermia is level 1 evidence for hypoxic ischemic encephalopathy after cardiorespiratory arrest. There are other indications not yet validated for the use of therapeutic hypothermia, such as ischemic stroke, traumatic brain injury, intracerebral hemorrhage, among others.

Objective: To present the case of a patient who was treated with hypothermia as a last resort in refractory vasospasm following SAH.

Patient and methods: Patient is a 57 y/o, right handed white female, who presented to the E.D. after an episode of sudden headache and stupor. CT scan on arrival revealed a Fisher 3 SAH, with a ruptured ACOM aneurysm on DSA. She evolved with progressive vasospasm, with tetraparesis and elevated velocities (200 cm/s) on TCD. CTA showed bilateral ACA and MCA vasospasm, with a PI demonstrating incipient ischemia of both ACA and MCA territories. After no response to common treatment protocols such as nimodipine, we decided to use moderate hypothermia for 8 days.

Results: Patient was treated with moderate hypothermia of 33° over 8 days. Doppler velocities decreased from 200 cm/s to 120 cm/s. After rewarming the patient presented with moderate tetraparesis and a frontal lobe syndrome, with a control MRI showing no additional ischemic lesions and regression of vasospasm in MRA.

Conclusions: Moderate hypothermia can be of help in the management of otherwise intractable severe vasospasm following SAH. Further clinical trials are warranted to investigate the full potential of this intervention.

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Abstract – WCN 2013

No: 2814

Topic: 4 – Neuro-critical care

Hemodynamic adjustment optimization with sympathomimetic agents in patients after severe traumatic brain injury (sTBI)

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Introduction: Sympathomimetics are the first agents for rapid correction of hemodynamics in patients with acute sTBI demanding optimal cerebral perfusion pressure maintenance.

Goal: To personalize the sympathomimetic application according to hemodynamic derangements guided by transpulmonary thermodilution (PiCCO) in patients with sTBI.

Methods: 58 patients with sTBI (GCS < 9) were enrolled in this study, six of them died. All patients received norepinephrine, dopamine, phenylephrine or combination of two agents for hemodynamic adjustment according to PiCCO monitoring measurements. Hemodynamic variables before and after sympathomimetic administration were estimated for therapy modification.

Results: Part of the patients (31%) had low arterial blood pressure and high heart rate due to systemic vascular resistance decrease. In this case the most effective was α -adrenergic agent administration

(phenylephrine). Hemodynamic profile in 15% of the patients was characterized by low cardiac output and heart rate. In these patients β -adrenergic agent (dopamine) provided the most appropriate correction of hemodynamics. Forty percent of the patients had mixed hemodynamic profile, which required both α - and β -adrenergic agent administration (norepinephrine). If norepinephrine was ineffective for heart rate, systemic vascular resistance and cardiac output correction we used a combination of two drugs: dopamine + phenylephrine (9%) or norepinephrine + phenylephrine (5%).

Conclusion: Advanced hemodynamic monitoring with PiCCO provides a reasonable and personalized selection of sympathomimetic agents in patients with acute sTBI.

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Abstract – WCN 2013

No: 2813

Topic: 4 – Neuro-critical care

Clinical syndromes of neurotransmitter dysfunction after severe traumatic brain injury (sTBI)

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Background: Brain functions are provided by different neurotransmitter systems, which help to maintain consciousness, movement activity, muscular tone and reflex regulation. Functional interaction between intact brain, balance between glutamate/GABA axis and dopamine/acetylcholine axis play a pivotal role in posttraumatic recovery processes.

Objective: To identify the main clinical manifestations of brain neurotransmitter system dysfunction and to evaluate its prognostic and therapeutic significance.

Patients and methods: 88 patients with sTBI (GCS < 8) entered this study in acute period (1–15 days) and 70 patients – in subacute period (18–70 days after trauma). Brain damage localization was verified by 1.5–3 T MRI in 79 patients. Plasma catecholamine and glutamate levels were measured in dynamics by HPLC.

Results: We have identified syndromes, which can indicate the preferential dysfunction in the main brain neurotransmitter systems: Glutamatergic Insufficiency (GIS) and Redundancy (GRS) Syndromes, Cholinergic Insufficiency Syndrome (CIS), Dopaminergic Insufficiency (DIS) and Redundancy (DRS) Syndromes. The first three syndromes had equal frequency in acute period of sTBI. In subacute period the most frequent was GRS and the less frequent – CIS. Plasma glutamate level was the highest in patients with GRS and the lowest in patients with GIS. The most unfavorable outcome was noticed in patients with CIS, the most favorable outcome – in patients with GRS.

Conclusion: It is well known that posttraumatic recovery often depends on regular choice of neuromodulators like glutamatergic, GABAergic, dopaminergic and cholinergic agents. Revealed clinical syndromes can be the basis for selecting a specific directed pharmacotherapy for brain function recovery after sTBI.

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Abstract – WCN 2013

No: 2790

Topic: 4 – Neuro-critical care

Functional results of surgery neurogenic heterotopic ossification in patients with severe traumatic brain injury: About 19 cases

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Introduction: Neurogenic heterotopic ossification is a frequent complication in brain injury patients. The functional impairment may be severe.

The aim is to determine functional outcomes of patients undergoing surgery for heterotopic ossification.

Materials and methods: This is a retrospective study of 19 patients with severe brain injury followed for neurogenic heterotopic ossifications between January 2010 and March 2013. Patients were evaluated before and after surgery with an epidemiologic profile, a range of movement and a functional assessment.

Results: There were 19 patients with 20 operated joints. The majority of the patients were male with 16 men, the average age was 31.5 years (19–44 years). The median duration of coma was 84.5 days (19–150 days).

Preoperative mobility: for the hip: 31° (0°–65°), to the elbow 28° (0°–80°), and the knee 45° (10°–80°). The functional assessment: walking and not sitting in 8 patients, not hand-back in 9 patients, not hand-neck in 8 patients and not hand-mouth in 7 patients.

All patients received a functional rehabilitation based on continuous passive motion in addition to functional work and surgery excision of heterotopic ossifications. There was a significantly increased mobility after surgical treatment, with a gain of average mobility of 34° at the hip, 51° at the elbow and 46° at the knee.

Discussion and conclusion: Neurogenic heterotopic ossification constitutes a disability in addition to various motor deficits, sensory or cognitive patient with severe brain injury. Surgery followed by appropriate rehabilitation has the goal of reducing pain and improving function.

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Abstract – WCN 2013

No: 2725

Topic: 4-Neuro-critical care

The role of CT-Angiography in diagnosing brain death

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Background: Due to its widespread accessibility CT-Angiography (CT-A) is a promising technique in detection of intracranial circulatory arrest in brain death (BD). Several studies assessed this tool but neither have standardised evaluation parameters been developed nor has information about specificity become available.

Methods: We conducted a prospective study between January 2008 and April 2012. Thirty patients were admitted to our University Hospital (16 males, 14 females, age 18–88 years) and underwent CT-A scanning at two occasions: immediately after the first signs of loss of brain stem reflexes and after definitive determination of brain. The results of CT-A were compared to transcranial Doppler ultrasonography (TCD) and electroencephalography (EEG).

Results: In 3 out of 30 patients, we observed a termination of contrast flow at the level of the skull base and foramen magnum in arterial scanning series before clinical determination of brain death. After clinical determination of BD, opacification of all vascular territories in arterial phase scanning was found in one case, but venous phase scanning revealed no blood return in internal cerebral veins (ICV). In all other cases, contrast filling ceased at level of skull base or below. Specificity of CT-A in detection of intracranial circulatory arrest was 90% and sensitivity was 97%.

Conclusion: CT-A is reliable and appropriate technical investigation to detect intracranial circulatory arrest in BD. Evaluation of contrast enhancement in arterial phase scanning seems to be more

reliable than in venous phase. An international consensus about a uniformly applied CT-A protocol for evaluation of BD should be established.

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Abstract – WCN 2013

No: 2716

Topic: 4 – Neuro-critical care

System for the assessment and communication of patients with disorders of consciousness (DOC)

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Patients with disorders of consciousness (DOC) normally stay in a rehabilitation institution for several months before they are released to professional home-care or to a private home – often without professional support. It is advantageous to have a system that can assess the cognitive functions of the patients in frequent intervals to show trends and allow patients to communicate if cognitive functions are still present.

To realize such a setup an EEG based system was developed that allows assessing cognitive functions and establishing an interface for communication. This was realized through

- (i) auditory evoked potentials (AEP),
- (ii) vibrotactile (VT) evoked potentials and
- (iii) motor imagery experimental protocols.

Both the auditory and vibrotactile experiments are designed to elicit a P300 response similar to a P300 spelling device. After the initial assessment with a positive P300 response, the patient can also be trained to use the system for communication. Motor imagery may also be used to control a brain computer interface (BCI). In this case, the patient has to imagine a right or left hand movement. This will result in an event-related desynchronization (ERD) and event-related synchronization (ERS) over the sensorimotor regions which are analyzed with the BCI.

This auditory, vibrotactile and motor imagery setup can assess whether the patient is able to follow instructions and to answer yes and not questions. Such a system is a new, practical EEG based device that can also work in real world environments to assess DOC patients and to establish communication.

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Abstract – WCN 2013

No: 2728

Topic: 4 – Neuro-critical care

Use of noninvasive ventilation at a neurological step down unit

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Introduction: The neurological step down unit (NSDU) is a specific unit for neurological patients that need intensive care, due to important dependency and continuous monitoring with regard to their moderate complexity. So far, there is no evidence of noninvasive ventilation (NIV) characterization in the NSDU.

Objective: Characterize the use of NIV in a NSDU at a private hospital.

Methods: A prospective and observational study, during an 8 month period after the inauguration of our hospital's NSDU and the beginning of physiotherapy service at this unit. The total number of admitted patients, number of physiotherapy prescriptions, patients profile, and the prevalence of NIV use were collected.

Results: During the study period, there were a total of 291 admissions, of these 137 (47%) were with physiotherapy prescriptions. Of patients with physiotherapy prescription, 24 (17.5%) used NIV. Patients that used NIV had a median age of 78 years (range of 18–98), 54% (n = 13) had admission diagnose of stroke, followed by 8% (n = 2) of brain tumor exeresis, 8% (n = 2) traumatic head injury, and 8% (n = 2) due to convulsive crisis. The median days of NIV use was 4 days (range of 1–31), 33% (n = 8) were tracheostomized, 54% (n = 13) used an orofacial mask and 13% (n = 2) used total face mask.

Conclusion: The use of NIV at a NSDU has a low prevalence, few days of use, with stroke as the most frequent diagnosis.

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Abstract – WCN 2013

No: 2654

Topic: 4 – Neuro-critical care

Zingiber officinale alters 3,4-methylenedioxymethamphetamine-induced neurotoxicity in rat brain

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Objective: The effects of this traditional herbal medicine on 3,4-methylenedioxymethamphetamine (MDMA) induced neurotoxicity have not yet been studied. The present study considers the effects of *Zingiber officinale* on MDMA-induced spatial memory impairment and apoptosis in the hippocampus of male rats. The spice *Z. officinale* or ginger possesses antioxidant activity and neuroprotective.

Materials and methods: Rats (200–250 g) were classified into three groups (control, MDMA and MDMA plus ginger). The groups were intraperitoneally administered 10 mg/kg MDMA, 10 mg/kg MDMA plus 100 mg/kg ginger extract, or 1 cc/kg normal saline as the control solution for one week (n = 7 per group). Learning memory was assessed by Morris water maze (MWM) after the last administration. Finally, the brains were removed to study the cell number in the cornu ammonis (CA1) hippocampus by light microscope, Bcl-2 by immunoblotting, and Bax expression by reverse transcription polymerase chain reaction (RT-PCR). Data was analyzed using SPSS 16 software and a one-way ANOVA test. In this experimental study, 21 adult male Sprague Dawley rats were used.

Results: Plus ginger group relative to the MDMA group (p < 0.001). Cell number increased in the MDMA plus ginger group in comparison to the MDMA group. Down-regulation of Bcl-2 and up-regulation of Bax were observed in the MDMA plus ginger group in comparison to the MDMA group (p < 0.05). Escape latency and traveled distances decreased significantly in the MDMA.

Conclusion: Of MDMA-induced neurotoxicity. Our findings suggest that ginger consumption may lead to an improvement.

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Abstract – WCN 2013**No: 2578****Topic: 4 – Neuro-critical care****Awakening after initiating levodopa in a thalamic hemorrhage**

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Background: The best therapeutic approach to conscious changes after severe brain damage (vegetative state or minimally conscious state) is not well established. However, some cases reported benefit of levodopa after traumatic brain injury.

Objective: To present a case of thalamic and intraventricular hemorrhage with response to levodopa.

Patients and methods: Clinical case.

Results: A 64-year-old man was referred to our hospital comatose (GCS 4), with a left thalamic hemorrhage with tetra-ventricular extension and obstructive hydrocephalus on CT. Acute treatment included insertion of right frontal intraventricular catheter and intraventricular fibrinolysis. At four weeks of follow-up, despite hematoma absorption, only a slight clinical improvement was observed. The patient was vigil, tetraparetic, spontaneously breathing by tracheostomy, with neither verbal response nor command obedience (GCS 8). He started levodopa therapy six weeks after stroke and two days later he started to follow moving objects with his eyes and to obey to simple motor commands. A progressive clinical improvement was noted in the next weeks permitting physical rehabilitation. At ten weeks of follow-up, the last one, the patient could express verbally simple words, answer to simple questions and do more complex motor tasks with a grade 3 motor strength (GCS 13).

Conclusion: This case presented a temporal relationship between levodopa use and improvement of consciousness state in a patient with a severe thalamic hemorrhage, although this association may be equivocal.

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Abstract – WCN 2013**No: 2058****Topic: 4 – Neuro-critical care****Phenytoin serum concentrations and factors affected after traumatic brain injury**

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Background: Phenytoin is an effective drug used for decreasing the risk of early post-traumatic seizures in patients with traumatic brain injury.

Objective: This study investigates phenytoin level and factors affecting seizure control in patients with traumatic brain injury.

Patients and methods: 122 patients with traumatic brain injury data were collected from Supprasithiprasong Hospital, Thailand, between May 2012 and January 2013. Phenytoin serum concentrations were measured in days 3–7 after started phenytoin. Two regimen of phenytoin had been taken; 100 mg q 8 h or 300 mg q 24 h with or without loading dose.

Results: 64 patients had total phenytoin concentrations in sub-therapeutic level (52.5%), 52 patients had phenytoin level in therapeutic range (42.6%) and 6 patients had phenytoin level in toxic level (4.9%). Eleven patients (9.0%) did not control seizures during the first 7 days. Five of them (45.5%) had phenytoin concentration in sub-therapeutic level, 5 patients (45.5%) had phenytoin concentrations in therapeutic range and 1 patient (9.1%) had phenytoin concentration in toxic level. Fever has correlation with phenytoin level ($p = 0.001$). Serum albumin concentration, severity of brain injury, and GCS score were not correlated with phenytoin level ($p > 0.05$).

Conclusion: More than half of the patients who were taking phenytoin for prophylaxis of seizures in patients with traumatic brain injury had phenytoin in subtherapeutic level. Phenytoin dosage should be adjusted appropriately to effective control seizure.

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Abstract – WCN 2013**No: 1178****Topic: 4 – Neuro-critical care****Acyclovir-induced nephro- and neurotoxicity: A case report**

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Introduction: Acyclovir is a highly effective antiviral agent available for treatment of herpes simplex virus and varicella zoster infections. Although acyclovir therapy is usually well tolerated; serious adverse effects such as acute renal failure and neurotoxicity have been reported usually following parenteral administration.

Case report: We present a case of acyclovir-induced nephrotoxicity and neurotoxicity that were reversible with cessation of treatment and hemodialysis.

Discussion: Monitoring of renal function is essential for an early diagnosis of acyclovir-induced nephrotoxicity. All patients who are treated with IV acyclovir should undergo a baseline BUN and creatinine evaluation before the regimen is started and should be monitored daily while treatment is ongoing.

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Abstract – WCN 2013**No: 2481****Topic: 4 – Neuro-critical care****Temperature modulates the neuronal response in the thalamus and the cortex in rats**

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Temperature fluctuations significantly impact clinical outcome after neurologic injuries. Although therapeutic hypothermia is used during cardiopulmonary bypass, and after hypoxic ischemia or stroke, the neuroprotective mechanisms underlying the temperature changes have not been fully elucidated. We electrophysiologically evaluated uninjured rat brains in response to mild hypothermia (32–34 °C) and mild hyperthermia (38.5–39.5 °C) by external systemic cooling/warming in rats (N = 6 each). For each rat, the EEG, and the neural spikes from the somatosensory cortex (CX) and the ventral posterolateral nucleus of the thalamus (TH) were continuously recorded after stabilization of temperature: normothermia at baseline (36.5–37.5 °C), hypothermia or hyperthermia, and normothermia after temperature modulation. The burst suppression ratio (BSR) in EEG, cortical and thalamic spike signals was computed using a custom algorithm. BSR significantly increased during hypothermia compared with baseline (EEG: $78 \pm 3\%/30 \pm 12\%$, $P = 0.010$; CX: $79 \pm 3\%/31 \pm 11\%$, $P = 0.011$; TH: $63 \pm 7\%/13 \pm 5\%$, $P = 0.003$). Under hypothermia, there was a marked decrease from baseline in the firing rate of spontaneous spike activity in the cortex ($16.3 \pm 4.8/7.4 \pm 1.8$ spike/s, $P = 0.017$) and in the thalamus ($3.3 \pm 1.1/1.5 \pm 0.4$ spike/s, $P = 0.046$). With peripheral nerve stimulation to evoke brain activity, the spike amplitude significantly decreased with hypothermia in cortical ($94.2 \pm 9.2/61.4 \pm 3.0$ μ V, $P = 0.014$) and

thalamic ($97.1 \pm 8.0/72.0 \pm 5.6 \mu\text{V}$, $P = 0.001$) neurons, with a prolonged response latency (CX: $9.3 \pm 0.5/14.4 \pm 1.2$, TH: $4.8 \pm 0.2/6.3 \pm 0.4$ ms, both $P < 0.01$). The increased BSR, reduced firing rate and spike amplitude, and prolonged response latency indicated that hypothermia suppressed the metabolism in the cortex and the thalamus, which might be one of the main mechanisms underlying neuroprotective benefits of hypothermia.

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Abstract – WCN 2013

No: 2507

Topic: 4 – Neuro-critical care

Chronic hydrocephalus after experimental subarachnoid hemorrhage

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Chronic communicating hydrocephalus is a significant health problem affecting up to 20% of survivors of spontaneous subarachnoid hemorrhage (SAH). The development of new treatment strategies is hampered by the lack of well characterized disease models. This study investigated the incidence of chronic hydrocephalus by evaluating the temporal profile of intracranial pressure (ICP) elevation after SAH, induced by endovascular perforation in rats.

Twenty-five adult male Sprague–Dawley rats were subjected to either endovascular perforation or sham surgery. Twenty animals survived SAH induction. At 7, 14 and 21 days after surgery ICP was measured by stereotaxic puncture of the cisterna magna. On day 21 T-maze test was performed. On day 23, the relative ventricle area was evaluated by histology.

On day 7 after surgery all animals showed normal ICP. Observing an ICP of 10 mm Hg as cut-off, 3 animals showed elevated ICP on day 14 and another animal on day 21. The absolute ICP values were significantly higher in SAH compared to SHAM animals on day 21. The incidence of ICP elevation was 40% in SAH animals. On day 21, results of T-maze testing were significantly correlated with ICP values, i.e. animals with elevated ICP showed a lower number of alterations and a delayed decision. Histology yielded a significantly higher relative ventricle area in animals with ICP elevation compared to animals without ICP elevation.

In conclusion, the current study shows that experimental SAH leads to chronic hydrocephalus, which is associated with ICP elevation, behavioral alterations and ventricular dilation in about 40% of the animals.

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Abstract – WCN 2013

No: 2479

Topic: 4 – Neuro-critical care

Therapeutic conundrum after diagnosis of Foix–Jefferson syndrome caused by intracavernous aneurysm

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Introduction: Intracavernous carotid artery aneurysms are usually symptomatic in the form of several neurological deficits based on their anatomic location. They are rarely associated with life-threatening complications, such as thrombosis and hemorrhage, commonly found in a ruptured form.

Case report: A 57 year-old woman was admitted with left eyelid ptosis, associated with moderate headache. Patient denied suffering from other illnesses. Relevant findings were the eyelid ptosis, ophthalmic paresis of the left third cranial nerve, of the abducens nerve and the ipsilateral trochlear nerve. Paresthesia and hypoesthesia were also found on the ophthalmic portion of the trigeminal nerve, without associated symptoms. Findings are compatible with the syndrome caused by carotid aneurysm compression of nearby structures, a cavernous-carotid fistula or a cerebral venous thrombosis. An angio-tomography showed aneurysmatic dilation of saccular multilobulated aspect located at the supraclinoid segment of the left internal carotid artery, with a maximum aneurysm diameter of 7.4 by 3.2 mm. Arteriography found a thrombosed aneurysm of the left petrous segment. Treatment was initiated with 100 mg of phenytoin every 8 h alongside nimodipine. The patient's condition remained stable; surgical intervention was not conducted due to the risk of thromboembolization.

Conclusion: Recognition of Foix–Jefferson syndrome alerts to the existence of underlying intracavernous carotid artery aneurysm that allows the neurologist to make an early diagnosis. The conundrum lies in whether the patient should be surgically or clinically treated. The patient had clinical monitoring and treatment before any serious adverse events, such as aneurysm rupture, whereas a surgical intervention would imply unnecessary risks.

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Abstract – WCN 2013

No: 2428

Topic: 4 – Neuro-critical care

EEG pattern of alpha activity and survival in comatose patients

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Purpose: To identify the factors determining survival in alpha coma. **Patients and methods:** Totally 21 patients in comatose state with alpha frequency patterns have been investigated. Patients were grouped as traumatic brain injury (7 cases—1st gr), basal ganglia hemorrhage (4 cases—2nd gr.), brainstem stroke (5 cases—3rd gr.) and diffuse hypoxic encephalopathy due to cardiac arrest (5 cases—4th gr.). Patients were rated by Glasgow Coma Scale (GCS). Spontaneous breathing, pupil reactions and gag reflexes were investigated. Brain lesion was assessed on conventional MRT (1.5 T). Brain electrophysiology was assessed by a 19 channel EEG apparatus. Non-parametric statistics was performed by SPSS-11.0.

Results: In the 1st group 5 patients revealed high amplitude non-reactive alpha frequency predominantly in fronto-temporal dimensions, while 2 patients showed reactive alpha frequency predominantly in parietal-occipital dimensions. In the 2nd group 3 patients showed high amplitude non-reactive alpha frequency in frontal regions and 2 patients revealed reactive EEG in dorsal parietal and temporal regions. In the 3rd group all patients showed non-reactive alpha frequency in fronto-temporal regions. The 4th group found 2 cases of reactive EEG in parietal-occipital region and 3 patients—non-reactive EEG in the same region. Three patients from the 3rd group and two patients from the 4th group were on artificial breathing. From all groups only 3 patients survived from coma. EEG patterns, brain damage location and size as well as brainstem reflex activity were entered in the logistic regression toward the outcome.

Conclusion: Apparently, in alpha coma the reactive EEG in parieto-occipital dimensions leads to more favorable prognosis.

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Abstract — WCN 2013**No: 2279****Topic: 4 — Neuro-critical care****Effects of prehospital neurologist assessment on appropriate delivery to neurology facilities: The stroke emergency mobile (STEMO) project**

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Objective: Neurological diagnosis is challenging in prehospital emergency medicine. Neurological symptoms vary widely and may be similar in different diseases. Pre-hospital decisions are important to deliver patients to hospitals with specialists' expertise when required. We aimed to evaluate whether appropriate delivery of patients with neurological diseases can be increased by neurologists who are directly engaged in prehospital care in the STEMO project in Berlin/Germany.

Patients and methods: STEMO is a specialized ambulance equipped with a CT-scanner plus point-of-care laboratory and staffed with a neurologist trained in emergency medicine. The effects of the STEMO implementation were evaluated in a prospective study comparing weeks with and without STEMO-availability. STEMO was deployed when the dispatchers suspected an acute stroke during emergency calls. Discharge diagnosis and destination were collected from hospitals to which patients were delivered.

Results: At the time of preliminary analysis, discharge documentation was available in 6012 patients (3117 in STEMO and 2895 in control weeks). 50% of all diagnoses were cerebrovascular, 22% were non-cerebrovascular but neurological and 28% were non-neurological. Patients with cerebrovascular respectively neurological (including cerebrovascular) diagnoses were less frequently delivered to hospitals without Stroke-Unit or without Neurology department when STEMO was deployed (6 versus 12%, $p < 0.01$ and 4 versus 6% respectively, $p = 0.02$).

Conclusion: Emergency assessment by a STEMO neurologist enhances the chances to be admitted to appropriate in-hospital care. Outcomes will be presented at the WCN.

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Abstract — WCN 2013**No: 2293****Topic: 4 — Neuro-critical care****Association of the *Shank3* gene mutations coupled with the 22q13.3 deletion syndrome**

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Background: Phelan–McDermid syndrome results from simple 22q13 deletions, ring chromosomes and unbalanced translocations. The gene responsible for the core neurological features is *SHANK3*, which is found to be mutated in patients with autism spectrum disorders and intellectual disability. The aim of the present study was to detect the chromosomal alterations and *SHANK3* mutations in Phelan–McDermid syndrome patients in Coimbatore region.

Methods: In the present study among 468 patients observed, only 8 patients were selected based on the clinical features, behavioural habits and standard questionnaire. The work was carried out in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki. Commercially available FISH probes and

PCR-SSCP techniques were used to detect chromosomal alterations and the mutations of *SHANK3* gene in patients.

Results: In our study, 22q13.3 deletion was observed in 5 patients and 2 patients had deletions of unbalanced translocation and one patient was mosaic for the 22q13.3 deletion with 85% of the cells showing deletion. Moreover 4 patients with nonsense de novo *SHANK3* mutation was detected and these results imply the mental status is related to sporadic occurrence of *SHANK3* gene complex multiple deletions.

Conclusion: Our findings conclude that 22q13.3 deletions may be a more frequent cause for the present study, and the *SHANK3* gene is involved in the Phelan–McDermid syndrome. Furthermore, our study supports haploinsufficiency of the gene *SHANK3*, which codes for a structural protein of the postsynaptic density, as a major causative factor in the neurological symptoms of 22q13.3 deletion syndrome.

doi:10.1016/j.jns.2013.07.1080

Abstract — WCN 2013**No: 1167****Topic: 4 — Neuro-critical care****Computational EEG analysis for critically ill patients based on the standardized terminology of the ACNS**

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Continuous EEG monitoring is an important tool to recognize clinically invisible deteriorations in critically ill patients. However, manually reviewing continuous EEG recordings requires substantial resources, which are often limited in intensive care units.

A computational algorithm for automatic analysis and trending of continuous EEG recordings from critically ill patients is being developed. Main Terms and most important Major Modifiers of the ACNS' standardized critical care EEG terminology are automatically determined and represented graphically, allowing reading of trends in the neurological state of patients.

Continuous EEG recordings from 65 patients from ICUs in three different clinics are used for algorithm development. Artifacts are removed using the PureEEG algorithm. Then EEGs are split into segments representing normal waves, discharges, or spikes. Localization and morphology of these segments are assessed, and finally the segments are reassembled into groups representing rhythmic activity, periodic discharges or spike-and-wave patterns (Main Term II). The localization of these patterns is determined (Main Term I) and finally frequency and amplitudes are determined.

In a visual analysis of results from the first algorithm version all patterns covered by Main Term II could be detected successfully. Results for localization, frequency and amplitude have been checked on a sample basis and were correct in most cases.

First results are very promising, but a quantitative performance analysis will be important, although methodologically demanding. We see a huge potential benefit from our algorithm analyzing EEGs from ICUs. Resources could be substantially saved, allowing for extended coverage of EEG monitoring in critically ill patients.

doi:10.1016/j.jns.2013.07.1081

Abstract – WCN 2013**No: 2093****Topic: 4 – Neuro-critical care****A rare presentation of brain abscess and ventriculitis caused by *Klebsiella pneumonia***

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Background: *Klebsiella pneumonia* is a Gram-negative, non-motile, encapsulated, lactose fermenting, rod shaped bacterium. This is a pathogen that causes opportunistic infections, mainly in immune compromised patients. Typical manifestations of *Klebsiella* infections are pyogenic liver abscess, meningitis, septicemia, pneumonia and endophthalmitis. In contrast, *K. pneumonia* is an uncommon pathogen of bacterial meningoenzephalitis.

Objective: Our aim is to report a case of a rare presentation of brain abscess and ventriculitis caused by *K. pneumonia*.

Patient and results: A 50-year-old female Korean was sent to our hospital three hours after acute onset of confusion. Her past medical history was depressive disorder. She had marked nuchal rigidity. The cerebrospinal fluid (CSF) was yellowish with pleocytosis (350/mm³; N/L = 96%), increased protein (536 mg/dl), and a low glucose level (7 mg/dl). Brain abscess and ventriculitis was found in the lateral ventricles on brain MRI. Culture of CSF was isolated *K. pneumonia*. She was treated with intravenous Carbapenem, Ampicillin, Vancomycin and 3rd Cephalosporin for two months and clinical symptom was fully recovered.

Conclusions: In our case, ventriculitis with periventricular inflammation and necrosis, and multiple brain abscesses were manifestations of *Klebsiella* meningoenzephalitis. To our knowledge, this is the first case report of *Klebsiella* meningoenzephalitis with brain abscess and ventriculitis in immunocompetent patient. As like this case, if there are brain abscess and ventriculitis in meningoenzephalitis, it should be considered diagnosis of *Klebsiella* meningoenzephalitis.

doi:10.1016/j.jns.2013.07.1082

Abstract – WCN 2013**No: 2035****Topic: 4 – Neuro-critical care****Valproate-induced hyperammonemic encephalopathy with triphasic waves: A case report**

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Introduction: Sodium valproate (VPA) is an effective antiepileptic drug used in neurology as well as in psychiatry. It is also associated with a hyperammonemic encephalopathy, when used in combination with other drugs.

Case report: We herein present a case of valproate-induced encephalopathy (VHE) in an epileptic patient who had subtherapeutic levels of valproate with subclinical hyperammonemia accompanied by triphasic waves on electroencephalography (EEG). We believe the hyperammonemia became clinically evident following diphenylhydantoin (DPH) loading.

Discussion: Hyperammonemia without clinical or laboratory evidence of hepatotoxicity is an important idiosyncratic side effect of VPA treatment. Clinical manifestations of VHE as well as EEG findings and hyperammonemia tend to normalize rapidly when VPA therapy is discontinued.

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Abstract – WCN 2013**No: 2005****Topic: 4 – Neuro-critical care****Vitamin D deficiency in neurointensive care: A retrospective observational study**

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Background: To date, vitamin D deficiency is frequently encountered in the general population. Critically ill patients have been found to be particularly susceptible to this condition and a poor vitamin D status is associated with higher morbidity and mortality. The prevalence of vitamin D status in neurointensive care is unknown.

Methods: In this retrospective analysis, we included 164 patients with available 25(OH)D serum levels hospitalized at the 8-bed neurointensive care unit (NICU) of the tertiary care center at the Medical University of Graz between 2008 and 2010.

Results: Patients were 61 ± 16 years old and mean SAPS2 was 25 ± 12 points. The main admission diagnoses included intracranial bleeding and stroke. Mean serum 25(OH)D level was 22.9 ± 12.3 ng/ml. Normal 25(OH)D levels (>30 ng/ml) were found in only 21.3% of all patients. 43.9% were vitamin D deficient (<20 ng/ml), while 34.8% were considered vitamin D insufficient (>20 and <30 ng/ml) by current definitions. 20.7% even had 25(OH)D levels below 12 ng/ml. All-cause hospital mortality was 10.4%. In patients with vitamin D deficiency, hospital mortality was significantly higher (15.3%, X² p = 0.03). However, ICU mortality and length of stay (LOS) in the NICU or hospital were not associated with vitamin D status.

Conclusion: A low 25(OH)D status is common in the NICU and is associated with higher hospital mortality. Future studies are needed to investigate if critically ill patients may benefit from vitamin D treatment.

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Abstract – WCN 2013**No: 1957****Topic: 4 – Neuro-critical care****Spontaneous spinal epidural hematoma after abrupt sneeze with prompt recovery of severe paraparesis without surgical treatment**

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Background: Spontaneous spinal epidural hematoma (SSEH) is a rare neurological condition which can lead to acute spinal cord compression.

Objectives: Symptoms of SSEH include sharp back pain and progression of muscle weakness. The diagnosis has to be done very quickly using magnetic resonance imaging (MRI). There is an indication for urgent surgical decompression, however, conservative therapy is supposed to be feasible in selective patients with early recovery. A relevant review of SSEH literature is included.

Methods: We present a 64-year-old female with history of hypertension and low dose of aspirin (100 mg daily) who had experienced severe low thoracic and back pain accompanied by severe paraparesis and urinary retention after abrupt sneeze.

Results: MRI revealed a posterior thoracic epidural hematoma extending from T6 to T11 vertebral level with spinal cord compression. Decompression was recommended but she refused surgery. Complete neurological recovery was observed within 24 h after SSEH onset. MRI one month later showed total absorption of hematoma.

Conclusion: Surgical decompression is still the main treatment option of SSEH, however, a conservative therapeutic approach with careful observation may be considered as a treatment of choice in some cases where surgery is refused (high risk or other reasons) and neurologic recovery is early and sustained.

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Abstract – WCN 2013

No: 1944

Topic: 4 – Neuro-critical care

Population pharmacokinetics of phenytoin in patients with traumatic brain injury

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Background: Phenytoin is commonly used to treat and prevent seizure after neurotrauma. However, many physiological changes occur in the neurotrauma patient, which alter the pharmacokinetics of drugs.

Objective: This prospective study investigated the population pharmacokinetics and the related factors that affect to pharmacokinetics of intravenous phenytoin in neurotrauma patients.

Patients and method: Patient data were collected from routine clinical visits between May 2012 and January 2013 at the neurotrauma wards of Sunprasithprasong Hospital, a hospital in the northeast of Thailand. Data from 122 patients aged between 12 and 79 years old were obtained in this study. Times of sampling were days 3–7 after starting with phenytoin, and about 3 serum samples/patients were taken. A 1-compartment model with nonlinear pharmacokinetics was fitted to the log-transformed concentration data using nonlinear mixed effect model (NONMEM). Gender, age, height, ABW, IBW, regimen, alcohol drinking, smoking, severity of head injury, depressed skull fracture, brain hemorrhage/hematoma, brain edema, brain contusion, midline shift, trauma organ involvement, brain surgery, fever, albumin level, LFT, and loading dose phenytoin before starting continuous regimen were 20 factors that were studied for the effect to pharmacokinetics of intravenous phenytoin in neurotrauma patients.

Results: The parameters of the pharmacokinetics base model were $K_m = 4.47$ mg/L, $V_{max} = 21.2$ mg/kg/day, and volume of distribution = 43.4 L. These finding actual body weight had affected on K_m , fever had affected on V_{max} , and regimen had affected on volume of distribution.

Conclusion: This study proposed population pharmacokinetics of phenytoin in patients with traumatic brain injury for appropriate dose adjustment.

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Abstract – WCN 2013

No: 1606

Topic: 4 – Neuro-critical care

The sedative effect of *Thymelaea lythroides* in Wistar rat

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Objective: Medicinal plants have played an essential role in the development of human culture. There are stimulating or calmative effects on the central nervous system. This study was performed to assess the sedative effect of the ethylic extract of *Thymelaea lythroides*

(*TI*) on Wistar rats. Prior to this we assessed the acute toxicity and determined the therapeutic doses of this extract.

Material and methods: The experiment carried out during this study is based on the gavage administration of *TI*'s ethylic extract with different doses (i.e., 200, 400, 500 and 5000 mg/kg). The sedative activity of the plant were observed and evaluated by the Hole Board and Open Field test. The results were analyzed using the ANOVA test followed by the Bonferroni test.

Results: The main results of this study showed that the lethal dose of the ethylic extract of *TI* ranged higher than the 5000 mg/kg dose. Additionally, compared to the control rats, the food and water intakes witness a significant decrease in the first week regarding the rats treated with the 5000 mg/kg dose, whereas the other doses showed no signs of toxicity. Furthermore, in comparison with the control rats, results of the psychotropic test conducted during this study showed significant decrease in the exploratory and locomotor activities of the rats treated with the different doses of ethylic extract. This decrease is similar to that of the reference molecule (Zepam).

Conclusion: These data showed that *T. lythroides* has a sedative effect.

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Abstract – WCN 2013

No: 1806

Topic: 4 – Neuro-critical care

Early Robot-Assisted Therapy in Patients with Stroke in Neurointensive Care Unit

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Background: Pulmonary embolism (PE) increases risk of unfavorable outcome and mortality in stroke.

Objective: To evaluate the impact of robot-assisted therapy in stroke patients on neurological outcome and rate of venous thromboembolism.

Patients and methods: This case-control study included 66 neuroICU patients (49 males, 17 females, median age 59.3) within 7 days from onset of ischemic and hemorrhagic stroke. We used NIHSS and GCS scores and assessed the rate of deep vein thrombosis (DVT) revealed with ultrasound scanning, the rate of PE and mortality from admission to Day 21.

Results: Patients were equally divided into two homogenous groups – Intervention and Control to receive standard stroke therapy plus daily robot-assisted arm and leg therapy (MOTomed letto 2) in Intervention group. Groups had similar stroke severity on admission (GCS: Me = 13 [LQ-10, UQ-15] vs. Me = 14 [LQ-10, UQ-15], $p = 0.11$; NIHSS: Me = 20 [LQ-16, UQ-29] vs. Me = 18 [LQ-15, UQ-27], $p = 0.5$ in Intervention and Control group, respectively). There was no significant difference in neurological outcome on Day 21 (GCS: Me = 15 [LQ-14, UQ-15] vs. Me = 15 [LQ-15, UQ-15], $p = 0.12$; NIHSS: Me = 11 [LQ-8, UQ-25] vs. Me = 15 [LQ-10, UQ-19], $p = 0.4$ in Intervention and Control group, respectively), in the rate of DVT and mortality on Day 21 (58% vs. 45%, $p = 0.147$; 39% vs 12%, $p = 0.058$, respectively). Rate of PE on Day 21 were higher in the Control vs. Intervention group (39% vs. 9%, $p = 0.029$).

Conclusion: Early robot-assisted therapy in severe stroke patients was associated with significant reduction of PE rate on Day 21, but did not influence neurological outcome and DVT rate.

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Abstract – WCN 2013**No: 718****Topic: 4 – Neuro-critical care****Amyloid- β in CSF predict neurological status and mortality after severe traumatic brain injury: Results of a pilot study**

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Objective: To assess amyloid- β_{1-42} (Ab42) concentrations and time-course in CSF and in plasma of patients with severe traumatic brain injury (TBI) and their relationship to injury characteristics, neurological status and clinical outcome.

Patients and methods: Twelve patients with severe TBI [Glasgow Coma Scale (GCS) ≤ 8] and 13 controls were included this pilot study. Paired CSF and plasma samples were taken from each TBI patient at admission and daily up to 7 days and Ab42 levels assessed by ultrasensitive digital ELISA. Data collected included demographic and clinical variables and survival 6 months post-injury.

Results: CSF Ab42 levels were significantly lower in TBI patients acutely after injury as compared to controls (median 105.9 vs. 537.6 pg/ml, $p < 0.0001$) with lower levels in patients who died than in those who were alive. Conversely, plasma Ab42 levels were significantly increased in TBI as compared to controls (median 17.02 vs. 7.289 pg/ml, $p < 0.0001$) with higher levels in patients who survived. A trend analysis using the Jonckheere–Terpstra test showed that both CSF and plasma Ab42 levels strongly correlated with mortality ($P < 0.0001$). CSF and plasma Ab42 concentrations within the first 24 h after injury did not correlate with TBI characteristics. A positive correlation between changes in CSF Ab42 concentrations and neurological status was identified, with Ab42 levels increasing as neurological status improved and decreasing when neurological status deteriorated.

Conclusion: These results suggest that determination of Ab42 may be valuable to obtain prognostic information in patients with severe TBI as well as for monitoring the response of the brain to injury.

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Abstract – WCN 2013**No: 1401****Topic: 4 – Neuro-critical care****Aneurysmal subarachnoid hemorrhage in stroke unit**

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Purpose: The aim of this study was to assess the features of meningeal syndrome (MS) in subarachnoid hemorrhage (SAH) patients depending on location of the cerebral aneurysm.

Materials and methods: We retrospectively analyzed clinical data from stroke unit of 25 SAH patients in age from 33 to 59 years old between January 2011 and March 2013. The relationship between symptoms severity, intracranial aneurysm location (by CT and angiography data) and Hunt–Hess scale grade was assessed.

Results: Aneurysm rupture as the cause of SAH was found in 71% of patients, in 29% the cause was unknown. The anterior

communicant artery location of ruptured intracranial aneurysm was found in 31%, internal carotid artery – 16%, anterior and middle cerebral arteries – 12% of each. Hunt–Hess scale scores were: 0 in 4%, 1 in 28%, 2 in 36%, 3 and 4 in 16% of each. The development of MS on the 1st day was found in 44%. MS time of developing was unknown in 66% of the patients due to later recourse in hospital. Nuchal rigidity was found in 96%. Symmetrical Kernig's sign was detected in 96%. Asymmetry of Kernig's sign and Brudzinski's sign was not found in all patients.

Conclusions: The location of ruptured aneurysm in most cases was detected in anterior communicant artery. Meningeal syndrome severity was not depended of aneurysm location. Mortality was in 20% of the patients due to cerebral edema and brainstem dislocation, 12% had subarachnoid hemorrhage rebleeding. 48% of all patients underwent cerebral aneurysm clipping.

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Abstract – WCN 2013**No: 1287****Topic: 4 – Neuro-critical care****Small-world characteristics of EEG patterns in post-anoxic encephalopathy**

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Background: Post-anoxic encephalopathy (PAE) has a varying outcome from death to good recovery. At present, it is possible to predict bad outcome using somatosensory evoked potentials in only a minority of the patients at an early stage. Network architecture, as can be quantified with continuous electroencephalography (cEEG) may serve as a candidate measure for predicting neurological outcome.

Objective: Using cEEG monitoring to detect the integrity of neural network architecture in PAE patients.

Material and methods: 19-Channel cEEG data was obtained from 56 PAE patients. Adjacency matrices of shared frequencies between 1 and 25 Hz of the EEG channels were obtained using Fourier transformations. Number of network nodes and connections, clustering coefficient (C), average path length (L) and small-world index (SWI) were derived. Outcome was quantified by the best Cerebral Performance Category (CPC)-score.

Results: Compared to non-survivors, survivors showed significantly more nodes and connections. L was significantly higher and C and SWI were significantly lower in the survivor group relative to the non-survivor group. The combination of number of nodes, connections, C and L showed the most significant difference and between survivors and non-survivors. At a specificity of 100% a sensitivity of 31% was present for predicting non-survival using this combination.

Conclusion: Our data shows that non-survivors have insufficient distribution and differentiation of neural activity for regaining normal brain function. These network differences, already present during hypothermia, might be used as an early prognostic marker. The predictive values, although already superior to current practice parameters, need to be further determined.

doi:10.1016/j.jns.2013.07.1091

Abstract – WCN 2013**No: 1274****Topic: 4 – Neuro-critical care****Early prognostic biomarkers in Guillain–Barre syndrome**

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Background: The clinical manifestations of Guillain–Barre syndrome (GBS) vary from mild neurologic deficiency up to tetraplegia with respiratory failure. The early correct prognosis of the clinical course might be helpful for the well-timed diagnostic of the life threatening disturbances.

Objective: To determine the prognostic value of biomarker neurofilament heavy chains (NfH), gliofibrillary acidic protein (GFAP) and tau heavy chains (hTau) in the clinical course of GBS.

Patients and methods: 35 patients, who fulfilled the diagnostic criteria of GBS, were included. The serum and cerebrospinal fluid (CSF) were collected before the beginning of therapy. CSF and serum NfH, GFAP and hTau levels were measured using a standard ELISA with analyst being blinded to all other data. All patients were divided into groups depending on symptoms of interest (respiratory failure, duration of ventilation, dysphagia) and their serum and CSF concentrations of biomarkers were compared. The statistic analysis was carried out using the Statistica 7.0, MedCalc 12.1.4.0.

Results: We determine the serum level of the NfH > 0.144 ng/mL which could predict the respiratory failure (AUC 0.804, $p < 0.0001$) and the serum level NfH > 0.094 ng/mL indicating the development of dysphagia (AUC 0.773, $p = 0.001$). We found the correlation between the duration of ventilation and serum ($R = 0.825$ $p = 0.003$) and CSF ($R = 0.820$ $p = 0.007$) GFAP levels. We also estimated prognostic hTau and GFAP levels in relation to prolonged ventilation.

Conclusion: Serum levels of NfH, GFAP and hTau can be prognostic markers in the clinical course of GBS.

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Abstract – WCN 2013**No: 750****Topic: 4 – Neuro-critical care****The experience of creating a mobile neurodiagnostic unit in a megalopolis (Moscow)**

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Background: An important factor in working out a comatose patient treatment plan is express neurophysiologic evaluation and assessment of the cerebral blood flow. It is evident that all the tests should be performed bedside and have high sensitivity and specificity.

Objective: To evaluate the performance of mobile neurodiagnostic unit in a megalopolis.

Material and methods: The special Mobile Neurodiagnostic Unit (MNU) was created in 1995. It is staffed with neurologists who can perform neurophysiology and neurosonology tests (EEG, TCD, EVP, duplex scan). The MNU, equipped with portable ultrasound scanner and encephalograph with EVP tool, works around the clock.

The MNU specialists go to Moscow hospitals to perform clinical examination and evaluate EEG, EVP and cerebral blood flow status for deep coma patients (GCS < 5). In case of suspected brain death (BD), the MNU experts take part in clinical consultations and perform ancillary tests.

Results: 4000 critically ill patients have been examined by experts of the MNU since 1995. Most patients suffered from stroke/SAH (68%), or TBI (28%). 3% and 1%, respectively, accounted for tumor and secondary brain damage. BD was confirmed more than 600 times. Since 1995, the average annual rate of BD diagnostics performed by the MNU has increased from 15 to 73 cases per year.

Conclusion: In a large megalopolis area, a highly professional mobile group able to make neurophysiological and cerebral blood flow assessment can improve the quality of neurocritical care without additional material costs, and increase the accuracy of BD diagnostics by using different confirmation tests.

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Abstract – WCN 2013**No: 999****Topic: 4 – Neuro-critical care****Fatal intracranial hemorrhage after anticoagulation in cerebral venous thrombosis**

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Background: The standard therapy for CVST is full dose of intravenous unfractionated heparin or subcutaneous low-molecular-weight heparin.

The rationale of anticoagulation therapy in CVST is to prevent thrombus growth, to facilitate recanalization, and to prevent deep venous thrombosis or pulmonary embolism. Controversy has ensued because cerebral infarction with hemorrhagic transformation or intra-cerebral hemorrhage is commonly present at the time of diagnosis of cerebral venous and sinus thrombosis, and it may also complicate treatment.

Presentation case: We present here a 52 year old male patient who reported sudden onset of nausea, vomiting, and severe headache, followed by tonic–clonic generalized seizures. On admission, physical examination was normal. Cerebral MRI revealed a left transverse sinus thrombosis associated with superior sagittal sinus thrombosis. With a diagnosis of CVST and following current recommendations for CVST treatment, LMWH (0.1 ml/kg/12 h) was administered. However, 24 h after heparin initiation, neurological deterioration was noted. The Cerebral Computed Tomography Scan showed a large left temporo-parietal hematoma with edema, mass effect on the ventricular system, and temporal pre-herniation. The anticoagulant treatment was stopped. A decompressive craniectomy with evacuation of the hematoma was performed. Control Cerebral Computed Tomography Scan revealed new infarct in temporal lobe, bilateral thalami and midbrain.

Conclusion: The results of some studies have been controversial, heparin remains the first line treatment option for CVST because it is probably safer and generally yields favorable trends of clinical outcomes shown in previous trials, but the risk of new bleeding is possible, as is illustrated by our fatal case.

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Abstract – WCN 2013**No: 901****Topic: 4 – Neuro-critical care****Isolated hypoglossal nerve palsy: A harbinger of serious underlying disease**

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Hypoglossal nerve palsy is not an uncommon finding in neurological diseases. Vascular, inflammatory, traumatic or space-occupying lesions

can affect hypoglossal nerve anywhere in its course from the nucleus in the caudal medulla, through the intracranial extramedullary portion of the nerve and hypoglossal canal, and finally into its extracranial termination.

Case 1: A 72-year-old woman was referred to the neurology department due to a sudden onset dysarthria. She had right breast cancer in 2003 and underwent mastectomy, local radiotherapy and adjuvant chemotherapy for regional metastasis. On neurological examination, her tongue was atrophy and deviated to the left side. The ocular fundi and other cranial nerves were normal. There were no long tract signs or signs of meningeal irritation. Brain MRI showed a subtle enhancing lesion involving the left occipital condyle with the destruction of the petrous apex and clivus.

Case 2: A 54-year-old woman with a history of headache for 2 months was admitted due to a sudden onset dysarthria. Her headache was located in the left temporal area and refractory to the medical treatment. Neurological examination showed atrophy with deviation of the tongue to the left. Otherwise were noncontributory. Brain MRI revealed an enhancing lesion involving the occipital condyle and clivus. The biopsy of regional enlarged lymph node showed the poorly differentiated metastatic squamous cell carcinoma.

Conclusion: We report two cases of isolated hypoglossal nerve palsy. Isolated hypoglossal nerve palsy is a rare finding but paralysis of this nerve may be the first sign of a serious underlying disease.

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Abstract – WCN 2013

No: 555

Topic: 4 – Neuro-critical care

Clomethiazole-induced hepatotoxicity as example of successful treatment with cooperation between clinical pharmacist and physician

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Background: Clomethiazole is being structurally related to thiamine and has been predominantly used for management of agitation, alcohol delirium and withdrawal and in rare cases for restlessness. There is little reported in literature regarding acute clomethiazole-induced hepatotoxicity.

Objective: A case of acute hepatotoxicity that occurred in a 71 year-old male patient with psycho-organic delirium who began clomethiazole therapy is described.

Patient and methods: A case report. The diagnostic and treatment procedures were reviewed. Clinical pharmacist was also introduced.

Results: Laboratory tests excluded other causes of acute liver injury. Liver enzymes returned to normal within one week of clomethiazole discontinuation. From the ratio S-AST/S-ALT it is possible to conclude that hepatocytes damage already occurred and recovered soon after clomethiazole discontinuation. To control symptoms of delirium rapid switching from clomethiazole to zuclopenthixol was advised by clinical pharmacist, therefore zuclopenthixol in small doses could be used in patient with clomethiazole hepatotoxicity.

Conclusion: Clomethiazole should be included among the causes of drug-induced hepatotoxicity. Open communication between the physician and clinical pharmacist on the patient's pharmacotherapy is necessary to achieve optimal clinical outcomes.

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Abstract – WCN 2013

No: 371

Topic: 4 – Neuro-critical care

Predictors of functional outcome of patients with Guillain-Barré syndrome in intensive care unit

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Background: Approximately one third of patients with GBS need to be admitted to the ICU because of respiratory failure. They are at risk for systemic complications with substantial morbidity and consequent mortality. There were only limited data about the morbidity of ICU GBS patients and its impact on functional outcome.

Objective: To analyze predictors for the functional outcome in consecutive GBS patients admitted to ICU.

Methods: Consecutive adult (≥ 15 years old) GBS patients admitted to the ICU during 11-year period from Jan 2000 through Dec 2010 were included. Using mRS (modified Rankin Scale) at 6 months, we divided and analyzed patients into favorable (mRS 0–2) and unfavorable (mRS 3–6) outcome groups.

Results: Of 43 patients, there were significant differences in age ($P = 0.007$), ICU stay ($P = 0.01$), intubation ($P = 0.003$), mechanical ventilation ($P = 0.0001$), tracheostomy ($P = 0.014$), comorbid conditions such as congestive heart failure ($P = 0.013$), complications such as acute renal failure ($P = 0.034$), abnormal liver function test ($P = 0.017$), cardiac complication ($P = 0.026$), pneumonia ($P = 0.014$), and sepsis ($P = 0.026$), a high score in the TISS-28 ($P = 0.0001$), and a high APACHE II score ($P = 0.007$) between favorable (25; 58.1%) and unfavorable outcome (18; 41.9%) groups. The mortality rate was 11.6% ($n = 5$). Main causes of mortality were sepsis and cardiogenic shock. In multivariable logistic regression analysis, TISS-28 (OR 2.55, 95% CI 1.06–6.10, $P = 0.036$) was independent predictor of unfavorable outcome.

Conclusion: The predictors for functional outcomes such as TISS-28 in GBS patients admitted to the ICU could help identify correctable factors, optimize medical treatment, and guide discussions of prognosis.

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Abstract – WCN 2013

No: 209

Topic: 4 – Neuro-critical care

Minocycline cannot protect neurons against bilirubin-induced hyperexcitation in the ventral cochlear nucleus

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Excitotoxicity has been suggested to play an important role in many central nervous system diseases, particularly in bilirubin encephalopathy. Minocycline treatment has been proposed to be one of the most promising potential therapies for excitotoxicity-induced neurological disorders. However, some key questions, such as the electrophysiological effect of minocycline on neuronal excitability and hyperexcitation in pathological conditions, require clarification. In this study, using patch-clamp techniques, we showed that bilirubin increased the frequency of both spontaneous excitatory postsynaptic currents (sEPSCs) and neuronal firing in isolated ventral cochlear nucleus (VCN) neurons at postnatal days 11–14 (P11–14) in rats but it did not affect the amplitude of sEPSCs or glutamate-activated (I_{Glu}) currents. However, minocycline had no effect on sEPSC frequency or I_{Glu} amplitude. Furthermore, minocycline pretreatment did not abolish bilirubin-induced sEPSC potentiation or neuron firing. These data

suggest that minocycline does not affect excitatory synaptic transmission or hyperexcitation induced by bilirubin in VCN neurons. From these results, we propose that the neuroprotective efficacy of minocycline, if it can protect neurons against neurotoxicity induced by substances like bilirubin, is mediated by either an alternative mechanism or downstream events post neuronal hyperexcitation. Certainly, additional investigation of the neuroprotective effects of minocycline is required before embarking on further clinical trials.

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Abstract – WCN 2013

No: 152

Topic: 4 – Neuro-critical care

A new percutaneous model of subarachnoid haemorrhage in rats

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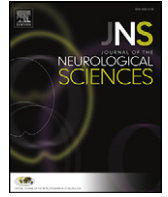
Objective: Describe the results obtained with a new percutaneous, intracisternal model of subarachnoid haemorrhage (SAH) in Wistar rats by a single injection of non-heparinised, autologous blood.

Methods: Once anaesthetized the rat was fixed prone in a stereotaxic frame. After identifying the projection of the occipital bone, the needle of the stereotaxic frame aspirated towards the foramen magnum until it punctured through the atlanto-occipital membrane and obtained cerebrospinal fluid. Autologous blood (100 µl) was withdrawn from the tail and injected intracisternally. This procedure was repeated in the sham group, injecting 100 µl of isotonic saline. On the fifth day post-intervention, the rats were anaesthetized and the brain was exposed. After a lethal injection of ketamine the brain was explanted and fixed in paraformaldehyde. Gross and microscopic inspection of the slices revealed the existence or non-existence of pathological findings.

Results: A total of 26 rats were operated on (13 in the SAH group/13 in the sham group). The average time between obtaining the blood and the start of the intracisternal injection was 10 (±1.2) seconds. The mortality rate was 16.12%. Intra- and extraparenchymal ischemic-haemorrhagic lesions were found in three animals (23.07%) – all from the SAH group – with ischemic neuronal cell injury detected in two of the three.

Conclusions: The new rodent model of SAH is easy to perform, with low mortality, minimally invasive, which makes it interesting for future studies on vasospasm-related delayed SAH complications.

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Topic: 5 - Dementia

Abstract – WCN 2013

No: 168

Topic: 5 – Dementia

Addenbrooke's Cognitive Examination standardized verbal fluency scores for differential diagnosis of AD and FTLD

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Background: The Addenbrooke's Cognitive Examination (ACE) is a commonly used cognitive screening instrument for the diagnosis of dementia.

Objective: To examine the differential impairment of ACE Standardized Verbal Fluency (SVF) scores for category fluency (CF) and letter fluency (LF) and measure their diagnostic utility for the differentiation of Alzheimer's disease (AD) and frontotemporal lobar degenerations (FTLD).

Patients and methods: ACE was administered to consecutive new outpatients attending a Cognitive Function Clinic.

Results: Of 285 patients administered ACE, final diagnoses included 114 AD patients and 16 FTLD patients. Examining scaled ACE SVF scores, LF > CF was found to have modest sensitivity (0.66) but poor specificity (0.44) for the diagnosis of AD. LF < CF was found to have excellent specificity (0.86) but poor sensitivity (0.25) for the diagnosis of FTLD. These figures were compared unfavourably with the ACE VLOM ratio for the same differential diagnosis: VLOM of >3.2 for diagnosis of AD had sensitivity of 0.76 and specificity of 0.76; VLOM ratio of <2.2 for diagnosis of FTLD had sensitivity of 0.31 and specificity of 0.90. The comparison of area under the receiver operating characteristic curves also favoured VLOM over SVF scores (0.80 vs 0.56).

Conclusion: Although verbal fluency has been described as the "ESR of cognition" and VF tasks are very sensitive in detecting dementia, ACE SVF scores showed no diagnostic advantage over VLOM ratios for the differential diagnosis of AD and FTLD in this patient cohort.

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Abstract – WCN 2013

No: 169

Topic: 5 – Dementia

Applause sign: Diagnostic utility in a cognitive function clinic

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Background: The applause sign (signe d'applause) has been observed in parkinsonian syndromes and cortical dementias.

Objective: To report the utility of the applause sign for diagnosis of cognitive impairment.

Patients and methods: The three-clap test was administered to consecutive new outpatients attending a Cognitive Function Clinic over a 5-month period (September 2012–January 2013).

Results: In 100 consecutive patients (M:F = 63:37; age range 20–88 years, median 59.5 years), 37 were demented by DSM-IV criteria and 20 had MCI by Petersen criteria. Nineteen had the applause sign, of whom 9 had a dementia syndrome and 6 had MCI. Of patients with a synucleinopathy (PD-MCI, PDD, DLB; n = 9), 5 (=55%) had the applause sign, which was also seen in patients with AD, alcohol-related dementia, and in 4 subjective memory complainers. Applause sign had poor sensitivity for the diagnosis of dementia (0.24) or cognitive impairment (0.26), but better specificity for these diagnoses (0.84 and 0.91 respectively).

Conclusion: In a prospective observational study of consecutive new cognitive clinic outpatients, the applause sign was not sensitive for a dementia or MCI diagnosis, but it was specific (i.e. its absence effectively rules out dementia or MCI). As in previous studies, the applause sign was not found to be specific to a particular disease.

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Abstract – WCN 2013

No: 210

Topic: 5 – Dementia

Age-associated changes of rat brain glutamate synthetase in rat brain Astroglial cells

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Glutamate (Glu) is known as an excitatory amino acid neurotransmitter which interacts with Glu receptors for basal excitatory synaptic transmission. Excessive levels of extracellular Glu in the brain are excitotoxic and could lead to neurotransmitter imbalance, neuronal death and several neurodegenerative processes. Astroglial cells remove extracellular Glu and convert glutamine by astroglial-specific glutamine synthetase (GS) enzyme. Several line of evidence indicated that aging induces many disturbances in the regulation of astroglial glutamate metabolism. Because age-related changes in the metabolism of DNA could lead to alter the brain protein synthesis, in this study the rate of DNA strand breaks, cytoskeletal glial fibrillary acidic protein (GFAP) and GS contents in astroglial cells isolated from rat brain were investigated. Three and 30 month-old Wistar rats were used. Astroglial cells were purified and morphological characteristics of the cells were confirmed and the rate of DNA strand breaks, GFAP and GS contents of the cell preparations were determined. The rate of DNA strand breaks was higher in cell preparation from aged rats, whereas the levels of GS on a protein basis were relatively lower in old animals as compared to that of young rats. However, GEAP content of the cells does not show a

significant change in aged rat brain, though there is indication of lower values compared with the young rat brain. Since, GS is the main enzyme of glutamate metabolism, it is concluded that GS might be considered as a therapeutic target for prevention of Dementia and Alzheimer's disease.

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Abstract – WCN 2013

No: 213

Topic: 5 – Dementia

The correlation between cognitive impairment and depression in patients on maintenance dialysis

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Patients with chronic kidney disease frequently show cognitive dysfunction. The association of depression and cognitive function is not well known in dialysis patients. We evaluated cognitive impairment, depression and their relationship between methods of dialysis, hemodialysis (HD) and peritoneal dialysis (PD). Fifty-six dialysis patients were recruited and their clinical and laboratory data were collected. The Korean version of Mini-Mental State Exam (K-MMSE) was applied to screen cognitive function of patients and the Korean version of Beck Depression Inventory (K-BDI) was used for detecting depression. The average age of participants was 54.2 ± 10.2 years and 29 (51.8%) were female. The average dialysis vintage was 4.2 ± 3.8 years. PD group showed significantly higher K-MMSE score (27.8 ± 2.9 vs. 26.1 ± 3.1 , $p = 0.010$) and lower K-BDI score (12.0 ± 8.4 vs. 20.2 ± 10.4 , $p = 0.003$) compared with HD group. The percentage of depression was higher in HD group (51.7% vs. 18.5%). There was a negative correlation between cognitive function and depression in both groups. Depression and education level were the independent predictors for cognitive impairment in multivariate analysis. The cognitive impairment was closely correlated with depression. It is important to detect cognitive impairment and depression early in dialysis patient with simple bedside screening tools.

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Abstract – WCN 2013

No: 279

Topic: 5 – Dementia

Dementia, delirium and aggressive behaviour (AB) among hospitalised elderly

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Background: Aggressive behaviour (AB) is common among hospitalised patients. Most available literature is focused on younger patients¹. We investigated the association of dementia and delirium with AB in the inpatient setting among a cohort of elderly subjects and the outcome of their index admission.

Method: We carried out a retrospective, observational study between January 2010 and January 2012 using a standardized electronic reporting system in a tertiary acute hospital in sub urban Australia.

Results: 145 episodes of AB among elderly (N = 93) accounting for 23.0% of all reported AB were identified. (4.9/1000 admissions, age 81.9 ± 6.3 years, male 61.5%). Delirium (92.3%) and dementia (60.4%) were strongly associated. Antipsychotics (66.6%), Benzodiazepines (9.6%) and a combination of the two (12.9%) were commonly used to manage AB. 20% had to be physically restrained. In hospital mortality rate was 10.3%, better than most reported rates for delirium². Residential care placement (39%) and the mean length of stay (15.5 ± 11.2 days) were comparable to reported outcomes of delirium².

Conclusions: AB in elderly inpatients is strongly associated with delirium and dementia. Frequently reported associations¹ among younger patients are not common in the elderly counterparts. There is a window of opportunity to act when the majority of these patients demonstrate milder AB prior to its escalation. Elderly hospitalized patients demonstrating AB have unfavourable outcomes but our institution had lower hospital mortality compared to most reported outcomes of delirium². The frequency of residential care placement and length of stay was comparable to reported outcomes of delirium².

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Abstract – WCN 2013

No: 267

Topic: 5 – Dementia

Dietary patterns and risk of dementia in an elderly Japanese population: The hisayama study

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Background: There are no previous reports that assessed the association between dietary patterns and the risk of dementia in Asian populations.

Objective: We investigated dietary patterns and their potential association with the risk of incident dementia in a general Japanese population.

Methods: A total of 1006 community-dwelling Japanese subjects without dementia, aged 60–79, were followed up for 15 years (median). The reduced rank regression procedure was used to efficiently determine their dietary patterns. The estimated risk conferred by a particular dietary pattern on the development of dementia was computed using a Cox proportional hazards model.

Results: Seven dietary patterns were extracted, of which dietary pattern 1 was correlated with high intakes of soybeans and soybean products, vegetables, algae, and milk and dairy products, and low intake of rice. During the follow-up, 271 subjects developed all-cause dementia. Among them, 144 had Alzheimer's disease (AD) and 88 had vascular dementia (VaD). After adjusting for potential confounders, the risks for the development of all-cause dementia, AD and VaD, were reduced by 0.66 (95%CI 0.46–0.95), 0.65 (95% CI 0.40–1.06), and 0.45 (95% CI 0.22–0.91) among subjects in the highest quartile of score for dietary pattern 1 as compared to those in the lowest quartile.

Conclusion: Our findings suggest that higher adherence to a dietary pattern was characterized by a high intake of soybeans and soybean products, vegetables, algae, and milk and dairy products, and a low intake of rice is associated with a reduced risk of dementia in the general Japanese population.

doi:10.1016/j.jns.2013.07.1106

Abstract – WCN 2013**No: 270****Topic: 5 – Dementia****Neurocognitive impairment (NCI) in HIV-1 infected adults in Sub-Saharan Africa: A systematic review and meta-analysis**

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Background: In SSA estimates of HIV related neurocognitive impairment (NCI) and the effect of ART on it have varied. A study was conducted to obtain the prevalence, burden and impact of ART on NCI.

Materials and methods: We searched Medline and other databases for relevant English language publications up to June 2012. Prospective studies in adults from SSA reporting HIV status, utilization of ART and presence of NCI using the IHDS were selected. Study quality was assessed using standard criteria. Meta-analysis used random-effects model to derive estimates and meta-regression was done to assess the effects of age, gender, education and CD4 cell counts on NCI. Publication bias and sensitivity analyses were conducted.

Results: 16 studies from 7 countries in SSA were included. The frequency of NCI pre-ART was 42.37% (95% CI = 32.18–52.56%) and 30.39% (95% CI = 13.17–47.61%) among those on ART for ≥6 months. We found no significant associations between age, gender, CD4 cell counts and years of education with NCI. The patients on ART were less likely to have NCI compared to HIV infected pre-ART patients with OR of 0.36 (95% CI = 0.19–0.69). In longitudinal studies with same patients followed before and ≥6 months after ART, the OR of NCI after ART compared to pre-ART was 0.23 (95% CI = 0.14–0.37). The combined burden of NCI among pre-ART and on-ART patients was estimated at 8,121,910 (95% CI = 5,772,140–10,471,680).

Conclusions: HIV strongly predisposes to NCI leading to a huge burden in SSA. Scale-up of ART can substantially reduce NCI in the region.

doi:10.1016/j.jns.2013.07.1107

Abstract – WCN 2013**No: 254****Topic: 5 – Dementia****Complementary and alternative medicine use among patients with cognitive impairment in Singapore**

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Introduction: Complementary and Alternative Medicine (CAM) is defined as medical interventions and therapies that are not taught widely in medical schools or available in hospitals (Eisenberg et al., 1993). Examples of CAM include Acupuncture and Yoga. Information of CAM use among patients is crucial for healthcare professionals to play an active role in their patients' treatment plans. However, there is limited information on CAM usage among patients with cognitive impairment in Singapore. Hence, this study is aimed to determine the prevalence and patterns of CAM use. In addition, relationship between CAM use and demographic characteristics, and Health Locus of Control (HLC) were also investigated.

Methodology: Fifty caregivers of patients with cognitive impairment were recruited using the purposive sampling method, from Neurology outpatient clinic in Singapore General Hospital (SGH) from November 2012 to January 2013.

Materials: An 11-item survey questionnaire, which collected information about participant's CAM usage and demographic information, was used in the study. The caregiver's Multidimensional Health Locus of

Control Scale (MHLC), adapted from Wallston, Stein, & Smith's (1994) MHLC scale – Form C, was also used in this study to assess the participant's beliefs about their relative's health condition.

Results & summary: 42% of the participants reported using CAM, of which 57.1% did not inform their doctors about their relative's CAM usage. The study found that majority of the users obtained information of CAM from low-credible sources such as family, friends and internet. The present study also found that female caregivers were more likely to use CAM for their relative's health condition.

doi:10.1016/j.jns.2013.07.1108

Abstract – WCN 2013**No: 256****Topic: 5 – Dementia****The effect of an APOE polymorphism on cognitive function depends on age**

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Objectives: APOE E4 polymorphism is a genetic susceptibility marker of Alzheimer's disease. However, it remains controversial whether this polymorphism is related to cognitive function in general population. We aimed to evaluate a cross-sectional association between the APOE E4 genotype and cognitive function, and whether this association may differ by age.

Methods: Cognitive function was assessed using the Korean version of modified Mini-Mental State Examination (K-mMMSE) in 10,371 Koreans aged 45 to 74 yrs in Namwon City. According to the APOE E4 status, all participants were classified as non-carriers, heterozygotes, or homozygotes. Participants were also categorized by age; 45–54, 55–64, and 65–74 years. **Cognitive impairment was defined as scoring below the 25th percentile according to age, sex, and education.** Multiple linear and logistic regression models were used to evaluate the association between APOE genotypes and cognition.

Results: The frequency of APOE genotypes in the study population was 0.4, 10.1, 1.1, 72.9, 14.7 and 0.8% for E2E2, E2E3, E2E4, E3E3, E3E4, and E4E4, respectively. Compared to the APOE E4 non-carriers, the heterozygotes and homozygotes showed 1.3% and 7.3% lower K-mMMSE scores at 65–74 yrs and 0.8% and 4.6% higher scores at 45–55 yrs, respectively. The E4 homozygotes had a higher risk for cognitive impairment (OR = 2.38, 95% CI = 1.13–4.98) at 65–74 yrs compared to the non-carriers, but showed lower risk (0.19, 0.03–1.48) at 45–54 yrs.

Conclusions: The present study demonstrates that the effect of APOE E4 on cognitive function depends on age, with positive correlation until late adulthood and negative correlation in the elderly.

doi:10.1016/j.jns.2013.07.1109

Abstract – WCN 2013**No: 237****Topic: 5 – Dementia****A randomized placebo-controlled trial of quetiapine for the treatment of behavioral and psychological symptom of dementia in Alzheimer's disease patients**

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Objectives: This randomized, double-blind, placebo-controlled trial examined the efficacy and safety of quetiapine in the treatment of

aggression, agitation, and psychosis in elderly nursing-home patients with dementia.

Methods: We conducted a 12-week, rater-blinded, randomised study of 80 outpatients (72–85 years) with behavioral and psychological symptom of dementia(BPSD) (assessed by NPI baseline score), who received low dose quetiapine (12.5–50 mg/day). Primary efficacy measures were: Neuropsychiatric Inventory (NPI); secondary efficacy measures: Clinical Global Impression (CGI), Cohen–Mansfield Agitation Inventory (CMAI), and Mini-Mental State Examination (MMSE). Safety evaluations included the incidence of extrapyramidal symptoms (EPS) and adverse events (AEs).

Results: Seventy-six of 80 patients were evaluable for efficacy (80 were evaluated for safety), while 4 patients discontinued (4 lost to follow-up). Seventy-six patients received quetiapine (mean dose 29 mg/day). NPI scores decreased significantly from baseline to week 12 ($P < \text{or} = 0.05$ vs. baseline). Most patients (72.5%) experienced clinical improvement (CGI-Improvement scores); quetiapine reduced agitation (CMAI scores); and there was no cognitive impairment (MMSE scores). There were no significant EPS. There were no cerebrovascular AEs or deaths.

Conclusions: Treatment with low-dose(mean dose 29 mg/day) quetiapine resulted in significant improvement in aggression, agitation, and psychosis associated with dementia and it was generally well tolerated (including no cognitive impairment) in the treatment of BPSD in elderly patients.

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Abstract — WCN 2013

No: 217

Topic: 5 — Dementia

Iatrogenic cognitive impairment in elderly patient in Russia

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Introduction: Most iatrogenic cognitive impairment (ICI) can be explained and can be attributed to polypharmacy in elderly, drug interactions, and altered pharmacokinetics.

Objective: To evaluate iatrogenic effects of drugs in elderly patients.

Material and Methods: The treatment of 180 patients aged >65 years who sought medical care in FSBI of Internal Medicine with complaints of cognitive impairment (CI) with the diagnosis of ICI in 2011–2012 was analyzed. All patients who received at least 6–8 drugs per day prescribed by the doctor about the current somatic pathology during long-term period (more than 2 month) were included in the analysis.

Results: It was found that the most adverse effects on cognitive function caused by benzodiazepine derivatives were use to improve sleep and sedation — in 34%, the same effect for the neuroleptic group with 4.1%. Antidepressants and antiepileptic drugs which were used in order to reduce pain caused ICI in 12.6% of cases. Clinical picture was almost identical to Alzheimer's disease and after cessation of treatment ICI persisted for 2–3 months. There was a growth of CI in patients receiving drugs for internal, ophthalmic and neurological disease (CHD, diabetes, constipation, glaucoma, ulcers, Parkinsonism, vertigo, others) even with minimal anticholinergic activity — in 49.3% of patients. An effect similar to the action of atropine was typical for prednisolone, digoxin, nifedipine, and dipyridamole in few cases.

Conclusions: Our findings show that mental confusion was the most frequent symptom of ICI. The decrease in the severity of CI after withdrawal of the drug indicates ICI. General practitioners should consider the possible iatrogenic effects in the treatment of elderly patients.

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Abstract — WCN 2013

No: 170

Topic: 5 — Dementia

Subjective memory complaints: Is family history of dementia a risk factor?

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Background: Ascertaining family history of dementia is a routine part of cognitive assessment.

Objective: To measure the frequency of cognitive impairment (dementia or mild cognitive impairment) in patients with or without a positive family history of dementia.

Patients and methods: Family history of dementia was ascertained in consecutive new outpatients attending a Cognitive Function Clinic.

Results: Of 139 patients, 43 reported a positive family history of dementia but only four satisfied criteria for autosomal dominant disease (3 or more affected family members in at least two generations). Of 63 patients with dementia or MCI, 14 (22.2%) had a positive family history, all familial disease (at least one first degree family relative affected). Of 76 patients with subjective memory complaints (neither dementia nor MCI), 29 (38.2%) had a positive family history (either autosomal dominant disease or familial disease). Risk ratio for subjective memory complaint patients having a positive family history of dementia was 1.72 (95% confidence interval = 1.00–2.96). The null hypothesis that the proportion of patients with a positive family history of dementia was the same in cognitively impaired and non-impaired groups was not rejected, although a trend was observed ($\chi^2 = 3.41$, $df = 1$; $0.1 > p > 0.05$).

Conclusion: Positive family history of dementia may be one stimulus for patient concerns about memory, leading to consultation and onward referral of individuals with subjective memory impairment. These individuals may be sensitized to the symptoms of memory impairment, including memory lapses, by their family history.

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Abstract — WCN 2013

No: 220

Topic: 5 — Dementia

Effects of gender on two clinical signs (attended alone and head turning) of use in the diagnosis of cognitive complaints

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Background: Previous studies have shown that the attended alone sign is very sensitive for the absence of cognitive impairment, whilst the head turning sign is very specific for the presence of cognitive impairment.

Objective: To report the effect of gender on the attended alone sign and the head turning sign in diagnosis of cognitive complaints.

Patients and methods: Data from studies of consecutive new outpatients attending a Cognitive Function Clinic were analysed by patient gender.

Results: In a study of the attended alone sign in 726 consecutive patients (M:F = 383:343), the null hypothesis that the proportion of patients attending alone did not differ significantly by gender was not rejected ($\chi^2 = 0.02$, $df = 1$, $p > 0.1$). Diagnostic accuracy data by gender showed similar sensitivity and specificity in males (0.91 and 0.49) and females (0.96 and 0.40). In a study of the head turning sign in 246 consecutive patients who attended with an informant (M:F = 123:123), the null hypothesis that the proportion of patients with the head turning sign did not differ significantly by gender was not rejected although a trend was observed ($\chi^2 = 3.26$, $df = 1$, $0.1 > p > 0.05$). Looking at the diagnostic accuracy data by gender, test sensitivity and specificity were better in females (0.72, 0.98) than in males (0.58, 0.91).

Conclusion: In prospective observational studies, gender does not seem to influence the attended alone sign. The head turning sign may possibly be of greater diagnostic utility in female patients.

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Abstract – WCN 2013

No: 120

Topic: 5 – Dementia

Analysis of polymorphism in Brain-derived neurotrophic factor gene in sporadic Alzheimer's disease in Han Chinese

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Objective: To investigate the association between the C270T polymorphism of the Brain-derived neurotrophic factor (BDNF) gene and sporadic Alzheimer's disease (SAD) in Han Chinese.

Methods: The polymerase chain reaction-restriction fragment length polymorphism (PCR-RELP) was used to detect BDNF genotypes of 55 SAD patients and 80 controls.

Results: Gender or age had no significant difference in SAD and control group ($\chi^2 = 0.072$, $p = 0.789$; $t = 1.494$, $p = 0.137$, respectively). The genotype distribution for the control group was not significantly deviated from the Hardy-Weinberg equilibrium ($\chi^2 = 0.167$, $p = 0.682$), and the frequency distributions of the two groups' genotype and allele were not significantly different ($\chi^2 = 0.219$, $p = 0.640$; $\chi^2 = 0.211$, $p = 0.646$, respectively).

Conclusion: There is no association between the C270T polymorphism of BDNF gene and SAD in Han Chinese.

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Abstract – WCN 2013

No: 111

Topic: 5 – Dementia

Behavioural Assessment of the Dysexecutive Syndrome (BADS) on the context of normal and pathological aging

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Several authors advocate the need to assess executive functioning (EF) as a robust determinant of the functional status of the subject and as a potential marker of dementia. However there are few available instruments that allow the assessment of different components of the EF in the context of aging, both normal and pathological. The present work aimed to determine the psychometric properties of the Behavioural Assessment of the Dysexecutive Syndrome (BADS) in the context of aging in a Portuguese sample.

Our sample comprised 33 subjects with more than 65 years divided in two groups: Control Group ($n = 22$), made of cognitively intact subjects; Dementia Group ($n = 11$), composed of patients with probable diagnosis of Alzheimer's disease (NINCDS-ARDA Criteria). We have applied the BADS, the Dementia Rating Scale and the Geriatric Depression Scale to both groups.

The BADS revealed good levels of acceptance, internal consistency, discriminative and criterion validity. These results favor the use of this instrument in the context of aging and dementia. It may be very useful in monitoring EF and in the detection of dementia.

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Abstract – WCN 2013

No: 93

Topic: 5 – Dementia

Posterior Cortical Atrophy – a possible cause for visual complaints

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Background: Posterior Cortical Atrophy (PCA) is a rare subtype of Alzheimer's dementia accounting for 5% of its cases. It refers to a clinical syndrome in which complex visual processing is progressively disrupted due to a cognitive neurodegenerative disorder, clinically distinct from typical Alzheimer's, but sharing similar neuropathological features. It involves the posterior parts of brain used in visual and gestural activities, with variable signs and symptoms observed in Gerstmann and Balint syndromes. Language, memory and insight remain relatively preserved until late in the course.

Clinical case: A 71-year-old woman was referred to our outpatient clinic after 4 years of unspecific visual complaints affecting predominantly the left visual field. She was followed and investigated during 2 years and no primary relevant ophthalmological pathology was found. A neurological consultation was required and several cognitive defects were detected: left hemineglect, prosopagnosia, simultanagnosia, finger agnosia, and mild left-right disorientation. She also had agraphia and alexia, without verbal language dysfunction. Her episodic memory was mildly impaired. The brain MRI revealed bilateral parieto-temporal-occipital cortical atrophy predominantly affecting the right hemisphere.

Conclusion: It is relevant to recognize the PCA as a type of dementia that usually presents first to the ophthalmologist due to its visual impairments. Since visual complaints can occur following any lesion affecting the visual pathways from the retina to the occipital lobe or adjacent processing visual cortex, a full neuropsychological examination should be performed whenever there is doubt about the significance of the visual impairment, especially when no obvious ophthalmological pathology is found.

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Abstract – WCN 2013

No: 73

Topic: 5 – Dementia

The diagnostic utility of cerebrospinal fluid alpha-synuclein analysis in dementia with Lewy bodies – a systematic review and meta-analysis

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Objectives: Dementia with Lewy Bodies (DLB) is difficult to distinguish from other dementias due to heterogeneity in clinical presentation and phenotypic overlap. Our aim was to investigate the diagnostic utility of cerebrospinal fluid (CSF) alpha-synuclein analysis in differentiating between DLB and other dementias.

Methods: We systematically searched MEDLINE, EMBASE, PsychINFO and Web of Knowledge up to May 2012. Studies were included if they utilized reproducible quantification methods of CSF alpha-synuclein levels and included patients from a representative spectrum, classified using international consensus criteria. Random effects model was used

to calculate weighted mean difference (WMD) and 95% confidence intervals between DLB and other groups.

Results: A total of 11 studies, comprising 2019 patients were included. 7 studies were evaluated in a meta-analysis. Mean CSF alpha-synuclein concentration was lower in DLB patients compared to those with Alzheimer's disease (AD) [WMD -0.24 ; 95% CI, -0.44 , -0.03 ; $p = 0.02$]. No significant difference was identified between DLB patients compared to those with Parkinson's disease [WMD 0.05 ; 95% CI, -0.17 , 0.28 ; $p = 0.65$] or other neurodegenerative conditions. Mean CSF alpha-synuclein levels were significantly lower in DLB patients compared to the non-synucleinopathies [WMD -0.25 ; 95% CI, -0.44 , -0.06 ; $p = 0.01$]. No significant difference was found between DLB and synucleinopathies [WMD 0.05 ; 95% CI, -0.17 , 0.27 , $p = 0.65$].

Conclusion: CSF alpha-synuclein may be of diagnostic use in differentiating between DLB and AD. However, no definite conclusion can be made due to assay heterogeneity, poor reporting standards and study design. We emphasize the need for adherence to reporting protocols in future studies.

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Abstract — WCN 2013

No: 74

Topic: 5 — Dementia

Novel prion protein gene mutation at codon 196 (E196A) in a septuagenarian with Creutzfeldt–Jakob disease

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Creutzfeldt–Jakob disease (CJD) is a rare and rapidly progressive neurodegenerative disease in the central nervous system, which may occur in inherited, acquired (variant and iatrogenic), or spontaneous (sporadic) forms. Genetic CJD is caused by a point mutation of the prion protein (PrP) gene (PRNP) on human chromosome 20. Here we report a 76-year-old CJD patient found unexpectedly to harbor a novel mutation in PRNP. Routine clinical investigations were undertaken to elucidate the cause of the rapidly progressive dementia and neurological decline manifested by the patient. Both neuron-specific enolase (NSE) and 14-3-3 protein in the cerebrospinal fluid (CSF) were positive. Magnetic resonance imaging (MRI) diffusion-weighted image (DWI) revealed ribbon-like high signals in the bilateral frontal, insular, occipital and parietal cortices, suggestive of CJD. Electroencephalography showed typical periodic synchronous discharge (PSD). Continuous deterioration of clinical symptoms was observed, and typical manifestations including myoclonus and akinetic mutism gradually appeared. CJD was clinically diagnosed based on the above characteristic findings. Interestingly, a point mutation of PRNP at codon 196 (E196A) was detected. To our knowledge, we are the first to report the point mutation of PRNP at codon 196 (E196A) in patients with CJD.

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Abstract — WCN 2013

No: 75

Topic: 5 — Dementia

Facilitative effects of ginkgolide B and velvet antler polypeptide on cholinergic progenitor differentiation

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Objective: We aim to investigate the effects of ginkgolide B (GKB) and velvet antler polypeptide (VAP) on the generation and differentiation of cholinergic progenitors.

Methods: Telencephalon cells were cultured. GKB, VAP and the combination of them were added into the culture medium. Cell culture was terminated on day 4, and then the SP kit (Boster, China) was used to mark the ChAT and BrdU positive cells. The data of these positive cells were analyzed among groups.

Results: Compared to the control group, the neurospheres of experimental group were bigger and the neurites were longer and stronger. More cells migrated from neurospheres. Most cells of ChAT positive distributed on the edge of neurospheres; only a small amount scattered among the neurospheres. The number of ChAT positive neurons in experimental group was higher than that in the control group, especially the GKB and VAP combined group. The VAP group had the most BrdU positive cells.

Conclusion: GKB and VAP, especially when used together, could facilitate the generation and differentiation of cholinergic progenitors efficiently.

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Abstract — WCN 2013

No: 9

Topic: 5 — Dementia

Antidepressant treatment improves estrogen levels and cognitive function in postmenopausal anxiety/depression patients

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Rationale and objectives: The current study investigated the role of antidepressant treatment on the level of sex hormones and cognitive function in post-menopausal anxiety/depression patients.

Methods: A total of 82 post-menopausal patients who were diagnosed with anxiety/depression disorder and were not receiving hormonal replacement therapy participated in a prospective, 6-month, open-label naturalistic study. Patients were divided into an antidepressant treatment group (44 cases) and a non-antidepressant treatment (control) group (38 cases). We collected demographic data and tested sex hormone levels and assessed the extent of psychological disorder and examined cognitive function.

Results:

- 1) The E2 level at six-months post-treatment was significantly increased, conversely, FSH and LH levels were significantly decreased, in the antidepressant treatment group compared with pre-therapy or the control group ($P < 0.01$).
- 2) HARS-14 and CHDS scores decreased significantly more, MoCA scores significantly improved in the antidepressant treatment group than in the pre-therapy or the control group ($P < 0.01$).
- 3) E2 levels were strongly correlated with MoCA and CHDS or HARDS-14 scores at pre- and post-therapy in the antidepressant treatment group. E2 levels maintained a significant association with improvement in CHDS at post-therapy, even we controlled for age, severity of depression, and MoCA scores, ($R = 0.35$, $P = 0.038$).

Conclusions: The antidepressant Paroxetine improved cognitive function in postmenopausal anxiety–depression patients, possibly by increasing endogenous estrogen discharge or delaying degeneration of the gonads.

doi:10.1016/j.jns.2013.07.1120

Abstract – WCN 2013**No: 311****Topic: 5 – Dementia****'Lexical' alexia in a Chinese semantic dementia patient**S.K.S. Ting, S. Hameed. *Neurology, National Neuroscience Institute, Singapore, Singapore*

Semantic dementia (SD) is characterized by losing the ability to understand the meaning of words and losing semantic knowledge. Reading disorder or surface dyslexia or 'regularization' error is typically described in English speaking patients. However, to date little is known about the reading disorder in Chinese semantic dementia patients. We describe a case of a Chinese SD patient who completely lost the ability to read Chinese characters even with common low stroke high frequency words. This suggests that Chinese character reading is likely mainly reliant on visual ventral pathway (orthography), in keeping with temporal lobe atrophy seen in SD, rather than phonological pathway.

doi:10.1016/j.jns.2013.07.1121

Abstract – WCN 2013**No: 128****Topic: 5 – Dementia****Effects of green tea consumption on cognitive dysfunction: An exploratory clinical study**K. Ide^a, N. Wakamiya^a, M. Park^a, N. Takuma^b, S. Fujii^b, A. Nakahara^b, T. Suzuki^b, J. Nakase^c, Y. Ukawa^c, Y.M. Sagesaka^c, H. Yamada^a. ^aGraduate School of Pharmaceutical Sciences, University of Shizuoka, Shizuoka, Japan; ^bWhite Cross Nursing Home, Higashimurayama, Japan; ^cCentral Research Institute, ITO EN, Ltd., Shizuoka, Japan

Background: In rapidly aging societies worldwide, the number of patients with disorders marked by cognitive dysfunction, such as Alzheimer's disease, is gradually increasing; however, thus far, no fundamental curative therapy has been established. Green tea, whose major constituents are catechins and theanine, is known to have various health benefits for humans. However, the effects of green tea consumption on cognitive dysfunction remain to be clinically verified.

Objective: To conduct a clinical study to investigate the effects of green tea consumption on cognitive dysfunction.

Patients and methods: Twelve elderly nursing home residents (2 men, 10 women; mean age, 88 years) with cognitive dysfunction (Mini Mental State Examination [MMSE] score: <28) were recruited. Written informed consent was obtained from the patients and caregivers. The patients were asked to consume green tea powder (2 g/day [containing 227 mg of catechins and 37 mg of theanine]; manufactured by ITO EN Ltd., Tokyo) for 3 months. Cognitive function was assessed by performing MMSE, and serum lipid levels were measured.

Results: The MMSE scores (mean \pm S.D.) significantly improved after the intervention (before intervention, 15.3 ± 7.7 ; after intervention, 17.0 ± 8.2 ; $p = 0.025$), especially for the category recent memory. Regarding serum lipid levels, low-density lipoprotein (LDL)-cholesterol levels remained unaltered, but triglyceride levels decreased significantly (before intervention, 124 ± 80 mg/dL; after intervention, 103 ± 57 mg/dL; $p = 0.041$).

Conclusion: These results suggest that green tea consumption may be effective in improving cognitive function, and additional confirmatory long-term controlled studies are needed.

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Abstract – WCN 2013**No: 308****Topic: 5 – Dementia****Fast and slow conversion to Alzheimer's disease in mild cognitive impairment: Role of cerebrospinal fluid biomarkers**M. Tondelli^a, A. Chiari^a, M.A. Molinari^a, R. Bedin^a, G. Bonifacio^a, T. Trenti^b, P. Nichelli^a. ^aNeuroscience Department, University of Modena and Reggio Emilia, Modena, Italy; ^bClinical Pathology-Toxicology, Ospedale Civile S. Agostino-Estense, Modena, Italy

Introduction: It is well known that CSF biomarkers help discriminating preclinical AD from age-associated memory impairment and other forms of dementia. Little is known about the ability of these biomarkers to predict speed of progression to AD in MCI patients.

Objective: To examine whether demographic, neuropsychological and CSF parameters at the preclinical stage of AD can be used to discriminate between slow and rapid converters to AD in patients with MCI.

Materials and methods: A group of 71 MCI patients was recruited from our Memory Clinic according to clinical criteria. We investigated baseline CSF and neuropsychological biomarker patterns between groups of MCI patients stratified by later diagnoses of conversion to AD or other dementia. In addition, MCI patients who converted to AD (cMCI) were classified in slow and rapid converters (< or > 18 months).

Results: cMCI show at baseline lower A β 1-42 as compared to patients who remained stable MCI or who developed other dementia. The subgroup analysis between slow and rapid converters confirmed the role of A β 1-42 to discriminate between the 2 groups: rapid converters showed already at baseline lower A β 1-42 levels as compared to slow converters. Survival analysis confirmed that cMCI patients with lower value of CSF A β 1-42 at baseline had significantly shortest median dementia-free survival.

Conclusion: Our results suggest that A β 1-42 may be related to speed of conversion to AD in MCI patients and it could be useful in the stratification of cMCI population to support patients' workup in clinical setting.

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Abstract – WCN 2013**No: 186****Topic: 5 – Dementia****Music therapy using flash song therapy for outpatients with juvenile Alzheimer's disease**M. Nakamura^a, M. Iizuka^b. ^aNeurology, Kyoto Medical Center, Kyoto, Japan; ^bBrain Nerve Center, Kyoto Medical Center, Kyoto, Japan

Background: Music therapy is applied to dementia as a non-pharmacological treatment; there is some evidence of its beneficial effects. However, most reported studies have been focused on patients in nursing homes.

Objective: To examine the effects of music therapy using Flash Song Therapy for dementia patients at home and visiting our hospital.

Patients and methods: Eight patients with juvenile Alzheimer's disease (MMSE 0–23, average 10.5) were included in this study; their cognitive functions, behavioral and psychological symptoms of dementia (BPSD), ADL, QOL and caregiver burden were evaluated before and after 8 and 16 private sessions. We used Flash Song Therapy, a method of music therapy that we developed. In this method, patients sing their favorite songs, usually more than twenty, one after another. Just when the first verse of one song ends, the introduction of the next song begins; the quick changes of songs and rhythms stimulate their brains and bodies and fill them with pleasure and satisfaction.

Results: The scores of BPSD and QOL improved significantly after the sessions, and some patients showed improvements in cognitive function. The tempo of gait tended to increase after each session. Some families and caregivers reported that the patients became calm, joyful and able to speak about themselves confidently, among others.

Conclusion: Music therapy using Flash Song Therapy could be one clinical approach for outpatients with dementia.

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Abstract – WCN 2013

No: 530

Topic: 5 – Dementia

Utility of neuropsychological tests in the diagnosis of amnesic mild cognitive impairment and Alzheimer's disease or other forms of dementia

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Aim: To show objective cognitive decline and estimate the severity of dementia and to determine whether dementia severity and the atrophy predict subsequent cognitive deficiency in dementia.

Background: The identification of variables that accurately predict progressive cognitive decline in dementia has important clinical implications and is essential to apply potential treatments. Amnesic mild cognitive impairment patients form a heterogeneous population that constitutes a higher risk for dementia development.

Methods: We recruited 5 AD patients and 3 amnesic mild cognitive impairment (aMCI) patients. Magnetic resonance imaging (MRI) was applied to all subjects for evaluating the atrophy. We applied a neuropsychological test battery including Wechsler memory scale (WMS), verbal fluency, verbal learning, dual similarities, clock drawing test, Benton face recognition, Hooper visual organisation test, visual memory test, visual learning and MMSE.

Results: 5 AD patients confessed cognitive problems with the worst cognitive score on memory test (encoding, recall, retrieval) and impaired visuospatial skill (Benton Facial Recognition) related with medial temporal or posterior cortical lobe atrophy. The 3 amnesic mild cognitive impairment patients confessed cognitive problems on executive dysfunction (and lesser on memory impairment) and severe visuospatial and visuo-constructive disabilities and psychomotor slowing.

Conclusion: AD patients presented progressive episodic memory impairment and impaired visuospatial skills (progressive cortical visual and visuospatial impairments if posterior cortical atrophy was present). aMCI patients may develop AD, but also other forms of dementia. Clinical correlates of cognitive disorders must be frequent in dementia.

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Abstract – WCN 2013

No: 508

Topic: 5 – Dementia

Involvement of the cholinergic basal forebrain nuclei in spinocerebellar ataxia type 2 (SCA2)

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University, Würzburg, Germany; ^dDepartment of Neurology, Philipps-University of Marburg, Marburg, Germany; ^eDepartment of Medical Genetics, University of Pecs, Pecs, Hungary; ^fDepartment of Neurology, J.W. Goethe University, Frankfurt/Main, Germany; ^gDepartment of Pathology and Medical Biology, University Medical Centre of Groningen, Groningen, The Netherlands

Background: Spinocerebellar ataxia type 2 (SCA2) is a polyglutamine disease associated clinically with attention deficits, impairments of verbal memory, frontal-executive and visuospatial functions, as well as global cognitive decline. The possible morphological correlates of these cognitive deficits are unclear.

Objective: To analyze the neuropathological state of the cholinergic basal forebrain nuclei (i.e. medial septal nucleus, MS; nuclei of the vertical and horizontal limbs of the diagonal band of Broca, DBV, DBH; basal nucleus of Meynert; BNM) in SCA2 patients which are believed to be crucial for several aspects of normal cognition.

Patients and methods: Pigment–Nissl stained and AT8 tau-immunostained serial thick sections through the cholinergic basal forebrain nuclei of clinically diagnosed and genetically confirmed SCA2 patients and control individuals were analyzed.

Results: These analyses revealed a severe and consistent neuronal loss in all of the cholinergic basal forebrain nuclei (MS: 72%; DBV: 74%; DBH: 72%; BNM: 86%) of the SCA2 patients studied. Damage to the basal forebrain nuclei was associated with everyday relevant cognitive deficits only in our SCA2 patient with an advanced Alzheimer's (AD)-related tau pathology.

Conclusions: The findings of the present study (1) indicate that the mutation and pathological process of SCA2 play a causative role for the severe degeneration of the cholinergic basal forebrain nuclei and (2) suggest that degeneration of the cholinergic basal forebrain nuclei per se is not sufficient to cause profound and global dementia detrimental to everyday practice and activities of daily living.

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Abstract – WCN 2013

No: 501

Topic: 5 – Dementia

Cognitive function in patients with arterial hypertension and cerebrovascular complications of Uzbek people

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Objective: To study the state of cognitive function in patients with arterial hypertension (AH) and chronic cerebrovascular events (CCVEs).

Material and methods: 50 patients of both sexes were examined aged from 33 years to 73 years. Cognitive disorders were detected using some scales: MMSE and a battery of tests of frontal dysfunction – BTFD. All the patients were divided as follows: the first group – patients with hypertensive encephalopathy; the second group – patients with ET 2, the third group – patients with ET-3, and the fourth group – patients with GE effects of stroke.

Results: Thus, 85.7% patients of the first group were with normal cognitive function according to BTFD. Patients with mild cognitive impairment were 14.3%. In the second group the frequency of occurrence of false violations and symptoms of mild dementia according to BTFD were the same with similar data obtained in the study on an MMSE scale. Moderate cognitive disorders are almost 2 times less likely (21.8% vs. 35.8% $p < 0.05$).

In the third group consisting of patients with mild cognitive impairment according to BTFD was 70.8%. The lowest percentage included patients with dementia of mild severity (4.2% vs. 20.8%, $p < 0.03$).

In the fourth group consisted of patients characterized by differences in the absence of dementia, detection of dementia is 20% according to the MMSE and BTFD scales. That the results of both tests in 80% of patients had mild cognitive impairment.

Conclusion: The degree of cognitive impairment depends on the degree and duration of hypertension.

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Abstract – WCN 2013

No: 226

Topic: 5 – Dementia

Human prion diseases in Japan: A prospective surveillance from 1999

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Background: A nationwide surveillance system for human prion diseases (PrDs) was established in April 1999 (Nozaki et al.).

Objective: To describe the features of epidemiology, clinical manifestations and CSF biomarkers of human PrDs in Japan.

Patients and methods: Information on clinical, neuropathological, and molecular genetic data of patients suspected as having PrDs was analyzed by the Committee, from April 1999 to September 2012.

Results: We have obtained the information of 3664 patients. A total of 1894 cases (51.7%) of PrDs were identified, including 1452 cases of sporadic CJD (sCJD) (76.7%), 270 cases of genetic CJD (14.3%), 79 cases of GSS (4.2%), 4 cases of FFI (0.2%), 81 cases of dura mater graft-associated CJD (dCJD) (4.3%), and 1 case of variant CJD (vCJD) (0.1%). The overall annual incidence rate was 1.01 cases per million person-years. Genetic analysis was performed in 1375 patients, and revealed that 351 cases had a mutation in PRNP gene. The most frequent mutation was 151 cases of V180I mutation (43.0%), followed by 70 cases of P102L, 52 cases of M232R, 49 cases of E200K, 6 cases of P105L, 5 cases of D178N, and 3 of V180I + M232R. Autopsy was performed in

17% of the total 1458 patients. Of the total 144 dCJD cases in Japan, 81 cases appeared in this surveillance system. Only one case of vCJD was identified in 2005.

Conclusion: Human prion diseases in Japan were characterized by frequent occurrence of dCJD and relatively larger amount of patients with uncommon genetic prion diseases.

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Abstract – WCN 2013

No: 486

Topic: 5 – Dementia

Brain volumetry and emotional processing in demented patients

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Background: Emotional deficits and cerebral atrophy are both features of Alzheimer's disease (AD). Behavioral studies have provided evidence that AD patients have deficits in the recognition of emotional expressions, especially when the processing is concurrent with another task.

Objective: We examined the relationship between brain atrophy and deficits in the identification of emotional expressions under two attentional conditions: simple and double task (DT) performance.

Patients and methods: 40 subjects (27 healthy subjects, mean age = 71.70 ± 4.23 and 13 AD patients, mean age = 75.92 ± 6.64 years) had to identify 24 facial expressions enacting the six basic emotions under two conditions: identification only and identification concurrent with a Digit Recall trial. All participants underwent an MRI scan. All MR imaging was performed on a 3T Signa HD× MRI scanner (GE Healthcare, Waukesha, WI) using an eight-channel phased array coil. A 3DT1 sequence with a TR = 9.24 s, TE = 4.148 ms, TI = 650 ms, NEX = 1, matrix size = 512 × 512, 170 slices, resolution = 0.4688 × 0.4688 × 1, and flip angle = 12 was acquired for each subject. For each subcortical structure the volume normalized by the total intracranial volume was calculated.

Results: It is shown that the atrophy of subcortical structures in AD patients (amygdala, hippocampus and pallidum) correlates only with the simple identification task, and the atrophy of cortical structures correlates with both conditions (frontal lobe and parietal lobe).

Conclusion: The present findings represent a contribution to our understanding of emotional processing in demented patients. Specifically, we demonstrate how brain atrophy characteristic of AD patients affects the ability to identify emotional faces as a function of attentional demands.

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Abstract – WCN 2013

No: 476

Topic: 5 – Dementia

The mini mental state examination could be helpful for thromboembolic risk stratification in atrial fibrillation?

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Aims: To determine whether atrial fibrillation (AF) in stroke-free patients is associated with impaired cognition and to determine the correlation between thromboembolic risk and the cognitive impairment.

Methods: 218 patients with AF, free of stroke, were consecutively examined. All cases underwent physical and instrumental examinations. To investigate the cognitive status subjects underwent the Mini Mental State Examination.

Results: The subjects (218) (mean 70.1 ± 0.65 years; 59% W) were stratified according to ECG features into 3 groups: (I) those with de novo AF ($n = 6$; 2.8%), (II) those with paroxysmal AF ($n = 40$; 18.4%), and (III) those with chronic AF ($n = 172$; 78.9%). Cognitive status was found to be significantly different in the 3 groups: group I – 28.1 ± 1.9 ; group II – 25.9 ± 2.9 ; and group III – 24.9 ± 2.9 ($p < 0.01$). Thromboembolic risk (according to CHA₂DS₂-VASc) was similar between groups (3.1 ± 0.6 in gr.III vs. 2.9 ± 0.8 in gr.II and 1.9 ± 0.2 in gr.I, $p = 0.05$). There was a significant association between thromboembolic risk and the presence of cognitive disturbances (MMSE < 26) (8.7% low risk, 21.4% moderate risk, and 48% high risk, $p < 0.05$). Brain CT scan has shown multiple cerebral ischemic areas in 52% of patients with cognitive disturbances. Among 56 patients with CHA₂DS₂-VASc score = 1, twelve of them (21.4%) have had MMSE – 26 with lacunar or ischemic changes in 25%.

Conclusions: Cognitive dysfunction is common in patients with chronic atrial fibrillation. The MMSE, in addition to CHA₂DS₂-VASc, could help to identify patients with atrial fibrillation who need oral anticoagulation and those who need extra efforts to maintain effective optimal anticoagulation.

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Abstract – WCN 2013

No: 453

Topic: 5 – Dementia

Diabetes exacerbates Alzheimer's disease induced brain pathology. Possible neuroprotective effects of Cerebrolysin

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Diabetes mellitus is a devastating disease that affects central nervous system (CNS) function causing brain pathology. Recent studies indicate that Alzheimer's Disease (AD) may reflect brain diabetes and may have a strong link with the disease as predisposing factor. However, the details of a possible link between diabetes and AD are still not well known. Previous works from our laboratory showed that diabetes exacerbates brain pathology following heat stroke and traumatic brain or spinal cord injuries. Thus, there is a possibility that diabetes may also aggravate AD induced brain pathology. This hypothesis was examined in our lab using a rat model of AD using amyloid-beta protein (AbP) infusion in normal or diabetic rats. Our observations show that AbP infusion exacerbates brain pathology in diabetic rats as compared to normal control group. This indicates that diabetes has the capacity to enhance AD pathology. Interestingly, concomitant infusion of Cerebrolysin, a combination of the brain, glia and endothelial cell derived neurotrophic factors with small peptide fragments is able to thwart AbP induced AD pathology in low doses (2.5 to 5 ml/kg, i.v.) in normal rats. However, in diabetic animals high doses of the drug (10 ml/kg) or nanowired delivery of the compound (5 ml/kg) is needed to achieve effective neuroprotection. These observations suggest that

diabetes play a major role in AD pathology and modulate therapeutic efficiency of drug therapy.

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Abstract – WCN 2013

No: 426

Topic: 5 – Dementia

Frequency of occurrence of disorders of higher mental functions of vascular diseases of cerebrum and dynamics under influence of treatment

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Background: Disorders of higher mental functions of vascular diseases of cerebrum are essential. Characteristic of disorder in the sphere of higher mental activity and its dynamic condition under influence of treatment are important for daily life.

Objective: To study the specialties of disorders of higher mental functions of patients with cerebrovascular disease.

Patients and methods: 121 patients (26.4% men and 73.6% women) with an average age 63.6 ± 8.8 and with cerebrovascular disease were studied. Neuropsychological examination was made in all spheres of higher mental activity. After treatment, examination of dynamic condition of higher mental function was made.

Results: Disorders memory 88.4% of patients, attention 83.5%, somatosensory gnosis 81.9%, intellect 66.9%, auditory gnosis 59.5%, dynamic praxis 52.9%, speech 50.4%, optic gnosis 44.6%, counting 11.6%, writing 10.7%, and reading 8.3%. After treatment: disorder memory 40.5% of patients, attention 31.4%, somatosensory gnosis 18.2%, intellect 26.4%, auditory gnosis 13.2%, dynamic praxis 9.9%, speech 17.4%, optic gnosis 18.3, counting 5.0%, writing 6.6%, and reading 4.1%. Most changes: memory 88.4%, attention 83.5%, somatosensory gnosis 81.9, and intellect 66.9%. Least changes: counting 11.6%, writing 10.7%, and reading 8.3%. Spheres of higher mental activity which had considerable improvements: after treatment – somatosensory gnosis: before treatment 81.8%, after treatment 18.2%; auditory gnosis: before treatment 59.5%, after treatment 13.2%; and dynamic praxis: before treatment 52.9%, after treatment 9.9%.

Conclusion: For diagnostics of higher mental disorders of vascular diseases of cerebrum, the study of memory, attention, somatosensory gnosis and intellect is necessary. All spheres of high mental activities can be treated. Best results are possible in somatosensory gnosis, auditory gnosis and dynamic praxis.

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Abstract – WCN 2013

No: 593

Topic: 5 – Dementia

Alzheimer's disease CSF biomarkers predict cognitive decline in healthy elderly over two years

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Background: In some studies with elderly as control subjects, up to 30% of people developed cognitive decline in the next 2–3 years.

Aim: To check the ability of CSF biomarkers of Alzheimer's disease (CSF-BMK-AD) to make discriminations within a healthy group,

according to their cognitive development, at two years of obtaining the sample.

Material and methods: Between 2008 and 2010, 67 subjects without cognitive or behavioral disorders were included as a control group in a study of CSF-AD-BMK. In most cases, neuropsychological assessment was performed both at baseline and at follow-up two years later. CSF was obtained at the inclusion and analyzed by INNOTEST reagents, measuring $A\beta_{1-42}$, total-tau and P-tau_{181p} protein levels, as well as T-tau/ $A\beta_{1-42}$ and P-tau_{181p}/ $A\beta_{1-42}$ ratios.

Results: Two years after inclusion, 28 subjects were not able to be checked. Among the 39 remaining, 29 were cognitively stable and 11 showed a GDS score greater than 2. When compared both groups, we found significant differences in $A\beta_{1-42}$ protein level (820 vs 1359 pg/ml, $p < 0.003$), in the T-tau/ $A\beta_{1-42}$ ratio (0.40 vs 0.19, $p < 0.009$) and in the P-tau_{181p}/ $A\beta_{1-42}$ ratio (0.09 vs 0.04, $p < 0.003$).

Conclusion: In a group initially asymptomatic, CSF-BMK-AD seems able to discriminate between subjects depending on their cognitive evolution at the two years follow-up. Our results are consistent with the decrease of CSF $A\beta_{1-42}$ protein levels, as the first finding in preclinical AD.

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Abstract – WCN 2013

No: 428

Topic: 5 – Dementia

Serum levels of albumin–amyloid beta complexes are useful for monitoring of the progression of Alzheimer's disease

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Objective: Decreased amyloid β ($A\beta$) clearance from brain to blood may play a key role in the development of Alzheimer's disease (AD). $A\beta$ is normally bound to and transported by albumin in blood, thus possibly maintaining a constant concentration of free $A\beta$ in blood. We therefore hypothesized that decreased serum levels of albumin– $A\beta$ complexes may be associated with decreased $A\beta$ removal from brain to blood, resulting in $A\beta$ accumulation in the brain.

Methods: We performed a cross-sectional investigation of the association between serum levels of albumin– $A\beta$ complexes (SLAAC) and AD prevalence.

Results: The mean SLAAC was significantly lower in the AD group than in the control group. Decreased SLAAC was correlated with decreased CSF levels of $A\beta_{42}$ and elevated CSF p-tau ($r = 0.38$, $P = 0.0221$; $r = -0.43$, $P = 0.0090$, respectively), findings that have been shown to be associated with AD progression.

Conclusions: This novel method may be very useful for monitoring of the progression of AD.

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Abstract – WCN 2013

No: 678

Topic: 5 – Dementia

Effects of endogenous serotonin potentiation on passive avoidance learning and memory in rats

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Background: Activation or blockage of serotonin receptors can induce alterations in learning and memory. The physiologic role of serotonin in different models of learning and memory processes is not clearly understood.

Objective: In this study the role of endogenous brain serotonin on acquisition, consolidation and retrieval steps of passive avoidance learning and memory was investigated.

Material and methods: In this study, in the case groups, fluoxetine was administered either orally before training (group 1) or retention (group 2), or by intraperitoneal injection after training of passive avoidance learning and memory tasks (group3), while the control groups only received saline without fluoxetine. The data were analyzed by Student *t* test for parametrical data, and Mann–Whitney test for nonparametrical data.

Results: The number of learning trials for acquisition in the group receiving fluoxetine before training was more than control group. There were no significant differences in the step trough latency to the dark compartment and the time spent in the dark compartment between groups that received fluoxetine or saline after training (consolidation), or the groups receiving oral fluoxetine or saline before performance during the retention test.

Conclusion: It is concluded that, in the passive avoidance learning and memory task, potentiating serotonin function in the synapses could decrease acquisition ($P < 0.05$), but cause no significant effect on consolidation and retrieval ($P = 0.3$).

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Abstract – WCN 2013

No: 614

Topic: 5 – Dementia
Suicide in dementia

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Objectives: Suicide occurs more often in elderly and its risk is increasing with age. Suicide rates in men are three times higher compared with women. The reasons for suicide are predominantly mental or physical illness. Especially mood disorders and the assumption to suffer from degenerative dementia are risk factors. The aim of our study is to determine risk factors of suicide in our memory-ambulance.

Methods: We examined $n = 1000$ patients in our memory-ambulance between 2002 and 2012. All patients suffered from dementia (vascular dementia or dementia of Alzheimer's type). Testing by Mini-mental-status test (Folstein, 1990) revealed mild, or moderate dementia.

Results: In our sample, only one patient (0.1%) suffering from moderate dementia of Alzheimer's type took his own life by suicide.

Conclusion: We did not find any increased risk of suicide in our sample of $n = 1000$ patients suffering from dementia. We suppose that regular care and continuous consultations may prevent patients to execute suicide.

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Abstract – WCN 2013

No: 615

Topic: 5 – Dementia
Frequency of falls in dementia

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Objectives: A growing problem is falls in elderly. They are often characterized by spontaneous events, not caused by external reasons like accidents. Often falls happen due to sedating drugs or missing coordination in the night. The aim of our study is to compile events of falls in a sample of patients visiting our memory ambulance.

Methods: Included were n = 400 patients in our memory-ambulance (200 patients suffering from vascular dementia (VD), 200 patients with dementia of Alzheimer's Type (DAT)). The observational period was 36 months. Testing by Mini-mental-status test (Folstein, 1990) revealed mild, moderate, or severe dementia.

Results: Within 36 months we registered 120 accidents (falls) without any external reason. All accidents caused injuries requiring treatment. We found no correlation between frequency of falls and intensity of dementia or sex. Patients with VD suffered significantly more frequent falls compared with DAT-patients.

Conclusion: We suppose that the higher frequency of falls in VD patients could be attributed to a higher amount of multimorbidity in this group. DAT patients often seem to be more casual and therefore could prevent falls more effectively.

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Abstract – WCN 2013

No: 762

Topic: 5 – Dementia

Impact of homocysteine on cognitive disorders in hypothyroidism

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Background: Hypothyroidism morbidity in the Republic of Belarus comprises 55.3 per 100 thousand people. In the decompensation stage 64% of patients with hypothyroidism have cognitive disorders with varying intensity (M.E. Begin et al.). Their formation mechanism is connected with the metabolic disorders.

Objective: To determine the impact of homocysteine on the formation of cognitive disorders.

Patients and methods: The amount of examined patients with clinical hypothyroidism from 2010 to 2012 comprises 80 people (male – 5 people, female – 75), the average age was 50.5 (40.5–57.0) years.

There has been assessed neurological status, the results of neuropsychological testing and serum homocysteine levels by immunofluorescent analysis on the unit «Architect I-2000 SR» company «Abbott» with the help of panel firm «DRG International Inc.» (USA).

Results: There have been three subgroups identified:

- 1) with mild cognitive disorders – 41 people (62.5%), whose homocysteine level was 13.4 (11.0–16.9) µmol/L;
- 2) with medium cognitive disorders – 9 people (11.2%) with homocysteine level 16.8 (13.5–18.8) µmol/L;
- 3) control group of 30 people (37.5%) – without cognitive disorders with homocysteine level – 12.8 (10.6–15.6) µmol/L.

The comparative analysis of the homocysteine level in different age groups has revealed statistically significant differences only in patients with hypothyroidism with cognitive disorders between the ages of 41 to 50 years (U = 23.0; p = 0.044).

Conclusion: Mild cognitive disorders are an early presentation of hypothyroidism. Hyperhomocysteinemia affects the formation of

cognitive disorders in hypothyroidism in patients older than 40 years.

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Abstract – WCN 2013

No: 691

Topic: 5 – Dementia

Comparative study between clinical symptoms found in most common dementias

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Background: According to the literature, we found that the most common dementia in the world is Alzheimer's (AD) followed by vascular dementia (VD), frontotemporal (FTD) and disseminated Lewy bodies (DLB), each having some characteristic symptoms that can differentiate them.

Objective: Evaluate and compare the main symptoms of dementia found in the population.

Methods: The study was conducted in the Ambulatory of Cognitive Neurology, where patients were evaluated claiming loss of memory. These individuals underwent a full neuropsychological and complementary evaluation for the dementia diagnostic.

Results: A number of 867 patients were assessed by loss of memory. In this group we found 367 patients with Alzheimer's diagnosis, 135 with vascular dementia, and 24 with frontotemporal 14 for Lewy body disease.

For forgetting recent events: AD shows 354 (96.5%), VD 132 (97.8%) FTD 3 (12.5%) and LBD 13 (92.9%). To behavior change: AD was 60 (6.3%), VD 35 (25.9%), FTD 17 (70.8%) and LBD 3 (21.4%) and for personality changes: AD 44 (12%), VD 36 (26.7%), FTD 17 (70.8%), and LBD 4 (28.6%).

Depression: AD 82 (22.3%), VD 38 (28.1%), FTD 12 (50%), LBD 2 (14.2%). For hallucinations 31 (8.5%), VD 31 (23%), FTD 3 (12.5%) and LBD 13 (92.9%).

Conclusion: The frontotemporal dementia initially evolves without forgetting recent events (12%), however has major complaints of changes in personality and behavior (70.8%). Alzheimer's shows up with low capacity for behavioral changes (6.3%) and personality (12%). Depression affects more patients with frontotemporal dementia followed by vascular dementia. Hallucinations appear more in disseminated Lewy body dementia, followed by vascular dementia.

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Abstract – WCN 2013

No: 828

Topic: 5 – Dementia

Relationship between cerebral cortical lesion progression and clinical findings in MM1-type sporadic Creutzfeldt–Jakob disease

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The neuropathological findings of sporadic Creutzfeldt–Jakob disease (sCJD) include spongiform degeneration in the gray matter, gliosis with hypertrophic astrocytosis, tissue rarefaction, neuron loss, and prion protein deposition. Comprehensive neuropathologic, molecular, and

genetic analyses, as well as investigation of clinical features, including biomarkers, will increase our understanding of sCJD. We investigated the clinical course and cerebral cortical pathology of 38 MM1-type Japanese sCJD cases. The first purpose of this study was to propose cerebral cortical pathological staging of sCJD by using hematoxylin and eosin (H-E staining). The second purpose was to investigate the relationship between clinical findings and cerebral cortical pathology. In the present sCJD series, we observed varying degrees of widespread cerebral neocortical involvement, and severity tended to be associated with total disease duration. On the basis of our present pathologic findings, we re-propose a cortical pathologic staging scheme classified into 6 stages as follows:

spongiform change (Stage 1),
hypertrophic astrocytosis (Stage 2),
tissue rarefaction (Stage 3),
neuron loss (Stage 4),
status spongiosus (Stage 5), and
large cavity formation (Stage 6).

This staging can be used more simply as follows: Stages 1 and 2 regarded as mild, Stages 3 and 4 considered moderate, and Stages 5 and 6 regarded as severe involvement. Statistical analysis revealed a positive correlation coefficient between the average neocortical pathologic stage and total disease duration and a negative correlation coefficient between the average neocortical pathologic stage and brain weight.

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Abstract – WCN 2013

No: 715

Topic: 5 – Dementia

Convenience sample versus epidemiological sample for the study of mild cognitive impairment and Alzheimer's disease – Fewer differences than expected

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Background: The characteristics of cognitively healthy subjects and subjects diagnosed with Mild Cognitive Impairment (MCI) from the epidemiologically derived Sydney Memory and Ageing Study (MAS) sample were compared with those of the convenience sample used for the Australian imaging, Biomarkers and Lifestyle (AIBL) study of ageing, in order to better to understand relationships between selection methods and sample characteristics when studying MCI and risk factors for the future development of cognitive decline, MCI and Alzheimer's disease among healthy subjects.

Methods: Cognitive variables were harmonised and identical MCI criteria were applied.

Results: Cognitively healthy AIBL subjects were demographically advantaged but not different in cognitive function to cognitively healthy MAS subjects. For MCI subjects from both studies there were negligible cognitive or demographic differences when diagnostic criteria were applied identically to subjects from both studies.

Conclusion: Results from convenience samples may be more generalisable than expected.

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Abstract – WCN 2013

No: 739

Topic: 5 – Dementia

Value of dehydroepiandrosterone sulfate determination in the diagnosis of early forms of Alzheimer's disease

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Purpose: Establishing the relationship with cognitive performance level of DHEA-S with early form of Alzheimer's disease (AD).

Material and methods: The study included 30 patients (12 men and 18 women), aged 65 (average age 61.3 ± 5.7 years) from the presence of cognitive impairment, and 5 healthy individuals. Research was conducted by ELISA with the definition of the level of DHEA-S before and after the oxidation catalyst Fe²⁺.

Results and discussion: Clinic of AD in patients of the main group was characterized by the presence of organic neurological deficits and a progressive decline in cognitive function. Neuropsychological research on the MMSE results suggest cognitive functions: 21.8 ± 4.05 in the group of patients with asthma and 28.1 ± 0.8 in healthy individuals. Oxidation of serum leads to a sharp increase in DHEA levels in the control group (up to 5.2 ± 0.5; after 7.1 ± 0.5), whereas in the serum of patients with AD, improvement wasn't observed or was negligible (up to 2.1 ± 0.5; after 2.3 ± 0.3).

Conclusions: Determining the level of DHEA and the reaction of Fe²⁺ may be taken as one of the biochemical markers of diagnosis of early forms of AD, which can be recommended for screening diagnosis of early forms of AD.

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Abstract – WCN 2013

No: 85

Topic: 5 – Dementia

Clinicopathological study of dementia in a consecutive autopsy series of 83 patients

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Background and objective: The prevalence of dementia has been increasing lately. Here, we retrospectively assessed the difference in clinical diagnosis and pathological diagnosis of dementia.

Patients and methods: From a consecutive autopsy series of 83 patients at Oyamada Memorial Spa Hospital, Japan, we reviewed 58 patients (25 men, 33 women; age at autopsy, 79.7 years) who had been clinically diagnosed with dementia. All cases were examined postmortem at the Institute for Medical Science of Aging, Aichi Medical University, Japan.

Results: Clinical diagnoses were Parkinson's disease/dementia with Lewy bodies (DLB; n = 15), Alzheimer's disease (AD; n = 13), prion disease (n = 11), vascular dementia (VD; n = 6), progressive supranuclear palsy (PSP; n = 4), frontotemporal dementia (FTD; n = 2), Huntington disease (HD; n = 2), mixed type dementia (AD-VD; n = 2), corticobasal degeneration (CBD) + normal pressure hydrocephalus (NPH) (n = 1), multiple sclerosis (MS; n = 1), and Korsakoff's syndrome (n = 1).

Pathological diagnoses were DLB (n = 14), prion disease (n = 11), AD (n = 6), VD (n = 6), argyrophilic grain disease (AGD; n = 4), PSP (n = 4), CBD (n = 3), frontotemporal lobar degeneration (FTLD; n = 2), HD (n = 2), superficial siderosis (n = 2), DLB + AD + NPH (n = 1), amyloid angiopathy (AA; n = 1), MS (n = 1), and senile dementia of the NFT type (SD-NFT; n = 1). Besides the pathology of

degenerative diseases, cerebral amyloid angiopathy, traumatic damage, and cerebrovascular disease influenced the clinicopathological features.

Conclusion: Pathological research is indispensable for diagnosing neurodegenerative and dementia diseases, especially AD, DLB, PSP, CBD, FTL, AA, SD-NFT, and AGD. Here we demonstrated the importance of pathological diagnosis for cases of dementia.

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Abstract — WCN 2013

No: 534

Topic: 5 — Dementia

Different cognitive profiles between Parkinson's and Alzheimer's diseases screened by Montreal cognitive assessment. A multicenter study of Keio PD database

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Objectives: To demonstrate the difference in cognitive impairment between PD and AD, evaluated by Montreal Cognitive Assessment (MoCA) which is recommended as screening cognitive impairments of PD.

Methods: We registered PD and AD patients at 13 participating hospitals, and measured their cognitive functions employing Japanese versions of MoCA and MMSE.

Results: The subjects were 304 PD patients (age 70.6 ± 8.3, H&Y 2.7 ± 0.7, mean ± SD) and 97 AD patients (age 78.4 ± 5.9). The PD patients were grouped into tertiles according to MoCA score; the high (24–30 points, n = 108), middle (19–23, 98) and low (5–18, 98) score groups. The AD patients were grouped likewise; the high (17–24, 35), middle (12–16, 31) and low (3–11, 31) score groups. By comparing the AD patients of high score group with the MoCA score- and age -matched PD patients, the subscores of trail making test (26% in AD vs. 51% in PD, mean) and orientation (76% vs. 95%) were lower in AD, and the subscore of serial 7 subtraction (92% vs. 76%) was lower in PD (p < 0.05, HSD test). In these patients, the subscore of 3-stage command of MMSE was lower in AD (68% vs. 86%) discriminating AD from PD with sensitivity 0.80 and specificity 0.77.

Conclusions: Naturally AD patients presenting as dementia yielded a lower range of MoCA score than PD patients whose cognitive function varies from normal to dementia. Comparison between AD and PD patients with equivalent MoCA score revealed the different impairment patterns of MoCA and MMSE tasks, suggesting their utility for differential diagnosis.

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Abstract — WCN 2013

No: 923

Topic: 5 — Dementia

Near-infrared spectroscopy (NIRS) in the diagnosis of Alzheimer's disease

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Objective: The aim of this study was to assess activation of brain regions using NIRS in patients with Alzheimer's disease (AD).

Methods and patients: Seventeen probable AD patients 57–83 years of age with a mean MMSE score of 22.5 ± 4.1 and 16 normal subjects 57–88 years of age were investigated. All subjects participated in three activation tests. Test 1 consisted of the Kana (Japanese letter) pick up test in which the patients were instructed to select five specified letters from randomly arranged Kana letters (Task 1) and old Japanese tales written in Kana letters (Task 2). Test 2 was a modified Stroop test in which the subjects were required to identify the ink color (Task 1) and the color of Chinese letters (Task 2). Test 3 comprised verbal fluency tests in which the subjects were asked to repeat as many words as possible from a semantic category (Task 1) and a phonetic category (Task 2). We used the 16-probe, 46-channeled NIRS device (OMM 3000, Shimadzu Corporation, Japan) and evaluated the OxyHb concentration for the analysis.

Results: The average concentration of OxyHb in the normal subjects increased in the left lateral frontal channels in Test 1, the bilateral frontal to parietal channels in Test 2 and the left posterior frontal to parietal channels in Test 3. In the AD patients, the average OxyHb concentration decreased significantly in these channels in all tests.

Conclusions: Measuring the concentration of OxyHb using NIRS is useful for making a diagnosis of mild AD.

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Abstract — WCN 2013

No: 915

Topic: 5 — Dementia

Neuropsychological and cognitive profile of retired Filipino boxers

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Introduction: Boxing has been a popular sport in the country, more so in the past few years, with a Filipino boxer dominating the global professional boxing scene. In light of its fame, the risk factors of actual and potential chronic injury among Filipino boxers should be investigated. It has been known that repetitive brain trauma associated with boxing may produce progressive neurological deterioration and is associated with memory disturbances, behavioral and personality changes.

Objectives: This study aimed to describe the neuropsychological and cognitive impairment among retired Filipino boxers and its possible risk factors.

Methods: This is a descriptive study on the cognitive and behavioral profile of retired Filipino boxers using standard screening examinations such as the Neuropsychiatric Index, Symptoms Suggestive of Cognitive Impairment, Hospital Anxiety Depression Scale and Montreal Cognitive Assessment Scale.

Results and discussion: An increase in times knocked showed a possible relationship with a low MoCA score. However, the test being in English, language barrier is therefore a big factor. SSCI and HADS showed a relationship with the increased rate of being knocked out and with longer years since retirement. Development of cognitive impairment, and neuropsychologic and neuropsychiatric features is therefore strongly supported by an increased rate of being knocked out and a longer time after retirement, chronic traumatic encephalopathy being a potential consequence of repetitive brain trauma.

Conclusion: An increased rate of being knocked out therefore increases the risk of developing cognitive impairment, neuropsychologic and

neuropsychiatric features. Moreover, symptoms are manifested after longer retirement years.

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Abstract – WCN 2013

No: 913

Topic: 5 – Dementia

Neuropathological study in dementia-free centenarians

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Background: There are centenarians who are free of dementia despite the presence of high Alzheimer's disease pathology.

Objective: To assess the prevalence of "dementia-free" among centenarians, and to ascertain presence of dementia-related neuropathological changes in dementia-free centenarians.

Material and methods: Centenarians were evaluated with MMSE or Hasegawa dementia scale. The autopsied brains from thirteen dementia-free centenarians were studied with standard hematoxylin–eosin, Klüver–Barrera and Gallyas–Braak stainings, and immunostainings for α -synuclein, phosphorylated tau, amyloid- β ($A\beta$) and TDP-43. CERAD criteria of neuritic amyloid plaques, Braak and Braak staging of neurofibrillary tangles and Saito's staging of argyrophilic grains were used to assess the brains.

Results: Thirteen of 46 (28%) centenarians were cognitively intact. The average brain weight of dementia-free centenarians (1096.5 g) was heavier than that of 46 centenarians (1035 g). While they were clinically dementia-free, they were diagnosed neuropathologically as Lewy body disease (5 cases), argyrophilic grain disease (3 cases), senile dementia of the neurofibrillary tangle type (3 cases), and Binswanger's disease (2 cases). Although none of the cases was diagnosed as Alzheimer's disease, substantial amount of senile plaques (CERAD B) and neurofibrillary tangles (Braak IV) were observed. Only 4 cases with subtle age-related changes were diagnosed as "normal-aged".

Conclusion: Many dementia-free centenarians had neuropathological evidence of dementia. These results may indicate that dementia-free centenarians seem to have resistance to developing dementia despite the presence of subtle pathology of dementia.

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Abstract – WCN 2013

No: 847

Topic: 5 – Dementia

The pathogenic mechanisms induced by amyloid beta in neural stem cells

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Objectives: Many studies of biochemical and animal studies support the concept that amyloid beta ($A\beta$) plays a central role in the development of Alzheimer's disease (AD). Although a lot of efforts have been focused on the pathogenic mechanisms of $A\beta$, it is unresolved how $A\beta$ accumulates in the central nervous system and subsequently initiates the disease of cells. In this study, we investigated the clue of the pathogenic mechanisms of $A\beta$ by proteomics and antibody microarray. **Material and methods:** To evaluate the effect of $A\beta$ on neural stem cells, we treated primary cultured cortical neural stem cells with several doses of $A\beta$ for 72h. We detected several factors that may be associated with $A\beta$ treatment by proteomics and antibody microarray. An MTT assay and

BrdU cell proliferation assay were performed. And, western blotting for the evaluation of effect on intracellular signaling proteins was achieved. **Results:** Various viability tests showed that $A\beta$ decreased neural stem cell viability and cell proliferation in a concentration-dependent manner. After $A\beta$ treatment, it significantly decreased lactate dehydrogenase B (LDHB), high-mobility group box 1 (HMGB1), survival signals, including phosphorylated Akt, and glycogen synthase kinase-3 β . $A\beta$ also increased Heat Shock Protein (HSF70), Electron-transferring Flavoprotein (ETF), and death signals, such as phosphorylated tau (pThr231) and activated caspase-3.

Conclusion: These results suggest that several factors which we found by proteomics and antibody microarray could be the clues of $A\beta$ pathogenesis. Further studies should be conducted, with a focus on these factors.

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Abstract – WCN 2013

No: 855

Topic: 5 – Dementia

Calcium-sensing receptor antagonist (calcilytic) NPS2143 prevents amyloid- β_{42} secretion in postnatal human neurons

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Objectives: We investigated whether calcium-sensing receptor (CaSR) signaling plays any role in fibrillar (f) $A\beta_{25-35}$ - and soluble (s) $A\beta_{25-35}$ -elicited effects on $A\beta_{42}$ and $A\beta_{40}$ secretion by HCN-1A neurons.

Methods: Human cortical HCN-1A neurons were treated with f $A\beta_{25-35}$ or s $A\beta_{25-35}$ plus/minus the CaSR allosteric antagonist NPS2143. The intracellular accumulation and secretion of $A\beta_{42}$ and $A\beta_{40}$ were analyzed via ELISA and immunofluorescence microscopy.

Results: In the HCN-1A neurons, f $A\beta_{25-35}$ and s $A\beta_{25-35}$ stimulate $A\beta_{42}$ production and secretion through a CaSR-mediated control of $A\beta_{42}$ accumulation and release. A CaSR antagonist, the "calcilytic" NPS2143, significantly prevented f $A\beta_{25-35}$ - and s $A\beta_{25-35}$ -treated HCN-1A neurons from secreting an excess of $A\beta_{42}$. Instead the intracellular distribution and secreted amounts of $A\beta_{40}$ were not changed by an exposure of HCN-1A neurons to f $A\beta_{25-35}$ \pm NPS2143 or s $A\beta_{25-35}$ \pm NPS2143.

NPS2143 preserved the viability of the f $A\beta_{25-35}$ and s $A\beta_{25-35}$ exposed HCN-1A neurons, whereas in its absence the $A\beta$ s induce the death of about 25% of the neurons within 72 h.

Conclusions: These data suggest that after stimulation with f- or s $A\beta_{25-35}$ the $A\beta_{42}$ is the main species released from HCN-1A neurons. By negatively modulating Ab/CaSR signaling NPS2143 prevents the surplus release of $A\beta_{42}$ induced by f- or s $A\beta_{25-35}$ and exerts a protective effect on the survival of Ab-treated neurons. Furthermore they suggest that a CaSR inhibitor like NPS2143 blocking by a feed-forward mechanism of $A\beta$ self-induction might be a novel therapeutic drug for Alzheimer's disease.

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Abstract – WCN 2013

No: 1010

Topic: 5 – Dementia

MRI classification of idiopathic normal pressure hydrocephalus and its prediction of shunt effectiveness

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Background: With the rapid aging of Japanese society, medical care of the idiopathic normal pressure hydrocephalus (iNPH) becomes an important issue. We reported the clinical significance of disproportionately enlarged subarachnoid-space hydrocephalus (DESH) on diagnosis of iNPH in our multicenter prospective cohort study. DESH findings fulfill three criteria of tight high convexity, enlarged sylvian fissure and ventriculomegaly (Evans index >0.3). However, we noted some patients in which these criteria were equivocal.

Purpose: To make clear which MRI findings are favorable for prediction of shunt effectiveness, we classified the MRI findings of our iNPH patients into DESH, incomplete DESH and non-DESH. Incomplete DESH consisted of findings fulfilling two criteria with one equivocal criterion.

Methods: The present study included 83 patients [54 males and 29 females] aged 66–87 years [mean 77 years] in our hospital during the recent four years. All of them were treated with ventriculo[45 patients]- or lumbar subarachnoid[38 patients]-peritoneal surgery. Improvement was assessed at discharge.

Results: Among the eighty-three patients, DESH was noted in 55 patients, incomplete DESH in 24 patients and non-DESH in 4 patients. Patients with DESH showed improvement in 74.5%, incomplete DESH in 58.3% and non-DESH in 33.3%. Among patients with incomplete DESH, patients with ambiguity on tight high convexity showed improvement in 27.2%, while patients with tight high convexity showed improvement in 84.6%.

Conclusion: iNPH patients can be divided into three types of DESH, incomplete DESH and non-DESH. DESH and incomplete DESH with tight high convexity showed high predictability of shunt effectiveness.

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Abstract — WCN 2013

No: 273

Topic: 5 — Dementia

Effect of montelukast and its interaction with the protective effect of rofecoxib, caffeic acid in kainic acid induced cognitive dysfunction

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Introduction: Potential role of antioxidant and COX-2 inhibitors has also been well reported against memory impairment in different experimental models.

Objective: The present study aims to explore the potential role of montelukast (a cysteinyl leukotriene inhibitor) in concert with rofecoxib (COX-2 inhibitor) and caffeic acid (a 5-LOX inhibitor and potent antioxidant) against kainic acid induced cognitive dysfunction in rat.

Materials and methods: In the experimental protocol, kainic acid (0.4 μ /2 μ l ACSF) was given intrahippocampally (CA3 region). Memory performance along with the locomotor activity was measured on the days 10–14 and 1, 7 and 14 respectively. The brains were isolated on the 14th day; oxidative stress parameters and mitochondrial enzyme complexes were then estimated.

Results: Montelukast (0.5 and 1 mg/kg), rofecoxib (5 and 10 mg/kg) and caffeic acid (5 and 10 mg/kg) showed significant improvement in memory performance as compared to that in control (kainic acid treated). Further, two weeks of the drug treatments reversed all the biochemical indices and showed favourable mitochondrial protection. Combination treatments of montelukast with rofecoxib or caffeic acid showed significant synergism of their protective effect which was significant as compared to their effect per se.

Conclusion: Thus the present study is unique in that it shows the positive modulation of cysteinyl leukotriene receptor inhibition on COX and LOX pathways in the control of existing neuroinflammation.

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Abstract — WCN 2013

No: 953

Topic: 5 — Dementia

The targets in Alzheimer's disease are A β oligomers: Count them for diagnosis and destroy them for therapy

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Alzheimer's disease (AD) is a progressive neurodegenerative disorder. Several lines of evidence suggest a central role of amyloid- β -peptide (A β) in the pathogenesis of AD. More than A β fibrils, small soluble and prion-like A β oligomers are suspected to be the major toxic species responsible for disease development and progression. Therefore, these oligomers should be our major target for therapy and used as the most direct biomarker for diagnosis and therapy monitoring.

Diagnosis: The A β oligomer count in CSF of AD-affected and healthy persons as determined by our new ultra-sensitive surface-based fluorescence intensity distribution analysis (sFIDA) assay revealed a surprisingly clear distinction between both groups. All samples of the control group showed homogeneously low numbers of A β oligomers, while the samples of the AD group exhibited significantly higher levels of A β oligomers with high variability. The A β oligomer levels clearly correlated with the patients' mini-mental state examination (MMSE) scores [Wang-Dietrich et al., *J Alzheimers Dis.* 34, 985–994 (2013)].

Therapy: We present our newest in vitro and in vivo results on D-enantiomeric peptide derivatives that specifically address A β oligomers and convert them into non-amyloidogenic, non-fibrillar and non-toxic species without increasing the concentration of monomeric A β . We show that next to plaque load and inflammation reduction, oral application of the peptide improved cognitive performance of AD transgenic mice. Cognitive improvement was even observed upon treatment of very old mice [Funke et al., *ACS Chem. Neurosci.* 1, 639–648 (2010); van Groen et al., *J Alzheimers Dis.* 34, 609–620 (2013)].

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Abstract — WCN 2013

No: 963

Topic: 5 — Dementia

Vascular dementia risk factors: The role of diabetes and hypertension among younger-old and older-old patients

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Objective: The main objectives of this project were:

- 1) To identify the role of diabetes hypertension as predictor of vascular dementia among the young-old and older-old patients.
- 2) To identify the co-morbid psychological disorders and determine their contributing role in presentations of dementia symptom.

Participants and methods: 350 individual ages 55 and older participated in this study; the subjects were the clients of a local large primary care practice. The participants' medical file was evaluated in order to obtain information as to their vascular risk factors. The risk factors were mainly based on the present diagnosis of HTN, high cholesterol and diabetes. Several cognitive screening data were obtained from the patients.

Results: The most prevalent risk factors that were identified in this group of participants were hypertension, and then high cholesterol and finally diabetes. Age was highly correlated with these risk factors as well. The group that was at the highest risk with the three risk indicators of hypertension, high cholesterol and diabetes was the older age group. The group with long term hypertension (more than 10 years) also showed a great deal of cognitive impairment.

Conclusions: Preventing the vascular risk factors is essential for either delaying or preventing symptoms of dementia. In the U.S. we need to manage the underlying causes of the risk factors in order to be able to prevent dementia disorders. The current practice of symptomatic treatment of the vascular risk factors does not necessarily prevent development of dementia related disorders.

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Abstract – WCN 2013

No: 966

Topic: 5 – Dementia

Cognitive impairment – Cause or consequence of anticoagulation underuse?

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Aim: We have analyzed whether cognitive capacity influences the control of anticoagulation in patients with atrial fibrillation.

Methods: 218 patients with atrial fibrillation (AF) and no history of stroke, were consecutively admitted. All cases underwent medical examination. To investigate the cognitive status, subjects underwent the neuro-psychological rating scale – MMSE.

Results: The 218 patients (mean age 70.1 ± 0.65 years; 59% W) were stratified according to CHA₂DS₂-VASc score into 3 groups. Patients with high thromboembolic risk (150/218) have been selected for anticoagulation. About 21/150 patients have had contraindications, being switched to aspirin, and 129/150 have initiated anticoagulation with warfarin. The number of warfarin users has dropped by 7.8% (–10), in six months by 13.2% (–17), in 12 months by 38% (–49), and in 24 months the number of subjects continuing anticoagulation has dropped by 48.8% (–63). Of these patients, 43 (19.7%) had an MMSE score <24, suggesting dementia, and 60 (or 27.5%) had intermediate scores of 24 to 26. Low MMSE scores were associated with a low rate of anticoagulation use (60.3% among those who suspended treatment with warfarin vs. 33.3% of those who continued treatment). The frequency of cardiac decompensation was higher at a follow up of 2.1 years in those with MMSE < 26 (p < 0.05).

Conclusion: Cognitive dysfunction is common in patients with AF and is related to less effective anticoagulation and more cardiac events. The MMSE identifies patients with AF in whom extra efforts are needed to maintain effective anticoagulation.

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Abstract – WCN 2013

No: 818

Topic: 5 – Dementia

Gender specific differences in cognitive profiles of patients with Alzheimer dementia. Results of the prospective dementia registry Austria (PRODEM-Austria)

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Goals: Gender differences have been well documented for healthy individuals in several cognitive tasks. Recent investigations suggested that gender may be an important modifying factor in the development and progression of Alzheimer's disease (AD). Here we examined gender-specific differences in the pattern of cognitive dysfunction of patients with mild to moderate AD.

Methods: We examined 417 subjects (mean age 83 (+8.1) years, 59% females) of the prospective registry on dementia in Austria (PRODEM), an ongoing longitudinal multi-center cohort study, conducted in 12 Austrian memory clinics. We analyzed differences in the cognitive profile between male and female AD patients on the CERAD-Plus test battery.

Results: We found gender related differences in the neuropsychological domains of verbal learning, delayed recall, recognition and intrusion errors; the women tended to perform worse than men. There were no significant differences between the two groups in constructional praxis. Analyzing the effect of gender in the possible AD and the probable AD groups, we found gender differences in the probable AD group only in verbal learning performance. Controlling for stage and duration of dementia and the level of education there was still a significant effect of gender on verbal episodic memory.

Conclusion: There is an interaction between gender and cognitive function, most notable in verbal episodic memory; female patients with AD performed worse on verbal episodic memory than men. This indicates that the gender specificities of neuropsychological functions should be given careful consideration in clinical diagnosis of dementia.

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Abstract–WCN 2013

No: 1217

Topic: 5–Dementia

Agraphia in posterior cortical atrophy

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Introduction: The Posterior Cortical Atrophy (PCA) is a neurodegenerative syndrome that is characterized by progressive decline in visuospatial, visuoperceptual, literacy and praxis skills. We report two cases of PCA with severe writing disorders.

Case 1: A 60-year-old man, right-handed, had progressive visual disorders. The neuropsychological examination shows: apperceptive agnosia, visuo-spatial disorders, apraxia, alexia and agraphia, realizing Gerstmann's syndrome and an incomplete Balint's syndrome. The agraphia was of spatial type. Cerebral MRI showed parieto-occipital atrophy predominating on the left hemisphere.

Case 2: A 51-year-old man, right-handed, had progressive visuospatial and gestural disorders. The neuropsychological examination shows: left visual hemi-neglect, neglect alexia and severe apraxia affecting hands, feet and the trunk; agraphia was of apraxic type. Cerebral MRI showed bilateral parietal atrophy predominating on the right hemisphere.

Comments: We report two cases of PCA with different cognitive and neuroradiological presentations (Aharon-Peretz et al, 1999). An occipital form with visuo-perceptive disturbances and writing disorders of the spatial type, and a parietal type with predominant apraxic disorders with a writing disorders of the apraxic type. And a parietal form with predominating apraxic disorders, in this form writing disorders is of apraxic type.

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Abstract – WCN 2013

No: 1224

Topic: 5 – Dementia

White matter change and cognitive function are related to APOE status

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Background and objectives: The aim of our study was to examine whether the presence of cerebral white matter change (WMC) associated with the performance in various cognitive domains. Additionally, we investigated whether apolipoprotein E (APOE) genotype influenced the association between WMC and cognitive function.

Method: We subjected 230 persons who visited our memory clinic. All subjects underwent MMSE and comprehensive neuropsychological tests to evaluate the following cognitive domains: attention, language, visuospatial function, verbal memory, visual memory, and executive function. The presence and severity of WMC was rated using a Fazekas scale and categorized into three groups. Multiple linear regression models were conducted to explore the association of WMC with cognitive domains after adjustments for age, sex, and education. We also performed analyses stratified according to APOE genotype.

Results: Two hundred fourteen participants (93.0%) had WMC. 75.6% had a Fazekas total score ranging from 1 to 4 and 40 (17.4%) were in severe group. Severe WMC group was older than no WMC group. Hypertension, atrial fibrillation, and prior stroke history were frequently shown in the severe WMC group. Higher Fazekas score was shown lower Z score in MMSE and worse performance on attention, language, visuospatial function, and verbal memory. Sixty two (27.0%) had APOE ϵ 4 allele. After stratifying according to APOE genotype, we found that severe WMC was associated with MMSE, attention, language, visuospatial function in APOE ϵ 4 non-carriers, while no association between WMC and cognitive function in APOE ϵ 4 carriers.

Conclusion: The present study showed the association of WMC with cognitive dysfunction. Especially, WMC implicated the performance in multiple cognitive domain in the APOE ϵ 4 non-carrier group.

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Abstract – WCN 2013

No: 1220

Topic: 5 – Dementia

The benefit of physical activity on cognition: Results from the Austrian stroke prevention study

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Objective: Cognitive decline leads to disability and decreased quality of life. It is related to life-style decisions, diet and physical activity. Here we investigate the association VO₂ max as a measure of cardio respiratory fitness (CRF) and cognitive domains, as well as global cognition in a large population based cohort. Additionally we investigate mediation by MRI correlates of brain atrophy and vascular degeneration.

Methods: The cohort consisted of 730 healthy participants (mean age 65 ± 7 years, 58% females) of the Austrian Stroke Prevention study. Cognitive function measurements include single tests, as well as composite scores for specific domains, including memory, conceptualization, visuopractical skills and attention / speed and global cognition (g-factor). CRF was assessed by performing a graded exercise stress test setup on a treadmill ergometer, and is expressed as VO₂max. Brain MRI was used to measure brain parenchymal fraction (BPF) and white matter lesion load (WML load). Multiple linear regression was used to test the association.

Results: VO₂max was significantly associated with almost all cognitive domains, (p < 0.05), as well as global cognition g (p < 0.01). The associations were independent of age, sex, educational level and life-style (smoking, BMI) and vascular risk factors (hypertension, diabetes, cholesterol) and ApoE genotype). White matter lesions and brain atrophy partly mediate the described effect of cardio-respiratory fitness on cognition in general, and in on each individual domains.

Interpretation: The results support the protective role of cardio-respiratory fitness on cognition, which may ameliorate age-related cognitive decline and thus disability in old age.

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Abstract – WCN 2013

No: 1172

Topic: 5 – Dementia

Using cognitive qEEG to characterize disease severity in the prospective dementia registry Austria (PRODEM)

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Quantitative EEG (qEEG) changes in Alzheimer's disease (AD) include frequency slowing, altered synchrony and reduced complexity. So far, qEEG alterations have been used to distinguish AD or prodromal AD from controls. The current study validated these qEEG metrics by assessing their relationship with cognitive functioning in patients with probable AD. Moreover, we defined an overall qEEG metric by independent component analysis. We studied 103 AD cases from the multi-centric study PRODEM-Austria. Their mean age was 73.1 years and mean dementia duration was 27 months. All patients underwent EEG recording in resting and cognitively active state. EEG analysis included automated artifact removal, manual selection of artifact-free epochs for qEEG assessments and calculation of six different markers in the domains frequency, synchrony and complexity. Quadratic regressions were calculated to determine the magnitude of variance of MMSE scores that was explained by each EEG metric and by the combination factor. Analyses were adjusted for age, sex, duration of dementia, and educational level. Relative theta power during face encoding explained 38% of the variance of MMSE scores, followed by relative alpha and beta1 power which explained 35% and 32%, respectively. No single measure of synchrony or complexity explained more than 30% of MMSE variance. With 28% variance explanation the combination factor was also inferior to frequency measures. Our data indicate theta power measured during face encoding to be most closely related to AD severity. Its role as a qEEG marker to predict AD progression needs to be further determined in longitudinal studies.

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Abstract – WCN 2013

No: 1236

Topic: 5 – Dementia

Anticardiolipin antibodies, cognition, and brain magnetic resonance imaging

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Background and purpose: The presence of anticardiolipin antibodies (aCLs) has been associated with vascular occlusive events. The role of aCLs as a risk factor for stroke has been a matter of debate, and there exists scarce information on the relationship between aCLs and other cerebral disorders. Reports exist for seizures, chorea and subtle

cognitive dysfunction. We further explored the association between aCLs and cognition and evaluated the relationship between aCL titres and brain magnetic resonance imaging (MRI) findings in a large cohort of community-dwelling individuals.

Methods: The study cohort was drawn from the Austrian Stroke Prevention Study (ASPS). A total of 1895 subjects had a complete risk factor assessment and measurement of aCLs titres in serum. Participants were classified as aCL positive if either the IgG or IgM aCL titres were elevated (IgG > 21 U/ml, IgM > 12 U/ml). All subjects were also categorized based on the quartile distribution of IgG and IgM isotype titres. All underwent cognitive testing by the Mini Mental State Examination (MMSE) and a random sample of 947 participants also underwent brain MRI.

Results: aCL positive participants performed worse on the MMSE. IgG but not IgM isotype titres related to worse performance on the MMSE. There existed no significant association with vascular brain abnormalities including white matter lesions, lacunes and atrophy.

Conclusions: These data support the view that in normal elderly persons increasing IgG aCL titres relate to global cognitive dysfunction. It is unlikely that structural brain lesions are responsible for this finding.

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Abstract – WCN 2013

No: 900

Topic: 5 – Dementia

Are practice effects an important confounder of longitudinal studies of HIV-associated neurocognitive disorders?

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Background: The diagnosis of HIV-associated neurocognitive disorders (HAND) is based on neuropsychological testing. Scores on neuropsychological tests tend to improve with repeated administrations—the practice effect. Longitudinal studies of HAND may be confounded by the practice effect. We determined the magnitude of the practice effect among HIV-uninfected individuals in Kenya.

Methods: A comprehensive battery of seventeen neuropsychological tests designed to diagnose HAND was administered to 100 HIV-uninfected individuals in 2009. We traced these individuals, performed HIV-antibody testing, and re-administered the neuropsychological tests. Raw scores were converted to Z-scores, and practice effect was defined as the difference between the Z-scores at initial and repeat administrations. A clinically significant change was defined using the Reliable Change Index (standardized Z-score difference $\geq |1.64|$).

Results: Of 100 individuals initially studied, we located 69 of whom 47 participated. The median interval between testing was 98 weeks [Range: 83–130]. Mean age was 30 years, 26% were female, 26% had less than primary school education, and 24% had more than secondary school education. One individual was found to be HIV-infected and was excluded. Overall, practice effects ranged from –0.29 to 0.75 and, among the 33 individuals without neurologic or psychiatric co-morbidities, ranged from –0.26 to 0.72. Practice effects were significantly different from the null for Color Trails, Grooved Pegboard and WAIS-3 Digit Symbol. Between 3 and 9% of HIV-uninfected individuals had clinically significant improvements or declines.

Conclusions: Practice effects may be an important confounder of longitudinal studies. Further research in larger and more representative populations in sub-Saharan Africa is essential.

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Abstract – WCN 2013

No: 663

Topic: 5 – Dementia

Aerobic physical activity stabilizes cognitive status in patients with dementia

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Background: Only symptomatic medical therapy with limited efficacy is presently available for dementia. There is insufficient evidence to date regarding the benefits of physical activity in patients with dementia and its impact on the rate of cognitive decline.

Objective: To evaluate the effect of aerobic physical exercise on the cognitive status in patients with dementia.

Patients and methods: 312 residents with dementia in long-term care facilities were included. The MEC scale (*Mini-Examen-Cognoscitivo*; cutoff score for dementia: 24/35), the Spanish-validated version of the MMSE, stratified individuals into two groups according to severity: mild (MEC 19–24; n = 48) and moderate–severe (MEC \leq 19; n = 264). Individuals were randomly allocated to two groups: control-care group and treatment group consisting of a 15-month supervised-training program of daily moderate aerobic physical activity (recumbent bicycle-heart-rate-integrated system). Cognitive status was measured by the change from baseline-to-final (15 months) in the total score on MEC, Neuropsychiatric Inventory (NPI) and Fuld Object Memory Evaluation (FOME). T-student and 2-way ANOVA statistics were applied.

Results: The mean work load/week was 106.67 min. In the control-care group (n = 208) the baseline/final scores (mean \pm sd) worsened significantly (p < 0.05 for all values): Mild (MEC: 22.00 \pm 1.55/ 20.36 \pm 1.33; NPI: 5.38 \pm 2.65/ 8.10 \pm 6.95; FOME: 14.00 \pm 13.04/ 11.50 \pm 9.09). Moderate–severe (MEC: 11.43 \pm 2.85/ 14.08 \pm 3.03; NPI: 14.30 \pm 15.56/ 19.74 \pm 15.46, FOME: 8.46 \pm 10.23/ 5.52 \pm 2.01). Treatment group (n = 104) scores remained stable: Mild: MEC: 22.00 \pm 1.59/ 22.66 \pm 0.98 (ns); NPI: 5.63 \pm 6.32/ 5.01 \pm 4.49 (ns); FOME: 13.50 \pm 12.67/ 18.00 \pm 11.31 (p < 0.05).

Moderate–severe: MEC: 11.75 \pm 3.03/ 11.62 \pm 2.84 (ns); NPI: 12.24 \pm 10.10/ 10.78 \pm 6.29 (p < 0.05); FOME 4.36 \pm 8.59/ 6.34 \pm 5.09 (ns).

Conclusions: Moderate aerobic physical activity showed benefit in stabilizing the cognitive status in patients with dementia.

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Abstract – WCN 2013

No: 1266

Topic: 5 – Dementia

Optimising disease state fingerprint for diagnosing frontotemporal lobar degeneration

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Background: Disease State Index and its graphical counterpart Disease State Fingerprint form a novel tool that combines data from different

sources, helping the clinician in the diagnosis and follow-up of dementia diseases. We have shown its applicability in the diagnosis of Alzheimer's disease (AD) and Frontotemporal dementia (FTD).

Objective: The tool was applied to classify FTD cases in comparison with AD and control cases.

Methods: The study cohort contains 40 FTD cases, 59 AD cases and 21 controls. The Disease State Index combined data from a wide battery of neuropsychological tests, volumetric MRI and single photon emission computed tomography (SPECT) parameters, cerebrospinal fluid biomarkers and the APOE genotype.

Results: We applied DSI for classifying patients between FTD, AD and controls groups. The data showed that FTD patients could be differentiated with a high accuracy, sensitivity and specificity from controls (0.97, 0.95, 1.00) and AD (0.82, 0.80, 0.83), and AD patients could also be differentiated with a high accuracy from controls (0.96, 0.95, 1.00). Additionally we present one example FTD case compared with controls, AD and FTD population data.

Conclusion: The results suggest that combining data from several sources (neuropsychological tests, MRI, SPECT, APOE and cerebrospinal fluid markers) with the Disease State Index can help differentiate normal status from disease and also FTD from AD. This study also suggests the implementation of SPECT and a full-scale battery of neuropsychological studies, which may help particularly in the differential diagnosis between AD and FTD.

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Abstract — WCN 2013

No: 1290

Topic: 5 — Dementia

Variation in risk factors of dementia among four elderly groups of hospitalized patients

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Objective: To examine variation in risk factors that contribute to dementia among four elderly cohorts by race and gender.

Background: We examined the 2008 Tennessee Hospital Discharged database for vascular factors that play a role in both stroke and dementia. Risk factor for dementia was examined for black and white patients aged 65+.

Methods: Four race-gender groups of patients — white males (WM), black males (BM), white females (WF), and black females (BF) were compared for prevalence of dementia and stroke. A logistic model predicted dementia in each group separately and used several vascular factors affecting dementia directly or indirectly through stroke.

Results: 3.6% of patients hospitalized in 2008 had dementia and dementia was higher among females than males (3.9% vs. 3.2%, $p < .001$), and higher among blacks than whites (4.2% vs. 3.5%, $p < .000$). Further, BF had higher prevalence of dementia than WF (4.2% vs. 3.8%, $p < .001$); similarly BM had more dementia than WM (4.1% vs. 3.1%, $p < .001$). In Logistic regression models, however, different patterns of risk factors were associated with dementia in four groups: among WF & WM, stroke, hypertension, CHF, and diabetes predicted dementia. Among BF & BM, only stroke and diabetes were related to dementia.

Conclusions: Aggressive management of risk factors (hypertension and diabetes) may subsequently reduce stroke and dementia hospitalization.

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Abstract — WCN 2013

No: 1292

Topic: 5 — Dementia

Diagnostic validity comparison between NIA criteria for CSF Alzheimer's disease biomarkers and other new criteria

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Background: In recent NIA criteria for Alzheimer's disease (AD) diagnosis, CSF biomarkers (BMK) have been accepted as evidence of pathophysiological process, for research purposes. Nevertheless, using it for diagnosing mild cognitive impairment (MCI) due to AD with high probability, they show a low sensitivity.

Objective: To compare the diagnostic validity of NIA criteria and our own new criteria, that take into account tau/A β proteins ratios, because they have showed their ability to predict dementia in preclinical AD.

Material and methods: Between 2008 and 2010, 157 MCI patients were included, after lumbar puncture, for measuring A β ₁₋₄₂, T-tau and P-tau₁₈₁ CSF proteins and the ratios T-tau/A β ₁₋₄₂ and P-tau₁₈₁/A β ₁₋₄₂. We used NIA and our own criteria for the evaluation of AD CSF BMK results. Using this analysis we consider a high probability of MCI due to AD, when 3 or more variables were abnormal. We calculated the diagnostic validity for every criteria.

Results: After a two year follow-up, 49 MCI patients remained clinically stable, 78 developed AD and 16 developed other dementia. Using NIA criteria and our own criteria, the BMK for AD showed: sensitivity 35% versus 76%; specificity 89% versus 66%; predictive positive value 79% versus 73% and a predictive negative value of 53% versus 69%.

Conclusion: The use of our criteria for AD CSF BMK assessment should be interesting for early detection of AD, because they show a higher sensitivity than NIA criteria.

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Abstract — WCN 2013

No: 1299

Topic: 5 — Dementia

Discriminability in recognition memory in amnesic mild cognitive impairment and mild Alzheimer's disease.

A preliminary study

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Clinical memory tests that included a yes/no recognition format provide scores only for the total number of target items correctly endorsed. However, many patients with cognitive impairment present a strong "yes" response bias in recognition tests, yielding both high hit and false alarm rates because they cannot reject distractor items. The purpose of this study is to assess recognition memory performance in a sample of cognitively normal adults, amnesic mild cognitive impairment (a-MCI) and Alzheimer's dementia (AD) participants. The sample included 45 a-MCI, 51 AD participants, and 43 controls. Individuals completed the Rey Auditory Verbal Learning Test as part of a larger neuropsychological battery. Yes/no recognition performance was analyzed using signal detection theory. Discrimination indices between targets and distractors (d' — how effectively the individual could differentiate targets

from distractors) and response bias (c – tendency to produce more yes or more no responses) were calculated.

Yes/no recognition showed significant group differences for d' , $F(2) = 83.26$; $p < .001$, and for response bias (c), $F(2) = 6.05$; $p = .003$. Both the impaired groups performed more poorly than controls on discrimination indices ($p < .001$) and response bias ($p = .03$), but there were no differences between the MCI and dementia groups.

Results suggest that the a-MCI and AD groups exhibited significantly poorer recognition discrimination accuracy and a significantly more liberal response bias than the control group. These two recognition memory components can enhance the clinical characterization of an individual's recognition memory performance.

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Abstract – WCN 2013

No: 1289

Topic: 5 – Dementia

CSF Alzheimer's disease biomarker validity for early differential diagnosis of mild cognitive impairment patients

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Background: The early differential diagnosis between mild cognitive impairment (MCI) patients who will remain stable from those who will develop Alzheimer's disease (AD) or other dementia remains a critical issue in clinical setting. CSF AD biomarkers (BMK) have been accepted as evidence of pathophysiological process of AD, in recent NIA criteria.

Objective: To calculate the diagnosis validity of CSF AD BMK in our MCI patients, after a two year clinical follow-up.

Material and methods: Between 2008 and 2010, 157 MCI patients were included, after the realization of a lumbar puncture, for measuring $A\beta_{1-42}$, T-tau and P-tau₁₈₁ CSF proteins, and the ratios T-tau/ $A\beta_{1-42}$ and P-tau₁₈₁/ $A\beta_{1-42}$. As cut-offs, we considered 500 pg/ml, 450 pg/ml, 70 pg/ml, 0.5 and 0.07, respectively. Innotech reagents from Innogenetics were used.

Results: After a two year follow-up, 49 MCI patients remained stable, 78 developed AD and 16 developed other dementia. We excluded in the analysis 14 patients because of death, no data or other diagnosis. When we calculated the variable diagnostic validity for AD, they showed the highest specificities for $A\beta_{1-42}$ (79%) and P-tau₁₈₁ (80%) and the highest sensitivities for ratios T-tau/ $A\beta_{1-42}$ and P-tau₁₈₁/ $A\beta_{1-42}$ (84%).

Conclusion: CSF AD BMK showed a high validity for early diagnosis of AD in MCI patients. Our longitudinal study confirms data from the literature, outlining the value of the ratios tau/ $A\beta_{1-42}$ for this purpose.

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Abstract – WCN 2013

No: 1185

Topic: 5 – Dementia

Butyrylcholinesterase genotype in Alzheimer patients not treated with rivastigmine

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In advanced stages of Alzheimer's disease (AD), butyrylcholinesterase (BuChE) progressively replaces acetylcholinesterase (AChE) in the hydrolysis of acetylcholine. This applies particularly in AD patients with the genetic variant of the BuChE wild-type, which has a higher hydrolysis rate than the frequent BuChE K-variant and is associated with a faster progression of dementia. Rivastigmine is the only commercially available AChE inhibitor, which also inhibits BuChE. Thus it may have advantages over the other AChE inhibitors, particularly in patients with BuChE wild-type.

We determined the proportion of patients with BuChE wild-type in AD patients not treated with rivastigmine.

In a multicentric study, the BuChE genotype was examined in AD patients with progressing dementia, who had no antidementive treatment or were under treatment with either donepezil, galantamine or memantine. The study was conducted in 126 AD patients (48 men, 78 women) at ages between 57 and 94 years (mean/SD: 79.2/7.3 years); MMSE score ranged between 4 and 27 (mean/SD: 17.8/5.7). The patients were treated with either donepezil ($n = 58$), memantine ($n = 29$) or galantamine ($N = 22$); 17 patients were not under antidementive medication. The BuChE wild-type was found in 95 (75.4%) of the patients, in 89 (70.6 %) in the heterozygous and in 6 (4.8%) in the homozygous genotype.

We found the BuChE wild-type in 75% of AD patients with progressing dementia, who were not under treatment with the BuChE inhibitor rivastigmine. In these patients, a switch of antidementive medication to rivastigmine or the initiation of rivastigmine treatment seems to be advisable.

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Abstract – WCN 2013

No: 1161

Topic: 5 – Dementia

A 2-year follow-up study of anti-dementia medications in patients with first diagnosed dementia from community based population

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Background: Alzheimer's disease and other type of dementia are characterized by progressive deterioration of cognitive function and ADL (activity of daily living). Anti-dementia medications such as cholinesterase inhibitors and NMDA receptor antagonist may alter the natural history of dementia.

Objective: The purpose of this study is to analyse the effect of anti-dementia treatment for 2 years in patients with first diagnosed dementia from community based population.

Methods: Participants were dwelling in Eunpyeong-gu, Seoul, Korea in 2008–2009 and going through a screening test using Korean Mini Mental Status Examination (K-MMSE). Detailed neuropsychological tests were performed for the people on the score K-MMSE below than 1.5 SD matched for age and education. The patients diagnosed with dementia were treated by antidementia medications, including donepezil, galantamine, rivastigmine and memantine for 2 years and performed K-MMSE 2 years later. A total of 121 demented people participated in this study.

Results: Seventy eight patients were AD, 30 were VD, 7 were mixed dementia, and 6 were categorized in other dementia. Memantine was used for 42 patients, donepezil for 41, galantamine for 17, rivastigmine for 16, and mixed medication for 5 patients. Mean score of initial K-MMSE was 15.4 ± 5.3 and mean score of two years follow up K-MMSE was 14.8 ± 6.4 , which showed statistically insignificant difference.

Conclusion: Our results showed that there was a short term beneficial effect of the use of anti-dementia medications and delay of a natural

history of dementia. Even though dementia is a progressive neurodegenerative disease, anti-dementia medications are continuously needed.

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Abstract – WCN 2013

No: 87

Topic: 5 – Dementia

Study of vestibular dysfunction in frontotemporal lobar degeneration

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Background: Vestibular imbalance might be involved in the onset of gait disorders that are frequently noted in patients with frontotemporal lobar degeneration (FTLD).

Objective: In view of this, a caloric test was performed to examine vestibular dysfunction in FTLD patients.

Patients and methods: Electro-nystagmography was performed on 9 FTLD patients (69.8 ± 5.4 years) and 9 healthy volunteers (70.9 ± 6.3 years). An air caloric stimulator was used to provide stimulation for vestibular testing, and the eye movement was recorded before and during the visual suppression (VS) test. The frequency and amplitude (slow phase and fast phase) as well as the mean velocity (slow phase and fast phase) of eye movements were measured as the parameters of induced nystagmus. Then, a statistical analysis was performed to examine the following three factors: stimulation sites (right ear, left ear), stimulation temperatures (24 °C, 46 °C), and the timing of stimulation (before or during the VS test).

Results: No significant difference was detected between any of one of the FTLD patients and the healthy volunteers in the slow-phase velocity of nystagmus before VS test. On the other hand, a significant decrease was noted in VS (%) in the FTLD patients, compared to the healthy volunteers ($p < 0.05$).

Conclusion: A severe degree of disturbance of visual suppression was noted in FTLD patients. Findings of the present study suggest that FTLD patients might develop vestibular dysfunction related to cerebral lesions, which might be involved in the onset of gait and balance disorders.

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Abstract – WCN 2013

No: 1130

Topic: 5 – Dementia

The conceptual enrichment therapy: Decelerating word meaning loss in semantic dementia

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Background: A central limitation of interventions for treating anomia in semantic dementia (SD) is the lack of generalisation of the re-acquired learning which remains rigid and context-dependent. The conceptual enrichment therapy (COEN) is a new strategy aiming at

decelerating the progressive loss of word meaning in SD at same time as it generates better generalisation of new learning compared to standard rehabilitation naming therapies (SRNT) (Suárez-González et al. submitted 2013). COEN exploits still-meaningful elements in the semantic network of the target item by strategically linking them to the item in the training phase in order to enhance learning and therefore the possibility of generalisation.

Objectives: To replicate previous results in which COEN learning demonstrated a better generalisation effect than SRNT and to investigate the inter-therapy differences in curves of learning and forgetting.

Patient and methods: A patient with mild SD underwent two different treatments (COEN/SRNT). Following training, generalisation was measured by three different tasks in addition to standard picture naming.

Results: Generalisation effects were significantly better after COEN on description to naming ($p < 0.001$) and naming to description ($p = 0.005$). Interestingly, 6 weeks after the end of the training patient was still able to provide significant information about most of the items trained with COEN but not SRNT (COEN 80%, SRNT 10%).

Conclusions: This second study confirms that treatment based on COEN may boost generalisation of learning in SD by providing a partial restoration of the item's meaning through a semantic knowledge restoration approach.

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Abstract – WCN 2013

No: 1120

Topic: 5 – Dementia

Use of pramiracetam in elderly patients with mild cognitive impairment and arterial hypertension

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Mild cognitive impairment and arterial hypertension are commonly found states in elderly patients and it is also well known that arterial hypertension is a risk factor for developing vascular or neurodegenerative cognitive impairment. Drugs with nootropic activities potentially can change cognitive status in such patient group.

Aim: To determine possibilities of cognitive state correction in patients with mild cognitive impairment (MCI) and arterial hypertension (AH) by oral intake of pramiracetam (Pramistar).

Methods and subjects: 25 elderly patients with MCI and AH were examined prior to and following one month of Pramistar treatment, 1200 mg daily. General clinical and neurological examinations, MMSE, WMS-R, FAB, GDS, State-Trait Anxiety Inventory (STAI), qEEG, and cognitive evoked potentials were performed.

Results: The performed treatment has shown positive improvements for the MCI and AH patients in WMS-R subtests: Attention (visual memory span ($p < 0.002$), digit span ($p < 0.002$)); short-term memory (logical memory ($p < 0.002$), verbal paired associates ($p < 0.01$) and visual reproduction ($p < 0.01$)) as well as long-term memory (logical memory ($p < 0.002$) and visual reproduction ($p < 0.01$)).

In FAB subtest, improvements occurred in conceptualization, motor series (programming) and inhibitory control ($p < 0.01$).

Anxiety ($p < 0.03$) and depression ($p < 0.04$) rates were reduced.

Reduction in delta rhythm power in multiple leads as well as an increase in beta rhythm power in separate leads ($p < 0.05$) had taken place. According to cognitive evoked potentials, peak latencies for N2 and P3 significantly reduced while their amplitude values increased.

Conclusion: Pramiracetam (Pramistar) has shown positive effects in elderly patients with MCI and AH.

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Abstract – WCN 2013**No: 1100****Topic: 5 – Dementia****Co-morbid medical conditions among patients with vascular dementia: A case-control study from a national managed care database**

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Objective: To compare the prevalence of medical co-morbid conditions between patients with vascular dementia (VaD) with a control group without VaD, from the National Managed Care Benchmark Database (IHCS).

Methods: The prevalence of comorbid medical conditions was compared between patients with VaD (defined by ICD-9 codes) and controls using data from the IHCS, a fully de-identified, HIPAA compliant database made up of more than 35 managed care health plans within the US and covering seven census regions. Matched case-control method was used to compare medical comorbidity. Controls were matched to cases by type of health plan and pharmacy benefits on an 18:1 ratio.

Results: Among the 488,091 patients 60 years or older with full year of eligibility, (from January 1st to December 31, 2010), there were 725 patients with VaD, 57.3% of them were women. A concurrent diagnosis of Alzheimer's disease and/or unspecified dementia was exclusion criteria. In general, VaD patients had more cerebral degeneration [odds ratio (OR) = 21.5, 95% CI = 7.0–66.1]; Parkinson's disease (OR = 13.6, 95% CI = 4.0–46.1); cerebrovascular diseases (OR = 12.6, 95% CI = 5.0–31.7); septicemia (OR = 6.5, 95% CI = 2.7–15.5); hypotension (OR = 4.6, 95% CI = 2.0–1052); atherosclerosis (OR = 4.6, 95% CI = 2.6–8.1); injuries (OR = 4.0, 95% CI = 2.6–6.1); heart failure (OR = 2.8, 95% CI = 1.6–4.7); lung diseases (OR = 2.4, 95% CI = 1.6–3.9); COPD (OR = 1.8, 95% CI = 1.2–2.7); cardiac dysrhythmias (OR = 1.7, 95% CI = 1.1–2.6) and urinary diseases (OR = 1.6, 95% CI = 1.1–2.3) compared to controls.

Conclusions: The present study confirms that medical comorbidities are frequent complications of VaD and physicians should be alerted to their presence in patients with VaD.

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Abstract – WCN 2013**No: 1487****Topic: 5 – Dementia****Stressful life events and development of Alzheimer's disease**

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Background: The sporadic nature of AD suggests that aside from biological determinants, environmental factors such as stress play a role in the development of disease.

This study aims to analyze the relationship between stressful life events and AD.

Methods: We studied 118 patients with diagnosis of AD. Medium age was 73 years. Meantime elapsed from the initial symptoms was 2.4 years.

A control group of 81 healthy individuals was studied.

A questionnaire looking for stressful life events in 3 years before diagnosis of AD was performed to patients, caregivers and controls.

Results: In the AD group, 85 patients (72%) presented a history of significant stressful life events, 2.1 years (SD 1.4 years) before the onset of symptoms.

The most common findings in the AD group were: couple death (24 cases), child death (15 cases), history of assault (21 cases), history of car accident (11 cases).

Other stressful situations were marked financial problems, bereavement, retirement, adaptive changes due to migrations and diagnosis of severe somatic disease in the family.

In the control group, only 21 individuals (26%) recognized similar previous stress factors.

Conclusions: We observed an association between stressful life events preceding the onset of dementia in a high percentage of our patients.

Stress could trigger the degenerative process in AD and growing evidences suggest a dysfunction in neuroendocrine and immune system.

According to our results, we can establish a relationship between several stressful life events and the onset of dementia. It is an observational finding and does not imply direct causality.

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Abstract – WCN 2013**No: 1488****Topic: 5 – Dementia****Cognitive decline progression prevention in patients with cerebrovascular diseases**

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Vascular disease of the brain is generally accompanied by focal neurological symptoms along with the development of cognitive memory impairment, that in place may lead to severe degree of dementia. The purpose of this study is to evaluate therapeutic and preventive effects of Alzepil (donepezil) on cognitive impairment in mild multi-infarct dementia in patients with arterial hypertension (AH) and cerebral atherosclerosis (CA).

Material and methods: 32 patients were divided into two groups comparable in all studied parameters. They were all prescribed Alzepil 5 mg/daily for the period of 28 days.

Results: Significant improvement in memory ($p = 0.01$), abstract thinking and reaction time was observed in patients treated with Alzepil, as evidenced by the improvements in EEG. Therapeutic and preventive effects on cognitive disorders in patients with AH and CA were evaluated. Comparison of scores on MMSE and Khachin scales, showed statistically significant improvement for all clinical symptom severity in patients on Alzepil. Meanwhile, in the placebo group, only partial positive trend was observed.

Conclusion: The investigations indicate that, after repeated treatments by Alzepil with 6 month intervals, patients with mild cognitive impairment amid AH and CA, show a steady improvement of the subjective status of patients. In addition, persistent increase in the memory function of attention, abstract and practical thinking were observed. Results of the study suggest that a course of Alzepil (donepezil) treatment in patients with early forms of vascular dementia provides regression of cognitive impairment, and has a stabilizing effect on the overall process, preventing its progression.

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Abstract – WCN 2013**No: 1437****Topic: 5 – Dementia****Selective bilateral thalamic necrosis following carbon monoxide poisoning: Case report**

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Introduction: Carbon monoxide (CO) poisoning is common and probably underdiagnosed. Delayed neuropsychological sequelae (DNS) commonly occur after recovery. On magnetic resonance imaging (MRI), the most common site of abnormality is the globus pallidus. The thalamus can be affected also but in association. To our knowledge, selective lesion of thalami on MRI has not been described.

Case report: We describe the case of a patient presenting with memory dysfunction, disorientation and apathy. He had no cardiovascular risk factors. Mini Mental State Examination estimated to 21/30. The interrogation revealed a CO poisoning 15 days ago. The lesions of the thalami were hypo-intense T1, hyperintense T2 and FLAIR with restricted diffusion.

Discussion: The most frequent symptoms of DNS after CO poisoning include delirium, amnesia and cognitive dysfunction. In our case, the dementia was secondary to bilateral thalamic necrosis. The selective thalamic involvement following CO exposure is rare. CO in particular has a propensity to affect the globus pallidus. The whole basal ganglia can be affected, or the putamen, caudate nucleus, and thalamus may be involved in isolation. Abnormalities of the brain following CO poisoning are shown with greater sensitivity by MRI. Necrosis of the basal ganglia is seen, typically showing low T1 and high T2 signals, but could be observed with high signal on T1 and T2 suggesting haemorrhage, often with restricted diffusion.

Conclusion: Our report suggests that selective thalamic involvement is rare but may occur after CO poisoning.

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Abstract – WCN 2013**No: 1014****Topic: 5 – Dementia****Aphasic syndrome in dementia with Lewy bodies (DLB): Is language impairment one of the features of DLB?**

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Background: Logopenic aphasia has recently been suggested in DLB.

Purpose: To discuss language impairment in DLB.

Patient 1: An 83-year old man started to stumble on words, use non-words and has difficulty in understanding for a year. Subsequently, he showed confusion with fluctuations, delusions and shuffling gait. Initial evaluations revealed logopenic plus jargon aphasia. Brain atrophy and hypoperfusion were accentuated at the left temporo-parietal regions on MRI and SPECT. Phosphorylated (P-) tau and beta-amyloid 42 (A β 42) in CSF suggested concomitant Alzheimer's (AD) pathology.

Patient 2: A 90-year old women presented reduced verbal output, phonological errors, impaired comprehension and shuffling gait for two months. Initial assessment revealed logopenic aphasia. Atrophy and hypoperfusion were detected at the left temporo-parietal areas. MIBG myocardial scintigraphy showed mild uptake decrease.

Patient 3: A 75-year old man became confused with multiple tasks for six months and then had difficulty in word finding and comprehension. Logopenic aphasia was detected at presentation. Atrophy and hypoperfusion were noted at the left temporo-parietal areas. Uptake on

MIBG scintigraphy was slightly decreased. P-tau and A β 42 in CSF suggested AD pathology.

Patient 4: A 67-year old woman presented amnesia and mis-identification syndrome for a year. At presentation, she showed mild cognitive decline without linguistic problems. Abnormal MIBG scintigraphy suggested DLB. SPECT showed that bilateral frontoparietal uptake decreases. Two years after the onset, the patient developed progressive apraxia of speech.

Conclusion: Clinical DLB may present logopenic aphasia and rarely apraxia of speech. Co-pathology of AD may be associated.

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Abstract – WCN 2013**No: 1334****Topic: 5 – Dementia****Nanotechnology for early-stage diagnosis and treatment of Alzheimer's disease**

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Dementia of Alzheimer's type (AD) is a loss of brain function that occurs with certain diseases. It affects memory, thinking and behavior. Scientists believe that changes in the brain may begin 10–20 years before symptoms appear and AD is diagnosed. The need to diagnose the devastating disease at an early stage is critical to manage and treat AD. Unfortunately, the lack of validated biomarkers limits the possibility of the earlier stages of AD. Recent advancements of nanotechnology and nanomaterials in biodetection/bioanalysis may provide an alternative excellent method to help research identify the biomarkers for the disease. The advantages of nanomaterials stem from nanostructures' high surface to volume ratio which enables multi-bioreceptors to bind on the surface of transducer at nano-scale. The nanoparticles as DNA carriers enable million-fold improvements over ELISA sensitivity. Recently, the gold nanoparticles were used to detect the amyloid beta-derived diffusible ligands (ADDL). The finding was made possible by combining ADDL-specific monoclonal antibodies. According to our opinion it is very important to develop multifunctional nanomaterials which are able to deliver genomic sequences and act as a sensor to recognize the targeted biomarkers. Such proposed multifunctional system will lead to a multifunctional bioassay for early-stage diagnosis of AD.

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Abstract – WCN 2013**No: 1309****Topic: 5 – Dementia****A flash on contemporary society**

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Excessive preoccupation, denial, neglect or even exhibition of flaws in appearance are expressions of a dysfunctional body image (Fiori P, Giannetti LM, 20). This may prelude the appearance of behavioral and cognitive dysfunction (Fiori P et al., Rankin KP et al., 2003).

Although the borderline is wider than frank pathological manifestations, the subtle pervasive features of such condition may have individual and social consequences.

The aim of our observational and interventional study is early identification and treatment of such condition. The diagnosis is performed by neuropsychological examination on subjects under the age of 65 years. Preliminary results of our study show:

- A high directiveness, peculiar of males, as well as a high transcendence, predominantly present in females, may not be positive factors concerning self realization, relational competence, social integration and development, compatible with environmental resources and worldwide shared strategy.

- Comorbidity of a dysfunctional body image with addiction, eating disorders, psychoses, epilepsy, cognitive deterioration, vascular diseases.
- Misdiagnosis, scarce compliance, especially in the case of instable socio-economical conditions, lack of educational programs and political leadership.

Body image is the cornerstone of our fitness. Its dynamic steadiness depends on genetic factors, modality of attachment, emotional and affective experiences, personality (temperament and character), behavioral repertoire, cognitive development, environmental, cultural and social factors. Although it is modified by life events, aging and diseases, the response to such challenge is different and depends on cognitive reserve, which has to be preserved and transmitted to new generations.

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Abstract – WCN 2013

No: 1314

Topic: 5 – Dementia

Clinical implications of hippocampal vulnerability patterns in tauopathies

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Background: Deposits of phosphorylated tau in the hippocampus is a hallmark of several neurodegenerative diseases (NDD), which show different, but partially overlapping cognitive symptoms. The hippocampus has several subregions involved in different aspects of cognition.

Objectives: The aim of the study was to evaluate vulnerability patterns of tau pathology in the hippocampus in different NDDs.

Methods: We investigated tau-AT8 immunoreactivity in the neuropil, neurons, astrocytes and oligodendrocytes in 24 hippocampal subregions in individuals above 75 years, including Alzheimer-related pathology with different Braak and Braak stages (AD; total 40 cases), argyrophilic grain disease (9 cases), progressive supranuclear palsy (7 cases), corticobasal degeneration (CBD, 7 cases), Pick's disease (8 cases), globular glial tauopathies (5 cases) and astroglipathies of the elderly (12 cases). Using mathematical modelling, we analysed the constellation of the AT8-immunoreactivity for every disease.

Results: Our model demonstrates specific constellations of tau pathology in the hippocampus. There are regional and cellular differences: Tau-immunopositive oligodendrocytes in stratum oriens of the hippocampus is highly indicative for the CBD; Tau-immunoreactivity in the dentate gyrus (DG) granular cells is suggestive of a non-AD disorder; and, the DG was highly vulnerable in tau-astroglipathy of elderly.

Conclusions:

- 1) Distribution of hippocampal tau immunoreactivity can predict the neuropathological diagnosis;
- 2) These observations have implications for the spreading of disease;

3) Neuropsychological examinations evaluating distinct functions related to hippocampal structures might be useful in the clinical practice to distinguish these disorders.

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Abstract – WCN 2013

No: 1296

Topic: 5 – Dementia

Argentina-ADNI: Preliminary report on CSF biomarkers

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Argentina has launched the first South American ADNI, (Arg-ADNI) effort to acquire data comparable to those gathered throughout other WW-ADNI centers. It is important to share data across the international research community, to evaluate whether clinical, neuroimaging, genetic and molecular characteristics in aging Argentines are similar to those observed in other global ADNI sites. The purpose of this abstract is to describe baseline profiles of 62 (9 controls, 37 mild cognitive impairment (MCI), and 16 Alzheimer's disease (AD)) Arg-ADNI participants.

Compared to individuals with AD, controls and MCI subjects evidenced higher MMSE and Categorical Verbal Fluency scores. MCI and AD cases scored lower than controls on delay and recognition trial RAVLT. APOE ε4 allele possession was prevalent among 33% (n = 7) MCI and 33% (n = 4) AD patients. Levels of CSF biomarkers corresponding to an AD profile including Aβ1-42, T-tau and P-tau, Aβ1-42/P-tau ratio and AD profile were not different between MCI and AD subgroups. Based on CSF biomarkers, 25% (n = 7) MCI patients showed biomarkers of neurodegeneration without positive markers of amyloid accumulation.

After one year of recruitment efforts, this is the first Arg-ADNI report. Arg-ADNI is a growing sample with ongoing recruitment of MCI and AD patients and controls. Follow-up of this important sample, particularly related to CSF and brain biomarkers, will allow better characterization of AD in Argentina, and perhaps point to unique environmental factors influencing brain health in South America. Clinical and biochemical follow up of MCI patients with neurodegenerative profiles and without amyloid accumulation may provide important insights.

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Abstract – WCN 2013

No: 1661

Topic: 5 – Dementia

Intranasal delivery of curcumin–donepezil nanoemulsion for brain targeting in Alzheimer's disease

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Objectives: The objective of the present work was the development of curcumin–donepezil nanoemulsions for delivery to brain by intranasal route. The combination therapy was based on cholinergic replacement hypothesis in combination with anti-amyloid and anti-inflammatory approach for better management of Alzheimer's disease via intranasal route for better brain targeting.

Methods: Nanoemulsions were prepared using aqueous titration technique using components selected based on solubility studies. They were characterized for various physicochemical parameters like globule size, zeta potential, refractive index, transmittance and *ex vivo* diffusion studies using sheep nasal mucosa. Pharmacokinetic studies of developed formulations were carried out in Wistar rats by intranasal and intravenous administration. The *in vivo* studies of developed nanoemulsions were carried out in streptozotocin induced Alzheimer's model. Various behavioural parameters like radial arm maze, elevated plus maze and one trial passive avoidance test were carried out. Biochemical parameters like acetylcholinesterase activity, glutathione, SOD were estimated.

Results: The developed nanoemulsions had particle size less than 50 nm suitable for intranasal delivery. *In vivo* pharmacokinetic studies revealed that higher drug concentration was achieved in brain via intranasal route compared to intravenous route and clearance was slower. Behavioural tasks showed improved memory and learning in group treated with nanoemulsions compared to pure drugs. Acetylcholine levels were significantly improved in brain of nanoemulsion treated group. Oxidative stress was much lower in animals treated with combination therapy.

Conclusions: The intranasal strategy of delivering acetylcholinesterase inhibitor with a neuroprotective and anti-amyloid drug is a promising strategy for the management of Alzheimer's disease.

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Abstract – WCN 2013

No: 1618

Topic: 5 – Dementia

Leptin and the risk of progression to Alzheimer's disease among Chinese older adults with amnesic mild cognitive impairment

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Background: Previous cross-sectional studies have shown that Alzheimer's disease (AD) patients have lower circulating leptin levels than non-demented older adults. There is a paucity of data on the risk of leptin and subsequent AD development in older adults with mild cognitive impairment.

Objective: The aim of the study was to investigate the relationship of the serum leptin level to subsequent risk of progression to AD among Chinese older adults with amnesic MCI (aMCI).

Patients and methods:

Design: One-year prospective cohorts study;

Setting: Ambulatory setting.

Subjects: Chinese older adults, aged 55 to 93 years old, with aMCI by the Petersen's criteria.

Measurements: Baseline demographic, clinical factors, and serum leptin level. All subjects were followed for one year. AD was diagnosed by the NINCDS-ADRDA criteria for probable AD.

Results: 131 Chinese older adults with aMCI were recruited and followed up for one year. 13.7% (n = 18) of them progressed to Alzheimer's disease by the end of one year. Older adults who progressed to AD had significantly a lower mean serum leptin level than that of stable MCI subjects (mean ± SD leptin levels = 5.18 ± 7.40 versus 9.72 ± 8.04 ug/L, respectively).

Conclusion: In Chinese older adults with amnesic MCI, having a low baseline serum leptin level predicts an increased risk of progression to Alzheimer's disease.

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Abstract – WCN 2013

No: 670

Topic: 5 – Dementia

Psychological impact of young onset dementia in Singapore

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Background: Young onset dementia (YOD) carries significant psychological burdens for patients and their caregivers. Previous Asian studies have focused on the geriatric age-group. This is the first study to examine the psychosocial impact of YOD in Singapore.

Objectives: The aim of the study was to investigate the psychosocial effects and perceived needs of YOD patients and their caregivers in Singapore.

Methods: A random sample of YOD patients (age ≤65 years old) attending a memory clinic in a tertiary setting was identified. Diagnosis of dementia had been made by a cognition neurologist.

2 independent physicians conducted phone interviews with patients and caregivers. A questionnaire on psychological impact and perceived needs was used. Case-notes were reviewed for demographic data.

Results: 12 patients (5 AD; 5 FTD; 1 VaD; 1 Hashimoto's) and 10 caregivers were interviewed. Patients' mean age was 59 (51–64); mean MMSE 18 (9–27). Caregivers comprised spouses (8) and children (2).

Emotional responses could be staged into pre-diagnosis, diagnosis, post-diagnosis and future. Caregivers verbalised frustration from the pre-diagnosis period. A few expressed relief at receiving diagnoses that explained patients' behavioural changes. Others experienced helplessness from loss of a functional family member. Prognostication to prepare for care needs and legal matters was found difficult. Associated perceived needs could be further classified into resource-related, rehabilitation, medical and psychological needs. At least 5 identified a lack of age-specific psychoeducation interventions and community services.

Conclusion: YOD patients and their caregivers experience significant emotional upheavals. Healthcare providers need to acknowledge and address this psychological impact. Further nationwide studies are underway.

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Abstract – WCN 2013

No: 1614

Topic: 5 – Dementia

Cerebrospinal fluid biomarkers in Chinese Alzheimer's disease patients

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Background: There is a paucity of data on the validity of cerebrospinal fluid biomarkers in diagnosing Alzheimer's disease (AD) in Chinese patients.

Objective: The aim of the study was to investigate CSF tau, p-tau, Aβ42 and Aβ oligomers concentrations in Alzheimer's disease (AD) patients.

Patients and methods: We recruited 14 patients with AD and 8 patients with non-AD conditions from the Memory Clinic of Queen Mary Hospital. CSF samples collection by lumbar puncture and cognitive assessment were done at baseline. We did CSF tau, p-tau, Aβ42, and Aβ oligomers proteins assays with commercial ELISA kits.

Results: We found that AD patients had higher levels of CSF tau and p-tau, but lower levels of Aβ42 level than non-AD patients (p = 0.014, 0.003, 0.034, respectively; Mann-Whitney U test). Moreover, AD patients had lower Aβ42/tau and Aβ42/p-tau181 ratios than non-AD patients (p = 0.001 for both). High CSF Aβ

oligomer levels (>1.98 pmol/L) were more common in AD than non-AD patients (35.7% versus 0 %; $p = 0.021$, Chi-square statistics). In ROC curve analyses, the area-under-curve (AUC) of CSF levels of A β 42, tau, p-tau181, A β 42/tau and A β 42/p-tau ratios ranged from 0.777 to 0.929. For the diagnosis of AD, the ratio of A β 42/tau had high sensitivity and specificity of 86% and 88%, respectively. The A β 42/p-tau ratio was the most specific for the diagnosis of AD (100% specificity), while retaining a good sensitivity of 79%.

Conclusion: In conclusion, the A β 42/tau and A β 42/p-tau ratios were sensitive and specific diagnostic biomarkers of AD in our Chinese population.

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Abstract – WCN 2013

No: 1657

Topic: 5 – Dementia

Actions to stop driving in different types of dementia: Data on 8850 patients from the Swedish dementia quality registry (SveDem)

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Background: Executive functions impairment, visuospatial dysfunction, behavioural disturbances and problems with orientation affect fitness to drive in dementia patients. We investigate if the reporting of driving unfitness by physicians differs with different dementia diagnoses.

Patients and methods: Totally, 8850 patients from SveDem were included. The registry holds information on 8 different dementia diagnoses (ICD-10). The primary outcome was agreement with patient to quit driving or report the patient to the driving license authority with a recommendation of license suspension. Variables such as age, gender and type of dementia were considered in the model.

Results: The mean age was 76.6 (8.0) years with a majority (55.1%) of males. An agreement with the physician not to drive was obtained in 80% of cases. In 705 (8.7%) cases the patients were reported to the Swedish Transport Agency. The highest and lowest rates of reporting were recorded in frontotemporal dementia (FTD) (23.3%) and dementia with Lewy bodies (DLB) (4.9%), respectively. After controlling for age, MMSE score ($P = 0.002$) and gender ($P = 0.001$), FTD [$P = 0.015$], DLB ($P = 0.004$) and Parkinson's disease with dementia (PDD) ($P = 0.014$) were significantly associated with driving license suspension.

Conclusion: SveDem data demonstrated that for the majority of dementia patients an agreement with the physician not to drive was reached. In less than 9% the driving license was reported to authorities. Of those, the majority of patients suffered from FTD. The loss of disease awareness in these patients may contribute to difficulties in reaching an agreement not to drive resulting in suspension of the driving license.

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Abstract – WCN 2013

No: 1554

Topic: 5 – Dementia

Correlation between epileptiform activity and cerebrospinal fluid biomarkers in Alzheimer's disease

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Background: Alzheimer's disease (AD) is associated with an increased incidence of unprovoked seizures. Epileptiform activity in AD has long been recognized as a secondary process resulting from advanced stages of neurodegeneration and aging-related cofactors; actually some evidence suggests that seizures may contribute to cognitive decline in AD.

Objective: The aim of the study was to investigate the relationship between the qualitative electroencephalographic pattern and cerebrospinal fluid biomarkers (CSF) in AD patients.

Methods: 141 outpatients with AD underwent a neuropsychological examination, conventional EEG recording and lumbar puncture for assessing tau protein, phosphorylated-tau protein (p-tau) and Ab₁₋₄₂ amyloid protein values. According to a visual qualitative EEG analysis, all participants were assigned to the following categories: Type 1. normal; Type 2. slow; Type 3. epileptiform.

Results: 37 patients showed the type 1 EEG pattern, 59 the type 2 and 45 the type 3. Statistical analysis indicated that patients with epileptiform EEG pattern have p-tau and tau/Ab₁₋₄₂ ratio values significantly higher than patients showing type 1 EEG pattern. This finding was independent from age, estimated disease duration and MMSE values of each EEG group. A correlation between the progressive reduction of MMSE, and respectively, the increase of both tau and p-tau values, and the decrease of Ab₁₋₄₂ was also found; patients with type 3 epileptiform EEG pattern showed a significant correlation between the reduction of Ab₁₋₄₂ and the decrease of MMSE.

Conclusions: Our findings support the hypothesis that aberrant excitatory neural activity may represent a primary upstream mechanism in the genesis of cognitive decline. Therefore, qualitative EEG analysis integrated with CSF biomarkers may be a valid tool to define prognosis of AD and possible therapeutic implications.

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Abstract – WCN 2013

No: 1688

Topic: 5 – Dementia

Symptoms and progression of Alzheimer's disease among South-East Asian population: A Pakistani study

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Background: Alzheimer's disease is one of the emerging health issues all around the world involving both the developed as well as the developing countries.

Objective: The aim of our study was to determine the prevalence and clinical correlates of extrapyramidal signs in outpatients with probable Alzheimer's disease.

Methods: This study was conducted at the Peshawar Medical College teaching hospital Pakistan over a period of 1 year from August 2011 till July 2012. A total of 38 patients presented in the medicine outpatient departments having symptoms of the disease and meeting WHO diagnostic criteria for probable Alzheimer's disease were recruited in the study.

Results: 10% of the neuroleptics-free patients were free of extrapyramidal symptom. The most common types of extrapyramidal symptom presented in our patients were hypomimia (54%), difficulty in talking (49%), bradykinesia (43%), postural instability (36%), abnormal gait (29%), and rigidity (21%), respectively. The mean duration of the disease in patients appearing with extrapyramidal symptom was found to be 2.12 ± 1.23 years. The presence of extrapyramidal symptom increases proportionally with the progression of the disease and cognitive and functional decline.

Conclusions: After controlling for duration of the disease, the use of neuroleptics is found to influence the appearance of extrapyramidal symptom in patients with Alzheimer's disease. Patients who presented extrapyramidal symptom at initial examination appeared to deteriorate faster, mainly cognitively, but also functionally.

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Abstract — WCN 2013

No: 919

Topic: 5 — Dementia

Rationale and study design of a MRS study to explore the effects of Souvenaid on brain metabolites in Alzheimer's disease

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Background: Synaptic dysfunction is a major contributing factor to the development of cognitive impairment in individuals with Alzheimer's disease (AD). Synapses consist of neuronal membranes largely composed of phospholipids. The formation of neuronal membranes is controlled by nutritional precursors and cofactors that are obtained from the circulation. The medical food Souvenaid®, containing the specific nutrient combination Fortasyn® Connect (FC)¹, has been formulated to support synapse formation and function in patients with AD by promoting phospholipid and membrane formation. It has been demonstrated that Souvenaid improves memory in mild AD patients.

Objective: The main objective of the present Magnetic Resonance Spectroscopy (MRS) study is to investigate to what extent Souvenaid affects brain phospholipid metabolism.

Patients and methods: A total of 30 drug-naïve patients with mild AD (MMSE ≥ 20) will be enrolled in this double-blind randomised controlled study (NTR3346) to receive either Souvenaid (containing FC) or an isocaloric control product (without FC) for a 4-week period. At baseline and after 4 weeks, participants will undergo phosphorus (³¹P)-MRS measurements to study phospholipid turnover. Additional proton (¹H)-MRS measurements will be conducted to assess neural integrity. Main outcome parameters are brain levels of the phosphomonoesters phosphocholine and phosphoethanolamine, and of the phosphodiester glycerophosphocholine and glycerophosphoethanolamine.

Results: The enrollment for the study started in October 2012. Currently, fifteen patients have been randomised.

Conclusion: The effects of Souvenaid on brain phospholipid metabolism are investigated using ³¹P-MRS. Furthermore, ¹H-MRS measurements will provide additional information on its effects on neural integrity.

¹Souvenaid and Fortasyn are registered trademarks of N.V. Nutricia.

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Abstract — WCN 2013

No: 1591

Topic: 5 — Dementia

In vivo visualization of tau pathology in Alzheimer's disease patients by [¹¹C]PBB3-PET

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Background: Neuroimaging results of brain amyloid beta by PET have provided many novel insights in the pathomechanisms of Alzheimer's disease (AD). Tau is another pivotal protein associated with neural dysfunction in AD, and *in vivo* tau estimation would be an important biomarker for AD research. We have developed a novel PET radioligand [¹¹C]PBB3, which shows high affinity and selectivity for tau deposits.

Objectives: To investigate the spatial characteristics of [¹¹C]PBB3 and PET in human with both normal and impaired cognition.

Patients and methods: Participants were patients with AD and age-matched healthy controls (HCs). [¹¹C]PBB3 was intravenously injected and dynamic scans were acquired over a 70 min. Standardized uptake value ratio (SUVR) image was created by the cerebellar cortex as reference region, and visually assessed in each subject. Additionally, cerebral amyloid deposition was estimated with amyloid-binding agent, [¹¹C]PIB and SUVR images were created from 50–70 min post-injection PET scan.

Results: All HCs were [¹¹C]PIB-negative, and all AD patients were [¹¹C]PIB-positive. SUVR images of [¹¹C]PBB3-PET demonstrated high accumulation of [¹¹C]PBB3 in the medial temporal cortex of all AD patients, in contrast with relatively low [¹¹C]PIB binding in the same region. Distribution of [¹¹C]PBB3 binding observed in AD patients extended to the limbic system and subsequently to the neocortex as a function of the disease severity.

Conclusions: The [¹¹C]PBB3 binding region in AD patients is in accord with the prior neuropathological findings and was distinct from [¹¹C]PIB binding region. Therefore, we conclude that [¹¹C]PBB3-PET can effectively visualize human brain tau deposition *in vivo*.

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Abstract — WCN 2013

No: 1596

Topic: 5 — Dementia

Socioeconomic burden of young onset dementia in Singapore — A pilot study

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Background: The socioeconomic cost of dementia is significant for patients, caregivers and society. Few studies have examined the impact of young onset dementia (YOD) in Asia, and none in Singapore.

Objectives: To determine the socioeconomic impact of YOD in Singapore.

Methods: A random sample of YOD patients (age ≤ 65 years old) attending a memory clinic in a tertiary setting was identified. Diagnosis of dementia had been made by a cognition neurologist.

2 independent physicians conducted phone interviews with patients and their caregivers on separate occasions using a set of questionnaire on financial burden. Case-notes were reviewed for patients' demographics and medical history.

Results: 12 patients (5 Alzheimer's dementia; 5 fronto-temporal dementia; 1 vascular dementia; 1 Hashimoto's-associated dementia) and 10 caregivers were interviewed. Mean age of patients was 59 (51–64); mean MMSE 18 (9–27). 7 patients reported premature loss of employment with average loss of employment of 4.6 years. This resulted in an estimated average monthly income loss of SGD\$2992 (\$0.1–6.5 k/month).

Caregivers reported varying degrees of indirect income loss including reductions of working hours. Most were unable to quantify exact

income loss; 1 reported a monthly loss of SGD\$4000 resulting from a job change.

Direct monthly medical cost averaged SGD\$480 (medication \$142; medical consultations \$85; out-of-pocket expenses \$253). Majority expressed a need for financial aids for medications and care.

Conclusion: Patients and caregivers of YOD are burdened with significant direct and indirect costs. Further studies are underway to exact nationwide socioeconomic burden, and rationalize resource allocation for YOD in Singapore.

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Abstract – WCN 2013

No: 1543

Topic: 5 – Dementia

The etiology of moderate cognitive disorders (MCD) in a working-age population according to epidemiological studies

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Background: Early diagnosis of pre-dementia contributes to the timely administration of pathogenetic therapy to delay or stop the progression of cognitive functions disorders.

Objective: To evaluate the prevalence and etiology of the MCD syndrome in the working-age population of Ulyanovsk.

Materials and methods: In the framework of the study the open population of Ulyanovsk in the age of 40–59 years underwent screening. MCD syndrome was determined in accordance to the modified R.Petersen and J.Touchon diagnostic criteria; the etiology of MCD syndrome was defined by anamnesis data, the results of the clinical examination, ultrasonic research of cerebral vessels, and brain MRI.

Results: We surveyed 300 people: 86 (15,93%) men, 214(25,78%) women. MCD syndrome was diagnosed in 71(23,67%) subjects. In 48(67,61%) patients the cause of this syndrome were vascular diseases (hypertension, and atherosclerosis), confirmed by ultrasound duplex scanning of brachiocephalic arteries. 7 patients (9,86%) had cerebral vascular disorders and diabetic encephalopathy. In 4 (5,63%) cases MCD syndrome was a consequence of vascular diseases and craniocerebral injury. 5 patients (7,04%) had a cerebrovascular disease and memory weakness, which was associated with an operation, performed under general anesthesia. There were isolated cases: posttraumatic encephalopathy, paraneoplastic syndrome, diabetic encephalopathy, a combination of vascular causes and myasthenia gravis in remission, a combination of such factors as hypertension and consequences of viral encephalitis.

Conclusions: The MCI syndrome was diagnosed in 20% of the working-age (40–59 years) population of Ulyanovsk. The main etiological factors for the development of the syndrome were hypertension and atherosclerosis.

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Abstract – WCN 2013

No: 1552

Topic: 5 – Dementia

Early-onset dementia (EOD). Demographic and etiologic characteristics of 170 patients

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The purpose is to investigate demographic profile and etiology distributions of EOD in a prospective memory centre cohort.

401 cases of dementia were diagnosed between January 2000 and May 2011; 170 were EOD, with age onset less than 65 years. Dementia diagnosis was based on complete neurologic and somatic clinical examination, and neuropsychological assessment. All patients had cerebral imaging. Routine laboratory tests were performed in all cases. More analysis was made when a specific etiology was suspected. We used DSM-IV criteria for diagnosis of dementia, NINCDS-ARDA criteria for Alzheimer's Disease (AD), NINDS-AIREN criteria for Vascular Dementia (VaD), and the usual criteria for diagnosis of Lewy Body and Fronto-Temporal Dementia (FTD), Progressive Supranuclear Palsy and Cortico-Basal Dementia.

EOD represented 42.4% of dementias, with 90 men and 80 females (range, 21 to 64 years). The mechanism of dementia was degenerative in 78 cases, including 61 AD (36% of EOD cases) and 8 FTD, vascular in 33 cases (19,4%) with 8 cases of Sneddon's Syndrome, infectious in 26 cases (15,3%) with 19 cases of Neuro-Syphilis and 4 cases of Creutzfeldt-Jakob disease, and inflammatory in 11 (including 4 cases of Neuro-Behcet). Other etiologies were found in 16 cases, and the cause was undetermined in 5 cases.

Our study shows a large diversity of etiologies in EOD, that differ in accordance with age onset. Before 50 years, we find essentially infectious and inflammatory diseases. After 50 years, AD and VaD are the most frequent. EOD needs exhaustive investigations for a rapid diagnosis and potential treatment.

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Abstract – WCN 2013

No: 326

Topic: 5 – Dementia

Application of AD7C-NTP ELISA kit on Alzheimer's disease diagnosis

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Objective: To develop urine AD7C-NTP diagnostic kit, analyze and evaluate clinical application on AD.

Methods: Immunogenicity AD7C-NTP peptide fragments had synthesized by solid-phase methods, then immunized animals and prepared antibodies. After matching screening, mouse antibody as packets antibody, while biotin-labeled rabbit antibody as testing antibody, horseradish peroxidase labeled avidin, the urine AD7C-NTP ELISA detective method was established. The morning urine samples of 121 AD patients and age-matched control group of 118 were collected.

Results: AD7C-NTP antibodies were identified. Mouse anti-AD7C-NTP antibody ELISA titer was 1:8000, and rabbit anti-AD7C-NTP antibody ELISA titer is 1:32,000; WB detected human brain specimens found a single band. In the relative molecular mass of 41,000, the sensitivity of ELISA methodology was 0.5 µg/L, the linear range was 0–10 µg/L, normal reference value of ≤1.5 µg/L, the average recovery rate was 100.2%, approved of CV 3.8%, 4.5%, and inter CV 7.6%, 6.8%. AD group [2.250(0.43–8.62) µg/L] was higher than control group [0.82(0.47–2.77) µg/L, P < 0.0001]. The positive rates of AD group and control group were 89.3% and 15.3% respectively, the sensitivity was 89.3% and specificity was 84.7%.

Conclusions: Using the self-designed synthetic peptide fragment and immunizing animals, preparing AD7C-NTP antibodies successfully, establish an ELISA detective method of urine AD7C-NTP on AD diagnosis. This ELISA diagnostic kit is high precision and sensitive, and can be used as an assistant examination in clinical diagnosis of AD.

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Abstract – WCN 2013**No: 1540****Topic: 5 – Dementia****Voltage-gated potassium channel antibody encephalopathy mimicking Creutzfeldt–Jakob disease**

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Introduction: Rapidly Progressive Dementias (RPDs), in which the sporadic Creutzfeldt–Jakob disease (CJD) is the prototype, requires an urgent and extensive workup. Voltage-gated potassium-channel encephalopathy (VGKC-E) is an auto-immune disorder with variable clinical presentations, including a RPD that mimics CJD.

Case report: A 75 year-old female presented with 6 months history of progressive cognitive abnormalities dominated by behavior, orientation and memory dysfunction with unsteady gait. She also presented continuous diarrhea and later developed myoclonic and generalized tonic-clonic seizures. Past medical history was irrelevant.

Neurologic examination revealed a complete dysfunction in multiple cognitive domains (orientation, attention, immediate/recent memory, calculus, writing, construction ability, insight/judgment and abstract thinking – MMSE-Score 2/30), apathy, frontal release signs, axial and appendicular rigidity, retropulsion falls and myoclonic jerks involving face and limbs. Laboratory screening showed hyponatremia. Anti-neuronal antibody testing came positive for VGKC-antibody LGI1 (Leucine-rich-glioma-inactivated-1). CSF analysis revealed mild lymphocytic pleocytosis, mild proteinoraquia and negative 14-3-3 protein.

Brain-MRI showed mesencephalic and anterior temporal lobe atrophy. EEG revealed left fronto-temporal slow and paroxysmic activity.

The patient was treated with high-dose methylprednisolone and intravenous immunoglobulin achieving a favorable recovery (MMSE-Score 22/30, resolution of diarrhea and hyponatremia).

Discussion: RPD remains one of the most challenging neurologic entities. Despite presenting criteria for CJD, the positivity for a VGKC-antibody (LGI1) and immunotherapy-response confirmed an immunologically-mediated dementia. Because VGKC-E is frequently misdiagnosed as a neurodegenerative or prionic disease it remains unrecognized. We present this case to emphasize the importance of VGKC-antibodies testing in the studying of RPD, thus treating what was apparently untreatable.

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Abstract – WCN 2013**No: 548****Topic: 5 – Dementia****The effect of physiological aging on spatial navigation in a computerized human analog of the Morris Water Maze**

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Objectives: Our recent findings derived from a real-space human analog of the Morris Water Maze (hMWM) suggested that physiological aging without pronounced hippocampal atrophy or vascular changes is associated with mild spatial navigation decline, which is less profound than in Alzheimer's disease (AD). The aim of this

study was to characterize spatial navigation performance and spatial learning in two different periods of the old age with an inexpensive and easy-to administer computerized hMWM.

Methods: Healthy participants without cognitive deficit on standard neuropsychological testing: 24 young (18–26 years), 24 young-old (60–70 years) and 20 old-old (71–84 years) without vascular changes and pronounced hippocampal atrophy on brain MRI underwent spatial navigation testing in the computerized hMWM, which has the advantage to test separately hippocampus dependent (allocentric) and parietal lobe dependent (egocentric) navigation.

Results: The old-old group was impaired in allocentric navigation compared to the young ($p \leq .001$) and the young-old ($p = .007$) groups. The young and young-old groups did not differ in allocentric navigation ($p = .077$). There were no differences in egocentric navigation among the groups ($p's \geq .189$). The learning effect was preserved in all groups ($p's \leq .001$) with no between-group differences in learning ($p \geq .180$).

Conclusions: Our results in the computerized hMWM supported our previous findings from the real-space setting that allocentric spatial navigation decline is age dependent unlike the egocentric and is manifested after the age of 70. Spatial learning seems to be preserved in contrary to prodromal AD, which makes this test be a useful screening tool for evaluation of individuals at risk of AD.

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Abstract – WCN 2013**No: 1750****Topic: 5 – Dementia****Healthcare professionals judgement of presence or absence of delirium using the 4at and CAM tools – A survey**

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Background: All adult patients with risk factors admitted to hospitals in the UK should be screened for delirium with a focus on preventing delirium (NICE). The diagnosis of delirium should be made by a trained healthcare professional using a validated tool such as confusion assessment method(CAM). The 4 As test (4AT) is a new screening tool for delirium and cognitive impairment and can be administered by any healthcare professional without the need for training. We surveyed the healthcare professionals in our hospital on their judgement of absence/presence of delirium using the short CAM and 4AT tools and sought their preference for wider use in hospital.

Methods: We devised 2 hypothetical patient histories with hypoactive and hyperactive delirium including essential data for application of the tools. We provided healthcare professionals from various subspecialties without any specific training on delirium these questionnaires along with the 4AT and CAM tools and asked them to judge for the presence or absence of delirium.

Results: 39 healthcare professionals completed the survey, 34 of the 39 questionnaires were completed fully while the remaining 5 were completed partially. Hypoactive delirium was correctly identified by 84% using the 4AT and by 97% using the short CAM. Hyperactive delirium was correctly identified by 78% using the 4AT and by 50% using the short CAM. There was higher preference for 4AT(57%) for wider use as a tool for screening.

Conclusions: Our survey shows that there is need for training of healthcare professionals in the application of tools for screening delirium.

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Abstract – WCN 2013**No: 1048****Topic: 5 – Dementia****Dementia diagnoses in the young are based on inadequate diagnostic evaluations – A nationwide register-based study**

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Background: The aim of this nationwide register-based study was to evaluate the quality of the diagnostic work-up in young patients registered with a diagnosis of dementia in routine clinical practice.

Methods: Two hundred patients were randomly selected from 891 patients aged ≤65 years registered with a dementia diagnosis for the first time in 2008. Through medical record review, 3 raters evaluated the completeness of the work-up on which the dementia diagnosis was based, using evidence-based dementia guidelines as reference standards. Data were compared to results from a previous similar study in the elderly Danish population.

Results: In total, 159 medical records were available for review. One third of the patients had been diagnosed with dementia during an acute admission. In total, dementia diagnosis was confirmed by the raters in 48% while 60% of dementia subtypes were confirmed. Thirty percent of the patients did not meet diagnostic criteria for dementia. An acceptable diagnostic work-up of all items was performed in only 19 patients. Most of the patients had been diagnosed with dementia by neurological or psychiatric departments, but supplementary diagnostic tools were only rarely used.

Conclusion: Our findings indicate that the diagnostic work-up of dementia in young patients in the secondary health care sector may not meet the basic standards set by evidence-based clinical guidelines. Consequently, patients may be either over-diagnosed with dementia or denied the treatment and care they need for their specific neurodegenerative disorder. Our findings call for increased competency in diagnosing dementia in young patients in the future.

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Abstract – WCN 2013**No: 524****Topic: 5 – Dementia****Safety and efficacy of ORM-12741 on cognitive and behavioral symptoms in patients with Alzheimer's disease: A randomized, double-blind, proof-of-concept study**

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Background: ORM-12741 is a highly potent and selective alpha-2C adrenoceptor (AR) antagonist that has demonstrated efficacy in rodent models suggesting beneficial effects on both cognitive and behavioral symptoms in AD.

Objectives: To evaluate safety, tolerability and efficacy of ORM-12741 as add-on therapy in patients with Alzheimer's disease (AD).

Methodology: This was a phase IIa, randomized, double-blind, placebo-controlled, parallel-group, multicentre, proof-of-concept study of 100 moderate AD patients (MMSE scores 12 to 21) with behavioral symptoms (Neuropsychiatric Inventory (NPI) score of ≥15). Patients were randomized to two flexible dose levels of either 30 to 60 mg or 100 to 200 mg of ORM-12741 or matching placebo twice daily for 12 weeks as add-on to their stable cholinesterase

inhibitor therapy (± memantine). Efficacy was assessed primarily by computerized cognitive tests from the CDR System. The NPI was used to assess behavioral and psychological symptoms.

Results: Statistically significant treatment effects were noted for ORM-12741 on the composite CDR measures Quality of Episodic Memory (p = 0.03) and Quality of Memory (p = 0.013) as well as on NPI Caregiver Distress (p = 0.034) favoring ORM-12741 over the 12 week treatment period with no significant differences between the two dose groups. However, a positive trend was noted for both Quality of Working Memory and NPI total score favoring the low dose group. ORM-12741 was generally well tolerated in the study.

Conclusion: Significant positive effects of ORM-12741 on several composite measures of memory in moderate AD patients as add-on therapy over 12 weeks were observed suggesting further study in longer term trials.

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Abstract – WCN 2013**No: 1741****Topic: 5 – Dementia****High density lipoprotein cholesterol in patient conversion with MCI in dementia**

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Objectives: The aim of this study was to investigate the impact of HDL-cholesterol in conversion to dementia for MCI patients.

Material and methods: We recruited 365 patients in families with medicine cabinets, patients with age of over 65 years, who met the criteria for MCI and evaluated their global cognitive performances at baseline and annual for the next three years. The diagnosis of dementia was established using DSM IV TR criteria.

Results: There were 365 cases of MCI incident, 194 male patients and 171 female patients. The mean conversion rate during follow-up period was 18.9% for Alzheimer's disease (AD), 11.8% for vascular dementia (VD) and 13.2% for Alzheimer's disease with cerebrovascular disease (AD with CVD). In our study, the relative risk of conversion of the patients with low HDL-C was 1.67 in dementia, 2.56 in AD, 4.25 in VD and 2.67 in AD with CVD; for the patients with normal HDL-C was 0.41 in dementia, 1.10 in AD, 0.12 in VD and 0.27 in AD with CVD; and for high HDL-C patients was 1.52 in dementia, 1.48 in AD, 2.21 in VD and 2.13 in AD with CVD.

Conclusion: Conversion in dementia was associated, in our study with low HDL-C. The high values of HDL-C associate a low risk of conversion in AD of patients with MCI comparative with conversion in VD and AD with CVD; in patients with unconverted MCI the normal HDL-C is dominant.

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Abstract – WCN 2013**No: 1610****Topic: 5 – Dementia****Memory aid – Computer based working memory training in elderly with mild cognitive impairment (MCI). A randomized, controlled trial**

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Background: Mild cognitive impairment (MCI) is a condition characterized by memory problems more severe than normal cognitive changes due to old age, and less severe than dementia. Reduced working memory (WM) is regarded as one of the core symptoms of an MCI-condition. Recent studies have indicated that WM can be improved through computer based training.

Objective: The objective of the study is to evaluate if working memory training is effective in improving the working memory in elderly MCI-patients. Further, to evaluate if cognitive training relates to structural changes in the white and gray matter of the brain, assessed by structural MRI.

Patients and methods: The proposed study is a blinded, randomized and controlled trial that will include 90 elderly patients from a Memory Clinic diagnosed with MCI. The groups will be randomized to either training or a placebo version. The intervention is computerized working memory training performed for 45 min over 25 sessions. Neuropsychological assessment and structural MRI will be performed before, after and at 6 months after training.

Results/conclusion: Currently there is no known treatment available for MCI, and few studies on specific cognitive training in MCI-patients have been performed. The proposed study has received funding and starts in September 2013. If computer based training results in positive changes to memory functions in MCI patients this may represent a new, cost-effective treatment. Secondly, evaluation of training induced structural changes to grey or white matter may improve our understanding of the mechanisms behind effective cognitive interventions in MCI patients.

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Abstract – WCN 2013

No: 1835

Topic: 5 – Dementia

Clinical and genetic aspects of Moroccan patients with Alzheimer's disease

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Background: Alzheimer's disease (AD) is the most common cause of dementia in the elderly. In Morocco it affects almost 30000 persons (WHO).

In this first study in Morocco we aimed:

- At evaluating the clinical aspects of Moroccan patients with AD.
- At evaluating the genetic contributions of APP exon 16 and exon 17 mutations in familial and sporadic cases.

Patients and methods: 6 families (8 cases) and 17 sporadic cases presenting at neurology department of the CHU IBN ROCHD, Casablanca, Morocco.

- Clinical methods: Cognitive function assessment, brain imaging and laboratory tests.
- Genomic methods: Genomic DNA extracted from blood. PCR and direct sequencing of the APP exons 16 and 17 were done.

Results:

- Clinical results: All patients had cortical atrophy; 48% of the patients that are either sporadic or family forms have a MMSE score of less than 10 and are affected by severe dementia, 28% by a moderately severe dementia and 12% are slightly insane.
- Genetic results: In APP exon 16, 2 novel frameshift mutations were detected in 4 family cases. No mutations in APP exon 16 have been

found in 17 sporadic cases. In APP exon 17, in all patients, 5 novel frameshift mutations were identified. Only one novel splicing mutation was detected.

Conclusion: High correlation between imaging and neuropsychological testing. The more the cortical atrophy is prominent, the more the neuropsychological performances are reduced. Moroccan patients with AD have novel frameshift mutations that induced a complete alteration of the amino acid sequence of a protein, which may lead to disease.

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Abstract – WCN 2013

No: 1483

Topic: 5 – Dementia

Suppression of spike frequency by α -synuclein oligomer in neocortical pyramidal neurons

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Background: Dementia with Lewy bodies (DLB) is pathologically characterized by the formation of Lewy bodies and neuronal death throughout the brain. α -Synuclein is a major component of Lewy bodies, and is proposed to play a central role in the pathogenesis of DLB. Recent evidence implicates soluble oligomers rather than insoluble fibrils as the toxic species. These data suggest the hypothesis that soluble α -synuclein oligomers could accumulate intracellularly and alter neuronal activities; however it has not been elucidated.

Objective: To investigate the effect of α -synuclein on neuronal excitabilities in neocortical pyramidal cells.

Material and methods: α -Synuclein protein was introduced into pyramidal neurons through whole-cell patch pipettes in mouse frontal cortex slices. The following four kinds of internal solution are applied; α -synuclein (α -SN; 1 μ M), dopamine (DA; 10 μ M), α -synuclein co-incubated with dopamine at 37 °C for 3 days (α -SN + DA), and vehicle solution.

Results: The application of α -SN + DA reduced spike firing by depolarizing current injection. Neither resting membrane potential, spike width, nor spike afterhyperpolarization had differences between these groups. Western blot analysis revealed that α -SN + DA included higher order oligomers compared with α -SN.

Conclusion: α -Synuclein oligomers lower spike frequency, which may cause the downregulation of neocortical activities in DLB.

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Abstract – WCN 2013

No: 1818

Topic: 5 – Dementia

Isolated frontotemporal dementia in valosin containing protein gene mutations

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Background: Frontotemporal dementia with inclusion body myopathy and Paget's disease of bone is a rare, autosomal dominant disorder caused by mutation in the gene valosin-containing protein (VCP). It seems that only 3% have FTD as an isolated phenotype.

Case report: A 52-year-old woman dedicated housewife progressively lost interest in her child and husband, making dinner and eating

without waiting for them or speaking with them. Family reported other abnormal behaviors: she started going to funerals of people she didn't know, trying to look at the dead's face; she entered every bathroom that she saw to wash her hands. Her family noticed speech problems in finding names and after 2 years her speech became incomprehensible. She developed swallowing problems and urinary incontinence. 10 years later she was referred to our hospital. Past personal and family histories were irrelevant. Examination revealed a profoundly apathetic and indifferent woman with repetitive motor behavior (raising from the chair), frontal release signs, retrocollis, asymmetric parkinsonism, useless left arm, and general atrophy without fasciculations. MRI showed important frontotemporal atrophy. EMG was normal but she wasn't able to collaborate. Paget's disease was excluded. Genetic test revealed a VCP mutation not described yet but probably pathogenic (c.811 + 2 T>C, intron 7). We are waiting for familial consents to perform genetic tests and muscular biopsy to exclude myopathy.

Discussion: This is the first Portuguese VCP mutation with a detailed clinical examination.

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Abstract – WCN 2013

No: 1814

Topic: 5 – Dementia

Fluidity changes in the forebrain membranes isolated from mice bearing experimental Alzheimer's disease

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Background: Membrane structure may play a mean role in the development of dementia. Therefore, it was important to determine membrane fluidity alterations for the development of AD-like pathology. **Objectives:** In this work changes in fluidity of membranes isolated from the microsomal and synaptosomal fractions obtained from forebrain of mice bearing experimental Alzheimer's disease induced with olfactory bulbectomy and developed by I. Nesterova et al. [1] were studied.

Materials and methods: Membrane microviscosity was measured by electron paramagnetic resonance (ESR) with 2,2,6,6-tetramethyl-4-capryloyl-oxypiperidine-1-oxyl as lipidic and 5,6-benzo-2,2,6,6-tetramethyl-1,2,3,4-tetrahydro- γ -carboline-3-oxyl as near-proteinic probes. Rotational diffusion correlation time, characterizing membrane components microviscosity, was calculated from the obtained spectra of the ESR.

Results and conclusions: We found out staged changes of forebrain cell membranes fluidity, accompanying AD. These alterations correlate with the stages of «clinical» indicator changes. The main disturbance of the fluidity took place in membranes of near-protein regions. The obtained results testify the lipid peroxidation regulation system failures. Thus, the membrane structure plays an important role in the development of AD, and, taking into account these structural changes, one can better understand the course of dementia, and significantly improve the disease therapy. It was revealed that the membrane structural state was disturbed already at an early stage of experimental pathology development, modeling of AD. Hence, lipid bilayer fluidity changes can be used for early diagnostics of Alzheimer's disease.

We believe that our results confirm the validity of Alzheimer's disease model based on olfactory bulbectomy proposed by N.V. Bobkova.

References

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Abstract – WCN 2013

No: 1815

Topic: 5 – Dementia

Microtubule-associated tau protein serum level in patients with different neurodegenerative diseases

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Microtubule-associated axonally-derived phosphoprotein tau is essentially involved into neurotoxic degenerative mechanisms: aggregation of its specific sets into filamentous inclusions is a common feature of intraneuronal and glial fibrillar lesions in numerous neurodegenerative disorders: Alzheimer's disease and Tauopathies. Tau-associated genes, proteins and Tau-phosphorylation were also abnormal in some forms of multiple sclerosis (MS). The aim was to compare tau level in patients with temporal lobe epilepsy (TLE), relapsing remitting MS (RRMS) and amyotrophic lateral sclerosis (ALS).

Method: Patients with drug-resistant long-standing TLE (n = 49; 20–41 years), RRMS (n = 38; 27–46), ALS (n = 5; 43–58) and controls (n = 16; 25–35) were investigated. Tau was measured by immunofluorescent method using monoclonal anti-mouse antitau-2 antibodies also specific for phospho-Tau. Results were expressed as optical density (OD = Log₁₀F₀ / F₁) units of FITC-labelled binding sites.

Results: Serum tau concentration (mean ± SD) under baseline conditions compare to controls was in TLE = 0.38 ± 0.041 vs. controls = 0.04 ± 0.002; RRMS = 1.02 ± 0.085 (P > 0.01; t = 4.69); ALS = 0.58 ± 0.06 (P > 0.01; t = 3.8). TLE that showed average tau levels had long-standing pharmaco-resistant course (mean duration ± SD: 18.5 ± 11.6) with psychiatric complications using old generation antiepileptic drugs where tau might reproduce secondary neurodegeneration process associated with epileptic lesion. RRMS and ALS (mean duration ± SD: 7.5 ± 2.23 and 14.2 ± 8.31, respectively) detected high tau-mediated toxicity associated in RRMS with relapses severity, and in ALS associated with cognitive decline progression as a part of tau-mediated neurodegeneration; eighteen RRMS patients formerly had brain injury (10), meningo/encephalitis virus (7) and chicken-pox (1).

Conclusion: Study revealed high level of hyperphosphorylated tau in RRMS and ALS where it plays role in neurodegeneration associated with diseases severity and progression.

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Abstract – WCN 2013

No: 1820

Topic: 5 – Dementia

Characteristics of Parkinson's disease dementia in Southern Tunisia

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Objective: To determine the prevalence, characteristics and factors correlated with the occurrence of dementia in patients with Parkinson's disease (PD) in Southern Tunisia.

Patients and methods: We recruited and examined 102 unrelated Tunisian patients with sporadic and familial PD. The mean age at onset was 64.9 years (range = 40–86) and 57.8% were men. Neuropsychological evaluation was conducted and included the Unified Parkinson

Disease Rate Scale I “UPDRS I”, Mini Mental State Evaluation “MMSE” and Frontal Assessment Battery “FAB”.

Results: Cognitive impairments were reported by 39.2% patients or their families as a symptom of the disease. The frequency of cognitive impairment was variable depending on the scale used. Based on the UPDRS I, cognitive impairments were present in 62.75%: mild to moderate in 55.88% and severe in 6.86%. Global cognitive efficiency was impaired in 36.26% while dysexecutive syndrome was frequent (56.86%) even in some patients showing normal intellect. There was no correlation between cognitive impairments and some clinical and epidemiological features, such as duration of the disease, age at onset, and the nature and dose of the treatment used. The occurrence of cognitive impairment was strongly correlated with the presence of depression ($p = 0.001$) and an advanced stage of the disease (Hoehn and Yahr > 2.5 , $p = 0.002$).

Conclusion: We conclude that cognitive impairments are often underestimated. MMSE is less insensitive than FAB to detect cognitive impairments due to PD. Depression and severe disability are the main factors that precipitate cognitive decline in PD.

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Abstract – WCN 2013

No: 1863

Topic: 5 – Dementia

Mirror neurons in Alzheimer's disease

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Mirror neurons are special neurons because they fire both when an action is performed, and when an action is observed. They play a major role in learning, are a key to empathy, and allow social interaction. Is the Mirror Neuron System in Alzheimer's Patients Dysfunctional? Never being published, the answer to this question could open a door to new possibilities in treating Alzheimer's disease by changing the approaches of treatment.

The approach was to use EEG testing. There were two groups, the control group and Alzheimer patients. During an EEG test, each participant was shown a video which consisted of a series of simple movements, followed by faces that expressed various emotions. The participants were first asked to just observe the video, and then when the video was replayed, they were asked to perform the activities.

22 Alzheimer patients and 16 control age matched participants were tested. There was no suppression of the mu range brain wave activity in the Alzheimer's group during observation and performance of the activities. The control group exhibited constant suppression of the mu range brain wave during observation and performance of the activities. These results strongly suggest that both the sensory motors and mirror neurons in Alzheimer's patients are dysfunctional.

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Abstract – WCN 2013

No: 1884

Topic: 5 – Dementia

Influence of vascular risk factors on the evolution of Alzheimer's disease

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Background and objective: Although hypertension, diabetes, dyslipidemia, and congestive heart failure are now considered as vascular risk factors (VRFs) of Alzheimer's disease (AD), it is still unclear that the

VRFs may influence on the effect of treatment with acetylcholine esterase inhibitors (AChEI) in AD patients. We investigated the influence of VRFs on the course of cognitive decline in AD patients.

Subjects and methods: The present study was based on 174 patients (59 men and 115 women) who were diagnosed as having a probable AD according to the NINCDS-ADRDA criteria. Their mean age was 75.8 ± 5.8 years. All patients were on donepezil hydrochloride, and underwent laboratory testings, MRI and neuropsychological evaluation including MMSE. The evolution of cognitive function was evaluated by comparing the MMSE scores between the baseline and 2-year follow-up. The influence of VRFs was analyzed statistically in relation to the evolution in MMSE score.

Results: 122 patients (non-responders) showed deterioration in MMSE score, whereas MMSE score was stable in 52 patients (responders). The baseline MMSE score ($p < 0.01$), body mass index ($p < 0.01$), and HDL-cholesterol negatively correlated with the clinical deterioration. The baseline MMSE score positively correlated with education and alcohol consumption ($p < 0.05$) and negatively correlated with female gender and possession of ApoE4.

Conclusion: The present results indicate that good nutritional status in addition to the efficient management of VRFs may contribute to the stabilization of cognitive function in elderly AD patients who are on treatment with AChEI.

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Abstract – WCN 2013

No: 1056

Topic: 5 – Dementia

Cyclophosphamide-responsive cerebral amyloid angiopathy-related inflammation. A case report and review of the literature

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Background: Cerebral amyloid angiopathy related inflammation (CAA-I) is a rare disease, defined by the deposition of amyloid proteins within the leptomeningeal and cortical arteries associated with vasculitis or perivasculitis. The inflammation typically responds well to steroid therapy.

We report a patient with CAA-I who improved with cyclophosphamide but not with steroid therapy.

Objective: A 78-year-old female was presented with progressive cognitive dysfunction and right hemiparesis. Magnetic resonance imaging (MRI) of the brain revealed high intensity area in bilateral frontal lobe on T2-weighted images (T2WI) and fluid-attenuated inversion recovery (FLAIR) showing leukoaraiosis. Brain biopsy was performed, revealed with no significant findings except edematous change. By the initial steroid pulse therapy, the white matter lesion and her symptoms improved moderately. 3 months later, consciousness disturbance and right hemiparesis worsened again, and the white matter lesion expanded on MRI. Steroid pulse therapy was performed again, but her condition got worse gradually and became akinetic mutism. T2* on MRI showed microbleeding, and analysis of the APOE genotype showed $\epsilon 4/\epsilon 4$. She was diagnosed as CAA-I and two periods of cyclophosphamide pulse therapy were performed. After the first cyclophosphamide pulse therapy the white matter lesion, consciousness disorder, and hemiparesis improved immediately. She was discharged from the hospital fully mobile.

Conclusion: In Japan, although steroid therapy for CAA-I is already known and common, CAA-I which resists steroid therapy should be treated by cyclophosphamide therapy on the early stage of the disease.

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Abstract – WCN 2013**No: 1870****Topic: 5 – Dementia****Common genetic variants are associated with severity of cognitive decline in Korean Alzheimer's disease**

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Background and objective: Recent genome-wide association studies reported evidence that some single nucleotide polymorphisms confer genetic risk for Alzheimer's disease. Whether these risk variants for AD also associated with severity of cognitive functions has not been studied. We aimed to find out correlations between severity of cognitive function and SNPs which were associated with risk of AD.

Methods: We prospectively recruited 211 patients diagnosed as AD from March 2010 to July 2011. They tested detailed neuropsychological evaluation which examined attention, language and related functions, visuospatial functions, verbal and visual memory, frontal and executive functions, K-MMSE, GDS, B-ADL and CDR scale. Percentiles of each neuropsychological test for age- and sex-matched controls in Korea were obtained. 86 SNPs in CR1, BIN1, LRAT, CD2AP, EPHA1, CLU, MS4A, PICALM, ABCA7, CD33 and APOE genes were previously reported to be associated with AD in GWAS. Multiple regression analyses in additive model were performed using SAS version 9.1.3.

Results: In multiple regression analysis adjusting for age at neuropsychological evaluation, sex, disease duration and education, 4 SNPs were identified with P values of $<5.0 \times 10^{-4}$: rs11803956 (CR1) in K-MMSE score, rs2075650 (APOE) in percentile of total score in phonemic fluency and percentile of RCFT copy score, and rs3752232 (ABCA7) in RCFT copy score.

Conclusions: We found significant genetic influences on global and specific cognitive domains. These results that risk variants for AD associated with severity of cognitive decline could strengthen the genetic effect for the pathogenesis of AD.

doi:10.1016/j.jns.2013.07.1211

Abstract – WCN 2013**No: 1942****Topic: 5 – Dementia****New aspects in the study of regional forecasting of post-stroke dementia in Uzbek population of Aral Sea region of Uzbekistan**

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There is a critical ecologically unfavorable situation in the Aral Sea and worsening health outcomes require urgent modern complex of preventive measures.

The purpose – to explore new aspects of regional forecasting of post-stroke dementia.

Material and methods: As part of screening we used standardized methods for detection of post-stroke dementia. All 113 participants were representatives of the Uzbek nationality.

Results: 28 patients suffered an acute ischemic stroke, 17 – hemorrhagic, 41 – transient ischemic attacks, and 27 – with the consequences of stroke. During magnetic resonance tomography (MRT) examination the presence of subcortical lesions of leucoareosis at white substance of the frontal lobes, nucleus basalis (globus pallidum) was revealed. There are 7 factors that determine the

relationship of psychoorganic disorders with comorbid syndromes: F1—is associated with atherosclerosis and hypertension disease; F2—TIA and diabetes; F3—encephalopathy with convulsive syndromes; F4—with the type of cerebrovascular accident; F5—a risk Parkinsonism; F6—ischemic stroke and coronary heart disease; and F7—angiopathic impairments in obesity. Graduation and the numerical values of risk factors were identified After that, according to a formula, the prognostic factors D1 and D2 with biofeedback was determined. During the identification of $D2 \geq D1$, it revealed a high risk of subcortical dementia in 78 participants.

Conclusions: The new approach will increase the accuracy in prediction of post-stroke dementia and help to choose adequate therapeutic tactics.

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Abstract – WCN 2013**No: 1935****Topic: 5 – Dementia****A β augments mCa²⁺ independent mros-mediated shift of lethal transient mitochondrial permeability transition to its permanent mode in narp cybrids**

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Mitochondrial dysfunction is a hallmark of amyloid β -peptide ($A\beta$)-induced neurodegeneration of Alzheimer's disease (AD). This study investigated whether mtDNA T8993G mutation-induced complex V inhibition, clinically associated with neurological muscle weakness, ataxia, and retinitis pigmentosa (NARP), is a potential risk factor for AD and the pathological link for long-term exposure of $A\beta$ -induced mitochondrial toxicity and apoptosis in NARP cybrids. Using noninvasive fluorescence probe-coupled laser scanning imaging microscopy and NARP cybrids harboring 98% mutant genes along with its parental 143B osteosarcoma cells, we demonstrated that $A\beta$ -augmented mitochondrial Ca^{2+} (mCa²⁺)-independent mitochondrial reactive oxygen species (mROS) formation for a cardiolipin (CL, a major mitochondrial protective phospholipid)-dependent lethal modulation of the mitochondrial permeability transition (MPT). $A\beta$ augmented not only the amount but also the propagation rate of mROS induced mROS formation to significantly depolarize mitochondrial membrane potential ($\Delta\Psi_m$) and reduce mCa²⁺ stress. $A\beta$ -augmented mROS oxidized and depleted CL, thereby enhances mitochondrial fission and movement retardation, which promoted the NARP-augmented lethal transient-MPT (t-MPT) to switch to its irreversible mode of permanent-MPT (p-MPT).

Interestingly, melatonin, a multiple mitochondrial protector, markedly reduced $A\beta$ -augmented mROS formation and therefore significantly reduced mROS mediated depolarization of $\Delta\Psi_m$, fission of mitochondria and retardation of mitochondrial movement to stabilize CL and hence the MPT. In the presence of melatonin, $A\beta$ -promoted p-MPT was reversed to a protective t-MPT, which preserved $\Delta\Psi_m$ and lowered elevated mCa²⁺ to sublethal levels for an enhanced mCa²⁺-dependent O_2 consumption. Thus, melatonin may potentially rescue AD patients associated with NARP symptoms.

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Abstract – WCN 2013**No: 1941****Topic: 5 – Dementia****Dual phases of respiration chain defect-augmented mros-mediated mCa²⁺ stress during oxidative insult in normal and rho 0 rba1 astrocytes**

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Mitochondrial respiratory chain (RC) deficits, resulting in augmented mitochondrial ROS (mROS) generation, underlie pathogenesis of astrocytes. However, mtDNA-depleted cells (rho 0) lacking RC have been reported to be either sensitive or resistant to apoptosis. In this study, we sought to determine the effects of RC-enhanced mitochondrial stress following oxidative insult. Using noninvasive fluorescence probe-coupled laser scanning imaging microscopy, the ability to resist oxidative stress and levels of mROS formation and mitochondrial calcium (mCa^{2+}) was compared between two different astrocyte cell lines, control and rho 0 astrocytes, over time upon oxidative stress. Our results showed that the cytoplasmic membrane becomes permeated with YO-PRO-1 dye at 150 and 130 min in RBA-1 and rho 0 astrocytes, respectively. In contrast to RBA-1, 30 min after 20 mM H_2O_2 exposure, rho 0 astrocytes formed marked plasma membrane blebs, lost the ability to retain Mito-R, and showed condensation of nuclei. Importantly, H_2O_2 -induced ROS and accompanied mCa^{2+} elevation in control showed higher levels than rho 0 at early time point but vice versa at late time point. Our findings underscore dual phase of RC-defective cells harboring less mitochondrial stress due to low RC activity during short-term oxidative stress but augmented mROS-mediated mCa^{2+} stress during severe oxidative insult.

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Abstract – WCN 2013

No: 1919

Topic: 5 – Dementia

The epha4-elicited signaling governs the stability of amyloid precursor protein fragments through a Lyn-mediated pathway

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Alzheimer's disease is the most common dementia afflicting the elderly in the modern society. This disease is a result of the neurotoxicity elicited by abnormal amyloid- β ($A\beta$) protein aggregates. $A\beta$ is generated by the γ -secretase-catalyzed proteolysis of amyloid precursor protein (APP) C-terminal fragment (APP- β CTF) that is released by the β -secretase cleavage of APP. Recent evidence suggests γ -secretase's substrate β CTF and its metabolite APP intracellular domain (AICD) could exert harmful effects on cells, suggesting that the proteolytic products of APP, including $A\beta$, β CTF, and AICD, could play a pivotal role in neuronal viability. Here, we demonstrated that ligand-activated EphA4 signaling could govern the proteostasis of β CTF, AICD, and $A\beta$ independent of γ -secretase activity. The inhibition of EphA4 by Dasatinib, a receptor tyrosine kinase inhibitor, effectively suppressed the EphA4-induced accumulation of β CTF and AICD. This EphA4-elicited accumulation of β CTF and AICD was mediated by a Lyn-dependent pathway whose activation could in turn phosphorylate EphA4 to constitute a positive feedback in governing the proteostasis of β CTF and AICD. Furthermore, EphA4 signaling could regulate the ubiquitin-proteasome system to control the proteolysis of β CTF and AICD. In conclusion, our data delineate an EphA4-Lyn pathway that is essential for the metabolism of APP and its proteolytic derivatives, providing novel pharmacological targets for the development of anti- $A\beta$ therapeutics for AD.

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Abstract – WCN 2013

No: 1916

Topic: 5 – Dementia

Neuroanatomical correlates of behavioural and psychological symptoms in Alzheimer's disease: Voxel based morphometric study

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Background and purpose: In addition to cognitive decline, behavioral and psychological symptoms of dementia (BPSD) are of equal importance in the clinical profile of the disease in patient with Alzheimer's disease (AD). Therefore, in this study, we investigated the relationship between neuroanatomical structure and subtype of BPSD in AD.

Methods: Voxel-based morphometry was used to correlate gray matter (GM) derived from T1-weighted MRI images of 57 patients with Alzheimer's disease and specific neuropsychiatric symptoms and behaviours measured by the Korean version of Neuropsychiatric Inventory.

Results: The BPSD of AD are associated with atrophies of specific brain region. Delusion, hallucination, anxiety, euphoria, apathy, disinhibition, irritability, aberrant motor behavior, night time behavior, and appetite change were associated with GM density loss in specific cerebral region, significantly.

Conclusion: In this study, we confirm an atrophy of particular cerebral regions correlated with the subtype of BPSD.

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Abstract – WCN 2013

No: 1927

Topic: 5 – Dementia

Impacting quality of life and cognition in dementia patients through dance movements: Everyday waltzes

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Introduction: Literature has shown that physical health deterioration parallels cognitive decline in dementia patients. However, programs for physical rehabilitation are few and mainly target specific motor skills. Using a creative dance movement program designed for the elderly demented, we hypothesize, firstly that, it will lead to improved physical skills and enhanced perceptions of quality of life, and secondly, it will lead to improvements in objective measures of cognitive skills.

Method: 10 subjects participated in the 6-sessions workshop based on inclusion criteria. They were diagnosed with dementia as per DSM IV criteria, had verified objective cognitive impairments, were mobile, and had no other health issues.

Outcome measures were the Alzheimer Disease Quality of Life Inventory (ADQoL) and the Neuropsychiatric Inventory (NPI). These inventories were administered before the start of the program and on the day of the last session.

Results: In the ADQoL, we found significant difference between pre and post total quality of life score as rated by participants [$t(9) = 2.849$, $p = .019$]. There were no significant differences between pre and post ADQoL perceptions by caregivers.

In the NPI, caregivers reported a general decrease in the frequency and the severity of inappropriate behaviors following the workshop but with only one significant finding in total frequency and severity scores [$t(9) = 3.077$, $p = .013$].

Conclusion: Everyday waltzes program helped improve physical skills which in turn led to perceptions of increased quality of life and improved behaviors.

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Abstract – WCN 2013**No: 1950****Topic: 5 – Dementia****The medical food Souvenaid improves memory performance and preserves functional connectivity in mild Alzheimer's disease (AD)**

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Objective: Souvenaid®, containing the specific nutrient combination Fortasyn®Connect¹, is designed to improve synapse formation and function in patients with Alzheimer's disease (AD). The efficacy of Souvenaid is investigated in the clinical trial program.

Patients and methods: The program includes: 1) 12-week Souvenir I RCT in drug-naïve mild AD patients; 2) 24-week S-Connect RCT in mild-to-moderate AD patients using AD medication; 3) 24-week Souvenir II RCT² in drug-naïve mild AD patients; 4) 24-week Souvenir II open-label-extension (OLE) study; 5) 24-month LipiDiDiet RCT³ in prodromal AD; and 6) Mode of action studies: a) electroencephalography (EEG) and magnetoencephalography outcomes in Souvenir II; b) MRI and CSF measures in LipiDiDiet; c) magnetic resonance spectroscopy and d) FDG-PET.

Results: Souvenirs I and II showed that Souvenaid improved the primary outcome memory performance. The Souvenir II OLE showed a significant continued improvement of the memory outcome throughout 48 weeks. S-Connect did not show an effect on cognition in more moderate AD patients using AD medication. All studies showed that Souvenaid is well-tolerated, with a high compliance (≥93%). Electroencephalography (EEG) measures in Souvenir II assessed neuronal activity and thus indirectly synaptic activity. Significant EEG differences suggest that Souvenaid preserves functional connectivity and brain network organization, supporting the hypothesis of changed synaptic activity.

Conclusion: These results suggest that Souvenaid is most efficacious in early AD and warrant long-term trials in the early AD process.

References

- [1] Souvenaid and Fortasyn are registered trademarks of N.V. Nutricia.
- [2] Partly funded by NL FND project N°10003.
- [3] Funded by the EU FP7 project LipiDiDiet, Grant Agreement N°211696.

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Abstract – WCN 2013**No: 1953****Topic: 5 – Dementia****Comparison of rural and urban dementia prevalences in two countries of central Africa: The EPIDEMCA study**

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The number of estimates of dementia prevalence in low-income countries has increased during the last years, but few concerned Africa compared to Asia or Latin America continents. The aim of this survey was to compare urban and rural prevalence of dementia in two Sub-Saharan African countries.

A multicenter population-based study was carried out in Central African Republic (CAR) and Republic of Congo between 2011 and 2012 including both urban and rural sites in each country. Participants aged ≥65 years old were interviewed using the Community Screening Interview for Dementia (CSI-D), the GMS-AGECAT and the CERAD's 10 word list. Elderly with low performance to the CSI-D (<24.5/30) were then clinically assessed by neurologist and underwent further psychometrical tests. DSM-IV and NINCDS-ADRDA criteria were required for dementia and Alzheimer's disease diagnoses.

Overall, 2004 elderly were interviewed in both countries, including 475 in Nola (rural CAR), 500 in Bangui (urban CAR), 529 in Gamboma (rural Congo) and 500 in Brazzaville (urban Congo). Among them, respectively 183, 162, 284 and 149 had to be examined by a neurologist at the second stage, in order to establish DSM-IV dementia diagnosis. Preliminary results showed a DSM-IV dementia prevalence at 8.8% (CI95% [6.4–11.8]) in Nola and at 6.0% (CI95% [4.1–8.4]) in Bangui. Data processing is still ongoing for Congo. DSM-IV dementia prevalence will be presented for each study area.

This first comparison of rural/urban dementia prevalence in Africa will add to the available figures from Sub-Saharan Africa and low-income countries.

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Abstract – WCN 2013**No: 1605****Topic: 5 – Dementia****Developing early functional neuromarkers for abnormal cognitive decline: Multimodal brain imaging approach in MCI**

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Introduction: The capacity to accumulate information over time is crucial to our functioning in an ever-changing world. As individuals age, the ability to store, process, and manipulate information drops. Recently, in young healthy subjects, we showed that brain uses a distributed and hierarchical network of brain areas to process information over time. Here we study information processing under deficient cognitive state – amnesic mild cognitive impairment (a-MCI).

Objectives: One of the most common characteristics of cognitive impairment is difficulty with comprehension. We intended to map, in a-MCI patients, the topographical organization of temporal scales using an ecologically relevant auditory stimulus – a real-life story. In addition, we assumed that studying healthy elders, who are at high-risk for cognitive dysfunctions, will enable to determine neuromarkers of predisposition to disorder. Moreover, since this control group is not symptomatic, results will allow us to discriminate between critical and non-critical differences between patients and controls.

Methods: The fMRI data were analyzed using inter-subject correlation approach. The time-courses within each brain area in patients were estimated against matched controls.

Results: Among healthy elders, we observed widespread sensitivity to the content of the story through pSTG, TPJ, supramarginal gyrus, parietal lobule, precuneus, and prefrontal cortex. Among a-MCI patients, remarkably similar response reliability was found at the earliest stages

of processing (along STG up to Wernicke's area) only. In higher-order areas, reliability of responses dramatically disrupted.

Conclusions: Findings pointed to a possibility that MCI may impact higher-order cortical regions, while regions involved in early processing remain unaffected.

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Abstract — WCN 2013

No: 1963

Topic: 5 — Dementia

Autologous anti-A β antibodies in CAA-ri: New biomarker for detection of amyloid-related imaging abnormalities (ARIA) during A β -disease modifying therapies for AD

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Background: Cerebral Amyloid Angiopathy-related inflammation (CAA-ri) is a rare disease characterized by vasogenic edema and multiple cortical/subcortical microbleeds, shearing several aspects with the recently defined Amyloid-Related Imaging Abnormalities (ARIA) during Alzheimer's disease (AD) passive immunization therapies.

Objective: We investigated the putative role of anti-A β autoantibodies as the primary actor of the inflammatory reaction rising in these two conditions.

Methods: By a multicenter case-control study in 80 subjects, using a novel ultra-sensitive technique (patent application pending), we evaluated the anti-A β autoantibodies concentration in the CSF of CAA-ri, CAA, AD, MS and healthy-control subjects. Levels of circulating A β 40, cA β 42, tau, P-181 tau and APOE4 genotype were also investigated.

Results: We confirmed a direct involvement of anti-A β autoantibodies during the course of CAA-ri, demonstrating that their concentration is specifically increased during the acute phase and progressively reduced with clinical and radiological remission. Moreover, a strong correlation with the increased mobilization of cA β 40 and cA β 42 during the acute phase was shown, followed by their return to control levels and reduced tau and P-181 tau after remission.

Conclusions: Our data strongly support the hypothesis that the pathogenesis of CAA-ri is mediated by a selective autoimmune reaction against cerebro-vascular A β , directly related to autoantibody concentration and cA β overloading. Given the similarities between spontaneous ARIA which develops in CAA-ri and that induced through immunization strategies, anti-A β autoantibodies in the CSF may be proposed as a valid alternative for the diagnosis of CAA-ri and as a novel biomarker to monitor ARIA during the ongoing amyloid-modifying therapies for AD.

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Abstract — WCN 2013

No: 2022

Topic: 5 — Dementia

Clinical and neuroimaging profile of probable Creutzfeldt Jakob disease; case series of four patients

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Background: Creutzfeldt Jakob disease is a rare neurodegenerative disorder, a spongiform transmissible encephalopathy characterized by rapidly progressive dementia, myoclonus, ataxia, pyramidal, extrapyramidal and visual abnormalities. It is universally fatal with almost hundred percent mortality. Creutzfeldt-Jakob disease (CJD) is one of the subtypes of prion diseases, classified as hereditary, infectious and sporadic disorders.

Objective: In recent past, the role of diffusion weighted imaging has been established in Sporadic Creutzfeldt Jakob disease due to presence of characteristic findings of cortical ribboning and hyperintense basal ganglia structures. Our objective is to highlight the clinical description and magnetic resonance findings of 4 patients diagnosed as Sporadic Creutzfeldt Jakob disease.

Patients and methods/material and methods: The clinical details and neuroimaging features of four patients diagnosed as Sporadic Creutzfeldt Jakob disease were described, admitted consecutively in tertiary center of North India in span of one year.

Results: Our 3 patients were female and one was male. The age ranged from 42–65 years (mean 55.5 years). Electroencephalography of all patients showed periodic spike and wave complexes. The neuroimaging findings were cortical ribboning of grey mantle and hyperintense signals of basal ganglia structures. All patients succumbed to illness within a year from onset of illness.

Conclusion: Creutzfeldt Jakob disease is an underdiagnosed, fatal neurodegenerative disorder. The awareness of CJD among physicians is essential to differentiate from other causes of treatable dementias. The magnetic resonance imaging should be used as an additive tool for diagnosis of Creutzfeldt Jakob disease.

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Abstract — WCN 2013

No: 1979

Topic: 5 — Dementia

Cognitive changes in idiopathic normal pressure hydrocephalus

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Background: Idiopathic normal pressure hydrocephalus (iNPH) is a syndrome that is characterized by cognitive decline, gait disturbance and impairment in bladder control (Adam's triad), and is associated with ventriculomegaly in the absence of elevated cerebrospinal fluid (CSF) pressure.

Objective: Diagnosis can be challenging due to its varied presentation and overlap with other disorders common in the elderly. The neuropsychological characteristics of iNPH have been much debated during recent years. This study investigates the neuropsychological features in patients with iNPH.

Methods: On the basis of Clinical Guidelines for idiopathic Normal Pressure Hydrocephalus, 8 participants were diagnosed with definite iNPH. We examined the scores of the Frontal Assessment Battery (FAB), Behavioural Assessment of the Dysexecutive Syndrome (BADS) and the Mini-Mental State Examination (MMSE). Patients with iNPH were assessed before and six months after shunt operation. The cerebral blood flow (CBF) was measured by N-isopropyl-123-P-iodo-amphetamine

single photon emission computed tomography, and the perfusion patterns of the cerebral cortex were measured based on three-dimensional stereotactic surface projection (3D-SSP) Z-score images.

Results: The score of FAB improved better than that of MMSE. Three patients showed significant improvement in BADS test. These changes were shown to be sensitive to damage to the frontal function. 3D-SSP analyses of SPECT images revealed that regional CBF in prefrontal cortex and Sylvian fissure decreased in patients.

Conclusions: Damages of periventricular structures, such as frontal subcortical and cortical areas, are closely connected with impairments of iNPH. We suggest that the frontal dysfunction should be used in the evaluations of iNPH.

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Abstract – WCN 2013

No: 2048

Topic: 5 – Dementia

Are the cortical cerebral microbleeds (CMB) present in dementia with Lewy bodies?

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Objective: To identify the location and number of CMB in dementia with Lewy bodies (DLB) and to estimate the influence CMB on cognitive decline in DLB.

Patients and methods: We examined 33 patients with DLB ((mean age 73.5 years, 20 (67%) males) and 51 patients with Alzheimer's disease (AD) with cognitive decline. MRI was performed by 1.5 T MR tomography. CMB were estimated in T2*GRE. Patients with DLB (16 (49%)) had hyperintensity in T2 reflecting vascular leukoencephalopathy which was estimated by the Fazekas scale. Neuropsychological testing was carried out by Montreal Cognitive Assessment scale (MoCA), Addenbrooke's Cognitive Examination (ACE-R), Clock Drawing Test, verbal fluency test, and visual memory test (SCT).

Results: CMB in DLB were found in 13 (39%) patients. The total number of CMB were 70, 40 (57%) – subcortical. Patients with cortical CMB had less memory score (3 ± 2) by ACE than patients with deep CMB. Deep CMB were found in 10 cases with severe vascular leukoencephalopathy. Most cortical CMB were observed in AD – 340 (96%). Patients with AD and CMB had significantly lower score of ACE-R (less than 3.7) than in DLB ($p < 0.01$). Significant differences in MoCA had been not detected.

Conclusion: We have not found cortical CMB in patients with DLB by MRI 1.5 T. The absence of cortical CMB in patients with DLB might be the additional criteria of diagnosis DLB. This observation allows to propose the presence of cortical CMB in patients with clinically diagnosed DLB to point out to the coexistence DLB with AD.

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Abstract – WCN 2013

No: 2116

Topic: 5 – Dementia

The diagnostic utility of ^{99m}Tc-HMPAO SPECT imaging in early onset dementia: A retrospective audit from a regional cognitive disorders clinic

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Background: Single photon emission computed tomography (SPECT) using the radiopharmaceutical agent ^{99m}Tc-hexamethylpropyleneamine (^{99m}Tc-HMPAO) provides whole brain perfusion imaging which is of potential diagnostic utility in the diagnosis of dementia.

Objective: A retrospective audit was conducted to determine the utility of HMPAO SPECT in a cohort of patients presenting to a tertiary centre early onset cognitive disorders clinic.

Patients and methods: 42 patients (23 males, mean age 59.6 y, range 37–80 y) reviewed over 1 year were studied. All were assessed clinically by a neurologist and a psychiatrist SPECT imaging was interpreted visually by a nuclear medicine specialist. Clinical diagnoses, Addenbrooke's Cognitive Assessment Revised (ACE-R) scores, SPECT, MRI and EEG findings were evaluated.

Results: 41 patients fulfilled clinical criteria for a dementia syndrome (mean ACE-R score 69/100). Clinical diagnoses included: Alzheimer's disease $n = 21$, posterior cortical atrophy $n = 4$, primary progressive aphasia $n = 4$, behavioural variant frontotemporal dementia $n = 3$, epilepsy $n = 1$, HIV $n = 1$, mixed dementia $n = 2$, and unclear aetiology $n = 6$. 78% of HMPAO SPECT scans ($n = 32$) that demonstrated regional blood flow abnormalities indicative of a dementia. 51% ($n = 21$) further differentiated between anterior and posterior dementia syndromes. In comparison, MR imaging detected abnormalities (vascular change, global/regional atrophy) in 48% ($n = 20$), and EEG (generalised slowing) in 14%.

Conclusion: Evaluation of regional blood flow abnormalities identified by HMAPO SPECT imaging appears a useful adjunct in evaluating patients with early onset cognitive impairment with favourable diagnostic sensitivity compared with other investigations. Quantitative methods of image analysis may enhance sensitivity, further aiding accurate diagnosis in this very challenging cohort.

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Abstract – WCN 2013

No: 2083

Topic: 5 – Dementia

A.C.E.S.I.D.E. group: Taking care of those who take care

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The A.C.E.S.I.D.E. group aims to provide psychological support to caregivers of patients with dementia. The purposes of the A.C.E.S.I.D.E. group are many: give caregivers a space in which they can think to themselves and deal with issues related to anxiety and mood disorders that may be a consequence of their caring tasks, and to find coping strategies in comparison with professionals and with other people in the same situation. We also want to inform the participants of what it means to be suffering from dementia illustrating the main symptoms and also inform them on key local services that constitute a rich resource. Participants are caregivers of dementia patients. Participation is voluntary, although it still requires continuity. The group consists of a number of participants, from 8 to 12. The meetings are 6 and their frequency is fortnightly. After the first meeting devoted to the project description and presentation of conductors and participants, subsequent meetings are devoted to the illustration of the main concepts related to dementia and the management of anxiety and stress, as well as of mood problems, which may occur in everyday life of the caregiver. In the first part of each meeting, nurses inform participants about local services and how to get more aid and economic assistance. Participants say they are always very satisfied and feel "privileged" to have the opportunity to attend a support

group. Generally there is a decrease in anxiety and activation of better strategies to combat stress.

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Abstract – WCN 2013

No: 2085

Topic: 5 – Dementia

Cardiovascular autonomic function in frontotemporal dementia, behavioral variant (FTDbv)

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Background: Patients with autonomic failure experience postural dizziness, syncope, and falls. Identifying symptomatic dysautonomia in dementia is of importance to ensure appropriate management and reduce risk of falls.

Objective: The aim of this prospective study is to identify autonomic dysfunction in patients suffering FTDbv.

Methods: Patients were recruited from the Department for Neurology, General Hospital, City of Linz. Clinical autonomic function tests were carried out according to Ewing's battery. Parasympathetic tests included resting heart rate variability, deep breathing (E/I ratio), and Valsalva ratio. Sympathetic function tests included blood pressure regulation on Valsalva, cutaneous cold stimulation, arithmetic exercise and 70° head-up tilt.

Results: History taking was often insufficient and could not replace autonomic testing.

One of 23 recruited patients had to be excluded due to atrial fibrillation. Mean age of 22 FTDbv patients was 66 ± 12 years, female/male: 13/9. In 8 patients (36%) autonomic dysfunction was demonstrated: 8 patients with sympathetic dysfunction of those 6 patients (27%) had manifested orthostatic hypotension (two related to medication). 4 had a parasympathetic dysfunction, in only one patient related to medication (beta blocker).

One patient suffered vasovagal syncope during tilt.

Conclusion: This is to our knowledge the first prospective study to elucidate autonomic dysfunction in FTDbv patients. Autonomic dysfunction is not uncommon and is present in about 1/3 of this cohort. Diligent history taking is of major importance in patients suffering autonomic symptoms; however in FTDbv patients, history taking is insufficient. Autonomic dysfunction might therefore be concealed if patients are not tested employing autonomic function tests.

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Abstract – WCN 2013

No: 2096

Topic: 5 – Dementia

Depression in dementia: Results from an ongoing dementia registry in Pakistan

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Background: With increasing life expectancy in Pakistan, dementia is becoming a growing public health concern. Depression is the most frequent psychiatric co-morbidity in dementia.

Objectives: To assess depression in dementia patients and identify the correlation between the Mini Mental State Examination (MMSE) scores and the Beck's Depression Inventory (BDI) scores in a tertiary care hospital in Pakistan.

Methodology: Out of 230 patients enrolled in our dementia registry, 92 were excluded because of MMSE scores ≥ 26 and 4 due to incomplete data, making the final sample of 134. A BDI score ≥ 17 was taken as clinical depression. SPSS v.16 was used for final analysis.

Results: Male to female ratio was equal. Mean age was 70.1 ± 9.6 years. 21.7% were uneducated. 73.9% were married. The majority lived with their spouse and children (46.4%) or in joint families (39.1%). Mean MMSE score was 16.5 ± 6.3 . 44.7% had moderate, 32.8% mild while 22.3% had severe impairment. Mean BDI score was 12.7 ± 9.2 . 29.1% had clinical depression. Out of these patients, 46.2% had moderate, 35.8% borderline and 17.9% severe depression; no case of extreme depression was observed. MMSE and BDI scores were significantly correlated showing that patients with higher MMSE scores had lower BDI scores.

Conclusion: Minority of our dementia patients had depression. This could be due to family support systems available for the majority of these patients. But with deterioration in the MMSE scores, risk of developing clinical depression increases hence dementia patients should have periodic evaluation for depression.

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Abstract – WCN 2013

No: 2113

Topic: 5 – Dementia

Association between orthostatic hypotension and mild cognitive impairment in the patients with benign prostate hypertrophy

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Background: Some reports suggested that orthostatic hypotension (OH) might be associated with the mild cognitive impairment (MCI). However, the prevalence of OH and MCI in the patient with benign prostate hypertrophy (BPH) has not been clearly demonstrated. We investigated the prevalence of OH and MCI in the patients with BPH and also analyzed the association between OH and MCI.

Methods: From April 2011, we recruited BPH patients who were ≥ 65 years old, and had taken alpha-blocker more than one year. Patients with previous history of OH, stroke, or dementia were excluded. All patients took the head-up tilt table test and the neuropsychological test.

Result: Until July 2012, 43 patients (age 69.6 ± 7.5 yrs) were finally enrolled to this study. Among them, 26 subjects (60.5%) had OH. The prevalence of MCI was 41.9% on MMSE and 67.4% on MoCA. There was no difference in the prevalence of MCI between OH group and control (On MMSE, 42.3% vs. 41.2%, $P = 0.94$; On MOCA, 65.4% vs. 70.6%, $P = 0.72$). The patients with OH had lower body mass index (BMI) than the control group (23.6 ± 2.6 vs. 27.2 ± 6.8 , $P = 0.04$). In the patients ≥ 70 years old, MMSE score was associated with BMI ($P = 0.041$) and systolic blood pressure ($P = 0.03$).

Conclusions: Prevalence of MCI and OH was high in the patients taking alpha blocker for BPH. OH did not seem to be associated with MCI.

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Abstract – WCN 2013

No: 1581

Topic: 5 – Dementia

Variety of clinical symptoms of logopenic progressive aphasia in Japanese patients

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Background: Logopenic progressive aphasia (LPA) is one of the entities of primary progressive aphasia. It was recently established and it is not yet sufficiently evidenced.

Objective: The objective of this study is to elucidate the implication of the symptoms, lesions and prognosis in Japanese patients with LPA.

Patients and methods: We investigated four Japanese patients fulfilling the criteria for diagnosis of LPA (Gorno-Tempini et al. 2011) with detailed neuropsychological examinations, MR imagings, and 3D-SSP analysis of ¹²³IIMP SPECT imagings (SPECT) for 4 to 7 years from the onset of aphasia.

Results: The patients were divided into two sub-groups: the parietal type and the temporal type. The patient of parietal type manifests prominent phonemic paraphasia, with relatively spared word finding ability, and also showed left parietal focal signs such as acalculia, constructional impairment and so on. The MRI and the SPECT findings showed the main lesion in the left parietal lobe, especially in the supramarginal gyrus. The patients of temporal type developed word finding difficulty with some phonemic paraphasia, word deafness due to the lesion in the posterior part of superior temporal gyrus. They also showed impairment of verbal short-term memory heralded by or concomitant with phonemic impairments. The MRI and the SPECT findings suggested the main lesion in the posterior part of the left temporal lobe.

Conclusion: LPA could be divided into two subtypes: the parietal type and the temporal type, which might be informative to know the prognosis and mechanism of the symptoms of the patients with LPA.

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Abstract – WCN 2013

No: 2133

Topic: 5 – Dementia

Wide spectrum of neurodegenerative pathologies in a prospective longitudinal community-based study

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Background: Neurodegenerative diseases are characterised by neuronal loss and cerebral deposition of proteins. The major proteins are amyloid-beta (Abeta), tau, a-synuclein, and TDP-43. There have been only few observations on the full spectrum of proteinopathies in the ageing human brain.

Objectives: To perform a comprehensive mapping of neurodegeneration-related proteins and vascular pathology in the brains of elderly individuals.

Methods: Cases were evaluated in a prospective longitudinal community-based study (VITA). Detailed neuropathological evaluation was performed following diagnostic criteria and by performing immunostaining for Abeta, tau, a-synuclein, and TDP-43 in several anatomical regions.

Results: 233 individuals (age at death 77–87) were examined. All brains (from individuals with and without dementia) showed some degree of neurofibrillary degeneration. Abeta parenchymal deposits were observed only in 160 (68.7%). Further pathologies included a-synucleinopathies (24.9%), tauopathies (23.2%), TDP-43 proteinopathy (13.3%), vascular lesions (48.9%), and others (15.1%; inflammation, metabolic encephalopathy, and tumours). TDP-43 proteinopathy correlated well with hippocampal sclerosis ($p < 0.001$) and AD-related pathology ($p = 0.001$). Abeta parenchymal deposits, tauopathy, TDP-43, and a-synucleinopathy showed significant association with dementia. The number of observed pathologies correlated with Braak & Braak stages and CERAD score.

Conclusions: Non-Alzheimer's Disease type neurodegenerative pathologies, and their combinations are frequent in elderly brains and should be considered both in diagnostic evaluation including biomarkers, and therapeutic trials. Addition of further pathologies may lower the threshold for symptomatic cognitive decline even with minor Alzheimer's Disease-related changes.

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Abstract – WCN 2013

No: 2136

Topic: 5 – Dementia

Randomized, double-blind, placebo-controlled, multicentre study to investigate memantine in the treatment of memory, concentration, and attention problems (subjective cognitive impairment)

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Background: Studies have shown that 25–50% of the population over the age of 65 in Europe suffers from subjective cognitive impairment (SCI). Different treatment options are used in this condition with doubtful scientific background.

The primary objective of the study was to investigate the efficacy and safety of 10 mg memantine once-daily in comparison to placebo in the treatment of memory, concentration, or attention problems (SCI) in the absence of dementia at week 12 with the Patient Global Impression of Change (PGI-C) scale.

Secondary objectives were to investigate the efficacy of 10 mg memantine as measured by the CogState computer-based neuropsychological test battery, Everyday Cognition39, and Hospital Anxiety and Depression Scale.

Patients and methods: This was an explorative analysis of data from a multicentre, randomized, double-blind placebo-controlled trial. 295 patients were randomized to memantine 10 mg once-daily or to placebo and treated for 12 weeks with a follow-up of 4 weeks.

Results: At week 12, the analysis of covariance of the PGI-C showed that the least square mean difference from placebo was 0.15 (90% CI: –0.02 to 0.32); this difference was not statistically significant ($p = 0.157$). Similarly, no statistically significant differences between the treatment groups could be shown for the secondary outcome measures.

In conclusion, one reason for no statistically significant difference between the groups could be that the subjects included might not have had a sufficiently manifested cognitive impairment to show an effect. Memantine was safe and well tolerated when administered to subjects with SCI in a dose of 10 mg, without previous up-titration.

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Abstract – WCN 2013**No: 2150****Topic: 5 – Dementia****The outcome of ventriculo-atrial shunt for idiopathic normal pressure hydrocephalus: Analysis of 393 cases**

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Background and purpose: Idiopathic normal pressure hydrocephalus (iNPH) has long been known as one of the causes of treatable dementia. Cerebrospinal fluid (CSF) shunt surgery is the only therapeutic modality for iNPH. As for the types of shunt surgery, ventriculo-peritoneal shunt (VP shunt) is prevailing and ventriculo-atrial shunt (VA shunt) is almost abandoned. However, VA shunt has many advantages. We have performed 466 VA shunts for normal pressure hydrocephalus since April, 2005. Among these patients, there were 393 patients with iNPH. The purpose of this paper is to show the outcome of VA shunt for iNPH.

Method: The 393 candidates for VA shunt were CSF tap test responders over 50 years old. The outcome was evaluated 3 months after the surgery depending on five categories; Excellent (improvement of modified Rankin Scale or improvement of mini-mental state examination over 3, or cease of urinary incontinence), Good (no improvement of mRS but decreased caregiver's burden or increased patient's activity), Fair (no or very slight improvement), Poor (worsen after VA shunt), or Dead (death within 1 month after VA shunt).

Results: Mean age was 78.1 years old and mean operation time was 36.6 min. There was no shunt infection. The outcome was as follows; Excellent (58.3%), Good (21.6%), Fair (16.5%), Poor (1.8%), Dead (0.5%), and Unknown (0.8%).

Conclusion: Despite the old age of the patients, the mortality rate was quite low and the shunt infection was zero. Our results suggest that VA shunt can be the first choice for iNPH.

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Abstract – WCN 2013**No: 2161****Topic: 5 – Dementia****Evolution of symptoms and signs over 6-months in subjects with possible dementia with Lewy bodies following DaTSCAN™ SPECT imaging**

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Background: The diagnosis of patients with dementia with Lewy bodies (DLB), can be clinically challenging, particularly with possible-DLB. In clinical practice, generally the diagnosis becomes clearer over time with the progression of symptoms and signs.

Objective: To compare baseline and 6-months follow-up clinical data of a cohort of possible DLB patients collected for a dopamine transporter SPECT imaging study.

Methods: One hundred and eighty seven patients with possible DLB (dementia + one core or one or more suggestive features excluding

DaTSCAN™ [Ioflupane¹²³I]) were randomized to have a DaTSCAN™ or to follow-up clinical care without DaTSCAN™. Diagnosis, DLB features and a comprehensive neuropsychiatric battery (Fluctuation scale, ACE-R, MMSE, NPI, UPDRS-III) were compared at baseline and 6 months. **Results:** Overall prevalence of DLB features was similar between the two study groups. At 6-months follow-up diagnostic category changed to non-DLB (26%) or probable DLB (26%). However, changes were more frequent in the DaTSCAN™ than in the control group (71% vs 16% $p < 0.0001$), particularly in those with abnormal images (87%).

Parkinsonism at baseline was more prevalent in probable DLB and was the only feature that evolved at 6-months follow-up (42% to 61%). UPDRS-III changes were in the same direction. There were no differences between the groups with regard to other clinical features at baseline and/or at 6 months.

Conclusion: In subjects with possible DLB the clinical evolution over 6-months without DaTSCAN™ did not impact the diagnosis. The DaTSCAN™ results in combination with other core and suggestive features contributed significantly to the diagnosis.

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Abstract – WCN 2013**No: 1387****Topic: 5 – Dementia****In Alzheimer's disease sleep impairment is correlated with cognitive decline, increased tau and decreased beta-amyloid CSF levels**

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Background: AD is primarily characterized by cognitive decline, but nocturnal sleep disturbances are also features of the disease and seem to be related to dementia severity.

Objective: To correlate in preclinical AD patients the objective macrostructural sleep parameters with MMSE and CSF biomarkers (tau and phosphorylated-tau proteins, beta-amyloid₁₋₄₂).

Patients and methods: Thirty-seven unselected patients affected by preclinical AD underwent MMSE, a 24-hour ambulatory polysomnography to evaluate the sleep-wake rhythm, lumbar puncture for the assay of CSF biomarker levels. Patients with sleep-disordered breathing were excluded from the study. The sleep macrostructural analysis was performed according to Rechtschaffen-Kales' criteria.

Results: In our AD patients, the increment of CSF tau levels correlated:

- i) positively with the NREM stage 1 sleep ($p < 0.05$) and the wakefulness after sleep onset (WASO) ($p < 0.01$);
- ii) negatively with sleep efficiency (SE) ($p < 0.01$) and REM sleep ($p < 0.05$). Moreover, the decrement of the CSF beta-amyloid₁₋₄₂ levels correlated with the decrement of REM sleep ($p < 0.05$) and the increment of the WASO ($p < 0.05$).

Finally, MMSE appeared statistically related to: i) the decrease of the SE ($p = 0.001$); ii) the decrement of the NREM stage 3 sleep ($p < 0.01$); iii) the reduction of the REM sleep ($p < 0.01$); and iv) the increment of the WASO ($p < 0.001$).

Conclusion: In AD patients higher tau and lower beta-amyloid₁₋₄₂ CSF levels are correlated with the impairment of sleep quality and continuity. Finally, our results suggest that in AD patients the cognitive decline is strictly related to the sleep dysregulation.

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Abstract – WCN 2013**No: 2208****Topic: 5 – Dementia****Cognitive assessment of adults with Down syndrome**

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Background: Cognitive deterioration, mood and behavioural changes suggest early dementia and are frequently observed in adults with Down syndrome (DS).

Objective: The aim of this study is to identify early signs of dementia in a Portuguese sample of adults with DS.

Patients and methods: Sixteen patients with DS, 25 to 50 years old, without treatable causes of dementia, were assessed at the beginning of the study (T1) and 12 months later (T2) using the Plymouth Dementia Screening Checklist (PDSC), Global Deterioration Scale (GDS), Mini-Mental State Examination (MMSE) and Dementia Questionnaire for People with Learning Disabilities (DLD). The data were statistically analysed.

Results: Most participants had low MMSE values, with no significant differences between T1 and T2. In T1, 2 individuals punctuated above the cut-off point on PDSC, particularly in memory and behaviour complaints. One individual was in stage 3 of GDS (mild cognitive decline) and had a low performance in cognitive and social domains of DLD. At follow-up, 7 participants had warning signs in PDSC, being memory the most compromised skill. Six of them had cognitive deterioration in GDS and altered long-term memory, behaviour, mood, activity and interest as measured by DLD. MMSE values were significantly related to these domains in T1 and T2.

Conclusion: Behaviour, mood and particularly memory should be assessed in DS when dementia onset is considered. PDSC was a useful instrument to predict dementia, further confirmed by GDS and DLD. MMSE identified cognitive deterioration signs in this sample and showed possible interference of mood and initiative.

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Abstract – WCN 2013**No: 1964****Topic: 5 – Dementia****Differential diagnosis of AD using PIB and FDG-PET in a memory clinic**

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Background: PET imaging using ¹⁸F-fluorodeoxyglucose (FDG) and ¹¹C-Pittsburgh compound B (PiB) has been proposed as biomarkers of Alzheimer disease (AD).

Objective: To compare the diagnostic performance of PET with the amyloid ligand PiB-PET to FDG-PET in discriminating AD from others in a memory clinic setting.

Patients and methods: In 106 patients who underwent brain MRI, PiB-PET and FDG-PET in our memory clinic, hypometabolism of the parietotemporal lobe and posterior cingulate gyrus in FDG-PET and cortical uptake in PiB-PET were evaluated as findings characteristic of AD. PiB scans were classified as positive or negative by 2 visual raters blinded to clinical diagnosis. FDG scans were visually rated as consistent with AD, and quantitatively classified based on the region of lowest metabolism relative to controls.

Results: The AD findings were observed in 86% of all AD patients with FDG-PET, 90.1% with PiB-PET, and 72% with both PET scans. Eight PiB negative cases had positive findings for AD in FDG-PET. 10

PiB positive cases had negative findings for AD in FDG-PET. PiB had a higher sensitivity for AD (87.2%) than FDG (79.5%) with similar specificity (PiB 81%, FDG 82%).

Conclusion: PiB and FDG showed similar accuracy in discriminating AD from others in a memory clinical setting. To use both PET improves the diagnostic accuracy than to use only one method.

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Abstract – WCN 2013**No: 2269****Topic: 5 – Dementia****Network analysis of intrinsic functional connectivity in the semantic variant of primary progressive aphasia**

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Background: The semantic variant (SV) of primary progressive aphasia is associated with focal, asymmetric, temporal lobe degeneration. However, language abilities must arise from the activity of distributed neural networks. Graph theoretical analysis allows examining the topology of complex network systems.

Objective: To examine the integrity of the functional brain connectome in SV.

Methods: Graph theoretical analysis was applied to resting state functional MRI data from 14 SV and 50 matched controls. Functional connectivity between 90 cortical and subcortical brain regions was estimated using bivariate correlation analysis and thresholded to construct a set of undirected graphs.

Results: Functional brain networks in SV did not show loss of small world properties, but were characterized by lower clustering coefficient, global efficiency and nodal degree, longer characteristic path length and higher assortativity compared with controls. SV did not show brain hubs in right superior frontal, inferior orbitofrontal, lingual gyri and cuneus, and in left anterior cingulate cortex and inferior temporal gyrus. Left precentral gyrus, supplementary motor area, bilateral middle frontal gyrus and thalamus were hubs only in SV. SV showed decreased nodal degree in left frontotemporal regions and right parahippocampal and middle occipital gyri.

Conclusions: SV functional networks are characterized by an imbalanced structure, with a loss of efficiency in information exchange between both close and distant brain areas. Local functional network organization is altered in SV. Focal degeneration of “classic” (temporal) language regions in SV may ultimately manifest in widespread connectivity disturbances elsewhere in the brain.

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Abstract—WCN 2013**No: 1810****Topic: 5—Dementia****Inherited prion disease due to 5-octapeptide repeat insertion**

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Background: Prion diseases are fatal neurodegenerative conditions which have sporadic, acquired or inherited aetiologies. All inherited cases have coding mutations in the prion protein gene (PRNP) – either point mutations or octapeptide repeat insertion mutations causing variable numbers of an octapeptide repeat element in the N-terminal repeat region.

Case history: A 56 year-old man was admitted to hospital following a fall. He had had symptoms for a minimum of six years which met standard criteria for the behavioural variant of frontotemporal dementia. There was a sub-acute decline in his condition during the admission and he became bedbound, incontinent and mute with marked myoclonus and limb rigidity. He died four months after admission. There is a family history of dementia.

Results: MRI brain showed generalised cerebral atrophy but no focal abnormality. Lumbar puncture constituents were normal. An initial EEG showed encephalopathy but a repeat had persistent generalised sharp waves at 2 Hz. Genetic testing revealed a five octapeptide insertion in the PRNP gene. Postmortem examination confirmed prion disease.

Conclusion: This case demonstrates that inherited prion disease can present with a long history consistent with frontotemporal dementia. The insertion mutation found in this case is identical to a previously reported case from Northern Ireland who presented with a very different phenotype and almost certainly these two cases are related.

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Abstract – WCN 2013

No: 2251

Topic: 5 – Dementia

Combining SPECT and EEG analysis for assessment of disorders with amnesic symptoms to enhance accuracy in early diagnostics

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Objectives: Comparative evaluation of regional brain perfusion measured by HMPAO-SPECT and quantitative Electroencephalography (EEG) analysis of persons with subjective cognitive complaints with (SCC+) and without (SCC–) minimal cognitive dysfunction, with amnesic mild cognitive impairment (aMCI) and such with Alzheimer's disease (AD).

Methods: A total of 662 patients were investigated because of suspected cognitive dysfunction. After exclusion of patients with other forms of dementia than DAT or relevant accompanying disorders, SPECT and EEG data from 31 SCC–, 50 SCC+, 99 aMCI, and 72 DAT (together 252) were analyzed. Relative cerebral blood flow of 34 anatomical regions was assessed with automated analysis software (BRASS). Besides SPECT, each patient has also undergone EEG, which was neither statistically evaluated on a group level, nor quantitatively assessed or correlated with the SPECT-results.

Results: The database is now completed and the comparative evaluation is ongoing the next 3 months. Therefore results will be demonstrated at the WCN 2013 congress.

Conclusion: The present project aims at assessing the diagnostic validity of new biomarkers by quantifying and combining SPECT- and EEG-data. The implementation of these methods in clinical practice

could improve accuracy of early diagnosis in disorders with amnesic symptoms.

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Abstract – WCN 2013

No: 2265

Topic: 5 – Dementia

I716F APP mutation associated with Alzheimer's and diffuse Lewy body disease

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Background: Neurodegenerative diseases (NDDs) are characterised by progressive loss of neurons and deposition of proteins in the brain. Mutations causing familial Alzheimer's disease (FAD) have been mainly described in three genes: amyloid precursor protein (APP), presenilin 1 (PSEN1) and presenilin 2 (PSEN2).

Objectives: To present clinical, neuroradiological, neuropathological, and genetic data on a patient with early-onset dementia.

Methods: Clinical follow-up, cranial MRI, neuropathology, including immunohistochemistry for phospho-tau, alpha-synuclein, prion protein, amyloid-beta, and phospho-TDP-43. Genetic analysis of the APP, PSEN 1 and 2, and PRNP genes.

Results: A 45-year-old man presented with psychiatric symptoms and later motor symptoms with myoclonus and tetraspasticity, followed by progressive dementia and terminally an apallic syndrome with epileptic seizures. The clinical course was 10 years. Similar symptoms were reported in the patient's brother. Neuropathological evaluation revealed neuronal loss predominating in the frontal and temporal cortices, basal ganglia, thalamus, and hippocampus. Tau-pathology comprised neurofibrillary degeneration corresponding to stage VI according to Braak and Braak. Alpha-synuclein-pathology consisting of Lewy-bodies and Lewy-neurites corresponded to Braak stage VI and met the criteria for the neocortical stage of dementia with Lewy bodies. Severe amyloid-b angiopathy and numerous amyloid-b plaques were seen throughout the cerebrum and cerebellum. Genetic analysis revealed an I716F mutation in the APP gene.

Conclusions: Similarly to sporadic NDDs, early onset dementias with a genetic background may be associated with multiple proteinopathies and complex clinical presentations.

This study was performed in the frame of the MUW-early-onset-dementia Consortium and was supported by the European Commission's 7th Framework Programme "DEVELAGE".

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Abstract – WCN 2013

No: 2266

Topic: 5 – Dementia

The role of neuroprotein in schizophrenia pathogenesis

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Background: Schizophrenia (SCZ) is a psychiatric disorder with both altered neurodevelopmental and genetic components. Despite several studies performed, the SCZ etiology is still unclear. Altered blood and mRNA expression levels of brain-derived neurotrophic factor (BDNF), promoting synaptic transmission and neural plasticity, in the brain of SCZ patients and genetic analysis suggest contribution of BDNF to disease pathogenesis.

Objective: To clarify the role of the BDNF in SCZ, protein levels and polymorphism in SCZ-affected and healthy subjects were evaluated.

Patients and methods: In this study, the BDNF blood levels in chronic patients with paranoid SCZ treated with antipsychotics, antipsychotic-naïve patients, and controls were evaluated by ELISA.

Results: Comparative analysis showed 1.45-fold decreased BDNF levels in SCZ when compared to controls ($p = 2.27E - 23$). Also, there was a significant difference in levels of BDNF between antipsychotic-naïve patients and controls ($p = 8.2E - 07$), whereas treated patients were not differed from antipsychotic-naïve ($p > 0.05$). With regard to gender, reduced BDNF levels in females compared to males both in patients ($p = 0.0018$) and controls ($p = 0.019$) were found. We compared the BDNF levels with regard to the Val66Met genotypes in SCZ patients and controls based upon our previous data. Relevant intragroup analysis showed increased BDNF levels in 66Met allele carriers when compared to Val/Val both in SCZ ($p = 0.0003$) and controls ($p = 4.22E - 05$).

Conclusion: To summarize, the results obtained suggest that decreased BDNF levels associate with 66Met allele carriers and might be involved in the pathogenesis of SCZ in Armenian population. All authors declare that they read and are aware of the content of abstract.

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Abstract – WCN 2013

No: 1928

Topic: 5 – Dementia

Predictors of response to the 13.3 and 9.5 mg/24 h rivastigmine patch: The optimizing transdermal exelon in mild-to-moderate Alzheimer's disease (optima) study

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Background: The OPTIMA study (NCT00506415) demonstrated reduced deterioration with 13.3 versus 9.5 mg/24 h rivastigmine patch in patients with Alzheimer's disease (AD). Treatment response can vary according to patients' individual characteristics. We conducted a responder analysis to identify the proportion of patients demonstrating a treatment response to each dose, and patient characteristics predictive of achieving a response.

Methods: Details of OPTIMA are published (Cummings et al., 2012). Patients meeting pre-specified decline criteria during treatment with 9.5 mg/24 h patch, entered a 48-week, double-blind (DB) phase, and were randomized to 13.3 or 9.5 mg/24 h. ADAS-cog and ADCS-IADL were co-primary endpoints. In this *post-hoc* analysis, the following criteria for achieving a treatment response at weeks 24 and 48 were applied: ≥ 4 points' improvement on ADAS-cog, and ≥ 4 points' improvement on ADAS-cog combined with no decline on ADCS-IADL.

Results: Demographics and baseline scores were comparable between patient groups (13.3 mg/24 h, N = 265; 9.5 mg/24 h, N = 271). At week 24, 25 versus 14% ($p = 0.001$) of patients receiving 13.3 and 9.5 mg/24 h patch, respectively, demonstrated ≥ 4 points' improvement on ADAS-cog, and 17 versus 7% ($p < 0.001$) demonstrated ≥ 4 points' improvement on ADAS-cog and no decline on ADCS-IADL. Of patients receiving 13.3 and 9.5 mg/24 h patch, respectively, at Week 48, 16% and 10% ($p = 0.020$) displayed ≥ 4 points' improvement on

ADAS-cog, and 8% versus 4% ($p = 0.023$) demonstrated ≥ 4 points' improvement on ADAS-cog and no decline on ADCS-IADL.

Conclusion: Titration of patients with mild-to-moderate AD to the high-dose (13.3 mg/24 h) rivastigmine patch (15 cm²) increases the likelihood of achieving a clinically-meaningful treatment response.

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Abstract WCN 2013

No: 1906

Topic: 5 – Dementia

Efficacy and safety of high-dose 13.3 mg/24 h rivastigmine patch in severe Alzheimer's disease with and without concomitant memantine use

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Background: The ACTIVITIES of daily living and cognition (ACTION) study demonstrated significant efficacy of 13.3 versus 4.6 mg/24 h rivastigmine patch on SIB and ADCS-ADL-SIV in severe Alzheimer's disease (AD). 61% of the study population was on concomitant memantine (regardless of dose or duration). Here the effect of concomitant memantine use on efficacy and safety of 13.3 mg/24 h patch was investigated.

Methods: ACTION was a 24-week, randomized, double-blind study in patients with severe AD (MMSE score 3–12). Patients randomized to 13.3 or 4.6 mg/24 h patch were subdivided according to whether they received ≥ 1 dose of concomitant memantine. Change from baseline at week 24 on SIB and ADCS-ADL-SIV was assessed. Safety evaluations included incidence of adverse events (AEs).

Results: Overall, memantine-treated patients, had a lower MMSE at screening (8.6 +/- 2.91 versus 9.2 +/- 2.88). At week 24, the 13.3 demonstrated significantly greater efficacy than 4.6 mg/24 h patch on SIB and ADCS-ADL-SIV in patients receiving concomitant memantine than in those not receiving memantine ($p < 0.05$ for all). The incidence of AEs was 71.4% with 13.3 mg/24 h patch and memantine, 79.7% with 13.3 mg/24 h patch without memantine, 74.7% with 4.6 mg/24 h patch with memantine, and 71.1% with 4.6 mg/24 h patch without memantine.

Conclusions: These data suggest benefit of 13.3 mg/24 h patch, regardless of concomitant memantine use, and demonstrate similar safety and tolerability of 13.3 mg/24 h patch in memantine-treated patients and those not receiving memantine.

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Abstract – WCN 2013

No: 2308

Topic: 5 – Dementia

Anxiety levels increase chronic musculoskeletal pain in Alzheimer's disease patients

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Background: Mood disorders in Alzheimer's Disease (AD) patients result in great difficulties in clinical practice in the assessment of pain and further treatment, therefore they should be early diagnosed and managed.

Material and methods: Twenty five AD patients, with anxiety and chronic musculoskeletal pain (experimental group) and thirty one age-matched patients with Alzheimer's disease and chronic musculoskeletal pain without comorbid mood disorders (control group) were examined. All participants were diagnosed with chronic musculoskeletal pain, according to their medical history and their medications (all of them took non-steroid anti-inflammatory drugs). The neuropsychometric evaluations were performed with the following tools: Geriatric Pain Measure, Patient Health Questionnaire, Pain Assessment in Advanced Dementia, Mini Mental State Examination and Pain Anxiety Symptom Scale.

Results: AD patients with comorbid anxiety disorders tend to report increased pain intensity more frequently compared to controls. Scores in fearful thinking and physiological responses scales of PASS were higher in female than male ($p = .014$), whereas scores in the cognitive anxiety scale of PASS have shown a highly significant positive correlation with the years of education ($p < .001$).

Discussion: According to the results of the present study, anxiety is a significant component and an important part of pain experience among patients with AD, even if they are not able to verbalize it; thus needs to be taken into consideration by the health professionals for the patient's management.

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Abstract – WCN 2013

No: 2390

Topic: 5 – Dementia

Catechin suppressed LPS-induced neuroinflammation in BV-2, C6 and NG108-15 cells by modulating CCL21 through PI3K-Akt

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Background and objectives: Excessive microglial and astroglial activation that leads to chronic neuroinflammation plays a vital role in the progression of Alzheimer's, Parkinson's and Huntington's diseases. Catechin or flavan-3-ol is mainly found in tea is reported to be neuroprotective. We investigated the regulation of neuroinflammation of LPS-induced neuroinflammation in BV-2, C6 and SHSY-5Y cells.

Material and methods: The expression of NO, ROS, PGE₂, TNF- α , IL-1 β , IL-2, IL-6, IFN- γ , iNOS, COX-2, CCL21 and the molecular pathways involved were determined by Flow cytometry chip beads array (CBA), Western blot and ICC. The protective effects of catechin on BV-2 and SH-SY5Y co-cultured model induced with LPS were further evaluated.

Results: Catechin downregulated iNOS expression leading to concomitant decreased of NO and ROS released. This was followed by reduction of PGE₂ production leading to downregulation of TNF- α , IL-1 β , IL-2, IL-6, IFN- γ and COX-2 expression. Catechin downregulated CCL21 and prevented I κ B α degradation and thus, prevented p65 NF- κ B translocation in BV-2 and SH-SY5Y cells. Withal, Catechin upregulated PI3K-Akt expression, suggesting activation of pAkt might be responsible in mitigation of neuroinflammation.

Conclusion: Suppression of CCL21 in BV-2 and C6 activation through PI3K-Akt by catechin conferred protection on SH-SY5Y cells against LPS-induced neuroinflammation. This finding further heightens the specific therapeutic potential of catechin for the treatment of neurodegenerative diseases.

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Abstract – WCN 2013

No: 2400

Topic: 5 – Dementia

The impact of neurological condition on driver distraction in a driving simulator experiment: Preliminary findings

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Background: Age-related neurological disorders may affect driver distraction, and medication may also impair driving. In the early stages, neurological disorders have little impact on daily life yet may significantly impact one's driving ability. Mild cognitive impairment (MCI), the prodementia stage of various dementing disorders, is associated with impaired driving to a small extent (Frittelli et al., 2009) and self-reported road accident involvement is correlated with future diagnosis of dementia (Lafont et al., 2008). It is unknown whether distraction affects the driving ability of neurology patients to the same extent as healthy persons. Because of the ageing population, the need to investigate this question becomes critical.

Objective: To present preliminary findings from a recently funded research programme, National Strategic Reference Framework (NSRF 2007-13, O.P. Thales), on the causes and impact of driver distraction in a driving simulator experiment.

Patients and methods: At least 90 patients with neurological disorders (MCI, mild Alzheimer's disease, Parkinson's disease) will be recruited, and will be compared with at least 60 middle aged and older participants. A neuropsychological battery that measures attention, memory, visual and executive functions, a neurological exam, and a detailed history of medication and sleep difficulties are included.

Results: Preliminary findings are presented on the impact of presence/severity of neurological disorder, use of medications, and sleep disorders on driving performance with and without distraction, under different driving conditions.

Conclusion: The neurological parameters that predict driving performance under different conditions are summarized and discussed. Preliminary recommendations are presented on criteria for safe driving.

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Abstract – WCN 2013

No: 2313

Topic: 5 – Dementia

Potential effect of amyloid imaging on diagnosis and intended management of patients with cognitive decline: Impact of appropriate use criterion

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Background: Published appropriate use criteria (AUC) by Johnson et al. (2013), provide guidelines for selecting patients for whom amyloid PET could be useful.

Objectives: To evaluate the impact of amyloid PET on diagnosis and intended management in a set of patients likely to meet AUC.

Methods: We examined 229 cases from a completed study of Florbetapir amyloid PET in patients undergoing or having recently completed an evaluation for cognitive decline in which AD was suspected. All cases had a provisional diagnosis, and an intended treatment/management plan prior to PET scan, and a final diagnosis and management plan post PET scan. Based on the retrospective review of prescan diagnosis and demographics, cases were classified as likely meeting AUC (AUC-like) or not.

Results: 140/229 (61%) subjects were AUC-like. The nonAUC cases included typical AD, MCI due to AD and cognitive decline without objective evidence of impairment (CD). 67/140 (48%) AUC-like cases were amyloid positive (A β +). Within the nonAUC group, the proportion A β + ranged from 6/22 (27%) in CD patients, to 23/34 (68%) in typical AD. Diagnosis changed after PET scan for 62% of AUC cases vs 43% of nonAUC cases. The proportion of patients with change in management plan was high (>85%) regardless of AUC category or diagnosis.

Conclusions: The AUC criteria exclude patients with a relatively high (typical AD) or low (CD) probability of an A β + scan. PET amyloid imaging altered diagnosis and management in patients selected according to AUC.

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Abstract – WCN 2013

No: 2364

Topic: 5 – Dementia

α -Lipoic acid protects against LPS-induced BV-2 activation and MPTP-induced toxicity in SH-SY5Y neuronal cells

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Background and objectives: α -Lipoic acid (LA), a natural dithiol compound is a powerful antioxidant that has been used to treat various neural disorders. Microglial activation has been implicated in chronic neuroinflammation leading to neurodegenerative diseases such as Alzheimer's and Parkinson's diseases. For the first time, the anti-inflammatory effects of LA and its possible mechanisms in LPS-stimulated inflammation in microglial BV2 cells and MPTP-induced toxicity in SH-SY5Y were investigated.

Material and methods: Cell viability was measured by using MTT assay. Flow cytometry chip beads array (CBA), Western blot and ICC were used to analyse NO, ROS, PGE₂, TNF- α , IL-1 β , IL-6, IFN- γ , iNOS, COX-2, CCL21 expression and the involvement of signalling pathways such as MAPK cascades, pMTOR-PI3K-Akt and NF- κ B. The protective effects of LA on BV-2 and SH-SY5Y co-cultured model induced with LPS and MPTP were further evaluated.

Results: LA significantly attenuated LPS-induced iNOS, NO, ROS, PGE₂, TNF- α , IL-1 β , IL-6, IFN- γ and COX-2 expression as shown in CBA and Western blot. LA also suppressed the expression of CCL21, a pro-inflammatory chemokine in both LPS-treated BV-2 and MPTP-treated SH-SY5Y. Moreover, LA inhibited I κ B α degradation and thus, prevented p65 NF- κ B translocation in BV-2 and SH-SY5Y cells. LA increased cell viability and rescued co-cultured SH-SY5Y cells from MPTP-induced toxicity and apoptosis.

Conclusion: The anti-inflammatory properties of LA prevented excessive microglia (BV-2) activation and thus, protected SH-SY5Y cells from LPS and MPTP induced toxicity by downregulating pro-inflammatory proteins through PI3K-Akt pathway. This suggests a therapeutic potential of LA for the treatment of neurodegenerative diseases.

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Abstract – WCN 2013

No: 1156

Topic: 5 – Dementia

Patient and caregiver adherence and persistence to the rivastigmine patch in a non-interventional clinical study

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Background: Rivastigmine transdermal patch provides smooth drug delivery and therefore increased tolerance at similar efficacy compared to the oral formulation. An earlier registration study for the patch (IDEAL) with 1059 caregivers demonstrated that more than 70% caregivers prefer rivastigmine transdermal patch to capsules.¹

Objective: To identify patient and caregiver-related factors associated with adherence, persistence and caregiver satisfaction with rivastigmine patch.

Methods: In this non-interventional trial, adherence and persistence were assessed after three and six months of patch treatment. Relevant factors from literature for persistence and adherence to medication were validated prior to study enrolment.

Results: 127 caregivers answered the questionnaires, 3 months (visit 1) and 110 caregivers after 6 months (visit 2) after first prescription of rivastigmine patch. Mean MMSE score of the patients was 20.8 after 3 months of treatment. Physician ratings for CGI-I scores for efficacy suggested patients as 'improved' (66.2%). Caregivers were adherent to patch use at both visit 1 and visit 2 respectively, ('never' omitted 74.8%; 78.2% or 'never' paused for a while, 73.2%; 74.5%) and agreed with ease of its application. Efficacy was rated from caregivers for visit 1 and visit 2 respectively, as 'satisfied' for memory (62.2%; 56.4%), activities (69.3%; 66.4%) and behavior and emotions (65.4%; 64.5%). Valuations of tolerability were deemed as 'good' (90.5% and 90%) at visit 1 and visit 2 respectively.

Conclusion: Caregivers achieved reasonably high adherence rates to rivastigmine patch. This may be linked to its good efficacy outcomes and favourable tolerability profile.

Reference

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Abstract – WCN 2013

No: 1169

Topic: 5 – Dementia

Ease of use of rivastigmine patch can help manage medication for caregivers and patients with Alzheimer's disease

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Background: Rivastigmine transdermal patch provides smooth drug delivery and better satisfaction to caregivers and patients compared to oral formulation. An earlier study demonstrated that more than 70% caregivers prefer the rivastigmine transdermal patch to the capsule.

Objective: To identify patient and caregiver-related factors associated with adherence, persistence and caregiver satisfaction to rivastigmine patch.

Methods: In this non-interventional, clinical and psychosocial study, caregiver or patient-related factors impacting on rivastigmine patch

adherence and persistence were assessed after three and six months of treatment. Relevant factors from literature for persistence and adherence on medication were validated prior to study enrollment.

Results: 127 caregivers answered the questionnaires at visit 1 (after three months of treatment). Most caregivers were female (70.9%). 50.4% female and 45.7 male patients had caregiver assistance. The mean age of caregivers and patients was 66 and 78 years respectively. A large proportion of patients (72.1%) were living at home and caregiver assistance provided within the family (spouse, 57.4%; daughter, 21.7%). Most of the patients and caregivers had concomitant diseases [heart disease, 33.1%; diabetes, 28.3%], [mobility, 26%; vision, 16.5%]. Patients either 'preferred' or 'highly preferred' the patch (37%; 26%) compared to liquids (17.3%; 6.3%), or pills (17.3%; 2.4%). Caregivers found patch application convenient and easy ('easy' and 'very easy' package opening, 89%, 'easy' and 'very easy' use by patients, 89%).

Conclusion: Ease of use of rivastigmine patch improves satisfaction of both caregivers and patients and may help enhance the quality of AD patients' assistance and management by the caregivers.

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Abstract – WCN 2013

No: 2294

Topic: 5 – Dementia

Long-term safety and efficacy of 13.3 mg/24 h rivastigmine patch in severe Alzheimer's disease: ACTIVITIES OF DAILY LIVING AND COGNITION (ACTION) study

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Background: The 24-week, randomized, double-blind ACTION study demonstrated efficacy of higher-dose 13.3 versus 4.6 mg/24 h rivastigmine patch on cognition and ADL, with comparable safety in severe Alzheimer's disease (AD). A 24-week, open-label extension assessed long-term safety and efficacy of 13.3 mg/24 h patch.

Methods: Completers of the double-blind study (13.3 or 4.6 mg/24 h patch) could continue into the open-label extension (13.3 mg/24 h patch). Incidences of adverse events (AEs), severe AEs (SAEs) and discontinuation due to AEs were reported. Change from baseline on ADCS-ADL-SIV and SIB, and ADCS-CGIC were assessed at week 48.

Results: 397 patients entered the open-label extension (197 continued on 13.3 and 199 up-titrated from 4.6 to 13.3 mg/24 h patch). Baseline characteristics were comparable. Similar rates of AEs (58% and 60%), SAEs (16 and 16%) and discontinuation due to AEs (11 and 12%) were reported in 13.3 and 4.6 mg/24 h, resp. Larger mean [SD] changes from double-blind baseline were observed in patients switched from 4.6 to 13.3 mg/24 h patch on ADCS-ADL-SIV (−4.6 [8.73]) and SIB (−7.0 [16.56]), than those who continued on 13.3 mg/24 h (−3.9 [8.00] and −4.7 [16.84], resp.). ADCS-CGIC scores were comparable.

Conclusions: There were no clinically relevant differences in safety between patients switched from 4.6 to 13.3 mg/24 h and those who continued on high-dose patch in the extension study. Greater decline was observed in patients switched from 4.6 to 13.3 mg/24 h patch at week 24, compared to those who received 13.3 mg/24 h patch for 48 weeks.

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Abstract – WCN 2013

No: 2382

Topic: 5 – Dementia

Cerebrospinal fluid protein and white cell count as potential markers for clinical progression in Alzheimer's disease

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Background: Neuroinflammation may have an important role in the pathophysiology and the clinical progression of Alzheimer's Disease (AD). CSF protein concentration (due to defects in the blood-brain barrier) and white cell count are potential markers of such processes.

Objective: To investigate whether an elevated CSF protein concentration or an elevated CSF white cell count at baseline can predict clinical progression from MCI to AD.

Patients and methods: Routine CSF parameters (white cell count, protein concentrations, albumin CSF/serum concentrations) as well as CSF-AB42, CSF-Total Tau were retrospectively analyzed in a consecutive cohort referred for cognitive evaluation and diagnosed with MCI.

Clinical progression was assessed by clinical history and cognitive tests (MMSE) administered at baseline and during follow-up.

Results: Thirteen MCI subjects (4 females, 9 men) were included (mean age 69.6, range 56 to 80 years), mean MMSE 28, range 26 to 30). Patients had a mean follow-up of 26 months (range 14 to 46 months).

Eleven patients had stable MCI during follow-up, and 2 patients progressed from MCI to AD. In 4 subjects an elevated CSF-protein (above 0.45 g/L) concentration was found, and in two of these the CSF/plasma albumin-coefficient was also increased (above 0.012). One subject had elevated white cell count (above 3).

None of the 2 patients with clinical progression had increased CSF protein concentrations nor elevated white cell count.

Conclusion: From these very preliminary data we found no association between CSF protein concentration or white cell count and clinical progression of MCI. Analyses on a larger cohort are on-going.

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Abstract – WCN 2013

No: 1566

Topic: 5 – Dementia

Various types of visual agnosia are present in Alzheimer's disease and mild cognitive impairment

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Background: Higher visual perception, which includes identification and recognition of faces, facial emotions and famous landmarks, is dependent on the medial temporal lobe structures that are affected early in the course of Alzheimer's disease (AD). However, examination of higher visual perception in patients with dementia due to AD

and patients with mild cognitive impairment (MCI) is still neglected in routine clinical practice.

Objectives: To determine whether famous faces identification (FFI), facial emotion recognition (FER), and famous landmarks identification (FLI) are impaired in patients with amnesic (aMCI) and non-amnesic (naMCI) MCI and mild AD.

Patients and methods: A total of 40 patients with aMCI, 27 patients with naMCI, 20 with mild AD and 38 controls were recruited. All subjects underwent standard neurological and neuropsychological evaluations as well as tests of FFI, FER and FLI.

Results: The subjects were similar in age and gender, but AD patients were less educated than other groups (p 's $\leq .016$). A one-way analysis of covariance adjusted for education was used. Compared to the control group, AD subjects performed worse on FFI ($p = .041$), FER ($p < .001$) and FLI ($p < .001$), whereas the aMCI and naMCI groups had significantly worse scores only on FLI ($p = .049$ and $p = .022$, respectively).

Conclusion: Patients with amnesic and non-amnesic MCI may have impairment of higher visual perception restricted to FLI, while patients with dementia due to AD may have more extensive impairment of higher visual perception. The results suggest that FLI testing may be a useful tool for identification of patients with prodromal AD.

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Abstract – WCN 2013

No: 2417

Topic: 5 – Dementia

Role of 5-HTTLPR polymorphism in vascular dementia

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Background: Early detection and accurate diagnosis of vascular dementia (VD) are important, as vascular dementia is at least partially preventable. Therefore there is a need to optimize and increase panel of diagnostic criteria for this disease. The 5-HTTLPR genetic variant of the serotonin transporter gene might also serve as a marker of vascular dementia.

Methods: The 5-HTTLPR polymorphism was genotyped in 272 subjects (110 males and 162 females) who fulfilled the diagnostic criteria for vascular dementia. Severity of disease was assessed by MMSE, HADS, clock drawing test and Orgogozo scale.

Results: Positive association was shown for males with SS genotype and early onset of vascular dementia: average age of onset for patients with SS genotype was 57 years, while for LL genotype – 63 years (Test M–W, $p = 0.031$). SS genotype was also associated with deterioration of cognitive functions: impaired attention ($p = 0.043$) and high results of clock drawing test ($p = 0.039$). No statistical difference was found for heterozygotes.

For females role of 5-HTTLPR polymorphism was less important. Some differences in severity of speech and language impairment were identified.

Conclusion: Based on this study the 5-HTTLPR polymorphism was shown as prognostic marker of vascular dementia for males. SS haplotype indicates an increased risk of early onset of VD and more severe progression.

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Abstract – WCN 2013

No: 2121

Topic: 5 – Dementia

Higher plasma concentrations of total carotenoids are associated with slower cognitive decline: Results from the Austrian Stroke Prevention Study

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Background: Antioxidant properties of micronutrients including vitamins and carotenoids may help to prevent cognitive decline.

Objective: To investigate the cross-sectional relationship between antioxidative micronutrients and cognitive function particularly in the domains of memory, attention–speed, conceptualization, visuo-practical skills and global cognitive function (g-factor) in the normal elderly. Additionally we investigate mediation by MRI correlates of brain atrophy and white matter lesions (WML) volume.

Patients and methods: The cohort consisted of 614 healthy participants (mean age = 66 years, 58% females) of the Austrian Stroke Prevention Study. Cognitive function measurements include composite scores of specific domains including memory, conceptualization, visual practical skills and attention–speed and g-factor. All subjects underwent brain MRI with assessment of brain parenchymal fraction (BPF, %) and measurement of WML volume (cm³). Principal component analysis constructed four nutrient plasma antioxidant patterns (NPAPs) from original set of ten antioxidants. Association between NPAPs scores and different cognitive functions was analyzed using multiple linear regressions by adjusting for age, sex, education (Model1) and by additionally adjusting for vascular risk factors such as hypertension, cardiovascular disease, diabetes, smoking, BMI, Apo 4 and HDL (Model2).

Results: We observed only NPAP-1 including β -cryptoxanthin, canthaxanthin, lycopene, α -carotene, β -carotene positively and significantly associated with conceptualization (Model1: $\beta = .043$, $p = .05$) and g-factor (Model1: $\beta = .068$, $p = .03$) when adjusted for age, sex, and education. The association remained significant when additionally adjusted for vascular risk factors (Model2: conceptualization ($\beta = .054$, $p = .02$), g-factor ($\beta = .068$, $p = .05$)). The effect of NPAP-1 on conceptualization and g-factor is not mediated by WML and brain atrophy.

Conclusion: Our result suggests the protective role of total carotenoids on cognitive function, which may ameliorate age-related cognitive decline.

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Abstract – WCN 2013

No: 2452

Topic: 5 – Dementia

PGRN and MAPT gene mutations cause of frontotemporal lobar dementia in Polish population

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Background: Frontotemporal lobar degeneration, the second most common form of presenile dementia, is clinically, pathologically and

genetically heterogeneous disorder. Up to 40% of FTD patients report family members with FTD supporting the important contribution of genetic factors to these diseases.

Objective: The aim of the study was to establish genetic backgrounds and frequencies of mutation causing FTD in the Polish population.

Patients and methods: The studied group consisted of 120 FTD patients (mean age of onset 63.6 ± 7.8 years), including 44 familial cases. All *PGRN* and *MAPT* exons with flanking intronic regions were sequenced. Analysis of hexanucleotide repeat expansion in C9ORF72 is in progress.

Results: We identified six different non-synonymous variants (*PGRN*: R418X, P439_R440fsX6, R433Q; *MAPT*: G55R, P301L, S305N). Among them two variants were new ones, three were previously reported as pathogenic mutations, and one variant was already known as non-pathogenic polymorphism. All coding non-synonymous variants were observed in familial FTD cases.

Conclusion: Causative mutations in *MAPT* and *PGRN* occur approximately in 4% of all FTD cases and in 11% of familial FTD cases in the Polish population. It could be concluded that *PGRN* and *MAPT* mutations are infrequent cause of FTD in Polish population. The symptoms of FTD are very variable and similar symptoms can result from mutations in different genes. Moreover the same mutation can present different symptoms in the same family.

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Abstract – WCN 2013

No: 2414

Topic: 5 – Dementia

Cognitive decline in older age and metabolic profile in middle age: Is there an association?

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Background: Cognitive impairment and dementia, either from vascular and degenerative origin, have been associated with metabolic disorders. However, there are no population-based studies comparing cognitive decline (CD) in older age and metabolic profile decades earlier.

Objective: To evaluate the association between CD in older age and serum metabolites (glycaemia, total cholesterol, HDL cholesterol, triglycerides, ALT) measured twenty years before.

Patients and methods: The MICOL cohort included a random sample of 2472 subjects recruited and examined in 1985–1986 and followed-up over 20 years until 2005–2006. In 2005–2006, 855 subjects of the cohort had ≥ 65 years and their cognitive status was explored using MMSE. Metabolic profile at baseline was compared in subjects with and without CD twenty years later.

Results: 797 subjects out of 855 answered MMSE, median score 29 (range 1–30). Six hundred eleven subjects (77%) had a MMSE score > 24 (normal), 127 (16%) between 21 and 24 (mild cognitive decline = MCD), and 59 (7%) < 20 (moderate/severe CD = MSCD). ANOVA analysis with Bonferroni correction showed that mean fasting glycaemia at baseline in MSCD group (115.8 ± 73.7 mg/dl) was significantly higher than fasting glycaemia in both normal (101.4 ± 20.8) ($p = 0.0005$) and MCD (99.7 ± 18.2) ($p = 0.0009$) groups. Total

cholesterol, HDL-cholesterol, triglycerides and ALT were not different between groups. Multinomial logistic regression adjusting for age, gender and educational level showed that high fasting glycaemia is associated with an increased risk of MSCD.

Conclusions: Cognitive decline in older age is associated with high fasting glycaemia, measured up to twenty years before, also controlling for main confounders.

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Abstract – WCN 2013

No: 2451

Topic: 5 – Dementia

DPPX-related rapidly progressive dementia

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In the past, rapidly progressive dementias were thought to be easily distinguished from more common “classical” slowly progressive dementias. More recently, diseases distinguished most commonly as causing “classical” slowly progressive dementias, including Alzheimer’s disease and dementia with Lewy bodies, have been shown to produce also rapidly progressive dementias, while diseases distinguished as causing rapidly progressive dementias have been shown to lead to slowly progressive dementias. This is also the case of paraneoplastic disorders. While in the past these disorders were considered as exclusively related to neoplasias, recent evidence suggests that many, if not the majority of them, represent in fact an autoimmune disorder rather than a paraneoplastic one. Accumulating evidence suggests that many autoimmune rapidly progressive dementias result from aberrant action of autoantibodies erroneously targeting synaptic proteins. Best described are autoimmune rapidly progressive dementias due to aberrant action of autoantibodies against voltage gated potassium channels and against N-methyl-D-aspartate receptor. Last year, rapidly progressive dementia due to aberrant action of autoantibodies against dipeptidyl-peptidase-like protein-6 was first described. This form of rapidly progressive dementia targets a subunit of Kv4.2 potassium channels, which play a role predominantly in regulating post-synaptic somatodendritic signaling. Here we present the first thorough description of clinical characteristics, results of cerebrospinal fluid and imaging analyses as well as neuropathological findings.

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Abstract – WCN 2013

No: 2478

Topic: 5 – Dementia

The effect of the neuroprotector NT-1505 on the structural parameters of mice membranes *in vivo*

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Background: NT-1505 influence on lipid composition and fluidity of the mice membranes was studied.

Objective: Lipid component of membranes plays an important role in the transmission, processing and storage of information in the cell. Activation of lipid peroxidation processes in AD leads to a change in the membranes’ structural organization. Disruptions in the regulation system work of membranes’ cell metabolism allow considering AD as membrane pathology. Cognition enhancer NT-1505 is the most promising substance for AD treatment, which modifies glutamate receptors functioning [1]. It was an apparent interest to study the influence of NT-1505 on the membrane structural state for the

estimation of the possibly favorable or unfavorable side effects during therapy. In this work the effect of neuroprotector NT-1505 on the lipid composition and fluidity of the mice membranes *in vivo* was studied.

Material and methods: The experiment was carried out on scrub mice. NT-1505 was kindly furnished to us by Bachurin S.O. Membrane microviscosity was measured by electron paramagnetic resonance (ESR) with 2,2,6,6-tetramethyl-4-capryloyl-oxypiperidine-1-oxyl and 5,6-benzo-2,2,6,6-tetramethyl-1,2,3,4-tetrahydro- γ -carboline-3-oxyl probes. Rotational diffusion correlation time calculated from the obtained spectra of the ESR as microviscosity. Lipid composition was determined by mass-spectrometry and TLC methods.

Results and conclusions: Obtained data shows, that NT-1505 reduces microviscosity of the synaptosomal membranes and modifies fluidity of the microsomal membranes. NT-1505 significantly influences lipid composition of the membranes. It was found out that changes in membrane structural state are preferred to be considered during therapy scheme development.

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Abstract – WCN 2013

No: 2324

Topic: 5 – Dementia

Molecular genetics investigation of 5HTR2A gene in frontotemporal lobar dementia

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Frontotemporal lobar dementia (FTLD), characterized by neuronal loss in the frontal and/or temporal cortices, includes behavioural variant of frontotemporal dementia (bvFTD), semantic dementia and progressive non-fluent aphasia subtypes. Mutations in the associated genes do not explain the majority of cases and the mechanism underlying the aetiology of FTLD remains unknown. The genes encoding for serotonergic (5-HT) components are good candidates for susceptibility factors, due to their role in human behavior modulation.

Our aim was to analyze the involvement of 5HTR2A gene in FTLD pathogenesis.

We have studied patients with probable diagnosis of FTLD, followed in dementia consultation, and healthy controls.

Analysis of 5HTR2A gene was performed by automated sequencing and statistical analysis using χ^2 and Fisher's exact tests (significance if $p < 0.05$).

We have found 17 sequence variations (13 known and 4 novel unreported) that possibly do not account for primary risk for FTLD, but may contribute for susceptibility.

The case-control association study suggests that the increase of G allele (rs3125; 3'UTR region) may be a risk factor for bvFTD in females ($p = 0.037$; OR = 2.5, CI 95%). Determination of mRNA levels has been initiated to further clarify its role in the disease.

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Abstract – WCN 2013

No: 1081

Topic: 5 – Dementia

Apolipoprotein E plasma level and genotype – Risk of dementia in 76,000 individuals from the general population

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Background: Alzheimer disease and other forms of dementia are devastating neurodegenerative diseases affecting more than 35 million people worldwide, with no currently available curative treatment. The apolipoprotein E (APOE) $\epsilon 4$ allele is a major genetic risk factor for dementia. In contrast, it remains unclear whether plasma levels of apoE confer additional risk.

Objective: We tested whether both plasma levels of apoE and APOE genotype associate with dementia.

Material and methods: We included 76,365 participants from the Copenhagen General Population Study and the Copenhagen City Heart Study; 1216 developed dementia. We measured plasma apoE levels and genotyped for rs429358 and rs7412, which combines into six common APOE genotypes ($\epsilon 22$, $\epsilon 32$, $\epsilon 42$, $\epsilon 33$, $\epsilon 43$, $\epsilon 44$).

Results: Multifactorially adjusted hazard ratios (HRs) increased from highest to lowest tertile of apoE levels and from $\epsilon 22$ to $\epsilon 44$ for Alzheimer disease (p for trend = 9.3×10^{-13} and 2.7×10^{-37} , respectively), and for all dementia (p for trend = 8.0×10^{-11} and 5.9×10^{-37} , respectively). After further adjustment for APOE genotype, apoE levels remained significantly associated with Alzheimer disease ($p = 0.005$), and after stratification for APOE $\epsilon 43$ genotype, apoE levels remained significantly associated with Alzheimer disease ($p = 0.04$), and with all dementia ($p = 0.02$). These associations were independent of cardiovascular risk factors.

Conclusion: Both plasma apoE levels and APOE genotype were associated with risk of dementia, and the association between plasma levels of apoE and dementia was independent of APOE genotype.

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Abstract – WCN 2013

No: 2473

Topic: 5 – Dementia

Thymoquinone improves cognitive function and increases α -7nAChRs expression in a rat model of neuroinflammation

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Background: Alzheimer's disease (AD) is a neurodegenerative disorder characterized by extracellular deposition of aggregated β amyloid protein ($A\beta$). Recently, correlation between nicotinic receptors (nAChRs) dysfunction, neurodegeneration and cognitive deficits was shown in AD. nAChRs have protective role against $A\beta$ toxicity while chronic inflammation damages cholinergic neurons and contributes to neurodegeneration. Thymoquinone (TQ) has anti-oxidant and anti-inflammatory effects besides its acetylcholine modulatory effects *in vitro*. However, effect of TQ on nAChRs *in vivo* needs to be clarified.

Aim: To investigate *in vivo* effects of TQ on cognitive functions and α -7nAChRs expression in a LPS model of neuroinflammation.

Methods: Rats injected with LPS (0.8 mg/kg) were divided into three groups for a five-day-treatment: TQ (5 mg/kg), TQ (10 mg/kg) and control group (0.08% Tween 80). Learning, memory and locomotor

activity were tested using Y-maze, object recognition and open field tests. Brain α -7nAChR protein level was measured by western blot.

Results: LPS significantly reduced alternation % in Y-maze, pattern separation in object recognition test and locomotor activity in open field test. TQ significantly improved these LPS-induced cognitive functions and locomotor activity impairment. 10 mg/kg TQ significantly increased α -7nAChRs protein level compared to the other groups.

Conclusion: This study demonstrates beneficial *in vivo* effects of TQ by improving learning, memory and locomotor activity in a model of chronic neuroinflammation. These effects were accompanied by significant increase in α -7nAChRs protein expression level. These results confirm the impact of neuroinflammation on the cholinergic signaling and suggest TQ as a potential promising treatment for AD.

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Abstract – WCN 2013

No: 2523

Topic: 5 – Dementia

Cerebral amyloid angiopathy presenting as a diffuse leukoencephalopathy

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Introduction: Amyloidosis is a heterogeneous group of disorders, with extracellular deposits of misfolded proteins, in a fibrillar configuration. Amyloid deposits in brain can take many forms: senile plaques (in Alzheimer's disease), amyloidomas (mimicking brain tumours) or cerebral amyloid angiopathy (CAA) by β -amyloid deposits in meningeal and cortical vessels. Spontaneous cerebral hemorrhage is the usual presentation of CCA. CAA presenting with subacute encephalopathy associated to diffuse white matter disease is extremely rare.

Clinical case: A 75 year-old man without relevant medical or toxic exposure history began with progressive attention and memory difficulty. One year later presented with moderate headaches, confabulation and urinary retention. Objectively had symmetrical head and limbs resting tremor, without pyramidal, sensory or cerebellar signs. Brain MRI showed bilateral fronto-temporal-insular symmetrical white matter lesions, with mass effect, extending to external capsules and sparing basal nuclei. Electroencephalogram suggested diffuse brain dysfunction. CSF had 0.71 g/L of proteins without cells. Metabolic, microbiological, immunological and occult malignancies investigation were negative. Brain MRI one month later showed lesion expansion. He didn't improve with methylprednisolone 1 g during 5 days. Brain biopsy diagnosed A β cerebral amyloid angiopathy. Patient died by an urosepsis 1.5 month after diagnosis.

Discussion: CAA manifested as diffuse white matter disease is a rare and challenging condition. An inflammatory process is thought to be related to the vascular deposits of A β in this presentation. We didn't identify an inflammatory process in the biopsy. In our case the patient didn't respond to steroids, the proposed standard treatment, and there was a fast worsening of the condition.

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Abstract – WCN 2013

No: 2560

Topic: 5 – Dementia

Synergistic effects of rTMS and cognitive training in Alzheimer's Disease

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Background: Current treatment options for Alzheimer's Disease (AD) show limited benefits. Transcranial Magnetic Stimulation (TMS) can be used to explore brain alterations and has a modulatory impact on brain plasticity and may therefore enhance the effects of therapy. We are evaluating a novel device, which combines rTMS with computerized cognitive therapy (NeuroAD™, Neuronix Ltd., Israel).

Objectives: Evaluate rTMS for enhancement of cognitive training effects.

Compare TMS-measures in mild AD and healthy controls (HC).

Patients and methods: Measures: Alzheimer's Disease Assessment Scale (ADAS-Cog), Clinical Global Impression of Change (CGIC), Activities of Daily Living (ADAS-ADL), motor threshold (MT), brain reactivity and plasticity, brain-scalp distances (BSD).

Patients received 6 weeks of daily active or sham treatment. During active treatment computerized cognitive training was combined with rTMS in an interweaved fashion. The six stimulated regions were related each with the treated brain sites.

Results: MTs in AD and HC were comparable, while baseline reactivity as well as maximum MEP change was significantly different. BSD was significantly reduced in M1 and was correlated with brain reactivity. While SICI and ICF were comparable, LICI was reduced in AD.

The real treatment group improved significantly in ADAS-Cog as compared to sham after treatment (real: -6.74 ± 3.25 ; sham: 0.66 ± 2.91). Non-significant improvement was observed in CGIC, while groups improved similarly in ADAS-ADL. Treatment groups did not change significantly regarding M1 reactivity and plasticity after training.

Conclusions: NeuroAD is a promising tool to enhance therapy effects.

Alterations in TMS-EMG measures are associated with brain atrophy.

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Abstract – WCN 2013

No: 2602

Topic: 5 – Dementia

Cholesterol and ultrasensitive C-reactive protein serum levels among patients suffering memory recall deficit

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Background: Metabolic syndrome patients have been shown to suffer arteriolar intracranial arteriosclerosis. This in turn leads to subcortical dysfunction expressed as memory recall deficit. So far, until now, memory deficit hasn't been studied among those patients with subtle cholesterol disturbances.

Objective: To present a series of cases of patients with serum cholesterol fraction levels and memory recall deficit.

Material and methods: 16 patients with complaints of subtle memory recall deficit undertook a routine medical checkup at Medica Sur Hospital Integral and Diagnosis Center, which included clinical laboratories tests and image studies assessments.

Results: It has been observed that memory recall deficit was more frequently reported in patients with low HDL cholesterol serum levels, rather LDL cholesterol serum levels. Ultrasensitive C-reactive protein serum levels were higher than 1.7 Mg/L. Only two patients

did not suffer sleep disturbances. All of them noticed difficulties in memory recall 5 years prior to the checkup.

Discussion: The current findings demonstrated that a mild memory deficit is not a late clinical expression in metabolic syndrome. The fact that low HDL cholesterol serum levels were more frequently found is a rare finding and it does not correlate to a specific lipoprotein phenotype.

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Abstract – WCN 2013

No: 2598

Topic: 5 – Dementia

Association between asymptomatic carotid stenosis and cognitive function: A systematic review

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Background: Asymptomatic carotid stenosis (CS), defined as the condition in which patients have carotid stenosis without a past history and current clinical evidence of any prior cerebrovascular event, and have traditionally been considered clinically silent. It is less clear whether asymptomatic CS itself is an independent risk factor for cognitive impairment.

Methods: We conducted a systematic review of literature using the Cochrane Library, MEDLINE, EMBASE and the China National Knowledge Infrastructure databases. We also searched the reference lists of relevant studies and review articles. Two reviewers used a standardised form to collect data and assess eligibility. The quality of study was assessed by the Newcastle–Ottawa Scale.

Results: A total of ten studies comprising of 763 participants in the CS group and 6308 in the non-CS group were included. All studies but one support the association between asymptomatic CS and cognition impairment. When examining the concomitant factors of asymptomatic CS and cognition in seven eligible studies, age (two studies) and reduced cerebrovascular reactivity (two studies) showed positive results associated with cognitive impairment.

Conclusions: These results suggest that rather than being clinically silent, asymptomatic carotid stenosis may be associated with cognitive function impairment, which needs to be further investigated with high-quality studies.

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Abstract – WCN 2013

No: 2623

Topic: 5 – Dementia

Age of onset of dementia is delayed by multilingualism and advanced by stroke and rural dwelling independently

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Background: Several lifetime experiences such as education, occupation, and intellectually stimulating activities delay the age of onset of dementia by improving the “cognitive reserve”. Midlife vascular risk factors and stroke, on the other hand are important factors that advance the age of dementia onset.

Objective: The study was undertaken to investigate the association between various lifetime experiences, demographic and midlife vascular risk factors on the age of dementia onset in India, a developing country characterized by a unique profile of demographic and lifestyle factors.

Patients and methods: Six hundred eight dementia patients diagnosed in a specialist cognitive clinic in Hyderabad, India were included. The association of the age of onset of dementia with potentially impacting variables was examined using ANOVA and logistic regression analysis.

Results: The men/women ratio was 424:224. The mean age of the onset of dementia was 63.8 years. 550 (84.9%) patients were literates, 149 (25.9%) patients were from rural areas. 161 had stroke. The age of dementia onset was found to be associated with gender, education, occupation, multilingualism, vascular risk factors, occupation and area of dwelling. Logistic regression analysis revealed that among all these factors, stroke ($\beta = .176, p = .001$) and rural dwelling ($\beta = 0.244, p < 0.001$) emerged to be independently associated with earlier age of dementia onset while multilingualism ($\beta = 0.12, p = 0.016$) was associated with later age of onset.

Conclusion: The age of dementia onset in India is impacted by the combined effect of three major factors unique to its setting, with stroke and rural dwelling associated with an earlier age of onset and multilingualism with a delayed age of onset of dementia in its population.

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Abstract – WCN 2013

No: 1375

Topic: 5 – Dementia

Is AD a medical notion of dementia worth keeping in neurosciences?

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Background: The eponym of AD has been around since 1910. Its diagnosis varies from senile dementia to senium praecox to MCI leading to AD as the most feared form of dementia.

Objective: Most neuroscientists have refused to admit such uncertainties. They prefer to:

- (1) perpetuate AD to keep it alive for contrast with other forms of dementia;
- (2) debate on the effect to seek a one-to-one cause–effect relationship, Amyloid-Beta Hypothesis;
- (3) advocate the intervention or prevention of AD in confusion with dementia, by using animal models to mimic AD;
- (4) take the liberty to assign lesion sites, unaware of Auguste's brain that had widespread atrophy, as she had four vascular disorders.

Methods: Dementia can be tested as the effect of brain atrophy; it is a cluster of behavioral alterations.

Results: Dementia is therefore neither a disease nor equivalent to AD. It is an illness. Plaques and tangles should be called Fischer's disease which results in simple dementia leading eventually to presbyophrenia, a dichotomy suggested by Fischer. MCI is a cheap reinvention of Fischer's dichotomy.

Conclusions:

- (1) AD is not worth keeping;
- (2) dementia can start from atrophy in any brain location, then spreads, because of wear and tear;
- (3) the cause is vascular or non-vascular in origin; the effect can vary, resulting in various behavioral alterations; and

- (4) there is no one-to-one cause–effect relation, as the brain atrophy spreads from cortical to cortical, cortical to subcortical, and/or subcortical to cortical regions.

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Abstract – WCN 2013

No: 2344

Topic: 5 – Dementia

The neuroprotective effects of lithium chloride (LiCl) on anxiety behavior in animal models of Alzheimer's disease

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Introduction: One of the neurological disorders associated with Alzheimer disease (AD) includes anxiety. Trimethyltin (TMT), as a tool to model AD in animals, is a potent neuro-toxicant which causes damage particularly in the hippocampus, having the potentiality of decreasing BDNF, while it increases TNF- α . Likewise, lithium has neuroprotective characteristics which decrease neuro-atrophy progress and prevent neuro-cell death, inducing expression of BDNF and decreasing that of TNF- α .

Method: In this study, the neuroprotective effects of LiCl on the treatment of Alzheimer-related anxiety were investigated. To begin with, the rats were subjected to AD using intra-peritoneal injection of 8 mg/kg of trimethyltin. The subjects were randomly divided into the following groups: non operated rats (control), TMT injection + saline administered rats (vehicle treated rats) and TMT injection + LiCl administered rats (Test group). The last step involved taking behavioral tests of open-field and elevated-plus-maze, trying to measure the anxiety of the rats. Then, hippocampal and serum levels of TNF α and BDNF were calculated.

Results: The results indicated that the rats with TMT injection showed more anxiety and that treatment with LiCl would produce a significant improvement in elevated plus maze and open field tests. With the presence of LiCl, the BDNF would be increased and the TNF would be decreased to a significant level.

Conclusion: Injection of LiCl had neuroprotective effects and could be used as a treatment for Alzheimer-related anxiety in an animal model.

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Abstract – WCN 2013

No: 2370

Topic: 5 – Dementia

Neuroprotective effect of lithium chloride on trimethyltin-induced learning and memory deficits in rats

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Objective: In order to determine the neuroprotective effect of lithium chloride (LiCl), the present study examined the effects of LiCl on learning and memory in a Morris water maze task and BDNF level of rats with trimethyltin (TMT)-induced neuronal and cognitive impairments.

Methods: The rats were randomly divided into the following groups: non operated rats (control), TMT injection + saline administered rats (vehicle treated rats) and TMT injection + LiCl administered rats (test group). Rats were administered saline or LiCl (20, 40 and 80 mg/kg, i.p.) daily for 2 weeks, followed by their training to the tasks. In the water maze test, the animals were trained to find a hidden platform in a fixed position during 5 days and then received a

60 s probe trial on the 6th day following removal of the platform from the pool.

Results: Rats with TMT injection showed impaired learning and memory of the tasks and treatment with LiCl (specially, in lower dose of LiCl) produced a significant improvement in escape latency to find the platform in the Morris water maze. Consistent with behavioral data, treatment with LiCl also slightly enhanced serum and hippocampal levels of BDNF compared to the vehicle treated group.

Conclusion: These results demonstrated that LiCl has a protective effect against TMT-induced neuronal and cognitive impairments. The present study suggests that LiCl might be useful in the treatment of TMT-induced learning and memory deficit.

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Abstract – WCN 2013

No: 2631

Topic: 5 – Dementia

Core cell cycle protein Mad2 is differently expressed in AD brains compared to age matched controls

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Background and objectives: Anaphase promoting complex (APC), BubR1 and Mad2 are proteins of the late phase of the cell cycle which have shown to express different functions than cell cycle control processes in post mitotic neurons versus mitotic cells. BubR1 and Mad2 proteins are inhibitors of the anaphase promoting complex (APC) thus regulating the anaphase cell cycle checkpoint control ensuring proper chromosome segregation and separation. Here we wish to elucidate the possible role of Mad2 protein in an Alzheimer's Disease (AD) brains versus age matched controls.

Methods: Immunohistochemical and Western blot analysis were used to evaluate the levels and expression patterns of Mad2 protein in the hippocampal region of AD brains compared to age matched controls.

Results: Our results show that Mad2 protein is differentially expressed, i.e. we found a decreased expression of Mad2 protein in AD brains compared to age matched controls.

Conclusion: The activity of APC is maintained by CDH1 in terminally differentiated cells. It is conceivable that Mad2 protein plays a role in the control of the APC activity in post-mitotic neurons. Our results show changes in expression in the AD brain which may lead us to the possible role of MAD2 in neuronal cell cycle re-entry, thus presenting a novel pathway to AD neurodegeneration.

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Abstract – WCN 2013

No: 2469

Topic: 5 – Dementia

From prevalent MCI to incident dementia as predicted by baseline neuropsychiatric symptoms: The Mayo Clinic Study of Aging

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Objective: To determine the incidence of dementia as predicted by baseline neuropsychiatric symptoms in subjects with prevalent mild cognitive impairment (MCI).

Design: Prospective cohort study.

Setting: The Mayo Clinic study of aging.

Participants: Baseline Neuropsychiatric Inventory Questionnaire data were available on 332 people with prevalent MCI who underwent at least 1 follow-up visit.

Method: The diagnosis of dementia was made by an expert consensus panel after reviewing neurological, cognitive, and other pertinent data. We calculated hazard ratios (HRs) and 95% confidence intervals (95% CI) by using a Cox proportional hazards model, with age as a time scale.

Main outcome measure: Dementia as measured by DSM-IV criteria.

Results: We prospectively followed the cohort to incident dementia for a median (inter-quartile range [IQR]) of 3.0 (2.5, 5.3) years. HR (95% CI) was calculated after adjusting for age, sex, education, and medical comorbidity. Baseline agitation (HR = 1.97; 95% CI, 1.13–3.42), nighttime behaviors (HR = 1.68; 95% CI, 1.02–2.78), depression (HR = 1.63; 95% CI, 1.10–2.41) and apathy (HR = 1.62; 95% CI, 1.03–2.54) significantly predicted incident dementia. Conversely, anxiety (HR = 0.93; 95% CI, 0.54–1.61) and irritability (HR = 1.00; 95% CI, 0.61–1.67) do not appear to be related to increased dementia incidence. Psychotic symptoms were rare and none of them reached statistical significance.

Conclusion: Among persons with prevalent MCI, the presence of baseline agitation, nighttime behaviors, depression and apathy predicted incident dementia. Anxiety and irritability were not significant predictors.

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Abstract – WCN 2013

No: 2586

Topic: 5 – Dementia

Profile and predictors of vascular cognitive impairment in African stroke survivors: The CogFAST – Nigeria study

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Background and purpose: Sub-Saharan Africa faces a growing prevalence of non-communicable diseases including stroke and dementia but little is known about the burden of stroke-related cognitive dysfunction. We assessed baseline profiles and predictors of vascular cognitive impairment (VCI) in Nigerian African stroke survivors participating in the Cognitive Function After Stroke (CogFAST) Nigeria Study.

Methods: We recruited 217 subjects (≥ 45 years old) comprising of 143 stroke survivors and 74 demographically matched stroke-free healthy controls. We obtained demographic, clinical and lifestyle information and assessed the cognitive status of the subjects at baseline three months after stroke. Standard neuropsychological tests included the Vascular Neuropsychological Battery which assessed executive function/mental speed, memory, language, and visuospatial/visuoconstructive functioning. Cognitive impairment and dementia were defined based on the AHA/ASA VCI guidelines and the DSM-IV criteria.

Results: Among the stroke survivors, 57 (39.9%) had cognitive impairment no dementia (CIND) while 12 (8.4%) were demented at baseline. Multivariate analysis revealed that older age ($p = 0.005$, OR = 1.05 (1.00–1.09)), and low education ($p < 0.001$, OR = 5.09 (2.17–11.95)) independently predicted cognitive dysfunction whereas pre-stroke daily intake of fish ($p = 0.022$, OR = 0.39 (0.15–0.89)) was inversely associated.

Conclusion: These results suggest high frequency of early VCI in older Nigerian stroke survivors. Apart from older age, educational level and pre-stroke diet were identified as modifiable factors. This emphasizes the vital role of education and healthy nutrition in ameliorating delayed brain injury after stroke.

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Abstract – WCN 2013

No: 2534

Topic: 5 – Dementia

Hospitalist care perceptions for inpatients with dementia

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In the United States, there are currently over 28,000 hospitalists actively practicing inpatient medicine. Hospitalists care for an ever increasing percentage of inpatients with Dementia in the US. This study is designed to understand how hospitalists are perceived in providing this care. A double blinded questionnaire scaled 1–10 surveying 42 nurses, 12 internists, and 4 neurologists regarding several aspects of inpatient dementia care was distributed. The surveys were collected in sealed envelopes and results independently verified. Patients of hospitalists received fewer medications for agitation, according to nurses (87%) and internists (63.2%), however were felt to receive more medications by neurologists (68%). Hospitalists were perceived to provide clear communication to families by nurses (83%) average communication by neurologists (74%) and average by internists (89%). Patients' overall status was felt to be unchanged by care provider, hospitalist vs non hospitalist compared by nurses (92%), improved by neurologists (78%) and worsened by internists (84%).

Based on these results, hospitalists are perceived to prescribe less medications for agitation by all other care providers, provide clear communication by nursing but average communication by internists and neurologists and the patients overall status is perceived as unchanged by nurses, improved by neurologists and worsened by referring internists.

Further studies to validate these perceptions are needed. Once validated, hospitalists can work to strengthen their perceived abilities in providing inpatient dementia care.

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Abstract – WCN 2013

No: 1225

Topic: 5 – Dementia

The Rowland Universal Dementia Assessment Scale (RUDAS) in an Arabic speaking population with limited schooling

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Background: We recently validated RUDAS in 242 Lebanese participants older than 65 years, 142 cognitively intact controls and 100 dementia cases. 50.4% are illiterate. With a cutoff of 22, the Arabic RUDAS demonstrated good sensitivity (84.0%) and specificity (84.5%), but a high false positive rate (15.5%, FPR). 99% of the

illiterate controls failed the RUDAS drawing test (a cube, 3 points on 30 points total).

We aimed to modify the RUDAS drawing test to improve the FPR among illiterate older people.

Method: The same participants were asked to draw two intersecting circles with the scoring rules: 1 point for 2 vaguely circular figures, 1 point for 2 clear circles, and 1 point for intersecting circles. Sensitivity, specificity, and FPR of the modified RUDAS were calculated.

Result: 32.3 % of illiterate controls failed to draw the circles. Overall, the modified RUDAS with a 22 cut-off had 77.0 % sensitivity (95% CI; 67.9, 84.2), 88.7 % specificity (95% CI; 82.5, 92.9), and 11.3 % FPR (95% CI; 7.1, 17.5). Among those who are illiterate (N = 123, 61 cases, 62 controls), the original RUDAS had 90.2% sensitivity (95% CI; 80.1, 95.4), 79.0% specificity (95% CI; 67.4, 87.3), 21.0% FPR (95% CI; 12.7, 32.6); Modified RUDAS had 83.6% sensitivity (95% CI; 72.4, 90.8), 83.9% specificity (95% CI; 72.8, 91.0), 16.1% FPR (95% CI; 9.0, 27.2).

Conclusion: The modified RUDAS is not as sensitive as the original RUDAS in screening for dementia among illiterate older people, but is more useful when higher specificity and lower FPR are preferred.

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Abstract – WCN 2013

No: 2474

Topic: 5 – Dementia

Report on cluster of three autopsy confirmed cases of sporadic Creutzfeldt–Jakob disease from Luxembourg including videos, imaging, genetics and neuropathology

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Background: Creutzfeldt Jakob disease (CJD) is a currently incurable, invariably fatal neurodegenerative disorder of misfolded proteins coined prions. The incidence of prion disease in humans is one in one million people per year with 85% of cases represented by sporadic CJD (sCJD), 15% by genetic prion diseases and less than 1% by acquired forms of CJD, including variant. Sporadic CJD can be further subdivided into six molecular classifications depending on the polymorphism at codon 129 (homozygous or heterozygous for methionine and/or valine on of the prion protein gene).

Objective: To report on a cluster of sCJD during a ten month interval in Luxembourg, population 517,000.

Patients and methods: We report on clinical, neuropsychological, laboratory, electroencephalogram, imaging, and neuropathological findings on three patients with rapidly progressive dementia (RPD) in a teaching hospital. Genetic analysis through blood testing was conducted at the German National Reference Center for Surveillance of Transmissible Spongiform Encephalopathies. Serial imaging was retrospectively reviewed by a blinded CJD expert.

Results: These cases represent a six-fold adjusted population based incidence of sCJD. Genetic analysis did not reveal any PRNP mutations, suggesting that all cases were sporadic, as none had any other risk exposures. Pathology ruled out variant CJD and confirmed likely sCJD. All patients had Type 1 prions. Two patients initially presented with motor symptoms were homozygous for methionine and one who presented with a neuro-cognitive disorder was heterozygous at codon 129 of PRNP.

Conclusion: This case series illustrates the workup of a cluster of RPD including international collaboration.

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Abstract – WCN 2013

No: 833

Topic: 5 – Dementia

Late-onset familial neuronal intranuclear inclusion disease presenting with dementia and unique MRI findings

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Background: Neuronal intranuclear inclusion disease (NIID) is a rare neurodegenerative disease associated with eosinophilic hyaline intranuclear inclusions in neuronal as well as visceral organ cells. As for its variable onset ages and symptoms, diagnosis cannot be made by clinical presentation alone and is solely dependent on pathological examination.

Objective: To describe a new NIID pedigree with characteristic radiological findings.

Methods: A 76 year-old woman had a 2-year history of progressive dementia, postural tremor in the upper extremities and retinitis pigmentosa (RP). MRI showed leukoencephalopathy with high intensity area along the arcuate fiber in diffusion-weighted image (DWI). Skin biopsy was performed for definitive diagnosis. Family members were screened for analogous clinical symptoms by Mini-Mental State Examination, neurological examinations, and color vision test. Seemingly affected individuals received MRI scan, nerve conduction study, and funduscopic examination for further investigation.

Results: Five affected members with at least 2 of the clinical features resembling those of the proband were identified from 3 consecutive generations. Proband's mother had dementia and tremor from her 70s. One sister had tremor, hyporeflexia and RP, while her 2 brothers had leukoencephalopathy, dementia, and tremor all starting from their 60s. One nephew had tremor, hyporeflexia, reduction in sensory nerve action potential and nyctalopia from late 50s.

Conclusion: In comparison with prior studies, late disease onset and DWI findings were the unique features of this pedigree. DWI could be a diagnostic tool for NIID patients with dementia, but needs accumulation of similar cases to prove its utility.

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Abstract – WCN 2013

No: 2515

Topic: 5 – Dementia

Associations of MRI white matter hyperintensities with cognitive functions, depression and quality of life

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Objective: White matter hyperintensities (WMH) are common findings on MRI and are associated with the risk of cerebrovascular diseases and cognitive decline. Associations of WMH with the most common cerebral complains (headaches, dizziness, tinnitus, memory decline) and such disorders as depression, lowering of quality of life and mild cognitive decline are not clear.

Methods: 284 subjects at age 40–59 were examined in cross-section population based study. Two groups were formed depending on finding or not WMH on MRI. Following surveys were performed – Luria's memory words test (LMWT), Munsterberg's correction test (MCT), SF-36, Hospital Anxiety and Depression Scale (HADS), and questionnaire for complains.

Results:

Group (1) – 124 women and 69 men mean age 48 ± 5 without WMH.
Group (2) – 55 women and 36 men mean age 53 ± 5 with WMH.

Group 2 was older ($p < 0.01$), sex distribution was similar. Memory decline by LMWT was more frequent in WMH group (52% vs 35%, $p = 0.01$). Operational integrity by MCT was lowered: 53% group 2 vs 46% group 1, $p > 0.05$. Cases with SF-36 results < 17 were in 78% of WMH group and 72% of the group without WMH, $p > 0.05$. HADS > 7 was in 31% vs 28% for anxiety and 22% vs 17% for depression, $p > 0.05$. Prevalence of dizziness and tinnitus was 52% vs 32%, $p = 0.002$; headaches – 76% vs 85%, $p > 0.05$.

Conclusion: WMH are associated with age, memory decline and dizziness/tinnitus complains. Headaches are less frequent in WMH patients, but this finding was statistically not significant.

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Abstract – WCN 2013**No: 2017****Topic: 5 – Dementia****High-dose 13.3 mg/24 h rivastigmine patch efficacy and safety in mild-to-moderate Alzheimer's disease with and without concomitant memantine use**

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Background: The OPTIMA study (Optimising Transdermal Exelon In Mild-to-moderate Alzheimer's disease; NCT00506415) demonstrated benefit of 13.3 mg/24 h rivastigmine patch on ADCS-IADL and ADAS-Cog in mild-to-moderate Alzheimer's disease. Here we investigated the effect of concomitant memantine use on efficacy and safety of 13.3 mg/24 h patch.

Methods: Details of OPTIMA are published (Cummings et al., 2012). In this retrospective analysis, patients randomized to 13.3 mg/24 h patch were subdivided to whether they received ≥ 1 dose of concomitant memantine during the double-blind phase. We compared change from baseline on ADCS-IADL and ADAS-Cog at weeks 24 and 48 using analysis of covariance with treatment, memantine usage, and treatment-by-memantine as factors. Safety evaluations included incidence of adverse events (AEs) and serious AEs (SAEs).

Results: 130 patients were included in this analysis (65 patients per group). The background characteristics including gender, race, age, BMI and MMSE were comparable between the 2 groups. Overall, 70% patients were female and 87% were ≥ 65 years.

Comparisons in the change from baseline on IADL and ADAS-Cog at weeks 24 and 48 showed no significant differences in memantine-treated patients and those not receiving memantine.

Incidence rates of AEs, AEs leading to discontinuation and SAEs in patients treated with the patch alone versus those receiving with patch and memantine were 66 vs 59%, 2 vs 6% and 9 vs 11%, respectively.

Conclusions: These data suggest that efficacy of high-dose rivastigmine patch remained unchanged regardless of concomitant memantine use, and demonstrate comparable safety profile of this patch in memantine-treated patients and those not receiving memantine.

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Abstract – WCN 2013**No: 2736****Topic: 5 – Dementia****Testamentary capacity and intention in patients with dementia**

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Background: There are many cases of contested wills due to a "lack of testamentary capacity". As the incidence of cognitive impairment of the elderly is increasing, the need of scientifically documented evaluations of their testamentary capacity is increasing too.

Objective: To explore the intention of patients with (mild or moderate) dementia in relation with their capacity to execute a will and how this intention can be measured/evaluated.

Material and methods: The literature relevant to the existing methods assessing the testamentary capacity of patients with dementia was reviewed. The scientific database searched was Medline and the keywords used were testamentary capacity, dementia, instruments, and intention.

Results: The testator should have the following features unaffected: memory, realistic perception of the value of his property, competence of the reality and intention of how and to whom he will dispose his properties.

Various models have been proposed for the assessment of the ability of patients with dementia to make their will. There are four common elements between the existing models:

- Expression of the patient's choices,
- Understanding of the information which is relevant to their choice,
- Evaluation of the significance of their choice, and
- Ability of rationalizing their decision.

Conclusion: Elderly people belong to a high-risk population in the area of competence evaluation. The challenge for the scientists is to develop a clinically applicable instrument for the quick and reliable assessment of the ability of people to make decisions about their own will, so as to preserve their autonomy.

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Abstract – WCN 2013**No: 2713****Topic: 5 – Dementia****Subjective memory complaints, depressive symptoms and cognition in patients attending a memory outpatient clinic**

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The goals of this study were to establish prevalence of subjective memory complaints (SMC) and depressive symptoms (DS) and their relation to cognitive functioning and cognitive status in an outpatient memory clinic cohort.

248 cognitively healthy controls and 581 consecutive patients with cognitive complaints who fulfilled the inclusion criteria were included in the study.

A statistically significant difference ($p < 0.001$) between control group and patient group regarding mean SMC was detected. 7.7% of controls reported considerable degree of SMC whereas 35.8% of patients reported considerable SMC. Additionally, a statistically significant difference ($p < 0.001$) between controls and patient group regarding Beck depression score was detected. 16.6% of controls showed clinical relevant degree of DS whereas 48.5% of patients showed DS. An analysis

of variance revealed a statistically significant difference across all four groups (control group, SCI group, naMCI group, aMCI group) ($p < 0.001$). Whereas 8% of controls reported considerable degree of SMC, 34% of the SCI group, 31% of the naMCI group and 54% of the aMCI group reported considerable SMC. A two factor analysis of variance with the factors cognitive status (controls, SCI group, naMCI group, aMCI group) and depressive status (depressed vs. not depressed) and SMC as a dependent variable revealed that both factors were significant ($p < 0.001$) whereas the interaction was not ($p = 0.820$).

A large proportion of patients seeking help in a memory outpatient clinic report considerable SMC, with increasing degree from cognitively healthy elderly to aMCI. Depressive status increases SMC consistently across groups with different cognitive status.

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Abstract – WCN 2013

No: 2753

Topic: 5 – Dementia

A critical role for molecular chaperones in Alzheimer's disease

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Background and objectives: Chaperones are one of the best examples of multifunctional proteins and their production against neurodegeneration may result from one or more of their activities in cells, perhaps in addition to their ability to inhibit fibril formation directly. Our objective was to explore if an excess of chaperone capacity in the cell can shift the equilibrium between amorphous and fibrillar aggregates and if the cells' proteolytic machinery can more efficiently turn over the toxic proteins. The area investigated may help explain chaperone suppression of neurotoxicity.

Material and methods: Expression pattern analysis of Hsp90/70 was determined by using Western Blot and immunohistochemistry on postmortem cortical tissues from Alzheimer's disease cases and aged matched controls.

Results: The results showed that AD samples contained significantly higher levels of the Hsp90/70 and this elevation was associated with the disease pathology. No cross reaction was observed between antibody used for Hsp90/70 (H9010/N27, respectively) and monoclonal antibody TG3. TG3 stains neuritic plaques and neurofibrillary tangles but does not react with tau from normal human biopsy tissue; therefore exhibiting its high degree of specificity for AD pathology.

Conclusion: Protein misfolding is believed to be the primary cause of Alzheimer's disease. Clearly, the effects of chaperones in multiple cellular pathways will have to be deciphered in order to understand which of these effects are primary and which are secondary in protection against neurodegeneration.

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Abstract – WCN 2013

No: 2341

Topic: 5 – Dementia

Mesugenin C mitigated LPS-induced neuroinflammation in BV-2 and NG108-15 cells through pMTOR-PI3K-Akt and ccl21 downregulation

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Background and objectives: Excessive microglial activation by inflammation leads to the progression of neurodegenerative diseases which results in neuronal cell death such as Alzheimer's disease. Mesugenin C (MC) was reported to be neuroprotective against H₂O₂-induced apoptosis in NG108-15 cells. Therefore, the anti-inflammatory effects of MC in LPS-stimulated inflammation in microglial BV2 and H₂O₂-treated NG108-15 cells were investigated.

Material and methods: The expression of NO, ROS, PGE₂, TNF- α , IL-1 β , IL-2, IL-6, IFN- γ , iNOS, COX-2, CCL21 and the molecular pathways involved were determined by flow cytometry chip beads array (CBA), Western blot and ICC. NG108-15 and BV-2 were co-cultured, stimulated with LPS and further evaluated. Signaling pathways were validated with inhibitors and siRNA CCL21 was used to assess CCL21 involvement in neuroinflammation.

Results: MC mitigated LPS-induced iNOS, NO, ROS and PGE₂ production leading to downregulation of TNF- α , IL-1 β , IL-2, IL-6, IFN- γ and COX-2 expression. MC upregulated the pMTOR-Rictor complex leading to the upregulation of PI3K-pAkt expression. Furthermore, MC prevented I κ B α degradation and thus, prevented p65 NF- κ B translocation in BV-2 and NG108-15 cells. Moreover, MC inhibited the CCL21 expression in both LPS-treated BV-2 and H₂O₂-treated NG108-15 cells resulting in the reduction of CCL21 chemotactic activity and mitigation of BV-2 activation. CCL21 siRNA transfection mitigated LPS-induced neuroinflammation, further proved that modulation of CCL21 by MC was indeed important.

Conclusion: MC protects NG108-15 by suppressing BV-2 activation through pMTOR-PI3K-Akt pathway and prevention of p65 NF- κ B translocation leading to downregulation of proinflammatory proteins and CCL21. This finding advocates therapeutic potential of MC for the treatment of neurodegenerative diseases.

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Abstract – WCN 2013

No: 2630

Topic: 5 – Dementia

Increased menin expression associated with neural apoptosis in the frontal cortex of SIV infected macaques

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Various types of neuronal damage have been reported in AIDS dementia complex (ADC). Human immunodeficiency virus (HIV) infected microglial/macrophage, not neuron directly in the brain. However, the damage of the neuron existed after the infection, in which the regulatory protein Tat of HIV-1 could be the critical factor. It has been recently reported that HIV-1 Tat transactivation requires menin, which hints that menin may be involved in the pathogenesis of ADC. However, the mechanism has not been elucidated completely. In this study, we report the up-regulated menin expression in the frontal cortex of SIV-infected macaques, especially in the neuronal nucleus by the double-labeling immunofluorescence, qRT-PCR and Western blot assay. The co-localization of menin and caspase3 is also observed in the frontal cortex of SIV-infected macaques. TGF- β , a cytokine associated with menin expression, is also detected increased in the frontal cortex of SIV-infected macaques. Furthermore, co-localization of SIV Tat and TGF- β appeared in increased TGF- β expressed vertebral neuron. In conclusion, these results indicate that

menin may be involved in the pathogenesis of ADC and the mechanism may be related to the pro-apoptosis of menin and the increased expression of TGF- β induced by SIV Tat. However, the precise mechanism needs to be further explored.

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Abstract – WCN 2013

No: 2831

Topic: 5 – Dementia

Early detection of abnormal prion protein in genetic human prion diseases now possible using real-time QUIC (RT-QUIC) assay

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Background: The definitive diagnosis of genetic prion diseases (gPrD) requires pathological confirmation. To date, diagnosis has relied upon the finding of the biomarkers in CSF, but many researchers have reported that these markers are not sufficiently elevated in gPrD, especially in Gerstmann–Sträussler–Scheinker syndrome (GSS). We recently developed a new in vitro amplification technology, designated “real-time quaking-induced conversion (RT-QUIC)”, to detect the abnormal form of prion protein in CSF from sporadic CJD patients.

Objective: In the present study, we aimed to investigate the presence of biomarkers and evaluate RT-QUIC assay in patients with gPrD, as the utility of RT-QUIC as a diagnostic tool in gPrD has yet to be determined.

Patients/method: 56 CSF samples were obtained from gPrD patients, including 20 cases of GSS with P102L mutation, 12 cases of fatal familial insomnia (FFI; D178N), and 24 cases of genetic CJD (gCJD), comprising 22 cases with E200K mutation and 2 with V203I mutation. We subjected all CSF samples to RT-QUIC assay, analyzed 14–3–3 protein and measured t-tau protein.

Results: The detection sensitivities of RT-QUIC were as follows: GSS (78%), FFI (100%), gCJD E200K (87%), and gCJD V203I (100%). On the other hand the detection sensitivities of biomarkers were considerably lower: GSS (11%), FFI (0%), gCJD E200K (73%), and gCJD V203I (67%). Thus, RT-QUIC had a much higher detection sensitivity compared with testing for biomarkers, especially in patients with GSS and FFI.

Conclusion/significance: RT-QUIC assay is more sensitive than testing for biomarkers in gPrD patients.

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Abstract – WCN 2013

No: 2945

Topic: 5 – Dementia

Blood-based biomarkers of Alzheimer's disease pathology and cognitive decline in non-demented elderly

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Background: There is an urgent need for Alzheimer's disease (AD) biomarkers that can detect the disease at the early pre-symptomatic stages. Case versus control study designs often ignore clinical heterogeneity in patients and neuropathology in controls resulting

in markers with doubtful clinical utility. Using well-established imaging phenotypes of AD pathology to derive biologically relevant biomarkers is an alternative approach. Neuropathological hallmarks of AD (B-amyloid plaques and neurofibrillary tangles) are commonly reported in brains of non-demented elderly individuals, suggesting these cases might represent preclinical AD and may be a good population in which to detect early AD biomarkers.

Objectives: To identify plasma biomarkers associated with AD endophenotypes in non-demented older individuals using two complementary longitudinal discovery-phase proteomic analyses.

Materials and methods: Two-dimensional gel electrophoresis coupled with mass spectrometry was performed on longitudinal plasma samples from non-demented older individuals exhibiting a range of C-PiB PET measures of amyloid load (n = 68). The relationship between protein concentrations and measures of brain atrophy, cognitive decline, and amyloid load were examined. A label-free LC-MS/MS approach targeted at low molecular weight proteins (<30kDa) was also performed on a subset of these subjects (n = 38) to further investigate this relationship.

Results: We have identified proteins associated with brain amyloid load, atrophy and cognitive scores, some of which are also capable of predicting future decline. Using longitudinal samples also allowed longitudinal changes in these proteins to be explored.

Conclusions: Our findings support previous reports that plasma biomarkers have the potential to be used as a screening tool for AD.

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Abstract – WCN 2013

No: 2997

Topic: 5 – Dementia

The effect of memantine in specific behavioural symptoms in ad outpatients. results of a Greek observational, open-label, 6-month study

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Objective: To assess the effectiveness of memantine treatment on specific behavioural symptoms of a Greek Alzheimer's Disease (AD) population.

Patients and methods: In this observational, open-label study, behavioural symptoms were evaluated by NPI Scale at baseline, 3 and 6 months.

Results: 1487 patients were included in the study. The NPI-items rated with the higher scores were “anxiety”, “depression/dysphoria” and “irritability/lability”, with scores (mean \pm SEM) of 3.0 (\pm 0.1), 2.8 (\pm 0.1) and 2.6 (\pm 0.1) points. In the “behaviourally disturbed population” (BDP), defined as patients having NPI Score > 0 in any of the symptoms: delusions, hallucinations and agitation/aggression, the symptoms with the higher scores were “anxiety” (3.4 \pm 0.1 points), “agitation/aggression” (3.3 \pm 0.1) and “irritability/lability” (3.2 \pm 0.1). All 12 NPI-items were significantly improved at 6 months (Friedman test, p < 0.001). The bigger absolute improvement was detected in “anxiety” and “depression/dysphoria”, by 1.6 points mean decrease in each item, while the higher percentage improvement was achieved in “agitation/aggression” and “irritability/lability” with 58% and 56% mean decrease over 6 months of treatment. In BDP, the higher improvements were observed, both in absolute and percentage values, in “agitation/aggression” and “irritability/lability” subscores (–2 and –1.9 points, 60.6% and 59.4%, respectively). 5.5% of patients prematurely discontinued treatment and only 0.3% of total population stopped due to adverse event (AE).

Conclusion: Memantine improved all 12 NPI behavioural items assessed in this observational study conducted in Greece in AD outpatients.

Relatively higher improvement was detected in “agitation/aggression” and “irritability/lability”. Memantine treatment was very well tolerated as only 0.3% of patients discontinued due to AE.

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Abstract — WCN 2013

No: 2992

Topic: 5 — Dementia

Discovery of peripheral biomarkers of alzheimer's disease pathology

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Background: Alzheimer's disease (AD) is the most common form of dementia in later life, affecting one in eight people by the age of 65 years. The development of minimally invasive peripheral markers that can better describe and predict the neuropathology of the disease, could greatly aid in overcoming the complexity of AD diagnosis.

Objectives: The aim of this study is to discover novel peripheral markers, that can predict Alzheimer's disease pathology.

Materials and methods: Peripheral biomarker discovery was undertaken in a cohort of extreme endophenotypes of pathological status, which were determined by the measurement of CSF Aβeta/Tau. Subjects were selected with very high CSF Aβeta/low Tau values (n = 25) or very low CSF Aβeta/high Tau values (n = 25). Isobaric tandem mass tagging (TMT) in combination with SDS-PAGE fractionation and LC-MS/mass spectrometry Orbitrap Velos Pro instrumentation were used to detect and quantify potential plasma biomarkers.

Results: A selection of 10 plasma proteins shows group-wise differences in their levels, according to CSF-measured pathology (Mann Whitney U Test, $P < 0.05$), and correlations of further 6 proteins with the degree of CSF-measured pathology are observed (Spearman's Rank Correlation $P < 0.05$). These include proteins identified in earlier studies as being associated with AD, as well as novel proteins not previously identified as being related to the disease and its pathology.

Conclusion: We have identified a number of plasma proteins as potential candidate markers of AD pathology in relation to CSF Aβeta/Tau. These markers include proteins related to the complement cascade and amyloidogenic processes.

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Abstract — WCN 2013

No: 2994

Topic: 5 — Dementia

The use of memantine in a Greek outpatient ad population. The effect of behavioural disturbances and other comorbidity

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Objective: To investigate the effectiveness of memantine treatment on Alzheimer's Disease (AD) patients' behaviour and cognition, in relation with common clinical parameters such as the presence of behavioural disturbances and concomitant diseases at baseline.

Patients and methods: This observational, open-label study was conducted in Greece, at an AD outpatient setting. Neuropsychiatric Inventory (NPI) and Mini-Mental State Examination (MMSE) were used for assessing behaviour and cognition. Patients were evaluated at baseline, 3 and 6 months and considered as “behaviourally disturbed population” (BDP) if at least one of the symptoms: delusions, hallucinations and agitation/aggression was present.

Results: 1487 patients were included in the study. Mean NPI score (\pm SEM) significantly improved from 24.5 (\pm 0.6) at baseline to 15.0 (\pm 0.4) at 3 months and 11.9 (\pm 0.4) at 6 months (Hotelling's test, $p < 0.001$). MMSE score accordingly improved from 17.1 (\pm 0.1) to 18.3 (\pm 0.1) and 18.7 (\pm 0.1) (Hotelling's test, $p < 0.001$). BDP patients (n = 987) improved significantly more than non-BDP population (n = 404) in NPI and MMSE (t -test, $p < 0.001$ for both scales). Patients were significantly more improved in both scales if they were behaviourally disturbed at baseline (t -test, $p < 0.001$) and if they were suffering from concomitant diseases (t -test, $p < 0.01$). Patients with concomitant diseases reported the significantly more adverse events (AEs, $p = 0.033$).

Conclusion: Memantine improved behaviour and cognition in Greek AD outpatients over a 6-month treatment. BDP and also patients with concomitant diseases showed greater improvement than non-BDP and patients without concomitant diseases respectively. AEs were few but relatively more in patients with concomitant diseases.

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Abstract — WCN 2013

No: 3000

Topic: 5 — Dementia

The effectiveness of memantine in a “behaviourally disturbed” AD population. Results from a Greek observational, open-label, 6-month study

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Objective: To evaluate the effectiveness of memantine treatment on behaviour and cognition of an Alzheimer's Disease (AD) “behaviourally disturbed” population.

Patients and methods: This observational, open-label study took place in Greece, in AD outpatients. Neuropsychiatric Inventory (NPI) and Mini-Mental State Examination (MMSE) were used for assessing behaviour and cognition. The “behaviourally disturbed population” (BDP), included participants having NPI score > 0 in any of the symptoms: delusions, hallucinations and agitation/aggression. Patients were evaluated at baseline, 3 and 6 months.

Results: 981 out of 1487 patients were classified as BDP. In this population, mean NPI score (\pm SEM) significantly improved from 31.1 (\pm 0.7) at baseline to 18.5 (\pm 0.5) at 3 months and 14.5 (\pm 0.5) at 6 months (Hotelling's test, $p < 0.001$). MMSE score also improved from 16.5 (\pm 0.2) to 17.9 (\pm 0.2) and 18.4 (\pm 0.2) (Hotelling's test, $p < 0.001$). Agitation/psychosis subscore (cluster of the 3 aforementioned NPI-items scores) significantly improved from 7.9 (\pm 0.2) to 4.6 (\pm 0.2) to 3.7 (\pm 0.2) at 3 and 6 months (Hotelling's test, $p < 0.001$). 5.3% of patients early discontinued treatment (only 0.2% due to adverse event—AE) and 4.6% of patients reported at least one AE.

Conclusion: Memantine improved agitation/psychosis NPI subscore and also total NPI and MMSE scores in Greek behaviourally disturbed outpatients over a 6-month treatment. Memantine was well tolerated as discontinuations and AEs reported were few.

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Abstract – WCN 2013**No: 2518****Topic: 5 – Dementia****Rapidly progressive dementia in the “Attikon” University General Hospital of Athens. A one-year experience**

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Introduction: Rapidly progressive dementia (RPD) is a rare neurological condition which progresses subacutely from days to months. A variety of possible causes of RPD have been reported, such as neurodegenerative, autoimmune, vascular, metabolic/toxic disorders, infections, malignancies, normal pressure hydrocephalus and psychiatric disorders.

Objectives: To describe the causes and the clinical presentation of RPD in the University General Hospital of Athens “Attikon” within a year.

Methods: From a total of 728 patients hospitalized in our department from January 2012 to February 2013, 21 patients presented with RPD. A detailed clinical and laboratory investigation was performed in all cases.

Results: The most common cause of RPD reported in our department was probable sporadic CJD (6 cases). Two patients were diagnosed with normal pressure hydrocephalus (NPH) and four patients developed RPD due to vascular etiology (2 patients with probable CADASIL, 1 patient with probable cerebral amyloid angiopathy and 1 patient with vascular dementia). The rest of the cases had various neurodegenerative disorders: probable AD (1 patient), DLB (1 patient), PSP (2 patients) and frontotemporal dementia (1 patient). One patient developed hydrocephalus and cognitive decline on the basis of *Mycobacterium tuberculosis* infection and 1 patient developed dementia probably due to chronic abuse of benzodiazepines.

Conclusion: RPD cases are not rare in a university general hospital. Besides CJD, neurodegenerative and secondary causes of RPD are frequent requiring a thorough investigation.

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Abstract – WCN 2013**No: 3010****Topic: 5 – Dementia****EEG microstates in Alzheimer's disease computed by continuous wavelet coherence**

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Background: Electrophysiological studies have demonstrated that the brain is active even in the absence of explicit input or output. Microstates represent subsequent coherent activation within the global functional brain networks. Continuous wavelet coherence is a measure of time-frequency changes in linear dependencies between EEG channels.

Objective: Areas of EEG microstates were evaluated in patients with Alzheimer's disease (AD). Differences in areas pertinent to each pair of electrodes and for frequency bands between patients with AD and healthy controls were calculated.

Patients and methods: EEG data were obtained for 110 patients with moderate dementia (MMSE score 10–19) and a control set of 110 age-matched, healthy subjects who had no memory or other cognitive impairment. Continuous wavelet coherence maps were computed for EEG records using the Matlab computational environment. The values higher than the threshold were considered as microstates corresponding to the coherent activation within the global functional brain networks. Areas of time-frequency plots for the delta, theta, alpha, beta and gamma bands were compared for both groups.

Results: Statistically significant differences were observed, particularly in the alpha, beta and gamma frequency bands of the frontal and temporal regions. The accuracy of distinction between healthy individuals and AD patients for the Fp2-T5, F8-T5, F7-O2, Fp2-O2 pairs of electrodes in the alpha band was more than 0.8.

Conclusion: These results can be interpreted as a decrease in the value of the parameter reflecting the ability of the system self-organization and thus suboptimal information processing clinically manifested as cognitive deficit.

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Abstract – WCN 2013**No: 2898****Topic: 5 – Dementia****Reproducibility of [18F]flutemetamol pet amyloid image interpretation**

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Objective: The reproducibility of [18F]flutemetamol PET amyloid image interpretation measured by inter-reader agreement (IRA) and intra-reader reproducibility (IRR), and quantitative test-retest variability was assessed, using GE's clinical trials data.

Methods: Blinded visual interpretations of color PET images from one Phase II study (Study 1) and two Phase III studies (Studies 2 and 3) were analyzed. In each study, five readers were trained to read [18F]flutemetamol scans via in-person training (Studies 1 and 2) or an electronic training program (Study 3). Readers then interpreted scans from healthy volunteers (HV), patients with Alzheimer's disease (AD), and amnesic mild cognitive impairment (aMCI) subjects as *normal* or *abnormal* for [18F]flutemetamol uptake. Included in each image set (to assess IRR) were randomly selected and inserted duplicates of ~10% of the images. In Study 1 only, intra-subject test-retest variability was calculated for five AD subjects who underwent two [18F]flutemetamol scans as the percentage difference between regional SUVR for a set of cortical regions.

Results:

Study 1: Across all groups, Fleiss' kappa for IRA (n = 67: 22 AD, 20 aMCI, and 25 HV) was 0.96 and IRR was 97.5%. Test-retest variability of regional SUVRs was 1% to 4%.

Study 2: Fleiss' kappa for IRA (232 aMCI patients) was 0.76 and IRR ranged from 86% to 100%.

Study 3: Fleiss' Kappa for IRA (80 aMCI patients) was 0.89, and IRR was 100%.

Conclusions: Across the three studies, blinded visual interpretations of [18F]flutemetamol PET amyloid images had high IRA and IRR. Regional SUVR determinations had low test-retest variability.

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Abstract – WCN 2013**No: 2940****Topic: 5 – Dementia****Rationale, design and preliminary baseline data of the ADEX study: The effect of physical exercise in Alzheimer's disease**

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Background: Exercise is hypothesized to improve cognition, physical performance, functional ability and quality of life, but evidence is scarce. Previous studies were of short duration, often underpowered and involving home based light exercise programs in patients with undefined dementia.

Objective: The aim of the ADEX trial is to establish whether aerobic exercise is effective in improving cognition as well as in reducing the prevalence of psychiatric symptoms among patients with Alzheimer's disease (AD). We present design and rationale of the study and preliminary baseline data from the participants included so far.

Patients and methods: The ADEX study is a multicentre, single-blind, randomized trial with two arms: an intervention group attending 16 weeks of continuously supervised moderate to high intensity aerobic exercise and a control group receiving usual care. Based on power calculations 192 patients with mild to moderate AD are to be recruited.

Results: A total of 121 participants (54 women and 67 men) have been randomized at the present time. Baseline characteristics are age 69.9 ± 7.4 years (50–90) and MMSE at baseline: 24.0 ± 3.7 (15–30). Drop-out rate is 20% and number of serious adverse events are 5, but only one was judged possible related to the study.

Conclusions: To our knowledge this is the first large scale controlled study to investigate the effects of supervised moderate-high intensity aerobic exercise. Complex interventions of this kind present unique challenges to study design and drop-out rates are known to be around 20–25%. Results are expected available in 2014.

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Abstract – WCN 2013

No: 2901

Topic: 5 – Dementia

The diagnostic value of Alzheimer's disease-related individual structural volume measurements using IBASPM

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Background: Despite the importance of hippocampal volume as a biomarker for AD and the availability of automatic measurement, the optimal cut-off of hippocampal volume using automatic segmentation is still unknown.

Objective: We validated the use of medial temporal volume measurements using the Individual Brain Atlases using Statistical Parametric Mapping (IBASPM), an automated volumetric method, for screening for Alzheimer's disease (AD). We also determined the age-specific, optimal cut-off values for hippocampal volumes.

Patients and methods: The study included T1-weighted magnetic resonance images (MRI) of 94 patients with AD and 51 normal controls. To determine age-specific cut-off values, we grouped subjects according to age. The hippocampal volumes of each group were measured using individual atlas-based volumetry with IBASPM.

Results: The normalized value of left hippocampal volume in patients with AD was significantly lower than that of the normal controls in all age groups. The areas under the curve (AUC) values of

the left hippocampus were also the largest in all age groups, indicating that AUC is the most useful factor of AD screening.

Conclusion: In conclusion, in clinical practice, measurement of left hippocampal volume using automatic measurement (IBASPM) assists in screening for AD, if used with the age specific cut-off value calculated in the present study. Left hippocampal atrophy is the best indicator in all age groups. Automated methods are faster and more feasible in clinical settings than manual segmentation, and provide a useful biomarker for AD.

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Abstract – WCN 2013

No: 2932

Topic: 5 – Dementia

Morphological alterations of purkinje cells of the flocculus of the cerebellum in early cases of Alzheimer's disease

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Introduction: Neuritic plaques, synaptic alterations and neurofibrillary tangles are the morphological hallmarks of Alzheimer's disease (AD), seen mostly in the hippocampus, the cortex of the cerebral hemispheres and the cerebellum at the advanced stages of the disease.

Material and methods: We studied the flocculus of the cerebellum of twenty cases at the early stages of Alzheimer's disease, applying rapid Golgi silver impregnation technique and electron microscopy. Control normal brains were similarly studied, in an attempt for correlative analysis of the findings.

Results: The morphological analysis of the flocculus of the cerebellum in AD, revealed no neuritic plaques and tau pathology. However a marked morphological alteration of Purkinje cells was seen, concerning mostly their dendritic arborization characterized mostly by a considerable shortage of dendritic branches, marked loss of dendritic spines, abnormal spines and decrease of spine density. In addition, numerous synaptic alterations between the parallel fibres, the climbing fibres and the Purkinje cell dendritic were revealed in electron microscopy. The number of the granule and Golgi cells was not substantially decreased, in comparison with normal controls. Synaptic pathology concerning mitochondria, synaptic vesicles and thickness of the postsynaptic membrane was frequently seen in the molecular layer of the flocculus in Alzheimer brains.

Conclusion: Extensive synaptic pathology occurs in the flocculus of the cerebellum at the early stages of AD, suggesting that the vestibule-floccular system is substantially early involved in the continuous process of dendritic and synaptic degeneration in Alzheimer's disease.

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Abstract – WCN 2013

No: 2938

Topic: 5 – Dementia

Default mode network in normal pressure hydrocephalus

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Background: Resting-state functional MRI (rs-fMRI) is a recent-developed neuroimaging method to examine the functional

connectivity of brain network. The default mode network (DMN) is one of prominent networks, which include the hippocampus, posterior cingulate (PCC), medial prefrontal, lateral temporal, and parietal cortices. Many studies found decreased DMN connectivity in Alzheimer's disease (AD) and suggested distinct involvement of DMN in different types of dementia.

Objective: We aimed to differentiate normal pressure hydrocephalus (NPH) from AD on the basis of neural connectivity with rs-fMRI, because these two diseases are sometimes difficult to discriminate only on the morphological examination.

Patients and method: Subjects diagnosed as having probable NPH (n = 9; mean age 77.4 ± 4.1 years, range 69–80 years), AD (n = 27; mean age 76.5 ± 5.0 years, range 66–86 years), and 57 healthy controls (HC, n = 57; mean age 67.3 ± 5.2 years, range 61–79 years, MMSE ≥ 27) underwent rs-fMRI. We used PCC as a seed region to determine the functional connectivity to other brain regions in DMN.

Results: The functional connectivity between PCC and other brain regions in DMN was decreased in both NPH and AD groups compared to HC group (p < 0.001), and there was no difference in the connectivity between NPH and AD groups.

Conclusion: Our findings indicate that the functional connectivity in DMN decreased in NPH as well as AD. However, it is difficult to differentiate NPH from AD on the basis of functional connectivity with rs-fMRI using seed-based analysis.

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Abstract – WCN 2013

No: 2834

Topic: 5 – Dementia

Cortical beta-amyloid and microstructural properties of the corpus callosum in patients with mild to moderate ad

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Background: Several studies have reported microstructural changes of the corpus callosum (CC) in AD. The pathological basis of these changes remains uncertain.

Objective: We investigated whether fractional anisotropy (FA) as a marker of microstructural properties of the CC is associated with cortical beta-amyloid measured by amyloid imaging.

Methods: Patients with mild to moderate Alzheimer's dementia (according to NINCDS-ADRDA criteria and ICD-10 criteria), were recruited.

Patients underwent 11C-PiB-PET for assessment of cortical beta-amyloid and MRI (Siemens Magnetom Trio 3T) including DWI for quantification of white matter microstructure. DWI data were fitted to the tensor model, and FA maps were created. Masks were manually delineated on the midsagittal slice of the FA maps, encompassing the anterior 1/6th of the CC (projecting to prefrontal cortex) and the posterior 1/4th (projecting to parietal, temporal, occipital cortices). For quantification of beta-amyloid standard uptake value ratios (SUVR) from prefrontal cortex, and parietal, occipital and temporal cortices were used in the analysis.

Results: Thirty-two patients (age, years: 69.2 (± 7.3); Gender (f/m): 14/18; MMSE: 24.8 (3.5)) were included in the study. Mean FA across subjects for anterior CC was 0.74 (± 0.038) and for posterior CC 0.71 (± 0.068). No significant associations between FA in anterior

CC and prefrontal 11C-PiB uptake or between FA in posterior CC and parietal-temporal-occipital cortex 11C-PiB uptake was found.

Conclusion: Cortical deposition of beta-amyloid does not seem to be the underlying mechanism of microstructural changes in the CC in AD patients. However, whether beta-amyloid may affect the CC through cortical atrophy remains undecided.

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Abstract – WCN 2013

No: 2836

Topic: 5 – Dementia

Being physically active is associated with improved executive function and processing speed but not memory: The LADIS study

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Background: Physical activity (PA) reduces the risk of cognitive decline, but may affect cognitive domains differently.

Objective: We examined whether PA may differentially modify processing speed, executive function and memory in a population of dementia-free elderly subjects with age-related white matter changes (ARWMC).

Patients and methods: Data from the Leukoaraiosis And DISability (LADIS) study, a multicenter, European prospective cohort study aimed at examining the role of ARWMC in transition to disability, was used. Subjects in the LADIS study were clinically assessed yearly for 3 years including MRI at baseline and 3-year follow-up. PA was assessed at baseline. Cognitive composite scores at baseline and 3-year assessment were used.

Results: Two-hundred-eighty-two subjects (age, y (mean (SD)): 73.1 (± 5.1); gender (f/m): 164/118); MMSE (mean (SD)): 28.3 (± 1.7) who had not progressed to dementia or MCI, were included. A multiple variable linear regression analysis with baseline MMSE, education, gender, age, stroke, diabetes and ARWMC rating as covariates revealed that PA was associated with better scores at baseline and 3-year follow-up for executive function (baseline: β:0.39, 95% CI: 0.13–0.90, p = 0.008; follow-up: β:0.24, 95% CI: 0.10–0.38, p = 0.001) and processing speed (baseline: β:0.48, 95% CI: 0.14–0.89, p = 0.005; follow-up: β: 0.15, 95% CI: 0.02–0.29, p = 0.02) but not memory. When including baseline cognitive score as a covariate in the analysis for 3-year follow-up scores, executive function remained significant (p = 0.04).

Conclusion: PA is associated with preserved executive function and processing speed but not memory in elderly dementia-free subjects with ARWMC.

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Abstract — WCN 2013**No: 3073****Topic: 5 — Dementia****Complete sequencing of the 7 mtDNA genes encoding for complex I subunits in frontotemporal lobar degeneration**

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Background: Mitochondria dysfunction and oxidative damage were reported in aging-related neurodegenerative diseases. A possible mechanism involves mitochondrial DNA (mtDNA) alterations impairing mitochondrial respiratory chain function. Contrary to other neurological disorders, frontotemporal lobar degeneration (FTLD) pathophysiology is still poorly understood, and etiology is often unknown. Recently, two mtDNA alterations were reported in a FTLD patient and an association with mtDNA haplogroup was proposed.

Objective: Sequencing the 7 *MT-ND* genes in FTLD patients.

Patients and methods: Total DNA was extracted from peripheral blood of 32 patients with probable FTLD recruited at Neurology Unit (CHUC). Analysis of 7 *MT-ND* genes sequences was performed by automatic sequencing and variants were submitted to in silico study. Statistical analysis was performed using Graph-Pad Prism 5.0.

Results: We found 80 different alterations in 30 patients (93.8%), including 68 polymorphisms (85.0%), 11 sequence variations previously associated to other diseases (13.7%) and 1 unpublished variant (1.3%). An analysis was conducted by dividing these patients in two groups: 1–5 sequence variants (15) and 6–13 alterations (15) per patient. There were no statistically significant differences when comparing both groups, according to age, gender, age of onset, disease duration, and CDR or MMSE results.

Conclusion: To our knowledge, this is the first report of complete sequence of the 7 *MT-ND* genes in FTLD patients. The high number of mtDNA alterations identified suggests a possible role of these variants in FTLD. However, further studies are needed to determine whether these variants are part of the etiology or an epiphenomenon.

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Abstract — WCN 2013**No: 3042****Topic: 5 — Dementia****Mutual information for diagnosis of AD**

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Background: Alzheimer's disease (AD) is associated with cognitive deficits due to neuronal loss, reduced brain neuronal connectivity and modifications to the functional organisation of the brain. Reduced connectivity might manifest as changes in the linear and non-linear synchronisation of collaborating parts of the brain.

Objective: Non-linear and linear measures of synchronicity in Alzheimer's disease (AD) were compared with those of healthy age-matched control subjects.

Patients and methods: Wavelet coherence (WC) was used to estimate linear synchronisation between electroencephalographic

(EEG) channels. Mutual information (MI) was applied to absolute values of complex wavelet coefficients in wavelet scales to estimate non-linear synchronisation. Synchronisation rates for a group of 110 patients with moderate Alzheimer's disease (MMSE 10–19) and 110 healthy control subjects were compared. Both parameters were estimated using the MATLAB computational environment.

Results: The most significant differences for an increase in mutual information were identified on the second and the third scale in the parietoccipital area, while for wavelet coherence the most significant difference was found between the frontal and parietal electrodes.

Conclusion: This newly proposed method utilises mutual information of complex wavelet coefficient absolute values in wavelet scales and demonstrates larger discriminatory values in Alzheimer's disease compared to wavelet coherence.

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Abstract — WCN 2013**No: 3011****Topic: 5 — Dementia****Comparison of complexity, entropy and complex noise parameters in EEG for AD diagnosis**

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Background: A non-linear analysis of EEG time series offers an alternative approach to understanding changes in Alzheimer's disease. Parameters used in this study are essentially a measure of the unpredictability, complexity or 'randomness' of the neural signal.

Objective: Complexity of EEG signal using fractal dimension, entropy and complex noise characteristics was evaluated in healthy population during aging and for a set of patients with Alzheimer's disease (AD). The threshold value of EEG complexity biomarker was estimated to distinguish patients with AD from healthy individuals.

Patients and methods: EEG data were obtained for 110 patients with moderate dementia (MMSE scores 10–19) and for a control set of 110 age-matched, healthy subjects who had no memory or other cognitive impairment. With the support of the MATLAB computational environment, EEG complexity was computed for 19 electrodes using Katz's and Sevcik's fractal dimension algorithms, permutation entropy and complex noise characteristics.

Results: AD patients and healthy individuals differed in EEG signal complexity. Using the threshold for separation of patients, we estimated the accuracy of 82% (Sevcik's FD), 72% (Katz's FD), 85% (color noise) and 70% (entropy).

Conclusion: These results suggest that characteristics based on the complex noise and Sevcik's fractal dimension provide the best discriminatory value for the non-linear measures of EEG complexity.

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Abstract — WCN 2013**No: 3064****Topic: 5 — Dementia****Cognitive deficit in Alzheimer's disease and during aging and self-organized criticality**

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Background: The hypothesis that the electrical activity of neural networks in the brain oscillate around critical points is very important, as many simulations show that the optimal efficiency of information processing is achieved at the critical point.

Objective: Complex noise with the power spectra dependence of the PSD = $1/f^\alpha$ frequency was evaluated in an aging healthy population and in patients with Alzheimer's disease (AD). Correlation between the alpha value and age and significance of difference between patients with AD and healthy controls were estimated.

Patients and methods: EEG data on 3845 truck drivers, 108 patients with moderate dementia (MMSE scores 10–19) and a control set consisting of 108 age-matched, healthy subjects who had no memory or other cognitive impairments were obtained. Power spectral densities (PSDs) of the EEG frequency components were estimated using the Welch method on the MATLAB computational environment. The slope α of the first degree polynomial that fitted the PSD (μV^2) data to frequency (Hz) in the log–log scale was estimated in the least squares sense.

Results: Aging correlates with a decrease of alpha towards values characteristic for the white noise in the most electrodes. The slope of the near linear decrease of the power spectra in the log–log scale shows statistically significant differences in the frontal areas of AD patients.

Conclusion: These results can be interpreted as a decrease in the value of the parameter reflecting the ability of the system's self-organization and thus suboptimal information processing clinically manifested as a cognitive deficit.

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Abstract – WCN 2013

No: 3083

Topic: 5 – Dementia

Progression of cognitive impairment in parkinsonian patients

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Parkinsonisms at onset can be associated to cognitive impairment. To describe the progression of cognitive deficits in patients with parkinsonism at onset.

We consecutively selected patients with parkinsonism and disease duration up to three years to take part in the Bologna-motor and non-motor prospective study on parkinsonisms at onset (Bo-ProPark).

All patients underwent, at baseline (T0) and after sixteen months (T1), neuropsychological assessment (Brief Mental Deterioration Battery, Stroop Test, Semantic Fluency Task, Beck Depression Inventory). Progression of cognitive impairment (CI) was defined as a drop of 1.5 SD in the corrected score for age sex and education. For all tests, except Barrage and Stroop, a positive score indicates CI progression.

35 patients were recruited. 27 patients were diagnosed as Parkinson Disease (PD), 2 as Progressive Supranuclear Palsy (PSP), 1 as Multiple System Atrophy (MSA), 1 as Cortico Basal Degeneration (CBD) and 4 as Parkinsonian Syndromes not otherwise specified (PS). At T0 13 patients (8 PD, 1 MSA, 1 CBD, 2 PSP and 1 PS) presented CI. The degree of CI was stable at T1. Global CI was detected in 2 PSP and 1 PD patients; executive function impairment was found in 5 PD, 1 DCB, 1 MSA and 1 PS, and memory impairment in 2 PD patients. Progression of cognitive deficits at T1 was observed in 5 PD patients with only executive functions.

CI can be observed in patients with parkinsonism at disease onset, independent from the clinical diagnosis. Progression of CI should be further evaluated as predictors of the diagnosis and prognosis.

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Abstract – WCN 2013

No: 3124

Topic: 5 – Dementia

A case of dementia secondary to subacute leukoencephalopathy by carbon monoxide

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Introduction: Delayed hypoxic–ischemic leukoencephalopathy described in 1976 by Ginsberg is a brain white matter demyelination phenomenon that occurred days or weeks after a hypoxic–ischemic injury followed by a complete recovery of the episode. Recovery is frequently observed in 75% of cases. We present the case of a patient exposed to carbon monoxide acutely, without impairment of consciousness that develops a subacute dementia syndrome.

Patients: A male patient of 52 years presented with a history of alcohol consumption, and an exposure to carbon monoxide for about 14 h. The patient was evaluated in the emergency room, with 24 h of observation and was discharged asymptomatic. One month later, the patient has alterations in daily living, he can't make paperwork, he can't work, and he gets lost on public roads. The patient was evaluated by neurology and highlights executive dysfunction without focal signs and without meningeal signs. The Addenbrooke's Cognitive Examination (ACE) was 60/100.

Material and methods: General exams are normal, HIV was negative, immunological tests were normal, cerebrospinal fluid thyroid tests were normal, and there was no asymmetry electroencephalogram without paroxysmal activity. Brain MRI shows typical leukoencephalopathy.

Results: The patient at 3 months shows a slight recovery of cognitive ability, still requires assistance to perform daily living and he cannot work. The ACE was 70/100.

Conclusion: A rare cause of dementia is subacute carbon monoxide poisoning, especially in this case, where exposure to carbon monoxide is brief and without impairment of consciousness of the patient.

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Abstract – WCN 2013

No: 3131

Topic: 5 – Dementia

Synaptic alterations in the claustrum in Alzheimer's disease: A Golgi and electron microscope study

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Alzheimer's disease is a heterogeneous neurodegenerative disorder of presenium and senium, characterized by a progressive decline of mental faculties, loss of professional skills, impairment of behavior, social performance and communication and various neurological manifestations. Since basal ganglia play an important role in cognitive functions, motor control and behavior, we attempted to study the pathological alterations of the claustrum whose enigmatic function may be related to cognition. We attempted to figure out the synaptic alterations in early cases of Alzheimer's disease. The morphological analysis is based on examination of twenty brains. Samples from the three parts of the claustrum, based on dorsoventral and anteroposterior axons, were processed for the Golgi technique and electron microscopy. The morphological and morphometric analysis revealed substantial neuronal loss and marked synaptic alterations mostly in the anterior

part of the claustrum. The dendritic arborization was decreased. Dendritic spines of the elongated and fusiform neurons were dramatically decreased. Mitochondrial alterations and fragmentation of Golgi apparatus were seen in a considerable number of neurons. Synaptic alterations were seen in all the parts of the claustrum, although the synapses were better preserved in the ventroposterior part than in the anterior dorsal one. Neurofibrillary tangles were frequently seen. Reactive astrocytosis was more prominent in the anterior part than in the middle and the posterior ones. The neuronal loss and the synaptic alterations in the claustrum may underline a broad cholinergic deficit in Alzheimer's disease and a substantial decline of cortico-subcortical cooperative activity in the early stages of Alzheimer's disease.

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Abstract — WCN 2013

No: 3173

Topic: 5 — Dementia

Telomere length distribution of circulating leukocytes in patients with Alzheimer's disease

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Background: A telomere is a structure containing repetitive DNA sequence located at the termini of human chromosomes. Telomere attrition has been observed in peripheral blood nuclear cells with aging. Older people have shorter telomeres in their somatic cells than young people. We have analyzed telomere length distribution of normal population and have shown that older people have less long telomeres and more short telomeres than young people. Telomere shortening is also accelerated by disease conditions such as Alzheimer's disease (AD). **Objective:** We analyzed telomere length distribution of AD patients to know if telomere attrition with aging is similar to that with AD in terms of telomere length distribution.

Patients and methods/material and methods: Female patients with AD visiting the outpatient clinic of Kyushu University Beppu Hospital, from May 2008 through March 2011 were enrolled.

Results: Thirty female AD patients were found to have normal mean telomere lengths (control; 6.5 ± 1.0 kb, AD; 6.1 ± 0.8 kb, $p = 0.13$), a decreased proportional amount of the longest telomeres (>9.4 kb) (control; $30 \pm 8\%$, AD; $24 \pm 8\%$, $p = 0.01$), increased middle-sized telomeres (control; $52 \pm 3\%$, AD $56 \pm 6\%$, $p = 0.02$), and an unchanged amount of the shortest telomeres (<4.4 kb) (control; $18 \pm 8\%$, AD; $20 \pm 9\%$, $p = 0.37$) in their peripheral leukocytes.

Conclusion: Though mean TRF of AD patients was not significantly different from that of controls, the longest telomeres (>9.4 kb) were decreased and the shortest telomeres (<4.4 kb) did not increase, which was different from the aging-associated change of telomere length distribution. AD-associated telomere attrition was dominantly observed in long telomeres.

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Abstract — WCN 2013

No: 3185

Topic: 5 — Dementia

Neuropsychiatric symptoms in mild Alzheimer's disease and mild cognitive impairment

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Background: Neuropsychiatric symptoms affect more than a third of dementia patients, and are associated with institutionalization and caregiver stress or care burden in patients with Alzheimer's disease (AD). They are also common in mild cognitive impairment (MCI).

The early identification of neuropsychiatric symptoms in MCI could improve further the prediction of the complex clinical course of the dementia illness, possibly helping improve treatment approaches, and ultimately the prognosis.

Methods: We included 101 outpatients of a memory clinic in Maryknoll Hospital (48 AD; 53 amnesic-MCI-multidomain) and 25 controls. Categorical disorders of depression and apathy were diagnosed with structured interviews. Symptoms were evaluated with the neuropsychiatric inventory (NPI). The odds ratios (OR) of patients having neuropsychiatric symptoms compared to controls were calculated with logistic regression, adjusted for sociodemographic and clinical variables.

Results: A large proportion of AD (62.5%), amnesic-MCI-multidomain (39.6%) patients had a depressed disorder. Apathy disorder was common in AD (68.7%) but less frequent in amnesic MCI-multidomain (9.4%). AD patients were five times more likely to have depression disorders (OR = 5.1, CI = 1.1–23.1) compared to amnesic-MCI-multidomain. After apathy and depression, the most prevalent neuropsychiatric symptoms in AD and MCI were anxiety, agitation, irritability, night-time behaviors, and appetite disturbances.

Conclusions: Clinicians should consider the relevance of neuropsychiatric symptoms in patients with cognitive disturbances, and incorporate a thorough psychiatric examination in the evaluation of patients.

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Abstract — WCN 2013

No: 3209

Topic: 5 — Dementia

Semantic verbal fluency categories in Brazilian Portuguese

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Background: Semantic verbal fluency is widely used as part of the cognitive examination. By far the “animals” category is more often used, and the performance for the other categories in the original list of this test is not clear, and some of them could be more useful in specific situations.

Objective: To investigate the performance of healthy Brazilian young adults in all 56 categories of the original list of semantic fluency.

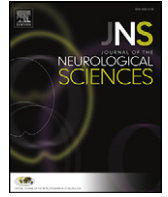
Patients and methods: 600 subjects (300 males and 300 females) were included (300 for São Paulo metropolitan area and 300 were evenly distributed in the other 5 areas).

Material and methods: All subjects are university students, both genders, aged 18 to 35 years, free of neurological and psychiatric diseases were submitted to all 56 semantic categories of the original verbal fluency test. They were divided by age in two groups (18–25 and 26–35) and four groups according to geographic area (São Paulo metropolitan area/north–northeast/central–central–south/south Brazil).

Results: There was no difference in performance between age groups, genders and among geographical areas. The perception of semantic category difficulty correlated with performance (low, intermediary and high difficulty). Statistical analysis was done using Mann–Whitney for comparison of age ranges for both genders and for genders for each age range. The procedures were approved by UNIFESP Institutional Review Board and all subjects signed an informed consent.

Conclusions: Semantic categories have varying levels of difficulty even for high educated young people. According to necessity categories could be selected for specific purposes.

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Topic: 6 - MS & Demyelinating Diseases

Abstract – WCN 2013cuo

No: 3213

Topic: 6 – MS & Demyelinating Diseases

Increased PPAR-gamma levels in the CSF of patients with multiple sclerosis

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Peroxisome proliferator-activated receptor (PPAR)-gamma, a ligand-activated transcriptional factor involved in the regulation of glucose and lipid metabolism, has gained interest as a possible therapeutic target in multiple sclerosis (MS) owing to its potent immunoregulatory effects and the therapeutic efficacy of its ligands in experimental autoimmune encephalitis (EAE). Elevated expression of PPAR-gamma has been detected in the spinal cord of EAE mice and in an in vitro model of demyelination; however, no reports have yet been available on the PPAR-gamma status in the central nervous system (CNS) of MS patients. Aiming to identify a possible alteration, the present study assessed the concentration of PPAR-gamma protein in the cerebrospinal fluid (CSF) of MS patients via ELISA method. We report a robust increase in the CSF levels of PPAR-gamma in MS patients ($n = 35$) compared to non-inflammatory controls ($n = 22$). This increase was independent of blood-CSF barrier dysfunction, but correlated with CSF cell count and IgG index, associating the observed increase with CNS inflammation. Controlling for potential confounders, CSF concentration of PPAR-gamma further showed a moderate but statistically significant association with clinical severity (EDSS). Corroborating with previous experimental findings, these results may contribute to our understanding about the role of PPAR-gamma in MS, and may implicate this protein as a potential CSF biomarker of clinical severity in MS.

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Abstract – WCN 2013

No: 942

Topic: 6 – MS & Demyelinating Diseases

Impact of tobacco smoking on mortality and life expectancy in multiple sclerosis patients

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0022-510X/\$ – see front matter.

Objective: To investigate life-style factors associated with increased mortality in a cohort of multiple sclerosis patients.

Method: Clinical and demographic characteristics of 968 individuals were obtained from a large population-based cohort of MS patients. Records were linked to the NHS death data to obtain data about death in Dec 2012 (index date). Patients were followed from the onset age to the death age or index date whichever occurred first. The impacts of sex, disease clinical course at the time of onset, 1 or more year exposure to treatment, onset age and smoking status (ever vs. never) were investigated using Cox hazard regression model.

Results: Of 923 patients with clinically definite MS and full data for analysis, 80 (47 male and 33 female) were deceased at the index date. The mean survival age was 77.3 years (95% CI: 75 to 79.5). Compared with ever-smokers, never-smokers lived almost 7 years less (81 vs. 74). Smokers were at higher risk for death, with a hazard ratio of 2.24 (95% CI: 1.30 to 3.85; $P = 0.004$) relative to never-smokers. Compared to the UK general population, the standardised mortality rates were 1.71 (95% CI: 1.37 to 2.13) for all patients, 2.64 (95% CI: 1.97 to 3.53) for ever-smoked patients and 1.14 (95% CI: 0.74 to 1.75) for never-smokers.

Conclusion: The higher mortality rates reported previously in MS population could partially have been described by patients' smoking habits. This survival analysis has determined that smoking is a significant yet preventable risk factor for death in patients with MS.

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Abstract – WCN 2013

No: 167

Topic: 6 – MS & Demyelinating Diseases

Endocrine disorders of patients with multiple sclerosis

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Introduction: While multiple sclerosis (MS) affects the nervous system, extra-neural pathology is also observed.

Objective detection of endocrine disorders and determination of its dependence on the degree of the disease severity.

Methods and materials: 85 patients were examined with relapsing-remitting type of MS clinical course. Severity of patient's state was rated on a scale: EDSS, FS Kurtzke. Mild severity – 2.33 ± 0.09 points (EDSS), moderate severity – 4.1 ± 1.08 , heavy severity – 6.3 ± 1.2 .

Results: Endocrine disorders were diagnosed of 25 (23.8%) patients. 17 (68%) patients developed mild MS and 8 patients (32%) – moderate MS severity. Cortisol level was 1.4 times more than normal of 10 (40%) patients. The correlation between stem disorders and cortisol levels ($r = -0.29$) was defined. 8 (32%) patients developed autoimmune thyroiditis in hypertrophic form. Thyrotropin hormone concentration

was 8 times higher than normal, the level of free thyroxine was 2 times less than normal. The correlation between hormone levels and severity of movement impairment ($r = -0.21$), cerebellar symptoms ($r = 0.55$), sensory disturbances ($r = -0.25$), pelvic disorders ($r = 0.74$) was established. 7 (28%) patients showed prolactin levels for 38.2% higher than normal. The correlation is established between hormone levels and the severity of movement impairment ($r = -0.61$), cerebellar symptoms ($r = 0.44$), stem disorders ($r = -0.35$), sensory disturbances ($r = -0.35$).

Conclusion: Endocrine disorders prevailed in mild severity of disease, were characterized by the presence hypercortisolemia, hyperprolactinemia, hypothyroxinemia, and an increased content of thyroid stimulating hormone. The correlation between level of thyroid stimulating hormone and EDSS score ($r = 0.43$) is established.

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Abstract – WCN 2013

No: 3080

Topic: 6 – MS & Demyelinating Diseases

TLR 3 ligand poly I:C does not induce encephalitogenic T cells nor essentially alter cytokine production in SJL EAE

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Background: Toll-like receptor (TLR) ligands have the ability to alter an autoantigen-specific immune response both when they are present during the priming phase as well as when they are present when the primed T cell reencounters its antigen, e.g. in the inflamed CNS.

Objective: Poly I:C, a viral mimic and ligand for TLR 3 is widely used as a paradigm for a viral infection. Since viral infections are both linked with the initiation of multiple sclerosis (MS) as well as with relapses, we here investigated the effect of poly I:C both as an adjuvant as well as a co-stimulant during PLP-specific recall.

Material and methods: PLPp 139–151 injected in CFA subcutaneously and systemic administration of Pertussis toxin were generated to induce EAE in female wild-type SJL mice. Alternatively, mice were injected with PLPp 139–151 and poly I:C as an adjuvant. PLPp-specific T cell responses were assessed by cytokine ELISPOT for pro- and anti-inflammatory cytokine production both in the spleen and in the inflamed CNS.

Results: We find that poly I:C as an adjuvant does not prime an encephalitogenic T cell population, nor does it essentially affect the PLPp-specific T cell cytokine signature during recall in the immune periphery or in the inflamed target organ.

Conclusion: We conclude that different TLR ligands can have fundamentally different effects on an ongoing CNS-autoantigen specific autoimmune process.

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Abstract – WCN 2013

No: 3121

Topic: 6 – MS & Demyelinating Diseases

Neuromyelitis optica and optic spinal multiple sclerosis are different diseases?

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Objective: To compare neuromyelitis optica (NMO) and optic spinal multiple sclerosis (OS-MS).

Method: Consecutive patients with NMO phenotype attended in Rio de Janeiro from 2009 to 2011 were classified in two groups: defined NMO according to Wingerchuk et al. (2006) (recurrent or monophasic) and optic spinal multiple sclerosis (OS-MS).

Results: We analyzed 99 patients: 66 were NMO-R, 8 NMO-M and 25 MS-OS. In NMO-R group, 91% were women, 65.2% African-Brazilian, mean age at onset is 13.03 years, median EDSS of 6.0 (2.0 to 10) in the last evaluation after median disease time of 8 years (2–35); the positivity of anti-AQP4 was 56.1%. In group NMO-M, 62.5% were women, 65.5% African-Brazilian, mean age at onset is 19.77 years, median EDSS of 3.0 (2.0 to 6, 0) in the last evaluation after 7 years of median disease time (3–14); the positivity of anti-AQP4 was 0.0%. In MS-OS group 84% were women, 16% African-Brazilian, mean age at onset is 8.12 years, median EDSS of 3.0 (1.0 to 6.5) in the last evaluation after a median disease duration of 8 years (3.0 to 27.0); the positivity of anti-AQP4 was 0%. There was a statistically significant difference between NMO and OS-MS groups concerning to race, long term disability, extension of the MRI vertebral lesion, positivity of anti-AQP4 and frequency of HLA DR2.

Conclusion: NMO and OSMS in Brazilian population are different demyelinating inflammatory diseases although they share the same clinical phenotype presentation.

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Abstract – WCN 2013

No: 3093

Topic: 6 – MS & Demyelinating Diseases

Demyelinating inflammatory idiopathic diseases in Latin America – A study of prevalent cases

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Background: Epidemiologic studies on multiple sclerosis (MS) and neuromyelitis optica (NMO) have shown that both diseases have particular world of geographic distribution according to population, Caucasians in MS and African ascendants and Asian in NMO. Considering the colonization of Latin America we can distinguish three main ancestries: Caucasians, Mestizos and Afro-descendant populations.

Objective: The aim of this study was to describe the frequencies of MS and NMO in different regions of Latin America with diversified ancestry.

Methods: It was asked of the physicians of MS treatment centers (one in Argentina, one in Paraguay, one in Venezuela and nine in Brazil) to inform all registered cases of idiopathic inflammatory demyelinating disease (IIDD) the frequencies of MS and NMO in following-up, as well as the ancestry of patients.

Results: In Argentina, among the 123 patients (99.2% Caucasians and 0.8% Asians) the frequency of MS and NMO were 93% and 1.6%

respectively; in Brazil, of the 1235 patients (69% Caucasians and 29% African descendants) the frequency of MS was 62.5% and NMO 13%; in Paraguay, of the 164 patients (70% Caucasians and 30% Mestizos) the frequencies were 79% MS and 7% NMO; and in Venezuela, of the 89 patients (64% Mestizos, 18% Caucasians and 9% African descendants), 47% MS and 33% NMO.

Conclusion: In the countries with the lowest number of Caucasians and greater population mixing, Mestizos and Afro descendants, as Venezuela and Brazil the NMO frequency is higher, reinforcing the observation recognized of the ethnic distribution of neuromyelitis optic among non Caucasian populations from tropical regions.

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Abstract – WCN 2013

No: 2936

Topic: 6 – MS & Demyelinating Diseases

Understanding mechanisms of axonal loss in non-optic neuritis eyes of MS patients

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Background: Axonal injury is a major cause of disability in MS. Numerous studies demonstrated loss of retinal ganglion cell axons in MS patients with no history of ON. However, the pathological basis of this loss at present is not clear.

Objective: To investigate links between loss of RGC axons and MS-related injury of anterior (outer retina) and posterior (optic tract and optic radiation) parts of the visual pathway.

Patients and methods: Thickness of temporal RNFL was analysed in 55 RRMS with no history of ON at least in one eye. Integrity of the outer-retina was assessed using electroretinogram (ERG) and measurements of outer retinal layers. Latency of the mfVEP indicated previous inflammatory demyelination along the posterior visual pathway. FLAIR T2 optic radiation lesions were identified using tractography.

Results: There was significant delay of photopic ERG b-wave, which correlated with loss of tRNFL. Significant reduction of tRNFL was also observed in patients who displayed functional (mfVEP delay or abnormal DTI) or structural (lesions) evidence of previous OR damage. No indication of OT lesions was found. Linear regression analysis explained 50% of the tRNFL variability. However, while contribution of ERG delay was significant, estimated predictive power of OR lesions was by far the largest.

Conclusion: Study result suggests dual nature of RGC axonal loss in NON eyes of MS patients. While strong tract-specific relationship between loss of tRNFL and OR damage advocates retrograde trans-neuronal process as a major factor, significant association of tRNFL thickness with ERG delay also implicates primary retinal pathology.

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Abstract – WCN 2013

No: 2939

Topic: 6 – MS & Demyelinating Diseases

Clinical consequences of persistently high-titer neutralizing antibodies in patients with multiple sclerosis – Results from the German reference lab

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Background: In patients with multiple sclerosis (MS) treated with interferon beta (IFNβ), neutralizing antibodies (NABs) may develop. If high-titer (ht) and persistent, these may limit the bioavailability of IFNβ and its clinical benefits.

Objective: To report NAB prevalences from the German reference lab identifying patients with persistent htNABs against IFNβ and to inquire about subsequent clinical decision making.

Methods: Serum samples sent in nationwide between January 2008 and June 2010 for NABs testing were subjected to ELISA screening for binding antibodies and to a luciferase reporter gene assay for quantitative detection of NABs. Treating physicians of patients with htNABs (>100 TRU/mL) detected in two independent samples were contacted by mail to provide follow-up information by means of a standardized questionnaire.

Results: 1990 serum samples were screened for the presence of NABs. NABs and htNABs were detectable in 22% and 4.8% of samples, respectively. Follow-up samples were provided in 11% and demonstrated consistent serostatus in 58%. NABs were lowest with IFNβ-1a i.m. compared to IFNβ-1b s.c. and IFNβ-1a s.c. (8.6% vs. 28.1% vs. 40.5%; $p < 0.0001$, chi-square). 25 patients with persistent htNABs were identified, of which 20 questionnaires were returned; therapy was amended in 16 patients: 75% were switched to glatiramer acetate (GA), 18.7% were escalated to more potent therapies.

Conclusions: The overall prevalence of persistent htNABs is low. When detected, the majority of treating neurologists opted to switch to GA rather than to continue IFNβ or to escalate therapy.

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No: 2763

Topic: 6 – MS & Demyelinating Diseases

MicroRNA, a critical regulator of an autoimmune demyelination

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Background: MicroRNAs (miRNA) are short, non-coding RNAs regulating expression of classical genes and emerge as a mechanism controlling immune reactions. However, a detailed role of miRNA in regulation of autoimmunity remains unclear.

Objective: To analyze microRNA involvement during the development of autoimmune demyelination.

Material and methods: We utilized an animal model of multiple sclerosis – experimental autoimmune encephalomyelitis (EAE). Sorted T helper (Th) cell from lymph nodes of C57Bl/6 mice immunized with MOG peptide 35–55 as well as from MOG-TCR transgenic mice were used for the analyses. We have performed a global miRNA profiling as well as individual miRNA assays during the development of the Th cell autoimmune recognition of myelin antigen.

Results: We have found a specific expression of miRNA in Th cell highlighting three miRNA to be upregulated: mmu-miR155, mmu-miR21 and mmu-miR301a. Mmu-miR155, mmu-miR21 and mmu-miR301a were also upregulated in brain infiltrating cells isolated from EAE. The changes in the above miRNA correlated with both in vitro and in vivo Th17 differentiation. Use of specific miRNA antagonists revealed that miR-301a contributed to the development of the Th17 subset via targeting of the interleukin 6/23-STAT3 pathway. The analysis of MS patients ($n = 48$) and controls ($n = 36$) revealed that stimulation of peripheral blood T cells lead to significantly higher overproduction of miR-301a in MS patients v. controls (2.9 v. 0.7 fold induction, $p < 0.015$).

Conclusion: We have identified a previously unknown critical role of miR-301a in regulation of autoimmune demyelination and development of autoreactive subset of Th cells.

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Abstract — WCN 2013

No: 2910

Topic: 6 — MS & Demyelinating Diseases

Fingolimod (FTY720) oral for the treatment of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP): Study design of the phase 3 FORCIP trial

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Background: CIDP is a chronic sensorimotor neuropathy causing significant disability. Fingolimod, a sphingosine-1-phosphate receptor modulator, completely prevented paraparesis, significantly decreased T-/B-cell, macrophage infiltration and demyelination of sciatic nerves in experimental autoimmune neuritis.

Objective: Evaluate efficacy in delaying disability progression (≥ 1 point increase from baseline on the adjusted Inflammatory Neuropathy Cause and Treatment (INCAT) disability scale), safety and tolerability of oral fingolimod 0.5 mg daily compared with placebo in CIDP patients.

Patients and methods: FORCIP study is a double-blind, randomised, multi-centre, placebo-controlled, parallel-group study in adult patients with CIDP and history of disease activity upon discontinuation of CIDP therapy. Patients are randomised (1:1) to either fingolimod 0.5 mg or placebo. The primary outcome is the time-to-first disability progression. The study has >90% power at a one-sided 2.5% significance level to detect a hazard ratio of 0.51; 111 events are anticipated in approximately 156–200 randomised participants for the final analysis. Participants fulfilling the disability event criterion will discontinue the study medication immediately and complete the study with a standard CIDP treatment. A group-sequential design with one planned interim analysis after 50 events for futility stopping (one-sided stratified log-rank test; futility boundary at p -value ≥ 0.2282) will be used to prevent patients from prolonged exposure to potentially inefficient study drug. The study otherwise will continue until the required number of events are observed, or up to a maximum of three years from the trial initiation.

Results and conclusion: This study will provide evidence whether fingolimod can delay disability progression in patients with CIDP.

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Abstract — WCN 2013

No: 2918

Topic: 6 — MS & Demyelinating Diseases

Discontinuation of natalizumab — Reasons and implications

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Background: Long-term treatment of multiple sclerosis with natalizumab is limited due to the risk of progressive multifocal leucoencephalopathy.

Objective: To evaluate reasons of treatment discontinuation and its effect on disease activity within 12 months after cessation of natalizumab.

Methods: We retrospectively analyzed data of 131 MS patients who discontinued natalizumab between 2007 and 2012. Patient data was taken from the Austrian natalizumab registry and completed by a questionnaire sent to 19 MS centers. Patients with data available both from the registry and the questionnaire were included.

Results: Duration of therapy exceeded 3 years in only 15.3%, while mean duration was 24.3 months (range 4 months–56 months). Most frequent reasons for discontinuation were: disease activity (14%), seropositivity for JC-virus (14.7%), and long-term treatment in JCV-seropositive patients (14.7%). Only 103 patients again started MS specific-therapies, 15 of them natalizumab. Annualized relapse rate (ARR) increased from 0.53 ± 0.97 on natalizumab to $1.2 (\pm 1.44)$ post-natalizumab ($p < 0.001$). Compared to pre-treatment levels (mean ARR 2.1), ARR after cessation was significantly lower ($p < 0.001$). Disease activity after 12 months exceeded pre-treatment levels in 14%, and was the same in 16.5% and lower in 69.4% of patients. At the beginning and end of natalizumab median EDSS was 3.5 and 3.0, respectively, and 12 months after cessation it increased to 3.5 ($p = 0.028$).

Conclusion: We could not find a rebound phenomenon within 12 months after cessation of natalizumab. While EDSS remained the same, ARR was significantly lower 12 months after discontinuation as compared to the pre-treatment year.

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Abstract — WCN 2013

No: 2956

Topic: 6 — MS & Demyelinating Diseases

The role of T-cells in the pathogenesis of neuromyelitis optica

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Neuromyelitis optica (NMO) is a severely disabling, demyelinating inflammatory disease of the central nervous system (CNS). The diagnostic hallmark of NMO is the presence of antibodies in the serum, which are specifically directed against Aquaporin 4 (AQP4), a water channel enriched on astrocytic endfeet at the glia limitans. These antibodies have been shown to be pathogenic. Once inside the CNS, they find their target on the surface of astrocytes, fix complement and initiate the complement mediated destruction of these cells.

We have previously shown in our animal-NMO-models that acute lesions with AQP4 loss can form when CNS antigen specific T cells initiate CNS inflammation, whereas no lesions can be induced with AQP4 antibodies only. Furthermore we could see that the antigen specificity had an impact on the speed of lesion formation, peripheral organ affection and recruitment of other inflammatory cells. We found that T cells which produce the largest lesions in AQP4 antibody positive rats were best reactivated within the CNS and produced the most IFN- γ . We then analyzed the effects of IFN- γ on the gene expression by microglia using microarray analysis. IFN- γ treated microglia produced complement factors, downregulated complement inhibitors and upregulated Fc γ r, and hence significantly contribute to the complement mediated destruction of astrocytes.

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Abstract – WCN 2013**No: 2955****Topic: 6 – MS & Demyelinating Diseases****The experience of the management of pregnant women with multiple sclerosis**T. Yakushina, V. Lizhdvoy. *Neurology, MRRICI, Moscow, Russia*

Due to the fact of the high percentage of women of the childbearing age among patients with MS, the problems of the management of pregnancy are given a special attention. The object of study is learning the characteristics of the management of pregnancy and partus of pregnant women with MS. Forty pregnant women with MS entered the study. 24 women received immunomodulatory treatment: Copaxone – 14, interferons – 6, Cladribine (in anamnesis) – 2, Mitoxantrone – 2. 24 women had normal pregnancy. 16 had abnormal pregnancy. There were no exacerbations of MS during the pregnancy. 26 women had partus in time. 7 women had premature labor. 10 women had caesarian section. The pregnancy of 6 women has been continuing. First woman, who was receiving Cladribine earlier, partuited the boy with low weight, plural congenital abnormalities. Second woman, who was receiving Cladribine earlier, partuited the normal child. 2 women, who were receiving Mitoxantrone earlier, had normal pregnancy and premature labor, partuited the children with low weight. The women, who was receiving Copaxone, interferons and no immunomodulatory treatment earlier, had normal pregnancy and partus. The management of pregnancy and partus isn't different from the general population. After immunomodulatory treatment the risk of development of complication and congenital abnormalities isn't different from the general population. After the immunosuppressant treatment the risk of congenital abnormalities increases.

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Abstract – WCN 2013**No: 2773****Topic: 6 – MS & Demyelinating Diseases****Next-generation sequencing for detection of mutations associated with rare disorders in patients meeting diagnostic criteria for primary progressive multiple sclerosis**

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Background: Rare Mendelian disorders have diverse phenotypes and many show variable expressivity. Given that some spastic paraparesis (SP) disorders show phenotypic overlap with primary progressive multiple sclerosis (PPMS), we hypothesized that sequence variants in these known disease genes confer susceptibility for progressive 'MS-like' phenotypes that meet McDonald criteria for PPMS.

Objective: To evaluate PPMS patients for sequence variants in genes associated with Mendelian causes of SP.

Patients and methods: DNA from 250 phenotyped PPMS patients are being interrogated using a custom next-generation sequencing panel (Nimblegen SeqCapEZ) of 72 genes associated with rare, progressive SP phenotypes. All identified variants were assessed by expert consensus and classified based on pathogenicity. Pathogenic and likely pathogenic mutations were confirmed by Sanger sequencing.

Results: 3/35 (8.6%) PPMS patients analyzed to date harbor previously reported heterozygous disease-causing mutations in genes associated

with dominant phenotypes that explain or significantly contribute to presenting phenotypes. Five patients (14.3%) had novel mutations predicted to be disease-causing in genes associated with dominant phenotypes, and six (17.1%) were heterozygous for known disease-causing mutations in genes associated with recessive disorders.

Conclusion: The results to date demonstrate that this novel sequencing strategy is effective at detecting variants in known SP genes among PPMS patients. We report identified mutations and the algorithms used to indicate pathogenicity. Data from this ongoing study may lead to further characterization of the clinical spectrum and prevalence of several rare Mendelian neurologic disorders, and provide guidelines for genetic evaluation of patients meeting PPMS criteria.

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Abstract – WCN 2013**No: 2751****Topic: 6 – MS & Demyelinating Diseases****Movement disorders in demyelinating diseases**B. Dönmez Colakoglu, E. Idiman, S. Ozakbas. *Dokuz Eylul University Medical Faculty, Department of Neurology, Izmir, Turkey*

Background: In multiple sclerosis (MS), movement disorders other than cerebellar tremors are rare.

Objective: The study aims to determine movement disorders in patients diagnosed with demyelinating diseases and to investigate clinical manifestations.

Patients and methods: 2783 patients are followed in our demyelinating disease outpatient clinic. Of these 2783 patients, those who presented to the demyelinating disease outpatient clinic in the past one year and had different types of movement disorders were evaluated.

Results: Last year, ten cases were diagnosed with movement disorders. Of them, seven had dystonia (three upper extremity dystonia, three paroxysmal dystonia and one writer's cramp), one Parkinsonism, one choreoathetosis and one postural tremor. Of these cases, seven were diagnosed with definite MS according to McDonald criteria and two with clinically isolated syndrome (CIS) and one with neuromyelitis optica. In two patients, MS symptoms began before the age of 18. While movement disorders were the initial symptom in one patient, in the other patients, movement disorders developed during the course of the disease.

Conclusion: Although tremor is the most common symptom in MS patients, movement disorders other than tremors can also occur, even as the first symptom. Movement disorders can occur not only in MS but also in other demyelinating diseases such as neuromyelitis optica. We consider that the careful evaluation and follow-up of these patients would be of great importance in order to reveal the relationship between multiple sclerosis and movement disorders.

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Abstract – WCN 2013**No: 2672****Topic: 6 – MS & Demyelinating Diseases****EAE disease prevention by a potent oral ROR γ t inverse agonist INV-17: A promising safe & efficacious ms treatment**A. Gaweco^{a,b}, S. Palmer^a, R. Shamilov^a, M. Fisher^a, J. Tilley^a. ^aInnovimmune Biotherapeutics, Inc., Brooklyn, NY, USA; ^bSUNY Downstate Medical Center, Brooklyn, NY, USA

Background: T helper 17 [T_H17] cells and its production of T_H17 cytokines play a critical role in the pathogenesis of MS and EAE. Retinoic acid receptor-related orphan receptor gamma t [ROR γ t] is a

nuclear hormone receptor that specifically regulates T_H17 cells by acting as a control switch for T_H17 differentiation and function. We have discovered proprietary novel lead chemical scaffolds of the INV-17 portfolio of small molecule ROR γ t inverse agonists.

Objective: To establish the *in vivo* proof of concept of the lead clinical compound candidate INV-17 in the chronic mouse EAE model.

Method: INV-17 was administered p.o. for 28 days as a prophylactic treatment in MOG_{35–55}-induced C57BL/6 mice to assess the therapeutic benefit of preventing disease in a chronic-progressive EAE model.

Result: Disease was successfully prevented in INV-17-treated mice with cumulative EAE scores (Scale: 0–5) of 3.3 ± 3.3 ($p = 0.008$) which is in striking contrast to 27.9 ± 6.2 of vehicle-treated animals with 100% disease incidence. Worse clinical score AUC of 26.50 ± 6.28 for the vehicle group was significantly different from 3.00 ± 3.00 ($p = 0.0097$) for the INV-17 group. INV-17 was well tolerated and INV-17-treated mice were unremarkable with optimal body conditions.

Conclusion: The superior safety and efficacy data following chronic oral INV-17 dosing provide compelling evidence of the effectiveness of ROR γ t inverse agonism and support advancing INV-17 into Preclinical/IND-enabling development stage. These findings highlight the potential promise of INV-17 as a safe & efficacious novel MS treatment.

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Abstract — WCN 2013

No: 2881

Topic: 6 — MS & Demyelinating Diseases

The prevalence of autoantibodies in ms patients in Riga East Clinical University Hospital, Latvia

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Introduction: Multiple sclerosis (MS), lupus erythematosus (LE), antiphospholipid syndrome (AFS) are chronic, immune-mediated, relapsing–remitting disorders. Neurological symptoms and MRI can have the same picture, but pathogenesis and treatment are different. **Objectives:** To analyse antinuclear antibodies (ANA), antiphospholipid antibodies (APA) and lupus anticoagulant (LA) in MS patients with specific symptoms and MRI findings and determination of specificity, sensitivity in MS differential diagnosis.

Methods: 31 MS patients from Riga East Clinical University Hospital were included in the study. The patients were in remission and did not get any disease modifying therapy before. The diagnosis was established in a classic way — with MRI by McDonald's criteria. Clinical symptoms, MRI, AFA IgG, IgM, LA, and ANA were analysed.

Results: Of 31 patients 17 (55%) were female with mean age of 34.4 ± 9.8 and male 14 (45%), were mean age of 36.1 ± 8.8 . All patients had changes in MRI according to McDonald's criteria. Spinal cord changes were detected in 90% of patients and optic neuritis in 59%. Spinal cord and optic nerve injury was noted more in females 55%. Positive autoantibody was detected in 1 female, who didn't have any clinical differences from other patients.

Conclusions:

1. ANA, AFA positivity is not frequent in MS patients with classic MS symptoms and MRI findings.
2. Autoantibody tests in MS patients are to be considered when the symptoms of the disease are atypical.
3. MS patients with positive autoantibody tests, should be further reevaluated by repeating the autoantibody tests and observing the clinical symptoms, in order to review of the diagnosis on time.

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Abstract — WCN 2013

No: 2838

Topic: 6 — MS & Demyelinating Diseases The spectrum of neuromyelitis optica

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Background: Neuromyelitis optica (NMO) is an idiopathic inflammatory demyelinating disease of the central nervous system with a predilection for the optic nerves and spinal cord with positive anti-aquaporin 4 antibody.

Objective: To review the clinical features of our patients who tested positive for anti-aquaporin 4 antibody.

Methods: Patients referred to our clinic with positive anti-aquaporin 4 antibody were reviewed.

Results: We reviewed 4 patients who had a history of transverse myelitis. 2 had a history of optic neuritis and both had recurrent myelitis and optic neuritis and were treated as mixed connective tissue disease and multiple sclerosis respectively. The remainder 2 had first onset transverse myelitis as their sole clinical finding. None were diagnosed as neuromyelitis optica initially. All patients had longitudinally extensive transverse myelitis (LETM) on MRI. All 4 tested positive for anti-aquaporin 4 antibody. All patients had their diagnoses revised. 2 had definite NMO while the rest were classified as NMO Spectrum Disorder (NMOsD) as they did not meet the criteria for definitive NMO.

Conclusions: All patients with longitudinally extensive transverse myelitis should be tested for anti-aquaporin 4 antibody.

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Abstract — WCN 2013

No: 2822

Topic: 6 — MS & Demyelinating Diseases

The effects of multiple sclerosis on women's sexuality: A controlled study

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MS is considered to have negative effects on women's sexual function. Sexual function of MS women seems to be associated with functional status and depression levels. Therefore, women with MS should also be evaluated with the suspicion of sexual dysfunction during the diagnostic and follow-up procedure. Mean age rates were 34.18 ± 8.23 in the study group and 35.32 ± 8.12 in controls ($p > 0.05$). While mean duration of complaints, and EDDS, BDI and total FSFI scores were 7.66 ± 5.47 years, 1.52 ± 1.43 , 18.20 ± 10.56 and 20.31 ± 8.91 in patients respectively, the scores of BDI and total FSFI were 7.61 ± 5.43 and 30.55 ± 4.29 in controls, respectively. Compared to controls, BDI was higher, and total and subscores of FSFI were lower in women with MS ($p < 0.001$). In MS patients, a negative correlation was present between EDDS, and BDI ($p < 0.001$, $r = -748$), total FSFI ($p < 0.001$, $r = -822$), and among FSFI subscores, desire ($p < 0.001$, $r = -779$), orgasm ($p < 0.001$, $r = -781$), satisfaction ($p < 0.001$, $r = -762$), pain ($p < 0.001$, $r = -711$), lubrication ($p < 0.001$, $r = -819$) and arousal ($p < 0.001$, $r = -812$). Fifty-one premenopausal women with MS were included into the study group, and 57 healthy premenopausal women constituted controls. Functional status of MS patients was evaluated with Expanded Disability Status Scale (EDSS); depression levels with Beck Depression Inventory (BDI); and, sexual functions with Female Sexual Function Inventory (FSFI). While

evaluating the data, mean, standard deviation, student's *t* test and Spearman's correlation analysis were used. To determine the sexual disability and related factors in women with multiple sclerosis (MS).

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Abstract – WCN 2013

No: 366

Topic: 6 – MS & Demyelinating Diseases

Disability improvement in the first case treated by Natalizumab in Libya

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A 31 year old Libyan lady presented with left lower limb cramps & sole numbness 6 years ago which spontaneously cured within 1 week. 4 months later she had the same symptoms & received IV steroids, then she was put on injectable Interferon-beta. She was stable & relapse free till Jan 2010 when she had left lower limb weakness & received IV steroids. 10 months later she had left side body numbness & left lower limb weakness & received IV steroids. 3 months later she had diplopia & right eye blurred vision with right 3rd nerve palsy followed by left lower limb weakness & received steroids, but neurological deficit & EDSS worse than before. 7 months later she had bilateral lower limb weakness & numbness up to the waist & received IV steroids. 4 months later she had left lower limb weakness, unsteadiness, right side body numbness with right side facial numbness & urinary incontinence & received steroids. At that time she started the 1st dose Natalizumab in Mar 2012 then once every 4 weeks till this month when she received the 13th dose. Clinically she had treatment ataxic paraparesis with right side hypoesthesia and urinary incontinence & we noticed that the patient started to improve clinically at the 3rd dose & the EDSS is getting better since then. Nowadays she has no neurological disability just mild past pointing in the left side & high DTR all over the body (Lt > Rt). MRI brain & cervicodorsal spine were done several times in each relapse and follow up MRI after Natalizumab treatment started which showed incredible improvement so far.

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Abstract – WCN 2013

No: 367

Topic: 6 – MS & Demyelinating Diseases

Retrograde diagnosis of tumefactive multiple sclerosis

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A 40 year old Libyan lady presented with acute severe headache & bilateral blurred vision with nausea & vomiting which happened 8 years ago. Clinically she had left temporal hemianopia & brisk reflexes all over the body with bilateral Babinsky sign. MRI brain showed large high signal intensity peripherally enhancing lesion in the right occipito-parietal region surrounded by edema and marked compression on the occipital horn of the lateral ventricle. They thought it was a tumour & surgical resection was done followed by an IV steroid therapy. Clinically post-operative she had permanent left temporal hemianopia only otherwise she was fully ambulatory without disability in spite of the huge brain tissue defect. 4 months later she had acute severe headache & MRI brain revealed recurrence of right occipital enhancing lesion with new involvement of the splenium of the corpus callosum. A 2nd operation was done with

resection of the new lesion & no clinical disability except left temporal hemianopia. A histopathology study showed nonconclusive inflammatory changes. 2 months later she had severe headache & bilateral blurred vision with tunnel vision. Clinically she had papilledema. MRI brain showed new hyperintense lesion noted in the left occipital region with marked enhancement in addition to the previous right occipital one & the splenium. IV steroids were given with dramatic improvement. 3 months later follow up MRI brain showed a decrease in size of the enhancing areas of both occipital regions after steroid therapy. 1 month later a stereotactic biopsy was done abroad & revealed a confirmed diagnosis of Multiple Sclerosis. IV Metoxantrone was started & she is stable till now.

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Abstract – WCN 2013

No: 2840

Topic: 6 – MS & Demyelinating Diseases

Disability assessment results of MS patients in Armenia

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Background: Multiple sclerosis (MS) is a degenerative, inflammatory, immune-mediated disease affecting 2.5 million individuals worldwide presenting the most common disabling disease of the CNS in young adults. The wide range of symptoms affects all aspects of physical and mental health. Unfortunately, no officially published up-to-date data exists on MS prevalence in Armenia.

Objective: The primary aim of the study is to establish association between the EDSS score and overall quality of life among MS patients. This study will also reevaluate the prevalence and incidence of MS in Armenian population, and assess the disability status and quality of life.

Methods: Cross-sectional study design was used to assess all patients with definite MS admitted to our clinic. The quality of life was measured by a Multiple Sclerosis Quality of Life (MSQOL)-54 questionnaire, which was translated and adapted for use with Armenian speakers. The Expanded Disability Status Scale (EDSS) evaluated disability. The data was analyzed using descriptive statistics, correlation and regression analysis.

Results: 45 MS patients were interviewed and underwent neurological examination. Mean age and duration of disease were 34 and 5.5 years respectively. Mean physical health and mental health composite scores were 105.5 and 54.4. Statistically significant correlation was established between (MSQOL)-54 physical composite score, mental health composite score and EDSS ($p < 0.01$) score implying that poorer QoL was associated with higher EDSS scores and more severe disability.

Conclusion: The findings being consistent with previous studies indicate that measures should be taken to improve the quality of life and disability status of these patients.

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Abstract – WCN 2013

No: 2849

Topic: 6 – MS & Demyelinating Diseases

Duration of multiple sclerosis on quality of life:

Preliminary results of Armenian study

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Background: Multiple sclerosis (MS) is an inflammatory, immune-mediated disease of the central nervous system, affecting and disabling young adults with complexity of manifestations varying from benign up

to devastating disease. It is obvious that the disease affects all aspects of the patient's mental and physical health negatively impacting on overall quality of life of a person.

Objective: The aim of the study was to study correlation between the quality of life and disease duration.

Methods: In this cross-sectional study all patients with definite MS included in 3 year long research were evaluated for quality of life measured by a Multiple Sclerosis Quality of Life (MSQOL)-54 questionnaire previously validated and adjusted for use with Armenian speakers and disability measured by the Expanded Disability Status Scale (EDSS). Descriptive statistics, correlation and regression analyses were included in statistical analysis.

Results: In total, 45 MS patients were interviewed and assessed. The mean age of the participants was 34 with male predominance (24/21). The average disease duration was 5.5 years. Mean physical health and mental health composite score were 105.5 and 54.4 respectively. Totals of each of the domains of (MSQOL)-54 were also calculated and analyzed. Statistically significant correlation was found between disease duration and (MSQOL)-54 physical composite score ($p = 0.001$) and mental health composite ($p = 0.003$) indicating that poorer QoL was associated with longer disease duration.

Conclusion: The findings suggest that the duration of the disease can be a significant clinical parameter for the quality of life in MS patients, and must be considered in patient evaluation.

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Abstract – WCN 2013

No: 2855

Topic: 6 – MS & Demyelinating Diseases

The probiotic therapy in patients with multiple sclerosis: Microbiological, immunological and clinical aspects

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Background: In recent years, the results of some pilot studies demonstrated the possible role of the intestinal microflora in the development of the demyelinating process in the CNS. Based on this, views were expressed that the correction of the intestinal dysbiosis possibly can be used for prevention and treatment of multiple sclerosis (MS).

Objective: To evaluate the effect of probiotics on intestinal microflora and some immunological parameters in patients with MS.

Materials and methods: We examined 40 treatment-naive patients with relapsing–remitting MS, conducting clinical, bacteriological and immunological studies in the dynamics. Patients with intestinal dysbiosis received therapy with probiotic "Bifidumbakterin forte".

Results: Intestinal dysbiosis was identified in 87.5% of patients with MS. The incidence and severity of dysbiosis were associated with the clinical severity of MS. In 71.4% of cases dysbiosis had clinical manifestations (dyspepsia). Dysbiosis was characterized by changes in both the indigenous and pathogenic flora: decreased levels of bifidobacteria and lactobacilli, changes in the content of *E. coli*, elevated levels of *Candida*, *Staphylococcus*, *Klebsiella*, and *Clostridium*. There were correlations between the greater severity of intestinal dysbiosis, and lower levels of T-lymphocytes, phagocytosis, oxygen-dependent metabolism of neutrophils, and increased levels of IgM and IgG. After a course of probiotic complete or partial regression of dysbiosis observed in 80% of patients; also there was a reduction of circulating immune complexes, CD-HLA-DR, initially increased helper–suppressor ratio, as well as reducing the severity of dyspepsia.

Conclusion: Probiotic therapy may be considered as a promising pathogenetic (immunomodulatory) and symptomatic therapy of MS!

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Abstract – WCN 2013

No: 2746

Topic: 6 – MS & Demyelinating Diseases

The influence of thyroid autoimmune reactivity in the course of multiple sclerosis

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Background: Many authors have noted a higher frequency of autoimmune thyroid disease occurrence in patients with multiple sclerosis (MS). The influence of the autoimmune response to thyroid antigens on the course of MS is very interesting.

Objective: The objective of this study was to determine the characteristics of MS in patients with thyroid autoimmune reactivity.

Patients and methods: 84 patients (16 males, 68 females) with clinical definite relapsing–remitting MS were examined. All patients had clinical remission. The average age was 37.1 ± 10.03 years (18–56 years). Severity of neurological deficit was assessed according to the EDSS. All patients underwent ultrasonography of the thyroid gland, serum for thyroid hormones (TG), and anti-thyroid peroxidase and thyroglobulin. All statistical analyses have been made using a standard statistical package.

Results: 17% of patients had elevated titers of antibodies to thyroid antigens. 26% of patients without thyroid pathology were the comparison group. These groups were comparable in age and gender composition.

The continuance of first remission in patients with thyroid autoimmunity was significantly shorter (1.4 ± 1.11 and 2.7 ± 3.81 years, $p = 0.048$). In patients with thyroid antibodies progression rate was 0.9 ± 0.75 score/year, in the comparison group it was 0.5 ± 0.34 score/year, $p = 0.03$. There was a statistically significant correlation between the levels of antibodies to thyroglobulin with the total number relapse, $r = 0.59$, $p = 0.01$.

Conclusion: The presence of the patient autoimmune reactivity to antigens of the thyroid can be a predictor of severe MS.

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Abstract – WCN 2013

No: 2766

Topic: 6 – MS & Demyelinating Diseases

Multiple sclerosis in Armenia: Quality of life and depression

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Background: Multiple sclerosis (MS) is an inflammatory demyelinating disease affecting the Central Nervous System, and is a leading cause of disability in young adults. Quality of life (QoL) usually decreases in MS patients. Many of them develop depression. Currently there are no accepted publications showing different aspects of MS in Armenia.

Objective: We aimed to evaluate QoL, its correlation with disease duration and type, depression and their intercoupling in MS patients.

Patients and methods/material and methods: 45 patients with different types of MS had been investigated. For all patients diagnosis was based on clinical symptoms and McDonald MRI criteria. Average age and disease duration were 34 and 6.2 years, respectively. Quality of life was assessed by Multiple Sclerosis Quality of Life (MSQOL)-54 questionnaire which includes both physical and mental health assessment scales. Beck's Depression Inventory (BDI) was used in order to diagnose depression.

Results: Average physical and mental health composite scores for MS patients were 82.06 and 54.4, respectively. Statistically significant correlation was established between (MSQOL)-54 physical composite and BDI scores, mental health composite and BDI scores, intimating that

decreased QoL was associated with higher BDI scores ($p < 0.001$). Long-term disease duration was associated with lower QoL. Decrease of QoL was directly proportional to the high degree of neurological deficit and frequency of new attacks.

Conclusion: Decreased QoL is primarily connected with long-term disease duration, degree of neurological deficit, frequency of new attacks and depth of depression. QoL must be considered in effective management of MS patients.

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Abstract – WCN 2013

No: 2770

Topic: 6 – MS & Demyelinating Diseases

Carbonyl proteins as marker of oxidative stress derived protein damage in neuroinflammatory and neurodegenerative diseases

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Background: The importance of oxidative stress in the genesis of neuro-immunological diseases has been discussed extensively. Carbonyl groups may be the end link of a series of metabolic processes leading to apoptosis and neuron damage.

Multiple sclerosis (MS) is the most common chronic neuro-inflammatory disease and a major cause for disability in young adults. Oxidative stress is driven by inflammation and leads to degeneration. By now, established markers of oxidative stress in CSF are rare.

The aim of the study is to detect carbonyl proteins in CSF in patients with MS, neurodegenerative diseases and healthy controls.

Materials and methods: CSF from diagnostic lumbar punctures was tested. To determine levels of oxidative stress in cerebrospinal fluid (CSF) Carbonyl Protein was measured by the Carbonyl Protein ELISA Kit (Immundiagnostik AG, Germany).

Results: Patients were divided in three subgroups (neuro-immunological disease, neurodegenerative disease, healthy controls).

Level of CP differed significantly between subgroups ($p = 0.025$) (Kruskal–Wallis-Test), mean values: neuro-immunological disease 630.8, neurodegenerative disease 756.1, and healthy controls 356.5.

Conclusion: In this pilot trial we tried to detect carbonyl groups (proteins?) as markers of oxidative stress. The results of this trial showed a significant difference in the mean levels of proteins in CDMS and neurodegenerative diseases in comparison to healthy controls. In conclusion, the trial showed that carbonyl proteins are able to serve as marker for oxidative stress. Larger trials have to show whether carbonyl proteins may be able to serve as biomarker for more severe course of disease or for developing CDMS in patients with CIS.

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Abstract – WCN 2013

No: 2771

Topic: 6 – MS & Demyelinating Diseases

NK cell expansion predicts treatment response to rituximab

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Background: Rituximab is used in a plenty of neuro-immunological diseases although it has not been approved for any neurological disease. Positive results have been published for relapsing–remitting Multiple Sclerosis (RRMS), Neuromyelitis optica (NMO) as well as case reports on

secondary progressive MS (SPMS), and myasthenia gravis (MG). Results in RRMS and NMO are impressive, whereas results are inconsistently in progressive MS and MG. Because of possible side effects of rituximab, biomarkers to predict clinical response to rituximab would be of particular interest. In rheumatoid arthritis, a favourable response to CD20 therapeutics was associated with an increase in NK. The aim of this study was to detect predictors of efficacy in patients with neuro-inflammatory diseases treated with rituximab.

Methods: We determined the levels of CD3+, CD3+CD8+, CD3+HLADR+, CD3–CD16&56+ and CD3+CD16&56+ cells in patients with neuro-immunological diseases.

Results: Overall fourteen patients were included in this study. Patients were grouped in responders and non-responders according to clinical disease progression during rituximab therapy. Responders showed a higher β and R2 for NK-cells in contrast to non-responders. The difference between these groups differed significantly ($p < 0.001$).

Conclusion: We found a statistically significant difference between responders and non-responders to Rituximab therapy in patients with neuro-immunological diseases. Responders showed a higher increase in the NK-cell population following initiation of Rituximab treatment as compared to non-responders.

Our results are in line with those in rheumatoid arthritis. Therefore we suggest that NK-cells might serve as a predictor of clinically favourable treatment response to Rituximab in patients with neuro-inflammatory diseases.

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Abstract – WCN 2013

No: 2774

Topic: 6 – MS & Demyelinating Diseases

Microna regulation of B cells in multiple sclerosis: MIR-320 and its specific targets

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Background and objective: B cells are strongly implicated in the pathogenesis of multiple sclerosis (MS). The role of microRNA (miRNA) in MS is not well understood. We studied if miRNAs regulate pro-inflammatory capabilities of B cells in MS.

Patients and methods: B cells and monocytes were separated from untreated patients with MS and age- and gender-matched control healthy subjects (CHS). Expression of 904 miRNAs was tested by microarrays followed by validation with real-time qPCR. Expression of molecules targeted by miRNAs was analyzed by Western blot, ELISA and Flow cytometry. Transfection experiments with miRNA inhibitors were conducted to prove the inhibitory effect of endogenous miRNAs on expression of its targets.

Results: Expression of miR-320 was significantly decreased in B cells of MS patients compared to CHS, $p = 0.014$, but not in monocytes. Expression of molecules specifically targeted by miR-320, Matrix metalloproteinase 9 (MMP-9), the protein implicated in disruption of blood-brain barrier, and Transferrin receptor protein 1 (TFRC), was significantly increased in B cells of patients. To test whether endogenous miRNA-320 inhibits MMP-9 secretion and TFRC expression, B cells from CHS were transfected with specific miR-320 inhibitor which led to increased TFRC expression and MMP-9 secretion to levels seen in patients with MS.

Conclusion: miR-320 expression is selectively decreased in B cells of patients with MS. This is associated with increased expression of miR-320-specific targets, TFRC and MMP-9, which promote cell proliferation and blood–brain barrier disruption, respectively.

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Abstract — WCN 2013**No: 2775****Topic: 6 — MS & Demyelinating Diseases****Assessment of natural course and prognosis of CIS: A prospective study on Iranian patients**

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Objective: To assess natural course of clinically isolated syndrome (CIS) and the role of some probable risk factors for the development of CDMS, in an Iranian population.

Materials and methods: In this prospective study, we studied 50 consecutive patients with CIS, presented in the neurology clinic of Rasul-e-Akram hospital in Tehran, from April 2007 to March 2011. We followed up each patient in 3-month intervals. The endpoint of the study was either diagnosis of CDMS or the end of considered period for follow up.

Results: We studied 50 patients with CIS (40 women, 10 men), age 29.0 ± 8.4 years. 76% of patients were between 20 and 40 years old. The most common presenting syndrome was optic neuritis (46%). Clinical conversion rate was 44%, during the follow up time 29.4 ± 11.4 months. It took 9.1 ± 9.6 months for CIS to convert to CDMS. The primary EDSS scores 1.3 ± 1.0 . Initial brain MRIs were space positive in 58% of cases, and initial cervical MRIs were normal in 45.8% of patients. In this study, we did not observe any statistical association between age at onset of disease, sex, CIS type, primary MRI findings, primary EDSS scores and progression of CIS to CDMS.

Conclusion: Since early diagnosis and treatment of MS are very helpful in proper management of the disease, it would be of great benefit to assess the likelihood of developing CDMS in every patient presenting with a first episode suggestive of MS.

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Abstract — WCN 2013**No: 2776****Topic: 6 — MS & Demyelinating Diseases****Correlation between retinal nerve fiber layer thickness and expanded disability status scale in Iranian patients with relapsing–remitting multiple sclerosis**

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Objective: To evaluate the peripapillary retinal nerve fiber layer (RNFL) thickness in relapsing–remitting multiple sclerosis (RRMS) and its association with disability scales.

Patients and methods: In this cross-sectional study, 69 patients with RRMS, with or without history of optic neuritis (ON), were coincidentally included. The patients' expanded disability status scale (EDSS) scores were evaluated by a neurologist. Peripapillary RNFL thickness was assessed by an ophthalmologist, using a Heidelberg Retinal Tomography (HRT) II device.

Results: 56 females and 13 males, with mean \pm SD age of 29.4 ± 4.1 (range = 17–59) years old, disease duration from 2 months to 20 years were studied. EDSS score measurements 2.83 ± 0.56 , mean of 2.69 and 2.86 for men and women respectively. Mean \pm SD of RNFL thickness of both eyes 0.223 ± 0.051 in all patients, 0.215 ± 0.053 in patients with history of ON, and 0.231 ± 0.051 in those without history of ON. Mean RNFL thickness were not statistically different in patients with and without history of taking interferon. We did not find any statistical relation between disease duration and mean RNFL thickness of both eyes, in all patients and patients with or without ON. There was no

significant correlation between EDSS and mean RNFL thickness of both eyes, in all patients and patients with or without history of ON.

Conclusion: Although not statistically approved, measurement of RNFL thickness appears to be a good strategy in evaluating the natural course of RRMS and the level of disability.

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Abstract — WCN 2013**No: 2755****Topic: 6 — MS & Demyelinating Diseases****A global survey on risk and occurrence of both domestic and work accidents in patients with multiple sclerosis**

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Background: Multiple Sclerosis (MS) can cause a reduction of functional independence in daily living, employment and community participation. Previous studies suggested that MS patients are more prone to incur in accidents, such as falls, during their lifetime. However, few data are currently available about domestic and work accidents.

Objectives: Our goals were reporting frequencies of main types of domestic (DA) and work accidents (WA) in MS, and assessing possible influence of cognitive and physical symptoms on their occurrence.

Patients and methods: We enrolled two cohorts of MS patients and matched control groups (CTRL). Thirty-one MS workers were interviewed about accidents and related risks on working places, while fifty MS subjects were chosen to respond about domestic traumas. Scales of neurological disability, cognitive impairment, mood and fatigue were also performed.

Results: While no statistical difference was found in the amount of WA and DA types between MS and CTRL, bumps emerged as the most recurrent type in MS. Significant correlation was found between bumps and Raven Matrices within MS DA group, while only risk of accident positively correlated with both cognitive fatigue and self-perceived depression in WA. Notably, EDSS and MSFC scores did not seem to influence WA and DA.

Conclusion: Our report showed the prevalence of bump, both in DA and WA MS cohorts. Interestingly, disability does not seem to play a major role in determining the event. Cognition, fatigue and self-perceived depression might be crucial in predicting the risk of WA.

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Abstract — WCN 2013**No: 2296****Topic: 6 — MS & Demyelinating Diseases****Safety and tolerability of BG-12 (dimethyl fumarate) in relapsing–remitting multiple sclerosis: Interim results from the endorse extension study**

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Background: BG-12 (dimethyl fumarate) demonstrated a positive benefit-risk profile in the Phase 3 DEFINE and CONFIRM studies.

Objective: To report interim safety results (as of March 30, 2012) from ENDORSE, an ongoing, 5-year extension of DEFINE and CONFIRM.

Patients and methods: Patients previously randomized to BG-12 240 mg twice (BID) or three times daily (TID) continued on the same dose in ENDORSE. Patients previously randomized to placebo (both studies) or glatiramer acetate (CONFIRM) were re-randomized 1:1 to BG-12 240 mg BID or TID. Analyses were performed according to treatment groups defined by whether patients were continuing BG-12 treatment or new to BG-12 treatment in ENDORSE.

Results: This analysis included 1,960 patient-years of follow-up. In the continuing groups, the incidence of adverse events (AEs), serious AEs, and discontinuations due to AEs was 80–82%, 10–12%, and 2–4%, respectively. In groups new to BG-12, these incidences were 75–85%, 8–16%, and 10–22%, respectively. The most common AEs ($\geq 10\%$ in any group) were MS relapse and nasopharyngitis for the continuing groups, and flushing, MS relapse, nasopharyngitis, diarrhea, and upper abdominal pain for the newly treated groups. Serious infection incidence was $\leq 2\%$ in each group. There were 14 malignancies (13 patients), diverse in type/location: six malignancies in patients continuing BG-12 and eight malignancies in patients newly treated. There were three deaths, none considered related to BG-12.

Conclusion: No new or worsening safety signals were identified for patients continuing BG-12. The safety profile for patients newly treated in ENDORSE was consistent with that in DEFINE and CONFIRM.

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Abstract – WCN 2013

No: 2276

Topic: 6 – MS & Demyelinating Diseases

Neuroradiological efficacy of oral BG-12 for relapsing–remitting multiple sclerosis (RRMS): Integrated analysis of the Phase 3 DEFINE and CONFIRM studies

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Background: Oral BG-12 (dimethyl fumarate) demonstrated positive clinical and neuroradiological efficacy and an acceptable safety profile in the Phase 3 DEFINE and CONFIRM trials.

Objective: To present the results of a pre-specified, integrated analysis of DEFINE and CONFIRM, conducted to obtain a more precise estimate of the therapeutic effect of BG-12 on MRI endpoints.

Patients and methods: Eligible patients were aged 18–55 years and had a diagnosis of RRMS (McDonald criteria) and an Expanded Disability Status Scale score of 0–5.0. In DEFINE, patients were randomized 1:1:1 to receive BG-12 240 mg twice (BID) or three times daily (TID) or placebo. In CONFIRM, patients were randomized 1:1:1:1 to receive BG-12 240 mg BID or TID, placebo, or glatiramer acetate (reference comparator). MRI was performed in a subset of patients at sites with validated MRI capability.

Results: A total of 1,046 patients (MRI cohort) were randomized and received placebo (n = 347), BG-12 BID (n = 345), or BG-12 TID (n = 354). At 2 years, BG-12 BID and TID reduced the number of new/newly enlarging T2-hyperintense lesions by 78% and 73%, respectively; new non-enhancing T1-hypointense lesions by 65% and 64%, respectively; and the odds of having more gadolinium-enhancing lesions by 83% and 70%, respectively, as compared to placebo (all comparisons $p < 0.0001$).

Conclusion: The results of the integrated analysis demonstrate consistent benefits of both dosing regimens of BG-12 on MRI activity. Alongside strong clinical efficacy and an acceptable safety profile,

these results suggest that BG-12 has the potential to become a valuable oral treatment option for RRMS patients.

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Abstract – WCN 2013

No: 2316

Topic: 6 – MS & Demyelinating Diseases

BG-12 effects on quality of life in relapsing–remitting ms patients: Integrated analysis of the Phase 3 DEFINE and CONFIRM studies

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Background: Oral BG-12 (dimethyl fumarate) demonstrated clinical and neuroradiological efficacy and an acceptable safety profile in the Phase 3 DEFINE and CONFIRM studies.

Objective: To report the results of a pre-specified, integrated analysis of health-related quality of life (HRQoL) endpoints in DEFINE and CONFIRM.

Patients and methods: HRQoL endpoints assessed in both studies were the Short Form-36 (SF-36) Physical and Mental Component Summary (PCS/MCS) scales, global assessment of well-being visual analog scale (VAS), and the EuroQOL-5D (EQ-5D) VAS. Higher scores indicated better HRQoL.

Results: A total of 2,301 patients were randomized to receive placebo (n = 771) or BG-12 240 mg twice (BID; n = 769) or three times daily (TID; n = 761). Physical and mental health and functioning were significantly improved with BG-12 versus placebo. At 2 years, mean SF-36 PCS scores increased from baseline by 0.47 (BID) and 0.43 (TID) versus a reduction of -1.05 (placebo; both $p < 0.0001$). SF-36 MCS scores increased by 0.31 (BID) and 0.63 (TID) versus a reduction of -0.60 (placebo; $p = 0.0246$ and $p = 0.0107$, respectively). BG-12-treated patients reported a significantly better sense of well-being and perception of health status than placebo-treated patients. Mean changes from baseline to 2 years with BID and TID versus placebo were -0.3 and $+0.1$ versus -4.0 for global well-being VAS (both $p < 0.0001$) and -0.90 and -0.31 versus -3.37 for EQ-5D VAS ($p = 0.0011$ and $p = 0.0002$, respectively).

Conclusion: BG-12 treatment resulted in significant improvements in physical and mental aspects of health and functioning, general well-being, and overall health status compared with placebo in RRMS patients.

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Abstract – WCN 2013

No: 2304

Topic: 6 – MS & Demyelinating Diseases

Gastrointestinal tolerability events in relapsing–remitting multiple sclerosis patients treated with BG-12 (dimethyl fumarate): Integrated analysis of DEFINE and CONFIRM

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Background: Oral BG-12 (dimethyl fumarate) demonstrated an acceptable safety profile in the Phase 3 DEFINE and CONFIRM studies. Common adverse events (AEs) included flushing and gastrointestinal (GI) events.

Objective: To further characterize GI AEs in patients treated with BG-12 240 mg twice daily (BID) in DEFINE and CONFIRM.

Patients and methods: A post-hoc, integrated analysis of the incidence, nature, severity, and management of GI AEs in DEFINE and CONFIRM was conducted, focusing on the initial treatment period (through Month 3). Management of GI AEs was evaluated by manual review of concomitant medications temporally associated with and indicated for each GI event.

Results: A total of 1,540 patients in DEFINE and CONFIRM were randomized to receive placebo (n = 771) or BG-12 BID (n = 769). The most common (≥5% patients) GI AEs during the initial treatment period were abdominal/upper abdominal pain, nausea/vomiting, and diarrhea; of these events, 91%, 95%, and 96%, respectively, were mild or moderate in severity, and 38%, 33%, and 26% were treated with symptomatic therapies. Symptomatic therapies included omeprazole, paracetamol, ranitidine, metoclopramide, domperidone, and loperamide (efficacy not assessed). Few patients (<1% placebo, 3% BG-12) discontinued treatment due to GI events.

Conclusion: GI AEs associated with BG-12 during the initial treatment period generally are mild or moderate in severity and infrequently lead to treatment discontinuation. Further studies are needed to evaluate the efficacy of individual management strategies.

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Abstract – WCN 2013

No: 2685

Topic: 6 – MS & Demyelinating Diseases

Case-report: Scleroderma with longitudinally extensive myelitis

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Background: There is an intrinsic relationship between systemic autoimmune diseases and myelitis. Most of them are reported with acute transverse myelitis, some of them with longitudinally extensive myelitis (LTEM) and by now in the Neuromyelitis optica spectrum disorders (NOSD) Researching in major scientific directories there are no cases reporting the association between acute myelitis and Scleroderma.

Objective: Describe an unreported association with Scleroderma and LTEM.

Patients and methods: A 53 year old man was admitted with progressive weakness in the last 24 h. Symptoms began with sudden pain and leg paresthesia and evolved to a complete medullary syndrome with paraplegia, bladder dysfunction and a sensory impairment by the level of T8. He was previously diagnosed with Scleroderma four years ago. He presented by Raynaud phenomena, sclerodactyly and telangiectasia in the face, chest, and upper and lower limbs. He presented a positive nucleolar pattern ANA. Lab tests have shown elevated proteins in CSF and elevated CRP and ESR. A full spinal MRI has shown a contiguous T2 increased signal beginning

from the bottom of the medulla oblongata to the level of T8 confirmed in the transactional view.

Results: The patient was submitted to a five-day course of Methylprednisolone followed by a course of Intravenous Human Hyperimmune Gammaglobulin. The patient was discharged with prednisone, colchicin and chloroquine. He returned 27 days later fully walking, without bladder or sensory dysfunctions. Antiaquaporin-4 IgG autoantibody was negative.

Conclusion: This case reported the association between Scleroderma and LTEM in the NOSD.

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Abstract – WCN 2013

No: 2707

Topic: 6 – MS & Demyelinating Diseases

Neurological symptoms in demyelinating disease mimicking an acute ischaemic stroke—Case report

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Background: The case presented here emphasizes the role of an accurate diagnosis for proper targeting of therapy.

Objective: Demyelinating disease should be considered even in older patients with focal and fluctuating neurological symptoms.

Patients and methods: In March 2013, a 50-year-old woman was admitted with acute left hemiplegia and severe dysarthria, which occurred three days ago. Ten days earlier the patient had intense vertigo, nausea, vomiting, diplopiae, speech difficulty and unstable gait. Neurological status on admission showed severe dysarthria, bilateral rhythmic nystagmus, left hemiplegia, vivid generalized muscle tendon reflexes with asymmetry in the right and positive Babinski sign on the right. In the following days MRI of the brain and lumbar puncture were performed. MRI of the brain showed hypersignal changes in T2 and FLAIR images. CSF electrophoresis showed mild immune activity in the central nervous system. VEP showed a prolonged latency in P100 on the right eye.

Results: Therefore, disseminated demyelinating process was diagnosed and intravenous methylprednisolone treatment was initiated (1 g/day for 5 days) with a subsequent progressive improvement of the neurological symptoms.

Discussion: Our case has shown that sudden, rapid and unusual neurological deficits can be observed in multiple sclerosis (MS). In such cases, there is a wide range of diseases in the differential diagnosis, including ischemic cerebrovascular events. The presented case demonstrates rare Multiple Sclerosis cases with late onset of the disease by the age of 50. The correct diagnosis of these two entities is essential for proper treatment.

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Abstract – WCN 2013

No: 2349

Topic: 6 – MS & Demyelinating Diseases

Memory impairment in patients with clinically isolated syndrome

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Background: Multiple sclerosis (MS) is a chronic disease affecting young adults, manifesting beside motor and visual deficits also by cognitive impairment, including memory. Recent studies indicated that cognitive impairment can be present already in patients with clinically isolated syndrome (CIS), who are at higher risk of developing CDMS (clinically definite MS) but specific studies focused on memory in CIS patients are lacking.

Objective: To assess whether verbal and non-verbal memory recall and recognition are impaired in patients with CIS.

Patients and methods: 14 patients on interferon- β fulfilling criteria for CIS and 14 healthy matched controls underwent brain MRI (magnetic resonance imaging) and detailed neuropsychological testing including tests of verbal (Rey Auditory Verbal Learning Test – RAVLT) and non-verbal (Brief Visual Memory Test Revised – BVMT-R) memory.

Results: Patients with CIS and controls did not differ in basic demographic characteristics. Patients with CIS were impaired on RAVLT-trials 1–6 ($p = .03$) and delayed recall after 30 min ($p = .024$), but not in recognition ($p = .789$). Patients with CIS were impaired on BVMT-R-trials 1 to 3 ($p = .001$), and borderline on recall ($p = .057$), but not in recognition ($p = .327$).

Conclusion: Our results suggest that impairment of both verbal and nonverbal memory recall may be present already in patients with CIS. The memory recognition is spared. There is ongoing a follow-up study to determine whether patients with CIS and more pronounced memory impairment are more likely to develop CDMS and to find structural correlates of memory impairment in patients with CIS on brain MRI.

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Abstract – WCN 2013

No: 2634

Topic: 6 – MS & Demyelinating Diseases

Cortical activation changes following botulinum-toxin treatment of leg spasticity in multiple sclerosis: Pilot study

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Background: Botulinum neurotoxin (BoNT) treatment affects multiple levels of the sensorimotor system and can relieve spasticity of lower limbs caused by multiple sclerosis. The aim of our functional magnetic resonance study was to evaluate cortical activation changes following botulinum-toxin treatment of leg spasticity in multiple sclerosis.

Methodology: Four patients (1 man, 3 women, mean age 46.5, SD 9.3 years) with multiple sclerosis affected with leg spasticity were studied. Patients performed repeated knee extension–flexion movements during brain functional MRI which was acquired in three sessions: before and 4 and 12 weeks after BoNT treatment into the spastic muscles. The change of leg spasticity was assessed using the Snow scale.

Results: BoNT treatment decreased leg spasticity across the group. fMRI pre-BoNT treatment showed extensive bilateral task-related activation of frontoparietal sensorimotor cortical areas, whereas post-BoNT treatment caused retraction to midline and contralateral sensorimotor cortex. Third examination after 12 weeks of BoNT treatment showed re-expansion to a similar extent as seen in the pre-BoNT session.

Conclusions and relevance: This pilot study suggests that relief of leg spasticity may be associated with temporary partial normalization of activation in primary and association sensorimotor cortical areas. Spasticity may be contributing to the documented compensatory overactivation of the sensorimotor system in multiple sclerosis.

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Abstract – WCN 2013

No: 975

Topic: 6 – MS & Demyelinating Diseases

Guillain–Barre syndrome in systemic lupus erythematosus:

Case report

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Systemic lupus erythematosus (SLE) is a chronic, inflammatory and autoimmune disease affecting various organs and neurological systems. Although peripheral polyneuropathies can be seen in 10–15% of SLE patients, Guillain–Barre Syndrome (GBS) has been rarely reported in SLE and usually has very poor prognosis. In this case report, a 32-year-old man who followed up with SLE and diagnosed GBS will be discussed.

The patient had numbness and weakness in his distal lower limbs around two weeks after his nausea, vomiting and cough symptoms. His neurological examination showed cranial and peripheral polyneuropathy findings. In his cerebrospinal fluid examination, there was no cell and the amount of protein was high (173 mg/dl). EMG examination showed mixed-type polyneuropathy findings that were affecting motor fibers. Antibody of GQ1b was (–). Intravenous immunoglobulin (IVIG) treatment was begun with 0.4 g/kg/day dosage. After 3 days of treatment, the patient got worse and was taken to the intensive care unit and intubated due to bulbar and respiratory problems. Treatment continued with pulse steroids for 5 days (1 g/day), 3 sessions of plasmapheresis and 2 sessions of cascade filtration and oral methylprednisolone therapy was started. Gaze palsy improved and tetraparesis began to decline after 3 months in the intensive care unit and the patient was extubated. Our case will be discussed because of association of SLE and GBS. This association rarely occurs and usually results in mortality.

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Abstract – WCN 2013

No: 2262

Topic: 6 – MS & Demyelinating Diseases

Prognostic value of magnetization transfer parameters in clinically isolated syndromes (CIS)

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Background: Magnetization transfer is a quantitative imaging technique which can assess tissue integrity. A clinically isolated syndrome (CIS) is the initial presentation form in multiple sclerosis (MS) patients.

Objective: To evaluate the use of magnetization transfer (MT) MRI in clinically isolated syndromes (CIS) in order to know the prognosis of the disease course.

To compare the prognostic value of MT parameters with conventional MRI markers like black holes and infratentorial lesions.

Methods: Patients with clinically isolated syndrome (CIS) were followed for 3 years.

Complete neurological explorations were performed every six months. 3 T MRI was performed at the diagnosis and after three years.

MTR histograms of whole brain, normal appearing white matter (WM) and grey matter (GM) were produced and calculated using JIM (Xinapse).

Results: 17 patients were examined (9 females). The baseline mean EDSS was 1.68 and 1.15 after three years. Only 8 patients had black holes during the study.

A significant correlation was found between baseline peak WM MTR at baseline and EDSS at the end of the study ($r: -0.82, p: 0.001$). The correlation between average MTR values at baseline and EDSS at the end was moderate ($r: -0.68, p: 0.01$). The EDSS at three years was also associated with the number of infratentorial lesions at baseline ($r: 0.75, p: 0.002$) and the black holes volume ($r: 0.82, p < 0.001$).

Conclusion: Our data suggest that MTR parameters at baseline are associated with disability progression. Black holes and infratentorial lesions are good markers for disease progression but they are not always present at the beginning.

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Abstract – WCN 2013

No: 1663

Topic: 6 – MS & Demyelinating Diseases

CNS demyelination associated with TNF α antagonists:

A pathologically proven case with an unusual presentation

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Introduction: Biological agents have revolutionized the treatment of several inflammatory diseases with a growing number of reports of the paradoxical induction of autoimmune reactions, including CNS demyelination. On the other hand, monoclonal antibodies have been linked to the development of progressive multifocal leukoencephalopathy (PML).

Case report: A 46-year-old man presented with right arm paresis and hypoesthesia progressing over 1 month. He had a medical history of seronegative spondylarthropathy, treated with TNF α antagonists (sequentially Etanercept, Adalimumab and Golimumab). The brain MRI showed a lesion involving the left periorlandic white-matter, with T2-hyperintensity, no mass effect, mild gadolinium enhancement and heterogeneous restriction on diffusion, and a smaller lesion on the right hemisphere. A PML was considered and Golimumab treatment was interrupted but there was clinical aggravation during the next 2 months. CSF cytochemistry was normal but oligoclonal bands were detected. JC virus DNA was not found on PCR analysis of CSF and urine. A brain biopsy of the main lesion was subsequently performed and showed white matter demyelination with macrophage infiltration, thickened vessels surrounded by T-lymphocyte infiltration and astrocyte hyperplasia, without viral inclusions. PCR analysis for JCV in brain tissue was negative. The diagnosis of CNS demyelination associated with TNF α antagonists was considered and the patient was treated with high dose corticosteroids followed by slow oral tapering with clinical and radiological amelioration and subsequent stabilization.

Conclusion: CNS demyelination associated with TNF α antagonists has a heterogeneous clinical and radiological presentation. Our case discloses a clinical picture that may simulate a PML.

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Abstract – WCN 2013

No: 2391

Topic: 6 – MS & Demyelinating Diseases

Clinical and epidemiological profile of Guillain–Barre syndrome in Tunisia

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Background: Guillain–Barre syndrome (GBS) is the most frequent cause of acute flaccid paralysis. The idiopathic form results from an immunological disorder usually following a recent infectious episode. Electrophysiological examination is useful to confirm the diagnosis and to determine the neuropathy type.

Objective: The aim of this study was to determine the clinical and epidemiological characteristics of GBS in Tunisia.

Patients and methods: We conducted a retrospective study using population-based information from medical records of 200 patients with GBS, hospitalized in the neurology department at the National Institute of Neurology in Tunis over a 10 year period.

Results: 194 patients showed typical GBS, 6 patients presented a Miller Fisher variant.

Mean age was 41 years. There were 28 children and 172 adults with a male predominance (130 men). The prevalence was highest in March and lowest in June.

The demyelinating form was diagnosed in 119 patients, axonal form in 30 patients and mixed form in 16 patients. CSF studies showed raised protein levels with no cells in 170 patients.

Intravenous immunoglobulin cure or plasma exchange was administered to 109 patients. All patients had vitamin therapy and motor rehabilitation.

A stay in the intensive care unit was necessary for 41 patients. 136 patients had improvement, 46 patients maintained a steady state. 11 patients worsened and 7 patients died.

Conclusions: The demyelinating pattern was the major electrophysiological subtype in our study. The clinical setting and determination of GBS pattern through electrophysiological examination are required to optimize therapeutic management.

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Abstract – WCN 2013

No: 2365

Topic: 6 – MS & Demyelinating Diseases

Mitochondrial dysfunction at the onset of neurological deficits in a neuroinflammatory model of MS studied using *in vivo* imaging

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Reversible neurological deficits in multiple sclerosis (MS) have been attributed to demyelination. Increasing evidence now implicates inflammation as a key player, but mechanisms remain unknown.

Objective: To assess the potential role of mitochondria in causing temporary loss of function in a model of MS.

Methods: Experimental autoimmune encephalomyelitis (EAE) was induced in adult female mice transgenic for expression of cyan fluorescent protein in axonal mitochondria. The spinal cord of symptomatic and asymptomatic animals was imaged at the onset of neurological deficit, or 6 weeks post-immunization. Fluorescent potentiometric dye (TMRM) was applied to the exposed spinal cord to reveal mitochondrial function.

Results: At the onset of neurological deficits, symptomatic animals were distinguished from time-matched asymptomatic and naive controls by fragmentation and loss of function of axonal mitochondria. The number of small mitochondria (1.5–3 μm) increased by ~54% ($p < 0.01$), and longer mitochondria (6–9 μm) decreased by ~49% ($p < 0.001$) compared with controls. 40% of the total mitochondrial mass was non-functional in symptomatic animals, vs. ~28% of mass in asymptomatics ($p < 0.01$), vs 0% in naives. Mitochondrial defects were mainly observed in areas infiltrated with T-cells and macrophages that were producing NO *in vivo*. Additionally, in inflamed areas activated astrocytes showed increased

glycolysis (phosphofructokinase-2.3+). In chronic animals with severe deficits ~51% of the mitochondrial mass was non-functional, although the inflammation had subsided.

Conclusions: Mitochondrial bioenergetic failure is implicated as a cause of neurological deficits at the onset of EAE, in association with increased glycolysis in astrocytes consistent with a mitochondrial energy insufficiency.

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Abstract – WCN 2013

No: 2572

Topic: 6 – MS & Demyelinating Diseases

Demyelinating disease affecting both the central nervous system (CNS) and peripheral nervous system (PNS): Study of 2 cases

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Background: Chronic inflammatory demyelinating polyneuropathy (CIDP) is a condition affecting the peripheral nervous system; however, it has been associated with central nervous system (CNS) involvement in rare conditions.

Methods: We report on two patients with a demyelinating disease affecting the central nervous system (CNS) and peripheral nervous system (PNS).

Results: The first case was 50-year-old man who presented a relapsing-remitting course in which CNS involvement preceded PNS involvement. He begun in 1985 to experience paresthesia in the four limbs followed 6 months later by left optic neuritis. The clinical examination then found pyramidal signs. He improved after steroid treatment. Since 1993, signs of peripheral nerve impairment appeared with repeated episodes of paresthesia, and asymmetric sensory motor signs predominating in distal segments. Clinical examination found areflexia predominating in the lower limbs. Nerve conduction study showed demyelinating motor and sensory neuropathy with persistent conduction blocks. CSF analysis showed albuminocytologic dissociation.

The second case was a 56-year-old man presenting since 2003 a CIDP with conduction block asymmetry predominant in the lower limbs. But the clinical examination found brisk reflexes. MRI examination documented few T2 high signals in brain white matter. No systemic inflammatory disease and no metabolic or inflammatory factor for peripheral neuropathy were found. An improvement in clinical status and neurophysiological parameters was observed after treatment with steroids and azathioprine (Imurel).

Conclusion: CIDP with conduction blocks may rarely be associated with central demyelination lesions. Our 2 cases are compared to rare similar ones in the literature and the favourable effect of steroid treatment is emphasized.

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Abstract – WCN 2013

No: 2256

Topic: 6 – MS & Demyelinating Diseases

Location and volume of multiple sclerosis lesions predict enhanced or decreased female sexual function

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Background: In Multiple Sclerosis, female sexual function (FSF) might change with affliction of brain areas involved in sexual function.

Objective: To evaluate correlations between FSF and site and size of MS-related magnetic resonance imaging (MRI) changes.

Methods: In 50 women with MS (37.0 ± 9.9 years), we determined cerebral and spinal MS-lesion-load and -location in T2-weighted 1.5 T MRIs, assessed the 19-item FSF-Index (FSFI) scores evaluating desire, arousal, lubrication, orgasm, satisfaction, pain (Rosen et al. *J Sex Marital Ther.* 2000;26:191–208), and calculated Spearman-rank correlations between FSFI-scores and MS-lesion-load in frontal, parietal, temporal, occipital, opercular, cingulate, thalamic, midbrain, cerebellar, pontine, medulla oblongata, and spinal cord areas (significance: $p < 0.05$).

Results: Direct correlations were found for FSFI-pain-scores with right prefrontal/orbitofrontal, primary-motor, parietal-cortical and bilateral temporal lesion-volumes, for FSFI-desire-scores with left frontal-premotor and thalamic lesion-volumes, and for FSFI-arousal-scores with right frontal primary-motor cortex and midbrain lesion-volumes. FSFI-lubrication-scores correlated inversely with right occipital middle/inferior gyral lesion-volumes, but directly with right prefrontal/orbitofrontal cortex, temporal, midbrain and left pontine lesion-volumes. FSFI-orgasm-scores correlated inversely with left temporal, and right occipital lesion-volumes, but directly with right frontal primary motor cortex, bilateral temporal, and midbrain lesion-volumes. FSFI-satisfaction-scores correlated inversely with occipital and directly with right frontal primary-motor lesion-volumes.

Conclusions: Surprisingly, intercourse-related pain decreased while desire and arousal increased in women with higher MS-lesion-load. In contrast, location and size of lesions determined whether there was improvement or deterioration of lubrication, orgasm, and satisfaction.

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Abstract – WCN 2013

No: 1545

Topic: 6 – MS & Demyelinating Diseases

IL-6, AQP4 and CX43 modulations by estrogen in a model of astrocyte-microglia: Insight to the NMO and pregnancy outcome

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Background: Neuromyelitis optica (NMO) is more frequent in women than men (9:1). Astrocytes, the main target in NMO, express Aquaporin-4 (AQP4) (with major role in NMO pathology) and Connexin43 (Cx43) (with unknown but probable impact on brain diseases). Recently, monoclonal antibody blocking IL-6 receptor was successfully applied to treat severe cases of NMO.

Objective: To investigate the role of 17-beta Estradiol (Es) on Cx43, AQP4 and IL-6 on a cultured model of astrocytes-microglia.

Material and methods: Primary astrocytes and microglia were prepared from postnatal Wistar rats (P0-P2). Cultures with about 5% (M5) or 30% (M30) of microglia were subjected to non-inflammatory and inflammatory condition and they were treated with 100 nM Es for 24 h. Cultures were evaluated for the expression of AQP4 and Cx43 expression by Western blotting (WB) and IL-6

ELISA. All experiments were performed at least four times from different primary cultures. Data were analyzed with GraphPad Prism using *t*-test (mean \pm SEM) and were considered significant with P_{value} of <0.05 .

Results: Cx43 ($73.49\% \pm 5.258$, $P_{\text{value}} = 0.009$) and AQP4 ($83.42\% \pm 3.27$, $P_{\text{value}} = 0.001$) were decreased in M5 conditions, while they were increased in M30: Cx43 ($122\% \pm 4.97$, $P_{\text{value}} = 0.0005$), AQP4 ($166\% \pm 27.15$, $P_{\text{value}} = 0.02$). IL-6 was decreased in M30 cultures ($P_{\text{value}} = 0.02$) but not in M5 cultures.

Discussion: We showed that, Es overexpressed AQP4 and Cx43 and decreased IL-6 in M30 cultures. Our *in vitro* study is in line with recent study in humans and supports the idea that pregnancy can worsen NMO. However, there is a need to understand the anti/pro inflammatory role of IL-6 in NMO with further *in vivo* experiments.

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Abstract – WCN 2013

No: 2029

Topic: 6 – MS & Demyelinating Diseases

Predictive factors of early visual recovery in patients with optic neuritis

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Background: Optic neuritis is an acute inflammatory disease of the optic nerve. Abrupt visual loss followed by spontaneous improvement over several weeks is a typical course of the disease. The Optic Neuritis Treatment Trial demonstrated the effect of intravenous corticosteroid. However, 10 to 15% of patients experience poor or slow recovery of vision.

Objectives: To elucidate predictive factors of early visual recovery in patients with optic neuritis treated by intravenous corticosteroid.

Patients and methods: Thirty-four consecutive patients with acute optic neuritis received intravenous corticosteroid were enrolled. According to the improvement of visual acuity during first 7 days, patients were divided into 2 groups – good response group ($n = 21$); Snellen visual acuities had more than 2 line increases at day 15, and poor response group ($n = 13$). Demographic, clinical, laboratory and radiological features were compared between the groups.

Results: The duration of symptom was numerically shorter in the good response group (12.5 ± 12.9 vs. 23.7 ± 48.7 days, $p = 0.433$) but not statistically significant. Demographic, clinical, and laboratory features did not show a significant difference between the groups. The optic nerve enhancement on orbit MRI was more frequent in the good response group (12 (57.1%) vs. 1 (7.7%), $p = 0.005$).

Conclusion: The effect of corticosteroid is the result of anti-inflammatory action. Gadolinium-enhancement of the optic nerve represents the degree of inflammation. The optic nerve enhancement on orbit MRI could be used as a predictive factor on early visual recovery in optic neuritis treated with intravenous corticosteroid.

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Abstract – WCN 2013

No: 2443

Topic: 6 – MS & Demyelinating Diseases

FAMVIR significantly improves the curative effect of the Normoxic compression in chamber in progressive multiple sclerosis

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Progressive MS is always accompanied by marked axonal injury with significantly reduced tissue respiration in the brain. A new protocol

for the therapeutic use of pressure, the so-called Normoxic Curative Compression (NCC), allows the recovery of the tissue respiration in the brain in stroke, trauma and neurodegeneration.

The aim of the study is to analyze the efficacy of Famvir in combination with NCC in progressive MS.

Materials and methods: In a main group comprising 81 patients with progressive MS, the patients received a course of NCC (1.1 ATA in air chamber for 20 min) in combination with Famvir. In control groups patients with progressive MS received either only Famvir or only NCC. The complex assessment of the patients included a dynamic of immune status, EEG, and acid–base blood condition.

Results: Maximum clinical effect was observed in the main group of patients who received combined therapy including anti-viral therapy, NCC and a short course of Dexason. A therapeutic effect was observed in reduced degree of disability, regression of neurological symptoms and changes in the course of MS. Only the main group of patients had a statistically significant decrease in the amount of B lymphocytes and apoptosis. The control group treated with Farmavir did not demonstrate a noticeable effect.

Discussion: The study confirms the viral nature of MS and demonstrates the necessity of complex therapy for MS to recover the tissue respiration in the brain and microcirculation. The use of the developed complex allows conversion of progressive MS into the remitting form of MS.

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Abstract – WCN 2013

No: 2551

Topic: 6 – MS & Demyelinating Diseases

Compensatory increase in mitochondrial trafficking induced by focal mitochondrial damage results in selective block of transport in small diameter axons

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Background: The role of impaired mitochondrial trafficking is increasingly recognised in the pathogenesis of peripheral neuropathies, but their behaviour *in vivo* is very poorly understood.

Objective: To study the effect of inflammation on mitochondrial function and transport, *in vivo*.

Material and methods: Mitochondria were observed by time lapse confocal imaging in the exposed saphenous nerves of anaesthetised mice with experimental autoimmune neuritis (EAN).

Results: The number of mobile mitochondria was significantly lower in animals with EAN compared with adjuvant controls or asymptomatic animals. At the onset of EAN, but not in control animals, we observed a number of small to medium diameter fibres ($3.34 \pm 0.61 \mu\text{m}$) containing focal accumulations of stationary mitochondria. Time-matched, asymptomatic animals with EAN showed few, if any such accumulations, but we were able faithfully to reproduce the focal accumulations by laser damaging (photo-bleaching) the mitochondria in these animals. The damaged mitochondria became depolarised, fragmented and stationary, presumably depleting the energy supply of the affected portion of the axons. Interestingly, the more proximal mitochondria in all photo-bleached axons started to move towards the damaged region in significantly increased numbers than before photo-bleaching ($p = 0.007$). In the larger axons alone the mitochondria passed unobstructed into the damaged field, repopulating it with healthy mitochondria. However, in smaller axons ($2.7 \pm 0.45 \mu\text{m}$), the arriving mitochondria stopped moving, seemingly obstructed by the damaged mitochondria.

Conclusion: Dysfunctional mitochondria obstruct axonal transport machinery resulting in failure of mitochondria to re-populate small

axons and thus rendering them more vulnerable to degeneration in some peripheral neuropathies.

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Abstract – WCN 2013

No: 2333

Topic: 6 – MS & Demyelinating Diseases

Endothelial dysfunction in patients with multiple sclerosis

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Background: The vascular component plays an important role in the pathogenesis of multiple sclerosis (MS). Few studies confirm the presence of endothelial dysfunction (ED) in MS and its role in the pathogenesis of the disease.

Objective: Identification of ED in patients with MS, an assessment of its relation with the course and disease modifying therapy (DMT) of MS.

Patients: 32 patients (22 female, 10 male), age 34.7 (± 2.03), with definite diagnosis of MS (McDonald criteria, 2005), disease duration – 8.1 (± 1.5) years (84% – relapsing–remitting MS (RRMS), 16% – secondary–progressive MS (SPMS)). 43.75% was naïve for DMT, 56.25% use DMT for 3.25 (± 0.6) years.

Methods: Blood level of von Willebrand factor (vWf) ELISA, statistical analysis – nonparametric statistic – φ^* Fisher.

Results: vWf values ranged from 0.6 to 2 U/mL (normal 0.5–1.5 U/mL), and was an average of 1.42 \pm 0.08 U/ml. 46.8% has above normal level of vWf – 25% without DMT, 15.5% with short term of DMT, 6.3% with long term of DMT and SPMS. In 15.6% the value of vWf was at the upper limit of normal (patients without DMT and patients with SPMS). Level of vWf in patients with RMS without DMT or with short term of DMT was significantly higher than in patients receiving DMT for more than 1 year ($\varphi^* = 2.48$, $p \leq 0.01$).

Conclusion: A significant proportion of patients with MS have ED. vWf depends on received DMT, and can be one of the predictors of the effectiveness of the therapy.

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Abstract – WCN 2013

No: 2539

Topic: 6 – MS & Demyelinating Diseases

Longitudinal assessment of the multiple sclerosis impact scale (MSIS-29) amongst A treated relapsing remitting multiple sclerosis cohort

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Background: The Multiple Sclerosis Impact Scale (MSIS-29) is a commonly used outcome measure of the physical and psychological impact of multiple sclerosis (MS) however long-term reliability has yet to be established.

Objective: To determine reliability of the MSIS-29 longitudinally in a treated cohort of relapsing remitting MS patients.

Patients and methods: A cohort of patients diagnosed with highly active RRMS on natalizumab were assessed every 6 months for 3 years. All patients completed MSIS-29 and Beck Depression Inventory (BDI). Physical assessment was measured using the Expanded Disability Status Scale (EDSS). Correlation between variables was investigated using the Spearman rho correlation coefficient and $p < 0.05$ was considered statistically significant. Only complete paired data sets were analysed.

Results: Comparison between the BDI and MSIS-29 psychological and EDSS and MSIS-29 physical showed strong correlation at 0 (n = 96), 12 (n = 78), 24 (n = 54), 36 (n = 30) months. The MSIS-29 physical score showed appropriate responsiveness when EDSS improved, remained stable or disimproved from time 0 to 36 months. Good correlation ($r = 0.564$, $p < 0.02$) was seen in change between the BDI and MSIS-29 psychological between 0 and 36 months.

Conclusion: The results demonstrate correlation between BDI and MSIS-29 psychological over time. EDSS and MSIS-29 physical scores correlate strongly at set time points and show a trend to longitudinal responsiveness. These findings support the longitudinal reliability of MSIS-29 as an outcome measure for MS patients.

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Abstract – WCN 2013

No: 1184

Topic: 6 – MS & Demyelinating Diseases

Adverse event profile of alemtuzumab over time in treatment-naïve patients with early, active relapsing–remitting multiple sclerosis (RRMS; CARE-MS I study)

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Background: In CARE-MS I, alemtuzumab reduced relapse rate by 55% vs. subcutaneous interferon-1a (SC IFNB-1a) ($p < 0.0001$) over 2 years in treatment-naïve patients with early, active RRMS.

Objective: Describe adverse events (AE) of alemtuzumab over time in CARE-MS I.

Patients and methods: Patients were randomized 2:1 to two annual courses of alemtuzumab 12 mg/day IV (Month 0: 5-day course; Month 12: 3-day course) or SC IFNB-1a 44 μ g 3 \times /week. Safety measures included monthly AE monitoring, clinical/laboratory measures, and patient/investigator education about autoimmunity.

Results: Of 563 patients, >90% per group experienced AEs, most of mild/moderate severity; few AE-related study discontinuations occurred (alemtuzumab, 0%; SC IFNB-1a, 2.7%). AEs were most frequent with alemtuzumab during Months 0 and 12, coinciding with treatment administration, largely attributable to infusion-associated reactions (IARs: Month 1, 85.9%; Month 13, 65.7%; non-IAR, Month 1, 44.7% vs. 60.4% SC IFNB-1a; Month 13, 29.3% vs. 30.9% SC IFNB-1a). Secondary autoimmunity increased over time after alemtuzumab and primarily comprised thyroid disorders (Year 1, 6.9%; Year 2, 13.8%); immune thrombocytopenia was uncommon (Year 1, 0.3%; Year 2, 0.8%). Infections decreased from Year 1 (56.6%) to Year 2 (47.3%); serious infections were uncommon (Year 1, 1.6%; Year 2, 0.3%). Two occult malignancies (thyroid cancer) were identified in alemtuzumab patients 11 months after Course 1 and 10 months after Course 2.

Conclusion: AE incidence was similar between groups, except in proximity to alemtuzumab administration, when excess AEs for alemtuzumab were mostly attributable to IARs. Increased incidence of secondary autoimmunity in Year 2 is consistent with previous studies.

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No: 1049

Topic: 6 – MS & Demyelinating Diseases

Alemtuzumab improves visual outcomes in treatment-naïve patients with relapsing–remitting multiple sclerosis (RRMS): Analysis from the phase 3 CARE-MS I study

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Background: Alemtuzumab demonstrated clinical efficacy in the treatment of RRMS. Sloan low-contrast letter acuity can identify MS-related visual dysfunction.

Objective: Compare alemtuzumab and subcutaneous interferon beta-1a (SC IFNB-1a) effects on visual outcomes in CARE-MS I.

Patients and methods: Patients (N = 581) were randomized to alemtuzumab 12 mg/day IV on 5 consecutive days initially and 3 consecutive days 12 months later or SC IFNB-1a 44 µg 3×/week. MS Functional Composite (MSFC; disability composite) and binocular Sloan (letter acuity) assessments were performed twice yearly, with combined scores creating a normalized 4-dimensional composite (MSFC + Sloan) that incorporates visual impairments in the overall disability assessment. Worsened or improved visual function was defined as ≥7-point decreases or increases, respectively, from baseline in Sloan scores, and ≥0.5-point changes in MSFC + Sloan scores.

Results: Mean changes in low-contrast (2.5%) Sloan scores from baseline were smaller with alemtuzumab vs. SC IFNB-1a (significantly at 12 months, $p = 0.0019$; non-significantly at 24 months, $p = 0.1205$). When Sloan 2.5% was added to MSFC, treatment differences significantly favored alemtuzumab at 12 ($p = 0.0001$) and 24 months ($p = 0.009$). Proportions of patients with worsened or improved Sloan 2.5% scores significantly favored alemtuzumab vs. SC IFNB-1a (worsened: 15.3% vs. 23.6%; improved: 18.5% vs. 13.4% at 12 months; $p = 0.029$). At 24 months, results again numerically favored alemtuzumab in terms of the proportion who worsened or remained stable, although not significantly ($p = 0.2832$).

Conclusion: Low-contrast letter acuity by Sloan charts favored alemtuzumab. Use of a visual component measure is likely to augment ability to detect treatment effects across a variety of clinical dimensions in MS trials.

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Abstract – WCN 2013

No: 2509

Topic: 6 – MS & Demyelinating Diseases Employment in multiple sclerosis

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Introduction: Multiple sclerosis (MS) is a chronic neurological condition affecting young adults during critical years of work life. Mobility-related symptoms, arm and hand difficulties could adversely affect independence, employment and quality of life. This study determined variables associated with unemployment and risk factors for the development of unemployment in people with MS.

Materials and methods: A case–control study was performed on patients followed by the Department of Neurology, Hospital Sahloul of Sousse. Demographic, medical and professional data were gathered using a questionnaire.

Results: A total of 45 patients were included in the study: 15 were working (group T1), 16 lost employment (group T2) and 14 had never worked (group T3). Hence, the employment rate was 33%, with an average time since disease onset of seven years. Lower educational level, disease progression, the presence of motor symptoms, and a worse EDSS were found to be negative factors. For group T2, the loss of the job is related to MS in 87.5% of patients. For group T3, disability represents 30.7% and the lack of suitable employment is 46.1%.

Discussion: In a recently published meta-analysis the mean unemployment rate in MS was 59%. Similar to many studies, lower educational attainment and the presence of motor symptoms predict the loss of employment. The progressive form predisposes also to employment loss. Other studies proved the role of a high EDSS scale as a predictive factor for unemployment which is consistent with our obtained results. Other reasons reported by people with MS for their loss of employment were workplace-related factors including insufficient flexibility of employment conditions.

Conclusion: These data suggest that low educational level, progressive form, high EDSS and presence of motor symptoms predict employment loss.

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Abstract – WCN 2013

No: 1174

Topic: 6 – MS & Demyelinating Diseases

Alemtuzumab treatment has no adverse impact on sperm quality, quantity, or motility: A CARE-MS substudy

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Background: Alemtuzumab is a humanized monoclonal antibody shown to reduce relapse rate and disability in patients with relapsing–remitting multiple sclerosis (RRMS) by selectively targeting CD52, a protein highly expressed on B and T lymphocytes. The male reproductive tract and mature spermatozoa express an isoform of CD52 and may be a target of alemtuzumab.

Objective: To evaluate the potential effects of alemtuzumab treatment on human sperm concentration, motility and morphology.

Patients and methods: CARE-MS phase 3 study patients received intravenous alemtuzumab 12 mg or 24 mg on 5 consecutive days at entry and on 3 consecutive days 12 months later. Semen substudy enrolment was optional. Samples were collected at baseline prior to treatment and at post-treatment Months 1, 3, and 6. Patients entering the substudy after initiating therapy provided samples prior to treatment at Month 12 (second course) and at Months 13, 15, and 18. Sperm count, motility, morphology, and anti-sperm antibodies were assessed in 2 ejaculate samples, separated by at least 48 h, at each time point.

Results: Of 13 alemtuzumab-treated patients in this substudy (12 mg, n = 10; 24 mg, n = 3), all had sperm concentration, percent motile sperm, and percent normal sperm within or above the range of baseline or normal values in ≥ 1 sample at each post-treatment time point. A sperm-binding antibody, likely alemtuzumab, was detected in 1 patient at Month 1 only, without adverse impact on subsequent sperm parameters.

Conclusions: These limited data suggest that alemtuzumab treatment has no adverse impact on sperm quality, quantity, or motility.

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Abstract – WCN 2013

No: 1783

Topic: 6 – MS & Demyelinating Diseases

Neurological manifestations revealing Gougerot Sjogren syndrome: 18 cases

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Introduction: Neurological manifestations in Gougerot Sjogren syndrome (GSS) are valued differently and are sometimes in front of the dry syndrome.

Purpose: To analyze the epidemiologic and clinical aspects of 18 patients having SGS revealed by neurological manifestations.

Patients and methods: We report 18 cases of neurological manifestations revealing SGS collected over a period of 8 years.

Results: There were 18 patients with a sex ratio 15 W/3 M whose average age was 44.14 years old. Peripheral nervous system manifestation occurred in 44.44% (axonal polyneuropathy in 37.5%, trigeminal and other cranial nerves neuropathies in 37.5%, multiple mononeuritis in 12.5% and anterior horn involvement in 12.5%). Central nervous system involvement was observed in 88.88% (chronic myelopathy, multiple sclerosis like manifestations, psychiatric manifestations).

All the patients received oral corticosteroids, cyclophosphamide was used in 5 cases. Evolution was good in 9 cases.

Discussion and conclusions: Neurological affection in the SGS is noticed in 8 to 70% of the cases according to studies. It is marked by clinical diversity dominated by peripheral neuropathies. The central affection is usually polymorphic. In our series, results were different. Isolated central affection was noted in 11 cases versus isolated peripheral affection in one case and was mixed in 7 cases.

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Abstract – WCN 2013

No: 190

Topic: 6 – MS & Demyelinating Diseases

Effect of teriflunomide on lymphocyte and neutrophil counts in patients with relapsing multiple sclerosis: Results from the Tower Study

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Background: Teriflunomide, a once-daily oral immunomodulatory therapy approved in the USA, Australia and Argentina for RMS treatment, selectively and reversibly inhibits dihydroorotate dehydrogenase, a key mitochondrial enzyme in de novo pyrimidine synthesis required by rapidly dividing B and T cells. Teriflunomide has the potential to limit over-activation of immune responses contributing to MS disease activity.

Objective: To assess lymphocyte and neutrophil counts in TOWER (NCT00751881).

Patients/Methods: Patients with RMS received once-daily placebo (n = 388), teriflunomide 7 mg (n = 407) or 14 mg (n = 370). Blood samples were collected at randomization, every 2 weeks for 24 weeks, then every 4 weeks, and at treatment end.

Results: Mean baseline lymphocyte and neutrophil counts were similar across groups. At Week 48, mean decreases were observed with a small dose response (mean change from baseline [CfB; SD] lymphocyte count: placebo, 0.02 [0.48] $\times 10^9/L$; 7 mg, $-0.19 [0.45] \times 10^9/L$; 14 mg, $-0.30 [0.45] \times 10^9/L$; mean CfB [SD] neutrophil count: placebo, $-0.03 [1.76] \times 10^9/L$; 7 mg, $-0.43 [1.58] \times 10^9/L$; 14 mg, $-0.50 [1.81] \times 10^9/L$). Magnitude of change was $\leq 15\%$ during the first 6 weeks and stabilized over time on treatment. The protocol specified that patients with neutrophils $< 1 \times 10^9/L$ were to permanently discontinue treatment. Discontinuations due to neutrophil count decrease and neutropenia AEs were reported in 1 (placebo), 5 (7 mg) and 9 (14 mg) patients. No cases of febrile neutropenia were reported and there was no evidence of link between neutrophil and lymphocyte count decreases and infection.

Conclusions: These data provide evidence that, despite small effects on lymphocytes and neutrophils, teriflunomide is an immunomodulator preserving protective immunity and maintaining immune surveillance.

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Abstract – WCN 2013

No: 1092

Topic: 6 – MS & Demyelinating Diseases

The impact of spasticity severity on healthcare utilization among MS patients: A large-scale six-year follow-up study

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Background: Over 80% of multiple sclerosis (MS) patients ultimately experience spasticity. Spasticity is associated with fatigue, pain,

impaired mobility and lower quality of life. The impact of spasticity on healthcare utilization (HCU) is poorly understood.

Objective: Assess association of spasticity and HCU among North American Research Committee on Multiple Sclerosis (NARCOMS) registry participants.

Patients and methods: NARCOMS maintains a self-report database of MS patients who volunteer health-related information through online or paper questionnaires at enrollment and semi-annually thereafter. Among 5172 individuals who completed surveys in 2005 and 2011, we evaluated the association of HCU with spasticity reported using the 5-point Performance Scales spasticity subscale (PSs).

Results: Responders were predominantly female (76.2%) and white (96.2%). In 2005 mean (SD) age was 52 (9.8) years, disease duration 13.7 (9.1) years, and 50.8% could walk unassisted. 18.8% reported no spasticity, 36.2% minimal, 17.5% mild, 15.7% moderate, 9.9% severe and 1.9% total. In the prior 6 months, 653 (13.2%) reported an ER visit while 358 (7.1%) reported hospitalization. Frequencies for both increased with spasticity severity ($p < 0.001$ for linear trend). Over 6 years, PSs improved or remained stable in 74.9% and worsened in 25.9%. Versus participants whose PSs score worsened, those with improved spasticity were less likely to report an ER visit (OR 0.83; 95%CI: 0.67–1.03) or hospitalization (OR 0.76; 95%CI: 0.59–0.99) even after accounting for disability and sociodemographic factors.

Conclusion: More severe spasticity is associated with greater HCU, emphasizing the adverse impact of spasticity and suggesting that effective treatment of spasticity might reduce HCU.

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Abstract — WCN 2013

No: 2462

Topic: 6 — MS & Demyelinating Diseases

Absence of delayed facilitation and post-exercise facilitation at early stage of multiple sclerosis: Evidence of altered cortical plasticity?

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Background: By means of transcranial magnetic stimulation (TMS) multiple sclerosis (MS) has been studied. Disabilities correlate with alterations of TMS parameters such as prolonged central motor conduction time, prolonged motor evoked potential (MEP) latencies and increased motor threshold (MT). TMS has also been used to explore movement-related cortical plasticity.

Objective: To compare cortical excitability in patients at early stage of MS versus healthy controls.

Material and methods: In each subject TMS of non-dominant hemisphere was used to define MEP amplitude and motor threshold (MT). Then subjects performed 3 blocks (30-s, 60-s, and 90-s duration) of a bimanual motor task (exercise condition). Amplitudes of MEPs elicited immediately after each block, and then after a 15-minute rest period were compared with baseline to evaluate the presence of post-exercise facilitation and delayed facilitation. Patients were not receiving any therapy and their Expanded Disability Status Scale score was between 1 and 2.5.

Results: MT resulted significantly higher in patients compared to controls. Compared with baseline, controls had larger MEP amplitudes after 30 and 60 s of exercise (post-exercise facilitation) and also after the rest period (delayed facilitation). In contrast, MEP amplitudes in patients were not significantly different from baseline after any of the exercise conditions or following the rest period.

Conclusion: Patients do not show the normal fluctuations of cortical excitability usually found after a bimanual finger motor task. These

results suggest an alteration in the cortical plasticity. We speculate that in SM patients capability to recover from functional impairments caused by demyelination is compromised.

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Abstract — WCN 2013

No: 2431

Topic: 6 — MS & Demyelinating Diseases

Transorbital sonography in the hyperacute stage of optic neuritis: A bicentric case-control longitudinal, blind study

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Background: Acute unilateral optic neuritis (ON) is known to be associated with an increased optic nerve sheath diameter (ONSD) as revealed by ultrasonographic investigations.

However, there is no consensus in the literature on the diagnostic cut-off of ONSD.

Objective: The goals of this study were to evaluate the accuracy/sensibility of US in the diagnosis of acute ON, and to assess the association between ONSD threshold and ON.

Patients and methods: A prospective blinded observational (case-control) bicentric study was performed. Seventeen consecutive patients with an acute non-compressive unilateral ON and onset of visual loss during the prior 10 days were included. We enrolled 20 healthy controls, matched by gender and age (± 5 years). Two experienced vascular sonographers, blinded to the status of case or control and to the affected eye, performed the study using B mode US. ONSD was defined as a sign of nerve thickening, if the difference in nerve diameter of 0.3 mm or more, compared with the contralateral side or controls.

Results: A significant ONSD increase on the affected compared to the health side, was found in all patients by both investigators ($6.6 \text{ mm} \pm \text{DS } 0.85$ versus $5.5 \text{ mms} \pm \text{DS } 0.66$ $p < .001$). ONSD was normal in all controls ($5.2 \pm \text{DS } 0.62$) mm and in all the non-affected eyes.

Conclusions: Our findings suggest that transorbital sonography has a very high sensitivity in recognizing ON, similar to that of the visual evoked potentials.

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Abstract — WCN 2013

No: 2460

Topic: 6 — MS & Demyelinating Diseases

Klüver–Bucy Syndrome in a multiple sclerosis patient

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Background: Multiple sclerosis (MS) can result in a complex net of neuropsychiatric symptoms. Klüver–Bucy Syndrome (KBS) is a rare complex behavioral syndrome. It's composed of psychic blindness, hyperorality, hypermetamorphosis, hypersexuality, disturbed dietary habits and placidity. It's widely related to temporal lobe lesions.

Objective: Report the rare association between Multiple Sclerosis and Klüver–Bucy Syndrome.

Material and methods: Case report.

Results: A 32 year old woman diagnosed with MS by the age of 20 started a multiple incipient behavioral change. She was already in Secondary-Progressive phase of the disease. She presents with placidity, hyperorality, hypermetamorphosis and a complex disturbed dietary habit. She used to avoid non-sweet and usually add sugar or sweetener in all kinds of food. The MRI showed diffuse cerebral atrophy especially in the temporal lobes with poor inflammatory lesion in both of them. Topiramate and valproic acid were introduced and controlled well the sweet compulsion. Anti-psychotics were tried in other behavioral symptoms.

Conclusion: MS is a complex disease composed of both inflammatory and degenerative lesions. Neuropsychiatric symptoms can be related to both of them. There is a complex role of cognitive and special behavioral symptoms that compose MS spectrum, as KBS is shown in this abstract.

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Abstract – WCN 2013

No: 2403

Topic: 6 – MS & Demyelinating Diseases

Disability due to multiple sclerosis in Ukraine (10 year experience)

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Multiple sclerosis (MS) is one of the leading causes of permanent disability due to neurological conditions in young adults.

Methods: 10 year (2003–2012) dynamics of MS prevalence and disability incidence rates for adult population (all per 100,000) was investigated. Disability grade (“invalidity” status) in Ukraine is set according to the Ministry of Health Order (no. 561, August 05, 2011) based on ICF (2001) principles. We compared MS prevalence (Ukraine Ministry of Health Statistics Dept.) and primary disability data structure due to MS and encephalomyelitis (ICD X codes G35, G36–37) in Ukrainian regions.

Results: Global panukrainian MS prevalence was gradually increased (48.0–47.8–48.4–49.1–50.0–50.1–50.9–51.6–52.9–54.6) as well as MS disability incidence (2.4–2.4–2.6–2.7–2.6–2.5–2.6–2.9–3.0–3.1). We also discovered (the data are not collected in clinical statistics) the relatively high rates of disability incidence due to demyelinating encephalomyelitis (total rates 1.3–1.2–1.3–1.1–1.2–1.2–1.0–1.0–1.1–0.9 for years 2003–2012 accordingly).

Conclusions: Further disability data investigation should be performed for understanding the correctness of demyelinating encephalomyelitis diagnostics and protection from MS false negative diagnostics followed by improper expert-rehabilitation tactic.

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Abstract – WCN 2013

No: 2321

Topic: 6 – MS & Demyelinating Diseases

Magnitude and concurrence of anxiety and depression among attendees at tertiary care neurology unit in Oman with multiple sclerosis (MS)

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Background: Anxiety and depression are commonly reported by persons with multiple sclerosis (PwMS) but no data, to our knowledge, have emerged from Arab Islamic population.

Objective: The study aims to investigate the prevalence of anxiety and depression among PwMS attending tertiary care hospital in Oman, Neurology Clinic, Sultan Qaboos University Hospital (SQUH). The characteristics such as socio-demographic and clinical variables were also explored.

Methods: Consecutive and consenting PwMS were subjected to the following measures.

The **Hospital Anxiety and Depression Scale** (HADS) was used to measure anxiety (cut-point > 7) and depression (>7).

Results: 54 patients with MS participated in our study and approximately 87% were on Interferon. The age range from 17 to 50 and the average age was 31.4 ± 8.84. In terms of gender, majority were female (males = 30% vs. 70% females). Approximately 50% of the attendees have acquired university or higher graduate education while 39% had completed secondary education. The rest acquired on the lower strata of education with one illiterate. The majority (69%) of attendees were married while 31% were single. Approximately 83% were employed, students or job-seekers. The majority of the patients had <4 in the EDSS. Prevalence of anxiety was 53%, depression 39% on HADS.

Conclusions: Anxiety and depression are common in PwMS among the attendees of tertiary care in Oman. Such psychosocial variables have been largely unreported from non-western population. As these variables are strong indicators of burden of MS, concerted efforts are needed to address such psychosocial dysfunctions in the algorithms of care for PwMS.

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Abstract – WCN 2013

No: 2394

Topic: 6 – MS & Demyelinating Diseases

Acute disseminated encephalomyelitis (ADEM) – probably triggered by *Borrelia burgdorferi* infection – with fatal end

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Background: Acute disseminated encephalomyelitis (ADEM) is a nonvasculitic inflammatory demyelinating condition that usually occurs following a viral, bacterial or parasitic infection, or even spontaneously.

Case presentation: We present a case of a patient of 48, who presented the onset of the actual disease with low-grade fever, fatigability, impaired gait and balance, symptoms debuted 2 months before he was admitted in our hospital. Neurological examination revealed bilateral cerebellar and pyramidal syndrome, thermo-algic hypoesthesia below T4 level and urinary incontinence. Cerebral MRI showed bilateral supra- and infra-tentorial demyelinating lesions, some of it with gadolinium enhancement and cervico-thoracic MRI revealed extended thoracic myelitis; lumbar puncture showed CSF with pleocytosis and highlights the presence of IgG anti-Borrelia antibodies. We considered the infection with *Borrelia burgdorferi* as a possible “trigger” factor in apparition of ADEM. The evolution was initially with improvement of symptoms on treatment with cephalosporin and high doses of methylprednisolone. After 2 months, the patient presented sudden motor deficit in the inferior limbs with impairment of the gait and slurred speech. Cerebral and spinal MRI showed more demyelinating lesions with larger diameters, some of them presenting gadolinium enhancement; most of lesions from the posterior fossa were confluated. Despite the treatment with antibiotics, methylprednisolone associated with IgG

IV immunoglobulin, the patient presented rapid deterioration of the neurological and general status, finally with death by cardio-respiratory arrest, after 4 days.

Conclusion: We presented this case because of the rare frequency of this disease, the peculiar evolution in two phases and the fatal end, unusual for ADEM.

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Abstract — WCN 2013

No: 1969

Topic: 6 — MS & Demyelinating Diseases

Percept: A prospective multicenter observational study on benefit/risk perception of natalizumab in neurologists and their patients in Germany

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Background: For decision making on NAT use, neurologists (NLs) and patients (pts) should be adequately informed on NAT efficacy and PML risk stratification.

Methods: PERCEPT investigates NAT benefit/risk perception and information levels of NLs and pts in routine practice. We present an interim analysis of BL data.

Results: Most NLs (n = 77) rated their MS expertise “excellent” (52.8%) or “very high” (41.9%). BL characteristics of 599 pts [means]: treatment in practices 76.3%; MS duration 10.3 years; NAT duration: 26.6 months; IS pretreatment: 9.0%; anti-JCV tested: 93.7% (45.5% positive). On visual analogue scales (1–25), NLs and pts rated MS as “serious threat” (NLs 18.3, pts 18.6), and PML as “moderate risk”: (NLs 8.11; pts 10.8).

98.5% of NLs rated NAT “very effective”; 48.4% tolerated a PML risk of 1/100, 18.8% up to 1/10. 80.5% of NLs rated the anti-JCV assay “clearly helpful”.

NL information level: correct answers regarding pivotal trial data on freedom from progression on NAT and placebo: 58.4% and 35.1%. Correct answers on PML risk during 1st/2nd + year of NAT: 97.3%/40.2%; on the risk factors IS pretreatment and JCV serostatus: 79.5% and 84.6%; on risk stratification 76.6%.

Conclusions: NLs and pts perceived MS as serious threat and PML as moderate risk. NLs had limited awareness of data on freedom from progression. Knowledge on risk stratification was generally good. The anti-JCV assay was regarded very useful. The PML risk tolerated by 50% of NLs was in the range for pts with 3 risk factors.

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Abstract — WCN 2013

No: 2051

Topic: 6 — MS & Demyelinating Diseases

Natalizumab long-term therapy for relapsing MS in clinical routine: Final results of the prospective observational multicenter study Tysabri® 24 plus

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Background: Natalizumab showed high efficacy in the pivotal trial AFFIRM and post-marketing studies; 37% of natalizumab patients remained free from disease activity for 2 years in AFFIRM. Patients infected with JCV have an increased PML risk on natalizumab. Additional risk factors include previous immunosuppressive therapy and natalizumab treatment >2 years.

Methods: In MS patients with ≥2 years of ongoing natalizumab therapy, Tysabri® 24 plus (T24plus) documented clinical outcomes, MRI, patient monitoring and anti-JCV serostatus in five 3-monthly visits.

Results: Patient characteristics at baseline (n = 939; 66.7% female) [means (±SD)]: age 40.5 yrs (±9.5); MS duration 10.7 yrs (±5.8); natalizumab (NAT) infusions until baseline 35.4 (±8.6); previous therapy with ≥1 immunomodulator: 92.4%, with ≥1 immunosuppressant: 19.2%.

During observation (321 days (±97)), clinical parameters evolved as follows: annualized relapse rate before NAT: 2.2 (±1.5); at baseline (after ≥2 yrs of NAT): 0.4 (±0.7); during T24plus: 0.18 (±0.6). Relapse-free pts in T24plus: 80.1%. EDSS before NAT 3.5 (±1.6); at baseline 3.4 (±1.8); during T24plus: 3.5 (±1.8). Cognition (DemTect) at baseline 14.8 (±3.5); during T24plus: 15.7 (±2.7). 71.7% of pts had ≥1 MRT. JCV serostatus was tested in 41.1% of pts (50.9% positive). 25 pts had serious adverse events, including 2 PML and 1 presumptive PML.

Conclusions: In patients with ≥2 years of natalizumab therapy T24plus revealed sustained high efficacy of natalizumab with effective reduction of the relapse rate and sustainably stable level of disability. Natalizumab safety profile was consistent with previous data.

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Abstract — WCN 2013

No: 1945

Topic: 6 — MS & Demyelinating Diseases

Incidence and prevalence of multiple sclerosis in Nordland County, Norway, 1970–2010

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Background: The risk of multiple sclerosis (MS) increases with increased latitude. Previous studies have reported an unevenly distribution of the disease in Norway, with a relatively low prevalence in the most northern parts of the country. The need for more accurate knowledge of the distribution of the disease in Norway motivated for a survey in Nordland County.

Objective: To describe trends in the occurrence of MS over a period of four decades.

Material and methods: All patients with MS living in Nordland County in the period 1970–2010 were identified by reviewing hospital charts. Point prevalence at the beginning of the decades was calculated. The average annual incidence was calculated for 5-year periods. The diagnostic criteria of Poser and McDonald (2001) were applied.

Results: The total crude prevalence rate on January 1st 2010 was 182.4 per 100,000. The annual incidence continuously increased from 0.7 per 100,000 in 1970 to -74 to 10.1 per 100,000 in 2005 to -09. The time delay from the first symptom to diagnosis has been stable from 1975 to 2010.

Conclusion: We report an increasing incidence and prevalence of MS in Nordland County. The county is a high-risk area for MS. The

increase could be due to altering in the diagnostic sensitivity, but could also reflect a genuine increase of the occurrence of the disease.

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Abstract – WCN 2013

No: 1560

Topic: 6 – MS & Demyelinating Diseases

Measurement and estimation of relaxation times in magnetic resonance imaging of brain tissue in patients with multiple sclerosis (MS)

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Contrast of conventional MRI Images is influenced by different parameters such as T1, T2 and proton density. Effect of these parameters together; sometimes cause invisibility of some lesion especially small ones in patient with multiple sclerosis. However, each brain tissue has specific values of T1 & T2 that usually change in different pathologic lesions, and can be diagnosed by first creating specific MR Images with contrast based on one of the relaxation times T1 or T2 only without the involvement of each other and the measurement of these relaxation times T1 & T2. Present study has been performed on 1.5 T MRI system. In the study, with optimization of interpulse time intervals (TE, TI & TR) to minimize errors, first T1 & T2 maps were computed from two images using spin Echo (SE) and inversion Recovery (IR) pulse sequences in some healthy volunteers as controls and patients with definite Multiple sclerosis. Then T1 & T2 values in 26 different regions of brain tissues were measured in controls and patients and finally compared together. Findings of this study indicate that not only T1 & T2 values in MS plaques increase considerably but also these values in normal appearing white matter (NAWM) increase meaningfully, whereas T2 value in normal appearing gray matter (NAGM) decreases meaningfully. As a result, increasing of T1 & T2 values in NAWM and decreasing of T2 values in NAGM can be considered as a diagnostic parameter of MS disease.

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Abstract – WCN 2013

No: 2281

Topic: 6 – MS & Demyelinating Diseases

The mesenchymal stem cells labelled by magnetic nanoparticles applying in experimental autoimmune encephalomyelitis

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Introduction: The application of mesenchymal stem cells (MSCs) labelled by magnetic nanoparticles in medicine attracts a great scientific interest.

Objective: To realize the control of MSC labelled by magnetic nanoparticle allocation in experimental autoimmune encephalomyelitis (EAE).

Methods: The laboratory animals (rats) with clinical signs of EAE were assigned into four groups: the control group with one fold saline solution injected, the second group injected with MSC, the third one with magnetic nanoparticles injected, and the fourth – MSC labelled by magnetic nanoparticles. As magnetic nanoparticles were used iron oxide 5.9 nm nanoparticles stabilized by oleic acid and triethanolamine. The clinical signs of EAE were estimated according to the International Criteria of Assessment of clinical signs

of EAE in laboratory animals. All animals underwent MRI control on the 1st, 10th, 20th, and 22nd days after procedure.

Results: The regression of clinical signs of EAE realized in shorter terms in the second and the fourth groups simultaneously ($p < 0.05$). The magnetic nanoparticles were detected by MRI in the brain of the animals only in the fourth group.

Conclusions: The application of MSC labelled by magnetic nanoparticles amplifies the possibilities of non-invasive control methods of cell technology application in CNS diseases.

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Abstract – WCN 2013

No: 2329

Topic: 6 – MS & Demyelinating Diseases

Natalizumab saturation: Biomarker for individual treatment holiday after natalizumab withdrawal?

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Background: More and more multiple sclerosis (MS) patients switch from natalizumab to fingolimod because of the risk of progressive multifocal leukoencephalopathy. The duration of the treatment holiday is still under debate referring to a possible recurrence of disease activity. **Aims of study:** To evaluate the prognostic value of natalizumab saturation on T cells for the recurrence of clinical and radiological disease activity.

Methods: Cell surface bound natalizumab saturation (in %) of CD8⁺ and CD4⁺ T cells from five MS patients was determined before initiation of fingolimod by flow cytometry and related to clinical and MRI outcome during a 6-month follow-up.

Results: In two patients with either clinical or radiological disease activity, the natalizumab saturation on CD8⁺ and CD4⁺ T cells was less than 30%, respectively. In contrast, the remaining three patients with the absence of disease activity had a median natalizumab saturation of 70% (range 59–79%) on CD4⁺ and 66% (range 52–68%) on CD8⁺ T cells.

Conclusions: The data of this pilot study indicate that clinical and radiological disease activity is closely linked to natalizumab saturation at the time point of switch. The determination of natalizumab saturation may be an essential tool to monitor cessation of natalizumab treatment.

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Abstract – WCN 2013

No: 2282

Topic: 6 – MS & Demyelinating Diseases

Switching from natalizumab to fingolimod

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Background: Natalizumab(N) is an efficient treatment for Multiple Sclerosis (MS). Unfortunately, recent data show that the disease activity returns after N discontinuation. Fingolimod (F), recently approved in highly active MS, might represent an alternative treatment after N suspension.

Objective: To evaluate the MS disease activity in patients who switched from N to F.

Patients and methods: We analyzed retrospectively 44 patients who switched to F (F-Group) after N suspension, and compared them with a group of 17 patients who restarted N after a drug holiday (N-Group). We assessed the recurrence of disease activity evaluating the relapse rate in 4 periods of time: before N (pre-N), during N (on-N), during wash-out (WO), and during the second treatment (post-N).

Results: In the F-Group: the number of patients experiencing relapses was 40 (90%) in pre N, 8 (18%) in on-N, 13 (29%) in WO, 4 (15%) in post-N. During WO the relapse-rate was significantly higher than on-N, as well as in post-N. There were no significant differences with the N-Group in any of the period considered.

Conclusion: Our experience shows that the beneficial effects of N rapidly deteriorated after its withdrawal. In our cohort the disease activity during treatment with F was not significantly different than during WO and on-N. However, it was still significantly lower than pre-N ($p = 0.02$), as we experienced with patients in the N-Group. Nowadays, an early resumption of alternative treatments after N suspension is recommended. F seems to be an option, but the optimal strategy is still to be determined.

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Abstract – WCN 2013

No: 1288

Topic: 6 – MS & Demyelinating Diseases

Birth outcomes of pregnancies fathered by men with multiple sclerosis

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Background: Studies have examined the association between multiple sclerosis (MS)-related clinical factors and birth outcomes in mothers but not fathers with MS. Autoimmune diseases in fathers have been associated with preterm and lower birth weight babies; poor sperm quality has also been reported in men with MS.

Objective: To investigate if paternal MS disease duration or disability (as measured by the Expanded Disability Status Scale) is associated with newborn birth weight or gestational age.

Patients and methods: The British Columbia (BC) MS database (containing >80% MS patients in BC, Canada) and the BC Perinatal Database Registry (capturing >99% of births in BC) were linked, along with the BC Vital Statistics Birth Registry to enable linkage of individual-level data for newborns, mothers and MS fathers in this retrospective cohort study. We investigated the association between paternal MS disease duration, disability (± 3 years of conception) and neonatal outcomes using multivariate regression models to adjust for confounding and familial effects.

Results: There were 202 singleton births fathered by men with MS (1996–2010). Mean birth weight of newborns was not associated with MS disease duration (adjusted $p = 0.20$) or disability (adjusted $p = 0.25$). Mean newborn gestational age was also not associated with MS disease duration (adjusted $p = 0.42$) or disability (adjusted $p = 0.32$).

Conclusion: We did not find an association between paternal MS-related clinical factors and birth outcomes. As one of the first studies investigating paternal MS clinical factors on birth outcomes, our findings are reassuring for fathers with MS and their families.

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Abstract – WCN 2013

No: 2060

Topic: 6 – MS & Demyelinating Diseases

Monitoring and management of MS patients starting natalizumab therapy in Germany – Final results of the prospective TYSTART study

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Background: In AFFIRM, 37% of natalizumab patients (pts) remained free from disease activity (placebo: 7%) over 2 years. JCV-infected pts have an increased PML risk on natalizumab, particularly after previous immunosuppression and natalizumab therapy >2 years. Natalizumab therapy requires structured monitoring.

Methods: TYSTART documented MS outcomes and monitoring in patients starting natalizumab (≥ 2 infusions received) in clinical routine. Clinical, MRI and laboratory parameters (JCV antibodies since 01/2012) were recorded at 5 three-monthly visits.

Results: Patient characteristics ($n = 258$; 65.9% women) baseline [mean (\pm SD)]: age 39.6 yrs (± 10.5); MS duration 8.3 years (± 7.0); natalizumab infusions before BL 3.9 (± 5.6); EDSS 3.6 (± 1.6); Fatigue Severity Score (FSS) 4.2 (± 1.7). 89.5% of patients had received ≥ 1 immunomodulator, 19.0% ≥ 1 immunosuppressant. MRIs before/at baseline showed: contrast-enhancing lesions: 2.9 (± 4.1) cerebral and 0.9 (± 1.5) spinal; T2 lesions: 11.5 (± 11.3) cerebral and 2.2 (± 2.7) spinal. Observation duration was 10.5 months (± 2.9). Common lab tests were transaminases (42.7–69.8% at the 5 visits) and blood counts (53.0–68.6%). 91.1% of patients had MRIs before/at baseline; 44.3% had 1–4 MRIs during study. 28.1% of 228 JCV antibody assays were positive. In patients with 12 months data, the EDSS remained stable (3.6 (± 1.7) vs. baseline 3.5 (± 1.7)); the FSS improved from 4.1 (± 1.7) to 2.2 (± 2.1). Four patients had serious adverse events, including 2 MS relapses and no PML.

Conclusions: TYSTART confirmed the high efficacy of natalizumab. The mean EDSS remained stable, the FSS improved during observation. No PML occurred. Monitoring comprised MRI and laboratory investigations including JCV antibodies.

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Abstract – WCN 2013

No: 2287

Topic: 6 – MS & Demyelinating Diseases

The meaning of herpetic infection in the progression of multiple sclerosis

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The aetiology of MS has multifactorial factor, in which the considerable part belongs to viruses.

Objective of study is learning the prevalence of herpetic infection of patients with MS, the influence of different types of herpes on the progression of MS.

Thirty patients with MS (28 – RR MS in the remission, 2 – PP MS) entered the study. Patients' age – 30.5 ± 8.1 ($M \pm \sigma$) years, the duration of the disease – 5.8 ± 4.4 years, the number of all exacerbations in time of disease – 4.0 ± 2.7 and EDSS 2.4 ± 1.2 . Enzyme-linked immunoelectrodiffusion assay and polymerase chain reaction to antigen of herpetic viruses (1, 2, 3, 4, 5, 6 types) were used.

All patients had herpetic infection. In the group with the virus (1, 4 or 5 type) in the replication phase ($N = 5$) EDSS = 2.8 ± 1.2 , the duration of the disease – 6.8 ± 4.6 years and the number of

exacerbations – 4.6 ± 3.1 . In the group with the latent virus ($N = 25$) EDSS = 2.4 ± 1.2 , the duration of the disease – 5.5 ± 4.4 years and the number of exacerbations – 3.7 ± 2.6 . Patients ($N = 15$) with latent 5th type had EDSS = 2.0 ± 0.6 , the duration of the disease – 5.6 ± 4.8 years and the number of exacerbations – 3.3 ± 2.2 . Patients ($N = 10$) with more than 2 types of herpes had EDSS = 2.8 ± 1.7 , the duration of the disease – 5.3 ± 4.1 years and the number of exacerbations – 4.5 ± 3.3 .

Patients with active herpes or with several types had more frank EDSS and more high number of exacerbations.

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Abstract – WCN 2013

No: 2163

Topic: 6 – MS & Demyelinating Diseases

BG-12 (dimethyl fumarate) and pregnancy: Preclinical studies and pregnancy outcomes reported during the clinical development program

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Background: No formal studies of BG-12 (dimethyl fumarate) were conducted in pregnant women, but pregnancies have occurred during the BG-12 clinical development program.

Objective: To present data from animal reproductive toxicology studies and pregnancy outcomes reported during the BG-12 clinical development program.

Patients and methods: Reproductive and developmental toxicology was evaluated in rats and rabbits given dimethyl fumarate during organogenesis or pregnancy and lactation. Subjects in BG-12 clinical trials were required to use contraception and discontinue drug in the event of pregnancy.

Results: There was no evidence of impaired fertility in rats or teratogenicity in rats and rabbits given dimethyl fumarate at doses that caused reductions in maternal weight gain. As of January 2, 2013 (data cutoff), 38 pregnancies in subjects exposed to BG-12 (37 MS patients, 1 healthy volunteer) and 14 pregnancies in placebo recipients were reported in clinical studies. In patients exposed to BG-12, 22 live births (65%), 3 spontaneous abortions (9%), and 9 elective terminations (26%) were reported; information is pending for 3 subjects and 1 subject was lost to follow-up. In placebo recipients, 9 live births (64%), 3 spontaneous abortions (21%), and 2 elective terminations (14%) were reported. No fetal abnormalities have been reported. The incidence of spontaneous abortion was consistent with the expected rate in the general population.

Conclusion: Based on limited data, no increased risk of fetal abnormalities or adverse pregnancy outcomes associated with gestational exposure to BG-12 during the first trimester has been observed. Further data will be collected through a pregnancy registry.

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Abstract – WCN 2013

No: 2270

Topic: 6 – MS & Demyelinating Diseases

Pregnancies after alemtuzumab treatment – Case report

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Introduction: Multiple Sclerosis (MS) is a chronic neurological disease, which is highly prevalent in women of childbearing age. Several new pharmacological treatments are now tested for relapsing remitting (RR) phase of the disease, but none of them is approved for use during pregnancy.

Alemtuzumab is a humanized monoclonal antibody that targets CD52 on lymphocytes and monocytes, and it seems to be an effective treatment for early multiple sclerosis.

Methods: Observational case report.

Results: A 27-year-old woman affected by MS, started treatment with alemtuzumab on May 2004 (EDSS 1.5). She received two intravenous cycles of alemtuzumab, last dose was on 11.05.2005. In April 2007 she became pregnant, and due to obstetric reasons in 10 January 2008 cesarean section was performed and a healthy male was born, birth weight 4140 g. In 2009 she became pregnant once again and in 18 March 2010 the next healthy son was born, birth weight 3800 g. Now our patient is in her third pregnancy. She is neurologically stable. There were no relapses since 2004, neither during the pregnancies nor in the post partum periode. Her EDSS is 1.5.

Conclusion: There is no long-term observation on a woman who becomes pregnant after alemtuzumab administration, but our patient can be an example of relative safety and efficacy of alemtuzumab.

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Abstract – WCN 2013

No: 2229

Topic: 6 – MS & Demyelinating Diseases

A case of very late onset multiple sclerosis

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The onset of multiple sclerosis (MS) after the age of 60 is very rare and classified as very late onset MS. The prevalence of very late onset MS ranges between 0.6% and 1.3%.

We report a 71-year-old Caucasian female presenting with monoparesis of her left leg. Fifteen days after her first symptoms she developed paresthesia on the upper thoracic region. Cerebral magnetic resonance imaging (MRI) showed multifocal white matter lesions with gadolinium enhancement and the most prominent one located at the pons and thoracic MRI revealed hyperintense lesion with gadolinium enhancement at the T3–4 level. Metastatic diseases, vasculitis, sarcoidosis and other demyelinating central nervous system disorders were ruled out with extensive tests. CSF analysis showed positive oligoclonal band type 2. She had a family history of MS in her daughter. Patient received intravenous corticosteroid therapy 1 g/day for 5 days and improved clinically. She didn't accept any disease modifying therapy and during 20 months follow-up period no new attack was observed.

This case has been presented to emphasize the possibility of multiple sclerosis in differential diagnosis of elderly patients with first onset multifocal neurological symptoms.

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Abstract — WCN 2013**No: 2242****Topic: 6 — MS & Demyelinating Diseases****Benefit 11: Long-term follow-up study of patients with clinically isolated syndrome treated with interferon beta-1b**

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Background: MS is a demyelinating disease that often begins as a clinically isolated syndrome (CIS). Long-term follow-up may provide important insights into outcomes for these patients; however, ascertainment is a challenge.

Objectives: To review the key findings over the 8-year study period of BENEFIT and to describe the design of the ongoing 11-year cross-sectional follow-up.

Patients and methods: 468 patients with CIS were randomized to either interferon beta-1b (early treatment) or placebo (delayed treatment) for 2 years or until CDMS. Analyses were conducted at 2, 3, 5, and 8 years. Comprehensive 11-year follow-up, including clinical, MRI, and OCT assessments, is ongoing through 2013–2014. To increase ascertainment in the BENEFIT 11 Study, the design includes telephone assessments as an alternative to face-to-face interviews and referrals from inactive to active sites.

Results: 93.6% completed the placebo-controlled phase. 84% and 75.5% completed 3 and 5 years and 60.7% gave consent for the extension, with a final ascertainment of 55%. Risk for CDMS was significantly lower in the early vs delayed treatment group (Year 2: 27% vs 45%, Year 3: 36% vs 51%, Year 5: 46% vs 57%, Year 8: 57% vs 66%). EDSS scores were not significantly different, and remained low in both groups (median 1.5 throughout the study).

Conclusion: Available follow-up of up to 8 years showed persistent advantages of early vs delayed treatment with interferon beta-1b in patients with CIS. The ongoing comprehensive reassessment at Year 11 will provide important information on disease course and long-term treatment effects.

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Abstract — WCN 2013**No: 2260****Topic: 6 — MS & Demyelinating Diseases****Autologous mesenchymal stem cell transplantation in patients with multiple sclerosis**

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Background: Autologous mesenchymal stem cell transplantation (AMSC) may provide a new pathogenetic treatment modality for a multiple sclerosis (MS) based on neuroprotective and immunomodulatory features of mesenchymal stem cells (MSCs).

Objective: To evaluate the efficacy of AMSC in patients with MS.

Patients and methods: Twenty-one patients with MS were enrolled in the study with median age of 35 years (range 19–47), median disease duration of 6 years (range 0.5–14). Thirteen patients were defined as relapsing-remitting MS (RRMS), 6 as progressive-relapsing (PR) and 2 as secondary-progressive (SP). Median baseline score of EDSS was 3 (range 1.5–6). Participants received a single intravenous infusion of autologous bone-marrow-derived MSCs no later than the third passage at the dose of $1-2 \times 10^6$ MSCs/kg. The clinical assessment included an EDSS evaluation, an MRI examination with gadolinium-enhancement, and optical coherence tomography (OCT).

Results: Median period of follow-up is 12 months (range 2–34). Confirmed neurological stabilization following AMSC was defined for the 14 patients, progression — for 6, improvement — for 1. Before AMSC 14 active lesions were revealed on baseline MRI scans and only 1 — on post-transplant MRI scans 9 months after AMSC. The number of relapses during a year before AMSC was 26, and 10 exacerbations were determined after AMSC. OCT revealed stabilization of RNFL thickness for 10 eyes, improvement — for 14, recovery — for 6.

Conclusion: Our results show that AMSC in MS is a clinically feasible and relatively safe procedure. Nevertheless, a longer follow-up period is needed.

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Abstract — WCN 2013**No: 2283****Topic: 6 — MS & Demyelinating Diseases****Severe thrombocytopenia in patient with multiple sclerosis during interferon-beta-1B treatment — Case report**

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Introduction: Interferon-beta-1B (Betaferon) is an established treatment for relapsing-remitting multiple sclerosis (MS) and haematological changes are commonly reported in clinical trials and clinical practice of this drug. Treatment with interferon-beta-1B led predominantly to asymptomatic dose-related reductions in white blood cells. At least two-thirds of patients affected by cytopenia experienced the onset of cytopenia within the first 6 months of therapy.

Methods: Observational case report.

Results: A 23-year-old man affected by MS, started treatment with interferon beta1B (Betaferon, 250 mg every other day, sc) on January 2011. Baseline blood examination and, in particular platelet count (276 GI/L) were normal. There was no history of any haematologic abnormalities or allergy. After 6 months of interferon-beta-1B treatment he developed thrombocytopenic hemorrhagic diathesis with platelet count 1 GI/L. Betaferon was immediately stopped. Patient was admitted to the hospital and received five infusions of packed red blood cells, and then steroids until platelet count was normalised. From February 2011 he is without steroids, and platelet count is 320 GI/L. Betaferon treatment was not.

Conclusion: Although haematological abnormalities are common and dose-related in patients with MS receiving interferon-beta-1B, the thrombocytopenia, especially so deep occurs quite rare.

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Abstract – WCN 2013**No: 2243****Topic: 6 – MS & Demyelinating Diseases****Uncommon ocular and visual signs of relapsing remitting multiple sclerosis**E. Kammoun, M. Zouari, E. Farhat, F. Hentati. *Neurology, National Institute Mongi Ben Hamida of Neurology of Tunis, Tunis, Tunisia***Background:** Ocular manifestations in multiple sclerosis (MS) are classic, including both visual and motor ocular systems. Optic neuritis remains the most frequent manifestation.**Objective:** To report two MS patients revealed by isolated homonymous lateral hemianopsia (HLH) and sixth cranial nerve palsy (SCNP).**Case 1:** A 29 year-old woman, who presented a sudden right visual field defect. Neurological examination revealed an isolated right HLH. CT scan demonstrated a left occipital hypodensity. The diagnosis of ischemic stroke was made. Brain MRI showed a large T2-weighted lesion in the left juxtacortical occipital region, associated to multiple T2-weighted lesions in the periventricular regions. The visual evoked potential was normal. Oligoclonal bands were present in the cerebrospinal fluid (CSF). MS was diagnosed and the patient was treated with intravenous methylprednisolone with clinical improvement.**Case 2:** A 30 year-old woman, who presented sudden horizontal diplopia. The neurological examination showed isolated left SCNP. The eyeground was normal. MRI showed a T2-weighted lesion of the sixth cranial nerve, associated to multiple T2-weighted lesions in the brainstem and spinal cord. Oligoclonal bands were present in the CSF. The diagnosis of MS was made with a complete spontaneously recovery within two weeks.**Discussion:** HLH is infrequent in MS, occurring in less than 1% of patients, due to the large required lesions in post-chiasmal visual pathway. Isolated sixth nerve palsy is a rare presenting sign of MS, with an incidence of 1.6%.**Conclusion:** The diagnosis of MS should be considered in young adults with HLH or ocular motor cranial nerve involvement.

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Abstract – WCN 2013**No: 2253****Topic: 6 – MS & Demyelinating Diseases****Rheumatoid arthritis and amyotrophic lateral sclerosis association: A case report and literature review**I. Bekri, M. Zouari, E. Farhat, F. Hentati. *Neurology, National Institute Mongi Ben Hamida of Neurology of Tunis, Tunis, Tunisia***Introduction:** Neurological complications are reported with rheumatoid arthritis (RA), involving especially the central nervous system. Degenerative motor neuron disease is unusual.**Objective:** The objective is to describe a rare association between RA and amyotrophic lateral sclerosis (ALS) and to discuss the relation between these two diseases.**Case report:** A 20-year-old woman presented rapidly progressive walking disorder and limb weakness without dysphagia and dysphonia. The physical examination showed signs of bilateral degeneration of upper and lower motoneurons involving the four limbs, without sensory disturbance and cranial nerve involvement. We noted also a deforming polyarthropathy of the hands. Electromyography found signs of impairment of the anterior horn of the spinal cord. Brain and spine MRI was normal. Serological test for rheumatoid factor was positive. The diagnosis of ALS associated with seropositive erosive RA was made.**Discussion:** ALS is a chronic disease of unknown cause. To date, only seven cases similar to our patient have been reported, most of them having long-standing but quiescent joint disease at the time of ALS onset. Our patient has an atypical bilateral presentation, without signs of medulla's involvement. There are other cases of ALS in combination with autoimmune diseases such as systemic lupus erythematosus, scleroderma, Wegener granulomatosis and Goujerot Sjögren's Syndrome. There is no evidence in favor of a common pathophysiological mechanism, and thus the possibility of a fortuitous association must be raised.**Conclusion:** The simultaneous presence of RA and ALS represents a difficult diagnostic challenge. There is no argument in favor of a shared physiopathological mechanism.

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Abstract – WCN 2013**No: 2181****Topic: 6 – MS & Demyelinating Diseases****A diagnostic challenge: Tumefactive demyelinating lesions, our clinical experience in Baskent University**S. Kibaroglu, U.S. Benli, E.D. Ciftci, M.K. Toprak, O.Y. Cakmak, U. Can. *Neurology, Baskent University, Faculty of Medicine, Ankara, Turkey***Background:** Tumefactive demyelinating lesions (TDLs) are a rare variant of multiple sclerosis (MS), characterized by the presence of large lesions in white matter (>2 cm) with mass like features and edema. Differential diagnosis of TDLs from other intracranial space occupying lesions is essential to avoid unnecessary interventions and treatments.**Methods:** Demographic, clinical, radiological and laboratory data of 6 patients with TDLs were retrospectively collected.**Results:** All patients were female with a mean age of 42.8 (33–57 years) and mean age at disease onset was 38.8 (31–47 years). The mean duration of disease was 4 years (range 1–10 years). All patients had solitary TDLs, five had tumefactive lesions at onset and one patient developed tumefactive lesion during the course of MS. One patient with RRMS developed two tumefactive relapses during follow-up. Clinical presentations varied but headache, visual changes, and sensory and motor symptoms were the most common. Brain MRI showed tumefactive lesions with mild to moderate edema and contrast enhancement. MR spectroscopy was performed in all cases and revealed findings suggesting demyelination.

CSF analysis was available for three patients. Oligoclonal IgG band was positive in one case. Biopsy was performed in one case, revealing nondiagnostic pathology; final diagnosis was TDLs. All patients responded to corticosteroids. Three patients were treated with interferon and one patient was treated with azathioprine.

Conclusion: Six patients with TDLs who had different clinical and radiological characteristics were presented with review of the literature with a special emphasis on spectroscopic investigation.

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Abstract – WCN 2013**No: 2171****Topic: 6 – MS & Demyelinating Diseases****Paraneoplastic neuromyelitis optica associated with stomach carcinoid tumor**T.M. Al-Harbi^a, M.F. Bin Falah^b. ^a*Neurology, King Fahad Specialist Hospital, Dammam, Saudi Arabia;* ^b*Neurology, Dr.Sulaiman Al-Habib Hospital, Manamah, Bahrain***Background:** Neuromyelitis optica (NMO), or Devic's Syndrome, is an autoimmune central nervous system demyelinating disorder affecting

primarily the spinal cord and the optic nerves. It is characterized by the presence of NMO antibodies, besides clinical and radiological findings. NMO and NMO-spectrum disorders have been reported in autoimmune disorders, such as systemic lupus erythematosus and Sjogren's syndrome. Rarely, they have been described as a paraneoplastic syndrome with cancers of lung, breast, and carcinoid tumor of thyroid.

Objective: To report a case with longitudinally extensive demyelinating lesion of medulla and cervical spinal cord, associated with carcinoid tumor of the stomach, high serum gastrin level, and positive serum anti-aquaporin-4 antibody (NMO IgG).

Case: A 40 year-old Saudi lady, who presented with severe nausea, vomiting, blurring of vision, vertigo, nystagmus and ataxia, was found to have a longitudinally-extensive demyelinating lesion at the cervico-medullary junction and cervical spinal cord on MRI. Her gastric endoscopy revealed carcinoid tumor of the stomach, and classic paraneoplastic antibodies in the serum and CSF were negative. She had extremely high serum gastrin level and high titer of NMO IgG autoantibody.

Conclusion: NMO may present as a paraneoplastic neurological syndrome associated with carcinoid tumor of the stomach.

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Abstract — WCN 2013

No: 2196

Topic: 6 — MS & Demyelinating Diseases

Atypical neurological presentation of GSS: Case report

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Introduction: Gougerot-Sjogren Syndrome (GSS) is an auto-immune exocrinopathy characterized by xerophthalmia and xerostomia. Neurological complications occur between 8.5 and 70%. Data concerning central nervous system (CNS) symptoms have been rarely described. We analyze the clinical, radiological and therapeutic aspects of a patient with CNS signs revealing primitive GSS and we discuss some particularities.

Case report: A 52-year-old woman was hospitalized for recurrent clinical focal signs of CNS involvement. It consisted on sudden left hemiparesis on 2009 and two sudden regressed episodes of aphasia on 2011 and 2013. Brain MRI revealed micro nodular lesions appearing as hyper-signal in T2 on periventricular white matter and frontal cortex. Both lacrymal and salivary gland secretions were affected. A high level of antinuclear antibodies to SSA was associated with inflammatory lesions in minor salivary glands biopsy. The diagnosis of GSS was established. An oral corticosteroid therapy (1 mg/kg/day) was prescribed during 6 weeks followed by progressive degeneration, with good outcome.

Discussion: CNS damages are less frequently associated with GSS's systemic damages, and are often inaugural. The pathophysiology remains unknown. Some authors suggest the pathologic role of the anti-SSA based on their higher frequency in patients with CNS manifestation. Neurological focal manifestations are the most observed, but deficit signs are polymorphic. Aphasia is rarely described. The onset may be sudden as stroke. The MRI can reveal T2 hyperintensities on periventricular white matter or juxtacortical, more rarely on the cerebral cortex. Treatment of GSS's neurological manifestations is usually based an oral corticosteroid with high dose.

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Abstract — WCN 2013

No: 2001

Topic: 6 — MS & Demyelinating Diseases

Onset of secondary progression and long-term prognosis in multiple sclerosis

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Background: Prognosis in multiple sclerosis (MS) remains largely unpredictable.

Objectives and methods: Among 806 relapsing-remitting (RR) patients from the London Ontario database, we assessed:

- 1) the effect of baseline features on the risk of secondary progression (SP);
- 2) the risk of becoming disabled (DSS 6–8) according to latency to SP.

Results: Longer latency to SP predicted lower risk of reaching DSS 6 (OR = 0.76 for 5 years and OR = 0.44 for 15 years of latency); same results were observed in patients matched for early (year-1 + year-2) relapses number. However, RR phase duration did not influence SP evolution. The latency to SP conversely related to the number of total relapses (1–2 = 8.8 years; 3–4 = 11.3 years; ≥5 = 12.2 years). Predictors of the risk of SP were male sex (HR = 1.41), older age at onset (age > 30years vs ≤20years, 21–30years HR = 0.52, 0.65, respectively) and high early relapse frequency (≥3 vs 1, 2 attacks HR = 0.63, 0.75, respectively). Disease duration affected the probability of becoming SP (OR = 1.07), increasing by 9% every 5 years. At baseline, OR for risk of SP yielded by 3 relapses were 3.6, 5.8, 9.5 in patients aged 20, 30 or 40 at onset respectively, and increased by 2-fold at 10 years (OR 7.5, 12.1, 19.8, respectively) and by 5-fold at 20 years (OR 15.6, 25.5, 41.4, respectively).

Conclusions: Prevention/delay of SP represents a potential therapeutic target. Early relapse number and age at onset can be used for selecting groups at higher risk of developing severe disability and requiring more aggressive treatments.

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Abstract — WCN 2013

No: 2105

Topic: 6 — MS & Demyelinating Diseases

Pediatric and adult multiple sclerosis — Comparison of magnetic resonance imaging characteristics at disease onset

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Background: Only few studies have compared MRI findings at disease onset between patients with pediatric- (POMS) and adult-onset multiple sclerosis (AOMS).

Objective: To compare the imaging characteristics of brain and spinal cord between children and adults at MS onset. Furthermore we evaluated the 2010 McDonald criteria in both groups.

Patients and methods: Retrospective analysis of initial MRI from 49 children and 43 adults. The following parameters were assessed: lesion distribution, number of T2 hyperintense, gadolinium-enhancing, and giant (≥ 2 cm) lesions.

Results: We found no significant differences in the number and distribution of cerebral lesions, as well as number and size of spinal lesions between the two groups. However, there was a tendency for children to have more frequently an infratentorial involvement (59% versus 44% of patients). Adults more frequently had enhancing spinal lesions (68% versus 28%), as well as predominantly only thoracal located lesions (25% versus 4%). At clinical onset the 2010 McDonald criteria were fulfilled as follows: dissemination in space (DIS) 89% in POMS and 86% in AOMS, dissemination in time (DIT) 53% in POMS and 44% in AOMS, and both, DIS and DIT, 51% in POMS and 44% in AOMS.

Conclusion: Between our pediatric- and adult-onset MS groups we did not find major differences on initial MRI presentation, although a trend was noted for higher percentage of infratentorial involvement in children. The adults showed more frequently contrast-enhancing spinal lesions. Regarding the 2010 McDonald criteria the demonstration of DIT and of both, DIS and DIT, was higher in children.

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Abstract – WCN 2013

No: 2101

Topic: 6 – MS & Demyelinating Diseases

Blink reflex test evaluation during immune-modifying therapy in multiple sclerosis patients

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The Blink Reflex Test (BRT) is an electrically-induced reflex used for evaluation of brainstem circuits. BRT helps locate the lesion, even to the subclinical manifestation at the early course of multiple sclerosis (MS). Electrophysiological techniques have an important role in demonstrating brain injuries that are clinically silent and could be performed in follow-up evaluations during remissions and exacerbations obtained with a little cost. Correlations between brain MRI and electrophysiological data from BRT have not been widely studied in the patients with MS.

The aim of the present study was to assess the changes of BRT in patients with MS and their relations to clinical and MRI brainstem findings before and during immune-modifying therapy.

Fifty-five (mean age 36.5 years) diagnosed according McDonald criteria MS patients were divided into two groups: group I – 31 patients with newly diagnosed MS and group II – 24 MS subjects receiving immunomodulatory therapy. Patients underwent clinical tests using Expanded Disability Status Scale (EDSS), BRT and MRI evaluation.

In group I, contrary to group II the R1 latency was abnormal in 54% vs 47% patients, respectively ($p > 0.05$); ipsilateral R2 latency was abnormal in 60% vs 47%, respectively ($p < 0.05$); contralateral R2 latency was incorrect in 63% vs 45%, accordingly ($p < 0.05$). These changes achieved in group II correlate with reduced gadolinium enhancing lesions in brain MRI and clinical state improvement.

Our findings suggest that BRT, mainly R2 latency, may have a role as prognostic tool to evaluate the effect of therapeutic immune-modifying procedure in MS patients.

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Abstract – WCN 2013

No: 2107

Topic: 6 – MS & Demyelinating Diseases

Functional and structural disruption of the precuneus contributes to cognitive impairment in pediatric multiple sclerosis

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Background: Cognitive impairment affects $\geq 40\%$ of pediatric MS patients.

Objectives: To combine structural and functional MRI to define the mechanisms responsible for cognitive impairment in pediatric MS.

Methods: Brain dual-echo, 3D T1-weighted and resting state (RS) fMRI scans were acquired from 35 pediatric MS patients and 16 matched healthy controls. Patients with ≥ 2 abnormal neuropsychological tests were classified as cognitively impaired (CI). Regional distribution of lesions and gray matter and white matter (WM) atrophy were assessed using SPM8. Functional connectivity (FC) of the default mode network (DMN) was assessed using an independent component analysis.

Results: Forty-five percent of patients were CI. Compared to cognitively preserved (CP), CI patients had a higher probability to harbor lesions in the right (R) thalamus, bilateral cingulum, R precuneus and bilateral parieto-occipital WM. Compared to CP, CI patients had atrophy of the R precuneus, left middle temporal gyrus, splenium of the corpus callosum (CC), posterior cingulum, WM close to precuneus, and R superior longitudinal fasciculus. CI patients showed a reduced DMN RS FC of the precuneus, whereas CP patients had an increased RS FC of the anterior cingulum. MRI findings correlated with the number of abnormal tests and performance at spatial, verbal memory and attention tests ($r = 0.42-0.70$, $p < 0.001$).

Conclusions: In pediatric MS patients, cognitive dysfunction is associated to structural and functional abnormalities of the DMN, mainly in posterior brain regions. Increased RS FC of regions of the frontal lobe might contribute to cognitive preservation.

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Abstract – WCN 2013

No: 1980

Topic: 6 – MS & Demyelinating Diseases

Iron and oxidative damage in the multiple sclerosis brain

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Background: Multiple sclerosis (MS) is a chronic inflammatory disease of the central nervous system (CNS) leading to oligodendrocyte destruction, demyelination, and neurodegeneration. Iron in the healthy human CNS is essentially stored within oligodendrocytes and myelin. However, excess or liberated iron has the ability to induce oxidative damage, and is therefore implicated in various neurodegenerative disorders.

Objective: We aimed to characterize the impact of iron liberation from degenerating oligodendrocytes and myelin on neurodegeneration in MS.

Materials and methods: Formalin-fixed, paraffin-embedded autopsy brain tissue of 33 MS cases, including 7 acute MS cases, and 30 controls was studied. For detection of non-heme iron, the diaminobenzidine-enhanced Turnbull blue staining was applied. The iron storage protein ferritin, the ferroxidases hephaestin and ceruloplasmin, and oxidized phospholipids as marker for oxidative stress (E06 epitope) were detected by immunohistochemistry.

Results: Compared to controls, MS brains showed a global reduction of iron in non-lesioned white matter oligodendrocytes and myelin. We observed a shift of iron storage and ferritin expression from oligodendrocytes to microglia/macrophages at active lesion edges. Upon iron accumulation, microglia became dystrophic and vanished towards inactive lesion cores, where iron was either completely removed or stored within astrocytes and axons. A minor proportion of active lesions harbored elevated amounts of iron in astrocytes and axons. Rarely, a direct colocalization of iron and oxidized phospholipids could be detected.

Conclusion: MS leads to a global iron loss in the brain white matter. Demyelination in MS leads to waves of iron liberation from intracellular stores, which may promote oxidative damage.

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Abstract — WCN 2013

No: 2045

Topic: 6 — MS & Demyelinating Diseases

Intrinsic damage to the cerebellar peduncles is associated to multiple sclerosis clinical disability

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Background: The cerebellum is a region frequently affected by multiple sclerosis (MS).

Objectives: To apply a voxel-wise analysis to metrics from diffusion tensor (DT) MRI tractography and T2 lesions of the middle (M) and superior (S) cerebellar peduncles (CP) to quantify their structural damage in MS, and to assess the relationship between CP damage vs global clinical disability, impairment of cerebellar (C) and brainstem (B) functional systems (FS).

Methods: Brain dual-echo and DT MRI sequences were collected from 172 MS patients and 46 healthy controls (HC). The EDSS score and degree of impairment of different EDSS FS were rated. Patients were dichotomized using CFS and BFS (impaired: FS \geq 1, unimpaired: FS = 0) and EDSS (ambulatory impaired: EDSS \geq 4.0, fully ambulatory: EDSS < 4.0). Using DT MRI tractography, probability maps of the SCP and MCP were produced. Voxel-wise analysis was used to assess the topographical distribution of damage along these tracts.

Results: Compared to HC, MS patients showed increased mean (MD), axial (AD), radial (RD) diffusivity and decreased fractional anisotropy (FA) of both tracts. Diffusivity abnormalities were more pronounced in impaired patients. Patients with EDSS \geq 4.0 had a higher probability of having T2 lesions in the MCP bilaterally, while patients with BFS \geq 1 had a higher probability of having T2 lesions in one small cluster in the left SCP.

Conclusions: In MS, the assessment of MCP and SCP damage, in terms of T2 focal lesions and diffuse white matter tract injury (probably due to Wallerian degeneration), contributes to explain global clinical impairment and impairment at single FS.

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Abstract — WCN 2013

No: 2053

Topic: 6 — MS & Demyelinating Diseases

Biochemical markers of CNS demyelinating diseases

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Background: Biochemical markers could be useful in diagnosing and monitoring of demyelinating diseases. Nowadays there is just one autoantibody specific to neuromyelitis optica (NMO).

Objective: Purposes of our study were to detect AQP4-IgG in patients with NMO-spectrum disorders, to investigate nitrites, nitrates (NOx) and nitrosothiols levels (RSNO) in serum of multiple sclerosis (MS) patients and to measure neurofilament heavy chain levels (NfH) in CSF/serum of MS patients compared with control group and ALS patients.

Patients and methods: The study concerned 16 NMO patients, 5 longitudinally extensive transverse myelitis (LETM) patients, 4 optic neuritis patients and 40 MS patients with different patterns of optic nerve and spinal cord involvement. Markers were also evaluated in 21 volunteers and 13 ALS patients. AQP4-IgG was detected by cell based assay. The measurements of NOx levels were done by the Griess method. Total RSNO levels were measured using 4,5-diaminofluorescein. NfH levels were determined by ELISA.

Results: 13 NMO and 1 LETM patients were positive for AQP4-Ab. In relapsing-remitting MS group NOx and RSNO were increased. In secondary progressive MS group only RSNO level was increased. RSNO was the only significant variable associated with spinal cord lesion. NfH level was higher in ALS group compared to secondary progressive MS patients.

Conclusion: AQP4-Ab can be detected in most of NMO patients and should be checked in case of NMO-spectrum disorder. RSNO seems to be a biomarker of spinal cord injury and NfH level reflects extensity of degeneration in demyelinating diseases.

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No: 2064

Topic: 6 — MS & Demyelinating Diseases

The results of visual evoked potentials in patients with neuromyelitis optica in mongolia

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Background: In comparison to the cases with multiple sclerosis cases of neuromyelitis optica are relatively common in Mongolia. These patients develop frequent episodes of retrobulbar neuritis which remains often refractory to high-dose methylprednisolone therapy.

Methods: We studied retrospectively the results of visual evoked potentials in 10 patients with clinically confirmed neuromyelitis optica before and after treatment of high-dose methylprednisolone for 5–7 days. We measured the P100 latency of response potential on the affected side. In 8 cases the thoracic MRI showed an extremely long (stretching from 5 to over 12 vertebra) inflammatory lesion of spinal cord. In 2 cases the MRI investigation was not possible due to financial problems.

Results: Before treatment in 7 (70%) cases of acute retrobulbar neuritis no P100 potential was available. In 3 (30%) cases the P100 latency was significantly prolonged. After treatment the P100 response potential recovered in 9 (90%) of 10 cases with P100 latency measurements of 80–119 ms, but with significant low amplitude and

signs of temporal dispersion. In 1 case the P100 latency increased despite the therapy from 189 ms to 237 ms.

Conclusion: Our measurements show that the acute retrobulbar neuritis episode in patients with neuromyelitis optica cannot be explained with mere demyelination. The absence of P100 response in acute relapse in most cases demonstrates the more prominent axonal damage of optic nerve. These severe causes of neuromyelitis optica in Mongolia should be studied thoroughly in future, and the management of relapse prophylactic treatment must be improved.

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Abstract – WCN 2013

No: 2069

Topic: 6 – MS & Demyelinating Diseases

Spinal cord expression of stathmin-1, SCLIP and SCG10 in experimental autoimmune encephalomyelitis (EAE)

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Background: The role of stathmins is the regulation of microtubule equilibrium and cytoskeleton reorganization. They alternate between assembly and disassembly of the cell with the purpose of correct formation of mitotic spindle.

Objective: We studied the stathmin expression in the spinal cord in EAE.

Material and methods: EAE mice were examined at three time-points (D10, acute, chronic) and compared with controls (D0). Stathmin1, SCG10 and SCLIP mRNA and protein expression was studied using Real Time-PCR and optical and confocal microscopy.

Results: There was reduced mRNA expression in acute phase of stathmin1 ($p < 0.05$); SCLIP was reduced both in acute and chronic phase of EAE ($p < 0.01$). An overall decrease of spinal cord stathmin1 and SCG10 protein expression was noticed in acute phase ($p < 0.001$, $p < 0.05$). However, our sub-analysis in the inflamed white matter showed increased expression of all stathmins ($p < 0.05$, $p < 0.001$, $p < 0.01$). Moreover, stathmin1 was predominantly expressed in NG2+ ($p < 0.001$) and O4+ ($p < 0.05$) cells whereas in lower levels in CNPase+ ($p < 0.05$) cells during the acute phase. SCG10 and SCLIP were expressed in axons already during the acute phase and throughout the entire EAE course though not in controls. In addition, inside the inflammatory lesions SCG10 was co-expressed with APP ($p < 0.05$), a marker of acute axonal damage whereas SCLIP followed the opposite course and was co-expressed with GAP-43 ($p < 0.05$), a marker of axonal regeneration.

Conclusions: The stathmin protein family kinetic in inflammatory demyelinating areas indicates their potential involvement in the underlying pathology and reorganization of the degenerative spinal cord during EAE.

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Abstract – WCN 2013

No: 2044

Topic: 6 – MS & Demyelinating Diseases

Connectivity patterns obtained by emulated vs. conventional resting state fMRI in clinical cohorts – Can parts tell the whole story?

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Background: Resting state (RS) fMRI has received considerable interest in clinical neuroscientific research, as it also allows concluding

on patterns of spatiotemporally consistent neural activity while the brain is at “rest”. With regard to this, Fair et al. (2007) suggested a method how existing datasets could be re-analyzed via extracting specific volumes of conventional (c) block design fMRI data to emulate (e) RS-fMRI.

Objective: We here aimed to investigate the similarity or diversity of results obtained with this approach (eRS) compared to cRS regarding connectivity network identification in a clinical cohort.

Patients and methods: 12 Multiple Sclerosis patients (MS, age: $x = 30.82 \pm 10.8$) and 18 healthy controls (HC, age: $x = 32.28 \pm 6.8$) underwent conventional task-related fMRI and cRS in the same imaging session. In the first step, the total cognitive paradigm has been treated as a RS-dataset, and both datasets were analyzed in total length. Then, the cognitive paradigm was truncated according to the criteria by Fair to emulate RS. The cRS dataset was truncated equally.

Results: Whereas visual inspection of seed based analyses of functional connectivity from the anterior cingulate cortex suggested almost identical patterns in MS patients and HC, higher-level within and between group analyses of eRS vs. cRS revealed distinct differences in connectivity networks.

Conclusion: Generally, using the eRS approach allows the identification of connectivity networks (similar to those obtained using cRS-data) also in diseased brains. However, results of higher-level contrasts yielded different results and thus have to be interpreted with caution.

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Abstract – WCN 2013

No: 2042

Topic: 6 – MS & Demyelinating Diseases

Predictive factors of physical component of quality of life in multiple sclerosis patients

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Background: Multiple sclerosis is a chronic progressive disease with multiple neurological and psychological impairments that can affect all aspects of quality of life.

Objective: The objective of this study was to investigate the predictive factors of physical component of quality of life in MS patients.

Patients and methods: 100 MS patients treated at the Neurology Clinic in Sarajevo were involved. Each patient underwent a complete clinical assessment, including disability status (EDSS), cognitive function (MMSE) and measurement of quality of life (MSQOL-54). Linear regression analyses were performed to identify significant predictors from sociodemographic and clinical characteristics in predicting MSQOL-54 physical and mental composite scores.

Results: The mean age of 100 patients (69% female and 31% male) was 39.88 ± 10.03 . The mean EDSS score was 3.57 ± 1.73 and 72% were of the relapsing–remitting clinical subtype. Linear multivariate regression analyses revealed that presence of pain ($\eta^2 = 0.29$), EDSS ($\eta^2 = 0.27$), sphincter disorders ($\eta^2 = 0.19$), employment status ($\eta^2 = 0.15$), type of disease ($\eta^2 = 0.10$) and age ($\eta^2 = 0.06$) proved to be main predictors of MSQOL-54 physical composite scores ($r^2 = 0.83$). Among the patients with relapsing–remitting type of disease number of relapses ($\eta^2 = 0.260$), EDSS score ($\eta^2 = 0.24$), sphincter disorders ($\eta^2 = 0.17$), presence of pain ($\eta^2 = 0.16$), employment status ($\eta^2 = 0.15$) and age ($\eta^2 = 0.08$) proved to be main predictors of MSQOL-54 physical composite scores ($r^2 = 0.87$).

Conclusion: Assessment of quality of life in MS patients provides additional information on the patient status. Management of pain

and sphincter disorders is highly important in treatment of MS patients.

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Abstract – WCN 2013

No: 2028

Topic: 6 – MS & Demyelinating Diseases

Reduced intrathecal IgG synthesis in the cerebrospinal fluid from natalizumab-treated patients with active multiple sclerosis

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Oligoclonal bands (OCB) have been reported to disappear from the cerebrospinal fluid (CSF) in natalizumab (Nzb)-treated patients with multiple sclerosis (MS) although OCB are considered to persist in MS. Detection of OCB is qualitative evidence of intrathecally produced IgG. We investigated if this Nzb-effect on intrathecal IgG synthesis can be quantitatively assessed by analyzing the fraction of intrathecally produced IgG (IgGIF).

We included 31 MS patients, 17 were untreated (first diagnosis) and from 14 patients CSF was collected to rule out progressive multifocal leukoencephalopathy (PML) during Nzb treatment. From 6 of the 14 Nzb-treated patients CSF data from the first diagnosis were also available. IgG and albumin concentrations of CSF and serum samples were measured by immunonephelometry and used to calculate the CSF/serum quotients and IgGIF (in %) according to Reiber (1998). Quantification of intrathecal IgG synthesis showed a reduced IgGIF in Nzb-treated patients compared to untreated MS patients ($p = 0.0026$). Intraindividual comparisons of baseline and follow-up data in the Nzb-treated group of 6 showed disappearance of intrathecal IgG production in 2 and an overall reduction of the IgGIF ($p = 0.018$).

Quantifying intrathecal IgG synthesis revealed a pronouncedly reduced IgGIF in Nzb-treated MS patients. This corroborates previous reports about disappearing OCB and suggests effectiveness of Nzb therapy on intrathecal IgG production. Since lumbar punctures were performed to rule out PML this observation is limited to Nzb-treated patients with active disease. Clarification of the underlying mechanisms will provide important knowledge about the clinical relevance of this finding and the pathogenesis of Nzb-associated PML.

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Abstract – WCN 2013

No: 2004

Topic: 6 – MS & Demyelinating Diseases

Incidence of multiple sclerosis in the region of Sarajevo before and after the war events

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Introduction: Multiple sclerosis is an autoimmune demyelinating disease and degeneration of axons, in the appearance of which stress has an important role. The goal is to determine the incidence of multiple sclerosis in the region of Sarajevo in the period before and after the war (1986–2012).

Material and methods: We analyzed all new cases of multiple sclerosis in the region of Sarajevo in the period January 1986–December 2012. Diagnostic criteria included anamnesis, clinical presentation, MRI

findings and evaluation of the spine, evoked potentials-visual and cerebrospinal fluid examination.

Results: During the period 1986–1990, there were 45 new cases of MS, in the period 1991–1995, there were 27 new MS cases, from 1996 to 2000 107 new cases were registered of MS, and from 2001–2005 there were 150 new cases of MS. In the period 2006–2010, there were 174 new cases of MS. In 2011 there were 34 new cases of MS and during the 2012, 41 new cases. Stress as provoking factor was present in the first five-year period at 11.1% in the second at 18.5%, in the third with 19.6%, in the fourth five-year period at 36.5% of cases and in the final at 43.8% of cases.

Conclusions: In the period after the war, the MS in the region of Sarajevo has increased in incidence. Multiple sclerosis in the region of Sarajevo is more frequent in women than in men, most frequently in the age 20–40 years. But it also tends to occur in a relatively advanced age, after 50 years.

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Abstract – WCN 2013

No: 1996

Topic: 6 – MS & Demyelinating Diseases

Multiple sclerosis prevalence study: The comparison of two coastal cities, located in the Black Sea and the Mediterranean Sea in Turkey

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Background: The studies about Multiple Sclerosis (MS) epidemiology have increased in the last years. The studies, we conducted in Turkey, which have been compared with new studies were made in Turkey. These studies show that there are also distinct variations between the same geographical areas.

Objective: We carried out this study in order to determine the prevalence and to compare the difference between Mediterranean and Black Sea regions.

Patients and methods: This descriptive, cross-sectional, door-to-door survey was carried out in the Mediterranean coastal city Gazipaşa and the Black Sea coastal city Artvin in 2012. All people, who live in the city, were visited and interviewed in their houses. The center town of Artvin and Gazipaşa had been screened by door to door. A population of 16.116 people was screened in Artvin and 13.451 of the people were screened in Gazipaşa. Poser criteria were used as diagnostic criteria in the study.

Results: Seven patients were diagnosed as clinically definitive MS with the definitive anamnesis and MR investigation. Female to male ratio was 1.33. The prevalence of MS was found to be 52.0/100.000 in **Gazipaşa**.

Three female patients were diagnosed as clinically definitive MS. The prevalence of MS was found to be 18.6/100.000 in **Artvin**.

Conclusion: According to the results, the countries in the Mediterranean area is also now in the high MS prevalence zone. More than 2 times of the difference was found between prevalences of Artvin and Gazipaşa. Our recent studies, conducted in Turkey, provide evidence that the latitude gradient in the distribution of MS is disappearing.

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Abstract – WCN 2013

No: 2019

Topic: 6 – MS & Demyelinating Diseases

Flow-cytometry and the detection of anti-natalizumab NAB in multiple sclerosis

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Background: Natalizumab-neutralizing antibodies (NAB) occur in approximately 5–15% of natalizumab-treated multiple sclerosis (MS) patients. In patients with persisting NAB natalizumab treatment has to be terminated.

Objective: Since high-titre NAB are strongly associated with persistence of NAB, we investigated if determination of saturation levels of alpha-4 integrins with natalizumab has the potential to discriminate early patients with NAB.

Methods: Cell-bound natalizumab and the natalizumab saturation of alpha-4 integrins on T-cells from 11 MS patients were detected using a monoclonal anti-human IgG4 antibody by flow cytometry before starting and after 4, 8, and 12 weeks of therapy. Natalizumab saturation (in %) was determined by relating median fluorescence intensities (MFI) of in vivo bound natalizumab to the MFI after in vitro incubation with saturating amounts of natalizumab. Determination of serum NAB was performed by ELISA.

Results: In 8 patients the median natalizumab saturation of T cells over 4, 8, and 12 weeks of natalizumab treatment reached 75% (range 65–86%; SD 6%) and was associated with no NABs. Three patients showed differences in the natalizumab saturation with a clear reduction in 2 patients after 8 weeks and a pronounced decline approximating baseline levels in all three patients after 12 weeks. High- and low-titre NAB were equally effective in reducing the cellular natalizumab saturation.

Conclusion: Monitoring the natalizumab saturation on T cells can serve as an early and sensitive method to identify patients with a reduced treatment effect due to NAB. Interestingly, both high- and low-titer NAB equally effectively reduced the cellular natalizumab saturation.

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Abstract – WCN 2013

No: 2012

Topic: 6 – MS & Demyelinating Diseases

Validation of a screening to detect cognitive and psychoaffective deficits in children and juveniles with multiple sclerosis:

The MUSICADO study

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Background: Neurocognitive deficits may already appear in children affected by multiple sclerosis (MS). The most vulnerable areas include attention, visual-perceptual abilities, executive functions, and language. However, these deficits are mostly underrecognized since there is a lack of specific screening tools which might be used during regular clinical visits for children and adolescents.

Objective: The aim of the MUSICADO-study is to validate a modified screening tool (MUSIC = MULTIPLE Sclerosis Inventory of COgnition) adapted for ADOLescents. Additional clinicometric methods are used to validate this measure together with a questionnaire for depression and fatigue.

Patients and methods: A number of 100 healthy controls and 100 patients, aged between 12 and 18 years will be included with and without treatment. The adopted psychometric tool MUSIC and additional methods to assess IQ, attention, visual-perceptual abilities, executive functions, verbal skills, quality of life, depression, and fatigue are performed at one

visit by a psychologist. The duration of the tests is about 2 to 2.5 hours. Currently, 2 study sites in Austria and 22 study sites in Germany are registered. The study has started in May 2012 and will end in December 2013 until the planned amount of patients has been included.

Results and conclusion: We aim to establish a screening tool to detect cognitive deficits and related comorbidities at an early stage of the disease. Such a tool may help to apply interventions timely.

A detailed study-protocol and data about the already included patient cohort are presented in this contribution.

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Abstract – WCN 2013

No: 1959

Topic: 6 – MS & Demyelinating Diseases

Extensive semi-automated gait analysis by Zebris FDM during treatment of multiple sclerosis associated gait disturbances with PR-fampridine

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The prolonged-release PR-formulation of fampridine (PR-fampridine; syn. modified or sustained-release fampridine, dalfampridine extended release) has been approved for the treatment of walking impairment in multiple sclerosis (MS) patients with walking disability. Pivotal trials showed a significant effect of PR-fampridine on the Timed 25 Foot Walk (T25FW). However, the relevance of this primary outcome measure and its correlation with gait improvement needs further evaluation.

To investigate the effect of PR-fampridine treatment on gait disturbance parameters obtained by an extensive semi-automated gait analysis was the purpose of this study.

Zebris FDM is a semi-automated gait analysis system which uses a 300 × 60 cm mat containing more than 22,500 pressure sensors. This analysis system has been evaluated broadly and is commonly applied by physiotherapists in rehabilitation centres. We included 20 patients with MS-associated walking disability (EDSS 4.0 to 7.0) and performed a gait analysis to determine gait velocity and variability of gait velocity before and during PR-fampridine treatment.

Treatment with PR-fampridine led to a sustained improvement of gait velocity and variability of gait velocity in 10 out of 20 patients for at least 6 months. Two patients had to discontinue treatment due to side effects (nausea, headaches) despite improvement. Seven patients showed no improvement after initiation of fampridine-SR. One patient had to stop treatment due to vertigo before assessment of gait. Both gait parameters obtained by semi-automated gait analysis correlated tightly with the T25FW test.

We observed a measurable positive effect of PR-Fampridine on MS-associated gait disturbances which is relevant in daily life in patients with gait disturbances.

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Abstract – WCN 2013

No: 1986

Topic: 6 – MS & Demyelinating Diseases

The relationship between the internal capsule DTI parameters and cognition in relapsing remitting multiple sclerosis (RR MS) patients

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Objective: To assess the relationship between cognition and internal capsule lesion distribution and frequency in patients with relapsing–remitting multiple sclerosis (RR MS).

Methods: We analyzed 37 patients with definitive diagnosis of RR MS and 37 gender-matched with the same age range control subjects. Conventional MRI and DTI were used to assess the relevance of brain lesion location for cognitive impairment all of the study subjects. The Rao Brief Repeatable Battery was administered to determine the cognitively impaired. Statistical comparison was conducted using repeated measures ANOVA type analysis to detect the relationships between the DTI and Brief Repeatable Battery test.

Results: We observed strong positive correlation between cognition and FA and ADC values which is in agreement with the observation of a statistically significant reduction in FA values for RR MS patients compared to the control group ($p < .0001$). Moreover, there exists a significant inverse relationship between the age FA values ($p = .0027$). On the other hand, we observed that ADC values are significantly higher for RR MS patients compared to the control group ($p < .0001$). Low scores of the Symbol Digit Modalities Test correlated with FA and ADC values in these regions.

Conclusions: This supports the notion of a functional (multiple) disconnection between gray matter structures, secondary to damage located in WM areas, as one of the most important mechanisms leading to cognitive impairment in MS.

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Abstract — WCN 2013

No: 1781

Topic: 6 — MS & Demyelinating Diseases

Balo's concentric sclerosis: Report of four cases and literature review

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Background: Balo's concentric sclerosis (BCS) is a rare variant of multiple sclerosis (MS), differing from typical MS in terms of initial presentation, symptom severity and radiological features.

Objective: We report four patients with BCS. The diagnostic process and the follow-up are described, with a brief literature review.

Material and methods: We studied four patients (three women and a man) affected by BCS. All patients underwent a brain and spine MRI, visual evoked potentials (VEP) and cerebrospinal fluid (CSF) study.

Results: The age of onset varied from 17 to 47 years. The symptoms at onset were: acute left-sided weakness and numbness in one patient, diplopia and progressive visual field restriction in two patients, and seizure and dysarthria in the last patient. Three patients had a history of relapsing–remitting symptoms, while one patient had a monophasic course. Expanded disability status scale (EDSS) scores were between zero and one. Brain MRI revealed in all patients, periventricular lesions with onion-like structure in T2 and FLAIR sequences with a diameter exceeding 2 cm. Spine MRI, revealed lesions in only two patients. VEP showed optic neuritis in one patient. The CSF study revealed the presence of IgG oligoclonal bands in one case. The patients improved after intravenous steroids and were diagnosed with MS according to MS criteria 2010.

Conclusion: BCS is classically considered as a severe variant of MS with a rapid and malignant course leading to severe irreversible disability or death. However, our patients had a mild to moderate disease course at follow-up. Few studies reported similar findings.

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Abstract — WCN 2013

No: 1985

Topic: 6 — MS & Demyelinating Diseases

A case of neurofibromatosis type I and relapsing–remitting multiple sclerosis association

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Background: Neurofibromatosis type I (NF1) associated with multiple sclerosis (MS) is a rare condition. Only thirteen cases have been described in the literature, especially with the progressive form of MS.

Objective: We report the case of a woman with a past history of NF1 who developed relapsing remitting MS.

Case report: A 31-year-old woman, diagnosed since childhood with NF1 on the basis of multiple “café au lait” spots and cutaneous neurofibromas, presented gait impairment appearing within 3 months. Neurological examination revealed ataxic gait associated with lower limb's spasticity, hyperreflexia and bilateral Babinski sign. Expanded disability status scale (EDSS) score was 2.5. Brain and spinal cord MRI showed bilateral T2-weighted lesions in the periventricular regions, corpus callosum, cerebellum and brainstem, with gadolinium enhancement. There were no IgG oligoclonal bands on the cerebrospinal fluid (CSF) study. MS was diagnosed referring to the 2010 Mc Donald's Criteria.

Discussion: NF1 is an autosomal dominant disorder; the mutated gene is located on chromosome 17q11.2. The gene for oligodendrocyte myelin glycoprotein (OMgp), a membrane glycoprotein which might be one of the target antigens of the autoimmune attack in MS, is embedded within intron 27b of the NF1 gene. Moreover, patients with NF1 with gross deletions may lose at least 11 functional genes, including OMgp. This association has been hypothesized to be related to mutations in the neurofibromin protein or (OMgp) gene.

Conclusion: Besides the genetic hypothesis, a casual relationship between MS and NF1 is possible with a higher risk of MS among patients with NF1.

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Abstract — WCN 2013

No: 1955

Topic: 6 — MS & Demyelinating Diseases

Prevalence of autoimmune Thyroiditis in an Italian multiple sclerosis cohort

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Background: Multiple sclerosis (MS) can be co-morbid with other autoimmune disorders, such as autoimmune thyroid diseases. In Western countries, the prevalence of Autoimmune Thyroiditis (AT) is about 5% in the general population (Bagnasco et al., 2006). In MS patients, the prevalence of AT tends to increase: in a Spanish cohort, it reaches about 11.8% (Munteis et al 2007). In Italian MS patients and, in particular, in the Campania region, this data is unknown.

Objective: To assess the prevalence of AT in a cohort of MS patients, all belonging to the Campania region; to compare the Expanded Disability Status Scale (EDSS) of patients with or without AT to evaluate the possible effect of thyroid function on the degree of disability.

Patients and methods: 82 (35 M, 47 F, mean age 41.6 years) consecutive MS patients who accepted to perform blood sample collection, thyroid echography and endocrinological visit in our centre. One of us assessed the EDSS in all patients.

Results: Out of 82 patients, 19 (21.95%) were diagnosed with AT. Mean EDSS value is 1.5 in patients with normal thyroid function and 2.0 in AT patients.

Conclusions: AT is much more frequent in MS patients than in general population and in Italian MS patients compared to other European series. Although the sample is not sufficiently numerous to identify a clear correlation between high values of EDSS and thyroid dysfunction, our data show a trend in that direction. So, management of MS people should include an endocrinological screening to verify thyroid function.

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Abstract – WCN 2013

No: 1886

Topic: 6 – MS & Demyelinating Diseases

Asymptomatic electrophysiologic carpal tunnel syndrome in diabetes: Ultrasonographic study

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Background: Carpal tunnel syndrome (CTS) is considered more common in diabetes mellitus (DM) patients. Asymptomatic electrophysiologic CTS is frequently found on screening nerve conduction study (NCS) for DM neuropathy, but the pathophysiology has been little known. We investigated the symptoms, electrophysiologic findings and median nerve ultrasonography (US) findings of DM patients, and compared those between no CTS, asymptomatic CTS and symptomatic CTS group.

Method: Thirty four hands with DM were prospectively assessed by three clinical scales, one electrophysiological scale and median nerve cross section area (CSA) through ultrasonography. The clinical scales adopted were the Boston carpal tunnel syndrome questionnaire (BCTQ), quantitative clinical scale by Simovic (Simovic clinical scale) and Historical-Objective (Hi-Ob) scale. The scale for electrophysiological assessment is the American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) classification. US was performed in all participants, and CSA of the median nerve at carpal tunnel inlet site was evaluated.

Results: There is no valid difference of median nerve bland scales between asymptomatic (2.36) and symptomatic (2.73) electrophysiologic CTS group. The median nerve CSA in non electrophysiologic CTS ($0.086 \text{ cm}^2, \pm 0.012$) was smaller than others ($p < 0.05$), and there is no valid difference of median nerve CSA between asymptomatic ($0.117 \text{ cm}^2, \pm 0.023$) and symptomatic electrophysiologic CTS group ($0.123 \text{ cm}^2, \pm 0.031$).

Conclusion: These findings suggest that asymptomatic electrophysiologic CTS in DM could be a prodromal phase of symptomatic CTS, because they may have the same pathophysiology. Therefore clinicians have to carefully monitor the DM patients with asymptomatic electrophysiologic CTS.

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No: 1905

Topic: 6 – MS & Demyelinating Diseases

Does OCT predict conversion to MS?

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Background: Optical coherence tomography (OCT) is a simple high resolution noninvasive technique to quantify the thickness of retinal

nerve fiber layer (RNFL) and an indirect measurement of axonal damage in multiple sclerosis (MS).

Objectives: To determine whether RNFL thickness predicts conversion to clinically definite multiple sclerosis (CDMS) in patients with clinically isolated syndromes (CIS) over the subsequent three years.

Methods: Patients with CIS (McDonald criteria 2005) were recruited in two months after the event and underwent a complete neurological examination. High field brain MRI and cervical MRI were performed within two months of the first attack and after three years. An ophthalmological evaluation including visual acuity and optical coherence tomography (Stratus) was done within the first week after diagnosis and after three years. The sensitivity and specificity of OCT to predict conversion to CDMS were analyzed.

Results: 23 patients with CIS with a mean EDSS of 1.72 were recruited and followed during three years. Eight patients did not have demyelinating lesions on baseline cranial MRI. 54.2% of all patients showed the presence of at least one quadrant of an optic nerve with a decreased RNFL thickness.

The presence of at least one quadrant of an optic nerve with a RNFL thickness at a $P < 5\%$ cut-off value had a sensitivity of 69% and a specificity of 60% for predicting conversion to CDMS in a period of three years.

Conclusion: Axonal damage in very early stages of the disease may be useful for predicting conversion in multiple sclerosis and useful for follow-up.

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Abstract – WCN 2013

No: 1940

Topic: 6 – MS & Demyelinating Diseases

Disease-specific mechanisms in cortical multiple sclerosis lesions

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Multiple sclerosis (MS) is an immune-mediated demyelinating disorder of the central nervous system. Traditionally, research focused on white matter lesions but within the last years, cortical lesions gained importance. Although inflammation appears to play a fundamental role in the formation of cortical MS lesions, disease-specific mechanisms leading to plaque-like primary demyelination and neurodegeneration are poorly understood.

We aim at identifying MS-specific mechanisms by combining neuropathological analysis of inflammatory and neurodegenerative diseases with gene expression studies.

For immunohistochemistry, archival formalin-fixed and paraffin-embedded autopsy tissue was used. Apart from cortical MS tissue, we also included cortical tissue from other inflammatory (tuberculous meningitis, Rasmussen's encephalitis, B-cell lymphoma, and meningitis) and neurodegenerative (Alzheimer's disease) diseases. Whole-genome microarrays were performed using microdissected cortical material from MS, tuberculous meningitis, and Alzheimer's disease patients and control cases.

Immunohistochemistry showed that despite complex and fulminant immune responses in many investigated diseases, primary demyelination was only present in MS. Using microarrays, we identified 301 genes being

differentially expressed in cortical MS lesions. Of these, 80% were assigned to inflammation, oxidative stress, tissue injury, DNA damage/repair and regenerative processes. Immunohistochemically, we confirmed that significantly more oxidatively damaged neurons were present in cortical MS lesions than in other examined diseases. Additionally, neurons and oligodendrocytes stained positive for DNA strand breaks. Interestingly, oxidative stress-mediated tissue damage seemed to be driven by NADPH oxidases rather than by nitric oxide synthases.

We show that MS-specific primary demyelination is driven by mechanisms of tissue injury that differ from any other investigated inflammatory and neurodegenerative disease.

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Abstract – WCN 2013

No: 731

Topic: 6 – MS & Demyelinating Diseases

Psychosocial complications of multiple sclerosis

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Background: Multiple sclerosis affects individuals diagnosed on both a physical and cognitive level, as well as an emotional one. Depression, anxiety, grief, other mental health concerns, and environmental factors all affect a patient's life with MS. Understanding this psychosocial impact is necessary to provide the best possible care for the MS patient.

Objectives: Recognizing the emotional flux of an MS patient is essential to learn how to both treat and prevent these challenges from impacting the patient physically. Psychotherapy provides insight into an MS patient's life. This work demonstrates the psychosocial challenges many patients face and analyzes how mental health and environmental problems can both be a precursor to and aggravated further by MS.

Patients and methods: Three case study examples of MS patients will be used to demonstrate psychosocial impact. All patients engaged in psychotherapy for at least one year to examine concerns related to their illness.

Results: The psychosocial impact on those living with MS is clear but inconsistent. These individuals along with many others will continue to need emotional support throughout their illness.

Conclusions: Examining the psychosocial component of patients living with MS allows medical professionals to take a more comprehensive look at treating the illness. While there is always no causation between emotional and physical health, the two often correlate. Research examining what happens physically to the patient when emotional health is exacerbated will go a long way in helping medical professionals understand the full impact of emotions on MS.

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Abstract – WCN 2013

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Topic: 6 – MS & Demyelinating Diseases

Identification of novel candidate autoantigens in multiple sclerosis by expression cloning

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Background: Multiple sclerosis (MS) is an immune-mediated disease characterized by demyelination of the central nervous system. In some part of MS lesions, IgG (antibody) deposition is found. Although many

components of the myelin sheath are suspicious for the targets of autoantibody, the real target molecules are unknown.

Objective: To elucidate whether there is a novel specific autoantibody in serum or cerebrospinal fluid (CSF) samples from MS patients using the SEREX (serological identification by cDNA expression cloning) and ELISA (enzyme-linked immunosorbent assays) techniques.

Material and methods: The phage expression library constructed from the U-87 MG glioblastoma cell-line was used in SEREX screening. Serum and CSF samples from 41 MS patients during the relapse or remission phases were obtained. Serum samples of 43 normal controls (NC) and CSF samples of 45 disease controls (DC) were used in ELISA.

Results: Immunoscreening of cDNA expression libraries with serum from a relapsing–remitting MS patient led to the isolation of several independent antigens including DDX39 (DEAD box polypeptide 39). The results of ELISA revealed that the levels of anti-DDX39 antibody were significantly higher in sera from MS patients than NC, but were not significantly different between CSF samples from MS patients and NC. There were significant differences in serum anti-DDX39 antibody levels during the relapse and remission phases.

Conclusion: Anti-DDX39 antibody is a possible target molecule for MS and could be used as a diagnostic biomarker. DDX39 may play some role in the pathogenesis of MS.

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Abstract – WCN 2013

No: 1883

Topic: 6 – MS & Demyelinating Diseases

Epidemiological analysis of multiple sclerosis in Goiás and in Brazil from 2008 to 2011

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Background: Multiple sclerosis (MS) is a chronic inflammatory demyelinating disease, autoimmune, confined to central nervous system. It affects about two times more women than men and causes great socioeconomic impact by having higher incidence among young adults, which may lead them to early failure.

Objectives: To describe and compare profile of patients hospitalized due to MS in Goiás and Brazil, according to sex and age, from 2008 to 2011, noting correlation with the literature.

Material and methods: An observational, descriptive, retrospective epidemiological approach with qualitative and quantitative data from hospital admissions in Brazil and Goiás, from 2008 to 2011, registered by Brazilian Health System based on database (DATASUS).

Results: Of the total hospitalizations due to MS in Brazil (6838), 2087 (30.5%) were in males and 4751 (69.5%) in females. Of this total, 4733 (69.2%) occurred between 20 and 49 years. Goiás had 219 hospitalizations, 58 (26.5%) in males and 161 (73.5%) in females. In terms of age, 164 (74.9%) were between 20 and 49 years.

Conclusion: The state of Goiás was responsible for 3.2% of admissions for MS in Brazil. In both scenarios MS predominates among women. In Brazil, there were 2.3 women for every man hospitalized by MS, while in Goiás, that relation was 2.8, which is equivalent to the literature. Age group was also in accord to the literature, varying from 20 to 49 years. The highest number of admissions occurred between 30 and 39 years, the age group with the largest percentage of economically active population in Brazil, justifying the impact of MS.

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Abstract – WCN 2013**No: 1848****Topic: 6 – MS & Demyelinating Diseases****Demyelinating peripheral neuropathy without conduction block in two patients with tumor necrosis factor α antagonist therapy**

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Background: Tumor necrosis factor α (TNF- α) antagonist has been successfully used in patients with various autoimmune diseases. Demyelination of the nervous system had been reported as a side effect occurring with these agents. However, polyneuropathy associated with TNF- α antagonist has been rarely described.

Objective: To report our two patients with demyelinating polyneuropathy receiving TNF- α antagonist therapy.

Patient and methods: Two patients (two women, patient 1 was 26 years old, patient 2 was 57 years old) who developed a demyelinating peripheral polyneuropathy during TNF- α antagonist therapy. Their clinical findings and electrophysiological results were retrospectively reviewed.

Results: Patient 1 was a 26-year-old-woman. She has polyarthritis for 4 years and received etanercept for 7 months. Weakness and numbness occurred on her distal and proximal extremities progressively. Dysphagia, dysarthria and diplopia were presented in parallel. She underwent intravenous immunoglobulin (IVIG) therapy (0.4 g/kg/day, 5 days) and improved completely in a month. Patient 2 was a 57-year-old-woman with a 6-year history of rheumatoid arthritis and started on infliximab therapy. Two months after initiating infliximab, she developed numbness in both hands and weakness in both lower limbs. She was treated with IVIG (0.4 g/kg/day, 5 days) and almost improved with remaining mild gait and sensory disturbance. The electrodiagnostic studies revealed demyelinating sensory and motor peripheral polyneuropathy without conduction block. Anti ganglioside antibody was absent in both patients.

Conclusion: Although previous cases after treatment of TNF- α antagonist revealed demyelinating peripheral polyneuropathy with conduction block, we hereby report novel findings in electrodiagnostic studies in two patients.

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Abstract – WCN 2013**No: 1866****Topic: 6 – MS & Demyelinating Diseases****Analyzing a regional MS registry and presenting the importance of early treatment after diagnosis with long term clinical course evaluation**

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Background: Between the year 1994 and 1995 the Immunomodulatory therapy (IMT) for patients with Multiple Sclerosis (MS) was introduced. The representative Pivotal studies for those therapies included patients with a long disease duration averaging five to seven years.

The introduction of IMT, following the Poser criteria for a diagnosis of MS, has resulted in a significant delay of access to therapy for many patients. Since 2001 several studies and publications (CHAMPS, ETOMS, BENEFIT, PreCISE) demonstrated the benefits of early MS treatment.

Objective: To present the importance of early treatment for MS patients after diagnosis with a long term evaluation of the clinical course.

Material and methods: We present a regional MS registry including 304 patients. 128 are considered an early therapy group where the

IMT started since 1995 within one year after the diagnosis of MS. 78 patients in this group received IMT within six months after the diagnosis of MS. Another subgroup is presented with 66 patients who received IMT from 1995 till 2005. The observation period of all these patients was between 1995 and 2011.

Results: The analyses of these patient groups demonstrate good compliance, adherence and no diagnosis revision. All patients showed good medication compatibility and tolerability, low Relapse-Rate after beginning of therapy and low disease progression measured by Expanded Disability Status Scale (EDSS).

Conclusion: Our study demonstrates the importance and benefits of early treatment, which is associated with a good clinical course of MS. MRI results from 1995 also demonstrated good diagnostic accuracy.

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Abstract – WCN 2013**No: 1865****Topic: 6 – MS & Demyelinating Diseases****Increased frequency of multiple sclerosis, the influence of environmental and immunological factors**

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Background: For decades, consistent geographical pattern of MS, with prevalence and increased risk in countries farther from Ecuador in both directions has been recognized.

Bolivia is situated in the central part of South America, between the meridians 57° 26' and 69° 38' longitude west of Greenwich and the parallels 9° 38' and 22° 53' south latitude.

Traditionally the frequency of multiple sclerosis in this area has been described as low; we have observed an increase in frequency in recent years.

Objective: To analyze the increase in the frequency of multiple sclerosis in a clinic for neurological diagnosis.

Material and methods: 16,779 medical records were reviewed in a clinic for neurological diagnosis from January 1992 to December 2012. Patients who met the clinical criteria and who had brain MRI compatible with multiple sclerosis were included.

Results: We identified 26 patients with multiple sclerosis. 19 female and 7 male, median age at diagnosis 31.6 years for women and 32 years for males. 1 (3.8%) case from 1992 to 1996, 3 (11.5%) cases from 1997 to 2001, 8 (30.7%) cases from 2002 to 2006 and 14 (53%) cases from 2007 to 2012.

Conclusion: Changes in environmental or immunological profiles which have been developed in our country may have contributed to the increased frequency of multiple sclerosis in the last 5 years.

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Abstract – WCN 2013**No: 1044****Topic: 6 – MS & Demyelinating Diseases****Cerebrospinal fluid oligoclonal IgG bands in Japanese patients with clinically isolated syndrome**

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Background: Oligoclonal IgG bands (OCBs) in the cerebrospinal fluid (CSF) in clinically isolated syndrome (CIS) predict multiple sclerosis (MS) development in European patients.

Objective: Evaluating the potential of OCBs in predicting MS development in Japanese patients.

Patients and methods: Thirty-four Japanese patients with CIS were consecutively enrolled in this prospective study conducted from Feb 2006 to July 2012 with a mean follow up period of 38 months. Anti-aquaporin (AQP) 4 antibodies were examined using a cell-based assay.

Results: Five patients developed MS (14.7%); of these, 2 revealed relapses of the different lesions and 3 revealed other lesions on brain MRI. Moreover 21 were classified as still having CIS (61.8%). The remaining 8 patients had a neuromyelitis optica-related disorder (NMOrd); 3 patients demonstrated anti-AQP4 antibodies, and 5 revealed a centrally located long spinal cord lesion on MRI (1 patient demonstrated both lesions). OCBs were observed in CSF at the first visit in 2 of 5 (40%), 3 of 21 (14.3%), and 3 of 8 (37.5%) patients who revealed conversion to MS, non-conversion, and NMOrd, respectively.

Conclusion: Two of 5 patients with CIS and OCBs developed MS for only for 38 months; moreover, NMOrd was observed in 8 of 34 with CIS (23.5%). This study suggests that CSF-OCBs could be helpful in predicting MS development in Japanese patients with a low incidence of OCB (47.6% of different our series) and that Japanese patients with CIS have a low risk of MS development.

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Abstract – WCN 2013

No: 1845

Topic: 6 – MS & Demyelinating Diseases

Impact of HLA DRB1*1501 gene and oligoclonal bands in multiple sclerosis. Clinical radiological/immunogenetic correlations

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Introduction: The aim of this study was to determine the value of immunogenetic risk factors and estimate their relationship with clinical and radiological characteristics and disability status of patients with multiple sclerosis in a population in northern Algeria.

Material and methods: This is a prospective study concerning 60 patients with MS. We noted the age, sex, and clinical manifestations since the beginning of the disease. Paraclinical analyzes systematically included in our patients, immunology of CSF, brain and spine MRI. We evaluated the frequency of HLA DRB1*1501.

Results: The mean age was 34.4 years with a sex ratio F/M = 2. The HLA DRB1*1501 was positive in 30% of cases, with an odds ratio of 2.3. In the HLA DRB1*1501 positive group, 38.9% were male and 26.2% were female. We observed the presence of oligoclonal bands in 85% of patients, 25% of the HLA DRB1*1501 positive group. No significant association between clinical and genetics has been found. The brain stem lesion on MRI was associated with the presence of HLA DRB1*1501 ($P < 0.01$). The relapsing multiple sclerosis represented 83.3% of the HLA DRB1*1501 positive group. The mean EDSS score was 1.2 in the HLA DRB1*1501 positive group and 0.91 in the negative group ($P = 0.41$).

Conclusion: The presence of oligoclonal bands in the CSF was associated with many exacerbations of the disease. The brain stem-lesion on MRI was significantly associated with the presence of HLA DRB1*1501.

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Abstract – WCN 2013

No: 1715

Topic: 6 – MS & Demyelinating Diseases

Uric acid serum levels change in the earliest phase of demyelization

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Objectives and methods: In order to examine the endogenous antioxidant values in the earliest phase of demyelization, we have determined uric acid (UA) serum values in the patients with clinically isolated syndrome (CIS) and relapsing remitting multiple sclerosis (RRMS), regarding their clinical disability, measured by Extended Disability Status Scale (EDSS), Magnetic Resonance Imaging (MRI), disease duration, gender and other parameters.

Results: The UA levels were lower in CIS and RRMS patients than in control group, whether male or female ($p < 0.05$). The UA levels were decreased in RRMS compared to CIS patients ($p < 0.05$). Regarding EDSS, MRI and disease duration, obtained values of UA were higher in both study groups in patients with lower EDSS, lower MRI lesion number and shorter disease duration ($p < 0.05$). The greatest significance in decreased UA levels was observed in female compared to male patients, in both study groups ($p < 0.05$). The results suggest negative linear correlation between UA levels and disease duration, EDSS and MRI in CIS ($p < 0.01$), with the same correlation between UA levels and disease duration in RRMS patients ($p < 0.01$). There was also significant correlation between UA levels and EDSS in RRMS patients ($p < 0.01$).

Conclusions: The obtained results point to the importance of endogenous antioxidants, UA, in the outbreak and course of neuroinflammation. This could be favorable for the new pathogenetically conditioned neuroinflammatory therapy concepts which do not initially rely only on immunomodulatory, but also on the antioxidative effects.

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No: 1519

Topic: 6 – MS & Demyelinating Diseases

Cerebrospinal fluid transferrin levels are reduced in patients with early multiple sclerosis

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Background: Abnormally high cerebral iron deposition has been described in multiple sclerosis (MS), which is mainly evidenced by applying advanced MRI techniques. Up to now it is not completely clear whether alterations of iron metabolism markers in body fluids are present in MS and if these markers are related to clinical and imaging parameters.

Objective: We aimed to investigate if iron metabolism markers in cerebrospinal fluid (CSF) and serum of patients with clinically isolated syndromes (CIS) and MS differ from those of control patients with other neurologic diseases of non-inflammatory aetiology (NC).

Methods: We included non-anaemic patients with CIS/MS ($n = 77$, CIS 62; MS 15) and 69 NC. None of the patients received any immunomodulatory treatment at time of lumbar puncture. We nephelometrically assessed serum levels of ferritin, transferrin and soluble transferrin receptor and CSF levels of ferritin and transferrin. The serum transferrin saturation was calculated from serum ferritin and transferrin levels.

Results: CSF transferrin levels were significantly reduced in CIS/MS compared to NC ($p < 0.001$). Higher CSF transferrin levels correlated with lower physical disability ($r = -0.3$, $p < 0.01$). There were no significant differences comparing CIS/MS and NC regarding CSF ferritin and serum ferritin, transferrin, soluble transferrin receptor and the transferrin saturation.

Conclusion: Our results indicate that CSF transferrin levels are altered in early phases of MS with some relation to physical disability. Further longitudinal studies are currently undertaken to investigate if CSF

transferrin levels are predictive of developing abnormally high cerebral iron deposition as measured by MRI.

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Abstract – WCN 2013

No: 1689

Topic: 6 – MS & Demyelinating Diseases

The benefits of visual evoked potential examination in the diagnosis and prognosis of multiple sclerosis

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Background: Visual evoked potentials (VEPs) have been used in the diagnosis of multiple sclerosis (MS). But, in the revised form of McDonald criteria it was not included.

Objective: We wanted to investigate the relation between the VEPs and the clinical types and the prognosis of MS.

Patients and methods: In 354 MS patients VEPs were evaluated. The relation between VEP results and neurologic findings at onset of the disease, IgG index, oligoclonal band positivity, EDSS scores at onset, 1st year, 5th year, 10th year and transition from clinical isolated syndrome to MS was determined. The patients were grouped according to their initial neurological findings such as optic neuritis, supratentorial involvement, brain-stem–cerebellar involvement and spinal involvement. The VEP findings were classified as monocular, bilateral pathology and no pathology.

Results: VEP abnormalities were more frequent in patients in optic neuritis group as expected. VEP abnormalities were less in the spinal group. In patients with bilateral VEP abnormality EDSS scores at onset, 1st, 5th and 10th years were higher than the patients with monocular abnormality and no abnormality. No relation was found between the VEP abnormalities and the clinical types of the disease and transition from clinical isolated syndrome to MS.

Conclusions: We suggest that VEP examination can predict the evolution of disability in MS patients.

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Abstract – WCN 2013

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Topic: 6 – MS & Demyelinating Diseases

Characteristics of headaches in cases diagnosed with neuro-behçet's disease

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Introduction and aims: Behçet's disease is a vascular-inflammatory chronic disease with an obscure pathogenesis. Primary neurological involvement is in the form of non-structural headache (migrainous HA), subclinical NBS, cerebral dural sinus vein thrombosis (extra axial), MSS involvement (intraaxial), neuro-psycho-behçet and peripheral nervous involvements.

Method: In this study, patients applying to the outpatient Behçet clinic between 2000 and 2013 were examined retrospectively. Headache severity was evaluated using VAS scales. The types of headaches of the patients diagnosed with headaches in Behçet's disease were made in compliance with the 2011 diagnosis criteria of the International Headache Society. Localization of headaches, the existence of aura, frequency, severity, duration of attacks, neurological examinations, accompanying diseases, clinic, methods of imaging, treatment, and treatments for prophylaxis in the acute and chronic period of the patients who applied to the clinic were reviewed.

Conclusion: Different from Behçet's syndrome, in the 50% of the cases it was found in compliance with intracranial increase in pressure. Lumbar function opening pressures changed between 250 and 350 mm of water. In the imaging examinations (cranial MRG, MRA) the existence of venous thrombosis (43%), and the ratio of parenchymal involvement (50%) were rather high.

Corticosteroid infusion is given to neuro-behçet patients during attacks and immunosuppressant treatment added following this. Nearly 90% of the cases benefited from the treatment given.

Discussion: Headaches are the most common neurological complaint of cases diagnosed with neuro-behçet. Cases were given appropriate treatment for their clinical findings and symptoms and nearly all of them showed full or partial remission.

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No: 1761

Topic: 6 – MS & Demyelinating Diseases

Characteristics of headaches in cases diagnosed with behçet's disease

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Introduction and aim: Behçet's disease (BD) is a chronic disease which progresses with the involvement of many organs and systems. It is a vascular-inflammatory disease with an obscure pathogenesis. Characteristics of headaches in patients diagnosed with Behçet's disease, their differences from other types of headaches, methods of imaging and treatment were planned to be assessed in accordance with literature.

Method: In this study, patients applying to the outpatient Behçet clinic between 2000 and 2013 were examined retrospectively. The types of headaches of the patients diagnosed with headaches in Behçet's disease were made in compliance with the 2011 diagnosis criteria of the International Headache Society. Localization of headaches, the existence of aura, frequency, severity, duration of attacks, neurological examinations, accompanying diseases, clinic, methods of imaging, treatment, and treatments for prophylaxis in the acute and chronic period of the patients who applied to the clinic were inspected.

Conclusion: It was found out that 24 of the 42 (57%) patients applying to the clinic had headaches. Approximately half of these cases had tension-type headaches (33%) and the other half (58%) had migraine-type headache. It was remarkable that most of the cases complained that their headaches were more severe and frequent during or before attacks.

Discussion: Characteristics and circumstances that need special attention were reemphasized for the treatment of these cases which were assessed with the existence of autoimmunity along with headache types.

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Abstract – WCN 2013

No: 1765

Topic: 6 – MS & Demyelinating Diseases

The clinical, electrophysiological and morphological peculiarities of sensory CIDP

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Background: Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired disorder of peripheral sensory and motor nerves. In 5–6% of patients with CIDP only sensory symptoms are present.

Objectives: Determination of the most sensitive tests to be performed in sensory CIDP and the most frequent clinical findings.

Patients and methods: We examined medical records of 16 patients with sensory CIDP according to the EFNS/PNS guideline (revised 2010). In all patients were performed: clinical evaluation, electrophysiological investigations and cerebro-spinal fluid (CSF) macroscopic/microscopic examination. A full routine blood biochemistry was done. Biopsy specimens of the sural nerve were prepared for light and electron microscopic examination.

Results: Sex ratio: 12 male and 4 female patients (55–79 years). Duration of progression from onset to maximal disability ranged from 4 months–8 years. CSF protein was elevated in 10 patients, ranging from 0.5–1.9 g/l, and normal in 6 cases. Motor and sensory nerve conduction abnormalities were present in 7 cases. Somatosensory evoked potential (SSEP) tests showed involvement of proximal segments or roots of sensory nerves in all patients. Sural nerve biopsy was performed in 5 cases. A definite decrease in the population of myelinated fibres was in all cases.

Conclusion:

1. Nerve conduction study is not a sensitive test to diagnose sensory CIDP, in 56% cases results were normal.
2. SSEP is a highly sensitive test to be used for identifying a possible CIDP.
3. Nerve biopsy is used mainly when electrophysiological studies fail to establish the diagnosis of CIDP.

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Abstract – WCN 2013

No: 1508

Topic: 6 – MS & Demyelinating Diseases

Estimation of the gene polymorphisms role in the progression of multiple sclerosis using MSSS method

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Background: The estimation of the MS progression is important for therapeutic and rehabilitation approach. Commonly used scale for disability evaluation EDSS is not reliable for assessment of progression so MSSS method was developed.

Objective: To explore the association of polymorphisms in arylsulfatase A (ASA) and glutathione S-transferase P1 (GSTP1) gene with disability and progression of MS.

Subjects and methods: The frequency of N350S and 1524 + 95 A-G polymorphisms associated with ASA-pseudodeficiency (ASA-PD), and of A313G and C341T polymorphisms in GSTP1 gene, was determined in 56 and 58 MS patients, respectively, using PCR-RFLP method. EDSS was used to estimate disability level and MSSS to estimate disease progression. Correlation between genotypes and progression was analyzed by Kruskal–Wallis test.

Results: Presence of one or both ASA-PD polymorphisms was determined in 13 patients, from which 9 had mild, 1 moderate and 3 severe disability. No correlation was found between ASA-PD genotypes and MSSS ($p > 0.05$). However, 10 polymorphism carriers had MSSS > 5 which indicates faster disease progression. 32 and 12 patients were found to be carriers of A313 and C341 GSTP1 polymorphism respectively. No correlation was found between investigated GSTP1 genotypes and disability ($p > 0.05$), however patients-homozygous carriers of mutated A313G genotype had significantly higher mean MSSS than patients with normal or heterozygous genotype ($p < 0.05$).

Conclusion: These results suggest that investigated polymorphisms may be associated with MS disability and progression. MSSS method

was proved to be useful for estimation of MS progression as well as for identifying factors affecting disease progression such as gene polymorphisms.

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Abstract – WCN 2013

No: 1507

Topic: 6 – MS & Demyelinating Diseases

Neurologic manifestations in inflammatory bowel diseases

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Introduction: Crohn's disease (CD) and ulcerative colitis (UC) are known as inflammatory bowel diseases (IBD). The association of IBD with neurologic involvement is rare (3%) and controversial.

Objective: To report neurologic manifestations in patients with IBD in order to address its clinical characteristics.

Patients and methods: We conducted a retrospective study over an 11-year period including all patients diagnosed with IBD and neurologic manifestations. Demographic data, neurologic examination, studies and imaging, and treatment were analyzed.

Results: We identified thirteen patients diagnosed with IBD and CNS symptoms: eight with CD and five with UC. Mean age was 51.1 years, mean age of onset of IBD was 38.1 and of neurologic symptoms was 43.3. Sex ratio was 2.5. Most of the patients developed neurologic manifestations after digestive symptoms appeared (11). Cognitive deficits were the most frequent manifestation observed (8 patients), followed by headache (4), epilepsy (3), cerebro-vascular disorders (2), movement disorders (2) and primary progressive MS (1). Two patients had peripheral neuropathy. CNS imaging showed cortical atrophy, white matter abnormalities or vascular lesions. Half of the patients have undergone surgery for their IBD.

Conclusion: The high frequency of cognitive deficits in our series constitutes a peculiar feature, in opposition to higher prevalence peripheral neuropathies, cerebro-vascular disorders (0.12–4%) and CNS white matter diseases in other series. The pathogenesis of neurologic manifestations of IBD is probably related to a common dysimmune basis. Neuropsychological assessment and CNS imaging are needed in patients with IBD for a better understanding of CNS involvement in those diseases.

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Abstract – WCN 2013

No: 1523

Topic: 6 – MS & Demyelinating Diseases

A new model of focal inflammatory demyelination in the neocortex of the rat

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Experimental Autoimmune Encephalomyelitis (EAE) is a widely used model which mimics many histopathological features of multiple sclerosis. However, there are some important disadvantages; firstly, the lesions commonly affect predominantly the spinal cords. Secondly the

lesion's exact localization is not predictable. We present a new technique of creating a focal lesion in the neocortex of the rat by cerebral open flow microperfusion (cOFM). cOFM is a minimal invasive sampling technique involving implantation of a probe into the neocortex without causing a glial scar encapsulation and enables direct access to the brain tissue. After a healing phase liquids can be introduced continuously into the brain tissue via the cOFM probe and sampling of interstitial fluid with cOFM allows monitoring of biomarkers and metabolites. Two weeks after probe implantation, the animals are subcutaneously injected with 100 µg MOG in incomplete Freund's adjuvant to induce a stable IgG antibody titer against MOG. Then 250 ng recombinant TNF-alpha and 150 U IFN-gamma in phosphate buffered saline are injected via the cOFM probe. This results in a selective disturbance of the blood brain barrier and an influx of the MOG antibodies. An inflammatory demyelinating lesion forms in direct vicinity of the probe.

Conclusion: Our new method enables us to predictably produce an EAE lesion in the neocortex of the rat. The cOFM probe allows monitoring of all stages of lesion formation.

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Abstract – WCN 2013

No: 1535

Topic: 6 – MS & Demyelinating Diseases

Pseudotumoral presentation of multiple sclerosis:

Report of seven cases

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Background: Multiple sclerosis (MS) with focal demyelinating lesions of the cerebrum mimicking a tumor of the central nervous system is a rare phenomenon.

Objective: To assess the clinical and radiological characteristics of pseudotumoral forms of MS.

Methods: Among the patients seen at the neurological department between 2000 and 2012, we identified all patients fulfilling McDonald Criteria and whose initial MRI (brain or spinal) presented a large tumefactive lesions. Only patients who have had onset since at least 5 years were included.

Results: Seven young women with tumor-like lesion on initial MRI (brain and/or spinal) are described. The average age of onset was 29 years. The beginning of neurological signs was progressive in four cases and acute in three cases. The main presentations included motor deficits in three cases, headache in two cases and a partial seizure in two cases. MRI showed a single large lesion in three cases and multifocal tumefactive lesions in four cases. The average duration of follow-up was seven years. In our study no biopsy and no surgery were undertaken. The favorable response to steroid therapy (4 cases), the spontaneous remission of symptoms (3 cases), the MRI control findings and the prolonged survival of patients supported the diagnosis of pseudotumoral MS.

Conclusion: This study illustrates the wide variety of MS presentation and underlines the importance of considering MS in the differential diagnosis of a brain mass.

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No: 1576

Topic: 6 – MS & Demyelinating Diseases

Cortical reorganization pattern as a predictor of clinical recovery after the relapse of multiple sclerosis

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Movement disorders are frequent syndrome in multiple sclerosis (MS), leading to severe disability. Early predictors of clinical outcome are important for optimization of rehabilitation therapy. Thus the aim of the study was to investigate cortical reorganization pattern during relapse and in follow-up as a predictor of clinical outcome.

Data were acquired from 25 MS patients, all right-handed, age ranged 19–50, during relapse characterized by unilateral light hand palsy, and in three months during persistent remission. All patients underwent full neurological examination; nine-hole peg test (NHPT) was additionally used to test hand function. Functional MRI (fMRI) was performed on 1.5 T scanner using simple movement paradigm for both hands.

fMRI data analysis showed differently directed changes during relapse: 15 patients had smaller primary sensorimotor cortex (SM1) activation during palsy hand movement in comparison with non-palsy (G1), 10 patients had larger activation (G2). All patients had NHPT palsy hand performance time increase in comparison with non-palsy: G1 (NHPT) ratio median 1.2; G2 (NHPT) ratio median 1.315. G2 palsy hand activation volume correlated with NHPT performance time ($r = 0.76$, Spearman's rho). In three months G1 still had smaller activation during former palsy hand movement in comparison with non-palsy; G2 former palsy hand activation decreased and was comparable with non-palsy. G1 NHPT palsy hand performance time in three months was still elevated (ratio median 1.09) and differed from G2 (Mann-Whitney U-test, $p < 0.045$), where NHPT performance time of both hands was comparable (ratio median 0.96).

Thus decrease of SM1 activation during relapse could be considered as more unfavorable pattern for recovery.

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Abstract – WCN 2013

No: 1702

Topic: 6 – MS & Demyelinating Diseases

Comparison of McDonald 2005, McDonald 2010 and Swanton criteria for prediction of conversion from clinically isolated syndrome to multiple sclerosis

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Background: In 2006, Swanton and colleagues modified the MRI criteria to simplify and speed the diagnosis. Recently the Panel has proposed new MRI criteria for the diagnosis of MS in patients with CIS.

Purpose: To compare the ability of the McDonald 2005, McDonald 2010 and Swanton's modified criteria to predict conversion CIS to clinically definite multiple sclerosis (CDMS) from baseline MRI findings. We also aimed to evaluate the accuracy of these new criteria for lesions dissemination in space (DIS) and time (DIT).

Patients and methods: 44 patients presenting with CIS included in the study, and followed up for at least 2 years. The sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV) and accuracy of MRI dissemination-in-space criteria were calculated.

Results: Overall conversion rate was 86.4%. Out of 44 patients, 38 converted to CDMS a mean of 10.1 months after onset of first clinical event. Swanton's modified criteria showed a sensitivity of 84%, a specificity of 23%, PPV is 88%, NPV is 25%, with an accuracy of 77%. Barkoff-Tintore criteria showed a sensitivity of 81%, a specificity of 50%, PPV is 91%, NPV is 30% with an accuracy of 77%. The McDonald 2010 modified criteria a sensitivity of 47.3%, a specificity of 83.3%, PPV is 94.7%, and NPV is 20%, an accuracy of 52.2%.

Conclusion: In conclusion, McDonald 2010 criteria seem to be simpler and more specific than previous criteria. Besides, lesions in corpus callosum and Gd-enhanced lesions have critical value to make an early diagnosis in CIS.

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Abstract – WCN 2013

No: 1691

Topic: 6 – MS & Demyelinating Diseases
Neuromyelitis optica in Brazilian woman with myasthenia gravis

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Background: Despite the rarity of myasthenia gravis (MG) and neuromyelitis optica (NMO), several cases or small series of patients with both disorders have been reported over the years. Here we describe one Brazilian patient with MG and NMO. To our knowledge, less than 50 cases have been reported in the medical literature.

Patient and methods: A Brazilian woman was diagnosed with generalized MG at age 16 years and underwent thymectomy one year after diagnosis. Ten years later she presented with leg weakness, sensory loss below T3 level and urinary retention. At age 27 years, she developed bilateral acute visual loss. Visual evoked response was absent on the left and of delayed latency on the right. At age 37 and 48 years, she had relapses of optic neuritis in the right eye.

Results: During the relapses, a cervical-dorsal lesion was detected on spinal cord magnetic resonance imaging (MRI), brain MRI showed non-specific white matter lesions and aquaporin-4 antibody was positive (titer of 1:640). The diagnosis of NMO was made according to the 2006 revised diagnostic criteria.

Conclusion: The exact contribution of thymectomy to the development of NMO is not clear. Future research is required to establish whether there is indeed a small long-term excess risk of developing autoimmune disease in individuals with MG who undergo thymectomy.

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Abstract – WCN 2013

No: 1679

Topic: 6 – MS & Demyelinating Diseases
Pattern of cognitive impairment and related factors in clinically isolated syndrome

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Background: The presence of cognitive dysfunction in a sizeable proportion of patients with clinically isolated syndrome (CIS) has been reported. The prevalence of the cognitive deficits usually encountered could vary with the clinical course of the disease.

Objective: The aim of the present study was to investigate the prevalence and pattern of cognitive impairment in CIS patients.

Patients and methods: 25 patients with CIS (mean age 36.3) included in the study. Age and sex matched 22 healthy control subjects (mean age 36.0) were also included. Cognitive performance was assessed by using a battery of tests consisting of Paced Auditory Serial Addition Test (PASAT), Stroop, Controlled Oral Word Association (COWAT), Rey Auditory Verbal Learning (RAVLT), Benton's Judgment of Line Orientation (BJLO), Trial making test (TMT), and digit span test (DST). Beck depression scale, Fatigue impact scale (FIS), and Multiple Sclerosis International Quality of Life (MUSIQoL) were administered.

Results: Globally, 21% of patients were found to be cognitively impaired at least two tests. When compared healthy controls, 44% of patients

failed in Stroop test. Impairment in COVAT was 56%. Impairment in RAVLT, BJLO, TMT and DST was relatively rare (10%, 9%, 5% and 12%, respectively). Impairment in PASAT was 12%. CIS patients were found more depressed ($p = 0.045$) and more fatigued ($p = 0.011$).

Conclusion: Our results supported the early onset cognitive impairment in demyelinating diseases. The most prominent impairment was found to be in verbal fluency (COVAT), attention and executive functions (Stroop). Fatigue (especially cognitive sub-group) found to be related with cognitive impairment in CIS.

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Abstract – WCN 2013

No: 1692

Topic: 6 – MS & Demyelinating Diseases
Interferon's flu-like syndrome and mental state: Possible interactions

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Background and objectives: Interferons-beta are currently the most used drugs for disease modifying of multiple sclerosis. One of the most common side effects of the therapy is a flu-like syndrome (FLS). FLS means a combination of at least 2 and the following adverse events: fever, chills, myalgia, arthralgia and sweating. The basis of the syndrome is probably on the change in the level of cytokines. But there are other factors that determine the severity and duration of the existence of this syndrome, in particular, the state of psychoemotional sphere.

Materials and methods: 90 patients (28 men and 62 women), RRMS, remission, age – 39.1–43.4 (95%CI) years, with disease duration of 117–156 (95%CI) months; interferon beta (i.m. and s.c.) use for 33.2–44.4 (95%CI) months; EDSS, Original adverse event questionnaire, Zung Depression and Anxiety Scale, Boyko's Euphoria Scale, Modified Fatigue Impact Scale. Statistical analysis – nonparametric statistic: R Spearman, U Mann-Whitney, φ^* Fisher.

Results and conclusion: FLS presents in 57 cases (63%) and is more common in women – 69% than in men – 50% ($\varphi^* = 1.75$, $p < 0.05$). Anxiety is significantly higher in group with FLS ($U = 569$, $p = 0.002$) and increases with the severity of FLS ($R = 0.35$, $p = 0.0005$). Depression ($R = 0.23$, $p = 0.034$) and asthenia ($R = 0.24$, $p = 0.025$) increase with the severity of FLS. Euphoria does not affect the severity of FLS ($R = -0.01$, $p = 0.92$).

Thus, the patient's mental state has a significant impact on the occurrence and severity of FLS, which may be important to determine the risk of the syndrome and improve patient compliance.

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Abstract – WCN 2013

No: 1672

Topic: 6 – MS & Demyelinating Diseases
Association between multiple sclerosis prevalence and environmental and genetic factors in Latin America and the Caribbean: An ecological study

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Objectives: To study the prevalence of MS and its associations with genetic and environmental variables and to estimate the proportion of the prevalence's variance explained by these variables through a statistical model.

Methodology: This is an ecological study. From a total of 33 studies, we selected 24 surveys for this ecological study. A model is proposed with data from 24 surveys, 12 countries, and 20 investigators. The surveys are from Mexico (28° of latitude north) to Patagonia (55° of latitude south). The variables under study are: the prevalence of MS, the latitude, the sun exposition, the ethnic group, the altitude, the temperature, and the Ultraviolet Radiations and the level of instruction.

Results: In the bivariate analysis we found an association between the MS prevalence and the latitude, the ethnic group, the altitude, and the temperature.

In the multivariate analysis using generalized linear models (GML) the best predictors were Latitude (second-degree polynomial), Ethnic group and Temperature (second-degree polynomial). The model accounted for 85.30% of the deviance.

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Abstract – WCN 2013

No: 1646

Topic: 6 – MS & Demyelinating Diseases

Tumefactive MS lesions under fingolimod: A case report and literature review

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Objective: The formation of atypical lesions in patients with multiple sclerosis (MS) on novel disease modifying treatments for MS like fingolimod is diagnostically challenging and pathophysiologically unclear. We report a patient who developed a tumefactive demyelinating lesion (TDL) 8 months after a switch from natalizumab to fingolimod and a second extensive lesion after another 6 months on continued fingolimod therapy.

Methods: We performed serial clinical and radiological assessments, and immunophenotyping of blood and CSF immune cells. Accessorily we present a literature review about recent similar cases.

Results: Clinical course and radiological findings were consistent with diagnosis of tumefactive demyelinating lesions. Immune cell phenotyping showed pronounced shifts in the immune cell composition related to fingolimod treatment. In addition, we observed a subset of highly differentiated effector cells (CD45RO^{neg}CCR7^{neg}) within the CD8+ T cell population which was about 2-fold enriched in the CSF compared to the peripheral blood.

Conclusion: Our observation adds further evidence for the development of atypical demyelinating lesions in some patients under fingolimod treatment. This notion may be due to a treatment-associated shift in the immune pathology of individuals with specific susceptibility.

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Abstract – WCN 2013

No: 1678

Topic: 6 – MS & Demyelinating Diseases

Impact of cortical lesions identified by routine 3 T MRI imaging on cognitive performance of patients with multiple sclerosis

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Background: Histopathologic studies have reported widespread cortical lesions (CLs) in multiple sclerosis (MS), however, in vivo detection by using routinely available pulse sequences is challenging.

Objective: We investigated the frequency and subtypes of CLs and their relationships to white matter lesion load and physical and cognitive disability.

Patients and methods: CLs were identified and classified on the basis of concurrent review of 3D FLAIR and 3D T1-weighted IR-SPGR 3 T MR images in 26 patients with MS. Twenty-five patients completed the MACFIMS cognitive battery. White matter lesion volume, CL number, and CL volume were assessed.

Results: Overall, 249 CLs were detected. CLs were present in 24/26 patients (92.3%) (0–30; 9.6 ± 8.8). Most CLs were classified as mixed cortical–subcortical (type I) (94.4%); the remaining 5.6% were classified as purely intracortical (type II). Subpial cortical lesions (type III) were not detected. White matter lesion volume correlated with CL number and CL volume (both $p < 0.001$). After controlling for age, depression, and premorbid intelligence, we found that CL number, CL volume, and white matter lesion volume correlated with the SDMT score ($p < 0.014$); CL number also correlated with the CVLT-II scores ($p < 0.043$). The EDSS scores correlated with CL number and CL volume ($p < 0.05$), but not with white matter lesion volume.

Conclusion: Our routinely available imaging method detected many CLs in patients with MS and was useful in their precise topographic characterization in the context of the gray matter–white matter junction. Routinely detectable CLs were related to physical disability and cognitive impairment.

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Abstract – WCN 2013

No: 1683

Topic: 6 – MS & Demyelinating Diseases

Effect of intravenous immunoglobulin treatment on postpartum-related relapses in multiple sclerosis

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Background: Multiple sclerosis (MS) mainly affects young women in their childbearing age.

During pregnancy the rate of relapses is decreased, especially in the third semester and increased in the first trimester postpartum. The result of some studies demonstrated beneficial effect of intravenous immunoglobulin (IVIG) on postpartum relapses since the relapse rate did not increase postpartum.

Objective: The aim of the study was to evaluate the effects of IVIG and breastfeeding on postpartum relapse rate.

Patients and methods: Patients with MS, who become pregnant during the year 2007–2012 were included in the study. A total of 67 patients with MS with median age 34.5 ± 4.3 year and a median

disease duration of 7.9 ± 4.0 years and the median EDSS score of 1.47 ± 1.37 were included in the study. 40 out of 67 were treated with DMT before pregnancy. After delivery all our patients treated with IVIG for the first six months.

The patients were followed from two years before pregnancy to 2 years after delivery. Data on relapse rate, EDSS and breastfeeding were collected.

Results: The mean annualised relapse two years before pregnancy, during pregnancy and 24 months after delivery was 0.48, 0.16 and 0.43 respectively. The mean EDSS at the year of pregnancy was 1.47 and 24 months after delivery was 1.7. Breastfeeding has no effect on relapse rate and on EDSS progression.

Conclusions: Treatment with IVIG in the first six months postpartum effectively reduced the incidence of postpartum relapse. Breastfeeding has no effects on relapse rate and on EDSS.

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Abstract – WCN 2013

No: 1311

Topic: 6 – MS & Demyelinating Diseases

Epidemiology and mortality of MS: A nationwide population study in Taiwan

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Background: There are few nationwide population-based studies of multiple sclerosis (MS).

Objective: To describe the epidemiologic features and mortality of MS in Taiwan.

Material and methods: The catastrophic illness registry of the Taiwan National Health Insurance Research Dataset and the National Death Registry of Taiwan were used to estimate the incidence and mortality of MS during 2003 to 2007.

Results: A total of 832 incident MS patients (180 males, 652 females) were identified during 2003 to 2007. The mean age at diagnosis was 39.4 ± 13.9 (range 5–83) years. The incidence of MS in Taiwan was 6.79 cases (males, 2.86; females, 10.92) per million person-years. The incidence in individuals aged <20, 20–39, 40–59 and ≥ 60 years was 2.21, 8.83, 10.65 and 3.16 per million person-years. Period prevalence of MS during 2003 to 2007 was 28.1 per million people. There were 64 deaths (males, 16; females, 48) during the study period; 1-, 2-, and 5-year survival rates were 98.4%, 95.8%, and 90.6%, respectively. Crude mortality rate was 24.5 deaths per 1000-patient-years, without sex difference (log-rank test, $p = 0.480$). MS patients had a standardized mortality ratio (SMR) of 4.00 (95% confidence interval [CI], 3.11–5.08) for all-cause mortality, as compared with the national population in 2003. SMRs (95% CI) for male and female MS patients were 3.20 (1.89–5.09) and 4.36 (3.25–5.74), respectively.

Conclusion: MS is rare in Taiwan but entails 4-fold risk of all-cause mortality in comparison with the general population.

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Abstract – WCN 2013

No: 1628

Topic: 6 – MS & Demyelinating Diseases

Interferon-beta up-regulates suppressor of cytokine signaling 1 in T cells of multiple sclerosis patients

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Background: Although interferon-beta (IFN- β) is commonly prescribed for the treatment of MS, its mode of action is not fully understood. Previous work suggests a beneficial role of suppressors of cytokine signaling 1 (SOCS-1) in experimental autoimmune encephalomyelitis (EAE), a widely used animal model of MS. We postulate that IFN- β attenuates immune responses in MS via induction of SOCS-1 in CD4 and CD8 T cells.

Objectives: To assess SOCS-1 expression in T cells of MS patients and to evaluate anti-inflammatory functions of SOCS-1 in human CD4 and CD8 T cells.

Methods: We quantified SOCS-1 mRNA in CD4 and CD8 T cells from untreated or IFN- β -treated MS patients. We tested the impact of a SOCS-1 mimic, tyrosine kinase inhibitory peptide (Tkip) on CD4 and CD8 T cell responses to cytokines with pro-inflammatory properties using flow cytometry-based assays.

Results: We found that SOCS-1 is significantly more expressed by both CD4 and CD8 T cells from IFN- β -treated MS patients compared to untreated patients. Mimicking the effect of SOCS-1 in-vitro significantly reduced cell proliferation, and production of IFN-gamma (IFN- γ) and granzyme B by human CD4 and CD8 T cells.

Conclusions: Our results indicate that IFN- β increases SOCS-1 expression in CD4 and CD8 T cells. Our in-vitro assays show that SOCS-1 interferes with cytotoxic T cell functions and the production of the Th1 signature cytokine IFN- γ . Hence, IFN- β may indeed mediate anti-inflammatory effects via an upregulation of SOCS-1 in CD4 and CD8 T cells.

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Abstract – WCN 2013

No: 1630

Topic: 6 – MS & Demyelinating Diseases

Trigeminal neuralgia and multiple sclerosis: Quality of life, headache impairment and anxiety-depressive symptoms

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Background: Trigeminal neuralgia (TN) has been related with multiple sclerosis (MS) although its mechanism is still unclear.

Objective: To compare quality of life, headache impact and anxiety-depressive symptoms in MS-related TN and idiopathic TN.

Patients and methods/material and methods: MS-related TN patients from our MS clinic and idiopathic TN patients from the Headache clinic were given these tests: Headache Impact Test (HIT-6), Hospital Anxiety and Depression Scale (HADS) and the SF-36 Health Survey.

Results: Prevalence of MS-related TN was 3.7% (9 of 13 patients agreed to participate in the study). Female/male ratio was 2:1 in both groups, mean age was 59.6 years old (42–68) in the MS-related group and 52.1 (39–78) in the idiopathic TN group. During the study, gabapentin and oxcarbazepine were the most common drugs used in both groups.

Results of MS-related TN group: Expanded Disability Status Scale (EDSS) average score was 4.4 (1.5–8.5). HIT-6 score was 26.25, HADS-anxiety 5.4, HADS-depression 5.6, SF-36/physical terms 54.46 and SF-36/emotional terms 55.48.

Results of idiopathic TN group: HIT-6 mean score was 61.1, HADS-anxiety 10, HADS-depression 6.7, SF-36/physical terms 56.1 and SF-36/emotional terms 55.6.

Conclusion: Unexpectedly, both groups showed very similar SF-36 scores and HADS scores were higher in idiopathic TN. It is also remarkable that idiopathic TN group had higher headache impact in their lives (more than twice of MS-related TN group). It could suggest that maybe MS-related TN have different pain characteristics or maybe MS patients have different pain threshold than non-MS patients.

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Abstract – WCN 2013

No: 1326

Topic: 6 – MS & Demyelinating Diseases

A patient suffering relapsing demyelinating disease affecting both central and peripheral nervous systems

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Background: Multiple sclerosis is characterized by inflammation, demyelination and gliosis, involving the central nervous system (CNS) and sparing the peripheral nervous system (PNS). Chronic inflammatory demyelinating polyneuropathy is regarded as being restricted to the PNS as the immune response seems to be directed against an antigen confined to PNS proteins. However, patients presenting both central and peripheral demyelination have occasionally been reported. We report a case of patient with relapsing demyelinating disease of CNS associated with demyelinating polyneuropathy.

Case report: A 53 year old woman presented a 7 year history of recurrent neurologic symptoms (Diplopia, ataxia, optic neuritis) with complete regression after intravenous corticotherapy. Her neurological examination out of the relapses was normal except of the generalized areflexia. Brain and spinal cord MRI demonstrated some small size white matter hyperintensities T2 (<3 mm of diameter) with contrast enhancement. CSF was acellular, with protein level 0.89 g/dl without any oligoclonal bands. Her electroneuromyography revealed a severe demyelinating sensory motor neuropathy with conduction blocks. The diagnosis of chronic inflammatory demyelinating polyneuropathy was confirmed by a peripheral nerve biopsy. Body CT, PET, biopsy of the minor salivary glands, immunologic, biochemical and infectious investigations were normal.

Conclusion: Immunological reactivity against antigens common to peripheral and central myelin or a higher general susceptibility to autoimmune diseases, may explain why the demyelinating disease affected both the CNS and PNS.

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Abstract – WCN 2013

No: 1383

Topic: 6 – MS & Demyelinating Diseases

Do disease modifying treatments improve outcome? First results from the London MS Database

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Objectives:

1. To present first assessments based on 'The London MS Database'.
2. To review eligibility for Disease Modifying Therapy (DMT) in a geographically defined cohort of people with relapsing-remitting multiple sclerosis (pwRRMS).
3. To evaluate the effect of DMT on disease progression.

Patients and methods: Medical records of 94 pwMS living in the London Borough of Tower Hamlets were reviewed for DMT eligibility according to the Association of British Neurologists' 2001 criteria. Outcomes were (i) sustained use of a walking aid (cane) and (ii) onset of secondary progressive (SP) MS; assessed using Kaplan–Meier analysis.

Results: Fifty-five pwRRMS were eligible for 1st line DMT, 17 were not eligible, and 21 were excluded for incomplete information. Median time between MS diagnosis and eligibility was 6.3 years, and median follow-up was 6.5 years. Baseline EDSS was ≤ 2.5 in 70%. Thirty-three eligible patients (60%) started DMT after a median delay of 2.1 years. Nineteen (57%) commenced on β -interferon, and 9 (27%) on glatiramer acetate. Eighteen pwRRMS converted to SPMS; 14 started using a walking aid. Kaplan–Meier analysis did not reveal significant difference between treated and untreated patients with respect to sustained use of a cane, and conversion to SPMS.

Conclusions: The London MS Database is a new dataset recording disease relevant details in an ethnically heterogenic population. The characterization of the included pwMS is ongoing. Our preliminary analysis failed to detect an effect of DMT on two robust outcomes. Further data collection is underway to clarify whether this result is due to the limited sample size.

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Abstract – WCN 2013

No: 1360

Topic: 6 – MS & Demyelinating Diseases

Intravenous immunoglobulin therapy for Anti-aquaporin 4 antibody seropositive optic neuritis

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Background and objects: Neuromyelitis optica (NMO) is an idiopathic, severe, demyelinating disease of the central nervous system that preferentially affects the optic nerve and spinal cord. Anti-aquaporin 4 antibody seropositive optic neuritis (ON) including NMO spectrum disorders is often severe, bilateral and recurrent. The prognosis of seropositive ON is worse than for multiple sclerosis (MS). Intravenous methylprednisolone pulse therapy (IVMP) for MS is ineffective for NMO. Our study is to clarify the effect of intravenous immunoglobulin (IVIG) for acute seropositive ON.

Patients and methods: This was an open-label, non-randomized study design. Four patients with anti-aquaporin 4 antibody seropositive ON who did not respond to IVMP were consecutively enrolled. The average age was 43.8 years old and the sex was all female. Two of them were involved bilaterally and others were unilaterally. The visual activity in the affected eyes was severe to light perception only. MRI showed swollen optic nerve in all patients. Their visual activity did not recover after IVMP. IVIG was administered at a dose of 400 mg/kg/day for five days followed by oral prednisolone.

Results: Three of four patients showed a return to normal or near normal after IVIG. The fourth patient showed only a partial recovery.

Treatment of IVIG was generally well-tolerated by all patients without any serious adverse effects.

Discussion and conclusion: Our study suggests possible effect of early IVIG therapy in anti-aquaporin 4 antibody seropositive ON who fail to respond to IVMP. A large controlled trial is indicated to confirm these results.

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Abstract — WCN 2013

No: 883

Topic: 6 — MS & Demyelinating Diseases

Spectrum of idiopathic inflammatory demyelinating diseases of the CNS in a tertiary care hospital in India

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Background: Idiopathic inflammatory demyelinating diseases (IIDD's) of the CNS include broad spectrum of CNS disorders. There are very few studies of its kind in India.

Objectives: To study the clinical, radiological and laboratory profile of IIDD's of the CNS prospectively.

Material and methods: Patients fulfilling the criteria for IIDD's in our hospital between October, 2010 and January, 2013 were included. Clinical, radiological & laboratory features were evaluated.

Results: 56 patients fulfilling the criteria for IIDD's were included and analyzed. The mean age was 24.97 years and was lowest in acute disseminated encephalomyelitis (ADEM). The female to male ratio was 1.55:1. Females predominated in all subtypes except in ADEM & ATM. Clinical spectrum was highest with clinically isolated syndrome (CIS) (n = 16) followed by neuromyelitis optica (NMO) spectrum (n = 12), acute transverse myelitis (ATM) (n = 11), ADEM (n = 9) and multiple sclerosis (MS) (n = 8). In MS, motor weakness, subcortical white matter lesions, and VEP abnormalities were the common findings. Two rare variants of MS were diagnosed. In NMO spectrum, NMO is the most common subtype (n = 6). Cervico-dorsal involvement, and VEP abnormalities were the common findings. NMO IgG was positive in two cases. In ADEM, altered sensorium (n = 6), and supratentorial lesions (n = 9) were the common findings. In ATM, dorsal cord was involved in 8 cases with significant motor & sensory involvement. In CIS, isolated Optic Neuritis (n = 13) was the most common presentation.

Conclusions: IIDD's of CNS had a wide spectrum of disorders. The frequencies of these disorders seen are quite different from those seen in the West.

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Abstract — WCN 2013

No: 1441

Topic: 6 — MS & Demyelinating Diseases

Correlation of CSF and plasma antioxidative capacity in patients with different clinical phenotypes of neuroinflammation and clinical and radiological findings

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Objective and methods: Oxidative stress is revealed as main contributors in the pathophysiology of multiple sclerosis (MS). Analyzing sera and CSF of patients with clinically isolated syndrome (CIS) and those defined as relapse remitting multiple sclerosis (RRMS), we tested the hypothesis that there is major oxidative stress intensity even in

the earliest neuroinflammation in MS, defined as CIS, with possible differences in peripheral and CNS mechanisms against oxidative condition.

Results: The obtained results revealed an increase in malondialdehyde levels in plasma and CSF most prevailing in patients with severe clinical presentation ($p < 0.05$), parallel with positive correlation with patient's clinical score and radiological changes ($p < 0.01$), in both study group, compared to control values. Similar trend was observed testing catalase and total superoxide dismutase activity; measured activities were higher in CIS and RRMS patients in plasma ($p < 0.05$), parallel with an increased catalase activity ($p < 0.05$), followed by a decrease in superoxide dismutase activity in CSF ($p < 0.05$), compared to the control values. The positive correlations of these biomarkers and clinical score were obtained ($p < 0.01$), without similar ratios compared to the patients' radiological features ($p > 0.05$). All measured values were significantly higher in peripheral and CNS of CIS compared to RRMS patients ($p < 0.05$).

Conclusion: The obtained results might be useful in providing the earliest antioxidative treatment aimed to preserve total and CNS antioxidative capacity parallel with delaying irreversible neurological disabilities in MS progression.

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Abstract — WCN 2013

No: 1404

Topic: 6 — MS & Demyelinating Diseases

Efficacy of fingolimod treatment in relapsing multiple sclerosis patients in clinical practice: A multicenter experience

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Background: Fingolimod is the first oral disease modifying therapy (DMT) approved for the treatment of Multiple Sclerosis (MS). Post-marketing studies are important to confirm what were established in clinical trials.

Objective: To evaluate efficacy of fingolimod in a cohort of relapsing MS patients treated with fingolimod in a real clinical practice setting.

Methods: We identified patients who had been prescribed fingolimod using the MS registry in 3 MS clinics in Kuwait. Patients with EDSS ≤ 6 and at least 6 months of follow-up were included. Patients with progressive MS were excluded. Primary endpoint was the proportion of relapse free patients at 6 months. Secondary endpoints included mean change in EDSS and proportion of MRI activity (new T2 or Gadolinium-enhancing lesions).

Results: Out of 101 eligible patients, 61 were women and 40 men. Mean age and mean disease duration were 33.97 ± 9.70 years and $7.32 \pm .03$ years respectively. 78.2% patients received prior DMTs. Mean duration of fingolimod exposure was 16.04 ± 5.36 months. Proportion of relapse-free patients increased significantly (10.9% versus 78.2%; $p < 0.0001$) at 6 months. Mean EDSS score at baseline was 2.97 ± 1.51 , which decreased significantly to 2.08 ± 1.25 . At 6 months, 19.8% of patients were free from MRI activity compared to 86.1% at baseline ($p < 0.0001$). No serious adverse events were reported.

Conclusion: In clinical practice, fingolimod is effective in reducing disease activity and progression of disability over the observational period. The extent of efficacy was more pronounced than what was reported in the pivotal trials.

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Abstract – WCN 2013**No: 1417****Topic: 6 – MS & Demyelinating Diseases****Multiple sclerosis (MS) developing in a patient first diagnosed with chronic inflammatory demyelinating polyneuropathy (CIDP) – A case report**D. Sisak, Z. Pál, A. Iljicsov, D. Bereczki, M. Simó. *Semmelweis University Faculty of Medicine, Budapest, Hungary*

Background: It has been previously reported that demyelinating inflammatory diseases of the central (CNS) and peripheral nervous system (PNS) have common aetiopathogenesis, probably due to an autoimmune response against a common myelin antigen. However, to date, only a few case reports document the coexistence of CIDP and MS.

Objective: We describe a 34 year old woman with CIDP whose disease went into remission, and who developed MS years after the diagnosis of the polyneuropathy.

Patients and methods: Case report with a 15-year-long follow-up.

Results: The patient's first symptoms occurred at the age of 20 with paraesthesia of the limbs, and progressive weakness of the feet. Neurological examination, nerve conduction studies, as well as cerebrospinal fluid examination supported the diagnosis of CIDP. She received steroid treatment and plasmapheresis several times. The disease went into complete remission. Two years after the diagnosis of the polyneuropathy, the patient developed brainstem symptoms, and brain MRI showed multiple demyelinating lesions. Three years later spastic paraparesis, urinary retention, and double vision developed. MRI disclosed lesion characteristic for MS, and cerebrospinal fluid examination proved OGP. Immunomodulatory treatment was introduced. In the last four years, the patient had residual signs of MS only.

Conclusion: Here we demonstrate a rare case of sequentially occurring CIDP and MS. While most previously published similar reports demonstrated a demyelinating disease affecting the CNS prior to the occurrence of peripheral demyelination, we describe a case where the PNS was affected before the symptoms of MS appeared, furthermore, CIDP became inactive.

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Abstract – WCN 2013**No: 1426****Topic: 6 – MS & Demyelinating Diseases****Multiple sclerosis in men: A Tunisian hospital cohort**H. Jamoussi, K. Meriem, N. Ben Ali, S. Fray, M. Fredj, S. Blel *Charles Nicolle Hospital, Tunis, Tunisia*

Background: Symptom prevalence as well as characteristics differs between the sexes in Multiple Sclerosis (MS).

Objective: We describe on clinical course in a cohort of a Tunisian men affected by MS.

Patients and methods: We selected men affected by MS followed in the service of Neurology of Charles Nicolle Hospital (2000–2012). Patients had a neurological examination, a cerebral and medullar magnetic resonance imaging (MRI), an analysis of cerebrospinal fluid and visual evoked potentials (VEP). All the results were analysed.

Results: From 160 patients MS followed, 41(25 %) were males. Median age of the disease onset was 25 years. Clinical onset was monosymptomatic in 50% of cases (pyramidal sign in 50%). Of 78% of patients who are initially diagnosed with relapsing remitting MS, 32% had transitioned to secondary progressive phase. 22% had a primary progressive form. The median time to convert from EDSS 1 to EDSS 3 was 3 years and from EDSS 6 was 5 years. The lesions on MRI were supratentorial in all cases, subtentorial in 22% and cervical

in 33%. VEP were altered in 64% of cases and oligoclonal bands were found in 36% of patients. The EDSS was improved or stabilised after immunomodulator treatment in 2/3 of cases. The median time of follow-up was 7 years.

Conclusion: Gender appears to affect MS prevalence, clinical course, pathology and response to immunotherapy. The effects of gender need to be taken into consideration when designing and interpreting research relating to MS, particularly in the development of new therapeutic agents.

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Abstract – WCN 2013**No: 1121****Topic: 6 – MS & Demyelinating Diseases****Immunosuppressive treatment in relation to different phenotypes of clinical presentation of chronic inflammatory demyelinating polyneuropathy (CIDP)**C. Marcoci^a, V. Lisnic^b, V. Nemtanu^b. *^aNeurology, State University of Medicine and Pharmacy 'Nicolae Testemitanu', Moldova; ^bNeurology, Institute of Neurology and Neurosurgery, Chisinau, Moldova*

Chronic inflammatory demyelinating polyneuropathy (CIDP) is uncommon, but an important disease. Learning about diseases of the peripheral nervous system in general, the diagnostic strategies required to sort them out. The only population-based studies suggest a prevalence of one or two per 100 000, but these are probably underestimates because of lack of referral or recognition of cases. In our population-based study of 52 patients with CIDP in the Republic of Moldova, for the confirmation of the diagnosis we used the criteria of the American Academy of Neurology, later revised by EFNS and PNS criteria, MRC score, neuropathy score, and electrophysiological examination. In some cases nerve biopsy was performed. The objective of the study was the determination of the phenotypes of clinical presentation of CIDP and the efficiency of various methods of immunosuppressive treatment: corticosteroids, corticosteroids in association with azathioprine, corticosteroids in association with plasmapheresis and intravenous immunoglobulins, in relation to different phenotypes. Three phenotypes of clinical presentation of CIDP were distinguished: motor-sensory, motor and sensory. The motor-sensitive phenotype develops as a distal symmetric neuropathy with or without signs of central nervous system impairment and multifocal motor-sensitive neuropathy. Administration of corticosteroids is efficient in the short-term management of CIDP. Corticosteroids are not efficient in the treatment of motor phenotype of CIDP. Intravenous immunoglobulins significantly ameliorate the evolution of all the phenotypes of CIDP. The treatment of corticosteroids in association with azathioprine is cost-efficient in the treatment of symmetric motor-sensory and sensory neuropathies.

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Abstract – WCN 2013**No: 1134****Topic: 6 – MS & Demyelinating Diseases****Complement regulator factor H as a serum biomarker of multiple sclerosis: Correlation with disease state and MRI**L. Elnabil. *Neurology, Ain Shams Faculty of Medicine, Cairo, Egypt*

Background: Multiple sclerosis (MS) has a variable phenotypic presentation and subsequent disease course that, although unpredictable at disease onset, is of crucial importance in guiding interventions. Effective

and accessible biomarkers are required in order to stratify patients and inform treatment.

Aim of the work: To determine the predictive value of serum complement factor H in MS patients and if they correlate with magnetic resonance imaging (MRI).

Subjects and methods: We prospectively studied sixty patients with MS. They were subjected to full clinical and neurological assessment, laboratory tests, and MRI study. Thirty healthy subjects matched patient group with age and sex were selected as a control group.

Results: Serum complement factor H levels were highly significantly elevated in all MS patients compared to controls (p value = 0.000), with sensitivity and specificity 98.3% and 93.3%, respectively, with positive predictive value 98.33%. Within the RRMS group, serum complement FH levels in remission patients were higher than in control (P = 0.000) and lower than in relapsing group (p = 0.000). Moreover, serum complement FH concentration had a highly significant value as a surrogate marker for the prediction of relapse in MS patients with sensitivity and specificity of 79% and 90%, respectively (test cut-off value of relapse \geq 6.25 ng/ml). Serum complement factor H level was not related to the localization of lesion in MS patients with acute relapse.

Conclusion: Serum complement factor H may be an effective indicator of progression, an accessible biomarker and stratifying tool in determining disease course, providing objective evidence to help guide therapeutic decisions.

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Abstract – WCN 2013

No: 1192

Topic: 6 – MS & Demyelinating Diseases Memory deficits in multiple sclerosis subtypes

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In MS patients, episodic short-term memory (ESTM) is frequently affected. It is assumed that cognitive deficits are less severe in patients with a relapsing-remitting (RR) than in those with a secondary-progressive (SP), primary-progressive (PP) or progressive-relapsing (PR) course. This difference is attributed to a difference in neurodegeneration. We examined the relationship between episodic short-term memory and course of the disease.

ESTM was assessed by means of the computer-based Memory and Attention Test (MAT) in not-acutely-ill MS outpatients. Severity of neurological symptoms was determined by the EDSS. ESTM group differences were calculated with unpaired Student's t-tests before and after matching the groups for EDSS, age, sex and education.

We assessed 531 patients (349 women, 182 men) aged between 17 and 60 years (mean/SD: 39.7/9.8 years). The course of the disease was RR in 409 patients (77.0%), SP in 71 (13.4%), PP in 15 (2.8%) and PR in 12 (2.3%); a CIS was found in 24 patients (not further considered here). ESTM was worse in patients with an SP, PP or PR course compared to RR (p < 0.001). However, patients with SP, PP and PR were older (p < 0.05) and had a higher EDSS (p < 0.01). Recalculating group differences in the matched groups revealed no significant differences anymore.

The noticeable severity of memory impairment in patients with a progressive course compared to RR patients may not be specific for these forms of the disease, because after controlling for severity of neurological symptoms, age, sex and education it can no longer be ascertained.

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Abstract – WCN 2013

No: 1199

Topic: 6 – MS & Demyelinating Diseases Is multiple sclerosis a prothrombotic disease?

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Background: The exact prevalence and pathogenic role of anti-phospholipid antibodies (aPL) in multiple sclerosis (MS) remain unclear.

Objective: The aim of this prospective study was to evaluate the rate of aPL positivity in different MS phases in order to recognize their involvement in the pathogenesis of MS.

Patients and methods: We studied the reactivity for aPL including anti-cardiolipin, anti- β 2glycoproteinI, anti-prothrombin, anti-annexinV in healthy controls and MS patients in different phases: secondary progressive (SPMS), relapsing-remitting in relapse (REL) or remission (REM).

Results: We analyzed the sera of 60 controls and 100 consecutive MS patients (58 REM, 26 REL and 16 SPMS). The rate of positivity for at least one aPL was significantly higher in MS patients compared to controls (32 vs 7%, p < 0.0001), in particular for anti-prothrombin IgM (7% vs 0, p = 0.05), and in REL compared to REM and SPMS (53.8, 20.7, 37.5% respectively, p = 0.002), specifically for anti- β 2glycoproteinI IgM (26.9, 1.7, 6.3% respectively, p < 0.0001), anti-prothrombin IgM (15.4, 3.4, 6.3% respectively, p = 0.05) and IgG (19.2, 5.2, 0% respectively, p = 0.05). Likewise, the absolute values of anti- β 2glycoproteinI IgM (p < 0.0001) and IgG (p = 0.007), anti-prothrombin IgM (p = 0.007) and IgG (p = 0.01) were significantly higher in REL compared to REM and SPMS.

Conclusion: Our results showed a significant increase of anti-phospholipid antibodies in MS patients compared to healthy controls, with a strong association of anti- β 2glycoproteinI and anti-prothrombin antibodies with disease exacerbation. Based on our results and on review of literature, we supposed that MS may be a primarily prothrombotic disease.

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Abstract – WCN 2013

No: 1197

Topic: 6 – MS & Demyelinating Diseases Educational attainment moderates the effect of T2 lesion load and atrophy on cognition in multiple sclerosis

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Background: The relationship between MR-markers of disease-related tissue changes and cognition in multiple sclerosis (MS) is variable. Previous work suggested that greater educational attainment (EA) might moderate the negative impact of brain changes on cognition, but this has not been replicated in independent cohorts and restricted to a limited set of MRI-metrics so far.

Objective: To test the cognitive reserve hypothesis in a large sample of MS patients from a single centre (Graz), using an extended set of MRI-metrics.

Patients and methods: The cohort comprised 137 patients (33 CIS, 92 RRMS, 12 SPMS). Cognition was assessed by the "Brief Repeatable Battery of Neuropsychological Tests" (composite Z-scores of all

subtests). The following MRI-parameters were obtained at 3T: T2-lesion load (T2-LL), normalised brain volume (global brain volume loss), third ventricle width (TVW, regional volume loss), basal ganglia R2*-values (iron accumulation) and magnetization transfer ratios.

Hierarchical regression models served to identify the strongest MRI-predictor for cognitive function. To assess if EA moderates the effect of disease severity on cognition, MRI-metrics, EA and the interaction between EA and MRI-parameters were step-wise included in the model, after controlling for sex, age and disease duration.

Results: T2-LL was the strongest MRI-predictor for cognition. Higher T2-LL and increased TVW independently correlated with worse cognition. Higher EA predicted better cognition. These effects were moderated by interactions between T2-LL, TVW and EA. Higher EA reduces the negative effect of T2-LL and TVW on cognition.

Conclusion: In this cohort, higher EA attenuated the effects of T2-LL and TVW on cognition.

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Abstract – WCN 2013

No: 1268

Topic: 6 – MS & Demyelinating Diseases
Reversible splenial lesion in a Caucasian adult

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Background: Reversible lesion in the splenium of the corpus callosum (SCC) caused by various agents and conditions was previously defined, usually with mild encephalopathy/encephalitis without residual symptoms. In few days or weeks, complete radiologic regression is typical. Most cases are pediatric ones reported from South-East Asia.

Objective: To present a case of mild encephalopathy with reversible corpus callosum splenial lesion (MERS) in a Caucasian adult.

Methods: Case report.

Results: A 32-year old Caucasian male with unremarkable clinical history was referred because of severe headache and fatigue developed after 3 weeks of upper respiratory tract infection. Physical and neurological examination was normal. MRI of the head showed solid ovoid T2-weighted hyperintense lesion in the center of the SCC with restricted diffusion and without contrast enhancement. Laboratory tests, routine and immunoelectrophoretic examination of the CSF were normal. MERS was diagnosed based on the typical MRI finding and clinical presentation. CSF, serum, throat swab and stool were examined for different viruses (entero-, rota-, adeno-, influenza- and parainfluenza viruses and retrovirus) and bacteria (Borrelia, Mycoplasma, Chlamydia) with negative results. Fatigue and headache gradually improved. In two weeks, the size of the lesion in the SCC slightly increased but the high diffusion signal nearly resolved; complete radiographic regression was seen after 10 weeks on MRI.

Conclusion: Our case expands the population spectrum of MERS presenting the first case of a Caucasian adult. MERS can occur at any age and population, although South-East Asian predominance is remarkable.

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Abstract – WCN 2013

No: 1234

Topic: 6 – MS & Demyelinating Diseases
The innate immune receptor CD14 is important for lymphocyte migration across the blood–brain barrier in EAE

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Multiple sclerosis (MS) is a chronic autoimmune disease of the central nervous system, histopathologically characterized by inflammation, demyelination and gliosis. A disease-promoting role of the innate immune system has been proposed, based e.g. on the observation that innate immune receptors modulate disease severity of experimental autoimmune encephalomyelitis (EAE). Recent studies of our group provided first evidence for a key role of the innate immune LPS receptor (CD14) in the pathophysiology of EAE. CD14 deficient EAE mice showed increased clinical symptoms and enhanced infiltration of monocytes and neutrophils in brain and spinal cord. In the current study, we further investigated the cause of this disease aggravation and examined T cell activation and inflammatory cytokine production by FACS analysis and T cell migration capacity over the blood brain barrier by in vitro adhesion and transmigration assays. In the results, we observed a significantly increased migration of CD14 deficient lymphocytes across an endothelial monolayer. In contrast, we did not see any difference in T cell activation, cytokine production or lymphocyte adhesion to endothelial cells. Thus, the key innate immune receptor CD14 may play a pathophysiological role in the migration of inflammatory cells across the blood–brain barrier in EAE.

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Abstract – WCN 2013

No: 1255

Topic: 6 – MS & Demyelinating Diseases
VZV encephalitis in a multiple sclerosis patient treated with natalizumab

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Background: Natalizumab is a humanized monoclonal antibody approved for the treatment of patients with RRMS who have failed other disease-modifying therapies or who have aggressive disease. In MS clinical trials, herpes infections (varicella-zoster virus, herpes simplex virus) occurred slightly more frequently in natalizumab-treated patients than in placebo-treated patients (7.1% vs. 6%). In post-marketing experience, there have been three published cases of serious CNS infections due to HSV (one of which was fatal) and one report of VZV meningitis. Reactivation of latent herpes infections due to impaired immune surveillance on natalizumab therapy has been proposed.

Case report: A 46-year-old woman with a 10 year history of RRMS, previously treated with s.c. IFβ-1b, developed worsening headache and vertigo two days after receiving her 15th treatment with natalizumab in February 2013. Five days after symptoms onset a LP was performed, which showed elevated proteins (0,86) and WBC count (95 cells). PCR for VZV was positive whereas PCR for JCV was negative. Brain MRI showed no additional lesions or meningeal enhancement. The patient improved gradually after a 21-day course of intravenous acyclovir. Natalizumab treatment was discontinued and glatiramer-acetate has been introduced.

Conclusion: To our knowledge, this is the first report of a VZV encephalitis in a MS patient treated with natalizumab. Our case supports the hypothesis that treatment with natalizumab is associated with reduced immune surveillance resulting in the reactivation of latent herpes infections.

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Abstract – WCN 2013

No: 1212

Topic: 6 – MS & Demyelinating Diseases
CXCL13 CSF level inversely correlates with duration of disease in primary progressive multiple sclerosis

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Background: Neuroaxonal degeneration underlines PP MS, but the extent to which immune-mediated mechanisms operate is still unclear. Chemokines are important mediators in MS. The CXCL13 has an important role in recruitment to the CNS of B cells and RANTES(CCL5) for activated T cells and monocytes.

Objective: To examine the CSF and serum CXCL13 and CCL5 levels in PPMS, compare the results with RR MS and control group of other noninflammatory neurological disorders. Additionally we evaluated correlation between CSF and serum levels of studied chemokines with age, duration of the disease, IgG index and EDSS level.

Patients and methods: 12 patients with PP MS, 15 with RR MS and 13 controls have been included in the study. ELISA method was used for chemokine measurement. Kruskal–Wallis test and Spearman rank test served for statistical analysis.

Results: In PP MS CSF CXCL13 and CCL5 levels were significantly higher ($p < 0.005$) in comparison with those of control group. CSF CXCL13 level inversely correlated with the duration of the disease. In RR MS CSF CXCL13 level was significantly higher in comparison with that of control group. No significant differences in CSF and serum CXCL13 and CCL5 levels have been observed between PP and RR MS patients.

Conclusion: Results demonstrate the involvement of the CXCL13 and CCL5 chemokines in the immunopathogenetic mechanisms both in PP and RR MS patients. CXCL13 level inversely correlated with the duration of the disease in PP MS, but not in RR MS, showing in that aspect differences between these two forms of MS.

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Abstract – WCN 2013

No: 971

Topic: 6 – MS & Demyelinating Diseases Anxiety in patients with multiple sclerosis

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Introduction: Anxiety is common but often unrecognized and untreated disorder in patients with multiple sclerosis.

Objectives: The aim of our study was to determine the prevalence of anxiety in MS patients and to determine possible correlation with fatigue, sexual dysfunction and disorders in the fields of physical and emotional health.

Material and methods: In our study, we used Multiple Sclerosis Quality of Life-54 (MSQoL-54), Fatigue Severity Scale, and Hamilton Anxiety Rating Scale. Statistical data processing, we used the Pearson correlation test and Student's t test.

Results: We enrolled 120 MS patients (68 females and 52 males; average age 47.36 years, mean disease duration 17 years). Normal values for the Hamilton Anxiety Rating Scale were found in 24 patients (20%), while in 96 or 80% we found anxiety (8(6.6% – mild, 13.33% – moderate and 60% – severe). Statistically significant negative correlation was observed between anxiety and MSQoL54 Emotional Well-Being ($r = -0.563$, $p = 0.01$), Energy ($r = -0.540$, $p = 0.02$), the Physical Component Summary Score ($r = -0.601$, $p = 0.0001$) and Mental Component Summary Score ($r = -0.590$, $p = 0.01$). We find the positive significant correlation between values of Fatigue Severity Scale and anxiety ($r = 0.620$, $p = 0.0001$). Also we found statistically significant negative correlation between the Sexual Dysfunction and anxiety ($r = -0.418$, $p = 0.022$).

Conclusions: Anxiety has significant association with mental and physical health disorders, sexual dysfunction and fatigue in MS patients. Unlike other aspects of MS, anxiety is treatable and with proper treatment of these patients, the quality of their life could be much better.

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Abstract – WCN 2013

No: 1001

Topic: 6 – MS & Demyelinating Diseases Impact of smoking on mortality and life expectancy in multiple sclerosis

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Objective: To investigate the relationship between smoking and mortality in a cohort of Multiple Sclerosis patients.

Method: Clinical and demographic characteristics of 968 individuals were obtained from a large population-based cohort of MS patients. Records were linked to the NHS data to obtain data about death on Dec 2012 (index date). Patients were followed from onset age to death age or index date, whichever occurred first. The impact of sex, disease clinical type at onset, ≥ 1 year exposure to treatment, year of MS onset and smoking status (ever vs. never) were investigated using Cox proportional hazard model.

Results: Of 923 patients with clinically definite MS and full data for analysis, 80 (47 male and 33 female) were deceased at index date. Mean survival age was 77.3 years (95% CI: 75–79). Compared with ever-smokers, never-smokers lived 7 years less (81 vs. 74). Mean MS survival was 56.1 (95% CI: 53–59) in never-smokers and 43 (95% CI: 40–46) in ever-smokers. Smokers were at higher risk for death; hazard ratio 2.24 (95% CI: 1.30 to 3.85; $P = 0.004$) than never-smokers after stratifying by sex and controlling for onset age, initial type of disease (RR vs. PP) and ≥ 1 year exposure to treatment. Compared to UK general population, standardised mortality rates were 1.71 (95%CI: 1.37–2.13) for all patients, 2.64 (95%CI: 1.97–3.53) for ever-smokers and 1.14 (95%CI: 0.74–1.75) for never-smokers.

Conclusion: Mortality in MS is in part due to smoking. Smoking is a significant yet preventable risk factor for death in patients with MS.

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Abstract – WCN 2013

No: 123

Topic: 6 – MS & Demyelinating Diseases Lipid profile in interferon and glatiramer acetate using patients with multiple sclerosis

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Background: In studies focusing on MS patients that claim there is a change in lipid profile depending upon drugs being used has been found to have a difference in cholesterol and triglyceride levels. In our study, we aimed to compare the lipid profile of MS patients using interferon or glatiramer acetate of untreated MS patients and control group of migraine patients, and to investigate the relation retrospectively along with the disease progress in both groups.

Materials and methods: This study included 42 MS patients under treatment (group 1), 19 untreated MS patients (group2) and 15 migraine patients (group 3).

Sociodemographic features, Expanded Disability Status Scores (EDSS), disease characteristics of patients and the lipid profiles were recorded through a form. Lipid profiles were measured before treatment three times for group 1 with an average of 10 and After 18 months of treatment, lipid profiles were measured two times for group 2 and one time for group 3. SPSS for Windows 18 was used for statistical analysis.

Results: There was no statistically significant difference between the values of group one before treatment and after treatment. There was no correlation between these values and the EDSS's of patients.

There was no statistically significant difference between the second measurements of group 1 and group 2 ($p < 0.05$).

There was no statistically significant difference between all three groups before and after treatment ($p < 0.05$).

Conclusion: Our results showed that lipid profiles of patients don't change under MS treatment, and don't have relation with activity of disease.

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Abstract – WCN 2013

No: 1050

Topic: 6 – MS & Demyelinating Diseases

Features of the first clinical symptoms of multiple sclerosis in the population of Tomsk region Russian Federation

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Aims: Some symptoms of multiple sclerosis (MS) at different frequencies are found in the first episode and in the developed stage of the disease. The aim of this study was to receive a feature of the first clinical episode of MS in the Siberian region of Russian Federation.

Method and materials: The data of the patients with MS in the Tomsk region in the period from 2003 to 2012 have been examined.

Results: Often the first clinical symptoms of demyelination were disorders of the cranial nerves, especially the optic neuritis. Pyramidal disorders in patients were presented as central mono-, hemi-, para- and tetrapareses with varying severity. The sensation disorders were diagnosed as decrease in vibration, pain, touch, sense of temperature and position. In the separation of patients by gender, predominance of any symptoms, as the manifestation of MS, was not found. We compared the data of the first clinical symptoms of the disease in 2012 with 2003. The results of the study showed an increase in 2012 cases of optic neuritis by 11.6% and cases of sensation disorders by 5.5%.

Conclusion: Diagnosis of MS in clinical practice is difficult. Symptoms of the disease are similar the symptoms of other pathological processes in the nervous system and don't have particular features. Clinical features of the first demyelinating symptoms in MS patients may determine the prognosis of the disease progression, treatment response and further patients management.

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Abstract – WCN 2013

No: 845

Topic: 6 – MS & Demyelinating Diseases

Atorvastatin protects NSC-34 motor neurons against oxidative stress through the activation of PI3K, ERK and free radical scavenging

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Objective: Although statins, hydroxymethylglutaryl coenzyme A (HMG-Co A) reductase inhibitors, are generally used to decrease circulating cholesterol levels, they have also been reported to have neuroprotective effects through diverse mechanisms. However, recent reports have shown controversial results as to whether statins might be harmful in patients with amyotrophic lateral sclerosis (ALS). In this study we investigated the direct effect of atorvastatin on motor neuron like cells (NSC-34D) against oxidative stress.

Material and methods: To evaluate the effect of atorvastatin and/or hydrogen peroxide on NSC-34D cells, the cells were treated with several conditions. 3-(4,5-dimethylthiazol-2-yl)-2,5-diphenyltetrazolium bromide (MTT) assay and trypan blue stain were performed for the evaluation of viability. Free radical level and intracellular signaling proteins were evaluated with fluorescent probe 2',7'-dichlorodihydrofluorescein diacetate (DCFH-DA) and western blotting, respectively.

Results: Atorvastatin protected NSC-34D cells against oxidative stress in a concentration-dependent manner. This neuroprotective effect of atorvastatin was blocked by LY294002, a phosphatidylinositol 3-kinase (PI3K) inhibitor and FR180204, a selective extracellular signal-related kinase (ERK) inhibitor. Atorvastatin treatment increased the expression levels of p85aPI3K, phosphorylated Akt, phosphorylated glycogen synthase kinase-3 β , Bcl-2 and phosphorylated ERK which are proteins related to survival, and decreased the levels of cytosolic cytochrome and cleaved caspase-3, cleaved caspase-9 and Bax which are associated with death, in oxidative stress-injured NSC-34D cells.

Conclusion: We conclude that atorvastatin has neuroprotective effect against oxidative stress in motor neurons via the activation of the PI3K pathway, ERK pathway and free radical scavenging. These findings indicate that statins could be helpful in protecting motor neurons.

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Abstract – WCN 2013

No: 891

Topic: 6 – MS & Demyelinating Diseases

Progressive multifocal leukoencephalopathy in an immunocompetent patient

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Introduction: Progressive multifocal leukoencephalopathy (PML) is caused by the JC polyomavirus, which occurs in immunocompromised patients, HIV or patients under immunosuppressant treatment (natalizumab, rituximab ...), associated with a poor prognosis. Few cases of PML on immunocompetent patients have been reported.

Case report: Male, 75 years-old with no previously known diseases, consulting for rapidly progressive acalculia and visual field disorder that progresses with left hemiparesis and aphasia. The MRI showed a cortical-subcortical right parietal lesion, with no Gd enhancement, which progressed during admission. Given the rapid evolution of the lesion and the clinical symptoms, a brain biopsy was performed, showing atypical proliferation of oligodendrocytes and lipid macrophages. Further immunohistochemical tests showed positive polyomavirus antigens inside the glia. All of these features made the diagnosis of definite PML. HIV studies were negative. Study of lymphocytic population was normal. The CSF study conducted prior the biopsy showed a high number of copies of the JC virus. Due to progression of symptoms the patient finally died months later.

Conclusions: Only a few cases of PML on immunocompetent patients have been reported. Paradoxically, in these cases, the prognosis is

worse than in immunocompromised patients. PML should be included in the differential diagnosis of rapidly progressive white matter lesions, even in immunocompetent patients.

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Abstract — WCN 2013

No: 929

Topic: 6 — MS & Demyelinating Diseases

Susac's syndrome — A case with unusual cardiac vestibular and imaging manifestations

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Susac's syndrome is a rare, idiopathic, microangiopathy affecting the brain, retina, and cochlea.

Methods: Detailed description of a patient with Susac's syndrome.

Results: A 38 year old patient developed dizziness and transient right eye visual disturbance one day prior to his admission. He was examined in the hospital because of confusional state with restlessness that lasted 24 h. Three days after his admission he developed right eye cilioretinal artery occlusion. In the fifth day of hospitalization he developed bilateral asymmetric deafness, tinnitus and upbeat nystagmus. Brain MRI was normal. CT angiography was normal. LP showed significant albumino-cytologic dissociation. A detailed oto-laryngological and ophthalmological work up was performed. Of note was the persistence of significant bradycardia without additional cardiological abnormalities.

Work up which included brainstem evoked potentials, Otoacoustic-electrocochleography, caloric tests and electronystagmography was positive for a significant peripheral and central vestibular and auditory involvement. The patient was successfully treated with IV steroids and Immunoglobulins. Brain MRI following a repeated episode of vertigo 3 month later showed a diffuse T2 hyper intense, ill-defined, left hemispheric lesion superior to the Lateral ventricles not seen in previous two MRI scans.

Conclusions: This patient developed the typical clinical picture of Susac's syndrome. However, additional features are unusual: Persistent bradycardia, significant vestibular involvement of both peripheral and central origin, and the late appearance of atypical white matter lesions on MR scan. The possibility of microangiopathy involving the heart and central auditory and vestibular involvement in the absence of initial MRI findings is discussed.

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Abstract — WCN 2013

No: 737

Topic: 6 — MS & Demyelinating Diseases

In vitro persistence of saffold virus type 3 which is isolated from cerebrospinal fluid of the patient with aseptic meningitis

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Background and objective: Saffold virus (SAFV) was identified in 2007 as a novel human Cardiovirus. SAFV type 3 (JPN08-404) has been isolated from cerebrospinal fluid of the patient with aseptic meningitis. This finding is of interest since Theiler's murine encephalomyelitis virus (TMEV), which is the closely related virus, is known to cause a multiple sclerosis-like syndrome in mice. During demyelination, TMEV persists in mouse. Therefore, we studied the possibilities of the in vitro persistence of SAFV.

Material and methods: HeLa cells were used as host cells. SAF404 virus derived from infectious cDNA clone of JPN08-404 was used as a challenge virus. The cells persistently infected with SAF404 were analyzed by Western blotting, immunocyto-staining and plaque assay. To investigate the mechanisms of SAFV persistence, the treatment with anti-IFN antibody was performed. Furthermore, the virus binding assay in HeLa cells maintained with CS or FCS was performed.

Results: SAFV persistence was first demonstrated in this study. The persistence of SAF404 was not influenced by the treatment with anti-IFN- α/β antibody, although TMEV persistence was collapsed. SAFV persistence was collapsed by the maintenance with FCS. Furthermore, the virus binding assay indicated that the densities of the virus binding molecule(s) were significantly higher in HeLa cells cultured with FCS.

Conclusion: The present study demonstrated that SAFV is able to persist in human-derived cell line. Furthermore, it was suggested that SAFV persistence may depend on the receptor densities. The present findings of the SAFV persistence will be helpful for further studies on the SAFV pathogenicity.

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Abstract — WCN 2013

No: 539

Topic: 6 — MS & Demyelinating Diseases

Clinical inflammatory activity associated with response to second-line treatments in multiple sclerosis

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Background: Suboptimal outcome on first-line MS treatments often necessitate initiation of second-line treatment. There is limited information on how to choose a second-line agent.

Objective: We examined the ability of 11 candidate demographic and clinical factors to predict treatment response in an observational MS cohort.

Patients and methods: Patients treated with one of the four most commonly used second-line MS treatments in our center at the time of this study were selected: cyclophosphamide, daclizumab, mycophenolate mofetil, and natalizumab. Outcomes were time to subsequent inflammatory event (clinical relapse or new MRI activity) and occurrence of progression in disability confirmed at six months. The association between potential predictors and the time to events was investigated using univariate Cox proportional hazards regression models. An additional analysis regarding whether patients remained disease-activity-free on specified measures for two years after treatment start was also performed.

Results: 101 cyclophosphamide, 57 daclizumab, 83 mycophenolate and 140 natalizumab patients were included. In all treatment groups except natalizumab, and among studied factors, those generally known to be associated with inflammatory disease activity (younger age at treatment start, shorter disease duration, and higher baseline relapse rate) were associated with more disease activity on second-line treatments. A similar set of factors were negative predictors for a disease-activity-free course independent of treatment duration.

Conclusion: High baseline inflammatory activity is associated with suboptimal response to the studied drugs, except for natalizumab. Identification of underlying mechanisms may aid in further optimizing second-line treatment response in MS.

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Abstract – WCN 2013**No: 711****Topic: 6 – MS & Demyelinating Diseases****Bickerstaff brainstem encephalitis without ophthalmoplegia: A reappraisal of current diagnostic criteria**N. Yuki, B. Wakerley. *National University of Singapore, Singapore, Singapore***Background:** External ophthalmoplegia is a cardinal feature of Bickerstaff brainstem encephalitis.**Objective:** Clinical and immunological evaluation of “incomplete” Bickerstaff brainstem encephalitis.**Methods:** We studied 2 patients with post-infectious brainstem syndromes who presented at National University Hospital Singapore. Laboratory work-up included measurement of anti-ganglioside antibodies.**Results:** Both patients displayed hypersomnolence and cerebellar-like ataxia in the absence of external ophthalmoplegia and carried high serum titers of IgG anti-GQ1b antibodies, strongly indicative of Bickerstaff brainstem encephalitis.**Conclusion:** Ophthalmoplegia can be absent or incomplete in Bickerstaff brainstem encephalitis, and the absence of this clinical feature should not exclude Bickerstaff brainstem encephalitis from the clinicians' differential. Such cases of incomplete BBE could be defined as “ataxic hypersomnolence without ophthalmoplegia”.

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Abstract – WCN 2013**No: 703****Topic: 6 – MS & Demyelinating Diseases****Veins in plaques of multiple sclerosis patients – A longitudinal magnetic resonance imaging study at 7 Tesla**A. Dal-Bianco^a, G. Grabner^b, S. Hametner^c, M. Scherthaner^b, C. Kronnerwetter^b, M. Weber^b, C. Vass^d, K. Kircher^d, A. Reitner^d, E. Auff^a, K. Vass^a, H. Lassmann^c, S. Trattnig^b. ^aDepartment of Neurology, Medical University of Vienna, Vienna, Austria; ^bDepartment of Radiology, Medical University of Vienna, Vienna, Austria; ^cCenter for Brain Research, Medical University of Vienna, Vienna, Austria; ^dDepartment of Ophthalmology, Medical University of Vienna, Vienna, Austria**Background:** Multiple Sclerosis (MS) is a chronic inflammatory disease of the central nervous system associated with demyelination, axonal loss and neurodegeneration. MS plaques are generally found to be centered by a vein. The role of blood vessels gains again attention due to ultra-highfield magnetic resonance imaging (MRI).**Objective:** Susceptibility-weighted-imaging (SWI) at 7 Tesla MRI allows precise depiction of veins. The purpose of this study was to monitor the intraplaque veins over a period of 2 years in plaques and corresponding normal appearing white matter (NAWM) of patients with relapsing–remitting and secondary–progressive MS compared to age-matched controls.**Patients and methods:** Ten MS patients and nine age-matched control subjects were enrolled in a prospective 2-year follow-up annual clinical and 7 T-Flair-SWI-MRI protocol. Veins were manually segmented in plaques, their contralateral NAWM and in the white matter of controls. Proportion of veins was assessed by dividing the volume of veins by their according tissue volume. Non-parametric tests were used for area-wised comparison.**Results:** Established MS plaques showed a significantly higher proportion of SWI-detected veins compared to corresponding control tissue without significant change within 2 years. We found a trend of an increasing venous proportion in pre-plaque areas and a significant

increase of venous proportion in newly developed plaques compared to controls. Proportion of veins in the NAWM was in accordance with values of matched control-areas.

Conclusion: Inflammation-induced metabolic activity, vasodilatation and angiogenetic factors may contribute to the significantly higher proportion of intraplaque veins compared to control and NAWM tissue.

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Abstract – WCN 2013**No: 702****Topic: 6 – MS & Demyelinating Diseases****Changes of visual evoked coherence in patients with multiple sclerosis detected by EEG**P. Schwenkreis^a, M. Tegenthoff^b, O. Hoffken^a, P. Stude^a, M. Lenz^a, I. Nowak^a. ^aBerufsgenossenschaftliches Klinikum Bergmannsheil Bochum, Bochum, Germany; ^bNeurology, Berufsgenossenschaftliches Klinikum Bergmannsheil Bochum, Bochum, Germany**Background:** Central encoding of higher functions and decoding of sensory inputs result from task-specific neuronal networks in which processing of the information is based on synchronization depending on functional connectivity. In multiple sclerosis demyelination and axonal damage lead to altered signal processing of participating networks. Our aim was to detect disturbances of neuronal synchronization in the visual system.**Methods:** During stimulation a 12-channel-EEG was recorded from 23 subjects with relapsing–remitting MS (6 males, 17 females, average EDSS: 3.5) and 19 controls. Stimuli consisted in the conventional pattern reversal checkerboard with constant total luminance (full field, binocular, visual angle 6°, ISI 1 s, 300 stimuli). Single stimulus test series (between 200 ms before and 1000 ms after the stimulus) were segmented in 136 overlapping segments (delta t 8 ms) each of 128 ms duration to create a sliding analysis. We calculated conventional VEPs and area-specific spectral power and coherence (as a measure of functional coupling) between “intra”-occipital and between “intra”- and “extra”-occipital electrode positions.**Results:** VEPs and spectral power did not differ significantly between patients and controls. “Intra”-occipital coherence increased whereas the “intra–extra”-occipital coupling decreased. However, in patients the “intra”-occipital increase and the “intra–extra”-occipital decrease were significantly more pronounced. Furthermore the “intra”-occipital increase correlated with the activity and the “intra–extra” occipital decrease with duration of the disease.**Conclusions:** Coherence in patients shows a more intense intra-regional occipital coupling and an enhanced coherency decline to cortical areas not participating in information processing. These results indicate reorganization as a consequence of disturbed fiber connections to provide adequate short term resources of visual information processing.

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Abstract – WCN 2013**No: 829****Topic: 6 – MS & Demyelinating Diseases****Application of the modified McDonald criteria of multiple sclerosis in Chinese demyelinating disease patients**R. Li, S. Cheng, K.L. Shiu, E. Yeung, C.N. Lee, M. Au Yeung, C.M. Cheung, T.H. Tsoi. *Medicine, Pamela Youde Nethersole Eastern Hospital, Hong Kong, Hong Kong S.A.R.*

Background: The International Panel on diagnosis of multiple sclerosis (MS) has revised the McDonald criteria in 2010. There is scarce data on its application in the Chinese population.

Objective: To evaluate the application and usefulness of the 2010 McDonald criteria in Chinese patients with idiopathic demyelinating diseases.

Methods: All patients who have been seen for idiopathic demyelinating diseases in a single regional hospital from 1996 to 2012 were recruited. They were retrospectively analyzed based on their most updated diagnosis as of March 2013. The Poser criteria for clinically definite MS (CDMS), the 2005 and 2010 McDonald criteria for MS were applied to all subjects and compared.

Results: A total of 114 patients were recruited. After a mean disease duration of 8.0 years, 38 patients (33.3%) converted from clinically isolated syndrome (CIS) to CDMS; 40 (35.1%) to MS by the 2005 criteria; and 44 (38.5%) by the 2010 revision. 18 (15.8%) converted to neuromyelitis optica (NMO), and 4 (3.5%) to recurrent optic neuritis or brainstem disease. Of the 38 patients with CDMS, the mean conversion time from CIS is 26.9 months, but is shortened to 22.7 and 20.4 by the 2005 and 2010 criteria respectively. 26.3% of CDMS patients already fulfilled the 2010 criteria by the initial MRI. None of the patients with other recurrent demyelinating diseases, including NMO, fulfilled the 2010 criteria on presentation.

Conclusion: The latest revision of the McDonald criteria is applicable to the Chinese population. It can speed up the diagnosis of MS, and is highly specific.

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Abstract – WCN 2013

No: 790

Topic: 6 – MS & Demyelinating Diseases

Clinical neurological dynamic after intervention treatment in patient with multiple sclerosis (MS)

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Background: Etiology of MS is unknown. It is believed to be an autoimmune disease. MR venography and postmortem studies have demonstrated a topographic correspondence between MS plaques and cerebral venous abnormalities. In several epidemiological observations the prevalence of chronic cerebrospinal venous insufficiency (CCSVI) in MS ranges from 56% to 100%.

Objective: To evaluate effect of transluminal balloon angioplasty (TPA) on clinical course of MS in patients with CCSVI.

Material and methods: 26 patient with different forms of MS were included in the study: Relapsing-Remitting (RR) – 13; Secondary-progressive (SP) – 7; Primary-Progressive (PP) – 5; Progressive-Relapsing (PR) – 1 patient. In all those patients different degrees of stenosis were observed in proximal parts of jugular veins (right or left). TPA was performed in all cases. We have used several methods for assessment of patients before TPA and at follow up: Clinico-Neurological assessment (Kurtzke Expanded Disability Status Scale (EDSS) and the Multiple Sclerosis Functional Composite (MSFC) (every 6 month); MRI (every 6 month).

Results: In 3 cases – positive dynamic (2-MS-RR, 1-MS-SP); In 18 cases – stabile dynamic (11 – MS-RR, 4 – MS-SP, 4 – MS-PP); In 5 cases – unknown dynamic (3 – MS-SP, 1 – MS-PP, 1 – MS-PR).

Conclusion: Chronic CCSVI can play a certain role in the development of MS. It is recommended to perform selective venous angiography for patients with MS to diagnose CCSVI. In case of venous insufficiency TPA may appear an effective treatment strategy

for the patients with MS. Future randomized studies are warranted to establish the efficacy of this new treatment for MS.

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Abstract – WCN 2013

No: 794

Topic: 6 – MS & Demyelinating Diseases

Optical coherence tomography in differential diagnosis of neuromyelitis optica and multiple sclerosis

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Objective: Optical coherence tomography (OCT) is a novel noninvasive method and identifies the retinal nerve fiber layer thickness (RNFL) and macular volume. We evaluated OCT as a differential diagnostic tool in neuromyelitis optica (NMO) spectrum and multiple sclerosis (MS).

Method: OCT and visual function testing were performed in 25 NMO spectrum patients and 40 optic neuritis patients as a first clinical presentation of MS.

Results: RNFL thickness was 56.9 µm in patients with NMO while 89.3 µm in MS patients and 105.2 in normal controls ($p < 0.001$). Macular volume loss was more prominent in NMO patients.

Conclusion: Our results showed that RNFL thickness reduction was the most significant in NMO patients when compared to optic neuritis patients as a first symptom of MS. We suggest that retinal damage is very severe in NMO and OCT is a helpful tool to differentiate optic neuritis due to NMO than MS optic neuritis.

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Abstract – WCN 2013

No: 756

Topic: 6 – MS & Demyelinating Diseases

Biomarkers of the progressive changing of the brain plasticity in patients with relapsing-remitting multiple sclerosis: FMRI and in-vivo ¹H MRS study

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Background: From fMRI- and MRS-data, we obtain biomarkers of the progressive changing of the brain plasticity in RRMS-patients.

Objective: We quantify the pattern of motor pathway activation during the simple unilateral finger taping, and mild mechanosensory stimulation, and study cerebral metabolism in RRMS-patients.

Patients and methods: Two groups of patients are studied by fMRI and MRS, 1.5 T SIGNA (GE). The 1st group consists of 23 RRMS-patients (18f, 5 m, 23–39 yo). The 2nd group – 20 volunteers (12 m, 8f, 21–35 yo).

Materials and methods: FMRI images were obtained using EPI:TR/TE = 3000/60 ms. Model based on two step ICA analyses was done using GLM design matrix (FEAT, and MELODIC), FSL5.0. Six bilateral ROIs (supplementary motor area (SMA), precentral gyrus (PG), putamen (P), globus pallidus (GP), thalamus (T), and subthalamic region (SR)) were using for cluster analysis. To quantify the degree of asymmetric brain activation an asymmetry parameter $AP = (CL - CR) / (CL + CR)$ was calculated (where CL, CR-cluster volume from the left, and right ROI). Spectra are recorded in all above matched ROIs with SVSSTEAM:TR/TE = 1500/144 ms.

Results: In the 1st group (in PG and P) $AP = (-0.11$ to $0.09)$. In the SMA, GP, T, SR $AP = (-0.29$ to $-0.43)$ and $(0.15$ to $0.26)$ in the RRMS patients with right and left hand predominant symptoms. In

the 2nd group the symmetric activation in all brain regions evaluated with $AP = (-0.12 \text{ to } 0.037)$ were found. We have found the progressive decreasing NAA/Cr in the PG, and SMA, that is associated with poorer motor function.

Conclusion: fMRI and MRS data give us new quantitative approach for understanding metabolic basis of decreasing of the brain plasticity in RRMS-patients.

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Abstract – WCN 2013

No: 779

Topic: 6 – MS & Demyelinating Diseases

Electrophysiological evaluation of dysphagia in the mild or moderate patients with multiple sclerosis

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Objective: This study aims to evaluate the presence of subclinical dysphagia in mild MS patients using electrophysiologic methods.

Study design and methods: A prospective study of 51 patients with RRMS and 18 age-matched healthy adults. Electrophysiologic evaluations were performed on submental EMG activity during attack, after the attack and in remission period. Dysphagia limit was determined during as single and 50 ml of sequential water swallowing in all patients and control group to detect dysphagia objectively.

Results: Clinical dysphagia was found in 12% of patients while the electrophysiological dysphagia was encountered in 33% of MS patients. Dysphagia limit was in normal limits in normal subjects while it was pathological in 23% MS patients. Duration of swallowing signal of submental muscles in all MS patients was longer than in normal subjects ($P = 0.001$). The forebursts of sequential water swallowing was recorded significantly more often in patients in remission period than in patients in attack or after attack groups ($P = 0.003$). The total swallowing duration was prolonged in MS patients ($P = 0.001$). The existence of dysphagia was significantly higher among MS patients who had higher disease disability ($P = 0.001$).

Conclusion: This is the first study investigating subclinical dysphagia from the electrophysiologic aspect in RRMS patients. In agreement with our data, dysphagia is common in MS patients even in early stage or mild forms of the disease. The presence of subclinical dysphagia should be searched even in the absence of any clinical signs because dysphagia may impact the daily life negatively and increase the morbidity.

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Abstract – WCN 2013

No: 792

Topic: 6 – MS & Demyelinating Diseases

Two-stage disability progression in multiple sclerosis in Tunisia

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Aims: Analyse the curse profile of a 600 Tunisian patients with MS followed prospectively between 2000 and 2010.

Methods: We report the results of a prospective database based study of 600 patients in the National Institute Mongi Ben Hamida of Neurology of Tunis. These patients were classified MS according to Mc Donald criteria and followed regularly using a CRF and entered in a database created since 2000.

Results: The distribution between remittent relapsing (RRMS) and progressive form (PMS) joined the large reported data. The mean

duration from clinical onset to DSS 3 was 15.61 ± 3.16 years in the RRMS and 8.78 ± 0.43 years in the PMS.

The mean duration from DSS 3 to DSS 6 was 4.15 ± 4.23 years for the RRMS and 4.33 ± 3.93 years for PMS without significant difference ($P = 0.6$).

These results support the hypothesis recently reported in the literature, of the presence of two independent stages in the natural history of MS: an inflammatory stage and a second neurodegenerative stage which is independent from the focal inflammatory lesions. In addition, the mean duration from DSS 3 to DSS 6 was shorter in our study in comparison with the Europeans ones supporting the data that MS is more severe in Tunisian patients.

Conclusion: Multiple sclerosis in Tunisia is more severe than MS in European countries.

This concept of a two stages disability progression disease is supported by the present study.

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Abstract – WCN 2013

No: 613

Topic: 6 – MS & Demyelinating Diseases

Low dose aspirin for MS-related fatigue: Results of a pilot, double-blind, randomized trial

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Fatigue is one of the most common and disabling symptoms in multiple sclerosis (MS), occurring in more than 60% of patients during the course of the disease. Despite high prevalence of fatigue in MS patients, its pathophysiology was not understood completely yet and this would difficult introduction of new medication for this symptom. Wingerchuk et al. for the first time assess the effectiveness of Aspirin (ASA) on MS related fatigue and show high dose ASA (1300 mg) would significantly decrease fatigue in these patients. According to severe complications in high dose use of ASA, we try to evaluate low dose ASA (80 mg) for primary fatigue in MS patients.

This was a single-centre, randomized, double-blind and parallel trial with 1:1 ratio allocation of two treatment groups of low dose Aspirin (80 mg) and placebo in outpatients with MS-related fatigue.

Ninety five patients with MS were randomized among them, 76 patients enter final analysis. No significant effect in FSS and MFIS was found using ANOVA for repeated measures when missing value were replaced. When we examine absolute changes for MFIS score, significant difference between two groups was seen (P value = 0.047, Mean difference = -8.49003 , CI = -16.84576 to -1.13430).

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Abstract – WCN 2013

No: 628

Topic: 6 – MS & Demyelinating Diseases

Role of serum TRAIL level and TRAIL apoptosis gene in multiple sclerosis and their relation with brain atrophy

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Background: One of multiple sclerosis (MS) presumed pathological mechanisms is failure of apoptosis of autoreactive T lymphocytes.

Objectives: To determine the relationship between tumor necrosis factor-related apoptosis-inducing ligand (TRAIL) mRNA gene expression ratio and serum TRAIL levels with MS and brain atrophy.

Materials and methods: This study was conducted on 53 relapsing remitting MS Egyptian patients and 25 matched healthy volunteers. The expression of TRAIL on peripheral blood lymphocytes was analyzed by RT-PCR, serum levels of soluble TRAIL (sTRAIL) was determined by ELISA and MRI brain for measurement of black holes and the bicaudate ratio (BCR) as a measure of brain atrophy were done for all patients.

Results: The serum TRAIL level was lower in multiple sclerosis patients compared to the control but no difference in the TRAIL mRNA gene expression ratio. No significant correlation detected between the serum TRAIL level and the TRAIL mRNA expression ratio in either group. No statistically significant correlation was found between the serum TRAIL levels or the TRAIL mRNA expression ratio with the number of black holes or the bicaudate ratio.

Conclusion: Apoptosis of T lymphocytes is deficient in MS patients which can be implicated in the treatment. No difference between TRAIL mRNA gene expression ratio between MS patients and controls.

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Abstract – WCN 2013

No: 675

Topic: 6 MS & Demyelinating Diseases

Late onset multiple sclerosis (LOMS): A diagnostic challenge

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Background: LOMS is an unusual entity presenting after the age of 50, commonly with progressive course and motor symptoms.

Objectives: We describe two cases of LOMS with diagnostic delay based on standard tests.

Patients and methods: A 67 years old woman with a previous stroke, suffered diplopia attributed to ischemic neuropathy. She developed progressive paraparesis, spastic gait, ataxia and urinary urgency during the following five years. A 56 years old smoker man, with hypertension and dyslipidemia, was studied due to a gait disturbance for fourteen months. Right lower limb weakness, vibratory hypoesthesia and hyper-reflexia in lower limbs were detected.

Results: Laboratory blood tests were normal. Case1 had CSF oligoclonal bands. Visual Evoked Potentials were normal, but Somatosensory Evoked Potentials (SSEP) were consistent with demyelization at cervical levels in both. 1.5 T MRI showed non-specific subcortical hyper-intensities and cervical protrusions without spinal cord lesions. Finally, 3 T MRI was conclusive showing non-enhanced hyper-intensities at C3, C5, C6, D9 cord levels and periventricular white matter in case 1, and at C2 and D6 levels in case 2. Both improved with corticosteroids pulses.

Conclusion: LOMS frequently presents as primary progressive, with greater tendency to affect the spinal cord. It is characteristic the diagnostic delay due to comorbidities, progressive form and nonspecific MRI. In our cases, a high suspicion and the results of SSEP led us to complete the study with 3 T MRI. We remark that in LOMS, standard tools may be non-diagnostic, requiring more specific tests (3 T MRI and SSEP) to confirm the diagnosis and try specific therapy.

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Abstract – WCN 2013

No: 662

Topic: 6 – MS & Demyelinating Diseases

Antiphospholipid antibodies in patients with multiple sclerosis or neuromyelitis optica in Ishikawa prefecture, Japan

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Background: Antiphospholipid syndrome (APS) is characterized by arterial and venous thrombosis and the presence of antiphospholipid antibodies (aPLs), including anticardiolipin antibody (aCL) and lupus anticoagulant (LAC). However, some patients with multiple sclerosis (MS) continuously test positive for aPLs.

Objective: In this study, we investigated the frequency of aPL positivity in patients with both MS and neuromyelitis optica (NMO) without APS-related clinical events, such as arterial and venous thrombosis.

Subjects: Fifty patients fulfilled the 2010 inclusion criteria for MS in 2010 and 13 of those patients fulfilled the 2006 revised NMO criteria.

Results: Eighteen percent of the patients with MS and 15% with NMO were continuously positive for 1 or more aPLs. Although phosphatidylserine/prothrombin complex antibodies (PS/PT) were found in both groups, LAC and beta2-glycoprotein I (GPI)-independent aCL were only positive in patients with MS. When LAC was positive in patients with MS, it was only demonstrated with the kaolin clotting time assays, not the dilute Russell's viper venom time (dRVVT) assays. No patient in either group was positive for GPI-dependent aCL. Interestingly, 8% of patients with MS and 15% with NMO were temporarily positive for aCL or LAC.

Discussion: The frequencies of aPL positivity were similar to those in previous reports; moreover, all of the patients exhibited low titers. Interestingly, although PS/PT is a key antibody for LAC, all patients with MS and LAC positivity tested negative for PS/PT. This result indicated that unknown autoreactive antibodies other than aPL may have crossreactivity with LAC.

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Abstract – WCN 2013

No: 642

Topic: 6 – MS & Demyelinating Diseases

Arterial and venous hemodynamics in patients with multiple sclerosis

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Background: Chronic cerebro-spinal venous insufficiency (CCSVI) is a new controversial theory of multiple sclerosis (MS) pathogenesis. Though the role of venous insufficiency in MS has not been fully explored surgical procedures are offered to MS patients.

Objective: The aim of the study was to evaluate extracranial and intracranial arterial and venous flow in MS patients.

Material and methods: 120 MS patients (mean age 38.9 ± 8.7 years; median EDSS – 3.3 range 0–8; disease duration 1–27 years) were compared with 80 healthy controls (mean age 34.7 ± 7.2 years). Arterial and venous flow was examined using a Philips duplex ultrasound machine.

A subject was considered CCSVI if ≥ 2 ultrasonographic criteria (Zamboni's five criteria) were fulfilled.

Results: No MS patient and no control had CCSVI. Internal jugular vein stenosis (venous cross-sectional area $\leq 0.3 \text{ cm}^2$) was the only criterion found for CCSVI (no significant differences $p = 0.87$ between MS patients and controls). Arterial blood flow (mean velocity, pulsatility index) was similar in both group, $p = 0.35$.

Conclusion: Our findings indicate that CCSVI has no role in multiple sclerosis.

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Abstract – WCN 2013

No: 656

Topic: 6 – MS & Demyelinating Diseases

A study of total-tau and phospho-tau (181) in the cerebrospinal fluid of patients with multiple sclerosis

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Biomarkers connected with clinical parameters and the process of MS could be very important for understanding and managing the disease.

The aim of the present study was the determination of total tau (tTAU) and phosphorylated tau (pTAU -181) concentrations in the CSF of MS patients vs controls and the correlation with clinical parameters (age, gender, type of MS, disability and treatment).

We examined 50 CSF samples, 40 patients with definite MS, 33 females (42.6 ± 11.9 years) and 7 males (42.4 ± 10.0 years), as well as 10 controls, 3 females (43.3 ± 7.4 years) and 7 males (40.3 ± 15.5 years), who didn't suffer from MS or any acute inflammation. 12 of the patients suffered from Clinical Isolated Syndrome (CIS), 22 from relapsing/remitting (RRMS), 3 from primary (PPMS) and 3 from secondary progressive type of MS (SPMS). The samples were centrifuged after the lumbar puncture and freeze-dried at -80°C . We used the Elisa technique and the commercial available kits (Inno-genetics, Belgium).

Our study indicated increased tTAU-Ag levels in older patients ($\rho = 0.371$, $p = 0.024 < 0.05$), while in the controls' group was demonstrated a reversed connection ($\rho = -0.679$, $p = 0.022$). In both (patients' and controls') groups, females demonstrated higher levels of tTAU ($p = 0.011$). The study of pTAU-181 showed an important correlation between PPMS and decreased levels ($p = 0.038$), while patients with more than a year duration of MS demonstrated higher concentrations ($p = 0.045$).

Our results indicated that tTAU could be correlated with the age and the sex of MS patients and phosphoTAU 181 with the type and the duration of the disease.

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Abstract – WCN 2013

No: 588

Topic: 6 – MS & Demyelinating Diseases

Endocrinologic disorders in patients with multiple sclerosis

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Background: Extranervous lesions are observed in multiple sclerosis (MS).

Objective: To detect endocrine disorders in MS and to investigate their correlation to the disease severity.

Patients and methods: We examined 85 patients with relapsing-remittent MS of various severity. EDSS and FS Kurtzke were used to describe patients' state, hormone concentrations defined using laboratory methods.

Results: The following endocrine disorders were diagnosed in 25 (23.8%) patients with mild (17 (68%)) to moderate (8 (32%)) MS

severity: cortisol 1.4 times exceeding normal levels ($p < 0.05$) in 10 (40%) patients that correlated to brainstem disorders (FS3) ($r = -0.29$, $p < 0.05$), but no other neurological lesions. 8 (32%) patients developed autoimmune thyroiditis with thyroid stimulating hormone concentration 8 times higher than normal ($p < 0.05$), and that-one of free thyroxine 2 times lower than normal ($p < 0.05$), that correlated to the movement impairment severity (FS1) ($r = -0.21$, $p < 0.05$), cerebellar (FS2) ($r = 0.55$, $p < 0.05$), sensory (FS4) ($r = -0.25$, $p < 0.05$), and pelvic disorders (FS5) ($r = 0.74$, $p < 0.05$). The increased for 38.2% prolactin level ($p < 0.05$) correlating to the neurologic disorders ($p < 0.05$) was found in 7 (28%) patients.

Conclusion: The correlation between cortisol, TSH and thyroxine, and prolactin levels and EDSS score was established ($r = 0.43$, $p < 0.05$). The possibility of the neurological state improvement by correcting patients' endocrine profile will be the subject of our future investigations.

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Abstract – WCN 2013

No: 609

Topic: 6 – MS & Demyelinating Diseases

I.V. autologous bone marrow derived mesenchymal stem cell in multiple sclerosis, double blind randomized clinical trial: Preliminary report of safety

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Introduction: Adult bone marrow derived stromal cells (MSC) were shown to induce immunomodulatory and neuroregenerative effects and to induce neuroprotection in the animal models. A randomized clinical trial, semi cross over, phase I/II study to evaluate the safety and efficacy I.V. injection of autologous bone marrow derived mesenchymal stem cell in patients with multiple sclerosis has been designed.

Methods: 30 MS patients (18–55 years, $3 < \text{EDSS} < 6.5$) who were resistant to the approved DMDs, were randomly recruited. After bone marrow aspiration the patients were randomly divided into 2 groups: early treatment group and delayed treatment group. Mesenchymal stem cells were transplanted I.V. to the first group and for second one injected after 6 months. All followed for immediate and late side effects, relapses, EDSS, RAO Test, MRI, CSF markers and quality of life questionnaire at the Baseline and after 1th, 3th, 6th and 12th months.

Results: Up to January 2013, 15 patients were injected and 8 out of them had their second injection. After 6 months of follow up, any clinical or laboratory adverse events and immediate clinical reactions within the first 6 h and between 48 h and 6 months of follow up were seen. Some patients got better with EDSS improvement. 2 patients had relapses and in one patient 3 severe disabling relapses occurred within 5 months. We did not detect any new GAD + lesions on MRI.

Conclusion: The intravenous transplantation of autologous MSC is safe without any adverse effect. Efficacy should be proved at the end of the study.

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Abstract – WCN 2013

No: 610

Topic: 6 – MS & Demyelinating Diseases

Motor sequence learning in relapsing remitting MS patients

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Introduction: Motor learning is sub-served by two interrelated systems (explicit and implicit). However, attempts to guide learning commonly are done using explicit instructions concerning “how to” perform a movement task. We investigated the impact of explicit information (EI) on motor-sequence learning in MS patients.

Material and methods: 30 RRMS (24 female and 6 male; mean age: 29.5 ± 5.6 ; mean duration disease: 64.43 ± 48.38 ; mean EDSS: 1.5 ± 0.4) and 30 healthy subjects were randomly divided in 2 explicit and implicit subgroups.

Explicit subgroup was provided with EI and another subgroup was not. All subjects performed serial reaction time task (SRT). In this task four squares with different colors appeared on the computer screen and subjects were asked to press 1 of 4 keys corresponding to the appropriately colored square immediately after observing it with a retention test after 48 h.

Results: Patients performed this task significantly slower than healthy groups (mean RT in MS patients: 1356.57 ± 348.89 s and in the healthy subjects: 1018.60 ± 210.49 s, $P < 0.05$). Time of final blocks decreased significantly in both groups ($P < 0.05$). Motor learning in healthy subjects was higher and the implicit subgroup more than explicit in the patient group ($P < 0.05$).

Conclusion: Although patient groups performed this motor task slower than the healthy subjects, there was motor learning in the patient in both subgroups (implicit and explicit). In the patients implicit subgroups indicate better motor learning rather than explicit subgroups. Explicit information was detrimental for motor-sequence learning in RRMS patients.

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Abstract – WCN 2013

No: 606

Topic: 6 – MS & Demyelinating Diseases

Does exercise improve cognitive functions in multiple sclerosis? Report of a pilot study

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Introduction: Multiple sclerosis has a significant effect on cognition and its dysfunction is evident in about 40 to 60% of the patients. Various interventions including medications had no significant effect in improving this problem, so finding the new more efficient and available methods to overcome this problem may help the patients and enhance the quality of their lives. One of these methods could be exercise therapy, so we designed a pilot study in MS patients to evaluate the effect of it on cognitive functions.

Method and material: 17 (3 male and 14 female) definite MS patients with mean disease duration of 3 y and mean EDSS of 2.35, enrolled and started the selected designed physical exercises, assessed as a before and after semi-experimental intervention. All of them did balance and aerobic exercises 3 times per week for 8 weeks. Before and after of aforementioned exercises, screening cognition test (Rao et al.) in 6 domains was obtained.

Results: The patients did 22.5 sessions of exercises. Changes in all part of Selective reminding test (SRT), Symbol digit modalities test (SDMT) and one part of Paced auditory serial addition test (PASAT) were significant ($p < 0.05$). The result of (10/36) Spatial recall test, word generation list test and second part of PASAT were not significant.

Conclusion: The results showed that physical exercises could make significant changes in different aspects of cognitive tests in the patients with MS. However, more investigation and RCTs are needed.

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Abstract – WCN 2013

No: 608

Topic: 6 – MS & Demyelinating Diseases

The relationship between fatigue and the severity of disability, in multiple sclerosis

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Introduction: Fatigue in multiple sclerosis is a very common symptom, can be the early manifestation and encompasses physical, social and cognitive inefficiencies. There are different conflicting results in relation to fatigue and the level of disability in MS. The aim of this study is to demonstrate the relationship between fatigue and the severity of disability in patients with multiple sclerosis.

Methods: In this cross sectional study, 140 patients with multiple sclerosis (114 F/26 M, age: mean \pm standard deviation = $32/77 \pm 8/22$) were assessed in three subtypes (relapsing–remitting, primary progressive and secondary progressive). The fatigue and the severity of fatigue were assessed and validated by Persian version of the Fatigue Impact Scale (PER-FIS) and fatigue severity scale (FSS) respectively. The severity of disability was measured by the Expanded Disability Status Scale (EDSS). The PER-FIS consists of three subscales (cognitive, physical, social role) and allows a multidimensional evaluation. Data was analyzed by Spearman's rho rank correlation coefficient.

Results: The EDSS was correlated with total scores of PER-FIS ($r = 0/36$ $p < 0.0001$) as well as with the cognitive ($r = 0/17$ $p < 0.05$), physical ($r = 0/42$ $p < 0.0001$) and social ($r = 0/36$ $p < 0.0001$) subscales. Furthermore EDSS among three subgroups of participants has positive relationship with FSS ($r = 0/32$ $p < 0.0001$). However, neither total nor the subscales score of PER-FIS were not linked to EDSS.

Conclusion: These data confirmed that the degree of the neurological disability in multiple sclerosis is correlated with the degree of fatigue. But, in the different subtypes, the fatigue remains independent from EDSS.

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Abstract – WCN 2013

No: 556

Topic: 6 – MS & Demyelinating Diseases

The course of washout period at JCV seropositive RRMS patients after natalizumab withdrawal and planned switch to fingolimod

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Background: Natalizumab withdrawal at JCV seropositive RRMS patients may cause disease reactivation/rebound. The switch to fingolimod, as one of the therapy options, is preceded by the washout, for its duration there has not existed an unequivocal consensus so far.

Objective: To assess the course of the washout at JCV seropositive RRMS patients after natalizumab with planned switch to fingolimod.

Patients and methods: We realized such a switch in 9 patients. All patients taking natalizumab have failed first-line therapies, in average of 4.2 years, number of relapses the year before natalizumab at least 2. The patients had no relapses under natalizumab and no new lesions in the brain MRI performed every 6 months. Natalizumab was discontinued at the patients' request after the evidence of JCV-antibody at 5 cases (examined for the first time) average number of infusions 17. Seroconversion was recorded in 4 patients by an average of 33 infusions (23–48).

Results: The course of washout (average duration of 3.3 months) was different in individuals. An uncomplicated course of a 3-month washout was recorded in 2 ill. Between the 2nd and 3rd month of washout MRI reactivation of MS was observed in 1 patient, 5 patients had relapses, 1 rebound, with actually EDSS 7.0. Except this patient, the others continue the fingolimod treatment with stable clinical course.

Conclusion: Available news from the literature draw attention to shortening the washout despite the increased risks of PML. Practical experience in our small group of the ill supports these tendencies.

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Abstract – WCN 2013

No: 447

Topic: 6 – MS & Demyelinating Diseases

New oral treatment designed for clinical stabilization in multiple sclerosis

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It is mandatory to have a safe and effective treatment to stop the Multiple Sclerosis' progression. Previous clinical trials designed to stop MS progression have failed to demonstrate the ideal clinical stabilization and most of them have potential severe adverse effects. We administrated to MS patients a new oral treatment to stop its progression. This treatment called Cervô, contains four substances that have effect in controlling the most important known mechanisms of disease progression as: aberrant apoptosis, oxidative damage, mitochondrial degeneration, caspase activation and Mitogen-Activated Protein-Kinase (MAPK) activation, among others. We had previously demonstrated that it is safe to use Cervô in humans. We gave Cervô as monotherapy during the follow-up period to all the patients.

Results: We included 23 patients with MS. Age: 20 to 60 years old (mean 39.6 years, SD \pm 12.81), 16 female (69.6%), 7 male (30.4%). Basal EDSS score: 0–8 (mean 3.0). Follow-up period: 3–67 months (mean 30.35, SD \pm 21.75). No adverse effects were observed.

Clinical evaluation: There was no deterioration in EDSS score in 21 patients (91.30%), and 9 of them (39.13%) improved their basal UPDRS score. The mean EDSS score at 12 months was 2.62 (SD \pm 2.76), at 24 months 2.61 (SD \pm 2.77), and at 46 months 2.71 (SD \pm 2.92). Only 2 patients (8.69%) had worse UPDRS score.

Conclusions: Cervô is a new promising medication that according with this study may stop MS progression in most patients. This needs to be confirmed with a randomized multi-centric study.

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Abstract – WCN 2013

No: 455

Topic: 6 – MS & Demyelinating Diseases

Topical application of brain derived neurotrophic factor attenuates spinal cord trauma induced edema and myelin damage

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Influence of brain derived neurotrophic factor on spinal cord trauma induced edema formation and myelin damage was examined using immunohistochemical and ultrastructural investigations. A focal trauma to the rat spinal cord (right dorsal horn lesion, T10–11) induces degradation of myelin basic protein (MBP) immunoreactivity

in both spinal cord gray and white matter at 5 h. This decrease in MBP immunoreactivity was most pronounced in the vicinity of the injured spinal cord. A general expansion of the cord and swelling of gray and white matter is clearly evident. A significant increase in spinal cord water content occurs in the traumatized as well as in the adjacent rostral (T9) and caudal (T12) spinal cord segments. Ultrastructural studies show profound myelin vesiculation, membrane damage and edema in the spinal cord. Pretreatment with BDNF (1 μ g/ml solution) for 30 min (n = 5), 0 min (n = 5) and 2 min (n = 5) after trauma significantly reduced the MBP degradation, edema and myelin vesiculation. However, similar treatment with BDNF for 10 min (n = 5) and 30 min (n = 5) after injury did not induce a significant protection of myelin following spinal cord trauma. The most pronounced effect of BDNF induced neuroprotection was found when the growth factor was applied 30 min before trauma. These observations suggest that BDNF is neuroprotective in spinal cord trauma, if given before injury, indicating a potential role of the growth factor in treatment of spinal cord injury in near future.

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Abstract – WCN 2013

No: 546

Topic: 6 – MS & Demyelinating Diseases

Correlation of multiple sclerosis incidence trends with solar and geomagnetic indices: New roadmap for solving the puzzle of multiple sclerosis

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Background: Recently, we introduced solar related geomagnetic disturbance (GMD) as a potential environmental risk factor for multiple sclerosis (MS). We showed that it can describe MS prevalence distribution better than other geophysical factors and can explain special MS features such as birth month effect, relapsing–remitting nature and migration effect.

Objective: To test probable correlation between solar activities and GMDs with long-term variations of MS incidence.

Materials and methods: We studied the association between alterations in solar wind velocity (V_{SW}) and Planetary A index (A_p , a GMD index) with previously published data of MS incidence trend in Tehran by Elhami et al. and western Greece by Papanthanasopoulos et al., during the 23rd solar cycle (1996–2008), through an ecological–correlational study. Cross-correlation analyses were used for finding possible lead–lag relationships.

Results: We found strong correlations among MS incidence of Tehran with V_{SW} ($r_s = 0.665$, $p = 0.013$) with one year delay, and with A_p ($r_s = 0.864$, $p = 0.001$) with 2 year delay. There were very strong correlations among MS incidence data of Greece with V_{SW} ($r = 0.906$, $p < 0.001$) and with A_p ($r = 0.844$, $p = 0.001$) both with one year delay.

Conclusion: For the first time in the history of MS, a hypothesis has introduced an environmental factor that can describe MS incidence variations. Important message of these findings for researchers is to provide MS incidence reports with higher resolution, based on the time of disease onset, not the time of diagnosis. Then, it would be possible to conduct superposed epoch analyses to better investigate the validity of GMD hypothesis.

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Abstract – WCN 2013**No: 545****Topic: 6 – MS & Demyelinating Diseases****Geomagnetic disturbances may be environmental risk factor for multiple sclerosis: An ecological study of 111 locations in 24 countries**

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Background: We noticed that a hypothesis based on the biological effects of geomagnetic disturbances (GMD) has the ability to explain special features of multiple sclerosis (MS) such as the cause of its relapsing–remitting nature, the birth month effect and migration effect.

Objective: As areas around geomagnetic 60 degree latitude (GM60L) experience the greatest amount of GMD, the easiest way to evaluate our hypothesis was to test the association of MS prevalence (MSP) with “angular distance to geomagnetic 60 degree latitude” (AMAG60) and compare it with the known association of MS with geographical latitude (GL). We did the same with “angular distance to geographic 60 degree latitude” (AGRAPH60) as a control.

Material and methods: 111 MSP data from 1980–2010 published papers were retrieved and entered in the study. By meta-regression analyses, relationship of MSP with GL, AMAG60 and AGRAPH60 was evaluated separately. Models were compared by their adjusted R square (AR²) and standard error of estimate (SEE).

Results: In each continent, AMAG60 had the best correlation with MSP, the largest AR² and the least SEE. Merging both hemisphere data, AMAG60 explained 56% of MSP variations with the least SEE (R = 0.75, AR² = 0.56, SEE = 57), while geographic latitude explained 17% (R = 0.41, AR² = 0.17, SEE = 78.5) and AGRAPH60 explained 12% of those variations with the highest SEE (R = 0.35, AR² = 0.12, SEE = 80.5).

Conclusions: Results confirmed that AMAG60 is the best describer of MSP variations and has the strongest association with MSP distribution. These results supported the fact that GMD may be an environmental risk factor for MS.

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Abstract – WCN 2013**No: 285****Topic: 6 – MS & Demyelinating Diseases****The month of birth and the incidence of multiple sclerosis in Southern Iran**

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Introduction: Recent studies on seasonal time of birth of multiple sclerosis patients showed a spring peak and an autumn nadir. In this study we examined month of birth (MOB) and season of birth (SOB) and risk of multiple sclerosis in later life in Iranian population.

Patients and methods: During a case control study, in Fars, Southern Iran, between 2005 to 2012, more than 1500 patients with definite multiple sclerosis (according to Mc Donald's Criteria) in outpatient clinic were involved in the study. We randomly selected 2000 individuals from Fars province populations who were matched with case group in aspects of age and sex. The dates of birth and gender were identified for each patient and control. The results were compared in the groups.

Results: 1020 females and 538 males, age between 15 and 65 years were involved in the study. Female to male ratio in this study was 1.9.

Overall we detected that rate of birth among cases was significantly higher in March, April and October than controls. Moreover it seems that this rate is significantly lower in January in case group. We didn't detect any significant differences among cases and controls in time of birth in different seasons of the year.

Discussion: Such as European countries we detected that rate of birth among cases was significantly higher in March, April and October than controls. Possible explanation is decreased exposure to sun in the winter which leading to low vitamin D levels during pregnancy.

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Abstract – WCN 2013**No: 287****Topic: 6 – MS & Demyelinating Diseases****The effect of free testosterone on course, severity, disease activity and disability in the patients with multiple sclerosis**

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Introduction: Sex related difference in the course and severity of Multiple Sclerosis (MS) could be mediated by several sex hormones. This study aimed to investigate the relation between free testosterone concentrations and course, severity, brain damage and disability in Iranian patients with MS.

Methods: 37 women with MS and 25 healthy subjects were included in the study as case and control groups.

Free testosterone level was assessed by ELISA method. Brain MRI with and without contrast was performed. Expanded Disability status scale (EDSS) and MS subtypes in the patients were collected via a questionnaire.

Results: Serum testosterone was significantly lower in women with MS than controls (P value: 0.026). The free testosterone levels were not associated with EDSS, MS subtype and MRI findings.

Discussion: The hormone related modulation of pathological changes does not support the hypothesis that sex hormones play a role in the inflammation, damage and repair mechanism in MS, although serum level of these hormones in the patients was significantly lower than controls.

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Abstract – WCN 2013**No: 288****Topic: 6 – MS & Demyelinating Diseases****Significant increase in the prevalence of MS in Iran**

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Introduction: Iran was previously considered to have a low prevalence rate of MS patients, but this rate has significantly changed over time.

Methods: This study was performed on the bases of the new reports of Iran Ministry of Health and Medical education about the patients who registered to receive beta interferon in Iran. Although these data doesn't show the exact number of the patients with MS in Iran but these information may be considered as the least prevalence rate of these patients in each province.

Results: To the end of 2011, there were 34,605 MS patients who were registered by ministry of health in Iran. With consideration of Iran population in this year prevalence rate is 30.75 per 100,000

populations. 77% of the patients are females. Maximum prevalence rate was seen in Isfahan province (80 per 100,000) at the central parts of Iran and the minimum prevalence rate is seen in Sistan–Baluchestan province (6 per 100,000) which is located in southwest of Iran with a warm and dry climate.

Discussion: According to our new information and regarding to the previous studies it seems that the prevalence of MS in Iran and of its provinces is similar to what is seen in western countries.

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Abstract – WCN 2013

No: 316

Topic: 6 – MS & Demyelinating Diseases

Zebrafish as a tool for molecular diagnosis of Alexander disease

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Background: Alexander disease (AxD) is an inherited demyelinating disease that primarily inflicts upon the white matter of the central nervous system (CNS), and is caused by mutation in a gene encoding glial fibrillary acidic protein (GFAP). The pathognomonic feature of AxD is eosinophilic Rosenthal fibers (RFs) in astrocytes.

Objective: To develop a new method to determine if identified mutations found in a GFAP gene are disease-causing, because of the difficulty in procuring the CNS tissues from patients.

Patients and methods: We found a family with slowly progressive gait disturbance and dysarthria. MR images of the brain showed a marked atrophy of the medulla oblongata and upper cervical cord. Under the provisional diagnosis of adult-onset AxD, DNA sequence analysis of GFAP gene revealed D128N mutation with autosomal dominant transmission. There was no previous report on the formation of RFs in GFAP (D128N) mutant. Therefore, various human GFAP mutants including previously well-known R79C, R79H, R239C or R239H by site-directed mutagenesis were generated and their expression levels in HEK-293T cells by Western blotting were compared. GFAP (D128N) DNA was microinjected into zebrafish embryos.

Results: Expression levels of mutant GFAP proteins are comparable to that of wild-type GFAP protein. However, mutant GFAPs including induced more aggregations than wild-type GFAP in zebrafish embryos. Electron microscopy displayed dense osmophilic inclusions in the cells of zebrafish embryos.

Conclusions: This finding indicates that D128N is the disease-causing mutation that causes AxD. Therefore, zebrafish can be used as a tool for molecular diagnosis of AxD.

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Abstract – WCN 2013

No: 354

Topic: 6 – MS & Demyelinating Diseases

Relapsing Devic disease treated with rituximab (case discussion)

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A 33 year old Libyan lady presented with left eye blurred vision 13 months ago which was remitted within 1 week after oral steroid

usage, 3 months later she got bilateral lower limb numbness & paraesthesia followed by weakness, urinary retention & unsteadiness & those symptoms were cured partially after usage of IV steroids, then 2 months later she got severe neuropathic pain & allodynia all over the body with bilateral lower limb numbness which was cured partially after usage of IV steroids & oral pregabalin, clinically she has ataxic paraparesis with brisk reflexes all over the body & bilateral lower limb hypesthesia and allodynia up to the level of D4, with positive Lhermitte sign.

Investigations: MRI cervicodorsal spine showed single long continuous enhancing cord lesion extending from D4 to D12 (which is going with Devic disease) with another 2 small ovoid cervical spine lesions which are going with Multiple Sclerosis.

MRI brain showed 5 white matter demyelinating lesions in the brain 4 of them disappeared after steroids.

CSF Aquaporin 4 Antibodies were negative as well as the CSF oligoclonal bands, with normal CSF IgG index. VEP showed delayed p100 response on the left eye.

BSEP was normal.

Treatment: She has been treated with full course of IV rituximab with excellent improvement clinically & radiologically.

Conclusion: The patient has relapsing remitting neurologic disease with changing MRI brain lesions & continuous enhancing dorsal spine lesion extending from D4 to D12 which is suggestive of relapsing Devic disease in spite of negative CSF Aquaporin 4 Antibodies.

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Abstract – WCN 2013

No: 372

Topic: 6 – MS & Demyelinating Diseases

Acute compressive myelopathy mimicking transverse myelitis

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Background: Acute compressive myelopathy (ACM) is occasionally associated with fusiform T2 signal changes on MRI. When this occurs, the radiologic appearance can result in misdiagnosis as transverse myelitis, either idiopathic or as a NMO spectrum disorder. This misdiagnosis can delay appropriate surgical referral, and may result in inappropriate treatments.

Objectives: To reveal clinical and radiologic findings that facilitate the recognition of surgically treatable ACM mimicking transverse myelitis.

Methods: Consecutive patients with suspected longitudinally extensive transverse myelitis (LETM) were enrolled during a 5-year period. Among these LETM patients, we ultimately identified ACM by determining that lesions were caused by compressive myelopathy due to cervical spondylotic lesion. We reviewed all clinical and radiologic data of the ACM patients.

Results: We identified 8 patients with ACM from a total of 26 suspected LETM patients. The mean age of symptom onset was 55.6 ± 9.4 years in the ACM. 7 of ACM patients (87.5%) were men ($P = 0.09$). All 8 ACMs were treated with intravenous corticosteroids. 1 of ACM patients experienced improvement ($P = 0.001$). No ACM patient had recurrence. ACM showed limited lateral or posterior patterns ($P = 0.014$), more upper cervical cord involvement ($P = 0.004$), and multiple lesions ($P = 0.014$) than those of LETM. Focal contrast enhancement was identified in all ACM patients, and occurred at the point of maximal focal stenosis. These characteristic enhancement findings were not observed in LETM group.

Conclusion: The above characteristic clinical and radiologic findings may distinguish ACM from inflammatory myelopathies such as LETM and have therapeutic implications.

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Abstract — WCN 2013

No: 391

Topic: 6 — MS & Demyelinating Diseases

Antiretroviral treatment in patient with secondary progressive multiple sclerosis — Impact on disease progression and safety

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A 58-year old white female with multiple sclerosis was treated with antiretroviral therapy (ART) of raltegravir and lamivudine. Over two years prior to the commencement of ART the patient reported progressive worsening of weakness in right arm and legs requiring use of a walking stick, burning sensations in limbs and persistent fatigue. MRI showed areas of demyelination in the spine but without involvement of the brain. CSF analysis showed oligoclonal IgG bands. The patient was treated with interferon beta for seven years but it was stopped due to disease progression. No other disease modifying therapy had been used.

Methods: Neurological examination, EDSS, Multiple Sclerosis Functional Composite (MSFC), Multiple Sclerosis Quality of Life Inventory (MSQLI) and blood safety parameters were performed at baseline then monthly over six months of treatment. Z-scores for MSFC and MSQLI were calculated and adjusted to reference population.

Results: There was no significant change from baseline in the EDSS, MSFC and MSQLI scores. There were no side effects of ART reported by patient or detected by blood safety parameters. The patient reported marked improvement in fatigue during the first two months of treatment.

Conclusions: ART in this patient was safe and may have stabilising impact on disease progression and quality of life. A possible mode of action of ART in MS includes suppressive effect on MS-associated Human Endogenous Retrovirus and herpes viruses triggering MS. This is the first case of patient with MS receiving antiretroviral therapy with formal evaluation of disease progression and safety.

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Abstract — WCN 2013

No: 411

Topic: 6 — MS & Demyelinating Diseases

Polarization of macrophages and T cells to anti-inflammation in EAE mice treated with Fasudil

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Background: Macrophages play paradoxical roles in the pathogenesis of neurological disorders, exerting both inflammatory M1 and anti-inflammatory M2 phenotypes. However, besides macrophages,

the dynamic balance between effector Th1/Th17 and Treg cells also regulates the development and improvement of diseases.

Objective: To observe the therapeutic effect of Fasudil on EAE, and explore its mechanisms.

Material and methods: Mouse EAE was induced by MOG_{35–55} immunization and passive transfer of primed lymphocytes treated with Fasudil in vitro.

Results: Fasudil significantly ameliorated the clinical severity of EAE, and decreased the expression of CD4 and CD68 in spinal cords, accompanied by an improvement of demyelination and inhibition of inflammatory cells in both active and passive EAE models. Fasudil shifted inflammatory M1 to anti-inflammatory M2, being shown by inhibiting the expression of F_{4/80}CD_{16/32}, F_{4/80}MHC-II, and F_{4/80}P₂X₇ and the production of NO as well as increasing the expression of F_{4/80}CD₂₀₆ and F_{4/80}IL-10 and activity of Arg-1. Simultaneously, Fasudil significantly downregulated CD4⁺IFN- γ ⁺ and CD4⁺IL-17⁺ and upregulated CD4⁺IL-10⁺ in Fasudil-treated T cells which were delivered into animals via adoptive transfer. The polarization of M2 macrophages may be associated with the decrease of inflammatory cytokines IL-6 and TNF- α and the increase of anti-inflammatory cytokine IL-10.

Conclusion: Using a passive transfer model, we find that MOG-primed cells treated with Fasudil in vitro did not trigger EAE generation by controlling the specific polarization of macrophages and T cells, which may represent a therapeutic opportunity for treatment of MS. This study was supported by National Natural Science Foundation of China (81272163).

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Abstract — WCN 2013

No: 434

Topic: 6 — MS & Demyelinating Diseases

Perception of muscular effort in multiple sclerosis

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Background: Resistance exercise is effective in improving muscle strength and preventing muscle weakness in multiple sclerosis (MS) patients. Control of resistance training intensity based on perceived muscular effort is applicable to healthy individuals, yet there is no evidence of its utility for MS patients.

Objective: To compare perception of muscular effort in MS patients to healthy controls.

Material and methods: Based on their perception of muscular effort, twenty-five MS patients and twenty-eight controls adjusted static elbow extension tasks according to five levels on the OMNI-Resistance Exercise Scale. Elbow extension strength and muscle activity were measured via load cell dynamometer and surface electromyography (EMG) and related to each participant's maximal voluntary contraction (MVC) strength and muscle activity. Two-way analysis of variance was used to evaluate statistical significance.

Results: There were no statistically significant differences between MS patients and healthy controls, they produced similar relative torque values ($F_1 = 0.196$; $p > 0.05$) and extensor muscle activities ($F_{2,617} = 1.556$; $p > 0.05$) across all effort levels.

Conclusion: No differences were found in the perception of muscular effort in MS patients and the age-matched control group. Future studies should explore, whether rating of perceived exertion is an effective instrument to control resistance training intensity in MS patients.

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Abstract – WCN 2013**No: 56****Topic: 6 – MS & Demyelinating Diseases****Postural orthostatic tachycardia syndrome associated with multiple sclerosis**

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Background: Postural orthostatic tachycardia syndrome (POTS) is an autonomic disorder characterized by an exaggerated increase in heart rate that occurs during standing, without orthostatic hypotension.

Objective: The aim of this study was to determine if there is a difference in the frequency of postural orthostatic tachycardia syndrome (POTS) in patients with multiple sclerosis (MS) compared to patients with symptoms of orthostatic intolerance and with no evidence of MS or other neurological illness.

Methods: We analyzed data gathered from 293 (112 with MS and 181 without MS) patients who underwent the head-up tilt table test protocol. If POTS was identified the head-up tilt table test was repeated and supine and standing serum epinephrine and norepinephrine were determined.

Results: POTS was identified in 39 patients: 21 (19%) in the MS group compared to 18 (10%) in the non-MS group ($p = 0.035$). There was no difference between groups in the occurrence of POTS associated syncope ($p = 0.52$). There was no difference between groups in the epinephrine or norepinephrine in supine and standing positions. While both standing epinephrine and norepinephrine levels were significantly higher compared to laying levels in the non-MS group, only standing norepinephrine levels were significantly higher in the MS group.

Conclusions: The results of this study suggest that POTS is associated with MS.

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Abstract – WCN 2013**No: 66****Topic: 6 – MS & Demyelinating Diseases****Pathogenetic role of T helper 17 cells and cytokines in Guillain-Barré syndrome**

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Background: The role of T helper (Th)17 cells, a newly found Th cell subset, has been scarcely studied in Guillain-Barré syndrome (GBS). Emerging evidence points to a pathogenetic role of Th17 cells in GBS.

Objective: To further clarify the roles of these Th 17 cells and cytokines in the pathogenesis of EAN.

Materials and methods: The EAN model was induced by immunizing animals with P0 protein peptide 180–199 together with complete Freund's adjuvant and pertussis toxin. The dynamic expression of Th1/Th2/Th17/Treg cells and cytokines in GBS and EAN was studied.

Results: The clinical signs of IFN- γ KO mice were significantly more severe than those of WT controls, along with higher proliferation of splenic mononuclear cells in KO mice. At the peak of EAN, the proportion of IL-17A-expressing cells in cauda equina (CE) infiltrating cells, and the levels of IL-17A in sera were elevated in IFN- γ KO mice. We further investigated Th17 cells and IL-17A in GBS patients. Circulating Th1, Th17 and Th22 cells were significantly increased in GBS patients during the acute phase. Moreover, CSF and plasma levels of IL-17A and IL-22 were also elevated. The levels of Th17 cells

and IL-17 from CE infiltrating cells and splenic mononuclear cells as well as in serum paralleled the disease evolution. Compound 1, a synthetic compound that can specifically block the production of Th17 cells, suppressed the severity of EAN.

Conclusion: Th17 cells are a neuritogenic T helper cell subset in GBS and EAN.

doi:10.1016/j.jns.2013.07.1514

Abstract – WCN 2013**No: 257****Topic: 6 – MS & Demyelinating Diseases****Auditory evoked potentials and vestibular evoked myogenic potentials in evaluation of brainstem lesions in multiple sclerosis**

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Objective: The aim of this study was to determine the role of magnetic resonance imaging (MRI), auditory evoked potentials (AEP) and vestibular evoked myogenic potentials (VEMP) in the evaluation of brainstem involvement in multiple sclerosis (MS).

Patients and methods: Altogether 32 patients with the diagnosis of MS participated in the study. The following data was collected from all patients: age, gender, Expanded Disability Status Scale (EDSS) score, brainstem functional system score (BSFS) (part of the EDSS evaluating brainstem symptomatology), and involvement of the brainstem on the brain MRI. AEP and ocular VEMP (oVEMP) and cervical VEMP (cVEMP) were studied in all patients.

Results: BSFS, MRI, AEP, oVEMP and cVEMP involvement of the brainstem was evident in 9 (28.1%), 14 (43.8%), 7 (21.9%), 12 (37.5%) and 10 (31.0%) patients, respectively. None of the test used showed statistically significant advantage in the detection of brainstem lesions. When combining oVEMP and cVEMP 18 (56.3%) patients showed brainstem involvement. This combination showed brainstem involvement in greater percentage than BSFS or AEP, with statistical significance ($p = 0.035$ and $p = 0.007$, respectively).

Conclusion: VEMP is a reliable method in detection of brainstem involvement in MS. It is comparable with MRI, but superior to clinical examination or AEP.

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Abstract – WCN 2013**No: 166****Topic: 6 – MS & Demyelinating Diseases****Knowledge and attitude assessment of Iranian patients with MS receiving interferon beta**

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Background: There are few studies on patient knowledge in multiple sclerosis (MS), and there are only three published questionnaires. The purpose of this study was to describe MS patients' understanding of their treatments. Consequently, to provide guidance for developing educational programs for MS patients.

Method: A total of 425 randomly selected MS patients participated in this survey (response rate 85%). The knowledge levels of correctly using IFN beta and their attitude towards some aspects of medical care were measured using self-reported questionnaires which consisted of 25 items with validity of multidisciplinary panel and

pre-testing on 20 patients. The data were analyzed using descriptive statistical methods.

Results: Acceptably, knowledge and attitude internal consistency (Cronbach's alpha = 0.73, 0.71) and content validity were established. MS patients' knowledge of treatment with IFN beta was very low, however their attitudes were at a very high level. Female patients, self-injection ability, the higher educational level, normal functional status, longitudinal delay from the start of diagnostic workup to definite diagnosis, and being younger were related to higher level of

knowledge. Attitude was associated with functional status, family history of disease and the summary of knowledge variable (positive association).

Conclusion: Low levels of patients' knowledge disclose their needs for developing educational interventions, with considering key role of some other important factors on patients' attitude.

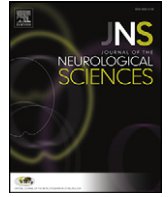
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Topic: 7 - Neuromuscular disorders

Abstract - WCN 2013**No: 191****Topic: 7 - Neuromuscular disorders****Cerebral hemodynamic changes in head-up tilt test in patients with orthostatic hypotension, postural orthostatic tachycardia syndrome and other orthostatic intolerance**

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Orthostatic hypotension (OH) and paroxysmal orthostatic tachycardia syndrome (POTS) are the common causes of orthostatic intolerance (OI). However the hemodynamic changes of cerebral flow in head-up tilt (HUT) test were not well known. The purpose of this study is to compare the cerebral hemodynamics in HUT test among patients with OH, POTS and other OI without OH and POTS (OOI). Two hundred and forty-three patients, who complained of orthostatic intolerance, were enrolled in this study. All patients performed the HUT test. Heart rate, blood pressure, and blood flow velocity (BFV) of the middle cerebral artery of the dominant hemisphere were obtained during supine position, just after and every one minute for 10 minutes after 80° HUT. Thirty-eight patients were included in group with OH, 28 patients group with POTS, and the other 117 patients group with OOI. We calculated the critical closing pressure (Pcrit). We also calculated the cerebral perfusion pressure (CPP) and cerebral vascular resistance (CVR). The changes of mean arterial pressure (MAP), BFV, Pcrit, CPP, and CVR between baseline and HUT were compared. The BFV were significantly decreased after HUT in all three groups, however the changes of BFV were not significantly different among three groups. The CPP and CVR were significantly decreased during head-up tilt in OH.

The BFV was decreased after HUT in all three OI groups. Although the MAP and CPP were decreased during HUT in OH, the CVR was also decreased, which showed the operation of cerebral autoregulation.

doi:[10.1016/j.jns.2013.07.1518](https://doi.org/10.1016/j.jns.2013.07.1518)**Abstract - WCN 2013****No: 142****Topic: 7 - Neuromuscular disorders****Tumor necrosis factor- α (TNF- α) is increased in the skin of patients with amyotrophic lateral sclerosis**

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Background and objective: Tumor necrosis factor- α (TNF- α) is a major inflammatory cytokine that elicits a wide range of biological responses and is implicated in the pathogenesis of neurodegenerative diseases. Skin studies from patients with amyotrophic lateral sclerosis

(ALS) have shown unique pathological and biochemical abnormalities. The lack of bedsores formation is considered characteristic.

Patients and methods: We undertook a quantitative immunohistochemical study of TNF- α in the skin from patients with ALS and controls.

Results: Immunohistochemistry for TNF- α demonstrated cytoplasmic activity in the epidermis and in some blood vessels and glands. The proportion of TNF- α -positive (TNF- α +) cells in the epidermis in ALS patients was significantly higher ($p < 0.001$) than in controls. There was a significant positive relationship ($r = 0.87$, $p < 0.001$) between the proportion and duration of illness in ALS patients, but there was no such relationship in control subjects. The optical density of TNF- α + cells in the epidermis in ALS patients was markedly stronger ($p < 0.001$) than in controls. There was a significant positive relation ($r = 0.70$, $p < 0.001$) between the immunoreactivity and duration of illness in ALS patients. In addition, there was an appreciable positive correlation ($r = 0.59$, $p < 0.01$) in ALS patients between the proportion of TNF- α + cells and the optical density of these cells.

Conclusion: These data suggest that changes in TNF- α identified in skin of ALS patients are likely to be related to the disease process and that metabolic alterations of TNF- α may take place in the skin of patients with ALS.

doi:[10.1016/j.jns.2013.07.1519](https://doi.org/10.1016/j.jns.2013.07.1519)**Abstract - WCN 2013****No: 259****Topic: 7 - Neuromuscular disorders****Sampling of the myelinated fibers for morphometric studies: How much is enough?**

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Background: There is no documented consensus if there is a sampling method that reproduces with reliability the complete evaluation of the endoneurial space of a determined nerve.

Objective: To investigate the viability of myelinated fiber sampling in normal nerves and on aging.

Methods: Female Wistar rats, aged 3, 6, 12, 18 and 24 months ($N = 6$ per group) had their right sural nerves prepared for epoxy resin embedding and light microscopy analysis. Myelinated fiber morphometry was performed with the KS-400 software and the sampling method consisted in the morphometric analysis of 30%, 50% and 70% of the microscopic frames generated from the fascicle and automatically selected by the software.

Results: Myelinated fibers and respective axons increased in size with aging, reaching the highest values at age 360 days. Assessment of 70% of the images showed no difference in average values or the histograms

distributions when compared with 100%, in all ages. Assessment of 30% of the images showed no differences on the average values for all parameters investigated but a difficult interpretation of the histogram shape in young (90 days-old) and old (640 and 720 days-old) animals was detected. Nevertheless, the 30% investigation of the myelinated fibers histograms was reliable in the adult animals (180 and 360 days-old).

Conclusions: The 30% sampling method is a reliable tool to assess the sural nerve myelinated fiber size and distributions in adult rats, reducing the morphometry time, effort and amount of data evaluated.

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Abstract - WCN 2013

No: 228

Topic: 7 - Neuromuscular disorders

Age and gender influence on motor unit number in healthy subjects estimated by multipoint incremental MUNE method

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Motor unit number estimation (MUNE) is a tool for approximation of the real number of motor units.

Objective: To evaluate the multipoint incremental MUNE method in healthy population, to analyze if aging/sex/side of dominant hand have influence on motor unit number and to assess reproducibility of MUNE by Shefner's modification.

Material and methods: 60 volunteers (mean age 47 yrs \pm 17.7) in 4 subgroups in the ranges 18–30; 31–45; 46–60; over 60 yrs. MUNE was calculated in abductor pollicis brevis (APB) and abductor digiti minimi (ADM) by dividing single motor unit action potential SMUP into supramaximal compound motor action potential (CMAP) amplitude.

Results: Test–retest variability was 7%. The mean value of MUNE for APB was 133.2 ± 43 ; for ADM 157.1 ± 39.4 . The significant differences between MUNE results were found between subgroups 18–30 and over 60 yrs and between subgroups 31–45 and over 60 yrs. MUNE results correlated with age of controls for both APB and ADM. SMUP, reflecting the size of motor unit, increased with age of controls only in APB. CMAP amplitude correlated inversely with age of controls in APB and in ADM. The correlation between MUNE/ CMAP amplitude of APB/ADM muscles and the age of females but not of men was found.

Conclusions: Multipoint incremental MUNE method in Shefner's modification is a non-invasive, easy to perform method with high reproducibility. The loss of motor neurons due to aging, appears to be more pronounced in healthy people over 45 yrs, especially in females, what could be caused by estrogen/androgens level alterations.

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Abstract - WCN 2013

No: 130

Topic: 7 - Neuromuscular disorders

Increased progranulin in the skin of amyotrophic lateral sclerosis: An immunohistochemical study

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Background and objective: It has been demonstrated that progranulin (PGRN) is a neurotrophic factor that enhances neuronal survival and axonal growth. Several lines of evidence have indicated that PGRN plays a role in the pathomechanism of amyotrophic lateral sclerosis (ALS). So far studies of ALS skin have shown unique pathological and biochemical abnormalities. The lack of bedsore formation in ALS patients is considered characteristic. However, there has been no study of PGRN in ALS skin.

Patients and methods: We made a quantitative immunohistochemical study of the expression of PGRN in the skin from 18 patients with sporadic ALS and 13 control subjects.

Results: Immunohistochemistry for PGRN demonstrated cytoplasmic activity in the epidermis and in some blood vessels and glands. Numerous PGRN-positive (PGRN+) cells were observed in the epidermis in ALS patients, which became more marked as ALS progressed. On the other hand, PGRN+ cells of the epidermis, dermal blood vessels and glands in control subjects showed a weak positive reaction. PGRN immunoreactivity of PGRN+ cells was markedly positive in the epidermis in ALS patients. The proportion of PGRN+ cells in the epidermis in ALS patients was significantly higher ($p < 0.001$) than in controls. There was a significant positive relationship ($r = 0.83$, $p < 0.001$) between the proportion and duration of illness in ALS patients.

Conclusion: These data suggest that changes of PGRN in ALS skin are related to the disease process and that metabolic alteration of PGRN may take place in the skin of patients with ALS.

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Abstract - WCN 2013

No: 131

Topic: 7 - Neuromuscular disorders

Decreased amount of collagen in the skin of amyotrophic lateral sclerosis in the Kii Peninsula of Japan

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Background: The Kii Peninsula of Japan, together with Guam and West New Guinea, has one of the highest incidences of amyotrophic lateral sclerosis (Kii ALS) in the world. There is a controversy whether the etiology is the same or not between sporadic ALS and Kii ALS. Skin studies from patients with sporadic ALS have shown unique pathological and biochemical abnormalities. However, there has been no report of collagen content of the skin of Kii ALS patients.

Objective: To measure collagen contents of the skin from Kii ALS patients.

Patients and methods: The skin tissues from 7 Kii ALS patients and 12 controls were studied by electron microscopy and their collagen contents were examined.

Results: On electron microscopy the most conspicuous finding in Kii ALS was the smaller diameter of collagen fibrils. The longer the duration of Kii ALS, the more marked these findings were. The collagen content per dry weight (mg) of the samples in Kii ALS was significantly decreased ($p < 0.001$) than in controls. In Kii ALS patients the more severely affected pathological samples showed the greater decrease. In addition, there was a significant negative correlation ($r = -0.88$, $p < 0.01$) between the collagen content and duration of illness in the Kii ALS patients, but there was no such correlation in controls.

Conclusion: These results indicate that the metabolism of skin collagen might be affected in the disease process of Kii ALS.

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Abstract - WCN 2013

No: 133

Topic: 7 - Neuromuscular disorders

Immunohistochemical studies of angiogenin in the skin of patients with amyotrophic lateral sclerosis

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Background: Angiogenin (ANG) is a member of the ribonuclease superfamily which is implicated in angiogenesis. ANG maintains normal

vasculature and thereby protects motor neurons from various stress conditions. It is suggested that ANG may play a role in the pathomechanism of amyotrophic lateral sclerosis (ALS). However, there have been no studies of ANG in ALS skin.

Objective: To make an immunohistochemical study of ANG in the skin of ALS patients.

Patients and methods: We made a quantitative immunohistochemical study of the expression of ANG in the skin from 20 patients with sporadic ALS, 20 patients with other neurologic or muscular disorders (control group A), and 20 patients without neurologic or muscular disorders (control group B).

Results: The nuclei of the epidermal cells showed a weak ANG immunoreactivity in ALS patients. These findings became more marked as ALS progressed. On the other hand, the nuclei in control groups A and B show a strong positive reaction. The optical density for ANG immunoreactivity of the nucleus in the epidermal cells in ALS patients was significantly lower ($p < 0.001$) than in control groups A and B. There was a significant negative relationship ($r = -0.82$, $p < 0.001$) between the optical density for ANG immunoreactivity of the nucleus and duration of illness in ALS patients.

Conclusion: These data suggest that changes of ANG in ALS skin are related to the disease process and that metabolic alterations of ANG may take place in the skin of ALS patients.

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Abstract - WCN 2013

No: 55

Topic: 7 - Neuromuscular disorders

The role of cervical and ocular vestibular evoked myogenic potentials in the follow-up of vestibular neuronitis

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Background: Vestibular evoked myogenic potentials (VEMP) are used in the assessment of patients with vestibular neuronitis (VN).

Objective: The aim of this study was to evaluate recovery of vestibular nerve function after superior vestibular neuronitis (VN) by vestibular evoked myogenic potentials (VEMP).

Methods: Twenty-six patients with the diagnosis of VN were included in the study. All patients underwent ocular VEMP (oVEMP) and cervical VEMP (cVEMP) recordings, at six days and six months from onset of symptoms.

Results: Of the 26 patients, 14 showed improvement on oVEMP at month 6 (group 1), and 12 showed worsening on oVEMP at month 6 (group 2). At the same time there was no change in the amplitudes of the cVEMP on neither healthy nor affected sides in both groups. Inability to perform Fukuda test and chronic white matter supratentorial lesions present on brain MRI were more frequent in patients with worse outcome on oVEMP ($p = 0.044$ and 0.045 , respectively). While involvement of the inferior branch of the vestibular nerve was not associated with oVEMP outcome, oVEMP latencies (N10 and P13) were associated with improvement or worsening of oVEMP amplitudes, showing that prolonged latencies correlate with 6 month improvement in oVEMP amplitudes (Pearson correlation -0.472 , $p = 0.041$ and Pearson correlation -0.580 , $p = 0.009$, respectively).

Conclusion: This study identified clinical, MRI and neurophysiological predictors of recovery in patients with superior VN and offers additional insight in better understanding the role of VEMP in diagnosis and prognosis of VN patients.

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Abstract - WCN 2013

No: 410

Topic: 7 - Neuromuscular disorders

Ischemic neuropathy in patients with peripheral arterial occlusive disease

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Peripheral arterial occlusive disease (PAD) is not only just a vascular disease but also a neuropathy and a myopathy. The neuropathy and myopathy in PAD may occur as a result of ischemic insult caused by arterial occlusion. We prospectively investigated the neurological and electrophysiological assessment to define the ischemic neuropathy and which factors affect the outcome of neurological and electrophysiological parameters in PAD. A total of thirty-two PAD patients (68.9 and their sixty-four lower limbs were enrolled. Primary outcomes were neurologic symptoms score (NSS) and neurological disability score (NDS) of lower extremities. Secondary outcomes were the data of nerve conduction study (NCS) of lower extremity – sensory nerve action potential (SNAP) amplitude and compound muscle action potential (CMAP) amplitude. The NSS and NDS of 64 lower limbs were 2.43 ± 1.77 and 6.9 ± 7.44 . Age and Fontaine classification were positively correlated and weight, BMI and ABI were inversely correlated with NSS and NDS. Height inversely correlated with NDS. Patients with female sex and distal arteries occlusion below knee showed increased NSS and NDS. Diabetes was associated with increased NDS and hypertension was associated with increased NSS. Age and Fontaine classification were positively correlated and weight and BMI were inversely correlated with SNAP amplitude of sural, superficial peroneal and medial plantar nerves and CMAP amplitude of posterior tibial and common peroneal nerves. Diabetes patients showed more decreased SNAP and CMAP amplitude than non-diabetes patients. Hypertension patients showed decreased CMAP amplitude in only common peroneal nerve. NDS and NCS could be good indicator to assess the ischemic neuropathy in PAD.

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Abstract - WCN 2013

No: 317

Topic: 7 - Neuromuscular disorders

OX40-OX40L expression in idiopathic inflammatory myopathies

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Background: The idiopathic inflammatory myopathies constitute a heterogeneous group of autoimmune diseases of skeletal muscle. OX40, member of the tumor necrosis factor receptor/tumor necrosis factor superfamily costimulatory molecules and its ligand, OX40L, are linked with the development of various autoimmune diseases.

Objective: To examine whether both OX40 and its ligand OX40L are expressed in idiopathic inflammatory myopathies and to investigate the types of inflammatory cells expressing OX40L.

Materials and methods: Immunohistochemistry was performed in limb muscle specimens from dermatomyositis, polymyositis and inclusion body myositis patients to analyze the expression of OX40 and its ligand OX40L. Double immunofluorescence labeling was performed to clarify the phenotype of inflammatory cells expressing OX40L.

Results: OX40 and OX40L expressing cells were observed in all subsets of inflammatory myopathies following a similar pattern of distribution. In polymyositis and inclusion body myositis inflammatory cells expressing the receptors invaded non-necrotic muscle fibers. OX40L expression was also found in endothelial blood cells in all dermatomyositis and some

polymyositis specimens. In all subsets of inflammatory myopathies OX40L was expressed by T cells (CD4+ and CD8+), macrophages (CD68+), B-cells (CD20+) and myeloid dendritic cells (BDCA1+). Plasmacytoid dendritic cells (BDCA2+) expressing OX40L were only found in dermatomyositis and polymyositis.

Conclusions: The simultaneous expression of both OX40 and its ligand OX40L in idiopathic inflammatory myopathies suggest that they might participate in disease pathogenesis. Expression of OX40L by different types of cells within the inflamed muscle implies that OX40–OX40L interaction may contribute in disease mechanisms through different pathways.

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Abstract - WCN 2013

No: 342

Topic: 7 - Neuromuscular disorders

Childhood onset nemaline rod myopathy: A report of two siblings

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Objectives: Nemaline myopathy (NM) is an uncommon disorder with inclusions known as nemaline bodies/rods presence in muscle fibers. Muscle weakness and hypotonia are apparent from the neonatal period. We report a rare presentation of NM seen in two siblings with similar symptoms. Both had slender physique, delayed motor milestones including delayed walking, normal language and cognitive milestones and difficulty in fast movements and change of posture. They had slowly progressive weakness. There was positive family history.

Methods: Investigations included CPK levels, motor-sensory conduction velocities, EMG studies, Muscle biopsy, MGT (Modified Gomori's Trichrome) stain and histo-chemistry studies.

Results: CPK levels were raised (279 U/l) with myopathic pattern of EMG with decreased motor unit potentials, recruitment with markedly decreased amplitude; mild decrease in motor conduction velocity in tibial-peroneal nerves in both with decreased ulnar nerve sensory conduction velocity in the girl and positive muscle biopsy. Both were diagnosed as Nemaline Rod myopathy.

Conclusion: Nemaline myopathy should be suspected in children with generalized hypotonia and progressive muscle weakness along with delayed motor milestones. This report highlights the importance of EMG & conduction velocity studies along with histo-chemistry & ultra-structural examination in diagnosis of NM in absence of availability of genetic studies.

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Abstract - WCN 2013

No: 352

Topic: 7 - Neuromuscular disorders

Subacute progressive ophthalmoplegia associated with dermatomyositis

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Background: Dermatomyositis is a systemic autoimmune disorder that primarily affects the skin and the proximal groups of limb muscles. Extraocular muscle involvement is a very rare manifestation of dermatomyositis.

Objective: Pathogenesis of ophthalmoplegia in the dermatomyositis patient is investigated.

Patient and method: The patient was a 53-year-old Japanese male who presented with myalgia and difficulty in climbing stairs. He developed progressive diplopia with eyelid edema and ocular pain two weeks after presentation. A femoral muscle biopsy and MRI scans of extraocular muscle and response of corticosteroid therapy were investigated.

Results: The physical examination revealed the presence of Gottron's papules on the dorsal surface of the hands, and a neurological examination showed symmetrical weakness of the proximal muscles, including the upper limbs muscles, and bilateral ophthalmoplegia with total limitations of eye movements. A muscle biopsy showed per fascicular atrophy with CD4 and CD8 positive lymphocyte infiltration into the interfascicular septae and within the fascicles. Orbital MRI scans revealed an increased thickness and high intensity signals of the extraocular muscles on fat-suppression T2-weighted images suggesting myositis. Corticosteroid therapy improved the muscle weakness, ophthalmoplegia and abnormal MRI findings. No myositis-specific autoantibodies, including anti-aminoacyl-tRNA synthetases or anti-signal recognition particles were detected in the serum.

Conclusion: The pathogenesis of ophthalmoplegia in the patient may be associated with inflammatory process of orbital muscles because of the orbital MRI images showing increased muscle thickness with abnormal intensity signal and dramatic efficacy of the steroid therapy on the ophthalmoplegia and abnormal MRI images.

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Abstract - WCN 2013

No: 283

Topic: 7 - Neuromuscular disorders

Guillain-Barré-like syndrome, as a rare presentation of adult T-cell leukemia-lymphoma: A case report

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Background: Guillain-Barré syndrome is an acute symmetric inflammatory polyradiculoneuropathy which commonly begins after an acute infectious process, immunization, or surgery. It has also been reported in association with some malignancies including lymphoma, small cell lung cancer and leukemia, but not as the primary presentation of Adult T-cell Leukemia/Lymphoma (ATLL) that is a non-Hodgkin T-cell Lymphoma linked to infection by the human T-cell lymphotropic virus type 1 (HTLV-1). The HTLV-1 has been endemic to certain parts of Iran like Khorasan province in the northeast since 1985 with 2.3% prevalence rate of infection. Thus, some rare neurologic complications due to this infection occasionally occur in this area that is worth to be reported.

Case report: In March 2009, a 21-year-old woman was admitted to Qa'em Hospital neurology ward in Mashhad having unilateral facial paresis and then developed progressive ascending flaccid tetraparesis with generalized areflexia. Electrodiagnostic studies revealed acute motor axonal polyradiculoneuropathy (variant of Guillain-Barré syndrome). Further evaluations revealed severe leukocytosis, increased erythrocyte sedimentation rate, increased protein content and presence of several lymphocytes in cerebrospinal fluid, and then the presence of HTLV-1 in serum and cerebrospinal fluid. Finally, biopsy of the enlarged lymph nodes resulted in the diagnosis of ATLL.

Conclusion: This case describes an extremely rare presentation of ATLL with a Guillain-Barré-like syndrome. Thus, in patients with manifestations resembling Guillain-Barré syndrome, it is important to consider

all the possible etiologies that can lead to this syndrome, and especially this is of value for diseases that are rare, but endemic, in specific geographic regions.

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Abstract - WCN 2013

No: 293

Topic: 7 - Neuromuscular disorders

Dilated cardiomyopathy in Becker muscular dystrophy

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Background: Becker Muscular Dystrophy (BMD) was first described in 1955 by a German Neurologist and Geneticist, Peter Emil Becker (1908–2000). The incidence of this rare X-linked muscular dystrophy is 3 in 100,000 live male births. The most common cardiac presentation is cardiomyopathy (occurs in 50% of cases) and conduction disorders. In the archives of the Philippine Heart Center, the most common etiology of dilated cardiomyopathy was found to be ischemic (59% of cases). In the same study, a non-ischemic cause of dilated cardiomyopathy was found in 41% of cases with following etiologies: idiopathic (77%), alcoholic (13%), myocarditis (2%) and peripartum (4%). Cardiomyopathy from BMD was rarely encountered.

Case: This is a case of a 27 year-old male who was diagnosed with BMD during his childhood. He developed gradual progressive muscle weakness with decrease in basal functional capacity. Diagnosis of dilated cardiomyopathy was based on the clinical and echocardiographic findings. He was later admitted due to ventricular tachycardia.

Diagnosics: The chest X-ray showed cardiomegaly with an increased cardiothoracic ratio. There was markedly elevated CK-MM at 4319 IU/L. Two dimensional echocardiography showed generalized hypokinesia with severely depressed systolic function (EF = 25% by Simpson's, 22% by Teicholz formula). There was dilated left ventricular cavity (LVMI of 132 g/m², LVEDD = 6.9 cm) and left atrium (LAVI = 29 ml/m²). The sphericity index was 1.23.

Conclusion: BMD is rare, however the knowledge of the disease progression including the development of cardiomyopathy and conduction disorders (atrial and ventricular arrhythmia) will aid the clinician in the management and enable the anticipation of probable sequelae.

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No: 560

Topic: 7 - Neuromuscular disorders

Piriformis syndrome and peroneal H-reflex

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Background: The electro-diagnostic studies defining piriformis syndrome was elaborated according to the peroneal H-reflex. The aim is to define electrical criteria with delay in latency and decreased amplitude during stress testing.

Objective: Combined to clinical exam, the peroneal H-reflex at stress testing are considered highly suggestive of piriformis syndrome.

Patients and methods: 19 patients suspected to suffer from piriformis syndrome after ruling out other causes of spinal affection were tested with EMG exam on both sides in neutral and dynamic positions. The amplitude and latency of the H-reflex on the peroneal nerve were studied and compared with the opposite painless side.

Results: The latency of the peroneal H-reflex at the normal side was delayed by 0.7 ms with the stress test. At the painful side, the delay was 5.4 ms, and when we excluded two patients with 2 ms delay confirmed not having piriformis syndrome, the average was 6 ms. The amplitude of the peroneal H-reflex at the normal side was decreased by 14% with the stress test, 70% at the painful side and 74% after excluding patients confirmed not having piriformis syndrome.

Conclusion: Our results confirm the importance of the peroneal H-reflex in diagnosing piriformis syndrome. The delay of latency at stress testing should be considered positive above 3 ms. At the same time the amplitude of the reflex has decreased more than 50% compared with the reflex in neutral position of the limb. Combined to clinical exam, these values are considered highly suggestive of piriformis syndrome.

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Abstract - WCN 2013

No: 562

Topic: 7 - Neuromuscular disorders

Correlation between oral corticosteroid therapy and present disease status in myasthenia gravis: A multicenter cross-sectional study in Japan

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Background: Patients with myasthenia gravis (MG) often do not achieve complete remission without immunotherapy, and many continue to have ADL difficulty due to insufficient improvement and long-term adverse effects of oral corticosteroids.

Objective: The aim of this study was to delineate the status of oral corticosteroid therapy in MG, and to elucidate the effectiveness of oral corticosteroids according to dosing regimen.

Methods: We evaluated 676 MG patients from 11 neurological centers. Disease severity was determined according to MGFA, QMG and MG composite. Clinical state following treatment was categorized according to MGFA postintervention status. We also completed the Japanese version of the 15-item MG-specific QOL scale (MG-QOL15-J). Multivariate regression analysis was used to determine the correlation between medication and clinical factors.

Results: Multivariate regression analysis revealed that the worst QMG and MG composite scores for the entire disease period had significant negative effects on the present disease severity. Achievement of minimal manifestation (MM) or better using maximum prednisolone (PSL) dose or reduced PSL dose by 25% or more after adding calcineurin inhibitors had significantly positive effects on the present QMG, MG composite and MG-QOL15-J. The maximum dose and administration period of PSL did not influence present disease severity, but administration period of PSL \geq 20 mg mg/day had significantly negative effects on the present QOL.

Conclusion: Achieving a status of MM or better by maximum PSL dose or reduced PSL dose combined with other agent may improve the present MG status and QOL.

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Abstract - WCN 2013

No: 563

Topic: 7 - Neuromuscular disorders

Effect of food on pharmacokinetics of 3,4-Diaminopyridine in rats and healthy volunteers

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Background: 3,4-Diaminopyridine (3,4-DAP) is commonly used to treat neuromuscular diseases such as Lambert–Eaton myasthenic syndrome and multiple sclerosis; however, the pharmacokinetics of 3,4-DAP is not well understood.

Objective: We aimed to investigate the effect of food on the pharmacokinetics of 3,4-DAP in rats and humans. In addition, we aimed to study the effect of food on the intestinal absorption of 3,4-DAP in rats.

Material and methods: We administered single 3,4-DAP doses of 10 mg/kg orally or 2 mg/kg intravenously to Wistar male rats (n = 4–7) and 10 mg 3,4-DAP to healthy volunteers (n = 5) in the fasting state and after meals. The intestinal absorption rate of 3,4-DAP was estimated by using a *in situ* closed loop method in rats.

Results: The areas under the serum concentration time curve for 3,4-DAP significantly decrease by food intake, and the corresponding bioavailability values were also significantly decrease from 25.1% ± 4.0% to 12.1% ± 0.8% (mean ± SD; P < 0.01) in rats. In the healthy volunteers, fasting might have reduced the time to reach the maximum serum concentration (C_{max}) of 3,4-DAP and elevated C_{max}. *In situ* close loop method, food reduced the intestinal absorption rate of 3,4-DAP by 5–10%.

Conclusion: These studies showed that food intake lowered the absorption of 3,4-DAP. Therefore, the administration of 3,4-DAP without food might be a candidate of improvement of the effect.

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No: 660

Topic: 7 - Neuromuscular disorders

Clinical picture of anti-MuSK-antibody-positive juvenile myasthenia gravis in Japan

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Objective: The present study aimed to determine the characteristics of anti-muscle-specific receptor tyrosine kinase (MuSK)-antibody-positive juvenile myasthenia gravis (JMG) (onset before age 15). Information on JMG is limited, and they are reported to be clinically and immunologically distinct from adult onset myasthenia gravis (MG).

Patients and methods: Four patients were positive for anti-MuSK-antibody among 28 patients collected nationwide from 2006 through

2009 for antibody assay in our institute. We investigated the clinical characteristics and measured IgG subclasses in these patients.

Results: All four patients presented generalized MG with relatively severe clinical course. There were no thymus abnormalities. IgG subclass was dominantly of IgG4 subclass.

Conclusions: The clinical characteristics and clinical course of anti-MuSK-antibody-positive JMG differ from general JMG or adult-onset MG.

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No: 612

Topic: 7 - Neuromuscular disorders

Baff (B cell activating factor) can be the clinical marker in myasthenia gravis

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Background: Myasthenia gravis (MG) is an antibody mediated disease in which the target autoantigen is the acetylcholine receptor (AChR) at the postsynaptic membrane of the neuromuscular junction. B cell activating factor (BAFF) is the potent B cell survival factor and is necessary for peripheral B cell differentiation and maturation. Excess BAFF promotes the survival and growth of autoreactive B cells. The previous studies have shown that serum BAFF levels in patients with MG are higher than in control subjects. BAFF may play an important role in the pathogenesis of MG.

Objective: To compare serum BAFF levels in patients with AChR antibody positive MG (AChR MG) and muscle specific kinase antibody positive MG (MuSK MG).

Patients and methods: We retrospectively analyzed 14 ocular AChR MG, 31 generalized AChR MG and 25 MuSK MG. We compared the serum BAFF levels with the clinical characteristics among 3 groups.

Results: The serum BAFF levels in generalized AChR MG (1559.6 ± 108.6 pg/mL) were significantly higher than in MuSK MG (568.1 ± 36.3 pg/mL) and ocular AChR MG (598.1 ± 65.9 pg/mL). There was a correlation between the serum BAFF levels and disease severity in AChR MG. The serum BAFF levels of the generalized AChR MG with thymic abnormalities (1536.4 ± 728.8 pg/mL) were statistically higher than the ocular AChR MG.

Conclusion: The serum BAFF levels are increased in patients with generalized AChR MG compared to MuSK MG and ocular AChR MG.

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Abstract - WCN 2013

No: 643

Topic: 7 - Neuromuscular disorders

Autosomal recessive limb girdle muscular dystrophy: Prospective study and characterisation of 280 cases by immunohistochemistry and immunoblotting

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Background: ARLGMD is a common disorder in Southern India due to high consanguinity.

Objective: To study pattern of ARLGMD in India.

Materials and methods: Prospective study of 300 cases seen between February 2010 and October 2012. All suspected cases of ARLGMD

attending the NMD clinic underwent phenotypic characterisation, muscle immunohistochemistry (IHC) and immunoblotting (IB).

Results: 280 cases biopsied, 226 had IHC and 176 of these IB. 54 patients excluded. Consanguinity—45.2%. 200 confirmed to have specific ARLGMD by IHC and/or IB. Commonest form: LGMD 2B (82/246—33.33%). Mean age of onset— 21.17 ± 6.32 (8–41 years). Mean duration— 7.24 ± 5.86 (1–36 years). Mean CK— 7966.7 ± 6029.6 IU/L (131–24037). All demonstrated dysferlin deficiency on IHC/IB. Second commonest—LGMD 2I (51/246—20.73%). Mean age of onset— 12.64 ± 7.17 (1–29 years). Mean duration— 8.54 ± 6.50 (1–27 years) Mean CK— 4059.56 ± 3210.8 (211–14667 IU/L). α -DG deficiency by IHC-9, IB-51. Third commonest LGMD 2C-F (35/246—14.23%). All confirmed by IHC/IB. Mean age of onset— 5.89 ± 3.45 (1–20 years). Mean duration— 4.56 ± 2.85 (1–12 years). Mean CK— 8688.31 ± 6113.86 IU/L (684–23577). Fourth group—LGMD 2A (25/246—10.16%). Age of onset— 15.52 ± 11.18 (2–41 years). Mean duration— 9.84 ± 9.33 (2–37 years). Mean CK— 2742.58 ± 2144.96 IU/L (286–9018). All confirmed by IB. Fifth group—LGMD 2G (8/246—3.25%). Mean age of onset— 12.38 ± 11.35 (5 to 40 years). Mean duration— 8.50 ± 6.87 (2–23 years). CK—718–9253 IU/L (mean \pm SD; 2574.4 ± 2847.52). Proximo-distal form with muscle atrophy, calf hypertrophy, foot drop. The course was slow in majority. All confirmed on IB. Last group—LGMD 2J-2 cases. Confirmed on IHC.

Conclusion: Our study confirms that LGMD 2B is the most common form of ARLGMD among our cohort. Further LGMD2I and 2G have a wider existence and may be among the common ARLGMDs in Indian population.

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No: 763

Topic: 7 - Neuromuscular disorders

The importance of follow-up thoracic imaging in myasthenia gravis patients

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Background: Approximately 20–30% of myasthenia gravis (MG) patients are accompanied by thymoma. In elderly patients, extra-thymic malignancies are also a matter of concern. Although most MG patients undergo thoracic imaging at the initial diagnosis, the follow-up protocol for thoracic imaging is not clear.

Objective: To elucidate the follow-up status of thoracic imaging in MG.

Patients and methods: From among 676 MG patients in 11 neurological centers, we evaluated 649 patients whose disease duration was over 1 year. The latest thoracic imaging (CT or MRI) after an interval of 1 year or more from the initial scan was evaluated in each case. The rate of abnormal findings and the details of findings in cases without thymoma (group N) and with thymoma (group T) were then investigated.

Results: Thoracic imaging follow-up was performed in 337 (51.9%) of MG patients studied (48.8% in group N, 74.1% in group T). The average interval was 7.3 years in group N and 7.8 years in group T. Abnormal findings were detected in 9.0% in group N and 25.2% in group T. Abnormal findings included breast cancer, swelling of lymph nodes and respiratory system infection in group N, while recurrence or enlargement of thymoma accounted for 76.9% of the abnormal findings in group T.

Conclusion: It is important to keep following up thoracic imaging in MG patients since the rate of abnormal findings is high even 7 years after onset.

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No: 719

Topic: 7 - Neuromuscular disorders

The initial electrodiagnostic signs that distinguish acute inflammatory demyelinating polyneuropathy from acute motor axonal neuropathy

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Background: Guillain-Barré syndrome (GBS) is divided into 2 neurophysiological subtypes, acute inflammatory demyelinating polyneuropathy (AIDP) and acute motor axonal neuropathy (AMAN). The early electrodiagnostic findings of AIDP overlap with AMAN, leading to the latter's under-diagnosis. Serial nerve conduction studies (NCSs) are more sensitive, but not always feasible.

Objective: To derive diagnostic criteria that distinguish AMAN from AIDP at presentation, by delineating their nascent electrodiagnostic features.

Patients and methods: We used at least 2 serial NCSs to categorise GBS patients into AIDP and AMAN. The initial NCS was scrutinized for features discriminating the 2 subtypes.

Results: Using serial NCSs, 30 GBS patients were divided into 11 AIDP, 14 AMAN, and 5 unclassified cases. The initial NCS, done at median 7 and range 2–14 days, showed prolonged distal motor latency in 7 AIDP and 8 AMAN patients. Six AIDP patients had slowing in non-entrapment sites and 4 had temporal dispersion (TD). Only 1 AMAN patient had these. Slowing at entrapment site was less specific, occurring in 8 AIDP and 3 AMAN patients. Three AIDP patients had conduction block (CB), all with slowing across the block. Seven nerves in 5 AMAN patients had CB, 5 without slowing.

Conclusion: The most specific initial signs of AIDP were slowing in non-entrapment sites and TD. CMAP reduction in at least 2 nerves without TD or slowing at non-entrapment sites and non-slowng CB predict AMAN. Using these features, 22 out of 30 patients were assigned to the right GBS subtypes; current criteria were correct in 14 patients.

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Abstract - WCN 2013

No: 761

Topic: 7 - Neuromuscular disorders

LGMD2I: Immunohistochemical and immunoblot technique assisted identification of 51 cases with both duchenne and becker phenotype

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Background: LGMD 2I is caused by mutations in fukutin-related protein gene (FKRP).

Objective: To describe the phenotype and findings of immunohistochemistry (IHC) and immunoblot (IB) in LGMD2I.

Materials and methods: Prospective study of 300 cases of ARLGMD seen at Neuromuscular Disorders clinic between February 2010–

October 2012. All underwent phenotypic characterisation, muscle histopathology, IHC and IB analysis. Age of onset <10 years taken as Duchenne phenotype; >10 years as Becker's phenotype.

Results: 280 cases biopsied, 226 had IHC and 176 of these IB. 54 patients excluded. LGMD-2I in 51 by IB. Nine of these 51 identified by IHC. M:F=4.1:1. Illness onset occurred in first to third decades (1–29 years; mean=12.64 ± 7.16). Exertional muscle pain and calf hypertrophy were prominent. Cardio-respiratory symptoms in 6 cases. Family history in 15. Duchenne phenotype—1, M:F=13:8. Symmetrical lower limb weakness in all. Proximal upper limb weakness—9. Hypertrophy of: Gastrocnemius—19, biceps—1, deltoid—2, triceps—3, extensor digitorum brevis—4. Ankle contractures—15, hip and knee contractures—3. Becker's phenotype—30. Onset in 2nd decade—76%. Lower limb proximal weakness—30. Proximal upper limbs weakness—9. Muscle hypertrophy: Gastrocnemius—25, deltoid and biceps—3, triceps—4, extensor digitorum brevis—16, hamstrings—1. Mean CK value=4059.56 ± 3210.82 IU/L. ECG-left ventricular hypertrophy—5/23. 2D-ECHO dilated cardiomyopathy with reduced left ventricular function—4/23, one had severe heart failure. The 156 kDa band specific for α -dystroglycan was absent in 38, severely reduced in 6, while band specific for dysferlin, Calpain, telethonin were present.

Conclusion: Our study confirms that LGMD-2I may be among the more common form ARLGMD in Indian population. It would be essential to identify them as this carries important implications for management and prognostication.

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Abstract - WCN 2013

No: 764

Topic: 7 - Neuromuscular disorders

Detection of limb girdle muscular dystrophy 2A (calpainopathy) by autocatalytic activity of calpain and by quantification of calpain bands by densitometry

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Background: LGMD 2A represents 9–30% of all AR-LGMD's. We describe clinical and Immunoblot (IB) features in 25 patients from South India with calpainopathy.

Materials and methods: This is a prospective case series from the Neuromuscular disorders clinic, NIMHANS. Patients with features of LGMD with autosomal recessive inheritance underwent phenotypic characterisation, muscle histopathology, immunohistochemistry(IHC) and IB.

Results: Among 200 patients confirmed to have AR-LGMD by IHC and / or IB there were 25 patients with LGMD2A. Diagnosis of LGMD 2A was confirmed by demonstrating the absence of 94 kDa (full length) and 60 kDa (degradation product) bands on IB in 15 patients, absence of autocatalytic activity of calpain-3 in 7 patients and quantitative reduction of calpain by densitometry in 3 patients. All had predominantly proximal muscle weakness at hips and shoulders. Mean age at onset was 15.5 ± 11.18 (2–41 yrs). Mean duration of illness was 9.84 ± 9.33 (2–37). The predominant presenting symptoms were difficulty in rising from floor, difficulty in running fast and tip toe walking. The other features include Scapular winging (72%), Biceps weaker than triceps (44%), Glutei weaker than iliopsoas (52%), Hip abductor sign (32%) and toe walking (52%). Mean CK value was 2742.58 ± 2144.96 (286–9018). Consanguinity was reported in 40%.

Conclusions: Calpainopathies formed 12.5 % of the AR-LGMD cases in our cohort. IB followed by quantitative estimation of calpain bands on

densitometry and/or by demonstrating loss of autocatalytic activity of calpain plays an important role in diagnosing calpainopathy and a subset of patients with normal calpain signal on IB require further analysis by densitometry or by demonstrating loss of autocatalytic activity of calpain.

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Abstract - WCN 2013

No: 769

Topic: 7 - Neuromuscular disorders

The impact of autoimmune thyroid disease on the course of myasthenia gravis

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Background: Autoimmune diseases often coexist in the same patients. The most common comorbid conditions with myasthenia gravis (MG) are autoimmune thyroid disease (ATD).

Objective: To determine the relationship between ATD and course of MG.

Material and methods: Examined population consisted of 343 consecutive patients with MG (236F, 107M) aged 4–89 seen at single centre. 83% were seropositive, in 6% antiMuSK Ab were detected. In all patients the concentrations of anti-TPO (thyroid peroxidase antibodies), anti-TG (thyroglobulin antibodies), anti-TSHR (thyrotropin receptor antibodies) and TSH level were measured. In doubtful cases the thyroid ultrasound examination was performed.

Results: ATD was diagnosed in 46 (13.4%) MG patients, 2–3 times more frequently than in general population. In 92 (26.8%) patients elevated thyroid antibodies were detected.

In patients with Graves' disease a higher incidence of ocular symptoms was noticed compared with a group of patients with positive thyroid antibodies and Hashimoto's thyroiditis ($p = .008$).

ATD prevalence was comparable in MG with early and late onset. There was no significant difference in the severity of MG and outcome in the group with and without ATD. In the group of patients with thyroid dysfunction of the nonautoimmune etiology trend towards more frequent incidence of the thymoma was stated as compared with ATD group and controls.

Conclusions: Our study provided the evidence of frequent co-existence of myasthenia with ATD. Except of a higher incidence of ocular symptoms in Graves' disease, ATD did not influence the clinical course or treatment results in our MG cohort.

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No: 771

Topic: 7 - Neuromuscular disorders

Swallowing and respiration relationship in amyotrophic lateral sclerosis

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Background: Difficulty in swallowing and respiratory muscle weakness are the most important factors in prognosis of ALS. We have studied swallowing function in ALS patient during inspiration and expiration separately.

Materials and methods: 25 patients diagnosed with ALS by El Escorial criteria were included in the study. Pulmonary functions were evaluated with sniff nasal pressure test. The presence of aspiration and swallowing dysfunction were checked with video fluoroscopy. Swallowing function of patients was checked with dysphagia limit accompanied by EMG.

Results: 25 patients were included in the study. The average inclusion time after diagnosis were 24.7 ± 27.3 months. 17 cases were in stage 3, 8 cases were in stage 4A and 2 cases were in stage 4B. 25 patients were checked for dysphagia limit and it was normal in 15 patients, abnormal in 10 patients. All cases could swallow 3 cm^3 liquid during the expiration. All patients with normal dysphagia limit, 3 patients with >5 dysphagia limit and 1 patient with ≤ 5 dysphagia limit could swallow during the inspiration. 6 of 7 patients with $\leq 5 \text{ cm}^3$ dysphagia limit could not swallow 3 cm^3 liquid during the inspiration.

Discussion: Absence of ability of swallowing in inspiration can use as a threshold in situation of swallowing dysfunction. We think, in which part of respiration patient can swallow is important in terms of dysphagia limit. We think ALS patients with $\leq 15 \text{ cm}^3$ dysphagia limit should be accepted as Stage 4A.

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Abstract - WCN 2013

No: 704

Topic: 7 - Neuromuscular disorders

Clinical and genetic characteristics of chronic progressive external ophthalmoplegia (CPEO)

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Background: Chronic progressive external ophthalmoplegia (CPEO) is a prominent clinical manifestation in mitochondrial disease, and can be caused by single or multiple deletions in mitochondrial DNA (mtDNA), as well as by mtDNA point mutations. Single deletions are usually sporadic, whilst multiple deletions arise due to mutations in nuclear genes. Here, we examine the distribution of the various mutations in a cohort of 290 patients with CPEO, and compare the clinical features exhibited by the various groups.

Methods: Muscle and blood, available from 203 of our patients, were screened for mtDNA deletions and for the most common mtDNA point mutations, m.3243A>G and m.8344A>G. Patients were also screened for GCG expansions in PABPN1.

Results: Single deletions in mtDNA were identified in 46% of the patients. Of these patients, 36% had CPEO only, 16% had CPEO plus proximal muscle weakness, while the remaining patients were affected by additional clinical features. Multiple mtDNA deletions were found in 24 patients (11.8%): of these, 21 (87.5%) had a mutation in POLG and 2 (8.3%) had a mutation in C10orf2. 3 common mutations in POLG were identified. Polyneuropathy is common in patients with multiple mtDNA deletions, but rare in single deletion patients. The m.3243A>G mutation was identified in 3.8% of the patients, and 1.7% of patients were positive for a GCG expansion in PABPN1.

Conclusion: Our genetic investigations in patients with CPEO and observations of the clinical features associated with the different causes of CPEO have allowed us to develop a diagnostic protocol for patients with CPEO.

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Abstract - WCN 2013

No: 697

Topic: 7 - Neuromuscular disorders

Complex diagnostics - entrapment neuropathy of the upper limb

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Background: Entrapment neuropathies of the upper limb are common problems. Despite all the modern diagnostics in rare case it is not possible to identify a clear correlation between clinical data and instrumental studies such as electroneuromyography (EMG), ultrasound (US).

Objective: Improving complex diagnostics with clinical, electrophysiological and US techniques.

Materials and methods: A total of 20 patients were examined with clinical diagnosis of entrapment neuropathy in upper limb, using a combination of EMG and US,

Result: The predominance of pain neuropathic component was observed in all patients. The pain is largely dependent on the psychological condition of the patients, half of patients had motor defect. In 85% of the cases provocative testing (Tinel's, Phalen's) proved positive, the severity of which was confirmed with EMG and US data. In 3 patients with suspected carpal tunnel syndrome (CTS) we have identified tenosynovitis in the thumb tendons, without symptoms of CTS, and one person manifested a combination of tenosynovitis and CTS. It was found that 10% of chronic entrapment neuropathy proved positive during provocative testing, severity of which was not confirmed by the clinical neurophysiological and US diagnosis.

Conclusion: Diagnosis of neuropathies is improved through a combined functional and morphological evaluation of the nerve by using EMG and US. But in rare cases confirmed diagnosis with clinical instrumental methods cannot be obtained at all. And besides neurological examination and modern instrumental methods the physiological condition of the patients should be taken into account for the selection of optimal and appropriate therapy.

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No: 695

Topic: 7 - Neuromuscular disorders

Role of occupational and environmental exposure to heavy metals in amyotrophic lateral sclerosis: A case-control study in Campania Region

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Objectives: Amyotrophic lateral sclerosis (ALS) is a progressive degenerative disorder of upper and lower motor neurons. Several evidences demonstrate the role of environmental factors in ALS onset. The aim of this study was to evaluate relationship between heavy metal occupational exposure and ALS in Campania.

Materials and methods: Blood samples of 34 ALS patients and 34 healthy age-, sex-matched controls were processed to assess serum concentrations of lead, aluminum and manganese by atomic absorption spectroscopy. Toxicological results were compared to Biological Reference Values and to working activity.

Results: Among ALS patients serum lead was increased in 5.88%, manganese increased in 41.17% specifically in truck drivers while aluminum decreased in 70.59% specifically in farm workers. Among healthy controls investigated lead was increased in 2.94%, manganese increased in 20.58% while aluminum increased in 17.64% and decreased in 8.82%. There was no blood levels altered value in 8.82% of patients and in 61.76% of healthy controls.

Discussions: Previous studies underlined role of heavy metals in neurodegenerative mechanism: lead interferes with calcium metabolism promoting axonal degeneration; manganese chronic exposure can determine manganese and parkinsonism; aluminum reduces calcium adsorption promoting demyelination.

Conclusions: Results suggest presence of heavy metals abnormal blood levels in ALS patients in Campania. We detected manganese increased mainly in ALS patients with late onset and aluminum decreased mainly in farm workers. Significant increase of lead was not found: it might be in agreement with longer survival in SOD1 G93A mutation transgenic mice exposed to lead in low concentrations according to increased VEGF expression in the ventral horn.

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No: 716

Topic: 7 - Neuromuscular disorders

Muscle magnetic resonance imaging (MRI) in patients with LGMD 2I confirmed by immunohistochemistry and/or immunoblotting

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Background: LGMD 2I is caused by mutations in fukutin-related protein gene. Muscle MRI is reported to demonstrate distinct findings.

Objective: To describe the muscle MRI findings in LGMD 2I.

Materials and methods: Prospective case series seen from 2012 to 2012. ARLGMD phenotype patients with absent α -dystroglycan expression on muscle immunohistochemistry and/or immunoblotting and thus having LGMD2I underwent MRI on 1.5 T machine. Protocol included T1W, T2W, STIR sequences. Fibrofatty replacement was evaluated using Mercuri score. Myoedema evaluated by new oedema score (3 grade scale).

Results: 23 LGMD2I cases. DMD phenotype in 4; BMD in 19. M:F:4.8:1. Mean age-22.13 \pm 13.08 years (6–37). Fibrofatty replacement: Gluteus maximus involved in-22 (95.7%); Gluteus medius and minimus-82% (17). Iliopsoas-81.8% (22). Posterior compartment of thigh-22 (95.7%); anterior-19 (87%). Long head of biceps femoris (BF)-22 (95.7%), short head of BF spared/hypertrophied-15 (65.2%). In anterior compartment of thigh vastus medialis and intermedius more involved than vastus lateralis. Adductor longus-8 (34.8). Adductor magnus affected in all, 6-stage 4; 3-stage 3. Spared and hypertrophied: Gracilis-20 (87%); Sartorius-18 (78.3%); Rectus femorus-13 (56.5%). Leg muscles: Tibialis anterior (TA) involved-7 (31.4%); peronei-17 (73.9%); soleus-18 (78.3%); medial gastrocnemius-15 (65.2%); lateral gastrocnemius-9 (39.1%). Tibialis posterior-spared in 22 (95.7%). Oedema assessment: Thigh: Anterior compartment-19 (82.6%); posterior-14 (60.9%); medial-9 (39.1%). Leg: Soleus-21 (87%); medial gastrocnemius-20 (87%); lateral gastrocnemius-17 (73.9%). Anterior compartment and peronei muscles showed less oedema.

Conclusion: Posterior and medial compartments of thigh were affected early and severely. Gracilis, sartorius, rectus femoris spared or hypertrophied. Peronei was more affected than TA. Medial gastrocnemius was more than lateral head. Myoedema was noted predominantly in anterior and posterior thighs, soleus, and gastrocnemii. Thus, muscle MRI shows distinct pattern in LGMD2I (FKRP) and could be a useful diagnostic tool.

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No: 717

Topic: 7 - Neuromuscular disorders

Magnetic resonance imaging findings in immunohistochemically confirmed sarcoglycanopathies

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Background: Sarcoglycanopathies (LGMD2 C,D,E,F) are a group of four genetically distinct but clinically similar muscular dystrophies. Scant reports present on MRI of muscle in SG's.

Materials and methods: Muscle MRI was performed in 1.5 T machine on Immunohistochemistry and/or Immunoblot confirmed sarcoglycanopathies (SG's). Protocol included T1W, T2W, and STIR sequences. 37 were muscles studied in each lower limb. Fibrofatty replacement was assessed using Mercuri scoring. Myoedema was evaluated on STIR.

Results: Twelve patients of SGs. M:F-2:1. Mean age-10.91 \pm 5.89 years (2–28). Fibrofatty infiltration demonstrated in: Gluteus maximus-12 (100%); Gluteus medius and minimus-9 (75.0%); Tensor fascia lata-10 (83.3%). Spared muscles: Rectus femoris-7 (58.3%); sartorius-8 (66.7%); gracilis-11 (91.7%). Thigh involvement: Posterior compartment-12 (100%); early and severe affection of long head of biceps femoris (BF)-12 (100%); short head in (66.7%); semimembranosus-8 (66.7%); semitendinous-5 (41.7%). Anterior compartment in 11 (91.7%); vastusmedialis-10 (83.3%) more often and severely affected than vastus lateralis and intermedius. Adductor magnus and brevis involvement-12 (100%); adductor longus-7 (58.3%). Leg muscle involvement: Anterior leg compartment-3 (25%). Soleus-7 (58.3%) with mild to moderate fatty infiltration; Medial and lateral gastrocnemius-7 (58.3%). Lateral compartment -10 (83.3%). Asymmetrical involvement in 50%. Tibialis posterior spared-11 (91.7%). Myoedema present: Gluteus maximus-2 (16.7%); gluteus medius and minimus-7 (58.3%); adductor longus-6 (50%); anterior leg compartment-3 (25%) which was segmental; soleus-9 (75%) and was diffuse and global; gastrocnemius-6 (50%). Tibialis posterior(mild)-1 (8.3%). Adductor magnus totally spared in all.

Conclusions: MRI revealed involvement predominantly of hip extensors, adductors and abductors along with severe affection of knee flexors. Vastus medialis more affected than other Vastii. Leg showed more affection of lateral followed by posterior compartment. Myoedema noted particularly in Gastro-soleus with sparing of Tibialis posterior.

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No: 688

Topic: 7 - Neuromuscular disorders

A retrospective review of autonomic screening tests conducted at a tertiary general hospital

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Background: Autonomic screening tests (AST) are commonly employed to assess the integrity of the sympathetic and parasympathetic systems.

Objectives: To describe the case mix, findings and usefulness of AST in a tertiary General Hospital setting.

Patients and methods: All AST performed in 2011–2012 were retrospectively reviewed. Referring diagnoses were classified into: orthostatic, neurologic, thermoregulatory and known/suspected disease assessment

(e.g. Diabetes mellitus, Parkinsonism). Abnormalities and final diagnosis were noted, along with medication and patient cooperation. The relevance of AST to the clinical problem was ranked on a 3-point scale.

Results: 162 patients underwent AST. Orthostatic dizziness was the most common referring diagnosis (53.1%) followed by known/suspected disease assessment (33.3%), thermoregulatory (8.6%) and neurological (4.3%). 47.5% of ASTs were abnormal, 40.7% were normal and 11.7% inconclusive. The most frequent abnormal final diagnosis was isolated orthostatic hypotension (OH) (27.3%). 25.5% of diabetics were eventually diagnosed with diabetic autonomic neuropathy. AST was very helpful in 56.2%, somewhat helpful in 32.7% and inconclusive in 11.1%. Among inconclusive ASTs, 61.1% were attributed to poor performance of test maneuvers, 16.7% to medications and 16.7% to both. All patients with irregular heart rhythms had inconclusive results.

Conclusion: AST was abnormal in more than half. The most common abnormality was isolated OH. Patients referred for known/suspected disease assessment were most likely to have abnormal AST. Conversely, patients with thermoregulatory problems were most likely to have normal AST. Poor performance of test maneuvers and irregular heart rhythm were confounders in the general population, while medication interfered with AST of Parkinsonism.

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No: 742

Topic: 7 - Neuromuscular disorders

Late onset post-radiotherapy brachial plexopathy

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Introduction: Risk of developing post-radiotherapy brachial plexopathy from breast cancer is less than 2%. It usually appears around 10.5 months (range 1.5–77) after the start of treatment and only in 25% of cases alterations in EMG are identified.

A case of brachial plexopathy developed 13 years after radiotherapy treatment.

Material and methods: Female patient, 66 years old with pain and muscle weakness in the distal portion of left arm and hand for 7 months. She had been treated 13 years ago for an infiltrating ductal carcinoma in the left breast with reconstruction and chemotherapy and subsequent radiotherapy.

Results: Electromyography: Left brachial plexopathy C7–D1.

MRI Cervical: C5–C6 degenerative disc disease without signs of CNS involvement.

MRI brachial plexus: Fibrosis post-radiotherapy encompassing left brachial plexus fascicles.

Laboratory study: Normal.

Conclusions: Post-radiotherapy plexopathy may develop years after completion of treatment probably due to the evolution of tissue fibrosis.

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Abstract - WCN 2013

No: 743

Topic: 7 - Neuromuscular disorders

Juvenile-onset Becker type myotonia congenita

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Introduction: Becker type myotonia congenita is a very rare (1:100,000) autosomal recessive disease which usually begins between the ages of 4

and 12 years old and its characteristics are defined by small patients series. CLCN1 genetic alterations that occur are multiple and usually heterozygous. We present a juvenile-onset Becker myotonia series.

Material and methods: Observational study of Becker type myotonia genetically diagnosed patients series. We analyzed the age of onset and clinical features.

Results: 3 patients (2 males and 1 female) with a mean age of onset of 1, 4 and 3 years old. Only one of them had family disease history. All had weakness at the beginning of the activity but without paralysis and with subsequent warm-up phenomenon. All of them had clinical and electrical myotonia. There was no clinical or EMG signs of muscular dystrophy. CPK levels were normal. Genetic studies identified a homozygous mutation in the male patient with family history, a heterozygous mutation for the female patient and 2 different heterozygous mutations for the gene CLCN1 in the other male.

Conclusions: Becker type myotonia congenita may have its onset in adolescence with similar symptoms than those described in earlier ages of onset. Genetic alterations are very variable.

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No: 744

Topic: 7 - Neuromuscular disorders

Could acetate Eslicarbazepina be effective in neuropathic pain?

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Introduction: Eslicarbazepina Acetate metabolite is a prodrug of an active Oxcarbazepine metabolite and with Carbamazepine, which share their sodium channels blockers mechanism. Theoretically it could be effective in patients with neuropathic pain.

Material and methods: An observational study in patients with neuropathic pain resistant to other drugs who are treated with Eslicarbazepina for 12 weeks. Pathology causing the pain, the effectiveness and side effects were analyzed.

Results: 15 patients (4 female and 11 male) with a mean age of 64.3 years. 3 patients with Trigeminal Neuralgia, 12 patients with non-diabetic PNP. Previous treatments: Pregabalin 80%, 33.3% Gabapentin, Carbamazepine 20%. Visual Analog Pain Scale Pre Eslicarbazepina treatment 6.6/10, post-treatment 3.3/10. Withdrawal of medication in 3 patients (20%) with PNP: 2 due to ineffectiveness and 1 due to diarrhea.

Conclusions: Acetate Eslicarbazepina could be effective in neuropathic pain. Clinical trials are needed to determine its effectiveness level and its potential use in monotherapy.

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Topic: 7 - Neuromuscular disorders

Clinical and immunological predictors of prognosis for Japanese patients with thymoma-associated myasthenia gravis

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Background: About 30%–50% of patients with thymomas also have autoimmune myasthenia gravis (MG). There are no immunological markers to predict the prognosis of patients with thymoma-associated MG (T-MG). Anti-Kv1.4 antibody, one of the striatal

antibodies, is specific for MG and related to severe symptoms and thymomas.

Objective: To evaluate the role of anti-Kv1.4 antibody as a predictor of prognosis among various clinical and immunological factors in T-MG.

Material and methods: We retrospectively investigated 56 Japanese patients with T-MG. Immunological factors including three autoantibodies and human leukocyte antigen DR alleles were investigated, as well as the clinical features of thymomas and MG. The factors associated with thymoma recurrence or MG relapse were examined by logistic analyses.

Results: Anti-Kv1.4 antibodies were detected in 28 patients, and thus the MG patients were divided into two groups: an anti-Kv1.4-positive group and an anti-Kv1.4-negative group. Five patients developed MG after the removal of thymoma. Patients with anti-Kv1.4 antibodies showed higher frequencies of thymoma recurrence and MG relapse compared to those without. The final outcomes of MG were more often unfavorable in the anti-Kv1.4 positive group than the negative group. Eleven patients died, but none died of MG. Anti-Kv1.4 antibody was a factor associated with thymoma recurrence, as well as Masaoka stage 4, World Health Organization type B3, and adjuvant radiotherapy in univariate logistic analyses. Multivariate analyses showed that anti-Kv1.4 antibody was the only independent factor associated with MG relapse.

Conclusions: Anti-Kv1.4 antibody is a useful predictor of the prognosis of Japanese patients with T-MG.

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No: 939

Topic: 7 - Neuromuscular disorders

The up-regulation of IL-1 β /TGF- β 1 and hypoxia induced in immobilization are related to the molecular mechanisms underlying muscle contracture

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Background/objective: This investigation was conducted to determine the molecular mechanism governing muscle contracture in immobilized rat soleus muscle.

Material and methods: Wistar rats were divided randomly into immobilization and control groups. In the immobilization group, both ankle joints were fixed in full plantar flexion with plaster casts for 1, 2, 4, 8, and 12 weeks. The right soleus muscle was immunostained for type I and III collagen and α -SMA (a myofibroblast marker). Additionally, the left soleus muscle was used to detect the expression of type I and III collagen, α -SMA, IL-1 β , TGF- β 1, and HIF-1 α mRNA by RT-PCR.

Results: Type I and III collagen, α -SMA, IL-1 β , and TGF- β 1 expression was significantly greater in the immobilization group than in the controls at each time point, whereas HIF-1 α expression was significantly greater in the immobilization group than in the controls at 4, 8, and 12 weeks. Furthermore, in the immobilization group, type I collagen, α -SMA, and HIF-1 α expression at 4, 8, and 12 weeks was significantly greater than at 1 and 2 weeks.

Conclusion: α -SMA, IL-1 β , and TGF- β 1 expression was significantly higher in the immobilization group than in the controls at 1 week. The

up-regulation of IL-1 β and TGF- β 1 may activate fibroblasts and promote their differentiation into myofibroblasts, and we suggest that these changes are associated with the increased expression of type I and III collagen. Additionally, the soleus muscle became hypoxic after 4 weeks of immobilization. Thus, we suggest hypoxia accelerates fibrosis and these alterations may influence the progression of muscle contracture.

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No: 938

Topic: 7 - Neuromuscular disorders

Effects of heat stress on glucocorticoid-induced myopathy

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Background: Glucocorticoids mainly induce atrophy of fast muscles. Heat stress inhibits disuse muscle atrophy by increasing the levels of heat shock protein (Hsp) 72 in fast muscles.

Objective: The purpose of this study was to investigate the influence of heat stress on glucocorticoid-induced myopathy.

Materials and methods: Eight-week-old Wistar rats were randomly assigned to the control (injected with 2 ml/kg saline 6 d/wk for 2 wk), dex (injected with 2 mg/kg dexamethasone 6 d/wk for 2 wk), and dex and heat (injected with dexamethasone and hindlimbs immersed in water at 42 °C 6 d/wk for 2 wk) groups. At the end of the experimental period, the extensor digitorum longus muscle was extracted. We examined the diameters of muscle fiber types I, IIA, and IIB; the number of myonuclei and capillaries per muscle fiber; expression of Hsp72; and concentration of insulin-like growth factor (IGF)-1.

Results: The diameters of all muscle fiber types, the concentration of IGF-1, and the number of capillaries per muscle fiber were significantly lower in the dex group than in the control group. The diameters of all muscle fiber types, the numbers of myonuclei and capillaries per muscle fiber, and the expression of Hsp72 were significantly higher in the dex and heat group than in the dex group.

Conclusion: Our results suggest that heat stress inhibits development of glucocorticoid-induced myopathy via inhibiting reduction of the numbers of capillaries and increasing the number of myonuclei and Hsp72 expression.

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Abstract - WCN 2013

No: 893

Topic: 7 - Neuromuscular disorders

Endocrine myopathies

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Background: Most endocrine disorders are now detected and treated at an early stage, and muscle abnormalities are more rarely troublesome or severe. Endocrine myopathy (EM) are often described in thyroid, parathormone dysfunction and vitamin D deficiency.

Objective: To report clinical, biological and histological findings of Tunisian patient with EM and to discuss diagnostic approach and patient's management.

Patients and methods: Over 20 years (1992–2012), 12 patients were followed up in our department for EM.

Clinical, biological, electrophysiological and histological findings are analyzed.

Results: They were 4 males and 8 females (mean age 33 years). All patients presented initially a proximal myopathy. Serum creatine kinase levels, electromyography and muscle biopsy were normal in all patients. Laboratory analysis revealed hyperparathyroidism in the first and second case, hypothyroidism in four cases. Six patients had low serum level of 25 hydroxy Vitamin D. Furthermore, radiological investigations confirmed osteomalacia in these patients. The myopathy improves with restoration of normal PTH levels, thyroidism and vitamin D replacement.

Conclusion: Proximal myopathy may rarely be displayed as the sole manifestation of endocrine disorders. However, it is recommended that endocrine myopathy should be taken into account during differential diagnosis of proximal muscle weakness mainly because they are treatable causes.

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Abstract - WCN 2013

No: 865

Topic: 7 - Neuromuscular disorders

Cerebrospinal fluid biomarkers of neurodegeneration in patients with juvenile and classic myotonic dystrophy type 1

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Aim: To analyze cerebrospinal fluid (CSF) levels of total tau (T-tau), phosphorylated tau (P-tau) and 42 amino acid form of β -amyloid ($A\beta_{42}$) in patients with myotonic dystrophy type 1 (DM1), and their possible correlations with cognitive and behavior manifestations in these patients.

Methods: Lumbar puncture was performed in 74 patients with DM1 (27 with childhood/juvenile (jDM1) and 47 with adult form (aDM1) of disease) and 26 control subjects who were subjected to orthopedic surgery. Sandwich enzyme-linked immunosorbent assay (ELISA) was used for measuring the levels of T-tau, P-tau and $A\beta_{42}$.

Results: CSF level of $A\beta_{42}$ was the lowest in patients with jDM1 and the highest in controls ($p < 0.05$). Tendency of T-tau and P-tau increase was observed in aDM1 patients compared to jDM1 and controls ($p > 0.05$). In both jDM1 and aDM1 patients, significant correlations were found between $A\beta_{42}$ and T-tau ($\rho = +0.81$ and $\rho = +0.67$, respectively, $p < 0.01$), as well as between $A\beta_{42}$ and P-tau ($\rho = +0.87$ and $\rho = +0.67$, respectively, $p < 0.01$). $A\beta_{42}$ /P-tau ratio decreased with age in aDM1 ($\rho = -0.30$, $p < 0.05$). Only level of $A\beta_{42}$ in CSF of jDM1 patients correlated with size of the CTG expansion ($\rho = -0.53$, $p < 0.05$). Only few correlations were observed between levels of biomarkers and neuropsychological testing.

Conclusion: Further studies with larger cohorts of DM1 patients are necessary. Identification of adequate biomarkers of brain involvement in DM1 is of great importance since central outcome measures are necessary for upcoming gene therapy clinical trials.

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Abstract – WCN 2013

No: 839

Topic: 7 – Neuromuscular disorders

Peripheral nerve safety measures in a multicenter, double-blind, randomized, placebo-controlled, dose-ranging study of fulranumab in patients with painful diabetic neuropathy

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Background: Chronic anti-nerve growth factor activity may adversely affect nerve function. To assess progressive neuropathy risk, intra-epidermal nerve fiber density (IENFD), nerve conduction velocity (NCV) and quantitative neurologic exam (Total Neuropathy Score-nurse, TNSn) were evaluated in a double-blind, placebo-controlled, phase-2 trial of fulranumab's efficacy and safety, in patients with diabetic neuropathic pain.

Objective: To evaluate the safety of fulranumab on IENFD and peripheral nerve function.

Patients and methods: Patients were randomized (3:2:2:3) to placebo or fulranumab (1-, 3-, or 10-mg; every 4 weeks subcutaneously).

Results: At the health authority's request, studies exploring anti-NGF therapy, were terminated early; only 77 patients were randomized. The efficacy and general tolerability of fulranumab from this study were previously reported. Change from baseline (95% CI) for IENFD and NCV was evaluated at screening, and weeks 13 and 53; for TNSn, every 4 weeks. There were no significant decrements in IENFD or NCV, and associated response amplitudes in any of the four nerves evaluated (ulnar and median sensory nerves; ulnar and peroneal motor nerves). There were also no significant deficits in TNSn composite measures or in its subcomponents (sensory, motor and autonomic symptoms; distal-to-proximal changes in pin and vibration sensibility). The analgesia observed in several patients, as determined by change in pain scores, was not associated with decrements in either IENFD or electrophysiology.

Conclusions: There is no evidence that, at the doses and for the period studied, fulranumab was associated with decreased nerve function. An adequately powered and longer duration study is required to confirm these observations.

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Abstract – WCN 2013

No: 1026

Topic: 7 – Neuromuscular disorders

Evaluation of Cost Of Illness (COI) and health care burden in Spinal Muscular Atrophies (SMA)

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Background: SMAs represent a group of fatal inherited disorders leading to progressive disablement with reduced working capacity and high health care utilization. Infantile forms significantly reduce life expectancy. There is no cure but new promising therapies are emerging. Clinical trials in patients are underway.

Objective: Development of these therapies is costly and implementation in SMA care may produce high expenditures. Despite their own cost, the potential contribution of innovative therapies could reduce the COI. In order to forecast the balance from a health economic perspective, our study aimed at assessing the current COI of the most relevant SMA subgroups.

Methods: Patients were questioned using an established German SMA patient registry. In order to determine the COI, a micro-costing method was used to examine the direct and indirect costs measuring the economic burden of SMA on patients, relatives and society.

Results: 300 patients comprising SMA subtypes 1 to 3 aged <0 to 65 years were included. In the light of current standard care the economic burden of patients, relatives and their families was analyzed. Relevant socio-political implications have been identified and compiled.

Conclusion: Our results provide the first step towards a systematic health economic assessment of innovative SMA therapies. They are essential for further cost utility analyses. Particularly, these results enable to speed up the preparation of payer negotiations with regard to pricing and reimbursement. In this context our results contribute to the facilitation of a smooth translation of innovative SMA therapies from their discovery to their implementation into standard care.

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Abstract – WCN 2013

No: 1054

Topic: 7 – Neuromuscular disorders

Understanding the pathomechanisms of inherited peripheral neuropathies

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Most genes for Charcot-Marie-Tooth (CMT) were identified through positional cloning or via a candidate gene approach. Different CMT phenotypes can be caused by mutations in the same gene, and conversely mutations in different genes may result in the same phenotype. This is further complicated by the fact that some mutations are private and occur in specific subtypes. Mutations in >20 genes cause primary alterations of the myelin sheath. Mutations in genes with a function in the axon however, result in axonal CMT phenotypes. Other mutations have been reported to cause intermediate CMT, with both myelin and axonal phenotypes. Mutations were also found in genes (amino-acyl tRNA synthetases, small heat shock proteins, and enzymes involved in lipid metabolism), where the resulting gene products have housekeeping functions and pleiotropic activities. Therefore, these genes were not the obvious candidates for peripheral nerve degeneration and it remains an enigma why the mutant proteins cause specific length-dependent degeneration of peripheral nerves. To identify peripheral nerve specific molecular pathways we will pinpoint differential protein-protein interaction networks starting from the ubiquitously expressed genes. The identification of interacting molecular partners and higher-order molecular complexes they form part of will provide novel insights in regulatory pathways in health and disease, and contribute to new candidate genes for CMT. Altogether, this will ultimately result in the identification of CMT gene networks which can provide insights in finding molecular targets for therapeutic intervention, not only for one type of CMT, but also for the more rare and/or complex phenotypes.

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Abstract – WCN 2013

No: 1019

Topic: 7 – Neuromuscular disorders

Which electrophysiologic parameter does correlate best with the neurologic severity of Bell's palsy?

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Background: Electrophysiologic tests have been used for evaluating facial nerve function in the patients with Bell's palsy. However, the associations between the electrophysiologic parameters and the severity of deficits have not been evaluated systematically.

Methods: This study included 37 patients with Bell's palsy of 7–10 days from onset. The subjects were divided into 3 groups according to House-Brackmann scale (HBS): mild (HBS 2), moderate (HBS 3) and severe (HBS 4 and 5). Blink reflex test (BRT), electroneurography (ENoG) and maximal nerve stimulation (MNS) were done in all subjects. We compared the means of parameters of each test in the 3 groups by ANOVA. The linear associations between the groups and all parameter of each 3 test were tested by linear trend contrast and quadratic trend contrast.

Results: In BRT, the linear association between the parameters and HB scale was found in latency of ipsilateral R1 response ($p = 0.003$), interside difference (ISD) of latency of R1 responses ($p = 0.008$), ISD of amplitude of ipsilateral R2 responses ($p = 0.001$) and contralateral R2 responses ($p = 0.001$). In ENoG, the linear association was seen only in amplitude ($p = 0.006$). However, all parameters including latency ($p < 0.001$), amplitude ($p = 0.01$), ISDs of latency ($p = 0.002$) and amplitude ($p = 0.002$) were significantly correlated with HB scale in MNS.

Conclusions: This study suggests that MNS is the best electrophysiologic test for the correlation with the severity of deficits in Bell's palsy of 7 and 10 days from onset. The parameter of ISD of amplitude of ipsilateral and contralateral R2 responses in BRT, amplitude in ENoG and latency in MNS were most significantly associated with the neurologic severity in each tests for Bell's palsy.

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Abstract – WCN 2013

No: 959

Topic: 7 – Neuromuscular disorders

Preoperative pulmonary function in relation to myasthenic crisis after thymectomy

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Background: Postoperative neuromuscular respiratory failure in patients who underwent thymectomy for myasthenia gravis (MG) may be associated with the preoperative pulmonary function (PPF).

Objective: To investigate whether the difference in PPF levels in MG patients who were successfully weaned from a ventilator after thymectomy was associated with myasthenic crisis after thymectomy (MCAT).

Methods: Seventy-one patients were retrospectively enrolled. We explored the relationship of PPF levels with the existence of MCAT occurring several weeks or months after surgery. The factors of PPF were as follows:

- measured FEV1, mFEV1,
- measured FVC, mFVC,
- mFEV1/mFVC,
- mFEV1/predicted FEV1 (pFEV1),
- mFVC/predicted FVC (pFVC).

PPF receiver operating characteristic (ROC) analysis to predict the occurrence of MCAT was assessed.

Results: MCAT occurred in 17 (23.9%) of MG patients after surgery. mFEV1, mFVC, mFEV1/pFEV1, and mFVC/pFVC were significantly correlated with the occurrence of MCAT, but, mFEV1/mFVC was not. ROC curve area for mFVC/pFVC was 0.902 (95% CI 0.808–0.960, $p < 0.0001$), with sensitivity, specificity, and negative predictive value (NPV) of 94.1%, 77.8%, and 97.7%, respectively, at a threshold of $< 80\%$ with 53.3% of 30 patients below this score. During 2 year follow-up, 97.6% of 41 patients with preoperative mFVC/pFVC $\geq 80\%$ had not experienced MCAT.

Conclusion: The preoperative mFVC/pFVC has high sensitivity and NPV for the prediction of MCAT. Therefore, before deciding on thymectomy in patients with MG, the preoperative measurement of PPF levels and stabilization of the unstable MG symptoms should be required for preventing MCAT.

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Abstract – WCN 2013

No: 805

Topic: 7 – Neuromuscular disorders

Distal hereditary motor neuropathy associated with multiple cranial nerve palsy

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Background: Distal hereditary motor neuropathy (dHMN) is a pure motor axonal neuropathy with autosomal dominant, recessive and X linked pattern of inheritance. The main symptoms of the disease are progressive distal muscle weaknesses and atrophy that affect firstly the hands and the legs. Additional signs like vocal paresis, pyramidal signs, facial weakness and diaphragm weakness can be observed. Here we report a Malian family with dHMN that started with cranial nerves palsy.

Case report: A 24-year-old Malian man born from consanguineous parents was referred for the evaluation of a slowly progressive disorder that started five years ago. The disorder was characterized by a facial weakness with bilateral eye closing weakness, dysphonia, dysarthria and dysphagia. He also presented a tongue atrophy, a weakness of the lower jaw muscle that prevented the complete closing of the mouth. Objectively, there is a distal muscular atrophy affecting the legs that led to a bilateral steppage when he walks. Pes cavus has also been observed. Later, weakness and muscular atrophy were noticed in the hands, particularly at the thenar muscle. Electrophysiological examination showed signs of axonal neuropathy.

Conclusion: Distal hereditary motor neuropathy (dHMN) associated with cranial nerve palsy is rarely reported in the literature. Among the cranial nerve palsy that has been associated with the disease no report concerns the motor branch of the V cranial nerve. Further studies must confirm recessive inheritance and contribute to the research of the gene involved in this particular phenotype.

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Abstract – WCN 2013

No: 1176

Topic: 7 – Neuromuscular disorders

Unique MRI brain imaging findings in patient with bulbar onset amyotrophic lateral sclerosis. A case report

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Introduction: Amyotrophic lateral sclerosis is considered as a progressive neurodegenerative illness that causes muscle weakness, disability, and eventually death, with a median survival of 3 to 5 years. Our case is about a young Pakistani male whose clinical presentation, electrophysiological and imaging studies were suggestive of amyotrophic lateral sclerosis.

Case description: This case is about a 36 year old Pakistani male who was working as a driver in Dubai for the last 10 years. He presented to the clinic with the history of mild dysarthria started around 6 month back without any other complaint followed by change in the quality of voice and having nasal twang. Now for the last one month he was feeling weakness in arms and legs.

On examination patient was conscious and alert but had severe nasal twang and gag reflex was absent. His tongue was full of fasciculation. His muscle bulk was reduced all over and there was obvious fasciculation on both arms and legs. Reflexes were brisk and plantar was up going bilaterally. On MRI brain, T2-weighted sequences showed symmetric bilateral hyper intensities along large myelinated pyramidal tract fibers in the posterior limb of the internal capsule and cerebral peduncle, extending along the corticospinal tract.

Discussion: Our case illustrates that amyotrophic lateral sclerosis can start with only bulbar weakness without any other system involvement and simple MRI brain can show finding suggestive of ALS at the start of disease as in our case.

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Abstract – WCN 2013

No: 1040

Topic: 7 – Neuromuscular disorders

Posterior interosseous nerve injury caused by occluded ganglion

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Background: Posterior interosseous nerve (PIN) injury can be diagnosed by correlating features typical to it. However, in some cases, dependence in these features may cause one to overlook important factors. Also, many overemphasize on differential diagnosis with lateral epicondylitis, which may lead to a narrow view in diagnosis and a resulting overlook in other causes of pressure increases that may cause PIN injury.

Case: A 69-year old right handed male shoemaker came complaining of weakness at his right wrist. He had been carrying shoes with the wrist flexed and pronated for several years, which has caused increased pressure inside the radial tunnel. Physical examination revealed decreased wrist and finger extension strength without any sensory changes. Plain radiography was normal, as well as the ultrasonographic evaluation for extensor tendinopathy. Electromyographic evaluations revealed abnormal spontaneous activities in the EIP, EDC, and APL muscles. Combination of clinical presentation and medical evaluations suggested PIN lesion, which correlated with the type of work activity. Physical therapy modalities were applied but symptoms were only aggravated. Finally, an MRI evaluation revealed a 13×6 mm sized ganglion at the supinator area with denervation myopathy of supinator and extensor muscles. Excision of ganglion resulted in improvement of symptoms.

Conclusion: Ganglion may arise where there is inflammation and damage of tendon and ligamentous structures. As with our case, symptoms of PIN lesions related to occupational activity can be caused by ganglion. MRI may be helpful in finding masses, though ultrasound may also be useful if held in mind during evaluation.

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Abstract — WCN 2013**No: 1099****Topic: 7 — Neuromuscular disorders****Brain and muscle independent involvement in myotonic dystrophy**

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Objectives: The aim of this study is to compare the degree of involvement in brain and skeletal muscle in a series of 25 molecularly-defined DM1 patients since there are few reports done in the same patients.

Methods: Twenty five DM1 patients were recruited for our study. Age at study, age at disease onset and disease duration were recorded. CTG(n) expansion was done in genomic DNA. Neuromuscular assessment in DM1 patients was performed by Muscular Impairment Rating Scale (MIRS). A morphometric study of muscle biopsies included the quantitative evaluation of fibre atrophy/hypertrophy factor of both fibre types. MRI imaging was done for White Matter Hyperintense Lesion (WMHL) and Tractography, MRI was also evaluated.

Results: Muscle morphometric analysis showed an increased atrophy factor (AF), AF was increased especially for type I fibres. Most patients had abnormal MRI imaging, showing scattered supratentorial, bilateral, symmetrical focal or diffuse WMHLs, with a typical temporo-insular diffuse subcortical pattern.

Patients harbouring larger CTG expansion had a more severe muscle impairment at clinical and histopathological level. We found no significant correlation between atrophy factors and MRI total lesion load of WMHL.

Conclusion: We conclude that greater expansion size is a risk factor for more extensive cerebral and muscle impairment, however we found that muscle and brain are independently involved possibly in relation to a different molecular mechanism involving either Muscle Blind-Like Protein 1 or 2 sequestration in respectively muscle and brain tissue.

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Abstract — WCN 2013**No: 980****Topic: 7 — Neuromuscular disorders****Quantitative analysis of dysferlin expression in peripheral blood mononuclear cells by flow cytometry as a screening tool for dysferlinopathies**

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Background: Recent advances in the diagnosis of dysferlinopathies have allowed dysferlin detection in peripheral blood mononuclear cells

(PBMC) by flow cytometry. However, there are only few studies with limited sample sizes using this new technique in which, the endogenous levels of dysferlin in healthy monocytes were not determined. Finding such parameter is a prerequisite for using flow cytometry as a diagnostic tool in dysferlinopathies.

Objective: To evaluate that the mean fluorescence intensity (MFI) for dysferlin in PBMC is a useful diagnostic marker for the detection of dysferlinopathies.

Material and methods: Blood samples were obtained from 54 healthy individuals and 17 patients clinically diagnosed with dysferlinopathy. PBMC were isolated by Ficoll-Histopaque centrifugation and immunolabeled using anti-dysferlin and anti-CD14-PerCP-Cy5.5 monoclonal antibodies. Samples were run on a FACSCalibur flow cytometer and analyzed using FlowJo software. The relative quantity of dysferlin was expressed as MFI and its best cut-off point was determined using the ROC curve.

Results: The dysferlin MFI best cut-off point was 111 fluorescence units with a 94.1% of sensitivity (only 5.9% false negative cases) and 75.9% of specificity (only 24.1% false positives cases). The positive predictive value was 55.1% and the negative predictive value was 97.6%, with an accuracy of 80.2% and a test prevalence of 23.9%.

Conclusion: The MFI cut-off point is a good marker to differentiate samples from patients and healthy individuals with an acceptable margin of error. Therefore, our study confirms the usefulness of this assay for screening and diagnosis of dysferlinopathies.

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Abstract — WCN 2013**No: 1094****Topic: 7 — Neuromuscular disorders****False positive diagnosis of amyotrophic lateral sclerosis: A two-year retrospective cohort study in turin**

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Objective: To verify how many cases of suspected ALS referred to the Turin ALS Center were confirmed as such or diagnosed differently.

Results: A total of 390 patients referred to our ALS Center from January 1, 2011, to December 31, 2012: 296 were diagnosed as definite or probable ALS and 27 as non-ALS MNDs. 67 cases were reclassified as having other disorders, the most common being cervical myelopathy and myopathies. The 296 patients meeting the eligibility criteria included 124 women and 172 men. The mean age at onset was 63.1 years (men 62.4; women 64.2). The presentation was spinal in 194 cases (65.5%) and bulbar in 99 (33.5%). The 27 patients meeting a diagnosis of non-ALS MNDs included 13 women and 14 men; the mean age at onset was 54.7 years (men 58.0, women 53.3). The 67 patients affected by other diseases included 42 men and 25 women; the mean age at onset was 53.7 years (men 51.8, women 56.9). The most frequent diagnosis made in men was motor neuropathy and myopathies while in women was myopathies and multiple sclerosis.

Male/female distribution showed no significant difference among the three groups ($p = 0.61$). Patients not confirmed as affected by SLA were significantly younger and showed generalized asthenia, atypical symptoms, symmetrical disturbances ($p < 0.05$) as presentation symptoms.

Conclusions: The diagnosis of ALS remains a matter of exclusion. The highest frequency of misdiagnosis was in younger men (26.8%; $p < 0.05$) and the most common confounding symptom was weakness of lower limb muscles (23.4%).

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Abstract – WCN 2013**No: 1101****Topic: 7 – Neuromuscular disorders****Isolated acute bilateral ophthalmoplegia as a form of anti-GQ1b syndrome - a case report and differential diagnostic considerations**

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Background: Isolated acute bilateral ophthalmoplegia is an uncommon occurrence, but it may cause differential diagnostic problems. It may be a manifestation of the Miller Fisher syndrome spectrum, which has been recently referred to as anti-GQ1b syndrome.

Objective: To present a case with acute bilateral ophthalmoplegia associated with anti-GQ1b antibody positivity, but without other typical signs of Miller Fisher syndrome (MFS). Differential diagnostic aspects are highlighted.

Patient: A 57-year-old female patient presented with an acute onset of diplopia, progressing to bilateral ophthalmoplegia within days. These symptoms were preceded by flu-like symptoms. Deep tendon reflexes were diminished, otherwise her neurological status was unremarkable, the typical triad of MFS was not observed. MRI of the brain was normal. Cerebrospinal fluid examination revealed increased protein content with normal cell count. Electrophysiological assessment showed normal sensory nerve action potentials, whereas low amplitude or absent sensory nerve action potentials are characteristic findings in MFS. Serological testing confirmed anti-GQ1b class IgG antibody positivity, supporting the diagnosis of MFS spectrum. The patient fully recovered within 3 months, without any specific treatment, showing the benign nature of the condition.

Discussion: Acute ophthalmoparesis may be a restricted form anti-GQ1b syndrome. In addition to classical MFS, further variants include Bickerstaff's brainstem encephalitis, pharyngeal-cervical-brachial paresis, and acute ataxic sensory neuropathy. Although acute ophthalmoparesis as a manifestation of anti-GQ1b syndrome is a benign condition, it may cause differential diagnostic problems with potentially more serious conditions needing prompt treatment, such as Wernicke's encephalopathy, ocular myasthenia, botulism, paraneoplastic brainstem encephalitis, or brainstem stroke.

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Abstract – WCN 2013**No: 1413****Topic: 7 – Neuromuscular disorders****“Reporting biomarker” development: Update in als patients treated with G-CSF -mobilized hematopoietic stem cells**

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Objective: Treatment development in neurodegeneration is demanding because of time slots for evaluating effects and multiple confounders. We selected prolonged open label “compassionate use” autologous BM

stem cell mobilization in ALS-patients to pre-validate biomarkers for disease modulation and safety evaluation.

Methods: 20ALS patients were treated with s.c. rec-hu-G-CSF, in a conventional 5/28 days or a 1/7 days outpatient regimen: 5 to 10 µg/kg BW were given daily plus Riluzole. Patients with a median age of 48 yrs. were evaluated by ALS-FRS-R each 4 wks. As reporting biomarkers we assessed

- (1) hypothenar muscle motor unit number estimates (MUNE, McComas), and
- (2) cranial MRI-DTI to delineate FAI-changes as sign of axonal damage/repair in motor cortex and pyramidal tracts,
- (3) BM function including stem cell differentiation profile, smears and blood counts, Burst forming and colony forming units.

Safety included abdominal sonography, determination of BW, pulmonary function, clinical chemistry.

Results: Both application modes were safe with no obvious difference in efficacy. Side effects were very mild, tolerance was good. No differences were detected when compared to an ALS population treated with Riluzole only. Clinical outcome revealed some longer stabilisations, unrelated to age or disease dynamics. Median overall survival in application mode **5/28** was 2.9 years. MUNE correlated to disease progression ($p < 0.01$), and DTI-FAI ($p < 0.003$), and showed increases in individual patients. DTI-/ FAI-values over time indicated decrease in most patients, in some minor improvement.

Discussion: Prolonged treatment with G-CSF is feasible and safe in ALS patients - prospective study data are needed. MUNE, DTI and BM function parameters are probably very useful biomarkers.

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Abstract – WCN 2013**No: 1229****Topic: 7 – Neuromuscular disorders****A missense mutation in the mouse TDP-43 gene leads to a gain of TDP-43 mediated splicing function: Implications for neurodegeneration**

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TDP-43 is a highly conserved member of the family of Heterogenous Nuclear Ribonuclear Proteins (hnRNPs). It is associated with neurodegeneration, constituting the major component found in neuronal cytoplasmic inclusion bodies from patients who suffered from Amyotrophic Lateral Sclerosis (ALS) and Fronto-Temporal Lobar Degeneration with Ubiquitinated Inclusions (FTLD-U/FTLD-TDP). Moreover, dominant missense mutations in *TARDBP*, the human gene encoding TDP-43, mostly located in its C-terminal, are causative of ALS and FTLD-U. Both the ectopic expression of human wild type TDP-43 and mutant alleles associated with ALS/FTLD-U have been reported to lead to neurotoxicity in different model organisms, including mice. This dose dependent toxicity of wild type and mutant TDP-43 alleles makes the mechanistic dissection of mutant toxicity challenging. Here, we characterize a novel mouse TDP-43 mutant line, carrying an ENU-induced point mutation in the mouse endogenous *Tardbp* gene. Using a variety of methodologies, including

RT-PCR, Western Blotting, Electrophoretic Mobility Assay and primary cell culture, we report that the TDP-43 M323K mutation (Methionine to Lysine at residue 323 of TDP-43) leads to perinatal lethality in homozygosity and to a “gain of normal splicing function” of TDP-43 in a dose dependent manner, which is not associated with increased protein levels or changes in its subcellular localisation. These results shed light on the effects of C-terminal mutation in TDP-43 and have potential implications for its role in ALS/FTLD-U.

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Abstract – WCN 2013

No: 1416

Topic: 7 – Neuromuscular disorders

Duchenne muscular dystrophy and epilepsy – rare comorbidity: Case report

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Background: Duchenne muscular dystrophy (DMD) is a form of muscular dystrophy that has been reported as an X-linked recessive disease caused by a defective dystrophin gene, with a rapidly deteriorating course. Association between DMD with central nervous system dysfunction in the form of mental retardation are well recognized, but correlation between DMD and epilepsy is reported in only a few studies.

Objective: We present a rare comorbidity of DMD and epilepsy.

Patients and methods: We report a 3.5-year-old boy with DMD diagnosis and generalized tonic-clonic seizures. He had no mental retardation or cognitive decline. He had febrile seizures in the first two years of life. His family history was insignificant.

Results: DMD was confirmed by polymerase chain reaction findings of Xp21. EEG showed generalized paroxysms of spikes and spike wave discharges. Repeated electroencephalography still had specific features, but without further seizure recurrence, that underlines the importance of careful electroencephalography evaluation in order to achieve earlier and better seizure control.

Conclusion: Although epilepsy and DMD has been described before as a rare comorbidity, with a total of only a twenty cases reported by different authors, there is evident increase in the prevalence of epilepsy in these dystrophin-deficient patients. So far, according to our best knowledge, this is the first case described in Serbia and countries around.

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Abstract – WCN 2013

No: 1412

Topic: 7 – Neuromuscular disorders

Idiopathic phrenic neuropathies: A case series and literature review

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Background: Phrenic neuropathies are important, and probably underdiagnosed cause of ortopnea, dyspnea and hypercapnic respiratory failure. However, the aetiology, optimal treatment and long term prognosis of “idiopathic” phrenic neuropathies are not known.

Objective: To try establish possible aetiology and long term prognosis of “idiopathic” phrenic neuropathies.

Patients and methods: Patients with “idiopathic” phrenic neuropathies were identified from this authors practice and by the literature search. Their clinical characteristics, treatments and long term outcomes were noted. Compatibility with neuralgic amyotrophy

diagnosis was established (typical arm motor nerve lesion findings – definite; trigger event or pain – probable).

Results: Unilateral/bilateral phrenic neuropathy was diagnosed in 10/9 in the authors series and in 18/40 patients from the literature search. In unilateral/bilateral groups 69% and 76% were men ($p > 0.05$), were $54 \pm 12/56 \pm 11$ years old ($p > 0.05$), 11%/22% had diabetes ($p > 0.05$), and 4%/31% hypertension ($p < 0.05$). Criteria of definite/probable neuralgic amyotrophy were fulfilled by 7%/61% of patients with unilateral, and in 20%/43% of patients with bilateral phrenic neuropathy ($p > 0.05$). Non-invasive ventilation was used by 22%, IVIg by 5%, steroids by 4% and valacyclovir by 4% of patients. Complete resolution occurred in 23%/14% ($p > 0.05$), and partial improvement in 18%/40% ($p < 0.05$) of patients with unilateral/bilateral phrenic neuropathy.

Conclusion: About two-thirds of patients with phrenic neuropathies fulfilled some criteria of neuralgic amyotrophy, and about one-half improved at least partially. Only few patients received adequate immunomodulatory and supportive therapy, partially also due to often too late diagnosis.

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Abstract – WCN 2013

No: 1449

Topic: 7 – Neuromuscular disorders

Possible toxicity of tuberculostatic agents in a patient with a novel TYMP mutation leading to mitochondrial neurogastrointestinal encephalomyopathy

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Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE) is a rare autosomal recessive multisystemic disorder caused by TYMP gene mutations. Here, we report on the first MNGIE patient diagnosed in Bulgaria who carries a novel homozygous TYMP mutation (p.Leu347Pro). The patient presented with gastrointestinal complaints, cachexia, hearing loss, ptosis, ophthalmoparesis, polyneuropathy, cognitive impairment and leukoencephalopathy on MRI examination of the brain. The patient's motor capacity declined significantly leading to wheelchair dependence several months following the administration of tuberculostatic treatment suggesting mitochondrial toxicity of these agents. The advanced stage of the disease and the poor medical condition prevented us from performing allogenic hematopoietic stem cell transplantation (HSCT). Early diagnosis is important not only for genetic counseling but also in view of the timely treatment with allogenic HSCT.

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Abstract – WCN 2013

No: 1465

Topic: 7 – Neuromuscular disorders

Stiff man syndrome associated with breast cancer about 2 cases

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Background: The stiff-man syndrome (SMS) is one of the syndromes of hyperactivity neuromuscular, it may be associated with neoplasias in paraneoplastic syndromes. We report 2 cases of stiff man syndrome.

Observation: Our patients were aged 54 years and 25 years, she had no particular history and had axial stiffness and rigidity with abnormal posture. Spasms were present.

The rest of the neurological examination was normal, no pyramidal signs, extrapyramidal disorders, sensory or cognitive disabilities.

MRI brain and spine were normal. Electromyography showed continuous motor activity of axial muscles. CSF was normal in our two patients. Anti-GAD could not be made. The physical examination showed a swelling of the breast with axillary lymph nodes, which led to a puncture and cyto-pathologic analysis revealing carcinomas. The antibodies anti-amphiphysin could not be measured due to lack of resources.

We observed an improvement in the clinical condition diazepam, baclofen and chemotherapy.

Conclusion: The stiff man syndrome associated with breast cancer is described in the literature, the diagnosis of this syndrome is not always obvious to the clinician and requires research associated pathologies.

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Abstract – WCN 2013

No: 1362

Topic: 7 – Neuromuscular disorders

Effect of carpal tunnel syndrome on ulnar nerve at wrist: Ultrasonographic and electrodiagnostic studies

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Introduction: Carpal tunnel is located close to Guyon's canal and increased pressure of carpal tunnel may affect ulnar nerve on Guyon's canal. The aim of study is to analyze the findings of electrophysiologic and ultrasonographic study in ulnar nerve in patient with carpal tunnel syndrome (CTS) and to verify the effect of CTS on ulnar nerve at wrist.

Methods: 12 hands of healthy volunteers (60.25 + 14.67 years) and 14 hands of patients with CTS (58.50 + 12.69) were enrolled. Electrophysiologic study of ulnar nerve was done in all participants. Cross sectional area (CSA) of median nerve in carpal tunnel and ulnar nerve in Guyon's canal was evaluated by ultrasonography. The findings of electrodiagnostic studies and CSA of ulnar nerve were compared in patients with CTS and control group. Correlation of CSA in median nerve and ulnar nerve was evaluated in patients with CTS.

Results: CSA of median nerve in ultrasonography of patients with CTS (15.62 + 6.47) was significantly large compared to control group (8.02 + 0.86). CSA of ulnar nerve in ultrasonography of patients with CTS and control group was 4.57 + 1.05 and 3.68 + 0.33, respectively. There was significant difference between these two groups ($p = 0.018$). Correlation of CSA of median nerve and ulnar nerve in patient with CTS was not observed, significantly. ($p = 0.088$). No other parameter in electrophysiologic study has significant difference between patients with CTS and control group.

Conclusions: Increased pressure leading to CTS may also affect ulnar nerve in Guyon's canal. Ultrasonography can be used as an assessment tool for the pressure effect of CTS on ulnar nerve.

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Abstract – WCN 2013

No: 1480

Topic: 7 – Neuromuscular disorders

Neuromuscular sarcoidosis: A retrospective study of 12 cases

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Background: Neuromuscular sarcoidosis accounts for 5–15% of cases of systemic sarcoidosis.

Objective: Determine the clinical characteristics of neuromuscular sarcoidosis.

Patients and method: We enrolled 12 patients with pathologically proven sarcoidosis with neuromuscular symptoms, who were referred to our hospital since 2006. We retrospectively reviewed their clinical records.

Results: Of the 12 patients, 4 had cervical myelopathy (3 intramedullary, 1 extramedullary), 4 had meningeal involvement (2 leptomeningitis, 2 pachymeningitis), 2 had neuropathy (1 spinal mononeuropathy multiplex, 1 recurrent hemifacial palsy), and 2 had chronic myopathy. Bilateral hilar lymphadenopathy and history of the uveitis were observed in 10 and 4 cases, respectively. None of the patients had cardiac involvement. Three had been already been diagnosed pathologically as having sarcoidosis before neuromuscular involvement. Three of the 4 patients with cervical myelopathy were diagnosed by transbronchial lung biopsy, whereas 3 of the 4 cases with meningeal involvement were diagnosed by meningeal biopsy. The mean level of serum angiotensin converting enzyme (ACE) was 23.8 ± 12.9 IU/L, and was not related to disease severity in the myelopathy and meningeal subgroups.

Conclusion: The neuromuscular presentations of neuromuscular sarcoidosis are varied and must be differentiated from other neurological disorders. The diagnosis of "isolated" neuromuscular sarcoidosis is quite difficult and is neglected because the biopsy is difficult to perform. In addition, not all of the neurological symptoms of pathologically proven sarcoidosis might be due to sarcoidosis. It is necessary, therefore, to develop new surrogate markers of the disease, instead pathological examination, for diagnostic purposes.

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Abstract - WCN 2013

No: 1395

Topic: 7 - Neuromuscular disorders

MR1 muscle fat quantification in ambulant patients with limb girdle muscular dystrophy 2A and correlation with clinical severity

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Background: Muscle MRI provides information on the distribution and evolution of limb girdle muscular dystrophies (LGMD). In the last years quantitative sequences like 3-point Dixon technique have been developed to measure muscle fat fraction (MFF).

Objective: To quantify MFF in lower limb muscles of ambulant LGMD A patients and to determine its correlation with functional scales.

Material and methods: Cross-sectional study of 10 ambulant patients with molecularly confirmed LGMD2A. Modified Gardner–Medwin–Walton and Vignos scales were assessed to determine clinical severity. A 3-point Dixon sequence (TE 2.3–4.6–6.9 ms, FOV 375, matrix 376/384, thickness: 2 mm, acquisition time: 5.10 min) was performed in a 1.5T Achieva Philips scanner. MFF was calculated drawing a region of interest covering cross-sectional area in three different slices per muscle (ten muscles in thigh, eight in calves). Correlations between clinical scales and MFF were determined with Spearman's rho coefficient.

Results: Mean age was 24.20 ± 10.58 years with 12 ± 6.67 years of disease duration. Mean clinical severity was 5.2 ± 2.3 and 4.2 ± 1.99

in Gardner–Medwin–Walton and Vignos scales. A significant correlation ($p < 0.05$) was detected between the mean MFF of all muscles and Gardner–Medwin–Walton ($r = 0.84$) and Vignos ($r = 0.60$) scale. This correlation was also found separately in the muscles of thigh (anterior $r = 0.83$, posterior $r = 0.75$) and calves (anterior $r = 0.65$, posterior $r = 0.82$).

Conclusion: Muscle fat content measured with 3-point Dixon MRI provides a high correlation with clinical severity of ambulant LGMD2A patients and could be a useful biomarker.

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Abstract - WCN 2013

No: 1233

Topic: 7 - Neuromuscular disorders

Time from onset to treatment and prognosis in patients with CIDP: A 3-year follow-up of 29 cases

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Background: Time from onset to treatment may be important for CIDP prognosis, although this variable has not been systematically assessed.

Objective: To investigate the association between time from onset to treatment and prognosis in CIDP patients.

Patients and methods: We retrospectively enrolled 29 consecutive CIDP patients [15 males, median age, 58 years (range, 15–82 years)] who were followed-up for 3 years after initial treatment. Twenty-seven patients fulfilled the EFNS/PNS definite criteria. Two patients categorized to probable CIDP were diagnosed using sural nerve biopsy. The primary outcome was good prognosis, which was defined as stable disease activity ≥ 2 years and Hughes grade of 0–1 at 3 years. Clinical characteristics, time from onset to treatment, and Hughes grade at initial treatment were evaluated as predictors of good prognosis.

Results: Patients were first treated with immunoglobulin infusion (57.1%), corticosteroids (32.2%), and plasmapheresis/immunoadsorption (10.7%). Twenty-five patients (86.2%) responded to the treatments. Patients showed monophasic (17.2%), relapsing (51.7%), and chronic (31.1%) clinical courses. Twelve patients (41.4%) showed good prognosis. Six of 8 (75.0%) patients treated < 8 weeks and 6 of 14 (42.9%) patients treated from 8 weeks to 1 year after onset showed good prognosis, whereas none of 7 patients treated ≥ 1 year after onset demonstrated good prognosis (Fisher's exact test, $P = 0.01$). Hughes grade at initial treatment was not significantly associated with good prognosis (Fisher's exact test, $P = 0.67$).

Conclusion: Time from onset to treatment was suggested to be important for CIDP prognosis, regardless of the disease severity at initial treatment.

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Abstract - WCN 2013

No: 1379

Topic: 7 - Neuromuscular disorders

Rare case of leprosy, where initial presentation is carpal tunnel syndrome

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33-year-old woman from village area of country, had history of right arm pain with tingling like sensation, especially aggravated in night time. Clinically suspected carpal tunnel syndrome confirmed with nerve conduction study. Apart patient did not have any other symptoms related to skin or other nerve involvement. 9 months post decompression surgery patient had developed hypo pigmented rash on forearm and leg, as well as trophic ulcer in hands. With no

apparent exposure to leprosy, clinical and histological evidence of tuberculoid leprosy was found. A particular characteristic of this case was leprous neuritis involving the median nerve which was diagnosed clinically as carpal tunnel syndrome, and scheduled for surgical treatment. Under treatment with dapsone and rifampicin, however, the condition cleared up completely.

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Abstract - WCN 2013

No: 1370

Topic: 7 - Neuromuscular disorders

Cholinergic neuromuscular hypersensitivity in musk antibody positive myasthenia gravis

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Background: The patients with MuSK antibody (MuSK-Ab) positive myasthenia gravis (MG) show distinct responses to acetylcholinesterase inhibitor (AChEI), such as less effective therapeutic response to AChEI and more frequent nicotinic side effects and negative result of diagnostic AChEI test. Some MuSK-Ab positive MG patients experience overt worsening after AChEI treatment. In addition, the compound muscle action potential with extradischarges (CMAP-EDs), electrophysiologic feature of cholinergic neuromuscular hyperactivity, may develop in the MuSK-Ab positive MG patients with usual therapeutic dose of AChEI.

Objective: We investigated the clinical and electrophysiologic features of cholinergic neuromuscular hypersensitivity in MuSK-Ab positive MG patients.

Patients and methods: We retrospectively reviewed the medical records and electrodiagnostic findings of seventeen MG patients (MuSK-Ab positive: 10, MuSK-Ab negative: 7) who underwent electrodiagnostic test before and after neostigmine test.

Results: The MuSK-Ab positive patients had higher frequency of intolerance to oral pyridostigmine bromide (50 vs 0%, $p = 0.044$) and lower maximal dose of oral pyridostigmine (90 vs 480 mg/day, $p = 0.023$) than the MuSK-Ab negative. The frequency of positive results of neostigmine test was significantly lower in the MuSK-Ab positive patient than in the MuSK-Ab negative (100 vs 40%, $p = 0.035$). The nicotinic side effects of neostigmine were more frequent in the MuSK-Ab positive patients (80 vs 14.3%, $p = 0.015$). The CMAP-EDs were more frequently developed after neostigmine injection in the MuSK-Ab positive patients than the MuSK-Ab negative (90 vs 14.3%, $p = 0.004$).

Conclusion: Cholinergic neuromuscular hypersensitivity is a distinct characteristic of MuSK-Ab positive MG.

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Abstract - WCN 2013

No: 1324

Topic: 7 - Neuromuscular disorders

Effect of Alpha-Lipoic Acid on the postural stability of patients with diabetic peripheral neuropathy

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Background: Diabetic peripheral neuropathy (DPN) is a common complication of diabetes mellitus, that affects the postural stability.

There are researches about the effect of Alpha-Lipoic Acid (ALA) on the reduction of pain and improvement of neuropathic deficits.

Objective: This study evaluated the effect of treatment with ALA on the postural stability in patients with type 2 DPN.

Patients and methods: Sixty patients and 20 healthy age-matched subjects took part in this investigation. All patients had a good glycemic control. The two schemes of treatment were applied: the first – with 600 mg ALA (5 day infusion and 60 day oral dose), and the second – ALA, benfotiamin, pyridoxine and cyanocobalamin together. The postural stability was evaluated using static posturography under two visual conditions (eyes open and eyes closed) on stable and soft surfaces. The investigations were made on the first, 5th and 60th day after the drug therapy.

Results: Before the treatment with ALA the all posturographic parameters, for both patient groups were significantly higher than in healthy subjects. After treatment the sway velocity and sway path decreased. The most pronounced decrease was observed in sway path during stance on foam support with closed eyes. The changes of the posturographic results after the combined therapy were better than with ALA only.

Conclusion: Treatment with combined therapy (ALA, benfotiamin, pyridoxine and cyanocobalamin together) showed stabilizing effect on the quiet upright stance, that leads to improvement of the quality of life of patients with type 2 DPN.

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Abstract - WCN 2013

No: 1355

Topic: 7 - Neuromuscular disorders

Classification of sporadic lower motor neuron disease in Jeju Island

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Objectives: The sporadic forms of lower motor neuron disease (LMND) constitute a heterogeneous group of diseases with largely unknown pathogenesis. Clinical, genetic, electrophysiological, and immunological findings can help to distinguish patients with LMND who never develop amyotrophic lateral sclerosis (ALS) from patients with typical ALS.

Methods: We studied the clinical and electrophysiological features of 23 patients with sporadic LMND in a cross-sectional study. Disease duration was more than 4 years to exclude the majority of patients with ALS. Based on the pattern of weakness, we classified patients into three groups: with generalized weakness (group 1); with non-generalized asymmetrical weakness of the arms (group 2); with non-generalized asymmetrical weakness of the legs (group 3).

Results: We identified two patients in group 1, eighteen patients in group 2, and three patients in group 3. Distinctive features of group 1 were an older age at onset, more severe weakness and muscle atrophy and more widespread abnormalities on needle EMG. In groups 2 and 3, age at onset was younger than patients in group 1. However, needle EMG findings also suggested a more widespread disease process. Retrospectively, the prognosis of sporadic LMND seems to be relatively good after a median disease duration of 8 years.

Conclusion: The clinical phenotypes of the different subgroups described in this study may help to differentiate the several LMND forms from each other. However, prospective studies are needed to investigate whether specific clinical or pathogenic variables may help to identify patients with a more benign form of LMND.

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Abstract - WCN 2013

No: 1114

Topic: 7 - Neuromuscular disorders

A rare case of an African–American male with AOA2

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Background: Ataxia with oculomotor apraxia type 2 (AOA2) is an autosomal recessive cerebellar ataxia caused by mutation of the *setx1* gene (SETX) on chromosome 9q34 and characterized by cerebellar atrophy, axonal sensorimotor neuropathy, oculomotor apraxia, and elevated serum alpha-fetoprotein (AFP).

Objective: We describe the case of a 32 year African–American male with classic clinical features of AOA2 with positive SETX gene mutation.

Methods: At birth the patient was delivered without complications and had normal development. At 7–8 years of age he developed difficulties with coordination and became wheelchair bound by age of 12. Since then he has been suffering from progressive cognitive impairment, polyneuropathy and hypercholesterolemia. However, family history is unclear. Neurological examination is significant for moderate cognitive impairment, dysarthria, dysconjugate gaze, jerk nystagmus and alternating strabismus. Motor exam revealed normal tone, reduced bulk in all the extremities and reduced strengths in bilateral lower extremities. Dysmetria on limb movements and marked ataxia were noted. Reflexes were absent in all the extremities.

Results: NCS revealed progressive sensorimotor axonal polyneuropathy and EMG showed evidence of acute & chronic denervation. MRI revealed marked cerebellar atrophy and degenerative changes in the cervical spine. Genetic testing was performed for AOA2 and Fragile X syndrome was positive for the SETX gene, confirming the diagnosis of AOA2.

Conclusion: AOA2 is a rare condition frequently seen in Europe, North Africa and West Indies. Most of the cases encountered in the US are of French descent. Our case is unusual in that the patient was of African–American origin.

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Abstract - WCN 2013

No: 1313

Topic: 7 - Neuromuscular disorders

Nerve ultrasound score in distinguishing chronic inflammatory demyelinating polyneuropathy from Guillain–Barré syndrome

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Background: GBS is defined as an acute-monophasic polyradiculoneuropathy, while CIDP may also present (16% of the cases) with a subacute-monophasic course. Their differential diagnosis is of great importance, as their therapies differ significantly.

Objective: The aim of this study was the development and evaluation of a new ultrasound score to distinguish these two nosologies.

Materials/methods:

Phase-1 (development):

20 GBS (mean-age 58.3y, SD ± 13.8; 10 women), 20 CIDP-patients (mean-age 56.7y, SD ± 13.5; 6 women), 75 healthy-subjects (mean-age 53.5y, SD ± 14.8; 30 women) underwent nerve-ultrasound, a mean

of 3.4 years (SD \pm 2.91) and 4.55 years (SD \pm 3.5) respectively, after onset. Using the acquired ultrasound data we developed a “Bochum-ultrasound-score” (BUS) summarizing:

- 1) the cross sectional area (CSA) of the ulnar nerve in Guyon's canal,
- 2) CSA of the ulnar nerve in upper arm,
- 3) CSA of the radial nerve in spiral groove,
- 4) CSA of the sural nerve between gastrocnemius muscle.

The patient received 1 point for every anatomic site of the “BUS”, where he showed pathological CSA enlargement (min = 0, max = 4).

Phase-2 (evaluation):

Further 21 GBS (mean-age 54.2y, SD \pm 12.6; 12 women), 10 CIDP-patients (mean-age 55.1, SD \pm 9.9; 3 women) underwent blinded for the diagnosis examination with the “BUS”, a mean of 2.3 years (SD \pm 1.5) and 3.8 years (SD \pm 2.7) respectively, after onset.

Results: The “BUS” showed a sensitivity of 90%, specificity of 90, 4% (PPV81, 8%, NPV = 95%) in distinguishing CIDP from GBS.

Highlight: 6 patients with subacute polyradiculoneuropathy (mean-age 52, 6, SD \pm 16, 47; 2 women), who presented in year 2012 to our department, underwent blinded for the diagnosis examination with the “BUS” a mean of 2.8 weeks (SD \pm 2.1) after onset. The “BUS” showed a sensitivity of 75%, specificity of 100% (PPV = 100%, NPV = 66%) in distinguishing subacute-CIDP from GBS.

Conclusion: The “BUS” seems to allow a reliable distinction of CIDP from GBS.

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Abstract - WCN 2013

No: 1635

Topic: 7 - Neuromuscular disorders

Plasma cortisol levels in amyotrophic lateral sclerosis

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Background: Amyotrophic lateral sclerosis (ALS) is associated with a significant distress, being possibly linked to changes in hypothalamic–pituitary–adrenal (HPA) axis activity or other stress–response systems. An early study reported a loss of cortisol circadian rhythmicity in ALS patients (Patacchioli et al, 2003), while more recently it has been shown a blunted cortisol awakening response, which correlated with the clinical status and depressive mood (Rozenaal et al, 2012).

Objective: To assay the circadian plasma cortisol levels in ALS and to study their relationship with the clinical phenotype and rate of disease progression.

Patients and methods: 122 ALS patients (Bulbar, 30; Spinal, 92; M/F = 1.73) and 103 controls (not affected by neurological or psychiatric disorders, free of drugs; M/F = 1.75) were recruited. Disease progression was scored with Δ FS. Plasma cortisol level (ug/dl) was assayed from fasting patients and controls at 8:00 and 20:00 using Elecsys® Cortisol Immunoassay System. Data were analyzed with the Mann–Whitney U test or, where appropriate, with the Kruskal–Wallis test.

Results: We found higher levels of morning cortisol in ALS patients than controls (morning: ALS, 15.2 [11.5–18.9] vs Controls, 11.4 [8.8–14.3], $p < 0.001$; evening: ALS, 7.5 [4.7–11.8] vs Controls, 7.9 [5.4–10.0], $p = 0.6$). The hormone's morning level was higher in the spinal-onset group (Spinal, 15.9 [11.9–19.0] vs Bulbar, 13.5 [10.1–18.6] vs controls, 11.4 [8.8–14.3], $p < 0.001$) and in patients with intermediate/rapid disease course.

Conclusions: Morning plasma cortisol level is increased in ALS and correlates with site of onset and rate of disease progression. Thus, a subgroup of ALS patients may have an altered HPA axis activity.

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Abstract - WCN 2013

No: 1613

Topic: 7 - Neuromuscular disorders

Lateral spread responses on facial motor nucleus suppression using intravenous diazepam in patients with hemifacial spasm

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Introduction: There has been a controversy between two hypotheses for the pathophysiology of hemifacial spasm: peripheral theory and central theory are two candidate. This study sought to investigate the pathophysiologic mechanism of hemifacial spasm.

Materials and methods: A total of 6 patients with hemifacial spasm were recruited. In experimental study, supraorbital nerve stimulation with orbicularis oris muscle recording study and lateral spread test were performed, then we applied intravenous diazepam 10 mg to the patients for facial motor neuronal suppression. Two ‘drug series’ were recorded, 10 min and 20 min after the subject had received a 10 mg diazepam intravenously.

Result: In all patients, orbicularis oris responses were appeared on supraorbital nerve stimulation with orbicularis oris muscle recording study and late responses were appeared on lateral spread test. After diazepam injection, on supraorbital nerve stimulation with orbicularis oris muscle recording study, latencies of orbicularis oris response were showed slowing tendency as time passes in the patients. On lateral spread test, however, in all patient, latencies of direct and late responses were consistent as time passes.

Discussion: In our present study, we inhibited facial motor nucleus using diazepam that is CNS depressant. If the site of abnormal cross transmission is in the facial nucleus, lateral spread response should be recorded with a wave form of delayed latency as our supraorbital nerve stimulation with orbicularis oris muscle recording study. The results suggest peripheral pathway, ectopic excitation–ephaptic transmission as the pathophysiologic mechanisms in hemifacial spasm.

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Abstract - WCN 2013

No: 1608

Topic: 7 - Neuromuscular disorders

Guillain–Barré syndrome in HIV infection – A case report

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Background: The acute inflammatory demyelinating polyradiculoneuropathy or the Guillain–Barré syndrome (GBS) was rarely described in HIV-infected patients. Recent studies suggest that the treatment with immunoglobulin (IVIg) not only improves the neurologic symptoms, but in addition to ART therapy, it also reduces the HIV reservoir.

Objective: We report the case of a 21-year-old man, with generalized motor deficit that interrupted his ART therapy 3 months ago. Five days before admission, he noted paresthesias in his limbs, followed by pain and progressive weakness.

Patients and methods: On neurological examination, he had 1/5 strength in the lower extremities, 3/5 strength in the upper extremities, diminished tendon reflexes, and decreased sensation in the lower limbs and dysphagia. Examination of the CSF revealed increased protein level and pleocytosis. Electromyographic findings suggested an acute demyelinating process with conduction block. A diagnosis of GBS was made and the patient received treatment with a 5-day course of 0.4 g/kg/d IVIG. He refused the ART therapy.

Results: After the treatment, the neurological status improved (3/5 strength in lower extremities, 4/5 in upper extremities, swallowing problems remitted). Interestingly, his CD4 and CD8 cell counts increased and the plasma viral load decreased.

Conclusion: IVIG has several effects on the immune system but the mechanism of action in HIV infection is unknown; recent studies suggest its utility as adjuvant therapy in HIV infection. The present case brings further evidence in favor of this hypothesis. Furthermore, the patient's immune status improved and the viral load decreased without any ART therapy.

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Abstract - WCN 2013

No: 1649

Topic: 7 - Neuromuscular disorders

Mutations in anoctamin 5 in limb girdle muscular dystrophy in Norway: Phenotypic variability and mutation spectrum

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Background: Recently, recessive mutations in the ANO5 gene have been identified as a major cause in limb girdle muscular dystrophy (LGMD) and distal muscular dystrophy with high CK-levels.

Objective: We aimed to investigate the prevalence of ANO5-mutations in Norway, and present clinical and biopsy findings of those with confirmed ANO5-mutation.

Material and methods: In the current study 53 Norwegian probands (31 males, 22 females) with myopathy of unknown etiology and high CK levels were tested for ANO5 mutations. Retrospectively, patients with confirmed ANO5-mutations were thoroughly investigated clinically, MRI of the lower limbs, muscle biopsies and EMG-findings were analyzed.

Results: In nine families, 11 patients with biallelic sequence variants in ANO5 were identified. Four of these patients were from North Norway and hence minimum prevalence in North Norway is estimated to 1/100,000. Seven patients were either homozygous or compound heterozygous for the most commonly found truncating mutation c.191dupA in exon 5. Mutations in ANO5 were exclusively found in male LGMD patients. The clinical symptoms were slowly progressive and affected mainly the lower limbs without any respiratory or cardiac affection. MRI of the lower limbs showed pathologic findings even in mildly affected patients. In EMG some patients only exhibited neurogenic changes.

Conclusion: LGMD2L, caused by mutations in the ANO5 gene, is a common cause of LGMD and might be the second or third most common cause behind LGMD2I and LGMD2A in Norway. LGMD2L

predominantly affects the lower limbs and muscle and MRI seems to be a valuable tool to detect muscle pathology.

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Abstract - WCN 2013

No: 1640

Topic: 7 - Neuromuscular disorders

Autosomal dominant Brown-Vialetto-Van Laere syndrome with UBQLN1 mutation

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Background: Brown-Vialetto-Van Laere syndrome (BVVLS) is a rare MND characterized by childhood onset, bulbo-pontine paralysis and neurosensory deafness. Recent genetic studies have demonstrated mutations in c20orf54 riboflavin encoding gene in most familial cases, recessive mutations on Chr. 8q24.3 in a Lebanese family and the UBQLN1 gene mutation E54D in a sporadic BVVLS patient.

Objective: To report the phenotypic and genotypic characteristics of an Italian family with autosomal dominant BVVLS.

Patients and methods: The family studied has two clinically-documented cases. The proband is a 15-year-old girl affected by BVVLS since she was 3. She is now tetraplegic and tracheostomized. The second case is the proband's maternal aunt. She was diagnosed with BVVLS at 20, but deafness and bulbar weakness started during adolescence. She died at 27 because of respiratory insufficiency.

For genetic testing, we obtained an informed consent from the proband's father. DNA was extracted from blood and the screening for c20orf54 and UBQLN1 genes was performed. Coding sequences of both genes were amplified and directly sequenced.

Results: A heterozygous substitution E54D in exon 1 of the UBQLN1 gene was found in the proband. No mutations in the c20orf54 gene were detected. Screening for UBQLN1 and c20orf54 gene mutations in available asymptomatic living relatives (the father and the brother) was negative.

Conclusions: In this study we describe the first family with an autosomal dominant form of BVVLS linked to a UBQLN1 mutation. The pattern of inheritance is autosomal dominant with incomplete penetrance and with variability of age of onset.

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Abstract - WCN 2013

No: 1173

Topic: 7 - Neuromuscular disorders

A report of two cases of Guillian Barre syndrome variants post dengue infection

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Background: Various viral infections have been known to precede the development of Guillian Barre Syndrome (GBS). Cytomegalovirus and Epstein Barr virus are commonly implicated. Influenza, parainfluenza, adenovirus, herpes simplex and varicella zoster virus have been rarely implicated. Post dengue GBS is an extremely rare neurologic complication of dengue with only few cases reported in literature.

Objective: To report two cases of post dengue GBS and to analyse their clinical presentation, electrophysiologic studies and the outcome.

Patients and methods: Both patients presented with limb weakness and their clinical examination was consistent with hyporeflexic/areflexic quadriparesis.

Patient 1 had history of ongoing fever of 4 day duration with thrombocytopenia and mild hepatitis. His dengue PCR was positive. His weakness recovered fully spontaneously.

Patient 2 had history of dengue infection a week ago with dengue IgM positive. She also had autonomic instability and bilateral facial weakness. She was treated with IVIG. She had partial recovery with residual weakness at discharge.

Results: Patient 1 had an acute inflammatory demyelinating polyneuropathy (AIDP/conventional GBS) with good clinical recovery.

Patient 2 had an acute motor-sensory axonal polyneuropathy (AMSAN variant of GBS) with partial recovery.

Conclusion: GBS is rare but known neurologic complication of dengue virus infection. It can present as different clinical variants of GBS. GBS should be considered if any patient with recent or ongoing dengue infection presents with weakness. Prognosis can be variable depending upon the pattern of nerve injury.

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Abstract - WCN 2013

No: 1680

Topic: 7 - Neuromuscular disorders

Parameters settings of NIV: Can they predict functional and survival outcome in ALS patients?

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Background: The lack of more specific tools, with low costs, which may be associated with the ALSFRS score to assist in analyzing the prognosis, is a constraint factor in the follow-up of ALS ventilated patients.

Objective: We analyzed potential predictors that can be related to rate of functional decline and survival measured by ALSFRS in ALS patients and disease duration.

Patients and methods: Prospective, comparative trial of 60 consecutive ALS patients (G1 = 29 dead, G2 = 31 alive), compliant to Non-Invasive Ventilation (NIV), followed up within the same period from January 2008 to May 2012. All patients were followed-up with ALSFRS, oximetry, Respiratory Function Test (RFT) and blood gases analysis once every three months. Primary outcomes included ALSFRS functional decline and disease duration; secondary outcomes included time of NIV, oximetry data and parameters settings at NIV adaptation.

Results: No clinical or laboratorial differences were observed between groups (dead or alive) for any variable at admission. Disease duration (in days) from symptoms was higher in G1 ($p = 0.46$), as well as duration to NIV ($p = 0.12$), but not significantly. These 2 variables correlate positively with maximal inspiratory pressure, IPAP and backup breathing rate. Multivariate Cox regression analysis showed that several parameters settings of NIV and data from RFT were associated with rate of functional decline in these patients.

Conclusions: For the first time, determinants of functional decline are significantly related to parameter settings of NIV equipment as well as to compliance data.

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Abstract - WCN 2013

No: 1548

Topic: 7 - Neuromuscular disorders

Cognitive and psychological profile in Tunisian children with Duchenne muscular dystrophy

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Background: In Duchenne muscular dystrophy (DMD), mutation of the dystrophin gene has long been recognized as a cause of intellectual impairment. However, underlying etiopathological mechanisms remain unclear.

Objective: To analyze cognitive and psychological profiles in Tunisian children with DMD and their correlation with causal mutations.

Patients and methods: Over 8 years (2005–2013), 18 boys were followed up in our department for a genetically confirmed diagnosis of DMD. Neuropsychological evaluation including full-scale intelligence quotient (IQ), memory assessment, verbal performances, attention processes, executive functions, mood and behavioral patterns. Correlation between dystrophin gene mutation and neuropsychological profile was analyzed.

Results: Mean age of patient was 11.16 years. Causal mutation predicted to lead to a loss of the Dp140 isoform has been found in 13 patients. Mean age at neuropsychological evaluation was 6.9 years. General intelligence assessments showed a mean IQ of 82 (range 50–110). Impairment in working memory was noted in 10 patients. Verbal performances and attention processes were altered in respectively 5 and 3 patients. Executive dysfunction was noted in 3 patients. Cognitive deficits were correlated to the loss of Dp 140 isoform in 9 patients out of 13. There was no evidence of cognitive declining with the progression of muscular deterioration.

Conclusion: Our results support emerging evidence of central nervous system involvement resulting in neuropsychological disorders in DMD. The loss of Dp 140 seems to be involved in the pathological mechanism underlying cognitive impairment.

Neuropsychological deficits should be systematically detected in DMD in order to improve patient's quality of life.

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Abstract - WCN 2013

No: 1512

Topic: 7 - Neuromuscular disorders

Extracorporeal photochemotherapy in treatment of myasthenia

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Aim: Estimation of extracorporeal photochemotherapy (EPCT) efficacy in myasthenic patients.

Material and methods: A total of 31 patients with generalized myasthenia were treated beyond exacerbation period. Muscular strength was evaluated according to 5-score scale. EPCT method was as follows: patients underwent interrupted leukapheresis procedure, which yielded, out of 1 L of patient blood, approximately 200–250 ml of leukocyte-enriched blood containing, on the average, $2.43\text{--}3.6 \times 10^9$ mononuclear cells. Prior to reinfusion, leukocyte suspension was UV radiated. Treatment course included 4 expositions (2 times a week with two expositions each time). Each patient received photosensitizing drug before each exposition.

Lymphocyte subpopulation, interferon status, and immunoglobulins were analyzed.

Results: Improvement was noted in 27 of 31 patients (87%). Statistically significant increase of strength was registered in mimic, neck and biceps muscles, musculus deltoideus, musculus gastrocnemius, and musculus iliopsoas ($p < 0.05$). Clinical effect was retained for 3–4 months. There is a 2-year-follow-up of a patient, admitted to the hospital with bulbar and oculomotor disturbances. During further regular treatment, bulbar disturbances disappeared, and mild eye double-seeing was cured after the first exposition. Lymphocyte subpopulations in general decreased, immune regulatory index CD4/CD8 normalized. Immunoglobulin levels decreased, mainly IgA (by 18%), and less-IgG (by 8,3%) and IgM (by 2%). Decrease of interferon status mean values was statistically insignificant.

Conclusion: Extracorporeal photochemotherapy is an effective and prospective method of myasthenia therapy. EPCT is an alternative method of medicamentous treatment of myasthenia, not requiring glucocorticoids or cytostatic drugs.

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Abstract - WCN 2013

No: 370

Topic: 7 - Neuromuscular disorders

Macrophages involvement in neurodegenerative process in motor neuron disease

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Amyotrophic lateral sclerosis (ALS) is incurable neurodegenerative disease characterized by death of upper and lower motor neurons. Neuroinflammation is pathological marker of many neurodegenerative diseases including ALS and is typified by activation and proliferation of microglia and infiltration of T cells into brain and spinal cord. Damaged motor neurons are able to activate complement system microglia and astrocytes, which can contribute to neurodegeneration. Activated microglia and astrocytes are found in close proximity to dying motor neurons. Their activation status and proliferation seemingly increases with disease progression. Aim of our research was to investigate pathologic changes of brain stem (BS) in ALS, we analyzed midsagittal sections of BS from 24 individuals with ALS and 29 controls by using conventional staining and immunohistochemistry with antibodies against CD68 and GFAP. Also we studied macrophagal phagocytosis in 67 patients with different ALS forms. Control group comprised from 122 healthy donors of same gender, age groups. The number of CD68-immunoreactive macrophages/microglia and GFAP-immunoreactive astrocytes were significantly higher in individuals with ALS than in controls in all areas of the BS except rostrum. Among the patients with ALS, the number of macrophages/microglia and astrocytes was significantly higher in isthmus than in rostrum. Multiple immunofluorescence labeling of selected biomarkers revealed different microglial phenotypes during culturing. These findings suggest that pathologic changes in the BS in ALS are present in posterior midbody and isthmus, where callosal motor fibers may traverse between two hemispheres. CD68 and GFAP immunohistochemistry are sensitive methods to detect those pathologic changes in routine paraffin-embedded specimens.

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Abstract - WCN 2013

No: 1763

Topic: 7 - Neuromuscular disorders

Symptoms and complaints of the patients with dm1 at the early stage

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Myotonic dystrophy type 1 (DM1) is an autosomal dominant disorder with dysfunction of multiple systems. Concurrent features of skeletal muscles, myotonia and weakness, do not make difficulties with DM1 patients in early and mild stage, and progress slowly. Consequently, the diagnosis of DM1 at the early stage is difficult and tends to delay. We analyzed main symptoms at onset and chief complaints at the first visit to clinic from the medical records of 67 patients with DM1 (41 males, 26 females). Mean age of their onset was 27.2 ± 14.1 . The main symptoms at onset were: weakness, 40; myotonia, 17; mental retardation, 8; dysarthria, 1; lumbago, 1. Mean interval from onset to their first visit to neurological clinics about DM1 was 11.6 ± 9.9 years. The chief complaints at the first visit to clinics were: weakness, 44; myotonia, 7; dysarthria, 2; lumbago, 2; genetic counseling, 2; apathy, 1; syncope, 1; no complaint, 8. Among 17 patients onset with myotonia, only 7 were with also myotonia as the chief complaint at the first visit to clinic. Among 40 patients onset with weakness, 33 were with weakness as the chief complaint at the first visit to clinic. Myotonia is a characteristic and important sign for DM1, which provides early diagnosis. In contrast, myotonia is less likely to be a reason for consultation to physician, suggesting that patients with DM1 are suffering more with weakness than with myotonia.

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Abstract - WCN 2013

No: 1752

Topic: 7 - Neuromuscular disorders

Clinical and neuroimaging features of familial C9FTD/ALS: A case report

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Background: The expansion of an intronic hexanucleotide repeat in chromosome 9 is the most frequent cause of ALS/FTD.

Objective: To describe the clinical and neuroimaging features of a patient with familial C9FTD/ALS.

Patients and methods: We describe a 45 year old patient with a family history of ALS and FTD (affected father and paternal aunt), in whom the C9ORF72 hexanucleotide expansion was detected.

Results: At 26 she started with progressive right hand weakness, followed by weakness of the upper left and, subsequently, lower

right limb. Inability to walk and bulbar symptoms are present since the age of 40. Additionally, personality changes, delusions and progressive cognitive decline started about 12 years after the onset of motor manifestations. Currently she has severe cognitive impairment with a delusional disorder and confabulations. Motor examination revealed tongue amyotrophy and fasciculations, dysarthria, tetraplegia, upper limb hyporeflexia and lower limb hyperreflexia, as well as marked generalized muscle wasting and diffuse, active denervation in the EMG. While the severe cortical atrophy – especially affecting the left temporal lobe – observed in 3-tesla MRI, was congruent with the predominantly left temporal hypometabolism showed through brain PET scan, the PET–MRI fusion image demonstrated a pattern of generalized cortical hypometabolism.

Conclusion: We present the detailed phenotypic characterization of a patient with *C9ORF72*-associated ALS/FTD. We highlight the potential contribution of PET–MRI fusion image to the understanding of this disorder, since it allows correcting the metabolic activity for the degree of cortical atrophy observed with MRI.

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Abstract - WCN 2013

No: 1714

Topic: 7 - Neuromuscular disorders

Anoctamin 5 myopathy: More patients, more phenotypes

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Background: Anoctamin 5 myopathies are autosomal recessive disorders resulting from mutations in the *ANO5* gene.

Objective: To present the expanding spectrum of the disease.

Patients and methods: We present here a cohort of patients diagnosed in Paris since 2011, when genetic testing became available in France. Patients tested include known but unexplained cases of muscle dystrophy and new patients diagnosed as possible anoctamin 5 myopathies on clinical grounds.

Results: 31 patients, stemming from 26 families, display two mutations of the *ANO5* gene, including 24 men and 7 women, the latter being less severely affected in a majority of cases. Mean age of onset was 32 years (range 10–50 years). Clinical pattern was LGMD2L in 22%, Miyoshi-type distal myopathy (MMD3) in 22%, proximo-distal weakness in 4%, exercise intolerance in 22%, isolated high-CKs in 16% of cases. Two further patients presented with a pseudo-Becker dystrophy and 2 female patients developed a severe, calpain-like limb-girdle myopathy. 6 patients presented with cardiac abnormalities on echocardiography, leading to medical treatment in 3 cases. CK levels were usually very high, ranging from 3.5 to 125 times the normal value. Muscle imaging showed a constant involvement of medial gastrocnemii. Most patients harboured heterozygous private mutations, except for the common c.191dupA duplication.

Conclusion: Anoctamin 5 myopathy appears as a major cause of muscle dystrophy, sharing similarities with dysferlinopathies. However, later onset, slower course and the presence of dysferlin on muscle immunostainings distinguish anoctamin 5 myopathies, as well as some particular phenotypes and mild heart involvement.

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Abstract - WCN 2013

No: 1744

Topic: 7 - Neuromuscular disorders

Is tracheostomy still an option in amyotrophic lateral sclerosis? Reflections of a multidisciplinary work group

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Question under study: Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease with a poor prognosis. Survival and quality of life of ALS patients have improved through the implementation of multidisciplinary approaches, the use of percutaneous gastrostomy and of non-invasive (NIV) or invasive ventilation. The question of whether or not to propose invasive ventilation (by tracheostomy: TPPV) to ALS patients remains a matter of debate.

Methods: The present study reviews the medical literature, the practice in Swiss and two large French ALS expert centres, and reports the results of a work group on invasive ventilation in ALS.

Results: Improved management of secretions and using different interfaces allows using NIV 24 h a day for prolonged periods and thus avoiding TPPV in many cases. TPPV is frequently initiated in emergency situations with lack of prior informed consent. TPPV appears associated with a lesser quality of life (QoL) and a higher risk of institutionalization than NIV. The high burden placed on caregivers who manage ALS patients is a major problem with a clear impact on their QoL.

Conclusions: Current practice in Switzerland and France tends to discourage the use of TPPV in ALS. Fear of a “locked-in syndrome”, high burden placed on caregivers, unmasking cognitive disorders occurring late in the evolution of ALS are some of the caveats when considering TPPV. Most decisions of TPPV are taken in emergency situations in the absence of advanced directives. One exception is that of young motivated patients with predominantly bulbar disease who “fail” NIV.

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No: 1656

Topic: 7 - Neuromuscular disorders

Spinal cord atrophy correlates with disease severity in amyotrophic lateral sclerosis

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Background: Biomarkers are needed to help in the diagnosis and long term care of patients with ALS. SC damage is a pathological hallmark of the disease, but there are few studies that investigated quantitative MRI of the SC as a biomarker in ALS.

Objective: To investigate spinal cord (SC) atrophy in Amyotrophic lateral sclerosis (ALS), and to determine whether it correlates with clinical parameters.

Methods: Forty-three patients with ALS (25 men) and 43 age-and-gender-matched healthy controls underwent MRI on a 3T scanner. We used T1-weighted 3D images covering the whole brain and the cervical SC to estimate cervical SC area and eccentricity at C2/C3 level based on a semi-automatic image segmentation protocol using a validated software (*Spineseg*). Acquisition parameters: TE = 3.2 ms, TR = 7.1 ms, flip angle = 8°, voxel size = 1.0 mm³ and FOV = 240 × 240. Disease severity was quantified with the *ALS functional rating scale*. SC areas of patients and controls were compared with Mann-Whitney test. We used linear regression to investigate the association of SC area and clinical parameters.

Results: Mean age of patients and disease duration were 53.1 ± 12.2 years and 34.0 ± 29.8 months, respectively. The two groups were significantly different regarding SC areas ($67.8 \pm 6.8 \text{ mm}^2$ vs $59.5 \pm 8.4 \text{ mm}^2$, $p < 0.001$). However, eccentricity values were similar in both groups ($p = 0.394$). SC areas presented a significant correlation with disease duration ($r = -0.585$, $p < 0.001$) and ALS functional rating scale ($r = 0.318$, $p = 0.037$).

Conclusions: Patients with ALS have SC atrophy, but no flattening and SC areas did correlate with disease duration and functional status.

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Abstract - WCN 2013

No: 1708

Topic: 7 - Neuromuscular disorders

Ultrasonography does not increase sensitivity of the carpal tunnel syndrome electrodiagnosis

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Background: Median nerve entrapment at the wrist (i.e., the carpal tunnel syndrome—CTS), is the most common neuropathy. In spite of its high sensitivity, electrodiagnostic (EDx) studies are negative in some patients with typical and bothersome CTS symptoms (i.e., paresthesias in at least 2 of the first 4 fingers pronounced during the night or in the morning).

Objective: To report utility of ultrasonographic (US) examination in this patient population.

Patients and methods: Three groups were studied: (1) patients with typical CTS symptoms and mild EDx abnormalities; (2) patients with typical CTS symptoms, but negative EDx studies; (3) asymptomatic volunteers. The cross-sectional area (CSA) of the median nerve at the pisiform bone and 1 cm proximal (the wrist) and 10 cm proximal (the forearm) were measured by a high-resolution US. The criteria for CTS diagnosis were the median nerve CSA at the wrist $> 10 \text{ mm}^2$, and wrist-to-forearm CSA ratio (WFR) > 1.5 .

Results: The median nerve CSA at the wrist was increased in 91% of 58 symptomatic patients with positive EDx, in 38% of 136 symptomatic patients with negative EDx, and in 35% of 48 asymptomatic controls. WFR was increased in 97%, 50%, and 48% of subjects, respectively. No statistical difference in CSA was found between patients with negative EDx and asymptomatic volunteers ($p = 0.46$).

Conclusion: US is not useful in patients with typical CTS symptoms, but negative EDx studies. EDx or US is equally useful to confirm CTS diagnosis.

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Abstract - WCN 2013

No: 1857

Topic: 7 - Neuromuscular disorders

Recurrent Guillain–Barre syndrome presenting with stereotypical manifestations, positive anti-ganglioside antibodies and prompt recovery

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Backgrounds: Compared to monophasic Guillain–Barre Syndrome (GBS), recurrent GBS (rGBS) is very rare and has been considered as a different entity. Here, we report incidence, clinical features, and laboratory findings of rGBS from single big university-based hospital in Korea.

Methods: We reviewed medical records and electrophysiological reports of GBS patients, from 2004 to 2010, in Inje University Busan Paik Hospital. By utilizing anti-ganglioside antibodies assay and nerve conduction study, subtypes of GBS were classified. Criteria of rGBS were adopted by those of previously conducted one study (Kuitwaard et al., 2009).

Results: Of 68 enrolled patients with GBS, three patients (4%) were proved to have more than two definite recurrences of GBS. All the 3 cases showed clinically stereotypical features, positive anti-ganglioside antibodies:

Case 1 showed 2 times of weakness in both arms and positive anti-GM1/anti-GD1b antibodies;

Case 2 presented with 3 times of sensory ataxia/cranial neuropathies and showed positive anti-GQ1b/anti-GD1b antibodies;

Case 3 had an identical distal leg weakness during 3 attacks and positive anti-GM1 antibody.

All the cases were promptly recovered since the nadir of symptoms.

Conclusions: rGBS is rare but a certainly existed entity showing clinically stereotypical manifestations, and remarkably prompt recovery. In a clinical aspect, differential diagnosis of rGBS from other chronic relapsing neuropathies is important in that whether clinicians should maintain immune-modulating therapies or not. Additionally, in a scientific aspect, the understanding of rGBS will be able to promote the clarification of both the precise immunopathogenesis and therapeutic targets.

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Abstract - WCN 2013

No: 1822

Topic: 7 - Neuromuscular disorders

Brain involvement in FKRP-related muscular dystrophy

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Introduction: FKRP mutations cause a muscular dystrophy which may present in the neonatal period (MDC1C) or later life (LGMD2I). Normal intelligence and brain imaging have been previously reported in FKRP-related muscular dystrophy, except in rare cases presenting with mental retardation associated with structural brain abnormalities.

Patients and results: We studied cerebral MRI in three unrelated children, aged 4–6 years, with FKRP-related muscular dystrophy presenting MDC1C and moderate mental disturbances. Brain MRI in the first patient with moderate muscular phenotype showed structural abnormalities of the posterior fossa with hypoplasia of the vermis and pons, multiple cerebellar hemispheric cysts, and abnormal white matter signal in the cerebellum and periventricular region. Moderate unilateral left vermis hypoplasia, few cerebellar cysts, mild pons hypoplasia, and left temporal atrophy were seen in the second case which had mild clinical picture. The last child had normal neuroimaging and severe phenotype.

Conclusion: MRI abnormalities are common in our patients with FKRP-related muscular dystrophy presenting at birth. They show no correlation with clinical features. Posterior fossa malformation may be associated with mental retardation.

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Abstract - WCN 2013**No: 1780****Topic: 7 - Neuromuscular disorders**
Bent spine syndrome due to myofibrillar myopathy

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Background: Camptocormia, as an isolated sign, could be a diagnostic challenge since it might be caused by several central and peripheral nervous system diseases, including Parkinson disease, dystonia, MSA, Alzheimer disease, ALS, CIDP, myasthenia and myopathies.

Objective: To describe a patient with camptocormia due to a myofibrillar myopathy (MFM) that is, to our knowledge, the second case reported in literature.

Patients and methods: A 75 year-old woman presented progressive trunk forward flexion since the age of 72 as her father, who showed same spine symptoms from the age of 50. The patient underwent clinical examination, laboratory exams, EMG, muscle MRI and biopsy.

Results: Clinical examination revealed camptocormia, mild paraspinal muscles weakness, “compensatory” posture with trunk hyperextension, flexion of legs and arms and tendency to touch lightly her thighs to walk more easily, resembling a “*geste antagoniste*”. There were no extrapyramidal signs. CPK level was normal and EMG of paraspinal muscles revealed a myopathic pattern. Muscle MRI showed fatty replacement of the paraspinal muscles. Biopsy of D5–D6 paraspinal muscle showed myopathic features with fiber size variability, internal nuclei, slight increase of connective tissue and some vacuoles; the Gomori trichrome staining revealed areas darkly stained within fibers. Immunohistochemistry studies showed an abnormal accumulation of desmin and α Bcrystallin. Electron microscopy confirmed myofibrillar disruption. A diagnosis of MFM was made.

Conclusion: Bent spine is predominantly described in the later stages of MFM, therefore represents an unusual sign at the disease onset. Among myopathies, MFM should be considered in the differential diagnosis of camptocormia.

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Abstract - WCN 2013**No: 1811****Topic: 7 - Neuromuscular disorders**
Distal myasthenia simulating radial nerve palsy: A case report

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Background: Distal limb weakness is an uncommon manifestation of myasthenia gravis, although it has been described both as a presenting symptom and during the course of the disease.

Objective: To report a patient previously diagnosed as having ocular myasthenia gravis who was attended because of acute distal weakness in his left hand.

Patients and methods: A 74-year-old male with a history of hypertension, cataract surgery and prostate adenocarcinoma was diagnosed as having seropositive ocular myasthenia gravis in November 2009. He subsequently presented bulbar involvement that required treatment with IV immunoglobulins, corticosteroids and pyridostigmine. Since then, he remained asymptomatic on treatment with pyridostigmine and

low doses of prednisone every other day. In September 2012 he developed acute dorsiflexor weakness in his left hand.

Results: Clinically, he had weakness of wrist extension, finger extension and finger abduction in the left hand. All other muscles (extraocular, facial, neck, trunk, upper and lower extremities) as well as the rest of the neurological examination were normal. Three-hertz repetitive stimulation with trains of 10 stimuli of the left radial, ulnar and median nerves demonstrated a significant amplitude decrement. The patient was treated with a gradually increasing daily dose of prednisone, with subsequent improvement of the hand weakness.

Conclusion: Although distal weakness is rare in myasthenia, it is recommended that this possibility should be considered in the differential diagnosis of patients presenting focal hand weakness in the absence of sensory symptoms and signs.

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Abstract - WCN 2013**No: 1024****Topic: 7 - Neuromuscular disorders**
Diagnostic usefulness of routine nerve conduction studies for the patients with carpal tunnel syndrome

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Background: Carpal tunnel syndrome (CTS) is the most common entrapment neuropathy. There are many additional electrophysiologic methods for diagnosis of CTS. We analyze routine nerve conduction studies to improve diagnostic simplification and sensitivity for CTS.

Methods: A total of 70 patients with clinically diagnosed CTS and 207 control subjects were enrolled. All subjects were examined by routine nerve conduction studies. Normal limits were derived by calculating the mean \pm 2 standard deviations from the data of the controls. The sensitivity and specificity of each test were calculated.

Results: The difference between median and ulnar sensory peak latency was the most sensitive parameter. The sensitivity and specificity of the median-ulnar sensory latency difference were 58.6% and 96.6% respectively. The sensitivity of other parameters was 55.7% for median nerve sensory peak latency, 55.7% for the median-ulnar motor terminal latency difference, 54.3% for the modified combined sensory index, and 52.9% for the median-radial sensory peak latency difference.

Conclusions: In our study, some parameters derived from routine nerve conduction studies, especially difference between median and ulnar sensory latency, could be more sensitive for diagnosis of CTS.

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Abstract - WCN 2013**No: 1827****Topic: 7 - Neuromuscular disorders**
Pan-cord brachial plexus injury after the mirpe for the correction of extreme pectus excavatum

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We report a case about brachial plexus injury after MIRPE that was confirmed by electrodiagnostic and imaging study.

Case report: A 23-year-old male patient had MIRPE (minimally invasive repair of pectus excavatum) for extreme pectus excavatum. To correct the deformity, two metal bars were inserted; the first bar was inserted at the sixth intercostal space, and the second bar was

inserted obliquely from the right fourth intercostal space through the left sixth intercostal space. On postoperative day 1, the patient complained weakness, mainly wrist extensor and numbness at whole right upper extremity. Under the impression of right brachial plexus injury, the patient received steroid therapy. 17 days after surgery, electrodiagnostic study showed right brachial plexus injury, involving pan-cord and right long thoracic nerve at clavicle level. MRI findings revealed signal changes of right brachial plexus at costoclavicular level and there was a difference in the space between first rib and clavicle on either side. The patient received physical therapy for 3 months. Strength of his right upper extremity and sensory symptoms was improved but, not fully recovered.

Conclusion: The correction mechanism of the MIRPE, deformed costal cartilages and sternum are lifted and transformed forcibly by the correction bar. So the abrupt elevation of sternum and rib cages resulted in altering space between first rib and clavicle and might have injured the brachial plexus. Brachial plexus injury is extremely rare complication of the MIRPE but it can be developed obviously considering anatomic structural alteration.

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Abstract - WCN 2013

No: 1874

Topic: 7 - Neuromuscular disorders

TITIN-CAP (TCAP) polymorphisms associated with LGMD2G among Indian patients with ARLGMD

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Background: This rare form of ARLGMD was first described from Brazil. It is mapped to chromosome 17q11-12. No reports are available from India.

Objectives: To perform genetic studies in 4 Immunoblot confirmed cases of LGMD2G.

Materials and methods: 300 cases of ARLGMD prospectively studied with phenotypic characterisation, muscle Immunohistochemistry (IHC), Immunoblotting (IB) studies. All the exons of TCAP were sequenced including UTR and flanking regions and sequence analysis performed. 100 age and ethnically matched controls were studied.

Results: Of 280 biopsies, 226 had IHC, 176- IB. Eight from 7 families had complete absence of 19 kDa band. Four belonging to 2 families revealed two pathogenic variations. Clinically: Mean age of onset was 12.38 ± 11.35 (5–40 years). Scapular winging was seen in all. Shoulder girdle and arms predominantly affected. Moderate distal weakness in two. In lower limbs: iliopsoas, gluteus medius, maximus preferentially involved. Hip adductors severely weak. Foot drop in all. Mean CK- 2574.4 ± 2847.5 IU/L (718 to 9253). Genetically: First family had a novel variation in three affected members, c.32C>A (p.Ser11X), resulting in a premature stop codon. Parents and unaffected siblings were heterozygous for same mutation. Second case had a reported homozygous 8 base pair duplication (c.26_33dupAGGTGTCG) resulting in frameshift and truncated protein (p.R12fsX31). All unaffected members were heterozygous for this mutation. These variations not identified in 100 age and ethnically matched controls.

Conclusion: This is the first study of TCAP mutations in this large sample size of ARLGMD from India. We have identified one novel and one reported functionally significant mutation, which can be used as diagnostic markers.

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Abstract - WCN 2013

No: 1482

Topic: 7 - Neuromuscular disorders

Factors affecting ultrasonographic findings of the ulnar nerve in ulnar neuropathy at the elbow

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Introduction: High-resolution ultrasound (HRUS) can demonstrate focal nerve enlargement in entrapment neuropathies. We aim to identify the factors affecting ultrasonographic findings of the ulnar nerve in ulnar neuropathy at the elbow (UNE).

Method: 54 arms of 49 patients (35 male, 14 female, mean age 48.9 years) with clinical and electrodiagnostic evidence of UNE were recruited in this study. 34 arms were electrophysiologically classified as axonal group and 20 arms were classified as demyelinating group. By using HRUS, the arms were also grouped into three categories according to the ulnar nerve movement during elbow flexion;

- 1) non-displacement (28 arms),
- 2) subluxation (11 arms),
- 3) dislocation (15 arms).

We measured cross sectional area (CSA) of the ulnar nerve at maximal swelling point and compared the results among the groups.

Results: The CSA of the ulnar nerve at maximal swelling point was significantly larger in displacement group than normal group; dislocation group 17.2 mm^2 , subluxation group 19.1 mm^2 , non-displacement group 12.6 mm^2 ($p < 0.01$). However, there was no significant difference in CSA of ulnar nerve between the axonal and the demyelinating group; axonal group 14.3 mm^2 , demyelinating group 16.0 mm^2 . There was no significant correlation between duration of symptom and CSA of the ulnar nerve.

Conclusion: As the type of the ulnar nerve displacement (non-displacement vs displacement), the difference of nerve CSA at entrapment neuropathy was existed at our study. Therefore, nerve swelling may be affected by ulnar nerve displacement.

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Abstract - WCN 2013

No: 1895

Topic: 7 - Neuromuscular disorders

Meralgia paresthetica: Topography of the sensory deficit

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Background: Meralgia paresthetica (MP) results from lesions of the lateral cutaneous nerve of the thigh (LCNT). This author observed that the sensory abnormality frequently differs from classical descriptions.

Objective: To evaluate the distribution of sensory abnormality in patients with MP.

Patients and methods: This was a prospective study. MP was considered present when the sensory loss involved at least part of the lateral thigh, there were no motor or reflex abnormalities in the leg, and electromyography of the quadriceps muscle was normal. The classic sensory distribution was considered to involve at least 50% of the length of thigh (iliac crest to upper patella), and did not extend beyond the anterior midline, nor below the lower edge of the patella. Sensory patterns in affected legs were categorized as: classic, bigger medially (beyond anterior midline), bigger inferiorly (below lower edge of patella), bigger both, or restricted (less than 50% of the length of the thigh).

Results: 40 patients (34 males) aged 34–73 years had MP. It was bilateral in 8 patients; total number of legs with MP was 47. Topographic

patterns: Classic 10 (21%); bigger medially 25 (53%), bigger inferiorly 5 (11%), bigger both 3 (6%), restricted 11 (23%).

Conclusions: Sensory loss in MP frequently differs from classic descriptions. The area is often larger, sometimes smaller. Explanations may include anatomic variation and the degree of damage of the LCNT.

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Abstract - WCN 2013

No: 1899

Topic: 7 - Neuromuscular disorders

Electromyography in a small city in Ecuador

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Background: EMG studies are helpful in diagnosis for many neurological diseases. There are not enough reports to take care about the necessities for a General Hospital in any region around the World.

Objective: To know the absolute number of patients who can be assessed by a novel EMG laboratory; correlate the true positive relation between the suspected diagnosis and the final report; try to avoid for the future, the excess of demand.

Results: During 2009 we studied a total of 209 patients, mean age 49 years (SD 17), 56.4% women and 43.5% men. Neurosurgery (34.4%), Rheumatology (22%), Traumatology (15%) and Neurology (12%) were the most frequent senders for the studies. The most common diagnosis included: neuropathic pathology (50.7%) in this item, the entrapment neuropathies were the most common diagnosis (carpal tunnel syndrome 34%); root lesions (27.7%) with compromise of the lower back in 58.6% (L4, L5 and S1 roots); and finally 17% of all were normal. 43% of cases had positive relation between the suspected diagnosis and the final report; 40% did not have a presumed diagnosis.

Conclusions: It is important to know the absolute demand for an EMG laboratory, and it is also necessary to establish an educational program for the medical staff in the sense of a better understanding of this kind of tests, the presumed diagnosis as a crucial tool for the electromyographer and of course for the patient because it is not a simple test and to avoid unnecessary discomfort.

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Abstract - WCN 2013

No: 1911

Topic: 7 - Neuromuscular disorders

Epidemiology of myasthenia gravis in Slovak Republic

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Background: 1785 patients were registered up to December 31, 2012 in the Centre for Myasthenia Gravis (MG) in Slovakia.

Objective: Analysis of MG epidemiological data in Slovakia (1978–2012).

Patients and methods: The diagnosis of MG was based on clinical and electrophysiological findings, positive AChR and MuSK autoantibodies.

Results: The prevalence rate on Dec 31, 2012: 241.8 per 1 million. The average annual incidence rate (2003–2012): 14.8 per 1 million. Sex: 1033 females, 752 males (1.37:1). The median age at MG onset: 54 years, females 48 years, males 62 years. Early-onset MG prior the age of 50 years: 800 patients, 605 females, 195 males (3.1:1). Late-onset MG after the age of 50 years: 985 patients, 428 females, 557 males (1:1.3). Infantile MG prior the age of 15 years: 102 children

(5.7%), 77 girls, 25 boys (3.1:1). AChR seropositive MG was diagnosed in 86%, MuSK positive in 3%, seronegative in 11% of MG patients. Ocular MG: 328 (18.4%), generalised MG: 1457 (81.6%) patients. Thymoma was histologically proven in 158 (8.9%) patients.

Conclusion: The prevalence rate of MG has increased over the past 35 years, because the mortality rates declined and MG patients have longer life span due to significant advances in the treatment. Since the 1980s a gradually increased incidence has been evident, mainly due to late-onset MG, giving a significant shift to older age at onset of MG. These findings are relevant for non-thymoma MG. The epidemiological data of thymoma-associated MG were constant.

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Abstract - WCN 2013

No: 1915

Topic: 7 - Neuromuscular disorders

Guillain-Barré syndrome: subtypes and predictors of outcome from India

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Background: Acute motor axonal variant of Guillain Barre syndrome has been reported to be commoner from South East Asia. The types of GBS however depend on the extent and timing of electrodiagnostic study. There is paucity of large study evaluating the subtypes of GBS and their outcome from South East Asia.

Objective: We report cliniconeurophysiological subtypes of Guillain Barré syndrome (GBS) and their outcome from Northern India.

Patients and methods: 328 GBS patients were categorized into acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor sensory axonal neuropathy (AMSAN), pure sensory (PS) and Miller-Fisher syndrome (MFS) based on nerve conduction study (NCS). The various patterns of GBS correlation with triggering factors, severity of illness, clinical characteristics, and 3 months outcome.

Results: The patients' mean age was 31.2 years. Clinically 204 (62.2%) patients had pure motor, 106 (32.3%) motor sensory, 16 (4.9%) MFS and 2 (0.6%) had pure sensory GBS. Based on NCS, 242 (73.8%) had AIDP, 44 (13.4%) AMAN, 15 (4.6%) AMSAN and 27 (8.2%) had equivocal GBS. AIDP patients were older, lesser disabled at admission and better outcome compared to AMAN. 11 (3.4%) patients died and 48 (14.6%) had poor outcome. The poor outcome was related to severity, dysautonomia and inexcitable motor nerves.

Conclusion: AIDP is the commonest subtype of GBS in Northern India. Patients with severe disability, dysautonomia and inexcitable nerves had poor outcome.

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Abstract - WCN 2013

No: 1409

Topic: 7 - Neuromuscular disorders

Development of service for patients with Motor Neuron Disease (MND) in Singapore

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Background: Observations made in clinical practice suggest that people living with MND need a comprehensive set of services that can meet their bio-psychosocial-spiritual needs. However, there is no known

literature that can inform the development of services appropriate for patients with MND living in Singapore, a multicultural and family-centered society.

Objective/method: Drawing from findings of our study that explored the needs of patients with MND and observations made in clinical practice, this presentation will attempt to describe services that can meet the needs of patients with MND in Singapore.

Results: Research findings and clinical observations point to the need to develop proactive services that are sensitive to the needs of patients and their caregivers.

It was found that “staging” the disease will better meet the needs of the patients as they progress along the disease trajectory.

At the earlier stage of the disease, timely supportive counseling for patients and their caregivers after disclosure of diagnosis is needed to manage impact of the disclosure. Counseling is also required to open up communication between patients and their caregivers. Paced education for patients and their caregivers through verbal communication and audiovisual aids, complemented by support groups, will benefit both patients and their caregivers.

At a later stage of the disease, there is a need to help

- 1) the patients work towards a respectful death and
- 2) their caregivers achieve a peaceful farewell.

Advance care planning can be a platform for patients and their caregivers to achieve the closure needed by both parties.

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Abstract - WCN 2013

No: 1976

Topic: 7 - Neuromuscular disorders

Brachial plexopathy due to localized axillary recurrence of breast cancer

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The brachial plexopathy in breast cancer patients has been reported to be due to loco-regional metastasis or radiation plexopathy. Localized axillary recurrence (LAR) is rare. The time to LAR after primary breast cancer is various from a few months to many years. We report a patient of brachial plexopathy due to LAR that developed twenty four years after radical mastectomy and radiation therapy for breast cancer.

A 58-year-old woman was admitted with left hand weakness, paresthesia and pain. The examination revealed edema, weakness and sensory change of left arm and hand. But there was no palpable mass in subclavian fossa or axilla. The NCS and EMG revealed left brachial plexus lesion (multiple levels of roots and trunks). Chest CT and breast sonography showed huge mass in the left axilla with invasion of left subclavian vessels. PET CT showed huge malignant mass along left axillary fossa without distant metastasis. We performed ultrasonography-guided gun-biopsy of mass and the pathologic diagnosis was invasive carcinoma with positive estrogen and progesterone receptors.

LAR of breast cancer is rare but can occur anytime after initial treatment. LAR is associated with poor prognosis because it is an independent risk factor for distant metastasis and death. We need to evaluate lymph node by sonography in the patient of breast cancer because the physical examination of the axilla can reveal false-negative finding in spite of huge mass.

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Abstract - WCN 2013

No: 1938

Topic: 7 - Neuromuscular disorders

Idiopathic inflammatory myopathies—Potential role of the immunoexpression of interleukin-35 in muscle biopsy

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Background: Interleukin-35 (IL-35) is a newly described cytokine that belongs to the IL-12 family. Although the experiments in mouse models showed immunosuppressive functions for IL-35, recent papers reported different expression pattern and rather pro-inflammatory properties of IL-35 in humans. Currently available biologic treatment for systemic rheumatic autoimmune diseases including the idiopathic inflammatory myopathies (i.e. polymyositis and dermatomyositis) targets specific cellular and molecular mechanisms; therefore, studies of various molecular pathways of immunopathogenesis in idiopathic inflammatory myopathies are of interest for future therapies.

Objective: To analyze immunoexpression of interleukin-35 in muscle biopsies of inflammatory and non-inflammatory myopathies to assess its pathogenetic and diagnostic value.

Materials and methods: Immunoexpression and its localization of IL-35 was studied by primary rabbit anti-human EB13 polyclonal antibody and mouse anti-human IL-12a(p35) monoclonal antibody in a series of 19 muscle biopsy samples of idiopathic inflammatory myopathies (9 dermatomyositis, 10 polymyositis). Results were compared to those obtained in 10 cases of non-inflammatory myopathies and in 10 control muscles biopsies.

Results: We showed localization of the p35 and EB13 subunits of IL-35 in B cells, T cells and macrophages of the inflammatory infiltrate in idiopathic inflammatory myopathies. No immunoreactivity was observed in healthy controls and in non-inflammatory myopathies.

Conclusion: Our data suggest pro-inflammatory properties of IL-35 in humans and a potential role in the pathogenesis of idiopathic inflammatory myopathies. The immunohistochemical expression of IL-35 might be also of diagnostic help in muscle biopsy.

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Abstract - WCN 2013

No: 1968

Topic: 7 - Neuromuscular disorders

Spinal pain syndromes: Psycho-emotional facets

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Background: Low back pain syndromes, especially the chronic ones, are the important link in neurology. Pain as personal experience develops not only due to physical pathology, but as consequence of person's attitude towards disease, previous experience, described in terms of sensory and emotional disorders.

Material and methods: We studied 23 patients with lumbar-sacral radiculopathies. Features of psycho-emotional state were studied using Hamilton scale and Aysenk questionnaire. Pain syndrome was assessed using VAS and DN4 scale.

Results: Disorders like depressive syndrome were observed among 58% patients. Pain syndrome according to VAS was 6.9 ± 0.3 in persons with

depressive signs and 5.5 ± 0.2 in persons without the latter. Among the persons with duration of the pain syndrome 2 and more months neuropathic elements developed that were assessed according to questionnaire DN4 (35%). Psycho-emotional disorders were very evident among patients with choleric and melancholic type (according to Aysenk questionnaire). Among persons with melancholic temperament course of pain syndrome was overlapped with depressive signs in 79%, among patients with sanguinic temperament it was not the case. Intensity of the pain syndrome was associated with the next trends: it was higher in persons with melancholic and choleric type (6.7 ± 0.2 and 6.3 ± 0.2 , respectively), and lower among persons with sanguinic and phlegmatic type (5.9 ± 0.2 and 4.8 ± 0.3 , respectively).

Conclusions: Study of the psycho-emotional features of pain perception, would contribute to optimization of the therapy and rehabilitation among the patients with chronic spinal pain.

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Abstract - WCN 2013

No: 2009

Topic: 7 - Neuromuscular disorders

Assessing motor units with an improved MUNIX

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Objective: Improvement and validation of the new non-invasive neurophysiological method MUNIX (motor unit number index). The reliability, practicability and inter-rater-variability of potential improvements to MUNIX were determined and compared to two established Motor Unit Number Estimation (MUNE) methods.

Methods: 40 healthy subjects and 18 patients with amyotrophic lateral sclerosis (ALS) were studied prospectively at single point or multiple points in time. MUNIX results were compared with incremental stimulation MUNE (IS-MUNE) at abductor digiti minimi muscles (ADM), and with spike-triggered averaging MUNE (STA-MUNE) at trapezius muscles (TRA). In contrast to the original MUNIX method, we recorded a continuous electromyogram during increasing muscle contraction to reduce the influence of both patient's compliance and investigator bias. Moreover, baseline correction for CMAP was implemented and the influence of the parameter settings (filters, number of data points, rectifying) was systematically studied.

Results: The best parameter setting includes high pass-filter 10 Hz, low pass-filter 3000 Hz, number of data points 1000, and rectification of the signals. This leads to an improved correlation between MUNE and MUNIX, up to 0.85 for ADM and 0.7 for TRA. The inter-rater-variability is expected to be considerable better than for IS-MUNE/STA-MUNE.

Conclusion: The improved MUNIX needs minimum patient cooperation, and provides stable results easily and quickly. Improved MUNIX is suggested as an excellent alternative to the established MUNE methods in distal and as well as in proximal muscles.

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Abstract - WCN 2013

No: 1987

Topic: 7 - Neuromuscular disorders

Clinical, ultrastructural study of trasportinopathy, a new nuclear envelope LGMD

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Background: LGMD are due to heterogeneous causes. Muscle histopathological, ultrastructural features of a large Italian-Spanish family with autosomal dominant LGMD, mapped to 7q32.1-32.2 (LGMD1F) were investigated.

Patients and methods: We collected the clinical history in 19 patients. We observed that the age of onset varied from 2 to 35 years, and occurred either in the upper or in the lower girdle; in 14 cases there was hypotrophy both in the proximal upper and in the lower extremities. The severity was not increased in successive generations. New findings were arachnodactyly, dysphagia and dysarthria; muscle biopsy histopathology was investigated in one pair of affected patients (mother 1 biopsy, her daughter 2 consecutive biopsies at 9 and 22 years).

Results: The daughter has a more severe clinical course, the first biopsy had only type 1 fiber atrophy while increased fiber atrophy was observed in the second biopsy. The mother had a compromised muscle histopathology (more muscle fiber variation, and autophagic changes by acid phosphatase stain). An abnormal sarcomeric assembly is the cause of progressive atrophy and myofiber loss. Electron microscopy revealed accumulation of myofibrillar bodies. Accumulation of myotilin and p62-positive aggregates was observed; autophagosomes were also present.

Conclusions: A defect in transportin-3 gene has been found as the cause of this disease, which represents a new mechanism of limb-girdle myopathy.

Our data seems to suggest a new type of nuclear envelop disease. It is possible that SR protein cannot migrate or be transported-in and -out of the nuclear membrane.

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Abstract - WCN 2013

No: 1282

Topic: 7 - Neuromuscular disorders

Lipid storage myopathies: Control of nutrition to prevent whole body catabolism

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Background: Lipid storage myopathies (LSM) represent a complex and potentially treatable group of metabolic myopathies with various ethiological mechanisms: defects of carnitine transport, CPT II defects, defects of Beta-oxidation, Riboflavin-Responsive Multiple Acyl-CoA Dehydrogenase (RR-MAD) deficiency due to ETF Dehydrogenase-deficiency and two types of Neutral Lipid Storage Disorder (NLSM-M, NLSM-I).

Objective: The lysosomal-autophagic pathway might be activated by starvation and plays an important role in both cellular and lipid catabolism. This pathway has not been so far studied in LSM.

Patients and methods: We investigated the Transcription Factor EB (TFEB), a master regulator of lysosomal biogenesis and various autophagy markers (LC3 and P62) by immunohistochemistry and Western Blot in six cases of LSM: one case of CPT deficiency, carnitine deficiency myopathy and RR-MAD due to ETF-dehydrogenase deficiency, and also two late-onset cases of Neutral Lipid Storage Disorders were studied.

Results: In LSM, except the case of CPT II deficiency, we observed activation of LC3 in a perivacuolar position and by Western Blot, while P62 was positive in atrophic fibers or in a more diffuse pattern in some fibers in cytoplasm or around lipid vacuoles, TFEB appeared activated in nuclei and in cytoplasm of late-onset NLSM.

Conclusions: We show that in LSM a transcriptional regulatory mechanism links the autophagic pathway to fatty acid cellular energy metabolism. It is well known that sudden crisis in LSM can be provoked by denutrition, with a still undetermined mechanism.

These observations can explain the deterioration of these patients during starvation and suggest control of nutrition to implement therapeutic strategies beside carnitine of riboflavin for disorders of lipid metabolism.

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Abstract - WCN 2013

No: 1992

Topic: 7 - Neuromuscular disorders

Brachial amyotrophic diplegia associated with a novel SOD1 mutation

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Background: Brachial amyotrophic diplegia (BAD) is an adult-onset subtype of lower motor neuron disease mainly affecting men, largely restricted to proximal arm and shoulder girdle muscles without involvement of lower limbs or appearance of pyramidal signs. BAD is considered a sporadic disorder although a SOD1 mutation (L106P) was recently described in a patient with BAD.

Objective: To describe the association between BAD syndrome and a novel SOD1 mutation in a small Italian family.

Patients and methods: A 72 years-old man presented with a 5 years history of weakness and wasting that began in the right arm and slowly progressed to the proximal contralateral arm. His 78 years-old brother complained in the last years a progressive difficulty in precise hand movements. Both patients performed full clinical and neurophysiological examinations and the direct sequencing of the five exons of SOD1 gene. No clinical information concerning their parents were available.

Results: In our family both affected members had clinical and electrophysiological findings of adult onset BAD syndrome with a history of selective involvement of upper limbs without pyramidal tract dysfunction. SOD 1 analysis disclosed in both patients a base substitution of adenine for thymine (GCT > GCA), at codon 140, known as the A140A 'silent' mutation since it doesn't change the correspondent encoded aminoacid (alanine). The same mutation has been previously described in few familial cases and in many sporadic ALS but it wasn't found in control population, suggesting its pathogenic role.

Conclusion: Our family pointed out the genetic heterogeneity underlying BAD syndrome.

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Topic: 7 - Neuromuscular disorders

Stapedius reflex testing demonstrates improvement of small muscle function with enzyme replacement therapy

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Introduction: Pompe disease primarily affects skeletal and cardiac muscles. It is difficult, assessing enzyme replacement therapy (ERT)

efficacy with end points based on changes in limb-girdle muscles, gait-endurance, and respiratory function, i.e. parameters depending on varying patient-performance. ERT-benefits might be more objectively shown by assessing function of very small muscles, such as the stapedius- and tensor tympani-muscles.

Objective: To assess whether stapedius-reflex-testing shows muscle-function-improvement with ERT in Pompe patients.

Methods: In four Pompe-patients, we determined stapedius-reflex-thresholds ipsilaterally in both ears one and two years after first dosage of biweekly ERT with alglucosidase alfa (Myozyme™, 20 mg/kg KG i.v.). Before ERT-onset, stapedius-reflex-thresholds were assessed in two patients. Muscle tension of stapedius- and tensor tympani-muscles was assessed by measuring acoustic impedance at the tympanic membrane in response to single bursts of 0.5, 1, 2, and 4 kHz tones. Reflex-thresholds above 90 dB indicate impaired reflex-thresholds.

Results:

Patient-1 (female, 46 years) had no responses before ERT-onset, reflex-thresholds of 98.8/87.5 dB (right/left ear) one year, and 91.3/87.5 dB two years after ERT-onset.

Patient-2 (male, 65 years) had reflex-thresholds of 98.8/93.8 dB before, 86.3/91.3 dB one year, and 90.0/91.3 dB two years after ERT-onset.

Patient-3 (female, 53 years) had reflex-thresholds of 93.8 dB (right) and no responses (left) one year, and 96.3/98.8 dB two years after ERT-onset.

Patient-4 (female, 69 years) had reflex-thresholds of 85.0/85.0 dB one year, and 80.0/81.3 dB two years after ERT-onset.

Conclusion: Stapedius-reflex-testing demonstrates impaired small muscle-function in untreated Pompe-patients. After one and two years of ERT, stapedius-reflex-thresholds improved, objectively demonstrating muscle-function-improvement with ERT.

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Abstract - WCN 2013

No: 2061

Topic: 7 - Neuromuscular disorders

A randomized controlled trial comparing botulinum toxin type A Xeomin® and dysport® for treatment of primary axillary hyperhidrosis

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Two non-bioequivalent toxins: Xeomin® and Dysport® are compared for the treatment of primary axillary hyperhidrosis in a randomized controlled double blind trial.

Methods: Twenty patients with primary axillary hyperhidrosis were treated with 50 units of Xeomin into one axilla and 150 units of Dysport into the other axilla. Both patients and the physician were blinded as to which axilla received which of the two toxins. Pain at the injection site has been evaluated by the VAS scale. The patients will be followed for the next 12 months to evaluate the difference in these two toxins in regard to side effects, time to the onset and the extent of reduction of sweating. All patients have completed the Hyperhidrosis Disease Severity Scale (HDSS) and the Dermatology Life Quality Index (DLQI) before treatment and at each follow up evaluation.

Results: No significant difference was observed between the two toxins regarding pain at the injection site. Preliminary data about side effects, time to the onset of reduction of sweating and amount of sweat reduction, as well as QoL data will be available after the first six months of follow up.

Conclusions: Axillary hyperhidrosis causes considerable emotional and social problems affecting the daily life. Xeomin, a BTX-A without

complexing proteins in its formulation, can be used safely and effectively for the treatment of axillary hyperhidrosis. To our knowledge, this will be the first study comparing the therapeutic effect of Xeomin and Dysport in the treatment of axillary hyperhidrosis.

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Abstract - WCN 2013

No: 2056

Topic: 7 - Neuromuscular disorders

Myogenic and endothelial progenitor cells in juvenile dermatomyositis

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Background: Juvenile dermatomyositis (JDM) is a rare immune-mediated microangiopathy characterized by a reduction of endomysial capillaries and perifascicular muscle atrophy. Whether muscle fiber injury induces regenerative processes in JDM muscle remains poorly investigated.

Objective: Our aim was to analyze expression of myogenic transcription factors involved in the regulation of reparative myogenesis as well as the frequency of endothelial progenitor cells in muscle biopsies from JDM patients.

Patients and methods: Markers of satellite cells (Pax7), proliferating (MyoD) or differentiating (Myogenin) myoblasts, and regenerating fibers (developmental Myosin) were studied by immunohistochemistry in patients with JDM (n = 7) compared to controls (n = 4). In addition, capillary density and numbers of endothelial progenitor cells within the endomysium were determined by double-immunofluorescence for CD34 and laminin.

Results: Myogenic regulatory factors (Pax7, MyoD and Myogenin) and developmental Myosin were highly up-regulated in perifascicular regions of JDM (n = 5). Within intrafascicular regions of JDM (n = 7), the number of Pax7+ satellite cells was equivalent to controls. Expression of MyoD, Myogenin and developmental myosin was rarely observed intrafascicularly in JDM and absent in controls. Loss of capillaries was pronounced in perifascicular regions of JDM (n = 5). Quantification of CD34+ endothelial progenitor cells within the endomysium, which are implicated in the process of neovascularisation, revealed no difference between JDM and controls.

Conclusions: Our results indicate induction of regenerative myogenesis in perifascicular regions of JDM, despite pronounced loss of capillaries. No evidence was found for compensatory neovascularisation.

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Abstract - WCN 2013

No: 2119

Topic: 7 - Neuromuscular disorders

An investigation into the quality of life of patients receiving surgical intervention for the treatment of chronic lower back pain

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Background: Lower back pain (LBP) is one of the most prevalent forms of chronic musculoskeletal pain and is the primary cause of disability in persons under the age of 45 years. LBP is a frequently

occurring health problem; often associated with substantial personal and community burden. LBP includes many symptoms that have a major adverse impact on individuals' lives, often resulting in a reduction in quality of life.

Objective: The current study aimed to evaluate the subjective wellbeing of patients prior to, and post-surgical intervention for the treatment of chronic LBP.

Patients and methods: The current study employed a longitudinal design. The Australian Unity Personal Wellbeing Index was utilised to assess quality of life in approximately 150 participants who were diagnosed with chronic LBP. All patients received either lumbar interbody fusion or decompressive laminectomy. Assessments were conducted at two time points; prior to surgery and within one week post-surgery.

Results: When compared to an Australian normative population (N = 1980), patients receiving surgical intervention for chronic LBP reported significantly lower quality of life across all domains investigated. Furthermore, patients' quality of life increased dramatically immediately post-surgery. The most dramatic improvement in quality of life was prevalent in relation to the domains of health and future security. Further results will be discussed.

Conclusion: Patients diagnosed with chronic LBP have significantly lower personal wellbeing than the general Australian population. After receiving surgical intervention, patients reported a statistically significant improvement in their quality of life. Further theoretical and clinical implications will also be noted.

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Abstract - WCN 2013

No: 2146

Topic: 7 - Neuromuscular disorders

Anterior horn cell and encephalopathic variant of Fabry's disease – The neuro-Fabry

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Fabry's disease has always been considered to be predominantly as of cardiac or renal type. Stroke due to vasculopathy and peripheral involvement of A-delta and C fibres with resulting neuropathic pain and autonomic dysfunction has been said to be the neurological involvement.

We report an unrecognised neurological variant of Fabry's disease.

A 33 year old, was seen in the neurology clinic with weakness, fasciculation and progressive wasting of initially the right hand and right upper limb over 8 months and progressed to involve the left upper limb. An electrophysiological diagnosis of anterior horn cell disease (motor neuron disease) was made. There is no relevant history other than a past history of encephalitis in his teens with mild cardiac involvement without a cause being found.

He, during the course of investigation developed encephalopathy that recovered in 3 weeks, spontaneously. Atypical demyelinating white matter disease on repeated contrast MRI of the brain and cervical cord without progression and persistently elevated cerebrospinal fluid (CSF) lymphocytes with normal levels of other parameters on repeated lumbar puncture were noted despite clinical recovery from encephalopathy. Extensive investigations were done to identify the cause of atypical central nervous system (CNS) demyelination, anterior horn cell disease with CSF lymphocytosis.

A dry blood spot and white cell enzyme studies on separate occasions showed low alpha-galactosidase A levels. Genetic study confirmed Fabry's disease with mutation c.299G>A, p.R100K.

Fabry's disease should be considered in the differential of CNS demyelinating disorders and encephalopathy or motor neuron disease with white matter changes.

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Abstract - WCN 2013

No: 1825

Topic: 7 - Neuromuscular disorders

Severe myotonia in juvenile myotonic dystrophy type 2 and sodium channel gene mutation

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Background: Myotonic dystrophy type 2 (DM2) is an adult onset muscular dystrophy caused by a dominantly transmitted (CCTG)_n expansion in intron 1 of the ZFN9 gene. Myotonia generally is mild and inconsistent in DM2. A severe or a juvenile onset myotonia may hide additional genes and/or modifying factors that need to be explored. An association between severe myotonia in DM2 and the presence of recessive chloride channel (CLCN1) mutation has been reported (Cardani R et al. 2012). A similar association with sodium channel gene (SCN4A) mutation has not been described.

Objective: To describe a family with atypical DM2 phenotype characterized by severe myotonia of early onset.

Case description: A 25 year old girl complained of hand cramping and difficulty to relax after activity. Neurological examination showed thenar percussion myotonia and mild distal weakness. EMG showed myotonic discharges in all muscles examined and genetic testing was positive for DM2. Genetic screening for CLCN1 did not identify any alteration in DNA sequence. The analysis of SCN4A gene revealed a SCN4A variant c.215C>T (p.Pro72Leu). Both SIFT and PolyPhen-2.2.2. (HumVar) predicted that this variant is pathogenetic. It may justify the atypical phenotype characterized by severe myotonia of the patient.

Conclusion: A severe and early myotonia in juvenile DM2 suggests the presence of modifying factors contributing to the phenotype. A screening of SCN4A gene is thus recommended in addition to CLCN1.

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Abstract - WCN 2013

No: 333

Topic: 7 - Neuromuscular disorders

Sonoporation delivered gold nanoparticles for myopathy treatment on rat model

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Background: Gold nanoparticles shown strong cardiomyoprotective effects, metallic nanoparticles enhance viability of a variety of neural cells.

Objectives: The aim was to test the myoprotective effects of gold nanoparticles on myopathy rat model, and to test the sonoporation effect to increase nanoparticles' delivery into myocytes.

Material and methods: Wistar 180–200 g rats (of each sex were selected on the basis of analogies (n = 30) were included for the

experiment to model statin-induced advance myopathy, registered by ultrasonography using transducers up to 12 MHz. We formed three groups of animals: the 1st group received colloid 20 nm nanogold (193 mg/ml for metal) injected into the injured muscle using 31 G needles under US guidance; the 2nd group of rats received additional insonation of injection locus in depth of 1 cm by 180 s by 130 Db 3–8 MHz ultrasound; and the 3rd group was the control. Morphological, laboratory, and ultrasound assessments were performed.

Results: In animals after nanogold injection regression of myopathy was registered clinically, on US. Light-optical microscopy defined in tissue samples the regression of muscle damage, vakuoli degeneration and myofibril lysis. Nanogold induced significant inhibition of Ca, Mg-ATPase activity in myofibrils after (1st vs 3rd group, p < 0.01). Electron microscopy demonstrated higher nanoparticle cumulation in myofibrils and mitochondria (P < 0.01 for both). Myopathy regression was clearly defined in all rats after sonoporation (1st vs 2nd group p < 0.05).

Conclusions: Gold nanoparticles have significant myoprotective effects, being a potential strategy for the treatment of muscular dystrophies, applicable for disperse localizations. Sonoporation is able to enhance gold nanoparticle delivery to myocytes in vivo.

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Abstract - WCN 2013

No: 2168

Topic: 7 - Neuromuscular disorders

A novel mitofusin 2 MFN2 gene mutation causing early onset Charcot-Marie-Tooth 2A disease: Genetic, clinical and MR spectroscopy characterization

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Background: Mitofusin 2 (MFN2) mutations are associated with axonal peripheral neuropathies known as Charcot-Marie-Tooth 2A (CMT2A). Even though central nervous system (CNS) involvement has been described in CMT2A, there are as to now no advanced neuroimaging study of patients with MFN2 mutations.

We describe a novel MFN2 mutation detailing its clinical presentation, and describing novel neuroimaging findings.

Materials and methods: A 7 yo boy with unremarkable family history underwent neurological assessment and gait analysis. Routine electrophysiological investigation (EMG and ENG) was consistent with axonal polyneuropathy. The MFN2 gene sequence was analyzed. Neuroimaging included standard morphological and spectroscopic (MRS) sampling.

Results: Sequence analysis showed a novel MFN2 mutation (c.2116A → C, T706P). Sequence analysis on the parents confirmed it as a de-novo mutation. The residue is situated between the transmembrane domain and the coiled-coil region close to the C-terminal. The clinical features included early onset with steppage, hyporeflexia and hypopallesthesia. Computerized gait analysis demonstrated primary distal lower limb involvement with club feet and accentuation of the hips and knee flexion. Standard MRI didn't reveal brain signal alterations. MRS showed an increase of NAA and Cho peaks.

Discussion: This novel mutation (T706P) is associated with early onset CMT2A. The MRI findings, in the absence of macroscopical brain signal alterations, show MRS data suggestive of whole brain biochemical involvement (elevated NAA, increased Cho). These findings, especially in such a young patient, confirm CMT2A as a disease involving both CNS

and PNS, and highlight the importance of MR spectroscopy as an objective indicator of disease.

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Abstract - WCN 2013

No: 2187

Topic: 7 - Neuromuscular disorders

Frontiers of amyotrophic lateral sclerosis cognitive assessment: The use of Eye-tracking and Brain Computer Interface in the eBrain project

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Background: Many patients affected by Amyotrophic Lateral Sclerosis (ALS) show cognitive alterations, especially regarding frontal executive functions. Cognitive assessment is problematic in moderate-severe stages of ALS, due to the presence of motor-verbal impairment. Recently, Eye-tracking (ET) and Brain Computer Interface (BCI) have been preliminarily used in ALS to administrate cognitive testing. However, an extended motor-verbal free neuropsychological (NP) battery is not yet available.

Objective: A recently funded project, “eBrain: BCI-ET for ALS”, aimed to evaluate the use of P300-based BCI and ET technologies to administrate cognitive testing in ALS.

Patients and methods: 28 ALS patients (mean age: 62.6 ± 11.8; mean education: 9.6 ± 3.6) and 30 healthy subjects (mean age: 56.2 ± 11.9; mean education: 13.7 ± 4.2) underwent a comprehensive motor-verbal free NP assessment, administered with both P300-BCI and ET. Moreover, clinical data were collected and usability of both devices was evaluated.

Results: Data showed significant differences between healthy subjects and ALS patients' performances in BCI and ET adapted measures of frontal abilities ($p < .05$); furthermore, a correlation between traditional NP assessment and BCI-ET one was found ($p < .05$), supporting the concurrent validity of the adapted measures. Finally, even if a higher perceived usability was overall observed for ET compared to BCI ($p < .05$), patients evaluated BCI as a positive and useful tool in order to compensate for the motor-verbal limitations.

Conclusion: These results support the good level of sensitivity and perceived usability of the BCI and ET-based NP assessments, offering promising insights on the use of such devices for the longitudinal cognitive testing in ALS.

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Abstract - WCN 2013

No: 2236

Topic: 7 - Neuromuscular disorders

Therapeutic effects of repetitive transcranial magnetic stimulation on clinical symptoms and signs of patients with HTLV1 myelitis

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Background: HTLV1 virus myelitis causes pain and paresthesia of the lower limbs, sphincter function impairment, and most disabling feature of spasticity. Current treatments for this disease have not been effective. Providing the patients with a more effective treatment can improve greatly the quality of life.

Objective: Studying therapeutic effects of repetitive transcranial magnetic stimulation (rTMS) on improvement of pain, spasticity and motor function of people suffering from HTLV1 myelopathy, one month after initiation of treatment.

Materials and methods: In this study 9 patients with confirmed symptomatic HTLV myelitis who had not responded satisfactorily to other treatments (methyl prednisolon, alfa interferone, muscle relaxants) were included. They received 5 sessions of rTMS with a frequency of 5 Hz and 100 times on each side at every session, in five successive days, stimulation sites were bilateral motor cerebral cortex responsible for movement of lower limbs. Spasticity, pain, and motor function were measured by Modified Ashworth Scale, Global Pain Scale, and Medical Research Council Scale, respectively. The data were analyzed by the Wicoxon test.

Results: Spasticity diminished significantly after one month of treatment. Patients showed improvement regarding pain and movement functions, though not statistically significant.

Conclusion: These findings suggest that rTMS can be used to reduce spasticity, as the most disabling feature of HTLV1.

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Abstract - WCN 2013

No: 2263

Topic: 7 - Neuromuscular disorders

Neuroanatomical patterns of central nervous system involvement in myotonic dystrophy type 1 and clinical correlates

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Background: Patients with myotonic dystrophy type 1 (DM1) often show cognitive impairment during the course of disease, beside the motor symptoms.

Objective: To investigate grey matter (GM) and white matter (WM) abnormalities in a large sample of myotonic dystrophy type 1 (DM1) patients and to assess their correlations with clinical findings.

Methods: We enrolled 51 DM1 patients [n = 14 with childhood-juvenile DM1; n = 37 with classic DM1] and 30 healthy controls (HC). Patients underwent clinical and neuropsychological evaluations and brain structural and diffusion tensor (DT) magnetic resonance imaging (MRI). Voxel based morphometry and tract-based spatial statistics were used to evaluate GM atrophy and WM microstructural damage in DM1 patients compared with HC. Regression analyses were performed to correlate GM and WM changes with clinical and cognitive variables.

Results: DM1 patients had moderate muscle impairment, sleepiness and depression as well as attentive-executive and visuo-spatial deficits. Voxel-wise analysis revealed significant and widespread GM atrophy and WM damage in DM1 group (both childhood-juvenile and classic patients) compared with HC. Significant negative correlation was found between Addenbrooke's Cognitive Assessment-revised orientation and attention sub-scores and MD increase along left corona radiata, internal capsule, inferior fronto-occipital fasciculus and SLF (temporal part) ($p < 0.05$).

Conclusions: Our data demonstrate the clinical relevance of GM and WM damages in a large sample of DM1 patients. These findings are in line with neuropathological studies showing severe central nervous system involvement in this multisystemic disease.

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Abstract - WCN 2013**No: 2264****Topic: 7 - Neuromuscular disorders****TNF- α is the critical molecule in critical illness myopathy; TNF- α antagonism prevented the neuromuscular changes in septic rats**

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Critical illness myopathy (CIM) is characterized with generalized and respiratory muscle weakness, accompanied with difficulty of weaning from mechanical ventilation and occurs mainly during sepsis after a 1 week intensive care stay. There is no effective treatment. During sepsis, TNF- α causes muscle breakdown, and also influences muscle excitability. This knowledge has led us think that TNF- α might be the critical molecule for CIM and CIM can be prevented by TNF- α decreasing agents pentoxifylline and etanercept. CIM have been searched with septic rats, induced by cecal ligation and puncture (CLP). TNF- α and IL-1 β levels were measured in plasma and muscle at the 2nd hour after operation. Nerve conduction studies (NCS) performed at the 48th hour. In one group Pentoxifylline was given 1 hour before the operation and for 6 days after. In the other group etanercept was given 1 day before the operation and repeated after 72 hours. Muscle biopsies were performed on the 6th day for histopathologic diagnosis and for detecting myosin level. This research showed that, both pentoxifylline and etanercept reduced mortality. Increased TNF- α levels of muscle revealed statistically significant difference from other groups in CLP group. NCS investigations revealed an early decrease in CMAP amplitudes in the same group. The main findings were fiber atrophy and basophilic fiber staining mainly in CLP group muscle biopsies. Myosin levels were lowest in CLP and CLP + Pentoxifylline group and similar with control group in CLP + Etanercept group. This study showed that TNF- α antagonism prevented the development of the neuromuscular changes in septic rats, and did not cause any increase in mortality rate.

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Abstract - WCN 2013**No: 2298****Topic: 7 - Neuromuscular disorders****Progress report on the development of new classification criteria for adult and juvenile idiopathic inflammatory myopathies**

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Background: Inadequate classification criteria for idiopathic inflammatory myopathies (IIM) limit clinical studies in myositis. An international,

multidisciplinary collaboration, the International Myositis Classification Criteria Project (IMCCP), was therefore established.

Objective: To develop and validate new classification criteria for adult and juvenile IIM and its major subgroups.

Patients and methods: Candidate variables were selected from published criteria and inclusion criteria in controlled trials of myositis. Comparator conditions resembling IIM were defined.

Clinical and laboratory data from IIM and comparators were collected from 47 rheumatology, dermatology, neurology and pediatrics clinics worldwide. Crude pair-wise associations among all measured variables and between each variable and clinicians' diagnoses were assessed. Explored approaches were:

1. Traditional: case defined by specified number of items from a set.
2. Probability score: case assigned a probability score by summing score-points associated with a set of variables.
3. Classification tree: case defined by decision tree.

Internal validation using bootstrap methods was performed.

Results: Data from 973 IIM patients and 629 comparators were obtained. Two probability score models were developed: Model 1 comprised clinical variables on muscles, skin, and laboratory measures; and Model 2 additionally comprised muscle biopsy variables. Model 1 performed nearly as well as Model 2 (specificity 87% vs 88%, sensitivity 89% vs 89%, and correctly classified 87% vs 89%). Both models performed equally or better than the classification tree that was developed and published criteria.

Conclusion: New classification criteria for IIM with readily assessable measurements and symptoms have been developed with generally superior performance compared with existing criteria.

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Abstract - WCN 2013**No: 2154****Topic: 7 - Neuromuscular disorders****HLA associations with mg in Saudi patients**

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Background: Myasthenia gravis (MG) is a rare autoimmune disease of the neuromuscular junction. MG has been shown to be associated with many HLA antigens in different populations. Recently using low resolution typing technique we found associations with HLA-A*23, B*08, B*18, DRB1*16 and DRB1*13.

Objectives: To further describe the association of HLA in Saudi MG patients using SBT.

Methods: We analyzed HLA associations using SBT in 75 Saudi MG patients and 1110 health controls.

Results: Our results supported our previous report with some new findings, B*08:01 (OR 3.58, p = 0.000), DRB1*16:01 (OR 6.82, P = 0.000). A*23:01, B*50:01, C*06:02 & DRB1*0701 were found to form the most common haplotype in our healthy controls. All these alleles were significantly reduced (protective) in the MG patients. No single allele of DRB1*13 was significantly associated with MG, however, collectively 13:01, 13:02 and 13:03 alleles were associated MG (OR 1.47, p = 0.056). Some of our new findings include HLA-C*07:02 and DQB1*05:02 association with MG. It is interesting to note that in Caucasoids A*01:01–B*08:01–C*07:01–DRB1*03:01 haplotype carries the highest risk for MG. Here we showed that a part of

this haplotype is present; B*08:01–DRB1*03:01 but not A*01:01 C*07:01.

Conclusions: Our results supported our previous report with some new findings. This finding may help in dissecting out the primary association with MG.

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Abstract - WCN 2013

No: 2405

Topic: 7 - Neuromuscular disorders

Low and high frequency repetitive nerve stimulation in the diagnosis of myasthenia gravis

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Background: Diagnosis of myasthenia gravis (MG) typically is made based on patient's history and clinical findings. However, the diagnosis must be verified by the electrophysiological investigation of neuro-muscular transmission. The repetitive nerve stimulation (RNS) is an electrodiagnostic technique used to assess the efficacy of neuromuscular transmission.

Objective: The objective of this study was to evaluate combined muscle action potential (CMAP) changes in response to high and low frequency rates of RNS in patients with MG.

Patients and methods: The correlation of muscle strength and CMAP changes were investigated in 104 myasthenic patients. The muscle strength was graded on a scale from 0 to 5. Two stimulating parameters were used throughout the experiments – the low frequency stimulation (LFS) and high frequency stimulation (HFS).

Results: It was found that in 15% of muscles with clinically detectable weakness the CMAP amplitude was decreased, by 21% in area, and by 8% in duration of the negative phase.

The positive correlation was observed between the negative phase of CMAP and the muscle strength and negative correlation was observed between the negative phase duration and the muscle strength.

Conclusion: We used RNS in a large cohort of patients as an additional tool for the confirmation of the MG diagnosis. We registered higher decrement in larger muscle groups which are more commonly affected clinically. However, smaller muscles, such as the abductor digiti quinti muscle are often preferable to investigate, because they create less discomfort for the patient and have fewer artifacts.

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Abstract - WCN 2013

No: 2277

Topic: 7 - Neuromuscular disorders

Altered somatosensory neurovascular coupling in patients with becker muscular dystrophy

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Background: In patients suffering from Becker muscular dystrophy (BMD) variable degrees of cognitive impairment have been reported in addition to the well-known muscular and cardiac symptoms. BMD is caused by mutations in the dystrophin gene, changing the cellular dystrophin complex and subsequent muscular blood flow. Few studies have addressed the mechanisms of cognitive impairment. Lack of dystrophin, however, causes secondary loss of vasoactive molecules such as nitric oxide, which is as an important player in brain vascular control. We hypothesised that dystrophin can be involved in neurovascular coupling and cerebral reactivity measured by functional MRI and EEG.

Objective: To study the baseline electrophysiological and fMRI BOLD-response to external somatosensory stimulation in BMD patients compared to age and sex-matched healthy controls.

Patients and methods: 17 men (mean age 38.5, range 25–63) with BMD and 8 healthy sex matched controls were included. Functional MRI (fMRI, BOLD) (3T, Phillips) and EEG with evoked potentials were performed during a somatosensory stimulation paradigm on the dominant median nerve.

Results: BMD patients showed a significantly ($p < 0.02$) decreased electrophysiological adaptation together with a decreased ($p < 0.01$) BOLD-response in the primary somatosensory cortex when compared to healthy controls.

Conclusion: The preliminary results suggest that patients with BMD have a fundamentally altered cerebral neurovascular coupling, which could impact cognitive function in BMD patients.

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Abstract – WCN 2013

No: 2379

Topic: 7 – Neuromuscular disorders

Two cases of POEMS syndrome: Initially misdiagnosed as CIDP

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POEMS syndrome (also known as Crow-Fukase syndrome) is a serious systemic disease characterized by polyneuropathy, anasarca, skin lesions and associated with osteoclastic bone lesions and with M-proteinemia. The diagnosis of POEMS syndrome is made robust by finding increased serum vascular endothelial growth factor (VEGF) in addition to the combination of the mentioned characteristic manifestations.

The diagnosis of advanced POEMS syndrome is not difficult when the combinations of characteristic manifestations are fulfilled. However, in the initial stages, only half of POEMS patients start with neuropathy. Neuropathy in POEMS syndrome is characterized by a subacute, progressive sensorimotor type of polyneuropathy with mixed features of demyelination and axonal degeneration.

However, nerve conduction study (NCS) results of POEMS syndrome occasionally fulfill the electrophysiological criteria for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), thus leading to the caveat of an often initial misdiagnosis of CIDP.

We herein present two female patients with POEMS syndrome.

Case 1: A 67-year-old woman was admitted to one university hospital with a 1.5-year history of progressive weakness and paraesthesia. Her weakness and paraesthesia initially affected his lower limbs, progressing within 2 months to her upper limbs.

Case 2: A 24-year-old woman was admitted to another university hospital with a 2-month history of progressive paraesthesia and muscle weakness affecting her lower limbs. Both cases were electrophysiologically diagnosed as CIDP, then treated with intravenous

immunoglobulin therapy without enough effects. After transfer to our university hospital, we paid attention to both cases of thrombocytosis. Both had hepatosplenomegaly, lambda type M-proteinemia, and significantly elevated serum VEGF.

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Abstract – WCN 2013

No: 2291

Topic: 7 – Neuromuscular disorders

Etiological consideration of 26 patients with dropped head syndrome

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Background: The dropped head syndrome (DHS) is caused by a wide variety of neurological disorders ranging from CNS degenerative diseases to peripheral neuromuscular problems. There have been few studies that identified causes of DHS for a large number of patients.

Objective: To elucidate the frequencies of individual causes in a case series of DHS patients.

Materials and methods: Patients presenting with DHS were extracted from the database of our institution for 13 years since 1999. Their medical and EMG records were retrospectively reviewed.

Results: Twenty-six patients were extracted (4 men and 22 women, age 71.1 ± 8.6 years). The final diagnoses were 13 myasthenia gravis (MG), 4 myositis, 1 myopathy of undetermined etiology, 3 amyotrophic lateral sclerosis, 3 Parkinson's disease, 1 dementia with Lewy body, and 1 with unknown etiology. One MG and 1 myositis patients were associated with Parkinson's disease, but they were diagnosed as myopathies by EMG. All but one MG patients were negative for AChR antibody, and MuSK antibody was also negative for all of 10 patients examined. They were diagnosed as MG by the abnormal single-fiber EMG (SFEMG) of the splenius muscle, together with lack of profuse fibrillation potentials and positive sharp waves in the concentric needle EMG (CNEMG) of the same muscle. Immunotherapies were attempted in 6 MG patients (5 IVIg and 1 steroid) and were effective at least transiently.

Conclusion: MG was the most frequent cause of DHS, followed by myositis. CNEMG and SFEMG of the splenius muscle were the key to their diagnosis.

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Abstract – WCN 2013

No: 2252

Topic: 7 – Neuromuscular disorders

Paraneoplastic sensorimotor polyneuropathy and inflammatory myopathy associated with Merkel cell carcinoma

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Background: Merkel cell carcinoma (MCC) is a rare and aggressive neuroendocrine malignancy. In the literature MCC has been very rarely associated with paraneoplastic neurological complications.

Objective: To report a case of paraneoplastic polyneuropathy and myopathy in a patient with MCC of unknown primary origin (MCCUP).

Patient and methods: A 65-year-old man was admitted to our department with progressive limb weakness, numbness and myalgias.

Physical examination revealed a left axillary nodular mass. On neurological examination he exhibited asymmetric, predominantly proximal tetraparesis with absent tendon reflexes. Brain MRI was unremarkable and spinal MRI demonstrated multiple levels of mild spondylosis. Cerebrospinal fluid examination revealed a normal cell count with increased protein content (77.2 mg/dl) and positive oligoclonal bands. Nerve conduction studies were consistent with a mixed sensorimotor polyneuropathy. Treatment with intravenous immunoglobulin was initiated followed by oral steroids, with subsequent gradual improvement of muscle strength. Biopsy of the axillary mass revealed an undifferentiated MCC. Screening for an underlying primary site and testing for anti-onconeural and anti-ganglioside antibodies were negative. Deltoid muscle biopsy revealed inflammatory myopathy and sural nerve biopsy demonstrated mixed axonal and demyelinating changes. The patient underwent complete regional lymph node resection followed by adjuvant chemotherapy, with significant clinical improvement, remaining relapse-free during a 2 year follow-up period.

Results and conclusion: Both the clinical presentation and the positive response of our patient to cancer treatment support the paraneoplastic origin of his neurological condition. MCC should be considered as a diagnostic possibility in patients presenting with peripheral neurologic syndromes with atypical features.

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Abstract – WCN 2013

No: 2330

Topic: 7 – Neuromuscular disorders

26 years' experience in myasthenia gravis in Iran

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Background: Myasthenia gravis is an autoimmune disorder affecting the neuromuscular junction. Signs and symptoms present a wide spectrum ranging from only ocular symptoms to a life-threatening respiratory failure.

Objective: This study was carried out to show the demographic characteristics of myasthenia gravis in Iran. Unusual presenting features of our patients were also taken into account.

Methods: The files of all myasthenia patients arriving to clinic during 26 years were studied on a retrospective basis; their demographic and clinical data were registered and duly analysed. Then, cases of congenital myasthenia were excluded from the study.

Results: Out of the 316 myasthenia gravis patients included in the study, 58.55% were female and 41.45% male. 272 patients suffered from the generalized form of myasthenia while the remaining 51 patients had only ocular presentations. 6 patients had atypical presenting features as the first manifestation (jaw-hanging, cervical weakness, dyspnea). Among those with the generalized type, 11% had suffered crisis and 13.6% experienced at least 1 episode of disease exacerbation. Interestingly the youngest and oldest ages of onset were 8 and 70 years, respectively. Out of the 128 patients who underwent thymectomy, 21.09% and 48.43% had pathology reports of thymoma and hyperplasia, respectively. Unfortunately 6 ladies and 12 men were taken by deaths, two of which died with pneumosepsis.

Conclusion: Epidemiologic findings of Myasthenia Gravis in Iran are more or less similar to those of other countries, but it is very important to bear in mind the atypical presenting features of this disease that can otherwise be misleading.

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Abstract – WCN 2013**No: 2311****Topic: 7 – Neuromuscular disorders
Sporadic Amyotrophic Lateral Sclerosis (SALS) and environment***H.A. Idrisoglu. Neurology, Istanbul University Faculty of Medicine, Istanbul, Turkey*

Background: The cause of Sporadic Amyotrophic Lateral Sclerosis (ALS) is not known. Studies associate toxic, dietary, infectious, neoplastic, and physical factors as underlying, predisposing or pathogenic influences.

Methods: 131 SALS patients are followed prospectively all of the patients investigated SOD1 mutation and blood and urine lead level. Also environmental exposure was assessed, in sporadic ALS patients.

Results: All of the patients are diagnosed definite ALS according to El-Escorial criteria.

81 of 131 patients who are diagnosed with definite ALS according to El-Escorial criteria are men. The others are women. Men–women ratio is 1.5/1. The age of onset of 131 definite ALS cases is between 15 and 73. The average age is 48.7. The beginning of age in men is 40.1. It is 49.7 in women. The average examination age in men is 49.1. It is 51.1 in women. 131 patients are separated to the specific types based on examination findings during examination. 16 of them are progressive bulbar paralysis (PBP), 72 of them are in PBP + spinal form, and 43 of them are in spinal form.

When we search blood and urine lead levels in all 131 patients and when we take family individuals. Of this patients as central group and compare them Blood and urine lead levels of this group are statistically and significantly found high. Blood lead level is $p < 0.001$, urine lead level is $p < 0.01$. We don't encounter abnormal (toxic) levels (0.80 mg/dl) in one of the cases.

All patients are followed prospectively.

Conclusions: We found blood and lead levels of SALS patients statistically and significantly high.

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Abstract– WCN 2013**No: 2373****Topic: 7 – Neuromuscular disorders
Parkinson, pesticides and environment***H.A. Idrisoglu. Neurology, Istanbul University Faculty of Medicine, Istanbul, Turkey*

Background: Parkinson's disease (PD) is the most common neurodegenerative movement disorder that is a consequence of premature death of dopamine-containing neurons in the substantia nigra. A number of observations have led to the hypothesis that environmental factors, including pesticides, play a significant role in the development of PD.

Method: 350 Parkinson patients applied to our clinic between the years 2008 and 2012. We achieved prospective follow up of these patients. In the result of this follow-up we determined that 35 patients were exposed to pesticide intoxication.

Result: Age interval of 35 patients was between 23 and 90. Mean age was 61.7. 33 of them were male and the other patients were female. We determined that 10% of 350 patients were exposed to toxic substances. The first sign of 8 patients was bradykinesias and the first sign of 27 patients was tremor. We determined pesticide intoxication in 19 patients, CO₂ intoxication in 2 patients, heavy metal exposure in 10 patients, CO₂ intoxication in 2 patients and electrical charging in 2 patients. Finally, we thought 10% toxic etiology risk factor. The other characteristics that we were interested in were male patients. The patients were male and PH signs were in association with tremor.

Conclusions: Pesticides and environmental factors and gene interactions seem to act together to increase PD risk.

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Abstract - WCN 2013**No: 2369****Topic: 7 - Neuromuscular disorders
Initial dip and bilobed N21 peak in the tibial nerve SEPs suggest predominant demyelination in the nerve root***T. Chiba^a, M. Sonoo^a, C. Oishi^b, K. Uchino^a, H. Okuma^a, H. Kurono^a, K. Hokkoku^a, Y. Hatanaka^a. ^aDepartment of Neurology, Teikyo University School of Medicine, Itabashi-ku, Japan; ^bDepartment of Neurology, Kyorin University School of Medicine, Mitaka, Japan*

Background: Sensory GBS or CIDP may be difficult to diagnose, especially when routine nerve conduction studies show unremarkable findings. Tibial nerve SEPs are useful because they can evaluate the proximal portions of the peripheral nerve. We noticed a previously-undescribed characteristic pattern in tibial nerve SEPs of such patients.

Objective: To document the significance of the “initial dip and bilobed N21 peak” pattern in the tibial nerve SEPs.

Material and methods: Subjects consisted of 4 patients presenting with this pattern. Their clinical and electrophysiological records were retrospectively reviewed.

Results: All patients presented with pure sensory or sensory-dominant symptoms. Routine nerve conduction studies did not show any sign of demyelination for all patients, except for the slightly reduced amplitude of the sensory nerve action potentials. The final diagnoses were sensory GBS in 2 patients, sensory CIDP in 1 patient, and sensory-dominant MADSAM in 1 patient. Tibial nerve SEPs revealed slightly delayed N8 and P15 components. The L1S-ICc (ipsilateral iliac crest) lead registered a positive dip 2 to 3 ms later than the P15 latency for all patients, and the N21 peak was either lost (1 patient) or showed a characteristic bilobed pattern (3 patients) with the later peak at a markedly delayed latency.

Conclusion: The positive peak must correspond to the “killed-end potential” suggesting the near interruption of the sensory conduction. The mechanism of bilobed N21 potential is unknown, but the recognition of this characteristic pattern will help to identify demyelination of the sensory nerve predominant at the root level.

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Abstract – WCN 2013**No: 2433****Topic: 7 – Neuromuscular disorders
Ultrasound diagnostics and amyotrophic lateral sclerosis***Y. Rushkevich, G. Zabrodets, S. Likhachev. Republican Research and Clinical Center of Neurology and Neurosurgery, Minsk, Belarus*

Background: Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease with progressive destruction of the central (upper) and peripheral (lower) motor neurons. One of the specific symptoms of ALS is the appearance of fasciculations, which may precede the development of muscle weakness and atrophy.

Objective: To explore the possibilities of ultrasound to detect muscle fasciculations in the differential diagnosis of suspected ALS.

Materials and methods: 150 patients were included (ALS, spinal amyotrophy, cervical myelopathy – 126 (84%), 11 (7%), 13 (9%) patients, respectively). Male/female – 93/37, age (median (25–75%)) – 56 (46, 62). The diagnosis of ALS was based on an analysis of clinical data electroneuromyographic El Escorial criteria (Brooks, 1998). All patients underwent electromyography (VikingSelect; Nicolet; USA). Ultrasound

diagnostics was carried out in the B-mode 12.8 MHz linear transducer with symmetric 2-side with the full relaxation of the muscles with the patient lying down. Additionally ultrasound diagnostics was performed on 45 healthy individuals.

Results: Generalized fasciculations on the trunk and extremity muscles related to the myotomes of the cervical, thoracic and lumbosacral levels were detected during ultrasound with ALS, spinal amyotrophy and myelopathy in 122 (96.7%), 1 (9.1%) and 1 (7.7%) patients, respectively. Fasciculations in ALS was characterized by severity and persistence in contrast to other pathology. In 60% cases of ALS generalized fasciculations by visual inspection were noted.

Conclusions: Early detection of muscle fasciculations generalization using ultrasound facilitates the differential diagnosis of suspected ALS.

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Abstract – WCN 2013

No: 2090

Topic: 7 – Neuromuscular disorders

Navigated transcranial magnetic stimulation in Motor Neuron Disease (MND)

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Background: MND is a set of disorders associated with the selective degeneration of both upper and lower motor neurons. Navigated transcranial magnetic stimulation (nTMS) is a tool for functional noninvasive brain mapping.

Objective: Assessment of the upper motor neuron functional state using nTMS.

Patients and methods: 31 patients with MND were included in the study. The mean age was 54.3 ± 12.0 years. Duration of illness ranged from 4 to 160 months (median – 12 months). 29 healthy volunteers with a mean age of 24.8 ± 3.8 years were studied as control group. nTMS (NBS eXimia Nexstim) was performed for all participants. Some parameters of TMS were detected: resting motor threshold (MT) and motor evoked potentials (MEP) at m. abductor pollicis brevis (APB). Also maps of APB cortical representation have been made.

Results: MT was significantly higher in MND patients in comparison to the control group in either hemisphere ($p < 0.001$). There was significant negative correlation between ALS FRS-R and MT, but this parameter didn't depend on disease duration. MND patients had significantly smaller MEP ($p = 0.004$ for right hemisphere and 0.009 for left one). More than all maps of cortical areas were smaller in patients with MND compared with healthy persons, but some patients with short duration of disease had extended maps.

Conclusion: Navigated TMS is an effective assessment of the upper motor neuron functional state in MND. Further researchers may help to reveal the pathogenic mechanisms of neurodegenerative process and the development of diagnostic and prognostic markers.

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Abstract – WCN 2013

No: 2453

Topic: 7 – Neuromuscular disorders

Hereditary sensory neuropathy presenting with only numbness of the extremities

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Background: Hereditary sensory neuropathy is a slow developing, autosomal dominant disease with sensory loss at the distal parts and autonomic disturbances. The definite prevalence is unknown, but it is estimated to be very low. The first onset is on the 2nd and 5th decades. Autonomic changes like sensory loss on the distal parts of the limbs, loss of muscle strength, skin ulcers and especially perspiration disorders are the clinical manifestations. Future complications that can cause serious morbidities can be prevented with early diagnosis.

Objective: In this report we would like to present a case with hereditary sensory neuropathy.

Patients and methods: A 20-year-old male patient presented to our outpatient clinic with complaints of numbness on the hands and feet. The patient stated 'stocking-glove' pattern of numbness. The complaints had become more evident in the last 2 years. On the neurological examination the patient stated stocking-glove pattern of hypoesthesia, and the DTRs were globally decreased. All other neurological findings were normal.

Results: The biochemistry tests were normal. Bilaterally, sensory nerve action potentials were absent on the electromyography (EMG) of the median, ulnar and sural nerves. The patient was diagnosed with sensory polyneuropathy.

Conclusion: We found this case worth reporting in order to remind the clinicians that hereditary sensory polyneuropathy is a rarely encountered disease that should be considered, with no other objective examination findings other than decreased DTRs.

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Abstract – WCN 2013

No: 2435

Topic: 7 – Neuromuscular disorders

Management of muscular dystrophies in Tunisian children

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Background: Muscular dystrophies (MD) are a heterogeneous group of inherited primary disease of muscle that share progressive weakness and dystrophic changes on muscle biopsy.

Objective: To report management approach in Tunisian children with MD and evaluate usefulness of a "Systematic protocol for children with MD".

Methods: Over 8 years (2005–2013), a "Systematic protocol for children with MD" has been used, for management of 86 children with MD. The protocol comprised 25 items including: individual case report form (CRF), diagnostic items, occupational, psychological, orthopaedic, cardiac and respiratory evaluations and corticotherapy. The degree of completion of each item has been analyzed.

Results: There were 57 Duchenne MD (DMD) and 27 recessive Limb-Girdle-MD (LGMD 2).

CRF have been fulfilled in all patients as well as diagnostic items. Psychological and occupational evaluations concerned respectively 84.5% and 97.6% of patients. Respiratory and cardiac evaluations concerned respectively 59.5% and 64.2% of patients. Orthopaedic management including: examination (80.9%), spine X-ray (100%), orthotic devices (92.8%) and wheelchair (36.9%) was planned each time it was needed. Corticotherapy was prescribed to 79.9% of patients. During follow up, 31 patients lost ambulation at mean age of 10.5 years. However, all patients showed improvement of quality of life.

Conclusion: Our study supports the usefulness of "Systematic protocol for MD" since it improves management and allows detecting complications at earlier stages. Knowledge of MD-specific complications and

implementation of anticipatory care have changed the standard of care, with an overall improvement quality of life of patients.

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Abstract – WCN 2013

No: 2471

Topic: 7 – Neuromuscular disorders

FGF21: A biomarker of neuromuscular diseases

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Background: Human fibroblast growth factor 21 (FGF21) is a 181 amino acid protein that belongs to the human FGF superfamily. The basic biological role of FGF21 is the regulation of the glucose and lipid metabolism.

Objective: Recently two observations were published where they showed elevated circulating FGF21 in human mitochondrial diseases, therefore it was suggested that FGF21 might be a biomarker of mitochondrial diseases.

Patients and methods: The serum level of FGF21 was determined by ELISA in blood samples from 20 healthy subjects, 15 patients with myotonic dystrophy type 1 (MD1) and 25 patients with mitochondrial diseases.

Results: Among healthy subjects serum FGF21 correlated with body mass index (BMI). Mean FGF21 level was significantly raised in MD1 compared to healthy subjects (424 ± 328 and 207 ± 165 pg/ml, respectively, $p < 0.05$, Mann–Whitney U test). Among mitochondrial patients FGF21 was elevated only in PEO (progressive external ophthalmoplegia) group (589 ± 496 pg/ml, $p < 0.05$, Mann–Whitney U test), but was not significantly altered in MELAS and myopathy patients. FGF21 correlated with serum creatine kinase (CK) and lactate levels, with clinical severity score as well as some biopsy findings (e.g. ratio of ragged red fibers and mitochondrial inclusions).

Conclusion: Our study implicates that serum FGF21 might be a biomarker for neuromuscular disorders. In contrast to the previous findings our results showed that elevation of FGF21 is not specific and not restricted to mitochondrial disorders. Further research is necessary to find out what neuromuscular disease groups are associated with abnormal FGF metabolism and to investigate the molecular pathomechanism.

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Abstract – WCN 2013

No: 2517

Topic: 7 – Neuromuscular disorders

Complex chromosomal rearrangements in a patient with oligozoospermia and Charcot–Marie–Tooth disease

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Background: Complex chromosomal rearrangements (CCR) occurring in phenotypically normal persons are rare, about 255 cases have been reported. Most familial cases have a normal phenotype with apparently balanced rearrangements while de novo cases usually are unbalanced or apparently balanced but with associated multiple anomalies as well as mental retardation.

Material and methods: A couple with fertility problems was investigated. He had oligozoospermia and Charcot–Marie–Tooth disease.

Results: Chromosomal analysis and fluorescence in situ hybridization with whole chromosome paint revealed that he had apparently balanced translocation between chromosome 2, 7 and 14. Array comparative genomic hybridization was normal.

Conclusion: To our knowledge, this is a new case of CCR identified in the human population and for the first time a CCR is identified in a patient with CMT.

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Abstract – WCN 2013

No: 2537

Topic: 7 – Neuromuscular disorders

Global proprioceptive resonance: Effects on neuromuscular and postural systems

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Background: The application of mechanical multifocal vibration (MFV) at targeted frequencies and short duration produces positive effects on bone structure, muscles and joints regulating neuromuscular response.

Objective: The purpose of this study was to investigate the effects of the global proprioceptive resonance (GPR) by MFV on muscle performance and body balance in healthy subjects.

Materials and methods: Sixty volunteers (26 males and 24 females, aged 19–25 years) underwent, in a randomized order, both the electromyography–electrogoniography (EMG–EGN) and stabilometry before the GPR and immediately after it. GPR was the ergonomic structure used in this protocol: it gives a psycho-physical release thanks to multifocal vibrations.

Results: The effects of GPR on the surface EMG of masseters and anterior temporalis muscles did not induce any statistically significant change, except for masseter muscles ($p < 0.05$). The results showed a significant improvement in the neuromuscular activity. Muscular activity mainly decreased, while the Freeway Space increased in 53% of cases: this was the evidence of a muscular release after GPR. From a postural point of view, there was an improvement in the load distribution and in the position of the barycenter according to the ideal axis. There were effects in body balance tests, too ($p < 0.05$).

Conclusions: In this preliminary study it was concluded that the GPR induced changes both in neuromuscular and in postural tests. Further and future studies should focus on evaluating the effects on orthodontic and temporomandibular disease patients, as well as the long term effects.

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Abstract – WCN 2013

No: 2519

Topic: 7 – Neuromuscular disorders

Prevalence of neuromuscular disorders in Italian Navy scuba divers: Personalized vs standard mouthpiece

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Background: Scuba divers deal with an extreme psychophysical effort during their activities showing neuromuscular, postural and craniomandibular disorders.

Objective: The aim is to determine the prevalence of temporomandibular disorders and the neuromuscular system variations in scuba divers of the Italian Navy with commercial (CM) and personalized

mouthpiece (PM), and to identify the risk factors for the development of TMD signs and symptoms before and after diving.

Material and methods: 40 males (23–30 years) undergone to neuromuscular tests, spirometric and postural exams before and after diving, and MRI and CBCT exams too.

Results: The prevalence of TMD symptoms after diving was about 93% with CM and 38% with PM versus a 32% before diving; with CM use the neuromuscular answers after diving (T3) were highly different from rest situation before immersion (T0) ($p < 0.001$); also postural aspects were different after diving ($p < 0.01$); with PM use there were no changes.

Conclusions: Scuba divers exhibiting TMD-related symptoms have met the greatest risk of developing TMJ dysfunction during and after the dive. Also in asymptomatic divers, before the dive, TMD was a common problem after it. Customized mouthpiece has reduced symptoms, significantly.

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Abstract – WCN 2013

No: 2553

Topic: 7 – Neuromuscular disorders

Increased prevalence of malignancy in mitochondrial disorders

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Objectives: There are indications that patients with a mitochondrial disorder (MID) develop more frequently malignomas or benign tumours than the general population.

Aims: To find out if the prevalence of tumours is increased in MID-patients and which of the malignomas or benign tumours are the most frequent.

Methods: Retrospectively evaluated were the charts of MID-patients for the presence of malign or benign tumours. MID was diagnosed according to the modified Walker-criteria.

Results: Among 475 MID-patients screened for tumours, at least a single malignoma was found in 65 patients (13.7%), and at least a single benign tumour in 35 patients (7.4%). Among those with malignancy, 22 were male and 43 were female. Among those with a malignancy 1 had definite MID, 9 probable MID, and 54 possible MID. The most common of the malignancies found in MIDs was breast cancer, followed by dermatological, gynaecological, and gastrointestinal malignancies. The most frequent of the benign tumours found in MID patients was the lipoma, followed by pituitary adenoma, meningiomas, carcinoids, and suprarenal adenomas. Compared to the general population, the prevalence of malignancies and of benign tumours was markedly increased. The female preponderance among those with a malignancy was explained by the frequent maternal inheritance of MIDs.

Conclusions: Adult patients with a MID, particularly females, carry an increased risk to develop a malignancy. Since malignancy is an important determinant for their outcome, these patients should be more accurately screened for neoplasms, not to overlook the point, at which an effective treatment can no longer be provided.

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Abstract – WCN 2013

No: 2561

Topic: 7 – Neuromuscular disorders

Monoclonal gammopathy of undetermined significance presenting with acute polyneuropathy

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Background: Monoclonal gammopathy of undetermined significance (MGUS) is a condition where protein M is generated by abnormal plasma cells. Polyneuropathy is present in 5–28% of the patients diagnosed with MGUS.

Objective: Our aim was to present a case with detected IgG paraproteinemia after sudden onset of walking disorder.

Patients and methods: A 61-year old female patient presented with difficulty in walking that started 2-months earlier, later developed numbness and loss of strength in her hands. Fifteen days later her walking completely impaired after which she became bedridden. On the blood workout her Hgb was 5.5 and 5 units of erythrocyte suspension was administered. On the neurological examination arrival, quadriparesia was present, especially on the extensor muscles of all four limbs, being more severe on the lower limbs. Hypoesthesia on the distal parts of all four limbs was present, DTRs were globally abolic and foot sole responses were bilaterally indifferent.

Results: The EMG performed was consistent with acute, severe sensorimotor polyneuropathy, more evident on the lower limbs. On the lumbar puncture, CSF protein level was –27. On the protein electrophoresis gammopathy was detected. On the serum immunofixation electrophoresis IgG-paraproteinemia was detected. The patient was started on IVIG.

Conclusion: MGUS is generally considered as a reason for chronic polyneuropathies and it is very rarely seen as an acute polyneuropathy. The possibility of gammopathies should be considered and protein electrophoresis was performed in the acute onset of polyneuropathies in the elderly. These types of symptoms might be the early sign of malignant diseases.

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Abstract – WCN 2013

No: 2590

Topic: 7 – Neuromuscular disorders

The attention to Duchenne muscular dystrophy

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Background: Duchenne muscular dystrophy (DMD) is a major genetic neuromuscular disease in childhood. Death eventuates commonly by the beginning of the third decade of life. Early diagnosis involves mainly a high degree of clinical suspicion, leading to biopsy or genetic assessment for confirmation.

Objective: This article provides a brief view on the state of this disease in Brazil, focusing on current diagnostic approach and the main challenges on the attention to patients with neuromuscular disorders.

Material and methods: We selected articles indexed in MEDLINE, PubMed, LILACS, BIREME, SCIELO and master degree and doctorate research databases looking for the average age of diagnosis, specialized clinical centrals and the general population knowledge of the disease.

Results: There is a severe delay on DMD diagnosis in Brazil, from 4 to 9 years after the first symptoms. In the whole country only two university hospitals had outpatient clinics solely dedicated to the care of DMD and less than 5% of the general population had heard about DMD.

Conclusion: The reported situation is caused by poor lay education, lack of genetic counseling services, professional unpreparedness and low investments in researches among others. Generally, such scenario does not differ from worldwide – with some particularities, though – and generates a costly care when DMD patients need respiratory aid. However, important progress is expected as methods of spreading

essential information and new specialized centers are developed in the country.

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Abstract — WCN 2013

No: 2632

Topic: 7 — Neuromuscular disorders

Are we really closer to improving the diagnostic certainty in ALS patients with Awaji criteria?

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Background: Amyotrophic lateral sclerosis (ALS) is a lethal disorder characterized by loss of upper and lower motor neurons. The Awaji criteria equate the diagnostic significance of neurogenic electrophysiological changes to the clinical signs of lower motor neuron dysfunction. They also increase the significance of fasciculation potentials (FPs). The aim of our study was to analyze whether the new criteria improve diagnostic certainty in ALS patients.

Material and methods: Medical records of 160 consecutive ALS patients who underwent electrophysiological examination of at least three anatomical regions were chosen for analyzing. The group consisted of 56.2% patients with classic ALS, 12.5% with progressive bulbar palsy, 3.1% with primary lateral sclerosis, 15.6% with progressive muscle atrophy and 12.5% with flail limb syndrome.

Results: Introduction of the FP aspect of Awaji criteria increased the number of electrophysiological involved muscles in 27.5% of patients and the regions involvement in 21.8% of cases. However, it was only able to improve diagnostic certainty of the ALS diagnosis in 1.25% of patients. The equalization of EMG findings with clinical symptoms increased the level of the diagnosis certainty in 5.6% of cases and decreased it in 3.75% of patients. Altogether, the use of Awaji criteria enabled to change diagnosis certainty in only 9.3% of ALS patients.

Conclusions: Awaji modifications are able to improve the diagnostic certainty in ALS diagnosis in some cases. The ratio for patients with improvement of diagnosis sensitivity depends on the clinical phenotype and is higher in cases with preferential upper motor neuron involvement.

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Abstract — WCN 2013

No: 1660

Topic: 7 — Neuromuscular disorders

A sporadic case of late-onset familial amyloid polyneuropathy with a monoclonal gammopathy

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Introduction: The Portuguese type of Familial amyloid polyneuropathy (FAP) may present with sensory-motor and autonomic neuropathy, cardiomyopathy, nephropathy and ocular involvement. Disease onset is usually before the age of 40.

Case report: A 77-year-old Portuguese female presented with burning and pain starting in the feet and progressing proximally, fatigue, anorexia, weight loss and diarrhea. These clinical features were gradually progressive since the age of 65. There was no family history of neurological disease. Examination showed sensory loss in a stocking-glove distribution and distal weakness, the tendon reflexes were abolished and the patient had difficulty in walking. Electromyographic testing revealed a severe axonal sensorimotor peripheral neuropathy. A monoclonal protein was identified in serum electrophoresis. The bone

scanning showed cardiac uptake of 99 m Tc-HMDP. Ophthalmological examination detected probable amyloid deposits in the right vitreous. The echocardiogram showed severe left ventricular hypertrophy. A diagnosis of primary amyloidosis (AL) was initially considered but the slow progression of the disease prompted screening for a pathologic transthyretin (TTR) mutation. The biopsy of abdominal fat pad did not reveal amyloid deposition and the genetic testing confirmed Val30Met TTR-FAP.

Conclusion: FAP has a clinical heterogeneity with respect to age at symptom onset and penetrance. The diagnosis of late-onset cases with no family history may be challenging. The coexistence of a monoclonal gammopathy with an axonal neuropathy does not necessarily imply a definitive diagnosis of AL amyloidosis as it can occur in a small proportion of patients with FAP.

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Abstract - WCN 2013

No: 2582

Topic: 7 - Neuromuscular disorders

A clinical analysis of 20 cases of chronic n-hexane intoxication

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Background: Chronic n-hexane exposure can result in n-hexane intoxication which is mainly characterized by a series of manifestations of peripheral nerve lesions. The disease was sometimes misdiagnosed as “unknown multiple peripheral neuropathy”.

Methods: Twenty cases of n-hexane intoxication were colligated between January 2000 and February 2013 in the National Institute of Neurology of Tunis.

Results: The mean age was 22 years (extremes: 11, 54 years), all men. Nineteen were glue-sniffer and one was n-hexane worker. Period of intoxication ranged from 9 months to 22 years. Ten patients suffered from limb weakness. Neurological examination showed peripheral neurogenic syndrom in eight patients associated with hypoesthesia in the lower limb in ten patients or with position errors in two patients. A cerebrospinal analysis was practiced in five cases and was normal in four cases or showed a high level of protein in one case. Electromyogram was practiced on thirteen patients. It showed axonal polyneuropathy in three cases, demyelinating polyneuropathy in two cases and axonal demyelinating polyneuropathy in eight cases. Neuromuscular biopsy was practiced on five patients. It showed giant axone in three cases. Neurogenic muscular affection and demyelinating neuropathy in one case and axonal neuropathy in one case. All patients were treated by vitamins and rehabilitation.

Conclusion: Diagnosis should be made according to the history of n-hexane exposure, the typical clinical manifestations of peripheral neuropathy. Therapeutic measures for peripheral neuropathy of other etiologies may be used and the prognosis is optimistic if correct diagnosis is made and further exposure stopped.

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Abstract - WCN 2013

No: 2511

Topic: 7 - Neuromuscular disorders

Clinical manifestations of chronic inflammatory demyelinating polyradiculoneuropathies: A retrospective study of forty cases

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Background: Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a rare peripheral neuropathy often leading to severe neurological disability.

Patients and methods: We conducted a retrospective study in forty patients, hospitalized between 2000 and 2012 at the National Institute of Neurology of Tunis and fulfilling the criteria of definite CIDP.

Results: The age of onset of the disease varied between 5 and 79 years with an average of 47 years. There was a male predominance with a sex-ratio of 2.73. Most patients presented with progressive lower limb weakness. Neurologic examination showed motor deficit in most patients (77.5%) and proprioceptive ataxia in 15% of patients. Cerebrospinal analysis showed a high level of protein in twenty-five cases (62.5%) and was normal in eight cases (20%). Electroneuromyogram showed a demyelinating neuropathy in 34 cases and a mixed polyneuropathy in six cases. Neuro-muscular biopsy was practiced in eight patients (20%) and showed demyelinating features in six cases (5%). Most patients had a good response to corticoids and immunosuppressive agents.

Conclusions: CIDP have various clinical and neurophysiological presentations which are important to recognize as they respond well to treatment.

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Abstract - WCN 2013

No: 2565

Topic: 7 - Neuromuscular disorders

Motor form of chronic inflammatory demyelinating polyradiculoneuropathy with conduction block: Clinical, electrophysiological and outcome study of 6 cases

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Background: Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a progressive or relapsing and remitting paralyzing illness. They are usually characterized by motor weakness of the limbs, sensory disturbances and distal areflexia. The protein concentration level in CSF is often high, neurophysiological and histological study finds signs of segmental demyelination. Besides the classical forms of CIDP, there are other clinical variants. Some of them are purely motor and can be a continuum of the motor neuropathy with conduction block.

Methods: We report 6 cases of mainly motor CIDP with clinical and electrophysiological study. Those 6 patients were recruited from between 1997 and 2011 among 50 patients with CIDP.

Results: There are 4 males and 2 females, of 7 to 50 years old. Within the electrophysiological study, signs of demyelination with conduction blocks were diagnosed in all cases. We noticed a high protein concentration level in CSF among 4 patients.

The inflammatory and immunological blood tests were normal for all patients.

Under 1 mg/kg/j oral corticosteroids: prednisone or methylprednisolone. Outcome was favorable in all cases. The follow-up duration is between 5 months and 6 years.

Conclusion: Usually in the literature, the first-line treatment for motor form of CIDP would be the IVIG. The particularity among our patients is the positive outcome reached under corticosteroids treatment thus this regime could be the first-line treatment for the motor forms of CIDP.

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Abstract - WCN 2013

No: 390

Topic: 7 - Neuromuscular disorders

VIII cranial nerve involvement and peripheral neuropathy in patients with Fabry disease

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Fabry disease (FD) is an X linked lysosomal storage disorder caused by deficiency of the — galactosidase A, affecting frequently the nervous system.

Our aim was to evaluate the frequency and characteristics of neuro-otologic and peripheral nerve involvement in the same group of patients.

We prospectively examined 36 consecutive adult patients with FD (11 males, mean age 34.7 years old, range 18–60; 25 females, mean age 36.1 years old, range 18–73).

Clinically 78% of patients reported acroparesthesias and 76% neuro-otological symptoms. In our patients 78% presented abnormal QST; within this subgroup 93% showed increased CDT and 43% had altered WDT.

The inferior division of the vestibular nerve (assessed by VEMPs) was abnormal in 45% of the patients. The superior division of the vestibular nerve (assessed by VNG) was abnormal in 51%. Audiometry showed sensorineural hearing loss in 58%. Neuro-otologically 90% of the patients presented a lack of neural or vascular distribution of their abnormalities known as “no Pattern”.

Conclusions: We identified symptoms of neuropathy in both heterozygous and hemizygous patients. The neuropathy predominantly affects A-delta and C small fibers.

The neuro-otological abnormalities, distributed without a vascular or neural pattern, were not only very common but also highly suggestive of scattered inner ear involvement.

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Abstract - WCN 2013

No: 2522

Topic: 7 - Neuromuscular disorders

CMAP parameters in the diagnosis of classic form CIDP, MADSAM, and DADS: Usefulness of Erb's point stimulation

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Background: CIDP is currently classified into several subtypes: classic form CIDP, multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), and distal acquired demyelinating symmetric neuropathy (DADS). In detecting these subtypes, it is often challenging for neurologists because motor conduction studies (MCS) are insensitive or nonspecific. Early diagnosis leading to prompt therapy may result in improved patient outcomes.

Objective: To determine the compound muscle action potential (CMAP) parameters along the motor nerve that best reveal the abnormalities in patients with classic form CIDP, MADSAM, and DADS.

Patients and methods: Subjects were 20 patients with classic form CIDP, 18 patients with MADSAM, 6 patients with DADS, and 35 normal controls. MCS of ulnar nerve were performed using standard surface electrodes for stimulation and recording. The ulnar nerve was stimulated on the wrist, below elbow, above elbow, arm, and Erb's point. Reference values of motor latency, duration, negative area, and negative peak amplitude of CMAP were calculated using 35 control

data. The frequency of an abnormal finding was calculated for the different CMAP parameters for each subtype.

Results: In classic form CIDP and MADSAM, CMAP parameters obtained from Erb's point stimulation were more often abnormal than CMAP parameters obtained from elbow or wrist stimulation. In MADSAM, CMAP parameters in general were less abnormal. In DADS, CMAP parameters revealed abnormality in 100% of the patients.

Conclusion: We suggest that CMAP parameters of Erb's point stimulation should be used to define the conduction along the entire motor nerve in detecting CIDP.

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Abstract - WCN 2013

No: 2441

Topic: 7 - Neuromuscular disorders

The follow-up of the patients with GBS treated with plasmapheresis

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Background: GBS is an acute polyneuropathy, a disorder affecting the peripheral nervous system. It can cause life-threatening complications, in particular if the respiratory muscles are affected or if there is autonomic nervous system involvement. The disease is usually triggered by an infection. Treatment consists of attempting to reduce the body's attack on the nervous system by plasmapheresis.

Objective: To evaluate the efficacy of PEX in GBS patients.

Methods: In our prospective study a group of 17 patients with GBS was evaluated before, immediately after PEX and 1 year later. The evaluation was made using the Hughes functional grading scale for GBS (from Hughes et al., Lancet 1978).

Results: We have evaluated the 17 patients. We have found an improvement in 14/17(82.35%) patients immediately after and 1 year after the PEX treatment. We haven't found an improvement in 3/17 patients. In 4 patients the progression of the neurological signs was stopped immediately after the first PEX procedure and in these patients we have found better results. We have found good results even in patients who require assisted ventilation (score 5 of Hughes functional grading scale for GBS). 3/17(17.64%) patients had an improvement even they haven't made all the PEX procedure because of complications.

Conclusions: The PEX treatment in GBS patients is efficient. The improvement consisted of good clinical outcome. In some cases the progression of clinical signs was stopped even after only one procedure.

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Abstract - WCN 2013

No: 1004

Topic: 7 - Neuromuscular disorders

Temporal cortical damage as a prognostic marker in ALS (amyotrophic lateral sclerosis)

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Background: ALS is a devastating neurodegenerative disorder of motor neurons. Prognostic biomarkers are urgently needed in ALS, and MRI studies are a promising candidate.

Objective: To find a MRI-based prognostic marker in ALS.

Patients and methods: 35 patients with ALS were evaluated with the ALSFRS-r scale and underwent brain 3T MRI scans. Seventeen patients were clinically reevaluated after 6.6 months. Cortical thickness and gray volume estimation were performed using the Freesurfer image analysis suite (<http://surfer.nmr.mgh.harvard.edu/>). A control group of 35 matched controls was used for comparison. Statistical analysis was performed using SigmaPlot version 12 (Systat Software, San Jose, CA) (uncorrected $p < 0.05$) and the Freesurfer suite (uncorrected $p < 0.0001$). Cortical thickness and gray volumes were compared between groups using ANCOVA, adjusting for subject's age.

Results: Patients presented cortical thinning of the precentral ($p = 0.004$), superior temporal ($p = 0.017$) and supramarginal ($p = 0.035$) cortices of the left hemisphere and paracentral ($p = 0.020$), precentral ($p = 0.001$) and supramarginal ($p = 0.050$) cortices of the right hemisphere. They also presented reduced gray volumes of the middle temporal ($p = 0.010$), postcentral ($p = 0.009$) and precentral ($p = 0.001$) cortices of the left hemisphere and paracentral ($p = 0.001$), postcentral ($p = 0.046$) and precentral ($p = 0.00032$) cortices of the right hemisphere. ALSFRS-r scale variation was associated to cortical thickness at the fusiform, entorhinal, temporal superior, middle and inferior gyri (right hemisphere) and temporal superior and inferior gyri (left hemisphere).

Conclusion: Patients with ALS have motor and non-motor cortical damage. Temporal cortex thinning is associated with faster disease progression, and might be a prognostic marker in ALS.

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Abstract - WCN 2013

No: 2706

Topic: 7 - Neuromuscular disorders

Autosomal recessive cerebellar ataxia: A clinical and genetic study

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Background: Autosomal recessive cerebellar ataxia (ARCA) contains more than 20 clinical entities and an even larger number of associated genes.

Objectives: To determine the frequency of the different ARCA forms.

Patients and methods: We studied 188 cases belonging to 117 families suspected of ARCA, coming from different regions of Algeria, between 2001 and 2012. Complete clinical examination, laboratory, imaging and electrophysiological investigations allowed us to obtain a detailed phenotype for each patient. The initial molecular investigation was the FRDA GAA expansion test. The non-Friedreich patients were then explored genetically according to their phenotype.

Result: The molecular diagnosis could be established in 67% of ARCA patients (126 patients): 52 patients were affected with Friedreich's ataxia (FRDA), 21 AOA2, 16 AVED, 12 AT, 9 ARSACS, 9 Joubert syndrome, 5 ARCA2, and 2 AOA1. Sixty two patients had no identified mutation.

Conclusion: This study in a large Algerian cohort of ARCA patients allowed us to determine the frequency of 8 ARCA entities. Friedreich's ataxia was the most frequent entity, as found in the majority of studies, followed by AOA2 and AVED. Ataxia with vitamin E deficiency with a founder effect mutation in North-Africa is one of the few ARCA that can be treated and therefore must be looked for in first instance after FRDA in Friedreich-like patients. Two families contributed to identify ARCA2, a new cerebellar ataxia entity. However 32% of ARCA patients remained not linked to known genes, such that new ARCA forms are expected in the near future.

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Abstract - WCN 2013**No: 2699****Topic: 7 - Neuromuscular disorders****Peripheral neuropathy in neurofibromatosis**M. Poloni^a, R. Barbò^b. ^aNeurosciences, HPG 23, Bergamo, Italy; ^bNeurosciences, ICH Gavazzeni, Bergamo, Italy

Neurofibromas are benign tumors which originate from peripheral nerves and are, together with others pathognomonic symptoms, a common finding in Neurofibromatosis 1 (NF1) conversely Neurofibromatosis 2 (NF2) or central neurofibromatosis is characterized by abnormal growth of Schwann cell tumors called schwannomas (SCH) at vestibular nerves, beyond meningiomas, ependimomas and SCHs of other cranial nerves or different Central Nervous System (CNS) tumors. NF1 is a genetic disorder transmitted thorough chromosome 17 due to neurofibromin alterations and abnormal tumors growth with rare Malignant Peripheral Nerve Tumors (MPNT), while NF2 is linked to merlin abnormalities and is at same genetically transmitted but through chromosome 22.

Peripheral neuropathy (PN) appears rarely in NF1, ranging from 0 to 4.3 % of patients in different casistics, while retains only instrumental evidence in NF2 where mononeuropathy, generally as a foot drop or a facial palsy, is relatively common.

Three interesting cases of important and distressing peripheral neuropathy, one affected by NF1 and two brothers affected by NF2, have been seen and studied at the NF Ambulatorial Center of Bergamo: we want to share the clinical and instrumental characteristics of these patients, discussing the differential diagnosis and arguing the different pathogenetic mechanisms.

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Abstract - WCN 2013**No: 2686****Topic: 7 - Neuromuscular disorders****Facial onset sensory and motor neuropathy: A neurodegenerative TDP-43 proteinopathy?**E.P. Bosch^a, B.P. Goodman^a, J.A. Tracy^b, P.J.B. Dyck^b, C. Giannini^c. ^aNeurology, Mayo Clinic Arizona, Scottsdale, AZ, USA; ^bNeurology, Mayo Clinic Rochester, Rochester, MN, USA; ^cPathology, Mayo Clinic Rochester, Rochester, MN, USA

Background: Vucic et al. (Brain, 2006) described four adult males with facial sensory loss gradually spreading to cervical and brachial dermatomes associated with LMN weakness of cranio-bulbar and upper extremity (UE) distribution. Autopsy suggested a neurodegenerative process affecting motor neurons and sensory ganglia.

Objective: We report clinical, electrophysiologic and pathologic features of six new cases of facial onset sensory and motor neuropathy or FOSMN syndrome.

Design and methods: A retrospective review of patients referred to Mayo Clinic from 2004 to 2012 identified six males who met the clinical features of FOSMN.

Results: The age of onset ranged from 55 to 63 years. The presenting symptoms, years after onset, were facial numbness (5), dysphagia (3), and masseter weakness (2). Sensory loss spread to scalp, neck, upper torso and distal UEs. Patients developed facial, masseter, bulbar, neck flexor/extensor and LMN weakness of UEs. Four patients died of complications 6 to 9 years after onset. Electrodiagnostic studies revealed blink reflex abnormalities, reduced sensory nerve action potentials in UEs, and chronic denervation mainly in cranial and cervical regions. Biopsies of greater auricular nerves revealed low grade axonal degeneration. One patient came to autopsy: Neuronal loss and gliosis with TDP-43 positive cytoplasmic neuronal

and glial inclusions were present in hypoglossal nucleus and cervical motor neurons.

Conclusion: FOSMN syndrome is a rare, progressive disorder affecting sensory and motor neurons in a cranio-caudal descending distribution. FOSMN syndrome joins sporadic ALS, frontotemporal lobar degeneration (FTLD-U) and other neurodegenerative disorders as a TDP-43 proteinopathy.

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Abstract - WCN 2013**No: 760****Topic: 7 - Neuromuscular disorders****Vulnerability of thenar muscle and sensory neuropathy in patients with amyotrophic lateral sclerosis (ALS)**C.-H. Kim^a, S.-Y. Kwon^b. ^aPhysical Medicine & Rehabilitation, Inha University, Incheon, Republic of Korea; ^bPhysical Medicine & Rehabilitation, Inha University Hospital, Incheon, Republic of Korea

Background: Among them thenar muscles were weakened earlier in amyotrophic lateral sclerosis (ALS).

Objective: To figure out whether that happens due to the early vulnerability of median nerve or susceptible overlapping peripheral neuropathy such as carpal tunnel syndrome (CTS).

Patients and methods: We selected 35 cases of ALS patients who had full electrophysiologic data and 50 age-matched control cases. We excluded pediatric patients and patients with incomplete medical records. Mean age of ALS patients was 61.7 ± 11.1 years, and that of control was 53.5 ± 13.9 years.

Results: In control subjects, comparison of median to ulnar CMAP amplitudes showed no statistical difference (median 7.7 ± 1.8 mV, ulnar 7.7 ± 1.7 mV, $p = 0.00$). Also, CNAP amplitudes showed a statistical difference (median 41.5 ± 18.5 μ V, ulnar 36.5 ± 17.3 μ V, $p = 0.04$), whereas sensory nerve conduction velocity (NCV) showed no significant difference (median 45.5 ± 4.0 m/s, ulnar 45.6 ± 7.6 m/s, $p = 0.95$) by T test.

In ALS patients, comparison of CMAP amplitude (median 3.9 ± 2.7 mV, ulnar 5.5 ± 2.4 mV, $p = 0.00$) and MDL showed significant difference (median 4.1 ± 0.9 ms, ulnar 3.1 ± 0.5 ms, $p = 0.00$) by T test. In contrast, comparison of CNAP amplitudes showed no statistical difference (median 22.0 ± 11.4 μ V, ulnar 19.9 ± 8.8 μ V, $p = 0.37$), and so did the sensory NCV (median 41.5 ± 7.6 m/s, ulnar 42.0 ± 5.9 m/s, $p = 0.58$) by T test.

Conclusion: We reconfirm that thenar muscles were more vulnerable than hypothenar muscles in ALS patients.

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Abstract - WCN 2013**No: 2664****Topic: 7 - Neuromuscular disorders****Dermatomyositis in elderly, disease that should be recognized – case report**M. Perovska^a, K. Majstorovic^b, A. Arsovska^c. ^aUniversity Clinic of Neurology Skopje, Ohrid, The Former Yugoslav Republic of Macedonia; ^bInstitute of Nephrology Struga, Ohrid, The Former Yugoslav Republic of Macedonia; ^cUniversity Clinic of Neurology Skopje, Skopje, FYROM – The Former Yugoslav Republic of Macedonia

Our aim was to present late onset of dermatomyositis as a chronic autoimmune condition, importance in differential diagnosis with other conditions and the possibility of underlying malignancy.

A 70 year old male was presented with history of symmetrical, proximal muscle weakness, muscle pain, cough, reduced appetite, remarkable weight loss, constipation, and erythematous forehead lesion. Clinical symptoms had subacute course three months prior to hospitalization, initially with erythematous forehead lesion, cough and constipation. The patient had noticed difficulty in climbing the stairs and a month later proximal upper limb weakness in difficulty overhead activities. The muscular weakness was gradually progressive. The patient has uncontrolled diabetes mellitus and he was complaining about hypesthesia in distal parts of lower limbs. Series of tests were carried out prior to hospitalization: complete blood count; EMG; LP; prostigmin test; cervical MRI; SEP; thyroid profile; and PSA. Bedside examination revealed erythematous forehead lesion, inspiratory crackles in the lung bases; proximal, symmetrical muscle weakness, muscle pain on grasping, generalized hypotony, proximal hypotrophy of upper limbs, and generalized areflexia. No fasciculations were seen in any of the muscle. We have made complete blood count; EMG; SEP; ocular fundus exam; chest radiography; and ECG.

With the results our case satisfied all the criteria for the definite diagnosis. Improvement was seen after we began the treatment with high-dose corticosteroids, supportive and symptomatic therapy.

It is essential to recognize early manifestations, investigate and differentiate dermatomyositis, initiate the therapy early, and search for malignancy. Further investigation is necessary.

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Abstract - WCN 2013

No: 2573

Topic: 7 - Neuromuscular disorders Cryoglobulinaemia and peripheral neuropathy

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Background: Cryoglobulinaemia is a systemic inflammatory condition characterised by immune complex-mediated small-to-medium-sized vasculitis. The most common type described in the literature is secondary to hepatitis C. It has a wide variety of presentations ranging from bruising, neuropathy, and hepatosplenomegaly to acute renal failure.

Objective: Here we described two cases of cryoglobulinaemia and peripheral neuropathy without HCV.

Patients: A 46 year old female and a 29 year old male without pathological history developed paresthesias, painful sensations and progressive distal motor weakness in the lower limbs. Neurological examination of the patients showed distal weakness and areflexia in lower limbs associated with superficial and proprioceptive sensory disturbance. Nerve conduction studies were suggestive of axonal sensorimotor neuropathy. Cutaneous muscular nerve biopsy showed a mild axonal neuropathy. Laboratory studies showed a detection of serum mixed (IgG–IgM) cryoglobulins in the first case and the presence of monoclonal cryoglobulins type IgG in the second, and excluded others' autoimmune and hepatitis markers. Our patients were treated and subsequently improved with only oral steroids in the first case, and with both oral steroids and immunosuppressive treatment in the second.

Conclusion: Neuropathy in our cases was the initial manifestation of mixed cryoglobulinaemia which was seen in half of the patients in the literature. Previous works reported that the peripheral neuropathy was more common with essential mixed cryoglobulinaemia and was generally of sensory type. Cryoglobulinaemia must be looked for in case of neuropathy.

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Abstract - WCN 2013

No: 1592

Topic: 7 - Neuromuscular disorders Agrin–Lrp4–MuSK signaling in passively transferred experimental myasthenia gravis rat

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Objective: Passively transferred experimental autoimmune myasthenia gravis (p-EAMG) rats can be induced by injecting serum from an AChR-immunized chronic EAMG rat. The present study aimed to clarify Agrin–Lrp4–MuSK signaling in the neuromuscular junction (NMJ) *in vivo* using immunohistochemical methods.

Methods: Injection of 250 µL of anti-AChR antibody-positive rat sera into normal female Lewis rats (n = 6) induced severe signs of p-EAMG including weight loss and AChR deficiency in the NMJs. By contrast, injection of 50 µL of the sera into normal rats (n = 6) induced mild signs of p-EAMG with no significant weight loss and AChR deficiency in the NMJs. Using the p-EAMG rats, expression levels of MuSK and Lrp4 proteins at the NMJ were analyzed by an immunohistochemical method.

Results: In the NMJs of p-EAMG rats with mild signs, high signal intensity of MuSK and Lrp4 was detected, but not in p-EAMG rats with severe signs and wild-type of rats.

Conclusions: Our data suggest that expression of Lrp4 and MuSK is up-regulated to oppose a disorder of neuromuscular function caused in rats with mild signs of p-EAMG.

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Abstract - WCN 2013

No: 2726

Topic: 7 - Neuromuscular disorders Myasthenia gravis and facioscapulohumeral muscular dystrophy (FSHD) co-existing in one patient – A case report

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Introduction: Some case reports describe co-existence of myasthenia gravis and facioscapulohumeral muscular dystrophy (FSHD) in one patient. It is unclear, if the association is coincidental or not. There is evidence that muscle fibre degeneration can cause innate immune responses and leads to the generation of autoantibodies against muscle proteins.

Case report: We describe a 66-year old male with onset of weakness of the proximal upper limb muscles at age 40. Family history was not available. At this time electrophysiological workup revealed myopathic changes in the upper extremities and muscular biopsy of the left deltoid muscle showed a neurogenic atrophy.

At the age of 45 the patient complained about a ptosis of the right eye and diplopia. A seropositive generalized myasthenia gravis was diagnosed and the patient underwent thymectomy.

On cyclosporine treatment the patient remained symptom free until the age of 61.

At this time neurological examination revealed moderate muscle weakness and atrophy of the scapulohumeral region. Creatine kinase was elevated between 200 and 300 U/l. A myopathic pattern of the muscles of the upper limb was found in electromyography.

Re-biopsy of the right deltoid muscle showed again a neurogenic atrophy.

Genetic testing on FSHD confirmed the diagnosis (10 copies of D4Z4-repeats).

Conclusion: In our case we found a coincidence of two diseases because ptosis and diplopia are not typical for FSHD and we observed an excellent response to immunosuppressive agents for the myasthenia related symptoms.

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Abstract - WCN 2013

No: 2782

Topic: 7 - Neuromuscular disorders

Mycosis fungoides and CIDP: A case report

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Background: Mycosis fungoides or granuloma fungoides, is the most common form of cutaneous T-cell lymphoma. It generally affects the skin, but may progress internally over time. Symptoms include rash, tumors, skin lesions, and itchy skin.

Most cases are sporadic and occur in people over 20 years of age, and it is more common in men than women. Treatment options include sunlight exposure, ultraviolet light, topical steroids, chemotherapy, and radiation.

Case report: A 53 year old male with history of Mycosis fungoides since 10 years PTA, came for his subacute progression of proximal muscle weakness that occurred 2 months before admission. Neurological examination revealed areflexia, proximal and distal muscle weakness, stokes and glove sensory disturbance, and loss of position sense. Electrophysiology study showed sensory motor polyneuropathy with conduction block without spontaneous activity. All lab tests for vasculitis and gammopathies were normal. Lp was done that showed elevated protein (90 mg/cc). The patient was diagnosed as CIDP and IVIG therapy was started.

Conclusion: Some complications like skin cancer, melanoma, colon cancer, Hodgkin lymphoma, and non-Hodgkin lymphomas have been reported but if polyneuropathy is a rare complication of disease or it is a concomitant state is still unclear.

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Abstract - WCN 2013

No: 2866

Topic: 7 - Neuromuscular disorders

Surface electromyography study of complete spinal cord injury patients. Preliminary results

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Introduction: Frankel A spinal cord injury patients have three different spinal cord functional segments detected by clinical examination: 1. – Proximal segment with motor and sensitive complete functional preservation until the first damaged metamer. 2. – Medial segment with motor and sensitive incomplete functional preservation. This segment should be not longer than three metamers, and 3. – Distal segment, with motor and sensitive null functionality. Muscular electric activity of these three segments is reported.

Methods: 9 patients with complete spinal cord injury assessed by two independent professionals underwent to a surface electromyogram of muscles innervated by last three metamers of the proximal segment, three metamers of the incomplete segment and three first metamers of distal segment. Registers of voluntary activity were taken with surface electrodes; and patients were awake but resting. Wave amplitudes were studied.

Results: The patients had normal electromyography registers in the proximal segment. Conforming the registers were obtained from muscles innervated for more distant metamers except the last preserved one, they showed progressive number lost of high activity waves, replaced by clusters of low activity waves. This tendency progressed on muscles innervated for the two metamers of distal segment, disappearing any electric activity at muscles innervated by the third metamer.

Discussion: Electrical activity in the two first metamers of the distal segment reveals that there is a subclinical connection between cortex and corresponding second motoneurons. Progressive increasing number of low activity waves supports that there is a diminishing number of motoneurons taken compensatory innervation of denervated miofibrillas.

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Abstract - WCN 2013

No: 2331

Topic: 7 - Neuromuscular disorders

Dermatomyositis in a patient with facioscapulohumeral dystrophy

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Background: The association of Facioscapulohumeral Dystrophy (FSHD) and Dermatomyositis is not described in the literature.

Objective: Case-report of patient with FSHD and Dermatomyositis.

Material and methods: Review of the clinical record.

Results: Female, 23 year-old, with family history of FSHD (mother, aunt and 2 cousins), apparently healthy until September 2011, when she started to develop a progressive tetraparesia, initially with asymmetry. The genetic testing for FSHD was positive. Five months after the initiation of the muscular weakness she presented a grade 2–3/5MRC tetraparesis, including cervical involvement and facial biparesia, and 2 months later she started with dysphagia for liquids. In May 2012 she was admitted to the Neurology Service because of clinical deterioration with dysphagia and loss of gait. The analytical review performed during this hospital admission showed: CK > 3000 IU, ANA1/1280 mottled pattern, Ac antiMi2+, EMG: findings compatible with necrotizing myopathy. A muscle biopsy showed changes compatible with an inflammatory myopathy. The patient suffered aspiration pneumonia leading to invasive ventilation. Concomitantly heliotropic rash and erythroderma developed at the knees and face with malar rash. Given the clinical picture and the results of diagnostic exams, the diagnosis of Dermatomyositis was proposed. She was treated with methylprednisolone, followed by prednisolone and parallel immune globulin with discreet improvement of motor function. Later, methotrexate was added as well as rescue therapy with 5 plasmapheresis sessions, because of the poor clinical evolution. Currently, she is on spontaneous ventilation, with improved distal muscle strength.

Conclusion: The presence of 2 rare diseases in the same individual hindered the diagnosis and delayed the start of immunosuppressive therapy.

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Abstract - WCN 2013**No: 2907****Topic: 7 - Neuromuscular disorders****Taurine ameliorates mitochondrial dysfunction and prevents stroke-like episodes in patients with MELAS**

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In 1966, Francis Crick predicted that the first anticodon (“wobble”) nucleotide recognizes the third codon nucleotide through non-canonical Watson–Crick geometry. To date, post-transcriptional modifications in tRNAs are thought to play critical roles in deciphering the genetic code. In MELAS patients harbouring the A3243G-mutant mitochondrial DNA, post-transcriptional taurine modification at the wobble nucleotide is deficient in the mutant mitochondrial (mt) tRNA^{Leu(UUR)}. Here, we show that addition of taurine to culture media ameliorated reduced oxygen consumption, decreased mitochondrial membrane potential, and increased oxidative stress in MELAS patient-derived pathogenic cells. Moreover, high dose oral administration of taurine (0.25 g/kg/day) completely prevented stroke-like episodes in two MELAS patients for more than nine years. These findings provide a new insight into our understanding of MELAS as putative RNA-modification disorders that lack the wobble taurine modification. The oral administration of taurine may be an effective therapy for MELAS.

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Abstract - WCN 2013**No: 1151****Topic: 7 - Neuromuscular disorders****Peripheral neuropathy (PN) and small fiber (autonomic) dysfunction in patients with Parkinson's disease (PD) and parkinsonism**

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Background: Recent studies have reported that large-fiber PN is common in patients with PD (Gondim, Ann Neurol 2010;68:973).

Objective: To evaluate the prevalence of large-fiber and small fiber neuropathy (PN) in patients with PD and parkinsonism.

Methods: We evaluated the presence of large and small fiber PN in 56 patients with parkinsonism in a tertiary outpatient clinic from Brazil (neurological exam, NCS/EMG and skin wrinkling test, SWT Teoh, JNNP 2008;79:835). The study was approved by the local IRB. Descriptive statistics, t-test and Mann–Whitney were used to compare the groups.

Results: 41 patients with PD (14 women, mean age: 62.7 ± 2.3 years, P < 0.05, mean disease duration: 8.4 ± 0.9 years) and 15 patients with other forms of parkinsonism [6 women, mean age: 48.2 ± 6.3 years, mean disease duration: 4.8 ± 1.2 years, 4 patients with Wilson's disease (WD), 3 with MSA, 3 with PSP/CBGM] completed clinical neuromuscular evaluation. NCS/EMG was performed in 12 patients (4 PD, 3WD, 2 MSA and 1 CBGM). Half of the patients with PD who underwent NCS/EMG had axonal sensorimotor PN, 1 had L5 radiculopathy and normal SWT and the other normal NCS/EMG and SWT. SWT was performed in 41 patients (32 PD, 9 parkinsonism). 18 out of the 32 PD had abnormal SWT while 4 out of 9 patients with parkinsonism had abnormal SWT.

Conclusions: Large fiber and small fiber PN are common in patients with PD, WD and other forms of parkinsonism. SWT may be used as a useful screening tool for evaluation of small fiber dysfunction in patients with PD.

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Abstract – WCN 2013**No: 1599****Topic: 7 – Neuromuscular disorders****Electrodiagnostic findings in a cohort of patients with Inflammatory Bowel Disease (IBD)**

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Background: Patients with IBD may experience a wide range of neuromuscular complications (Gondim, Brain 128:867, 2005).

Objective: To report the electrodiagnostic and clinical findings in a cohort of IBD patients.

Patients and methods: Clinical and electrodiagnostic findings of 51 patients with Crohn's disease (CD), 70 patients with ulcerative colitis (UC) and 50 controls (gastritis/dyspepsia/irritable bowel syndrome) were compared. Descriptive statistics and t-test were used to compare the different groups.

Results: In the CD group, 7 patients had small fiber neuropathy (SFN) and 12 large-fiber PN. Sural SNAPs and peroneal CMAPs were lower in the large-fiber PN group: 2.7 ± 1.6 versus 27.9 ± 3.2 (asymptomatic) and versus 24.5 ± 2.7 (SFN) uV (P < 0.05) and 2.2 ± 0.6 versus 6.2 ± 0.5 (asymptomatic) and 5.1 ± 0.4 (SFN) mV (P < 0.05). Median and ulnar sensory SNAPs and tibial, median and ulnar CMAPs were also decreased (P < 0.05). In the UC group, 8 patients had SFN and 12 large-fiber SM PN. Sural SNAPs and peroneal CMAPs were lower in the large-fiber PN group: 5.9 ± 1.5 versus 27.3 ± 2.3 (asymptomatic) and versus 31.5 ± 3.7 (SFN) uV (P < 0.05) and 3.2 ± 0.6 versus 5.3 ± 0.4 (asymptomatic) and 4.1 ± 0.2 (SFN) mV (P < 0.05). Median SNAPs and tibial, median and ulnar CMAPs were also decreased (P < 0.05). One of the patients also developed myasthenia gravis.

Conclusion: Excluding other contributory factors (medications, low B12, diabetes, hypothyroidism, diabetes or glucose intolerance), 7.1% of the IBD patients had SFN or large-fiber PN probably due to IBD. The most common form of PN in IBD patients is a mild distal symmetric axonal sensorimotor.

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Abstract - WCN 2013**No: 2442****Topic: 7 - Neuromuscular disorders****Seed-based and functional connectivity analyses as a tool for evaluating Amyotrophic Lateral Sclerosis (ALS) progression, a pilot study**

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Background: ALS is a severe neurodegenerative disorder of motor neurons. Candidates for prognostic markers are necessary to follow the course of the disease and possibly guide medical actions.

Objective: Investigate possible fMRI candidates: specific seed-based and functional connectivity analyses from resting state (RS) scanning.

Patients and methods: 30 patients and 24 controls were evaluated with the ALSFRS-r and severity scales. A BOLD signal time series from each RS-scanning of all subjects was obtained using a mask of Brodmann area 4 and the Matlab (MathWorks Natick, Massachusetts) toolbox Pickatlas (Functional MRI Laboratory, Wake Forest University). A correlation map with this signal was calculated for all voxels and all subjects. Correlation maps of control and patient groups were compared by two sample t-test. We also calculated correlation matrices for each subject of every voxel pair of Brodmann area 4. Then we determined a value of connectivity degree. We used General Linear Model to analyze connectivity degree and the clinical variables (ALSFRS-r, severity scale, variation and rate of variation of ALSFRS-r and of severity scale).

Results: We found no significant results in the application of these methods, considering a significance level of 0.01.

Conclusion: Further studies with greater groups may be necessary to confirm if there is no difference between groups analyzed by the cited seed-based and functional connectivity methods. Our results do not support follow-up of patients by the methods we used, nor differentiation between them and individuals from control group.

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Abstract – WCN 2013

No: 3026

Topic: 7 – Neuromuscular disorders

Subdermal needle electrode facilitates recording of lumbar N22 tibial SEP responses

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Background: When the tibial SEP shows a delayed scalp response, the N22 lumbar response helps distinguish if the delay is central or peripheral.

Objective: To determine if a subdermal needle electrode enables recording of N22 lumbar posterior tibial SEP responses when the surface recording shows no response.

Materials and methods: Over a two year period twenty-eight patients undergoing tibial SEP testing who had absent or poorly formed N22 lumbar responses using standard surface recording had the SEP study repeated at the same visit replacing the lumbar surface electrode with a subdermal needle electrode. Most of these patients were eventually found to have a variety of CNS, structural intraspinal or other disorders. Each patient consented to have a lumbar subdermal needle placed in an attempt to bring out the N22 lumbar potential.

Results: A reproducible N22 lumbar response was obtained in 14 of 28 subjects. Patients with lumbar responses were of similar age (mean age 62.0 vs. 59.4 years) and had comparable BMI values (mean value 29.0 vs. 30.4) to those without lumbar responses.

Conclusions:

- 1) A subdermal needle electrode facilitates recording of a reproducible N22 lumbar potential in the posterior tibial SEP study when the surface recording technique shows no convincing lumbar response;
- 2) Identifying a lumbar potential can help localize delayed conduction.

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Abstract – WCN 2013

No: 3029

Topic: 7 – Neuromuscular disorders

Lipid and protein oxidative damage in G93A mice, a model of amyotrophic lateral sclerosis

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Background: Amyotrophic lateral sclerosis is a progressive neurodegenerative disorder. Recent studies guide in question, a multisystem disease with a special predisposition for motor neuron. The SOD1 mutation found in some patients with FALS has been shown to result in an increase in free radical production that may cause systemic oxidative injury. In this study, we analyzed the extent of lipid and protein oxidative damages to the brain, heart, liver, muscle and spinal cord of transgenic FALS mice that overexpress the SOD1 mutation.

Material and methods: We used a colony of G93A transgenic mice divided in two groups: control and diseased both with 50 of 40 days and 50 of 100 days of life. We analyzed the concentrations of MDA-4-HDA and carbonyl, indicators of lipid and protein oxidation.

Results: In the preclinical stage (40 days old) no significant differences were found between levels of lipid or protein oxidation when we compared health versus sick animals. In the clinical phase (100 days old), values of lipid and protein oxidative damage in sick animals were higher in all tissues compared with healthy animals. Significant differences were only found in sick spinal cord lipid peroxidation values (diseased spinal cord 1.42 ± 0.24 vs healthy spinal cord 0.62 ± 0.03 ; Mean \pm SEM; $p < 0.01$).

Conclusions: Our results suggest that overproduction of free radical production caused by the G93A SOD 1 mutation leads to extensive lipid and protein systemic damage, above all in spinal cord, that may be involved in neurodegeneration.

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Abstract – WCN 2013

No: 3015

Topic: 7 – Neuromuscular disorders

Atypical Guillain–Barre syndrome presenting with isolated facial palsy and acral paresthesia

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Bilateral simultaneous facial paralysis is a rare clinical entity, which unlike the unilateral presentation, is seldom secondary to Bell's palsy. A 26 year-old-man presented with a one week history of bilateral facial weakness and acral paresthesia. Neurologic examination revealed bilateral complete lower motor neuron type of facial palsy, loss of deep tendon reflexes and diminished vibratory sensation. But motor examination was completely normal in both upper and lower extremities. Blood tests for full blood counts, urea and electrolytes, serum angiotensin converting enzyme (ACE) and the vasculitic screen were within normal limits. Tests were negative for Lyme disease, Herpes simplex, *Borrelia burgdorferi* and Oligoclonal bands. Lumbar puncture revealed albuminocytological dissociation. Enhancement of the left geniculate ganglion was confirmed by cranial MRI. The diagnosis of AIDP was made and he was initiated on a five day course of 30 g of intravenous immunoglobulin infusions from day 8. Incidence of bilateral facial palsy is about 1 per 5,000,000. The etiology of facial paralysis includes many

conditions such as congenital, traumatic, infectious, neurological, metabolic, neoplastic, toxic, vascular, and idiopathic. GBS presents as a progressive development of palsy of the voluntary muscles. In 27% to 50% of the cases, the facial nerve is involved. Isolated facial diplegia with no limb weakness has been described as a rare GBS variant. Usually in these cases, areflexia helps in distinguishing GBS as the underlying etiology. Prognosis is rather good and therapy consists of plasma exchanges and administration of IV immunoglobulins.

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Abstract — WCN 2013

No: 3023

Topic: 7 — Neuromuscular disorders

Late-onset non-thymomatous generalized myasthenia gravis

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Epidemiological studies have resulted in a change of concepts about the frequency of myasthenia gravis (MG) in the elderly population, with the implication that MG may be quite frequent in later years. In our MG database, there were 132 late-onset MG (LOMG, ≥ 50 years) patients whose symptoms started in the 10 years between 2001 and 2010 and who were followed for at least 2 years. Ninety-five patients had generalized MG without thymoma. Ten patients harbored a thymoma and 27 patients had ocular MG. In the generalized non-thymomatous LOMG group, there were 41 females and 54 males. Seventy-seven percent of the patients had onset before 70 years of age. Onset symptoms were ocular in 61%, bulbar in 17%, in the extremities in 13%; two patients had neck weakness and 7 had mixed onset symptoms. Anti-acetylcholine receptor antibodies (AChR Ab) were positive in 81 patients (85%), 5 were anti-MuSK Ab positive and 9 were double negative. The disease was severe (maximum MGFA-4 and -5) in 14 patients (15%) with intubation (MGFA-5) in 6 AChR Ab positive patients (6%). Forty-nine patients had MGFA-2 and 32 had MGFA-3. Thymectomy was performed in 12 patients. Prednisolone and/or azathioprine were administered in 95%. Sixty-six percent were PR or MM at the last visit. The maintenance prednisolone dose was 8.5 mg/day in these patients as compared to 22 mg/day in those with less well prognosis. In this study group, LOMG appears to be a relatively benign disorder with good prognosis although there are patients with severe symptoms and unfavorable outcome.

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Abstract — WCN 2013

No: 3004

Topic: 7 — Neuromuscular disorders

Assessment of ataxia in patients with Progressive External Ophthalmoplegia (PEO)

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Background: Progressive external ophthalmoplegia (PEO) comprise a clinical spectrum that varies from pure ocular myopathy to complex multisystem disorders, such as Kearns-Sayre syndrome (KSS). Ataxia is a frequent feature in patients with KSS.

Objective: Assess ataxia in patients with PEO/KSS, correlating with level of independence and quality of life.

Patients and methods: We evaluated the presence of ataxia in 15 patients with mitochondrial disease in the spectrum PEO/KSS and 38 controls. Ataxia was assessed with Brief Ataxia Rating Scale (BARS), International Cooperative Ataxia Rating Scale (ICARS) and Scale for the Assessment and Rating of Ataxia (SARA). Level of independence was estimated through Barthel index and quality of life with the World Health Organization Quality of Life (WHOQOL-bref).

Results: Scores obtained with the ataxia rating scales demonstrated a significant difference between PEO/KSS patients and controls ($p < 0.001$; Mann-Whitney test). However, only one patient reached the maximum scores, while the remaining ranged from 3–16 (BARS 0–30); 9–42 (ICARS 0–100) and 2–15 (SARA 0–40), indicating a mild to moderate ataxia. Nevertheless, no patients scored zero, compared to 27 controls. Correlation between ataxia scales demonstrated a positive correlation of moderate to high magnitude ($r = 0.60$ to 0.94 ; $p < 0.05$). There was no correlation between the presence of ataxia and level of independence or quality of life.

Conclusion: Our results suggest that ataxia is a frequent feature in patients with mitochondrial disease within the spectrum PEO/KSS. However, ataxia is only mild to moderate and did not influence the level of independence or the quality of life.

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Abstract — WCN 2013

No: 2968

Topic: 7 — Neuromuscular disorders

A case of painless ptosis due to isolated levator palpebrae myositis

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Background: Idiopathic orbital myositis is a nonspecific orbital inflammation primarily involving one or several extraocular muscles. The cardinal symptom is orbital pain exacerbated by eye movement accompanying with or without ptosis, diplopia, exophthalmos, and conjunctival injection.

Objective: To present a case of levator palpebrae myositis without any pain or other unpleasant feelings.

Patients and methods: A thirty-eight-year-old man visited our outpatient clinic with left ptosis five days ago. He complaint of myalgia and generalized weakness from a week ago, but he did not complain of diurnal variation or progression of symptom. Repetitive nerve stimulation test and thyroid function test were normal. Levels of angiotensin converting enzyme, acetylcholine receptor antibody, and anti-GQ1b antibody were normal. His visual acuity and light reflex was normal. There was no edema, exophthalmos, or orbital bruit. His brain MRI showed the swelling and enhancement of left levator palpebrae muscle. After oral steroid treatment, his ptosis was improving. After a week, his symptom was completely improved.

Results: The patient's neurological symptom could be commonly confused with other neurological disorders such as thyroid eye disease, neuromuscular junction disorder, and other autoimmune disorders. MRI could be the most important radiologic finding for diagnosis.

Conclusion: In this case, brain MRI may play an important role in the diagnosis of levator palpebrae myositis. Unlike the other cases, levator palpebrae myositis could be not painful.

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Abstract – WCN 2013**No: 2802****Topic: 7 – Neuromuscular disorders****Interferon gamma as a potential therapy for Friedreich ataxia**

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Background: Friedreich ataxia (FRDA) is the most common of inherited ataxias, affecting >20,000 individuals in western countries. It is caused by GAA triplet expansions in the first intron of the frx gene, that result in the insufficient production of the mitochondrial protein frataxin. Patients, usually children, experience progressive loss of motory coordination up to severe disability and premature death. Pathology is dominated by the degeneration and loss of dorsal root ganglia neurons. There is no approved therapy for FRDA.

Objective: We observed that interferon gamma was able to upregulate frataxin levels in cells derived from FRDA patients. We aimed at obtaining preclinical evidence of efficacy in an animal model of the disease.

Methods: FRDA mice, engineered to express only reduced amounts of frataxin, were treated for several weeks with interferon gamma or vehicle.

Results: Interferon gamma-treated FRDA mice showed improved performance on all the motory coordination tests performed, compared to vehicle-treated FRDA mice. Degeneration and neuronal loss were largely prevented in dorsal root ganglia and this was associated with a recovery in neuronal frataxin levels.

Conclusions: Treatment with interferon gamma, a drug currently approved for pediatric indications, shows potential as a therapy for FRDA. A phase II clinical trial, aimed at assessing tolerability of interferon gamma and the ability of interferon gamma to elevate frataxin levels in FRDA patients, is currently underway. Future clinical trials considering the possibility of a more targeted drug delivery by the use of implantable intrathecal pumps and addressing clinical endpoints, are being planned.

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Abstract – WCN 2013**No: 2996****Topic: 7 – Neuromuscular disorders****Using sensitivity, specificity and accuracy for detection of axonal neuropathy**

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Background: Digital signal processing and general mathematical methods of pattern analysis can be used in needle electromyography to detect motor unit potentials and to recognize various kinds of muscle diseases.

Objective: The goal of the paper is to contribute to various methods enabling discrimination of individuals with axonal neuropathy from normal cases using signals acquired from needle electromyography.

Patients and methods/material and methods: Data from a control set of 104 individuals and a set of 76 patients were used to validate selected methods for their separation and classification. Different kinds of signal features in time as well as frequency domains were studied to obtain the most reliable results.

Results: The proposed features used to detect individuals with axonal neuropathy were analyzed by specificity, sensitivity and accuracy of data obtained using the receiver operating characteristic

curves and confusion analysis. Accuracy higher than 95% was achieved for the given sets of individuals and for optimal criterion values proposed.

Conclusion: The proposed modified Willison amplitude together with statistical and spectral properties of signal components can be used to classify individuals into sets of healthy and neuropathic patients with sufficient accuracy.

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Abstract – WCN 2013**No: 2824****Topic: 7 – Neuromuscular disorders****Serum cytokine profiles of chronic inflammatory demyelinating polyneuropathy**

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Background: Chronic inflammatory demyelinating polyneuropathy (CIDP) is a heterogeneous disorder that has some subgroups characterized by the different clinical features.

Objective: To elucidate the pathogenesis of CIDP, we extensively investigate the cytokine profiles of CIDP ("typical CIDP" and "multifocal acquired demyelinating sensory and motor neuropathy" [MADSAM]), Guillain-Barre syndrome (GBS) and normal controls.

Method: We analyzed the sera of 30 subjects with CIDP, 9 with GBS and 18 healthy controls. Forty-seven inflammatory mediators were simultaneously measured with a multiplex bead-based ELISA on a Suspension Array System.

Results: TNF α , HGF, MIP1 β , MIP1 α and IL1b levels were increased in CIDP as compared with controls. Particularly, the serum levels of TNF α were significantly increased in the patients with "MADSAM", and the serum levels of HGF, MIF and MIP1 β in the patients with "typical CIDP". However there were no significant differences between the patients with typical CIDP and MADSAM. An increase in HGF, GRO α , and MIG, and a decrease of RANTES and SCF were found for GBS.

Conclusion: Serum concentrations of several inflammatory cytokines were increased in the patients with both subgroups of CIDP. These cytokines may play a role in the pathogenesis of demyelination and the breakdown of the blood-nerve barrier in CIDP.

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Abstract – WCN 2013**No: 2963****Topic: 7 – Neuromuscular disorders****Objective biological markers during the course of disease in amyotrophic lateral sclerosis**

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Background: The possibility to use objective biological markers during the course of the disease is of great importance to enable studies on treatment in ALS. The variable course of the ALS syndromes makes it necessary to have several objective biochemical markers of the degeneration.

Objective: To find stable, predictive and reliable biochemical markers related to the progression of ALS in patients with different courses of the disease.

Patients: More than 400 patients were followed by clinical and biochemical data through the course of disease. Blood and serum and

in most also CSF analyses were performed during the course of the disease. Both inflammatory and degenerative markers were studied.

Results: Serum analyses of degenerative biochemical analyses were of variable stability during the disease. In CSF, NFL was an important marker of disease activity, closely related to the different variants of ALS disease and their progression rate. Specific inflammatory changes were common in CSF. The course of the disease was predicted by early combined analyses of serum and CSF.

Conclusion: There were several simple and non-expensive objective parameters that could be used to predict the further course of the disease, in parallel with the clinical estimates of FVC and ALSFRS-R. The findings were important for the information and planning of support to patients and next of kin and pertinent to consider in interventional clinical studies.

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Abstract – WCN 2013

No: 2804

Topic: 7 – Neuromuscular disorders

Sphincter dysfunction in a case of cauda equina disorder: An atypical variant of autoimmune lumbosacral polyradiculoneuropathy

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Background: Sphincter dysfunction is not included in the diagnostic criteria of autoimmune lumbosacral polyradiculoneuropathy and its infrequent appearance can be attributed to autonomic impairment.

Objective: To describe an atypical variant of autoimmune lumbosacral polyradiculoneuropathy in the form of a cauda equina syndrome.

Patient: A 51-year-old woman presented with a 2 week history of back and leg pain, burning pain in her feet, numbness in her toes and saddle area and sphincter dysfunction. There was no history of diarrhea or surgery.

On examination muscle strength was normal, deep tendon reflexes at the knees and ankles were reduced, over the next few days were lost, and vibration sense in her feet was decreased.

Results: CSF analysis showed no albuminocytologic dissociation. Serological studies were negative for syphilis, Mycoplasma pneumoniae, Lyme disease and HIV. Coprocultures for Campylobacter jejuni were negative.

The levels of ACE, LDH and beta 2-microglobulin were within normal limits, and tumoral markers were absent.

Gadolinium-enhanced MRI of the lumbar spine showed thickening and enhancement of the cauda equina nerve roots, pointing towards an inflammatory etiology.

The demyelinating features revealed by the electrophysiological analysis and the high titres of antiganglioside antibodies confirmed the dysimmune etiology.

The patient was treated with intravenous immunoglobulins, which improved her symptoms.

Conclusion:

- After ruling out tumoral, compressive and infiltrative processes in a cauda equina syndrome we have to consider an inflammatory and autoimmune etiology.
- Spinal MRI is a highly valuable imaging method for the diagnosis, as reliable as nerve conduction studies and CSF analysis.

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Abstract – WCN 2013

No: 2785

Topic: 7 – Neuromuscular disorders

Chronic autoimmune sensorial neuropathy: A case report

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Introduction: The sensory neuropathies may present clinical heterogeneity with an acute or chronic onset. The clinical features can include sensorial loss, hyporeflexia or areflexia. In this report, we present a rare case with chronic autoimmune inflammatory demyelinating sensorial neuropathy accompanying polymyositis.

Case: A 33 year old female presented to our clinic with progressive postural instability, unable to walk without any support since 2 years. The patient had taken pulse steroid and a single dose of cyclophosphamide in another clinic, but her complaints had not improved with this therapy. For this reason, standard IVIg therapy had given for 5 days and the case had begun to start walking without any support. Our neurological examination revealed postural instability and no mobility without support, bilateral symmetric quadriparesia, hyperalgesia in distal extremities, impairment of sensation of vibration and position, absence of deep tendon reflexes and unresponsive bilateral reflex in sole skin. Blood, urine, CSF analyses and CT imaging were normal. EMG indicated severe sensorial and mild motor fiber neuropathy and with these findings, we considered the case as a chronic autoimmune sensorial polyneuropathy. IVIg therapy was given to the patient and clinical findings improved significantly. However, 2 weeks after IVIg therapy the complaints of the patient increased and then we decided to give IVIg therapy for every 2 weeks.

Discussion: IVIg therapy is an effective method for chronic autoimmune sensorial polyneuropathy unresponsive to steroid therapy. However, in rare cases unresponsive to standard monthly IVIg therapy, more often applications could be a beneficial option.

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Abstract – WCN 2013

No: 2943

Topic: 7 – Neuromuscular disorders

Genetic analysis of Japanese patients with motor neuron disease

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Background: Motor neuron disease (MND) is heterogeneous syndrome. Even in the same race and the same nation, there are regional differences in the prevalence and clinical features. Because some ALS patients with SOD1 mutation have only lower motor symptom clinically, we think that it is necessary to examine not only ALS patients but also lower MND patients to reveal the prevalence of the patients with ALS related gene mutations.

Objective: We aimed to reveal the genetic background in patients with MND in San-in district, Western Japan.

Patients and methods: We had clinically examined 32 MND patients in our hospital in 2007–2013. We got informed consent for genetic examination in 16 patients among them. They included five families with familial MND (eight patients). We did genetic test for SOD1, ANG, FUS, TDP-43, OPTN, C9ORF72 and SBMA using auto-sequencer.

Results: Three patients in a family had OPTN G478E mutation. Two of them had homozygous mutation and earlier onset of the disease than the patient with heterozygous. These three patients developed dementia and flexion of limbs. Two patients in another family had SOD1 L126S mutation. One patient with familial ALS

(FALS) had SOD1 c.379_380delTT mutation. One familial and two sporadic patients with lower MND had SBMA mutation. One FALS patient and six sporadic ALS patients had no mutations in this study.

Conclusion: We revealed SOD1, OPTN and SBMA mutations in our patients. Some of them had atypical features for ALS. We think these genetic tests are useful for the diagnosis of MND.

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Abstract – WCN 2013

No: 2731

Topic: 7 – Neuromuscular disorders

Preliminary report of Tropals study – A survey of amyotrophic lateral sclerosis in Africa

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Background: Epidemiological studies of Amyotrophic Lateral Sclerosis (ALS) in the tropics are rare and their methodologies, heterogeneous. Many questions arise as regards the characteristics of this disease in the tropics, especially in Africa.

Objective: Describe sociodemographical and clinical characteristics of ALS patients diagnosed in Africa.

Patients and methods/material and methods: TROPALS (<http://www.tropals.unilim.fr/>) is a multicentre observational cohort study. A shared methodology and an online data base that allows centers to collect data in a standardized and homogeneous way.

Results: 40 patients have been included to date in 5 centers (Benin, Mauritania, Senegal, Togo, Tunisia), 3 other centers are open (Burkina Faso, Gabon, Mali) and 6 are about to be open.

Mean age at diagnosis was 51.9 ± 13.5 years (2 cases less than 25 years), male/female sex ratio was 2.4.

First symptoms were mostly spinal (72.5%) and 80% ($n = 32/40$) of patients had Electroneuromyography for diagnosis purpose. At this time mean ALS FRS R was 32.1 ± 10.5 and 75% of patients presented atypical symptoms (mostly dysautonomic or sphincter problems).

After diagnosis, 97.4% of patients were prescribed an occidental treatment: Rilutek® ($n = 12$), physiotherapy ($n = 17$), or symptomatic treatment ($n = 12$). 21.6% of patients used a “traditional treatment” based on infusion-decoction for 2 of them and of unknown type for 6 patients.

Conclusion: More inclusions are needed to produce precise estimations. Follow-up data are currently being collected. Tropals study will allow us to improve the description of ALS characteristic prognosis of patients and comprehension of the disease in this continent.

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Abstract – WCN 2013

No: 2933

Topic: 7 – Neuromuscular disorders

The importance of imaging of the nerves in some cases of mononeuropathy

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Case 1: Forty year old female patient came with h/o pain in the left sciatic nerve distribution of 1 year duration. Her MRI lumbar spine and neurophysiological studies are normal. As the symptoms are continuous and partly responding to neuropathic pain medications, it was decided to do MRI imaging of the left sciatic nerve. The investigation showed neuroma of the sciatic nerve in the left mid thigh region.

Case 2: Fifty year old patient noticed acute finger drop in the right hand after heavy cooking. The MRI cervical spine was normal. As she c/o pain in the right elbow region and clinically there was a tiny lump in the palpation, it was decided to do neuroimaging of the right radial nerve around elbow region focusing on posterior interosseous nerve. The MRI showed a ganglion compressing the nerve resulted in the finger drop.

Case 3: Twenty eight year old gentleman had a trauma in the left midarm region two years back. He noticed a small swelling in the same region for the past one and half years. He noticed painful tingling sensation in the left ulnar nerve distribution for the past 2 months. His neuroimaging of the left arm showed a hemangioma in close relation to the ulnar nerve.

The neuroimaging of the peripheral nerves may be needed in some of the atypical cases of mononeuropathy to rule out underlying structural cause, which is important for further management.

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Abstract – WCN 2013

No: 3116

Topic: 7 – Neuromuscular disorders

Myasthenic crisis presenting with isolated bilateral partial vocal cord palsy

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Objective: To present a case of myasthenic crisis with acute upper airway obstruction and severe stridor due to isolated bilateral partial vocal cord palsy.

Case: We report the case of a 65-year-old man with a 3-month-history of acetylcholine receptor antibody positive generalized autoimmune myasthenia gravis (MG), who acutely developed severe stridor and speech difficulty. His medications included pyridostigmine 60 mg five times daily, azathioprine 75 mg once daily; the corticosteroids (prednisolone) had been recently increased from 30 mg once daily to 30 mg/60 mg on alternating days. During the preceding days he had developed an upper airway infection. Clinical examination at the time of exacerbation showed normal strength in the neck muscles and no proximal limb weakness. A flexible laryngoscopy was performed and revealed a near adduction position of the vocal cords with limited movement especially on the right side, without any upper airway edema. The patient remained self-ventilating and was transferred to the neurological intensive treatment unit for airway monitoring. He received a full dose of human IVIg (2 g/kg) over 4 days with significant recovery and clinical stabilization.

Discussion: Acute exacerbation of autoimmune MG can present with great variability and can selectively involve isolated muscles. In the case that we present, a myasthenic crisis emerged with inspiratory stridor due to isolated bilateral partial vocal cord palsy, without any signs of other muscle weakness.

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Abstract – WCN 2013**No: 2545****Topic: 7 – Neuromuscular disorders****Novel mutations for SPG 11 in a 25-year old woman with hereditary spastic paraparesis and evidence for upper extremity involvement**

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Introduction: Familial spastic paraplegia with thin corpus callosum is related to mutations in the genes SPG11, 15 and 18. Mutations in the SPG11 gene have also been described in amyotrophic lateral sclerosis with young onset and a relatively benign course.

Objectives: We present a patient with typical spastic paraplegia, with electromyographic evidence for upper extremity involvement.

Case report: A 25-year-old woman was admitted because of progressive gait disturbance and muscle cramps since 5 years. Her mother is healthy; the patient has no contact to her father.

The clinical neurological examination showed spasticity in both lower limbs with hyperactive reflexes and clonus, a bilaterally positive plantar reflex, a beginning pes equinus bilaterally, mild ataxia of both lower limbs and slightly saccadic eye movements. Sensory functions were normal. Neuropsychological testing revealed deficits in attention, memory and executive functions. MRI of the brain showed a remarkable thinning of the corpus callosum, MRI of the spinal cord was normal. Serologic testing revealed no evidence for HTLV1 or 2 infections. Nerve conduction studies were normal. Electromyography showed fasciculations and giant potentials consistent with denervation of motor units.

Genetic testing for SPG 3 and 4 were negative. Mutations were found in the SPG11 gene, probably consistent with a compound heterozygous state: Three heterozygous mutations were found: c.1621C>T, (known causal mutation) and c.5623C>T and c.6526T>C, which are novel mutations.

Conclusion: Upper extremity involvement and an overlapping phenotype with motor neuron disease may be present in familial paraplegia with thin corpus callosum and two novel SPG11 mutations.

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Abstract – WCN 2013**No: 3048****Topic: 7 – Neuromuscular disorders****Electrophysiological study of spinocerebellar ataxia and Friedreich ataxia's patients**

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The aim of our study is to examine the bioelectrical activity of nerves and muscles in spinocerebellar (SCA) and Friedreich's ataxia (FRDA) patients according to a standardized protocol, in order to obtain information about the severity and distribution of peripheral nerve involvement.

We examined 18 genetically diagnosed, older than 18 years of age SCA and FRDA patients. Control group was formed by 31 age-matched healthy individuals. We performed sensory and motor conduction studies, F responses, needle EMG of proximal and distal muscles, and motor unit number estimations (MUNE) of distal

muscles to upper and lower extremities of both groups. Descriptive statistics and group comparisons were done for each recorded parameter.

All sensory responses were low in amplitude ($p < 0.01$) in patients as well as sensory conduction velocity was slow ($p < 0.05$) for ulnar nerve. Median and peroneal motor amplitudes were also low ($p < 0.01$). Median, peroneal ($p < 0.01$), ulnar, and tibial ($p < 0.05$) motor conduction velocities were significantly slow. MUNE in m. abductor pollicis brevis and m. tibialis anterior muscles were significantly lower in patient group than healthy individuals ($p < 0.01$). Needle EMG evaluation revealed neurogenic involvement in 1/3 of patients.

Polyneuropathy syndrome is frequent in SCA and FRDA patients. Sensory nerves in lower extremities were predominantly involved however, signs of motor dysfunction were also notable. MUNE can provide quantitative information about motor nerve fiber and/or motor neuron involvement. Clinical and electrophysiological findings of peripheral polyneuropathy syndrome were more striking in atactic patients with proven genetic defect.

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Abstract – WCN 2013**No: 3074****Topic: 7 – Neuromuscular disorders****Central nervous system involvement in a patient with acute motor axonal neuropathy and antiganglioside antibodies**

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Background: Acute motor axonal neuropathy (AMAN) is a variant of Guillain-Barre syndrome (GBS), usually associated with antiganglioside antibodies and *Campylobacter jejuni* infection. Rarely, AMAN may coexist with signs of central nervous system involvement such as hyperreflexia or asymmetrical weakness.

Objective and methods: To report clinical and neurophysiological evidence of central nervous system involvement in a patient with AMAN and positive antiganglioside antibodies.

Results: A 64-year-old man was admitted to our department with upper and lower limb weakness, leg paraesthesias and urinary urgency, following a gastrointestinal infection. Neurological examination showed mild tetraparesis with increased tendon reflexes. Cerebrospinal fluid had a normal cell count with increased protein (84 mg/dl). Nerve conduction studies revealed motor axonal polyneuropathy. Serum testing demonstrated high titers of anti-GM1 and anti-GQ1b antibodies. *C. jejuni* was isolated from stool culture. The patient received a 5-day course of intravenous immunoglobulin with no improvement. In fact, leg weakness deteriorated and became asymmetric, whereas voiding required catheterization. Brain and spinal cord MRI was normal. Motor and somatosensory evoked potentials showed prolongation of both motor and sensory central conduction times, confirming central nervous system involvement. One month after symptom onset, weakness and gait difficulty started to improve. After one year of follow-up the patient still presents pyramidal signs and severe disturbance of micturition.

Conclusion: This is the first report demonstrating involvement of central motor as well as somatosensory tracts in GBS. The pathogenesis of central nervous system dysfunction may be associated with antiganglioside antibodies but this needs further investigation.

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Abstract – WCN 2013**No: 3168****Topic: 7 – Neuromuscular disorders****HLA-DRB1 and HLA-DQB1 allele association to myasthenia gravis in Sudan; an Arabian-African population**

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Background: HLA gene association to MG has been reported by many studies but the pattern of association varied in different populations and MG clinical subgroups.

Objectives: This study aimed at detecting the association between HLA-DRB1 and HLA-DQB1 genes in MG patients in Sudan.

Methods: Typing was performed at low resolution for HLA-DRB1 and DQB1 loci with PCR dependent sequence-specific primers (PCR-SSPs). HLA allele frequencies in 87 MG patients were compared with 153 none myasthenic controls using SPSS software.

Results: HLA-DRB1*08 was found to be positively associated with MG, OR = 2.65 (1.2–5.87) ($P < .01$). None of the HLA-DQB1 alleles showed independent association to MG. Two DRB1 genotypes, HLA-RB1*08DRB1*13 (OR 3.3, 1.2–8.7, $P < .05$) and HLA-DRB1*03DRB1*1001 (not found in controls, $P < .01$), were found to be associated with MG in general and with early onset MG (EOMG) in females. One HLA-DQB1 genotype was found to be associated with the risk of MG; HLA-DQB1*03DQB1*06 (OR = 3.3, 1.6–6.5, $P < .001$). Two HLA genotypes have shown very strong association to MG; HLA-DRB1*08, *13DQB1*03, *06 (OR = 7.14, 1.9–26.4, $P = 0.001$) and the genotype HLA-DRB1*03, *1001DQB1*02, *05 (not found in controls, $P < .01$). HLA haplotypes in the two risk genotypes were HLA-DRB1*13DQB1*06, HLA-DRB1*08DQB1*03, HLA-DRB1*03DQB1*02, and HLA-DRB1*1001DQB1*05. The genotype HLA-DRB1*08, *13DQB1*03, *06 was associated with an increased risk in EOMG ($P < .01$ in females, $P < .05$ in males).

Conclusion: We reported a pattern of HLA association unique to the Sudanese population, in which two HLA-DRB1 DQB1 genotypes can be used as susceptibility markers for MG. One of the risk genotypes included HLA-DRB1*03DQB1*02 haplotype that has been constantly reported as a risk haplotype in Caucasian populations. This strong association with certain HLA-DRB1 and DQB1 combinations suggests a role of dominant epitopes, or an effect of a linked recessive non-HLA gene.

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Abstract – WCN 2013**No: 3151****Topic: 7 – Neuromuscular disorders****An autosomal recessive CMT family with a new mutation in the NDRG1 gene**

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Background: HMSN-Lom/CMT4D is very rare, an autosomal recessive (AR) peripheral neuropathy with deafness and unusual neuro-pathologic features.

Objective: We report the clinical features of the first AR duplication copy number variation (CNV) of *NDRG1* causing HMSN-Lom/CMT4D (MIM#601455) in a large Turkish pedigree. The intragenic duplication CNV included exons 6 to 8 and caused a complete loss of function of *NDRG1*.

Patients and methods: There were four affected siblings from this consanguineous family. Two of them had the same clinical picture. One of them was different and did not have *NDRG1* mutation. The index case was a 30 year-old male, and his 24-year-old sister had delayed motor milestones. Involvement of lower and upper extremities became apparent in the first decade with hearing loss and glaucoma. The third affected member had a history of epilepsy and intellectual disability in addition to her more severe neuropathy phenotype.

Results: Their clinical examination showed typical features of peripheral neuropathy including muscle weakness and wasting, tendon areflexia, and skeletal and foot deformities. Nerve conduction studies (NCSs) showed absent sensory nerve action potentials (SNAPs) in all limbs except the little sister. Compound motor nerve potentials (CMAPs) recorded from distal muscles were also absent.

Conclusion: Clinical findings in our cases were consistent with HMSN-Lom (CMT4D) however further cases are needed to make a better genotype–phenotype correlation. To the best of our knowledge this is the first recessive duplication CNV in the literature and the third mutation in *NDRG1*.

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Abstract – WCN 2013**No: 3153****Topic: 7 – Neuromuscular disorders****Jitter estimation using concentric needle electrode on voluntarily activated orbicularis oculi and frontalis muscles in healthy control and myasthenic patients**

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Background: Single fiber electromyography (SFEMG) is the most sensitive clinical neurophysiological test for neuromuscular junction disorders, particularly myasthenia gravis (MG).

Objectives: No available data concerning jitter measurement in Sudan in addition to scarcity of data worldwide. The aim of this study was to estimate normal (reference) & abnormal jitters in voluntarily activated orbicularis oculi (OOc) & frontalis muscles using disposable concentric needle (CN) in the same subject [either control or myasthenic patients].

Methods: Prospectively 45 MG patients (21 males & 24 females, mean age 39.1 ± 16.9 years) & 62 controls (20 males & 42 females, mean age 43.2 ± 14.0 years) were included in the study. Jitter values were expressed as the mean consecutive difference (MCD) of 30 potential pairs in μ s.

Results: In the control group, the mean MCD, mean MCD for individual potential pairs & mean outlier jitter values (with upper 95% confidence limit-CL) for [OOc] were [26.9 ± 3.3 (32.0), 26.1 ± 8.9 (41.8) & 38.5 ± 5.7 (49.0) μ s respectively] & for [frontalis] were [27.1 ± 3.0 (31.3), 26.4 ± 9.4 (42.9) & 39.9 ± 5 (49.2) μ s respectively]. In the patient group the mean MCD & mean MCD for individual potential pairs for [OOc] were [50.9 ± 21.8 , 48.8 ± 36.8 μ s respectively] & for [frontalis] were [47.4 ± 13.9 , 45.8 ± 28.7 μ s

respectively]. Percent of patients with: [increased mean MCD above our own upper practical limit (95%CL), >10% abnormal pairs using our own 95% CL of individual MCD values & 95%CL of outlier values and >10% of individual pairs with block] were as follows: [87%, 95.6%, 91.1% & 42.2% respectively] for OOc & [91.7%, 100%, 88.9% & 40% respectively] for frontalis. Comparison between the mean MCDs of [OOc] & [frontalis] revealed no significant statistical difference [t-test] in the control & patient groups.

Conclusion: The suggested practical upper limits for mean MCD & for outliers were (32, 49 μ s) for OOc & (32, 50 μ s) for frontalis. The study is unique in that it compared between jitter values of two voluntarily activated facial muscles (OOc & frontalis) using CN in the same individual [recruiting large number of controls & patients] & determined the percent of patients who fulfilled the criteria for diagnosing a patient as having positive jitter test.

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Abstract – WCN 2013

No: 3160

Topic: 7 – Neuromuscular disorders

Neuromuscular junction autoantibodies in seronegative myasthenia gravis patients from Sudan

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Background: In myasthenia gravis (MG) autoantibodies target neuromuscular junction (NMJ) proteins leading to a decreased sensitivity to ACh with muscle weakness. 80–88% of MG patients have AChR antibodies (AChR-Ab⁺). It was suggested that AChR-Ab^{-ve} patients may have autoantibodies against other NMJ proteins or a more sensitive assay is required to detect AChR-Abs. Hoeh et al. (2001) detected muscle specific kinase antibodies (MuSK-Abs) in 70% of AChR-Ab^{-ve} generalized MG patients (GMG). Leite et al. (2008) detected clustered AChR-Abs by the cell based assay (CBA) in 66% of AChR-Ab^{-ve} patients. A considerable proportion of MG patients remained seronegative (SN-MG).

Objectives: To identify the nature of NMJ autoantibodies in AChR-Ab^{-ve} MG patients from Sudan and to relate it to their clinical presentation.

Patients and methods: 96 MG patients (77 GMG and 19 OMG) were randomly recruited from two national MG referral centers in Khartoum during the period 2010–2012. All patients were screened for AChR-Abs and MuSK-Abs by radioimmunoprecipitation assay (RIPA). Negative sera were screened for clustered AChR-Abs by the CBA.

Results: 62.5% (60/96) of patients were AChR-Ab⁺ by RIPA. Of the 36 AChR-Ab^{-ve} patients, 19.4% (7/36) were MuSK-Ab⁺, and 22.2% (8/36) were clustered AChR-Ab⁺. 21.9% (21/96) of patients remained SN-MG. MuSK-Ab⁺ patients (MuSK-MG) represented 25.9% (7/27) of AChR-Ab^{-ve} GMG with a female:male ratio of 1.3:1 and dominant bulbar symptoms (85.7%,6/7). Clustered AChR-Ab⁺ patients were all females (n = 8). SN-GMG were predominantly females (92.9%, 13/14)(P = 0.02). The incidence of an associated autoimmune disease (AUID) in SN-GMG females was 53.8% (7/13) compared to 17.6% (6/34) in AChR-Ab⁺ GMG females (P < .013), RR (95% CI) = 3.05 (1.3–7.38).

Conclusion: The incidence of MuSK-MG was within the reported detection rates (0–50%). The association between AUID and SN-GMG

suggests the involvement of sex and predisposition to autoimmune diseases in determining the pathogenicity in MG.

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Abstract – WCN 2013

No: 3162

Topic: 7 – Neuromuscular disorders

Concentric needle jitter using different stimulation methods in extensor digitorum communis and orbicularis oculi in healthy subjects and myasthenic patients

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Background: Single fibre electromyography (SFEMG) is the most sensitive clinical neurophysiological test for neuromuscular junction disorders, particularly myasthenia gravis (MG).

Objectives: No available data concerning jitter measurement in Sudan in addition to scarcity of data worldwide. The study aimed at measuring jitter values, estimating conversion factor[CF] & comparing between the standard methods of stimulation [voluntary activation (V-) & axonal stimulation (S-)] using concentric needle (CN), testing extensor digitorum communis (EDC) & orbicularis oculi (OOc) muscles in the same individual [either control or patients with MG].

Methods: Prospectively 45 MG patients (21 males & 24 females, mean age 39.1 \pm 16.9 years) & 62 controls (20 males & 42 females, mean age 43.2 \pm 14.0 years) were studied. Jitter values were expressed as the mean consecutive difference (MCD) of 30 potential pairs in (μ s). T-test was used for comparison.

Results: In the control group the mean MCD, mean MCD for individual potential pairs & mean outlier jitter values (with upper 95% confidence limit–CL) for [V-EDC] were [27.5 \pm 3.4 (33.5), 26.7 \pm 9.0 (43.3) & 39.3 \pm 5.1 (48.2) μ s respectively], for [S-EDC] were [21.2 \pm 2.9 (25.8), 21.0 \pm 8.1 (35.9) & 32.6 \pm 4.5 (39.6) μ s respectively], for [V-OOc] were [26.9 \pm 3.3 (32.0), 26.1 \pm 8.9 (41.8) & 38.5 \pm 5.7 (49.0) μ s respectively] & for [S-OOc] were [21.8 \pm 2.4 (25.6), 21.5 \pm 7.6 (35.3) & 32.4 \pm 3.5 (37.5) μ s respectively].

In the patient group the mean MCD & mean MCD for individual potential pairs for [V-EDC] were [46.8 \pm 18.4, 44.2 \pm 30.5 μ s respectively], for [S-EDC] were [27.4 \pm 10.4, 27.0 \pm 19.0 μ s respectively], for [V-OOc] were [50.9 \pm 21.8, 48.8 \pm 36.8 μ s respectively] & for [S-OOc] were [42.9 \pm 20.4, 38.5 \pm 25.4 μ s respectively]. V-EDC showed statistically significant higher MCD values than S-EDC in the controls [CF = 0.78] & patients [CF = 0.64]. V-OOc showed statistically significant higher MCD values than S-OOc in controls [CF = 0.82] but not statistically significantly higher than S-OOc in patients [CF = 0.92]. In the control group, comparison of the mean MCDs for [V-EDC/V-OOc] & [S-EDC/S-OOc] revealed no significant statistical difference. In the patients, comparison of the mean MCDs for [V-EDC/V-OOc] showed no significant statistical difference as well but revealed significant difference between [S-EDC and S-OOc].

Conclusion: The suggested practical upper limits for mean MCD & for outliers were (34, 49 μ s) for V-EDC, (32, 49 μ s) for V-OOc, (26, 40 μ s) for S-EDC and (26, 38 μ s) for S-OOc. The study is unique in that it compared between jitter values of the two standard techniques using CN in EDC & OOc muscles of the same subject [recruiting large number of controls & patients].

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Abstract – WCN 2013**No: 3184****Topic: 7 – Neuromuscular disorders****The component of EMG studies to predict outcome after transforaminal epidural steroid injection**

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Objectives: We investigated the predictive value of components of electrodiagnostic studies for outcome after lumbar transforaminal epidural steroid injection in 38 subjects with clinical lumbosacral radiculopathy.

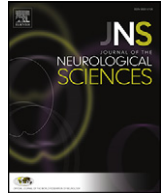
Method: In this retrospective study, visual analog scale (VAS) for evaluation of severity of pain, and Roland Morris Disability Questionnaire (RMDQ) and Oswestry Disability Index (ODI) for evaluation of functional outcome were evaluated in 38 patients.

Result: Among 38 patients who had undergone electrodiagnostic studies before injection, 28 patients were positive for lumbar radiculopathy and 10 patients showed negative results. There were

significantly greater improvements of VAS and ODI for patients with a positive lumbar radiculopathy as confirmed by electrodiagnostic study. However, when H-reflex abnormalities, ASA in paraspinal muscle, ASA in lumb muscle, and MUAP abnormalities were evaluated separately as an indicator of outcome, all of the individual components of electrodiagnostic studies failed to be the significant predictors of improvement based on outcome measure such as VAS, RMDQ, and ODI.

Conclusion: Based on this study, positive lumbar radiculopathy confirmed by electrodiagnostic study is a predictor of pain reduction and functional improvements after transforaminal epidural steroid injection for lumbar radiculopathy. Patients with positive electrodiagnostic study are more likely to show pain reduction and to show better functional recovery after transforaminal epidural steroid injection. However, H-reflex abnormalities, ASA in paraspinal muscle, ASA in limb muscle, and MUAP abnormalities did not predict pain reduction or functional outcome improvement in patients with lumbar radiculopathy.

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Topic: 8 - Headache

Abstract – WCN 2013

No: 3149

Topic: 8 – Headache

Intracranial hypotension is a rare cause of orthostatic headache: A review of the etiology, treatment and prognosis of 13 cases

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The aim of this investigation is to examine the causes, clinical picture, treatment, and prognosis of spontaneous intracranial hypotension, a rare cause of orthostatic headache, among the cases presenting in our clinic.

Thirteen cases (5 males and 8 females), diagnosed with spontaneous intracranial hypotension in our clinic between January 1st, 2009 and October 30th, 2011, were included in this study.

The presenting symptoms, treatment, findings on cranial magnetic resonance imaging, cerebrospinal fluid pressure measured at lumbar puncture (in available patients), and healing period of the patients were recorded. Five patients with orthostatic headache and accompanying symptoms were treated with bed rest, increase in oral fluid intake, intravenous hydration, and caffeine, and experienced a complete recovery. Complete recovery was observed in two patients (15.3%) within 10 days, in another two (15.3%) within 15 days and in one patient (7.6%) within 21 days. Headaches and other clinical symptoms significantly regressed within 30 days in four patients (37.6%) who received similar treatment, but a mild headache persisted intermittently during follow-up in these individuals. As the headache had not resolved after 30 days, an epidural blood patch was applied in these four cases (37.6%) and the clinical picture completely improved within 10 to 15 days.

Spontaneous intracranial hypotension should primarily be suspected in cases complaining about postural headache and contrast-enhanced cranial imaging should be performed. The presence of cranial nerve paralysis and pyramidal tract signs should be considered. Conservative treatments should be considered initially, however if conservative treatments fail, epidural blood patches must be applied.

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Abstract – WCN 2013

No: 3171

Topic: 8 – Headache

Association of cholecystokinin receptor 1 gene polymorphism and migraine

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Background: Cholecystokinin (CCK) is one of the most abundant neurotransmitter peptides in the brain. CCK coexists with dopamine in dopaminergic neurons, and modulates the release of dopamine in the nucleus accumbens. The CCK system is believed to be involved in pain processing. The aim of the study was to investigate the prevalence of -81A/G (rs1799723), -128G/T (rs1800908) and 984T/C (rs1800857) polymorphisms of the CCK-AR gene in migraine patients and controls.

Method: 144 migraine patients (ICHD III), mean age = 41.6 ± 12.5 y.o. and 197 healthy controls living in Moscow and the Moscow Region were included. SNPs were genotyped by a PCR-RLFP technique: PCR with “GenPak™ PCR Core” (Isogene Lab., Ltd.) and restriction with HinfI for rs1799723 and rs1800908 and with PstI for rs1800857 (ferments by SibEnzyme Ltd.).

Results: C-allele frequency in 984C/T was significantly higher in migraine patients, 0.479 ± 0.030 compared with controls, 0.154 ± 0.026 ($\chi^2 = 85.44$, $p < 10^{-10}$; OR = 5.0, 95% CI = 3.50–7.13). The -81A/G and -128G/T minor allele's frequencies didn't differ between groups: -81G = 0.049 ± 0.018 in the migraine group and 0.045 ± 0.015 in the controls; -128T = 0.045 ± 0.017 in the migraine group and 0.025 ± 0.011 in the controls.

Conclusion: This study is the first to report a more positive association of the C-allele of the CCK-AR gene 984T/C polymorphism in patients with migraine than in control subjects.

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Abstract – WCN 2013

No: 3178

Topic: 8 – Headache

Can vision influence trigeminal nociception? A study of the effect of visual cortex activation on the nociceptive blink reflex

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Background: In migraine, the link between cortical phenomena and trigeminovascular activation is not clear and thus, headache as well.

Objective: To search in humans for a possible functional connection between the visual cortex and the trigeminal nociceptive system by studying the effect on the nociceptive blink reflex (nBR) of repetitive transcranial magnetic stimulation (rTMS) applied over the visual cortex, and to compare healthy volunteers (HV) and migraine without aura patients (MO).

Methods: Fifteen bilateral nBR responses were recorded by stimulating the right supraorbital nerve in 22 HS and 13 MO before and after 1 Hz (15 min train) or 10 Hz (20 trains with a 15 s intertrain interval) rTMS. For comparison, we also performed the same study in HV after an 8 Hz visual flash stimulation.

Results: 1 Hz rTMS significantly decreased pain threshold ($p = 0.0019$, $p = 0.046$) in HV but not in MO. 1 Hz rTMS increased R2 AUC bilaterally ($p = 0.024$ and $p = 0.036$) in HV, but not in MO. The effect of flash stimulation in HV was the opposite of that of 1 Hz rTMS: increase in pain threshold, decrease in R2 AUC.

Conclusions: These results demonstrate for the first time a functional connection between the visual cortex and the trigeminal nociceptive system in humans. Interestingly, the modifications found after 1 Hz rTMS, supposed to inhibit the underlying cortex, are similar to those observed during migraine attack and in line with those described after CSD in animals. Why changes are not significant in MO could be related to the interictal dysexcitability found in migraine.

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Abstract – WCN 2013

No: 3179

Topic: 8 – Headache

Does trigeminal nociception influence the visual cortex: A study of the effects of supraorbital electro- or chemo-nociceptive stimulation

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Background and aims: In migraine pathophysiology, the link between the visual cortex and the trigeminovascular system might play a pivotal role. In a previous study we showed the top-down relation between the visual cortex and the trigeminal nociceptive system.

To search for the existence of a bottom-up relationship we studied the effect of electrical stimulation or application of capsaicin in the territory of the 1st division of the trigeminal nerve on pattern-reversal visual evoked potentials (PR-VEP).

Methods: We recorded 600 responses of PR-VEP at 3.1 Hz before and after trigeminal stimulation. The electrical stimuli were delivered over the supraorbital nerve for 20 min at 16 mA and 100 Hz in 7 healthy volunteers (HV). Capsaicin (cream 3%) was applied over the right frontal area for 1 h in 8 HV. We measured PR-VEP N1, P1 and N2 latencies, N1–P1 and P1–N2 amplitudes in six sequential blocks of 100 sweeps and habituation as the change in amplitude over the six blocks.

Results: The electrical stimulation increased the latency of P1 ($p = 0.027$) and N2 ($p = 0.042$) but left habituation unchanged. The capsaicin application increased habituation between the 1st and 2nd block of 100 responses ($p = 0.049$) but had no significant effect on latencies.

Conclusions: These results suggest that a bottom-up relation exists between the trigeminal (V_1) afferent nociceptive pathway and the visual cortex. Interestingly, an increase in PR-VEP habituation, such as that found after capsaicin application, also occurs during a migraine attack, where it replaces the interictal deficit of habituation.

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Abstract – WCN 2013

No: 3069

Topic: 8 – Headache

Verapamil side effects in our cluster headache population

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Background: Verapamil is an effective preventive therapy for cluster headache (CH). Side effects include constipation, nausea, asthenia, edemas, bradycardia and atrioventricular block.

Objective: Evaluate verapamil side effects, mainly cardiac, in CH patients.

Patients and method: Retrospective analysis of 84 patients with CH evaluated in the subspecialty Headache Clinic from May 2010 to June 2012. Demographic variables, types of CH [chronic (CCH), episodic (ECH)], acute and preventive therapies, and maximum dose of verapamil achieved, time of use, side effects and EKG characteristics, were reviewed.

Results: A total of 84 patients, 68 men and 16 women, mean age of 46 ± 12 years, were identified. Sixty patients were ECH and twenty four CCH. Seventy eight were treated with verapamil. The maximum daily dose of verapamil achieved was 500 ± 173 mg (mean \pm SD) in CCH and 344 ± 139 mg in ECH. Mean time of treatment was 67 ± 72 months and 36 ± 37 months respectively. Only 21 patients treated with verapamil had EKG on records. Eleven patients reported side effects due to verapamil and 7 stopped verapamil treatment, three of those with digestive and four with cardiovascular effects: three patients had hypotension and edema and one patient had a complete heart block which needed a transient pacemaker. Four other patients had asymptomatic bradycardia on EKG. All these patients had CCH.

Conclusions: Patients with CCH had more side effects due to verapamil. Cardiovascular side effects were related to higher doses and lengths of treatment. Although only one patient had a severe arrhythmia, we strongly recommend repeated EKG to avoid fatal complications.

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Abstract – WCN 2013

No: 3082

Topic: 8 – Headache

Headache-related disability in patients with systemic lupus erythematosus and primary Sjögren's syndrome

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Background: Headache in patients with systemic lupus erythematosus (SLE) has been considered common and has a strong weighting in the SLE disease activity index.

Objective: To investigate whether headache has more impact on SLE than on healthy persons, and also to compare headache-related disability in patients with another autoimmune disease, namely primary Sjögren's syndrome (pSS).

Patients and methods: Fifty-five patients with SLE, 51 pSS patients and 69 healthy subjects, all with primary headache, were included. Headache was classified according to the International Classification of Headache Disorders. The Migraine Disability Assessment Scale (MIDAS) and Headache Impact Test-6 (HIT-6) were used to assess headache-related disability. Depression was measured with the Beck Depression Inventory (BDI).

Results: SLE patients had higher HIT-6 scores (median = 51, range = 36–67) compared with healthy persons (median = 46, range = 36–72), $P = 0.02$, but no differences in impact scores were revealed between SLE and pSS patients (median = 54, range = 36–72), $P = 0.35$. More SLE patients had moderate to severe headache-related impact (HIT grades III–IV) compared with healthy persons (35% vs 17%, $P = 0.04$), but not with pSS patients (43%), $P = 0.43$. BDI scores were higher in SLE patients vs healthy persons, but not in SLE patients vs pSS patients. However, differences in headache-related disability between groups disappeared when adjusting for BDI and age.

Conclusion: SLE patients have more severe headache-related disability than healthy persons, but not more than patients with pSS. This may be related to more depression in these patients.

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Abstract — WCN 2013

No: 3066

Topic: 8 — Headache

Reduced sleep quality during the active phase of cluster headache

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Introduction: Cluster headache (CH) is a severe chronobiological pain-condition with recurring attacks of high-intensity headaches accompanied by autonomic symptoms and agitation. Many attacks occur at night which disrupts sleep but even patients who do not have nightly attacks complain of daytime sleepiness and poor sleep quality. A direct effect on sleep is obvious when attacks occur during the night but how daytime attacks affect sleep is not understood. There is a direct anatomical overlap of regions involved in the pathogenesis of headaches and those involved in the regulation of sleep.

Objective: We aimed to evaluate self-reported sleep quality in CH patients compared to a number of headache-free controls.

Patients and methods: Employing the Pittsburgh Sleep Quality Index we evaluated self-reported sleep quality in 129 patients suffering from CH and 50 headache-free controls.

Results: For all CH patients the average PSQI indicated poor sleep quality compared to healthy controls ($p < 0.01$). Comparing CH patients in active bout (attack within past month) with those outside of bout, sleep quality was significantly worse in the active group ($p < 0.01$).

Conclusion: It is possible that headaches occurring strictly during the daytime may affect sleep through an unknown pathway. That CH-patients also report poor quality of sleep in the inactive period of their disease suggests that the mechanisms leading to headache are reversible yet those leading to poor sleep quality are not. Poor sleep may thus be an epiphenomenon or an intrinsic part of the disease mechanism and need further detailed investigations.

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Abstract — WCN 2013

No: 3040

Topic: 8 — Headache

Cognitive function patients with idiopathic intracranial hypertension

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Background: Idiopathic intracranial hypertension (IIH) is a condition of raised intracranial pressure without identifiable cause. IIH primarily affects young obese individuals and the incidence is rapidly increasing. In addition to classical symptoms of headache, tinnitus and visual disturbances, patients often report impaired memory and concentration. Despite an obvious and serious threat to visual function; compliance with long-term treatment and instructed weight-loss is remarkably poor which could indicate frontal lobe impairment. However, cognitive impairment in IIH is only infrequently and inconsistently described in literature.

Objective: To investigate the cognitive function in a well-defined group of patients with IIH in a controlled design.

Materials and methods: We compared 31 patients (31.0 ± 11.1 years) with IIH and 31 healthy sex and age-matched (30.7 ± 11.6 years) controls by a comprehensive neuropsychological test battery consisting of validated computerized (CANTAB) and paper tests (Rey–Ostereich complex figure, trail making test A and B; letter and category fluency test; symbol digit modalities test and DART (Danish version of the national adult reading test)). Patients were tested in close relation to diagnosis and before any treatment was initiated.

Results: Patients with IIH performed significantly worse than controls in 8/10 of the initiated tests even though the premorbid intelligence level as measured by DART was similar for both groups. Especially domains controlling verbal fluency, processing speed and set shifting seem to be affected ($p \leq 0.01$).

Conclusions: The present case-control study indicates that IIH causes cognitive impairment and that a focused multidisciplinary approach including neuropsychological rehabilitation may supplement the treatment of IIH.

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Abstract — WCN 2013

No: 3104

Topic: 8 — Headache

Atypical liquor-hypotension syndromes

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Background: Liquor hypotension (LH) is a rare syndrome characterized by positional headache while in vertical posture and decreased CSF opening pressure during LP and characteristic MRI changes. Absence of certain cardinal symptoms however, may occur in some cases or atypical symptoms may appear and dominate the clinical picture, leading to differential diagnostic difficulties and delay in the treatment.

Objectives: To analyze the diagnostic difficulties and complications of LH.

Patients and method: A prospective analysis of LH cases admitted to our department of neurology between 2010 and 2013.

Results: 4 cases of LH were analyzed for their atypical features. In two cases typical clinical manifestations and imaging abnormalities were present. However, CSF opening pressure was normal, questioning the diagnosis. In one patient the CSF pressure was normal despite a documented CSF leakage.

In two other cases of LH subdural fluid collections were detected on CT scan. The complications in these cases were severe and misleading. The acute deterioration of the clinical status required urgent neurosurgical intervention.

Conclusions: Normal opening pressure does not exclude LH. LH is generally considered as a benign condition. Although severe complications are rare, subdural effusion or hematoma may indicate worse prognosis, questioning the benign nature of LH.

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Abstract — WCN 2013

No: 3113

Topic: 8 — Headache

Economic aspects of headaches (HA) in Russia

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Background and purpose: Being a major health problem, headache is also associated with a certain financial burden (which was equal to 140 billion rubles in 2011 for migraine alone). The HA management system should be improved and financial losses should be refused to solve the problem.

Material and methods: Data of Federal Service for State Statistics for 2011 and our own questionnaires for migraine patients were used to study the HA management in 6 Moscow and Moscow region outpatient clinics.

Results: The system of HA management was found to be ineffective. Total work input losses related to migraine were 47 million human-days. To solve the problem, a national program for HA management is required including education of the community, foundation of 9 multidisciplinary HA centers, training of physicians and nurses in HA management, education of local healthcare authorities in the field of social and economic aspects of HA with distribution of funds depending on needs of each region and application of effective treatment.

Conclusion: Medical care should consist of three levels: first – general practitioners give primary care to 80–90% of all HA patients; second – neurologists treat 10–15% of patients and the remaining 5–10% of patients will be treated in HA centers. The treatment in such centers is proved to be cost-effective with 10.3% economy of direct medical expenses as compared to the standard therapy. Losses of working days are reduced by half, saving about 10 billion rubles each year for migraine only.

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Abstract – WCN 2013

No: 2915

Topic: 8 – Headache

Clinical characteristics of migraine in Sudan

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Background: The International Headache Society had set certain criteria for the diagnosis and classification of migraine, however some studies reported that migraine has different presentations in different populations.

Objectives: To study the clinical characteristics of migraine in Sudanese families and whether it differs from that set by the International Headache Society.

Material and methods: A cross sectional study that involved 175 subjects from 12 large Sudanese families; Afroasiatics (Ja'afra, Jaa'lia, Shawiga, Kwahla) and Nilosaharan (Halfaween and Mahhs). 107 migraineurs were included in this study. Diagnosis of migraine was carried out with a validated questionnaire and confirmed by two neurologists.

Results: From the 175 who agreed to participate, 107 were migraineurs (70.1% females, 29.9% males). Both migraine with aura and migraine without aura co-exist in the same family and even in the same individual with the most common migraine type being MO (51.4%, N = 55), followed by MA + MO (32.7%, N = 35) and MA (15.9%, N = 17).

Unilateral location, presence of nausea, photophobia and aggravation by exercise showed significant results (P value = 0.000).

Conclusions: In the present study the presentation of migraine headache showed results similar to that described by the IHS in most of the criteria and differed in others. Co-morbidity with a number of diseases and migraine coexist.

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Abstract – WCN 2013

No: 2876

Topic: 8 – Headache

Position-triggered secondary paroxysmal hemicrania after medullary ischemia

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A 65-year-old male was admitted with a history of staggering vertigo in upright position starting several hours ago. His medical history was significant for hypertension. There was no relevant history of headache. Neurologic examination showed an inability to stand or walk because of propensity to fall with no paresis or paraesthesia and no further neurological signs. Cerebral MRI detected a subacute right medullary ischemia and no evidence for vertebral artery pathology. On the third day after admission the patient developed attacks of right-sided intense headache (more than 15 times/day) triggered by postural maneuvers. Pain attacks were accompanied by lacrimation and conjunctival injection of the right eye. The attacks of 5–7 min occurred only in upright position and could be relieved by lying down on the right side. Paracetamol, diclofenac, ASS, theophylline, methylprednisolone and oxygen were ineffective. Initiation of indometacin at 150 mg/d led to cessation of the pain attacks within 24 h, and could be tapered off 3 months later with rare reoccurrence of pain attacks.

Numbers of reports on secondary trigeminal autonomic cephalgias are increasing. Only few stroke-associated reports of paroxysmal hemicrania after medullary infarction including vertebral artery dissection have so far been described. Reports of triggered trigeminal autonomic cephalgias are also rare. There is only one case report provoked by postural maneuvers. It can be hypothesized that a lesion of the posterior spinocerebellar tract and the spinal tract nucleus of the trigeminal nerve triggered the hemicrania.

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Abstract – WCN 2013

No: 2919

Topic: 8 – Headache

Abnormal circle of Willis among migraineurs in Sudan

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Background: Several studies have shown a strong relationship between migraine and cerebral ischemia. The circle of Willis (CW) which is the main cerebral collateral pathway that allows flow redistribution in response to low perfusion is reported to be defective during migraine.

Objectives: This study aims to assess the structural characteristics of CW in Sudanese migraineurs.

Material and methods: This is a descriptive study including 54 migraineurs selected from a larger study (Migraine Profile in Sudan; a genetic, EEG and MRI study in 12 Sudanese pedigrees) and 17 apparently healthy volunteers. All subjects underwent three-dimensional MRA of the cerebral arterial circle.

Results: Eighty four point four percent (n = 38) of migraineurs have defective CW, while only 15.5% (n = 7) have normal configuration. Sixty point nine percent (n = 28) showed incomplete configuration of CW. Hypoplastic vessels account for 43.4% (n = 20), of these 17.4% (n = 8) have hypoplastic both PcoA. Furthermore, severe migraine is significantly associated with hypoplastic arteries of the posterior circulation (P value = 0.01).

Conclusions: Strong association has been observed between migraine and an abnormal configuration of the circle of Willis with hypoplasia of the both posterior communicating arteries significantly associated with severe migraine. This finding needs further confirmation as it will have an impact on the management and prognosis.

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Abstract — WCN 2013

No: 2925

Topic: 8 — Headache

Chronic headache and over-the-counter medication overuse: Prevalence and lifestyle factors

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Background: Chronic headache (CH) is defined as headache \geq 15 days a month for \geq 3 months. Many of those with CH overuse over-the-counter (OTC) analgesics.

Objectives: To estimate prevalence of CH in the Danish population; and to investigate associations between CH, OTC drug overuse, and lifestyle factors.

Methods: A representative sample of 129,150 individuals aged \geq 16 years from two regions of Denmark were invited to participate in the Danish National Health Survey of 2010. Participants were grouped into: no chronic headache (–CH), CH without OTC medication-overuse (CH–MO) and CH with overuse \geq 15 days per month (CH + MO). Prevalence of CH was calculated across demographic categories and weighted for stratified sampling and non-response. Health behaviors of interest were compared across the groups.

Results: The response rate was 53%. The crude prevalence of CH was 3.0% (weighted 3.3%), with a F:M ratio of 1.6:1. The prevalence of CH + MO was 1.6% (weighted 1.7%). CH–MO was higher in the youngest age group, while CH + MO peaked in the middle age group. There was an inverse relationship between CH prevalence and socioeconomic position. The CH + MO group had the highest proportion of people who **smoked** daily, were **sedentary**, with **BMI** over 30, and had high **stress** levels. **Alcohol** overuse was lower among those with CH. There were no differences in use of **narcotics** in the younger age group.

Conclusions: OTC analgesic overuse was seen in more than half of those with CH. Associations between unhealthy lifestyle and CH emphasize the need for lifestyle interventions concurrent with medical management of medication overuse headache.

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Abstract — WCN 2013

No: 2948

Topic: 8 — Headache

Headaches in public transport drivers

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Background: Headaches are the significant and frequent complaints, caused by many factors, with not always known pathogenesis and different clinical symptoms.

Objective: This work analyses the types and frequency of headaches among public transport drivers and tries to assess the importance of such factors as drivers' age and time of job.

Patients and methods: The study covered a group of 40 public bus drivers. Their average age was 43.2 (between 31 to 58 years). Their average job time was 11.6 years (between 2 to 33). The history of headaches was completed with the question set to objectify the answers of the examined people. The International Headache Society Classification (1988) was used to diagnose clinical forms of headaches.

Results: In the examined group the 15 (35%) drivers suffered from headaches of different clinical symptoms. Tension-type headaches were diagnosed in 7 (17.5%) examined persons. The other ones — symptomatic (posttraumatic) headaches or connected with vascular diseases (hypertension) or cranial neuralgia were diagnosed. There were no clinical deficits in the neurological examination in the examined group.

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Abstract — WCN 2013

No: 2900

Topic: 8 — Headache

Spontaneous intracranial hypotension in tuberous sclerosis: Is it a rare association or coincidence?

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Background: Spontaneous intracranial hypotension (SIH) is an important cause of orthostatic headache often associated with minor trauma, connective tissue disorders, and degenerative disc diseases. Tuberous sclerosis (TS), on the other hand, is an inherited neurocutaneous disorder complex involving multiple organs. We present a case of SIH with coincidentally diagnosed TS and speculate an etiological association.

Case: A 36-year-old male patient presented with severe headache and intermittent nausea since fifteen days, which was prominent in upright posture and refractory to analgesics. Neurological examination was normal except for the emergence of headache with erect posture. T2-weighted FLAIR MRI showed subdural hematoma, diffuse dural contrast enhancement and caudal herniation of cerebellar tonsils at the level of foramen magnum. Additionally, calcified subependymal nodules and cortical hamartomas were seen in T2-weighted FLAIR MRI. Further spinal and abdominal imaging demonstrated bilateral renal angiomyolipoma. The patient was diagnosed with definite TS based on the presence of two major criteria, and SIH based on characteristic clinical and imaging findings. Conservative treatment consisting of oral and intravenous hydration, excessive caffeine intake, strict bed rest, and oral theophylline (200 mg/day) provided partial clinical improvement in 18 days and complete improvement at two months.

Conclusion: There are no prior reports of SIH associated with TS according to our knowledge. The potential etiological or simply incidental association between these two diseases should be examined in further research studies. Clinicians should note this possible etiological association when encountered with orthostatic headache and any spectrum of tuberous sclerosis complex.

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Abstract — WCN 2013

No: 2987

Topic: 8 — Headache

Predictors of disability in patients with chronic migraine

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Background: Chronic migraine (CM) is an unfavourable outcome of migraine course. It is often associated with lower QoL and increased disability, and may be due to several factors, including comorbidity to depression, genetic terrain, and medication overuse. A personal factor like self-efficacy and an adequate level of social support may act as protective factors towards disability reduction.

Objective: To assess predictors of disability in patients with CM at the time-point of detoxification.

Patients and methods: In this cross-sectional study, disability was measured with the WHO-disability assessment schedule (WHO-DAS), disease activity with the MIDAS, mood state with the Beck depression inventory (BDI-2), self-efficacy with the global self-efficacy scale (GSE), and social support with the MOS-SSS (Social Support Survey).

Linear regression was performed to assess as to what extent disease activity, mood state, self-efficacy and social support predict disability.

Results: 189 adult patients were enrolled (82.5% women; mean age = 43.8 ± 12; mean MIDAS = 90.1 ± 69.2); mean WHO-DAS score was 31.5 ± 13.9. Linear regression model explains 34.7% of variance, with MIDAS ($\beta = .228$) and BDI-2 ($\beta = .492$) being independent predictors.

Conclusion: Data show that CM patients report high disability, compared to general population (approximately five-folds) and to other neurological diseases such as epilepsy (13.6), myasthenia gravis (18.5), Parkinson's disease (19.3), episodic migraine (21.3) or stroke (25.9). Disability in CM patients seems to be due to poor personal factors, while mood state had a great impact. These results suggest that treatment programmes should take into account mood states in the daily evaluation of CM patients.

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Abstract – WCN 2013

No: 2378

Topic: 8 – Headache

Prevalence and characteristics of headaches in Albanian adolescents

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Background: Headache is a common disorder in all age groups. No study has been carried out to evaluate headache in Albanian adolescents.

Objective: Assessment of headache presence and its characteristics in Albanian adolescents.

Patients and methods: 1380 (820 females, 560 males) high school students aged 16–18 years old (average = 17.1) were interviewed using an eight-item questionnaire for headache evaluation.

Results: 540 students (39.1%) declared headache episodes, females reporting more headache suffering than males. 385 females (47.0%) reported headache versus 155 males (27.7%). 310 students (57.4%) reported a generally stable headache pattern over the past months. Headache for longer than six months was reported by 101 (18.7%) students. 482 (89.3%) declared that headache somehow interferes with activities of daily living. Most reported headaches 5–15 days/month (n = 229, 42.4%) or 1–4 days/month (n = 193, 35.7%), but importantly 67 (12.4%) reported 15 days/months to daily headache episodes. If left untreated, headache would last for 1–4 h in 275 cases (50.9%), but in 82 cases (15.2%) it was reported to last for 4–24 h.

Analgesics were used <1 day/week by 227 (42.0%), 1–2 days/week by 231 (42.8%), >2 days/week by 62 (11.5%) and everyday by 20 (3.7%). Changes in senses immediately before headaches start appear in 193 cases (35.7%).

Conclusions: In the adolescent population, the most common self-reported headache types are tension-type headache (27.5%) and migraine (7.8%), both with a females to male ratio of 3:1. 42% of migraine-reporting adolescents refer aura. Chronic tension-type headache is reported in 4.9%, and medication overuse headache in 5.9%.

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Abstract – WCN 2013

No: 3046

Topic: 8 – Headache

Idiopathic intracranial hypertension presenting as unilateral papilledema: A case report

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Background: Idiopathic intracranial hypertension (IIH) usually presents with papilledema, headache, pulsatile tinnitus, transient visual obscuration, and diplopia. Both papilledema and IIH-related headache are typically bilateral, however asymmetrical or even unilateral localizations are rarely described in literature. Herein, we describe a rare case of a 63-year-old woman who presented with unilateral papilledema and was diagnosed as IIH.

Case report: A 63-year-old woman with a 12 month history of severe headache attacks, (lasting 20–45 min, with a frequency between two and four attacks per day, 4–5 days a week) and transient right visual obscurations. Fundoscopy showed absent venous pulsations with papilledema in the right eye, while the left fundus was normal. Both brain magnetic resonance imaging and magnetic resonance venography were normal. Lumbar puncture, done in the left lateral decubitus, revealed an opening pressure of 330 mm H₂O. Cerebrospinal fluid examinations, including cell count, glucose, total proteins, protein electrophoresis and Ig index were normal. In addition, viral serology and cultures were negative. On the hypothesis of IIH, we prescribed acetazolamide 750 mg/day and topiramate 100 mg/day.

Conclusion: The exact pathomechanism of unilateral papilledema is unknown in IHH. Several mechanisms have been suggested, like optic nerve sheath anomalies, variations of the trabecular meshwork of fibrous adhesions in the subarachnoid space surrounding the optic nerve, and anatomical difference in lamina cribrosa. Unilateral or apparently unilateral papilledema are sometimes signs of intracranial hypertension. Our case report highlights the difficult diagnosis of a highly asymmetric papilledema, and the possible delay of a neurological study.

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Abstract – WCN 2013

No: 3005

Topic: 8 – Headache

Stimulation of the sphenopalatine ganglion (SPG) for cluster headache (CH)-pathway CH-1 – Effectiveness and quality of life through 18 months

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Background: A multi-center study of a novel SPG neurostimulation therapy for chronic CH (CCH) has been conducted.

Objective: The pain and autonomic symptoms of CH result from activation of the trigeminal parasympathetic reflex, mediated through the SPG. We investigated long-term effectiveness of on-demand SPG stimulation in CCH.

Patients and methods: All patients met the ICHD-2 criteria for CCH and were implanted with the miniaturized ATI Neurostimulator. Pain response following SPG stimulation, attack frequency, and Quality of Life (QoL) were analyzed over 18 months. Acute responders achieved relief from \geq moderate pain, freedom from mild pain in \geq 50% of analyzable treatments. Frequency responders achieved a \geq 50% reduction in attack frequency for \geq 2 months.

Results: 43 patients were enrolled, 23 completed follow-up through 18 months following implantation, 10 have not reached 18 months and 10 were terminated (1 – regulatory, 5 – noncompliance, 4 – explanted). Of these 23, 91% responded to SPG stimulation. 81% were frequency and 52% were acute responders. 33% were both acute and frequency responders. Frequency responder reduction was 82% (range: 52–95%) from 15 baseline attacks/week (range: 5–25) to 2.8 (range: 0.4–9.7) and the \geq 50% continuous frequency reduction lasted 215 days (range: 75–408). 86% of patients experienced clinically significant improvements in QoL (SF-36v2 physical and/or mental scores), 70% experienced clinically significant improvements in headache disability (HIT-6) and 82% rated their overall satisfaction with the ATI Neurostimulation System as very good/good.

Conclusion: On-demand SPG stimulation using the ATI Neurostimulation System provides a lasting, clinically significant response and QoL improvements in CCH patients.

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Abstract – WCN 2013

No: 2982

Topic: 8 – Headache

Photophobia is associated with allodynia during migraine attack

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Background: Allodynia is a phenomenon of central sensitization that may lead to migraine transformation. Allodynia is associated with more functional disability and severity of migraine. It is well known that allodynia is positively related to headache frequency, attack duration, illness duration, and unilateral pain. On the contrary, little is known if some of the common accompanying symptoms of the migraine pain are more present in allodynic migraineurs.

Objective: To investigate if some of the common accompanying symptoms of migraine are more frequent in allodynic patients.

Materials and methods: We recruited 200 consecutive migraineurs. Other primary headaches, comorbidity and migraine prophylaxis were exclusion criteria. Each patient was interviewed following a structured questionnaire including the presence or not of photophobia, phonophobia, osmophobia, nausea/vomit, throbbing pain, unilateral pain, autonomic signs, and aura. Allodynia during the migraine attack was measured using the allodynia symptoms check-list 12 (ASC-12); χ^2 test with continuity correction or Fisher's exact test was used for categorical variables.

Results: Forty-two patients (22%) were non-allodynic, 58 (29%) mild-allodynic, 51 (25%) moderate-allodynic, and 49 (24%) severe-allodynic. Photophobia is significantly more frequent in moderate/severe allodynic patients than in non/mild allodynic ones ($P = 0.033$).

Conclusion: Allodynic migraineurs showed a more frequent association with photophobia during migraine attack. We suggest that cortical hyperreactivity and central sensitization may play a role in the light susceptibility of allodynic patients. Other studies are needed to support this hypothesis and clarify the underlying pathophysiological mechanism.

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Abstract – WCN 2013

No: 3036

Topic: 8 – Headache

Dexketoprofen trometamol in the acute treatment of migraine attack: A phase II, randomized, double-blind, crossover, placebo-controlled, dose optimization study

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Background: Migraine is a disabling disease that can significantly affect the quality of life of the sufferers.

Objective: To assess efficacy and tolerability of two doses of dexketoprofen trometamol, 25 mg (DKP25) and 50 mg (DKP50), compared to placebo for migraine acute treatment.

Material and methods: This randomized, double-blind, single-centre, cross-over, placebo-controlled study was performed in 93 patients with at least one migraine attack per month in the preceding 6 months. Participants were enrolled and randomized to DKP25, DKP50 and placebo. Primary endpoints were pain-free and pain relief was at 2 h after drug intake. The presence of accompanying symptoms and adverse events were also recorded.

Results: 76 patients (mean age = 40.5 \pm 10.9 and 61% female) completed the study. At baseline, the mean number of attacks/month was 3.7 \pm 1.3, with a mean duration of 15.4 \pm 13.5 h. Pain relief at 2 h after drug intake was significantly reduced by DKP50 vs placebo (33.8% vs 14.7%, $p = 0.0065$). Both DKP25 (57% vs 25%, $p = 0.0002$) and DKP50 (65% vs 25%, $p < 0.0001$) improved headache relief compared to placebo. Furthermore, both doses of DKP significantly reduced the functional disability at 2 h after drug intake (DKP25, 40% vs 24%, $p = 0.045$ and DKP50, 46% vs 24%, $p < 0.0004$). Both DKP dosages were well tolerated.

Conclusions: Both doses of DKP were effective and well tolerated, with no statistically significant differences in adverse events compared to placebo.

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Abstract – WCN 2013

No: 2868

Topic: 8 – Headache

The efficacy of greater occipital nerve blockade in chronic migraine with triptan overuse

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Introduction: In this study, we evaluate the efficacy of GON blockade in chronic migraine with triptan overuse headache.

Materials and methods: 54 patients were included in the study. These patients were divided into 3 groups. Group 1 received no GON blockade, triptans were decreased gradually and hydration was made. Group 2 received 1 session of GON blockade, triptans were decreased gradually and hydration was made. Group 3 received 3 sessions of GON blockade, triptans were decreased gradually and hydration was made. All patients had blockade with 2 ml of 1% lidocaine bilaterally. Severity of headache was evaluated by VAS. Number of days with headache per month and number of triptan use per month were recorded before treatment and 2–4 months after treatment.

Results: When we evaluate Group 1 according to the severity of pain, a significant decrease was present at the second month of treatment ($p = 0.020$) and this decrease continued at the fourth month ($p = 0.50$). The number of triptan use per month before treatment and after treatment did not show a significant difference. The number of painful days in Group 1 has decreased at the second month after the therapy ($p = 0.032$) but this decrease had ended at the fourth month ($p = 0.098$). When we evaluate Group 2 and 3 for all parameters, a statistically significant decrease was detected 2 and 4 months after treatment ($p < 0.05$ for all parameters).

Discussion: 3 sessions of GON blockade are more effective in reducing the pain severity, number of painful days and number of triptan use per month in patients that have chronic migraine with triptan overuse headache.

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Abstract – WCN 2013

No: 2859

Topic: 8 – Headache

The efficacy of 1 session and 3 sessions of greater occipital nerve blockade in migraine without aura

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Introduction: The blockade of the greater occipital nerve (GON) has been used in the treatment of migraine without aura (MWOA), tension-type headache (TTH) and cervicogenic headache (CH). In this study, we compare the efficacy of 1 session and 3 sessions of GON blockade in MWOA.

Materials and methods: 40 patients (9 male, 31 female) diagnosed with MWOA that had headache >4 days per month during the previous 6 months were included in the study. 20 patients had 1 session of GON blockade with 5 ml of 1% lidocaine (Group 1) and 20 patients had 3 sessions of GON blockade with 5 ml of 1% lidocaine (Group 2) bilaterally. Severity of headache was evaluated by VAS (Visual Analogue Scale) and number of days with headache per month, and duration were recorded before treatment and 6–12 weeks after treatment.

Results: When we compare both groups according to gender, age, pre-treatment pain intensity, duration and frequency, no significant difference was detected ($p > 0.05$ for all parameters). When we evaluate Group 1 according to the frequency and duration of pain before treatment and 6–12 weeks after treatment, there was no statistically significantly difference ($p: 0, 330$; $p: 0, 203$ respectively).

The severity of pain 6–12 weeks after the treatment has decreased ($p: 0, 001$; $p: 0, 013$ respectively). When we evaluate Group 2 for all parameters, a statistically significant decrease was detected 6–12 weeks after treatment ($p < 0,05$ for all parameters).

Discussion: 3 sessions of GON blockade are more effective in reducing the pain severity and frequency. Additional placebo-controlled studies that include larger study populations are needed to confirm our results.

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Abstract – WCN 2013

No: 2857

Topic: 8 – Headache

Age and pericranial tenderness during headache attacks

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Background: Recently it was known that pericranial tenderness (PCT) mechanisms play a role not only in tension-type headache (TH) but also in migraine, whereas we feel muscle fatigability or muscle pain more by aging. In our former study we have paid attention to PCT for migraine patients. According to The International Classification Of Headache II, THs are divided into TH with PCT and TH without PCT. Is migraine with PCT one condition of migraine by aging or a distinguished clinical entity toward migraine without PCT? **Objectives:** Using our data we reanalyzed the relationship between age and PCT in migraine patients.

Patients and methods: We studied PCT during headache attacks using questionnaires given to 363 headache outpatients (153 males, 210 females, with an average age of 47.3 years) from May 2007 to December 2007. All of them completed the questionnaires. We checked distributions of patients with PCT in migraine according to the each generation.

Results: Of 363 patient cases 105 had migraine (26 males and 79 females) and 81 had one or more areas of PCT. Each generation has almost the same rate of migraine patients with PCT. ($P < 0.01$, Mann-Whitney U test).

Conclusion: Our result suggests that migraine with PCT does not concern about aging and there is a possibility that migraine with PCT may be one clinical entity.

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Abstract – WCN 2013

No: 2862

Topic: 8 – Headache

The efficacy of greater occipital nerve blockade in chronic migraine, chronic tension type headache and patients having both headaches

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Introduction: In this study, we compare the efficacy of GON blockade in chronic migraine, chronic tension type headache and patients having both headaches.

Materials and methods: 61 patients (21 migraine (Group 1), 20 TTH (Group 2), both (Group 3)) were included in the study. All patients

had 3 sessions of GON (one injection per week for 3 weeks) blockade with 2 ml of 1% lidocaine bilaterally. Severity of headache was evaluated by VAS (Visual Analogue Scale) and number of days with headache per month, number of analgesic use per month and number of triptan use per month were recorded before treatment and 3 months after treatment.

Results: When we compare both groups according to gender, age, pre-treatment pain intensity, and frequency, number of analgesic use per month and number of triptan use per month; no significant difference was detected ($p > 0.05$ for all parameters). When we evaluate Group 1 and 3 for all parameters, a statistically significant decrease was detected 3 months after treatment ($p < 0.05$ for all parameters). When we evaluate Group 2 according to the frequency of pain, VAS and number of analgesic use per month before treatment and 3 months after treatment, a significant decrease was detected but there was no statistically significant difference in the number of triptan use per month ($p < 0.05$).

Discussion: GON blockade is more effective in patients with chronic migraine and patients with both migraine and chronic tension type headache, but response in TTH is less. Additional placebo-controlled studies that include larger study populations are needed to confirm our results.

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Abstract – WCN 2013

No: 2856

Topic: 8 – Headache

Sinus headache's relationship with primary headaches; clinical and radiological evaluation

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Introduction: Patients suffering from sinus headache (SH), may be able to meet the criteria for primary headaches (PHs) according to IHS criteria. In this study, we aimed to assess the clinical and radiological findings of sinus headache sufferers that meet the criteria of primary headache.

Materials and methods: 39 patients suffering from sinus headache were evaluated clinically and radiologically with paranasal sinus tomography. At CT scans; Lund–Mackay Grading System (LMS) scores, sinus abnormalities and rhinological pathologies were investigated. PHs that also meet the criteria of SH were detected.

Results: When findings are analyzed in terms of meeting the criteria of primary headaches (IHS criteria): 18 (46.1%) patients meet the criteria for migraine, 10 patients (25.6%) for tension type headache (TTH), and 2 patients (3.2%) for cluster headache (CH). Evaluation according to LMS scores did not show significant differences between the migraine group, TTH group, CH group and SH that do not meet the criteria for PH group ($p > 0.05$). All of the patients had radiological rhinological pathology. Groups that meets the criteria of PH had sinus disease more than septal deviation and concha bullosa.

Conclusion: Most patients with sinus headaches also meet the criteria for primary headaches. These primary headache patients mostly have radiological sinus disease. In the evaluation of patients with primary headaches associated with sinus pathology (in terms of

correct diagnosis and treatment), sinus CT findings are important as well as neurological and rhinological examination.

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Abstract – WCN 2013

No: 2846

Topic: 8 – Headache

Survey about headaches in patients with obstructive sleep night apneas

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Introduction: We enrolled 254 subjects, 173 men and 81 women, mean age of 56.8 + 15.0, suffering from obstructive sleep night apnea syndrome to evaluate the prevalence of headache, at the Department of Pneumology of the Teramo Hospital, in a period of 2 years.

Materials and methods: Apneas were graded as mild (AHI between 5 and 10), moderate (AHI between 11 and 20), severe (AHI greater than 20). Headaches were classified according to the ICHD-II criteria. Twenty patients (8.1%) referred a history of primary headache: 3 were affected by migraine (1.2%) and 17 (6.9%) by headache. One hundred and eighty (70.9%) had headache on awakening, with a greater frequency of breathing pauses during sleep, insomnia of central type and episodes of sweating. All underwent an interview with a standardised questionnaire on sleep features.

Results: The severity of obstructive sleep night apneas is related to the frequency of morning headache (frequency greater than in patients with insomnia), suggesting the relevant role of the hypercapnia consequential vasomotor phenomena.

Discussion: The study group has a high prevalence of morning headache confirming results of previous studies concerning both its high frequency and severity in patients with sleep disorders. The strength of our study is the large sample size assessed, and the detailed information collected on sleep disorders and other risk factors for headache.

Conclusions: Our study suggests the need for patients with morning headache to undergo careful screening for sleep disturbances related to breathing disorders, for a correct diagnostic and therapeutic approach.

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Abstract – WCN 2013

No: 2826

Topic: 8 – Headache

Contribution of twenty-one epidemiological factors to migraine disability

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Background: Migraine headache is one of the most important primary headaches. It is associated with a negative impact, reduction of life quality and disability. Epidemiology and impact of migraine on disability are only partially documented in Serbia.

Objective: This study is to determine whether socio-epidemiological parameters have an impact on migraine headache and potentially high scores on the MIDAS questionnaire.

Patients and methods: An unsponsored prospective study has lasted for 5 months. A total of the 543 patients who applied for examination at the Migraine Center and who are, according to the IHS criteria (International Classification of Headache Disorders, 2nd ed., 2004), diagnosed with migraine. 108 were included in the study (average age of 39.96 years, F:M = 88:40). The subjects were randomized. Each patient whose illness protocol number ended with number 5 or 0 is included in the study. The survey was conducted using a standardized closed questionnaire containing a set of 21 social and epidemiological questions (age, gender, qualification, employment, hours of work, professional satisfaction, marital status, housing, smoking, alcoholism, drugs, errors in diet, vegetarianism, hobby, family history, etc.) and a MIDAS questionnaire.

Results: By variance analysis, a statistically significant difference has been found in the score of the MIDAS questionnaire among subjects who smoke and non-smokers ($p < 0.05$). Participants who came into contact, on their workplace, with more than 10 individuals had significantly higher scores on the MIDAS questionnaire ($p < 0.05$), compared to those who came into contact with less than 10 individuals. Other parameters did not show a statistically significant difference.

Conclusion: This study showed that smoking may have an additional impact on the disability of patients with migraine headache, as well as the number of people whom they may come in contact with at the workplace.

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Abstract – WCN 2013

No: 385

Topic: 8 – Headache

Atypical presentation of Tolosa–Hunt syndrome treated with aggressive immunosuppression and radiotherapy

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We present an interesting patient with atypical Tolosa–Hunt Syndrome that was refractory to immunosuppression and needed radiotherapy. A 50 year old Caucasian male presented 3 years ago with gradual onset dull headache for 2 months. This evolved into cluster-like attacks with cranial autonomic features which responded to Sumatriptan. He developed diplopia due to a partial Rt 6th nerve palsy. There were no other visual or neurological symptoms and no systemic manifestations.

The ESR was 38, and CRP, haematology, biochemistry, glucose, lipids, TFT, autoimmune profile, vasculitic screen, treponemal serology, HIV, B12, folate and clotting were all normal or negative. CT scan of the brain was normal. CSF showed no cells and normal glucose, that the protein was raised at 0.78 (0.2–0.04) and the oligoclonal bands were negative. MRI of the brain showed a Rt cavernous sinus lesion that enhanced with contrast (see picture). MRA was normal as well as CT scans of the chest, abdomen and pelvis. Biopsies from the lesion showed non-specific changes, but no malignancy.

His symptoms were steroid responsive with resolution of the 6th nerve palsy, but he continued to have flare ups of severe headaches whenever the steroids were reduced below 50 mg o.d. He tried Cyclophosphamide (for 6 months) with some improvement. Mycophenolate and Methotrexate were tried without benefit. Subsequently, he had radiotherapy with marked improvement. He is back to work now on a small dose of steroids and continues to have occasional migraines.

The MRI lesion resolved (see picture).

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Abstract – WCN 2013

No: 2762

Topic: 8 – Headache

Clinic and Doppler ultrasonography correlation of reactivity of cerebral vessels in patients with chronic tension-type headaches

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The aim of the research was the investigation of the changes of hemodynamics and reactivity of cerebral vessels among the patients with chronic tension-type headaches by using Doppler ultrasonography with compression tests.

We have examined 39 patients. There were 27 patients with chronic headaches of tension in the first group. The second group consisted of 12 healthy people.

While doing research we detected that the patients with chronic tension-type headaches had normal indicators of CO = 1.2 and FAR = 0.79 (in the control group: CO = 1.25, FAR = 0.798). In the compression of carotid the elevation of linear blood flow velocity (LFV) was to 48.2% (to 42.2% in the control group). Postcompression elevation of LFV was in average of 27.3% in comparison to the primary level of LFV (38.2% in the control group). The CO in common arteries was 0.96 ± 0.01 and FAR was 0.6 ± 0.02 and the CO in carotid arteries was 0.72 ± 0.01 and FAR was 0.64 ± 0.22 respectively in the group of patients with chronic headaches of tension. In the control group the CO was 1.25 ± 0.02 and FAR was 0.79 ± 0.01 . As a result of our research it was detected that the changes of reactivity of cerebral vessels towards the decline of LFV were observed in the group of patients with chronic tension-type headaches. Paradoxical reaction of vessels was detected in 19 out of 27 patients with chronic headaches of tension. The data obtained testify compensatory derangement of autoregulation of cerebral vessels in chronic tension-type headaches.

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Abstract – WCN 2013

No: 2742

Topic: 8 – Headache

Cerebrovascular reactivity across the menstrual cycle in young healthy women

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Objective: Cerebral artery function across the menstrual cycle in healthy women may undergo periodic changes along with oscillations of circulating ovarian hormones. The purpose of the study was to assess the relationship of cerebrovascular reactivity in young healthy women in relation to changes of concentrations of circulating ovarian hormones throughout the menstrual cycle.

Subjects and methods: Nineteen healthy nulliparous, right handed, regularly menstruating women (age 23–25 years) underwent color-coded duplex sonography of common (CCA), internal (ICA) and external (ECA) carotid arteries on both sides. Peak systolic, mean and end-diastolic velocities were measured before and 10 min after intravenous administration of 1000 mg acetazolamide (ACE). Hemodynamic changes

in carotid arteries after ACE injection were assessed in relation to serum concentration of estrogen and progesterone on 5th, 13th and 26th days of the cycle, after standardization to a 28-day menstrual period.

Results: After ACE administration the flow velocity in the right CCA and ICA increased by 23% and 35% on 5th day, 12% and 31% on 13th day and 30% and 47% on 26th day respectively, and the changes were significantly larger in relation to insignificant changes on the left side ($F = 6.793$ and $F = 4.098$ respectively; both $P < 0.05$). Multivariable regression analysis showed significant associations between changes of blood flow velocity after ACE injection in the right CCA and ICA and ovarian hormones concentrations ($F = 3.828$, $P = 0.028$ and $F = 3.671$, $P = 0.032$ respectively).

Conclusion: Cerebrovascular reactivity changes across the menstrual cycle are associated with ovarian steroid hormones change, and are more pronounced in the right hemisphere.

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Abstract – WCN 2013

No: 2646

Topic: 8 – Headache

Headache prevalence in Uruguay: A general population study

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Headache is one of the most frequent disorders which lead patients to see a physician; however, there are no prevalence data in Uruguay.

The aim of this study was to know the global prevalence per year of headache and migraine in Uruguay.

A descriptive cross sectional study was performed including people age 15 and above who were interviewed over the phone by physicians with a special training in headache issues.

The diagnostic criteria of IHS were used for migraine diagnosis. The sample was selected by means of a simple random sampling, using the phone directory.

The population's response to the phone interview was very good, with a participation rate of 94.9%. The yearly global prevalence of headache was 58.4% (IC 95% 54.8–62.0), males = 50.8 (IC 95% 43.8–57.8) and females = 65.2 (IC 95% 61.2–69.2). The yearly prevalence of migraine was 6.23% (IC 95% 4.48–7.98), male = 1.5% (IC 95% 0.1–3.11) and female = 10.5% (IC 95% 7.9–13.1).

This paper provides the first epidemiological data on headache in Uruguay. Although there are many headache prevalence studies at international level, it is difficult to compare them due to differences in the populations studied, the methodology employed, the case definition and the prevalence period considered. Therefore, from the analysis of the above data, we can conclude that it is still necessary to make comparable epidemiologic headache studies.

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Abstract – WCN 2013

No: 1705

Topic: 8 – Headache

Neurological manifestations of cerebral venous thrombosis: Retrospective case study of 100 patients with our experience in Kuwait

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Objective: The aim of this study is to highlight the importance of neurological manifestations of cerebral venous thrombosis (CVT) for early diagnosis, management and our experience.

Materials and methods: We performed retrospective case analysis of one hundred patients with CVT, treated at neurology center, Kuwait, from January 2000 to Dec. 2012. The records of patients were retrieved and entered in a data sheet for review.

Results: One hundred patients were included in this study, with a male to female ratio of 1:1.7. The main clinical presentations were neurological. Neurological signs and symptoms were headache (90%), focal neurological deficits (62%) and seizures (42%). Papilledema with raised intracranial pressure was recorded in 30 patients (30%). The venous sinuses involved were superior sagittal sinus in 60% ($n = 60$) and transverse and sigmoid sinuses 63% ($n = 63$). Hemorrhagic venous infarctions were seen in 24% ($n = 24$). Sixty percent of patients recovered within 2–4 weeks and 20% of patients required ICU admission.

Conclusion: Headache and focal seizures were the common neurological manifestations. Early diagnosis and immediate treatment of idiopathic intracranial hypertension (IIH), may prevent the visual loss. Serum D-dimer level is useful for early diagnosis with sensitivity of 54.5%. Heparin is the gold standard treatment for CVT. Our study supports the use of heparin in all patients, even with intracerebral hemorrhage or hemorrhagic venous infarction. The use of steroids and osmotic diuretics as anti-edema measures were found useful, even-though no clear sufficient evidence in the literature.

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Abstract – WCN 2013

No: 2662

Topic: 8 – Headache

Migraine as the manifestation of acute myocardial ischemia

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Objectives: To evaluate migraine (Mg) as a clinical manifestation of acute myocardial ischemia (MI).

Methods: Study sample consists of 55 pts. with MI and headache divided in two groups: Gr. I ($N = 29$) MI with Mg and Gr. II ($N = 26$) MI with other types of headaches. The data was analyzed using Epi Info.

Results: Mean age in Gr. I was 54.06 ± 5.67 and 52.88 ± 4.84 in Gr. II. Headache history was 28.10 ± 12.19 ages in Gr. I vs. 12.55 ± 13.63 in Gr. II ($p < 0.05$) and severity 7.37 ± 2.59 in Gr. I vs. 3.11 ± 2.72 in Gr. II ($p < 0.05$). Headache as the clinical manifestation of MI was 55% in Gr. I and 20% in Gr. II ($p < 0.05$). The severity of headache in the beginning of MI was 5.5 ± 3.55 in Gr. I vs. 1.63 ± 2.97 in Gr. II ($p < 0.05$), during the MI in Gr. I was 6.0 ± 3.33 vs. 1.83 ± 2.99 in Gr. II, and the severity of headache after MI was 4.37 ± 3.43 in Gr. I vs. 1.02 ± 2.38 in Gr. II ($p < 0.05$). Headache in the beginning of MI was 55% pulsating, 20% throbbing and 25% dull in Gr. I vs. 70% dull and 30% pulsating in Gr. II.

Conclusion: Migraine type of headache could be one of the clinical presentations of myocardial infarction suggesting the possible common pathophysiological mechanisms. The clinical presentation of headache associated with acute myocardial infarction was more severe in intensity and pulsating quality. The link between migraine and myocardial infarction needs further investigation.

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Abstract – WCN 2013**No: 2661****Topic: 8 – Headache****Dexamethasone multifunctional role in spreading depression**B. Khodaie. *Neuroscience, Shefa Neuroscience Research Center, Tehran, Iran*

Spreading depression (SD) is a neural hyperexcitability, which spreads to adjoining area of brain cortical; slowly continues through the brain and changes neural electrical and metabolic activity. It has been proved that SD plays a dramatic role in some neurological disorder through the changes it causes. Previous studies applied various types of drugs through diverse pharmacological aspects to relieve SD wave distribution. However most have been abort due to their unpleasant side effects. In the present study we seek for dexamethasone's (Dex) multifunctional properties in order to protect cellular damage and then prevent even other neurological disorder accuracy following SD. Animals were randomly divided into five groups including CTRLs, sham, spreading depression and dexamethasone treatment group. Animals were anesthetized and a cannula as well as an electrode was established above the brain cortex. Intracerebral KCl was used to induce repetitive cortical SD for 4 continuous week. Following each induction memory retrieval tests (T-maze) were done. The brains were removed after 4 weeks. Histopathologically estimation was used to testify dexamethasone effect. In addition dexamethasone effects on blood brain barrier (BBB) to facilitate substance recovery in cerebral fluid were tested by Evans blue. By repetitive SD memory retrieval was impaired due to increment of dark neural seen in several regions of juvenile rat brain. Besides that brain electrical activity determination showed slake of neural activity after SD induction. Neural cell recoveries after SD due to increment of substance passage through BBB were promoted.

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Abstract – WCN 2013**No: 2603****Topic: 8 – Headache****Natural experimentation is a poor method for identifying headache triggers**J. Porter^a, D.P. Turner^b, T. Houle^b. ^a*Advance Neurology & Pain, Cornerstone Healthcare, Advance, USA*; ^b*Anesthesiology, Wake Forest School of Medicine, Winston-Salem, NC, USA*

Background: Headache patients naturally use the covariation of the presence-absence of triggers with headache attacks to assess the potency of triggers. The validity of this natural experimentation has never been investigated. This study examines this process using real-world conditions.

Objective: In this study we set out to determine whether individual headache sufferers can learn about the potency of their headache triggers (causes) using only natural experimentation.

Methods: The similarity of day-to-day weather conditions over four years, as well as the similarity of ovarian hormones and perceived stress over a median of 89 days in nine regularly cycling headache sufferers were examined. An arbitrary threshold of 90% similarity using Gower's index identified similar days for comparison.

Results: The day-to-day variability in just three headache triggers is substantial enough that finding two naturally similar days for which to contrast the effect of a different trigger (e.g., drinking wine versus not drinking wine) will only infrequently occur (e.g., 2 similar stressful days/month). If finding comparable days in one trigger is difficult, finding similar days using multiple triggers becomes increasingly difficult. If there were no covariation in 12 different triggers considered together, using a statistical simulation we would expect a similar day occurring only once every 684 days.

Conclusion: Although assessing the personal causes of headache is an age-old endeavor, many candidate triggers exhibit variability that may prevent sound conclusions without assistance from formal experimentation or statistical balancing.

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Abstract – WCN 2013**No: 2490****Topic: 8 – Headache****Primary stabbing headache: A new dural sinus stenosis-associated primary headache?**A. Ranieri, S. Montella, R. De Simone. *University Federico II, Naples, Italy*

Background: Primary stabbing headache (PSH) is a primary syndrome of unknown aetiology characterized by brief stabs felt in orbital, temporal and parietal areas, whose daily frequency largely varies. PSH incidence resulted 33/100000/year in clinical series with a 35.2% prevalence in a population study. Although considered an infrequent condition, a sinus stenosis-associated intracranial hypertension without papilledema (ss-IHWOP) is much more prevalent than believed and may represent a risk-factor for the progression of migraine, tension type headache and exertional, cough and sexual activity-associated headaches.

Objective: Test the hypothesis that PSH is a sinus stenosis-associated primary headache.

Patients and methods: We retrospectively investigated co-occurrence of sinus stenosis by MR-venography (MRV) in patients referring to our headache centre reporting PSH.

Results: Out of 50 consecutive subjects reporting PSH, 8 (6 F, 2 M) undergone MRV. All showed significant unilateral or bilateral sinus stenosis. Mean age at PSH onset was 35.3 ± 18.9 years. Median frequency of attacks/month was 14 (range = 4–30) with a median frequency of attacks/day of 4 (range = 2–20). Attacks lasted 1–2 s in all but one patient (5 s). Six patients described attacks in temporal or parietal areas, one at the top of the head, and one in occipital area. Seven patients were also diagnosed with migraine without aura and one had isolated PSH. Seven patients responded to indomethacin and one to topiramate (both drugs known to lower CSF pressure).

Conclusion: Our findings indicate that PSH is associated with sinus venous stenosis suggesting that an undiagnosed ss-IHWOP might be involved in PSH pathogenesis.

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Abstract – WCN 2013**No: 2446****Topic: 8 – Headache****Quality of life measured by the Comprehensive Headache-related Quality of life Questionnaire improves after successful detoxification of medication overuse headache**C. Ertsey^a, É. Csépany^a, G. Bozsik^a, I. Kellermann^b, B. Hajnal^b, E. Balogh^b, Z. Nagy^b, D. Bereczki^a. ^a*Department of Neurology, Faculty of Medicine, Budapest, Hungary*; ^b*Semmelweis University, Faculty of Medicine, Budapest, Hungary*

Background: Medication overuse headache (MOH) affects 1% of the population and is notoriously difficult to treat. The cornerstone of its treatment is detoxification supplemented by adequate preventive measures. Measuring quality of life (QOL) is an important means of assessing the impact of headache, and may be used as an endpoint in therapeutic trials. The Comprehensive Headache-related Quality of life Questionnaire (CHQQ) is a new headache-specific QOL instrument that has been validated in migraine and tension type headache.

Objective: To test whether detoxification of patients with MOH can improve the clinical characteristics of the patients' headaches and lead to a better QOL as measured by the CHQQ instrument.

Patients and methods: Patients fulfilling the diagnostic criteria of MOH underwent an inpatient detoxification program. Clinical data were collected using a detailed headache diary. CHQQ was completed at the beginning and at the end of the treatment period. We used nonparametric (Wilcoxon) tests while testing for differences between pre- and post-withdrawal variables.

Results: Ten patients (9 females; mean age = 40.5 ± 15.9 years) were enrolled. After detoxification the clinical characteristics (attack frequency, days with headache, duration and intensity of the attacks) improved significantly ($p < 0.04$). The total score and also the physical, mental and social dimensions of CHQQ showed significant improvements of headache-related QOL ($p < 0.01$).

Conclusion: In this sample detoxification resulted in the improvement of both the clinical characteristics and QOL of patients. Our data suggest that CHQQ may be useful in monitoring QOL in patients undergoing headache prophylaxis.

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Abstract – WCN 2013

No: 2538

Topic: 8 – Headache

A short battery to evaluate cognitive dysfunction in migraine

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Background: Cognitive symptoms during migraine attacks contribute to patients' disability and are sometimes not relieved by migraine acute medication. Measuring the impact of such symptoms is of increasing importance in clinical settings yet no objective measurement of impairment exists in clinical practice.

Objective: We developed a practical and short neuropsychological battery for migraine and tested the performance of migraine patients and controls and the learning effect on repeated short-term applications.

Methods: The battery was applied twice in 24 migraine patients outside an attack and 24 age and sex matched healthy controls, with an interval of one to two months between applications.

Results: The performance of migraine patients and controls in this battery was similar in both applications. In both groups, increased performance was observed in second evaluation that was significant in single test – the Stroop Interference test ($p = 0.002$, multiplicity corrected).

Conclusions: Migraine patients outside an attack show cognitive performance comparable to controls in a short battery testing mainly executive functions. Short-term repeated applications of this battery reveal a slight learning effect that must be recognized in its clinical application.

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Abstract – WCN 2013

No: 1830

Topic: 8 – Headache

Giant cell arteritis and varicella zoster infection: Concomitant causes of temporal headache

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Background: Giant cell arteritis (GCA) and varicella zoster virus (VZV) reactivation are serious but treatable causes of headache, and their relationship is complex. We report a case of concomitant zoster ophthalmicus and GCA.

Case report: A 72 year-old male presented with a week-long severe left fronto-temporal headache and scalp and temporal artery (TA) tenderness. Doppler ultrasound (DU) detected a hypoechogenic halo sign in both TA. GCA was presumed and corticotherapy started. He returned within 24 h, with a zoster rash affecting left ophthalmic nerve region. Valacyclovir was instituted, with improvement. Two weeks later he developed VZV encephalitis, with CSF inflammatory changes and VZV PCR detection. Right TA biopsy (TAB) did not show signs of vasculitis, nor VZV infection by PCR analysis, and corticoids were tapered. One week after corticotherapy withdrawal headache recurred, with left eye vision loss due to optic ischemic neuropathy. ESR and CRP were 98 mm and 13.7 mg/dL, respectively. DU detected recurrence of the halo sign, and corticosteroids for GCA were reintroduced, with sustained benefit after one year of follow-up.

Conclusions: Although VZV vasculopathy is known to mimic GCA, the reverse happened in our case. The initial diagnosis of GCA was reviewed by the coexistence of VZV ophthalmicus and encephalitis in a patient with negative TA biopsy. Corticotherapy withdrawal triggered GCA recurrence. Despite its limitations, DU provided important clues for correct diagnosis. Clinicians should be alert for the possible coexistence of GCA and VZV infection. This case reinforces the possibility of a common pathogenic relationship.

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Abstract – WCN 2013

No: 2516

Topic: 8 – Headache

Vertebral artery dissection mimicking status migrainous

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Background: Vertebral artery dissection (VAD) uncommonly presents as isolated headache. More rarely, it may simulate classical headache syndromes, including migraine. We report a VAD mimicking status migrainous.

Case report: A 41 year-old woman presented a 6-day history of left side throbbing headache, associated with nausea, photophobia and kinesiophobia. She had a past medical history of catamenial migraine and cigarette smoking. Pain persistence, relative refractoriness to analgesics and unusual occipital location (her pain used to be left frontal) led to seek medical evaluation. Observation was normal other than mild left neck tenderness. There was an episode of mild neck pain while doing abdominal crunch exercises, 2 days before headache onset. Head computed tomography (CT) scan and magnetic resonance imaging (MRI), including magnetic resonance venography (MRV), lumbar puncture and transcranial Doppler ultrasound were normal. A cervical Doppler ultrasound suggested a left VAD (V2 segment), and this diagnosis was confirmed by MRI, including cervical magnetic resonance angiography (MRA) and T1-weighted imaging (T1WI) fat-suppression axial sequence. Anticoagulation was initiated and there was progressive recovery.

Conclusion: Our patient's headache fulfilled diagnostic criteria for status migrainous, other than the fact that a secondary cause was identified. Persistent pain and localized tenderness motivated a search for secondary causes, leading to the diagnosis of VAD. This is to our knowledge the first description of isolated status migrainous as the presentation of VAD.

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Abstract – WCN 2013**No: 2410****Topic: 8 – Headache****Cerebral miliary tuberculosis: A case report**

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Background: Tuberculosis is one of the most common infectious diseases worldwide, and constitutes a public health issue in the developing countries. The percentage of extrapulmonary seems higher in people of black ancestry, women, and immunodepressed patients. Miliary tuberculosis is due to the lymphohematogenous spread of tuberculous bacillus from focal lesion. Stereotactic biopsy establishes the final diagnosis. In most cases, the diagnosis is presumptive on clinical and biological findings, and the progression under anti-tuberculosis drugs. We report here a case of an immunocompetent patient with cerebral miliary tuberculosis.

Methods: Neurological examination was done by a neurologist; brain CT-scan, blood chemistry and lumbar puncture were performed.

Results: A 29-year-old taxi driver, alcoholic but stopped two months ago, and smoker, came to our clinic for a progressive headache since five months associated with vesper fever, recurrent vomits, and dry cough. He had a deteriorated state without adenopathy and meningeal syndrome with a conserved consciousness.

Brain CT-scan showed multiple enhanced widespread micronodules with an edema caused by the lesion. Thoracic X-ray identified diffuse micronodular opacities in the two pulmonary fields. Cerebrospinal fluid study showed a pleiocytosis at 50/mm³ with lymphocytes at 85%, total proteins at 2.17 g/l, and a glycorrachie at 0.5 mmol/l. HIV test was negative.

Anti-tuberculosis treatment associated with corticoids led to progressive clinical and biological improvement.

Conclusion: Although uncommon, cerebral miliary tuberculosis should be considered even in immunocompetent patients before intracranial hypertension symptoms.

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Abstract – WCN 2013**No: 2468****Topic: 8 – Headache****Third molar tooth in the etiology of cluster type headache: A case report**

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Background: Headaches are one of the most frequently encountered health problems of multifactorial etiology. The fact that the trigeminal nerve has a role in the etiology of migraine and other headaches is being accepted in the recent years.

Objective: We wanted to report a case of unilateral headache accompanied by autonomic findings that meets the diagnostic criteria for cluster headache.

Patients and methods: A 30-year-old male patient complained of headache for about 6 years. His headaches occurred at certain periods of the year and relieved with painkiller but never completely recovered. His headaches were generally localized on the right side

of his face, occasionally on the left, generally occurring in the night and sometimes at day time. The pain would start behind his eye and on his forehead lasting for 1–2 h, accompanied by ptosis and rhinorrhea. The patient was previously diagnosed with cluster type of headache and gained benefit from oxygen therapy.

Results: On the panoramic dental x-ray images obtained on his controls, horizontally located third molar teeth were seen bilaterally, compressing the neighboring teeth. Especially on the right side, significant contact with the alveolar nerve and inflammation were detected. The complaints were thought to originate from the wisdom (third molar) teeth so they were bilaterally removed.

Conclusion: In the evaluation and treatment of headache etiologies it is of importance in patients with cluster type of headache to evaluate the alveolar branch of the trigeminal nerve, and not to forget the dental pathologies as a causing factor.

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Abstract – WCN 2013**No: 2425****Topic: 8 – Headache****Migraine and hypothyroidism: A possible new comorbidity?**

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Background: Migraine patients suffer from a variety of comorbid conditions.

Objective: The International Classification of Headache Disorders (ICHD-II) included in the secondary headache group a form attributed to hypothyroidism (HT). We assessed the prevalence of HT in migraineurs.

Patients and methods: We retrospectively evaluated 3718 patients diagnosed with primary headaches from 2005 to 2012.

Results: Overall, 98 cases (95 females and 3 males) with HT requiring hormone therapy were observed. Ninety of these cases (2 males) were migraineurs and 8 suffered from tension-type headache (TTH). Therefore, the prevalence of HT was 3.0% in migraine and 1.6% in TTH. In population-based studies the prevalence of HT in the general population resulted to be <1%. Interestingly, in our clinic-based survey HT occurred after migraine onset in 87 patients (96.7%), whereas it preceded migraine in 2 migraineurs and in 3 TTH patients. For the latter subjects headache attributed to HT was ruled out, due to headache persistence after levothyroxine treatment. For 52.0% of patients the headache showed a significant worsening after the onset of HT and hormonal replacement therapy. It is challenging to speculate whether the worsening could be attributable to the hormonal disorder, to levothyroxine treatment or both.

Conclusion: We found a high prevalence of HT in migraine, significantly higher than in the general population. HT should be considered as one of the various migraine comorbidities, even if possible pathophysiologic relationships remain unclear. In case of worsening of pre-existing migraine, thyroid function should be investigated to rule out a possible HT.

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Abstract – WCN 2013**No: 2419****Topic: 8 – Headache****Headache and sleep: A prospective 6 month study**

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Objective: The aim of this study was to evaluate patients with primary headache who had disturbances of sleep.

Background: Several studies have demonstrated a relationship between sleep disturbances and headache.

Design and methods: Diagnosis was made by the use of two special questionnaires, MIDAS and HIT-6. MIDAS and HIT-6 were applied in all patients at screening and at follow-up, six months after screening. Patients' diaries were provided for six months for documentation of headache and sleep disturbances.

Results: 96 patients, mean age of 37 years, were screened during six months. Overall, 71% (68) patients fulfilled criteria for any type of migraine (M), 27% (26) patients met criteria for any type of tension-tip headache (TTH) and 2% (2) patients met IHS criteria for migraine cluster headache (CH). From the 68 patients who met IHS 43% (29) met criteria for migraine with link between migraine and sleep disturbances. The 26 patients with TTH never have sleep problems due to headache. Both patients with CH have a relationship between headache and sleep disturbance. In group with diagnosis of M, migraine attacks occurred during the night or early morning were more intensive in all patients. Mean score on a pain scale of 0–10 were 8 (standard deviation = 2) in this group and MIDAS grade III (moderate disability) in 35% (24) patients and grade IV (severe disability) in 65% (44) patients.

Conclusions and relevance: Our study evaluated the relationship between sleep and headache.

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Abstract – WCN 2013

No: 2328

Topic: 8 – Headache

Effect of Yoga therapy on migraine patients: A clinical and cardiac autonomic study

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Background: Yoga is a traditional Indian psycho-philosophical-cultural method known to rectify the autonomic imbalance in various disorders. However, there is not much evidence regarding autonomic balance as measured by heart rate variability following adjuvant Yoga therapy in patients with migraine. We aimed to compare and evaluate the effects of Yoga therapy with conventional care versus conventional care by clinical outcome and autonomic function tests.

Methods: Sixty migraine patients were recruited & were randomized into conventional care and conventional care and Yoga therapy using a concealed allocation protocol. Yoga group received additional Yoga practice for 5 days a week for 6 weeks. Clinical assessment (frequency, intensity of headache, HIT-6) was done at baseline and at the end of intervention. An autonomic function test was done pre and post intervention.

Results: Yoga with conventional care group showed significant improvement in clinical variables. In addition there was statistically significant improvement in the cardiac autonomic parameters in patients receiving Yoga as adjuvant therapy.

Conclusion: Six weeks of conventional care and conventional care with Yoga intervention showed significant clinical improvement in both groups. Headache frequency and intensity were reduced more in Yoga group with conventional care than the conventional care group. Study also showed that Yoga therapy enhances the vagal tone and decreases the sympathetic drive, hence improving the autonomic balance. Thus, Yoga therapy can be effectively incorporated as an adjuvant therapy in migraine patients.

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Abstract – WCN 2013

No: 982

Topic: 8 – Headache

Transient headache and neurological deficits with cerebrospinal fluid lymphocytosis associated with IgM antibodies to the Epstein-Barr virus viral capsid antigen

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Introduction: Some authors have suggested that the syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL) results from an immunological response directed against a viral agent.

Case report: A 19-year right-hander old male with no previous history of disease, in particular of migraine, was admitted following two episodes of transitory aphasia combined to acute headache and vomiting. Emergency cerebral MR imaging with intracranial and cervical MR angiography did not reveal any vascular or parenchymal lesions. An electroencephalogram revealed the presence of non-epileptiform slowing on the left hemisphere. Examination of the cerebrospinal fluid (CSF) found 250 lymphocytes/ml, normal glucose levels and a minimal increase in protein concentration (0.54 g/l). A PCR performed on CSF with primers specific to HSV and EBV delivered negative results. In contrast, EBV serological test detected isolated IgM viral capsid antigen. Clinical symptoms disappeared within 24 h. EBV serological controls at 4 weeks showed an unchanged profile. Compilation of the clinical and laboratory features led us to pose a diagnosis of HaNDL.

Discussion: Our patient had increased IgM directed against the EBV VCA. These results can be ascribed to an EBV primary infection or to a non-specific reaction. We posit that a virus infection elicits an immune response that includes the production of antibodies.

Conclusion: Throughout this case report, we have argued about a possible abnormal immune reaction elicited by an infection that would trigger the development of this uncommon affection.

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Abstract – WCN 2013

No: 2338

Topic: 8 – Headache

Presence of headache triggers in migraineurs

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In people with migraine, headache triggers increase the probability of a migraine attack over a short period, usually shorter than 48 h. Very rarely the same triggers may provoke headache in persons who do not suffer from migraine. Migraineurs have one or more precipitating factors. Attacks may be provoked by endogenous factors and environmental factors that are in constant interaction and influence the genetic predisposition to migraine. Although literature data show that between 58% and 90% of migraineurs report some precipitating factors, their real effect has not been clearly established yet.

In our research that included 1022 subjects, 579 suffered from headache, and of these, 169 had migraine, 224 had tension-type headache, and 186 had other types of headache. The questionnaire about the characteristics of headache included a question related to the presence of headache triggers. Subjects could choose between one or more listed triggers or name another triggers.

The results showed the following possible headache triggers in migraineurs: consumption of certain foods (5.9%), consumption of certain beverages (9.5%), mental strain (58.6%), fatigue (45.6%), strong odors (9.5%), changes in atmospheric pressure (49.1%), menstruation (47.3% of all subjects with migraine, or 54% of women migraineurs), and other triggers (15.4%).

Our results show that mental strain, menstruation, changes in atmospheric pressure and fatigue are the most significant triggers of migraine headache. The effects of food and drink as precipitating factors are present but not significant. These findings are important for planning treatment and educating patients.

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Abstract – WCN 2013

No: 2376

Topic: 8 – Headache

Transcranial electro stimulation in the acute treatment of chronic migraine patients with lateral cerebral ventricles asymmetry

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Background: Transcranial electro stimulation (TES) is an effective acute treatment for chronic migraine (CM), one of the possible mechanisms being the increase of β -endorphin level. Pharmacologic CM preventive treatment seemed to be less efficient if a “benign” lateral ventricles asymmetry (LVA) was associated. The aim of our study was to estimate the efficiency of TES in the acute treatment of CM with pronounced LVA.

Methods: We conducted a study on 52 patients diagnosed with CM (ICHD-II, revised 2006) treated by TES (39 patients) and by an active placebo (AP) device (15 patients). LVA was graded based on ventricular frontal horns asymmetry index (AI): control group comprised 26 patients with CM + minimal LVA (AI = 1.0–1.24), the study group – 28 patients with CM + pronounced LVA (AI \geq 2.0). Pre- and post-treatment analyses of serum β -endorphin levels and headache severity on visual analogical scale were performed.

Results: There was a significant headache intensity decrease only in the control group (TES – 4.89 ± 2.71 vs. 2.74 ± 3.05 , $p < 0.01$ and AP – 4.71 ± 1.98 vs. 2.43 ± 1.39 , $p < 0.01$). Most of pronounced LVA patients (71.4%) demonstrated a decrease of post-treatment serum β -endorphin level instead of the expected increase (TES – 26.89 ± 12.83 vs. 19.63 ± 11.19 , $p > 0.05$ and AP – 33.45 ± 31.83 vs. 25.82 ± 34.63 pmol/l, $p > 0.05$).

Conclusions: Our results indicate a lack of efficiency of TES in the acute treatment of CM if a pronounced LVA is associated, that could suggest an eventual dysfunction of the opioid system in LVA patients with an unexpected reaction of diminishing β -endorphin level after TES.

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Abstract – WCN 2013

No: 2389

Topic: 8 – Headache

The prevalence of cluster headache in the elderly is higher in women than in men

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Background: Cluster headache (CH) is considered a disorder of young men, peaking at 30 years of age. It is well known that CH affects men more frequently than women, the gender ratio ranging from 4.4:1 to 2.5:1 in various studies.

Objective: We evaluated the patients with CH aged 65 years and older.

Patients and methods: For the last 16 years we have observed 254 patients suffering from CH. Out of these cases, 42 patients (16.5% of the whole population) were older than 65 years.

Results: In this group of elderly patients, 24 were females (57.1%) and 18 were males (42.9%). We diagnosed 4 patients with CH (only one bout, according to the International Classification of Headache Disorders), 24 with episodic CH, and 14 with chronic CH. The onset occurred in ages 35–44 years for 21.4% of cases, in ages 45–54 years for 16.7%, in ages 55–64 years for 28.6% and after the age of 65 years for 33.3%. Notably, in the latter subgroup, the females significantly prevailed, accounting for 78.6% of cases.

Conclusion: In CH patients over the age of 65 years, females represented the great majority of cases, in contrast with the evident male preponderance in the previous ages. Apparently peculiar to the female distribution, an increased frequency of CH appears to occur in middle-age and elderly patients. To our knowledge, we report the patient with the oldest age at onset (a 93-year-old woman) and the largest case series of CH elderly patients published in the literature to date.

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Abstract – WCN 2013

No: 2404

Topic: 8 – Headache

First experiences with onabotulinumtoxin A (Botox) in patients with chronic migraine in Czech Republic—preliminary data

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Background: Several studies have demonstrated an effect of onabotulinumtoxin A in patients with refractory chronic migraine.

Design and methods: ICHD-II criteria for chronic migraine were applied to symptoms described by headache patients. Diagnosis was made by ICHD-II criteria for chronic migraine. MIDAS and HIT-6 were applied in all patients at week 0 and at follow-up, three months and six months after first injection. Patients' diaries were provided for six months for documentation of headache episode frequency. All patients had poor headache control, poor quality of life, high disability scores and high acute medication intake. Subjects were treated with injections every 12 weeks of onabotulinumtoxin A (150 U). The primary endpoint was mean change from baseline in headache episode frequency at week 24. The second endpoint was mean change from baseline in HIT-6 and MIDAS at week 24.

Results: We reported preliminary data of 7 patients (2 males and 5 females), mean age of 56 years (median = 48 years), who fulfilled ICHD-II criteria for chronic migraine. Mean score on a pain scale of 0–10 were 8 (standard deviation = 2) at week 0, MIDAS grade IV (severe disability) at week 0 and HIT-6 mean score of 65 (median = 66) at week 0. Two patients (1 male and 1 female) interrupted treatment for lack of efficiency at week 12. The rest of patients still take the treatment. The subjects did not have any adverse events.

Conclusions and relevance: We need to treat more subjects with onabotulinumtoxin A for more experience.

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Abstract – WCN 2013**No: 2412****Topic: 8 – Headache****Cluster-migraine does exist**C. Lisotto^a, F. Mainardi^b, F. Maggioni^a, G. Zanchin^a.^aDepartment of Neurosciences, University of Padua, Padua, Italy;^bDepartment of Neurology, Hospital of Venice, Venice, Italy

Background: Cluster headache (CH) is a well definite primary headache. When attacks fulfill all but one of the criteria for CH, established by the International Classification of Headache Disorders (ICHD-II), probable CH should be diagnosed. This entity requires one of the following conditions:

- 1) attacks lasting >180 min,
- 2) attacks without local autonomic signs or restlessness,
- 3) sporadic (less than one every other day) attacks.

Objective: We evaluated the patients previously diagnosed with probable CH.

Patients and methods: For the last 16 years we have observed 254 patients suffering from CH. Out of these cases, 35 (19 males and 16 females) could not fulfill all the criteria for CH.

Results: In this population we could distinguish 4 different subgroups. They could be diagnosed with CH except for:

- 1) attack duration >180 min, ranging from 4-to-8 h (7 cases),
- 2) absence of local autonomic signs or restlessness (5 cases),
- 3) sporadic attacks, with no cluster periodicity (11 cases).

We could also identify a fourth subgroup of 12 patients without cluster pattern and attack duration >180 min, ranging from 3-to-5 h. The first three subgroups meet the criteria for probable CH. The fourth subgroup does not fulfill the criteria either for probable CH or probable migraine without aura (MO).

Conclusion: Patients sometimes present with clinical scenarios having characteristics of both MO and CH, but they do not fully meet ICHD-II criteria for either condition. These occasions provide diagnostic challenges and account for the controversial form of “cluster-migraine”, which was previously considered as a variant of CH.

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Abstract – WCN 2013**No: 1631****Topic: 8 – Headache****Sexual headache related to ginkgo glycosides**H. Akgun^a, M. Yucel^b, S. Tasdemir^c, S. Alay^c, O. Oz^c, U.H. Ulas^c, S. Demirkaya^c. ^aDepartment of Neurology, Etimesgut Military Hospital, Ankara, Turkey; ^bDepartment of Neurology, Kasimpasa Military Hospital, Istanbul, Turkey; ^cDepartment of Neurology, Gulhane Military Medical Academy, Ankara, Turkey

Background: The heart rate and the blood pressure rise during sexual activity. There are two mechanisms in sexual activity causing headache. It is stated that these two mechanisms are related to vascular structure constriction or muscle constrictions. Although there are many reasons of sexual headaches it has been stated that they may develop due to medicines. Especially, it has been proved that birth control pills cause sexual headaches.

Objective: In our manuscript we would like to report a sexual headache due to the use of ginkgo glycosides.

Patients and methods: A 29-year-old male patient presented to the neurology outpatient clinic with a complaint of severe headache

during sexual intercourse. The patient stated that in order to lessen his forgetfulness and strengthen his memory, upon the recommendation of his friend he used 19.2 mg ginkgo glycosides twice a day for 2 weeks. Two times during orgasm he had suffered severe, bilateral, throbbing headache. He had no headaches other than those two during the orgasm. His neurological examination was normal.

Results: The brain CT and MRI were reported as normal. The patient's medication was terminated and did not suffer a headache since.

Conclusion: Headache is one of the adverse reactions of ginkgo glycosides. However, being first case presenting as a sexual headache that we know of, we found it worth reporting.

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Abstract – WCN 2013**No: 2233****Topic: 8 – Headache****Cluster headaches: Underestimated medical problem in Armenia**H.R. Vekilyan^a, A.H. Karapetyan^a, E.M. Gevorkyan^b, H.M. Manvelyan^a. ^aNeurology, Yerevan State Medical University after M. Heratsi, Armenia; ^bNeurology, National Institute of Health, Yerevan, Armenia

Objectives: Cluster headaches (CH) are one of the rare forms of primary headaches. The prevalence of CH is less than 1% and mostly affects men. Despite the presence of precise diagnostic criteria of CH in the country, it still remains undiagnosed and as a result unsuccessfully treated. Although many studies of primary headaches are conducted in the world, data on both abundance and incidence of this type of headaches are still lacking in Armenia.

The aim of this study was to investigate prevalence, correct diagnosis and management of primary headaches, including CH, in Armenia.

Study design: 2000 special questionnaires (according to recommendations of International Headache Society) were created and distributed among matching population in the capital city, and rural area as well. In the study 489 (356 women/133 men) patients with primary headache (migraine, tension type and cluster headache) were included. Age of participants was 18–60 years.

Results: Data analysis revealed that 11 patients, or 2% of respondents, suffer from CH (7 males and 4 females). All of them had previous medical counseling, misdiagnosed and unsuccessfully treated as trigeminal neuralgia (9), or migraine (2). Correct diagnosis and appropriate medical management were paramount in effective treatment.

Conclusion: All together our data is matching to similar international studies, but with some prevalence in cluster headache. Another issue is that despite of wide distribution, many neurologists are mistaking cluster type headaches with trigeminal neuralgia or migraine. More educational work toward understanding and correct assessment of headaches must be done.

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Abstract – WCN 2013**No: 2228****Topic: 8 – Headache****Altered resting state functional connectivity in patients with headache and medication overuse**

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Background: Medication-overuse headache (MOH) is defined as headache that develops or significantly worsens during medication

overuse. Functional and structural alterations have been described in MOH, including hypometabolism in orbitofrontal regions (Fumal et al., 2006) and grey matter increases (Riederer et al., 2012) in brainstem regions.

Objective: In episodic migraine, altered functional connectivity (FC) between areas involved in pain modulation and nociceptive processing has been described. It was the aim of the study to investigate potential FC differences in MOH.

Material and methods: We recorded resting state (6 min, eyes closed) functional magnetic resonance imaging data from 17 healthy controls (HC, mean age: 41.6, range: 31–65 years) and 11 age-matched ($p = 0.54$) patients with MOH according to diagnostic criteria of the International Headache Society (mean age: 43.6, range: 31–61 years). Participants with other neurological- or severe psychiatric disorders were excluded. Voxel-to-voxel functional connectivity (FC) was applied to estimate (whole-brain) spatio-temporal correlations within- and between-groups ($p < 0.01$, FDR-cluster corrected)

Results: We found exclusively hyperconnectivity in HC in the right insular cortex, middle temporal gyrus, posterior thalamus, premotor cortex, middle cingulate cortex, orbitofrontal cortex, and periaqueductal gray (at $p < 0.05$, corrected). In contrast, no hyperconnectivity were observed in patients with MOH at all.

Conclusion: Hypoconnectivity between pain modulating regions, such as insular- and cingulate cortex and posterior thalamus, is consistent with dysfunction of pain modulating systems in MOH patients. Further, the decreased FC in the orbitofrontal cortex might indicate a disturbed reward system or a drug addiction phenomenon.

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Abstract – WCN 2013

No: 2206

Topic: 8 – Headache

Trans-cutaneous electric nerve stimulation in chronic migraine patients with tender points

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Background: The therapeutic effect of neuromodulation has been investigated thoroughly in patients with chronic daily headache and has shown promising results.

Objectives: In our case series study, we tried to study the effect of trans-cutaneous electric nerve stimulation (TENS) on the frequency and severity of headache days in patients with chronic migraine with tender points in their scalp.

Methods: Our 5 patients received 3–10 successive TENS sessions on their tender points. They did not change their prophylactic medications during the study period. Patients maintained a headache diary for one month before and after the stimulation sessions. Our primary outcome was reduction in headache days, while our secondary outcomes were reduction in severity, headache index and in the number of rescue medications.

Results: There were significant reductions in the number of headache days ($p = 0.043$), headache intensity ($p = 0.038$) and headache index ($p = 0.043$) with nearly significant reduction in number of abortive pills ($p = 0.068$) yet with insignificant reduction in attack duration ($p = 0.498$).

Conclusion: The use of tender nerves as a clinical determinant of the subsequent efficacy of peripheral neuromodulation seems to be a reasonable strategy to manage selected cases of chronic migraine.

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Abstract – WCN 2013

No: 2160

Topic: 8 – Headache

Clinical effectiveness of osteopathic manipulative treatment versus sham therapy and usual care in chronic migraine: 3 armed RCT

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Migraine is a common disorder with population prevalence in Europe of 6% in men and 18% in women. Recent studies documented controversy results in relation to the benefit of the application of OMT in migraine and those are even more unclear if CAM are considered.

The aim of this 3-armed RCT study is to determine the effectiveness of OMT on a sample of subjects affected by migraine evaluated using the HIT-6 questionnaire, drug assumption, days of migraine, pain intensity and functional disability.

The study was carried out in the Department of Neurology of Ancona's United Hospitals in the period between March and November 2010. 105 patients entered in the study and were randomly divided into three groups: OMT, sham therapy and triptans only. Patients received 8 treatments in a study period of 6 months. Data were analyzed by repeated-measures analysis of variance (ANOVA).

At baseline, groups were comparable in terms of biometric characteristics and severity of migraine. At the end of the study period repeated measures ANOVA showed that HIT-6 was significantly greater in OMT group. Drug assumption, days of migraine, pain intensity and functional disability scored significantly lower in the OMT and sham therapy groups compared to triptans group. Tukey's post-hoc comparisons showed that OMT was associated to an improvement of all outcomes measured compared to sham and triptans groups. These findings suggest that OMT can be considered a valid procedure for the management of patients with migraine.

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Abstract – WCN 2013

No: 1301

Topic: 8 – Headache

Efficacy of MAP0004 in treating subjects with severe migraine pain: A subpopulation analysis

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Background: The treatment needs of patients with severe migraine are often unmet by available therapies. MAP0004, an investigational product that delivers dihydroergotamine systemically via oral inhalation, was shown to be superior to placebo for the acute treatment of migraine in a phase 3 clinical trial.

Objective: This post hoc analysis of phase 3 data was performed to assess the efficacy of MAP0004 in treating severe migraine pain.

Patients and methods: This analysis included 366 subjects who treated migraine while experiencing severe pain during the double-blind period. Subjects reported pain levels at 10, 30, and 60 min and at 2, 4, 24, and 48 h after treatment.

Results: Subjects with severe migraine pain receiving MAP0004 experienced statistically significant pain relief compared with subjects receiving placebo at 10 min and at all subsequent time points. Significantly more subjects were pain free in the MAP0004 group than in the placebo group at 60 min and at all subsequent time points. Rates of sustained pain relief and sustained pain free for 2–24 h and 2–48 h

were statistically significantly higher in the MAP0004 group than in the placebo group. Headache recurrence over 24 h was reported by 6.2% of subjects with severe migraine pain receiving MAP0004 compared with 18% of subjects receiving placebo.

Conclusion: This post hoc analysis shows that MAP0004 was effective in the acute treatment of severe migraine pain in subjects enrolled in a phase 3 clinical trial.

Study supported by MAP Pharmaceuticals, Inc., a wholly owned subsidiary of Allergan, Inc.

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Abstract – WCN 2013

No: 2094

Topic: 8 – Headache

White matter microstructure abnormalities in pediatric migraine patients: In vivo measures of brain hyperexcitability?

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Background: Diffusion tensor (DT) magnetic resonance imaging (MRI) provides several quantities with the potential to disclose white matter (WM) microstructure abnormalities. Among these, fractional anisotropy (FA) reflects axonal integrity and fiber organization, mean diffusivity (MD) measures the overall magnitude of diffusion, axial diffusivity ($\lambda_{||}$) is associated with fiber density and axon intrinsic characteristics, whereas radial diffusivity (λ_{\perp}) reflects the degree of myelination.

Objective: To explore abnormalities of WM microstructure in pediatric migraine patients using DT MRI and two different methods of analysis.

Patients and methods: Using a 3.0 Tesla scanner, dual-echo and DT MRI scans were acquired from 15 pediatric migraine patients and 15 age-matched controls. Tract-based spatial statistics (TBSS) analysis and a DT probabilistic tractography of the major brain WM tracts were performed.

Results: Both TBSS and DT tractography analysis showed significant lower MD, $\lambda_{||}$ and λ_{\perp} of the brainstem, thalamus, fronto-temporo-occipital lobes (right cingulum, corpus callosum, optic radiation, fronto-occipital fasciculus and corticospinal tract, bilaterally) in migraine patients vs controls. Migraine patients also experienced increased FA of the left optic radiation. Reduced MD of the right cingulum was correlated with disease duration.

Conclusions: Pediatric migraine patients present diffuse WM microstructural abnormalities. Higher FA and lower MD, $\lambda_{||}$ and λ_{\perp} might be explained by repeated neuronal activation, that is likely to lead to cell swelling and might stimulate the activity-dependent myelo-modulation, or by the presence of higher neuronal and synaptic densities in migraine patients compared to controls. Both these mechanisms would reflect a hyperexcitability of the brain in migraine patients.

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Abstract – WCN 2013

No: 2080

Topic: 8 – Headache

The prevalence of primary headache disorders in Saudi Arabia

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Background: Epidemiological data on primary headache disorders in Saudi Arabia is lacking. Very little population-based information is available on migraine and tension type headaches from Eastern Mediterranean region or from other Arabic speaking countries. This was a project within the Global Campaign against headache.

Objective: To estimate the prevalence of common headache disorders in Saudi Arabia.

Methods: Country-wide population-based cross-sectional survey of the Arabic-speaking adults (18–65 years) living in Saudi Arabia. Interviewers were trained surveyors. Access was by mobile phone, with sampling by random-digit dialing. Diagnoses were made algorithmically: the Arabic translation of a previously developed questionnaire was validated in a sub-sample re-interviewed by a neurologist.

Results: Among 2421 respondents (male 62%, female 38%; mean age 32.2 ± 12 years), the observed 1-year prevalence of all headaches was 63%, of migraine 32%, (female 32.8% and male 25.7%), of TTH 27% and of medication-overuse headache (MOH) 2.7%. Odds ratio of migraine, TTH, and MOH for female compared to male were (OR = 1.9), (OR = 0.7) and (OR = 3.4), respectively.

Conclusions: Headache disorders, especially migraine, are highly prevalent in Saudi Arabia. The high prevalence of migraine in men requires further exploration. Possible causes include poor sleep habits and hot weather that are commonly experienced.

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Abstract – WCN 2013

No: 2114

Topic: 8 – Headache

Is gray matter atrophy a biomarker of migraine? A study in pediatric patients with migraine

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Background: Voxel-based morphometry (VBM) studies in adult patients with migraine have consistently demonstrated atrophy of several gray matter (GM) regions involved in pain processing. Whether abnormalities of these areas represent a phenotypic biomarker of this condition, or, are the consequence of the repetition of migraine attacks is still controversial.

Objective: To assess the regional distribution of GM and white matter (WM) abnormalities in pediatric patients with migraine using VBM. The correlations between regional volumetric abnormalities and clinical manifestations of the disease were also investigated.

Patients and methods: Using a 3.0 Tesla scanner, brain T2-weighted and 3D T1-weighted scans were acquired from 12 pediatric migraine patients (seven with visual aura and five without aura) and 15 age-matched controls. Volumetric abnormalities were estimated using VBM.

Results: Compared to controls, pediatric patients with migraine experienced a significant GM atrophy of several regions of the frontal and temporal lobes which are part of the pain-processing network. Increased GM volume was detected in the right putamen in pediatric migraine patients vs. controls. Compared to patients without aura, those with aura had an increased GM volume of the left fusiform

gyrus. No significant WM changes were detected. No correlation was found between GM abnormalities and disease duration and frequency of attacks.

Conclusion: Cortical abnormalities do occur in pediatric patients with migraine. The presence of these abnormalities early in the course of the disease, and the absence of correlation with patients' clinical characteristics suggest that they may represent a phenotypic biomarker of this condition.

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Abstract – WCN 2013

No: 2065

Topic: 8 – Headache

Headaches among medical students: Non-expected conclusions

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Background: The knowledge of the most common headaches is essential for every physician.

Objective: We decided to strengthen the importance of knowledge by making medical students pay attention to their own or their colleagues' headaches.

Methods: We interviewed 61 students, from the first, second and fourth grades. They answered a questionnaire about the characteristics of their headaches (if they suffered from those conditions). They also mentioned if they take medicines to prevent the headaches and what happens with their symptoms during the days they are submitted to routine examinations.

Results: Twenty-five percent of the students had migraine and 59% had tension-type headache. Among those suffering from migraine, only 7% were taking daily medicines to prevent the pain. Considering the whole group of students with headaches, interestingly, 25% improved during the examinations periods.

Conclusion: The percentage of medical students suffering from migraine and tension-type headache is not different from that of the general population. We have to stimulate the students to look for proper attention. This is particularly true for those with migraine. Indeed, less than 10% are taking medicines to prevent the crisis. The common idea that the examination period could trigger the symptoms of the headaches may not be true. One in four of our students actually improved in the days they were tested.

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Abstract – WCN 2013

No: 1971

Topic: 8 – Headache

A case of multiple sclerosis presenting as trigeminal neuralgia

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Trigeminal neuralgia is characterized by recurring paroxysmal and severe pain in the distribution of the trigeminal nerve. In multiple sclerosis it occurs more commonly than in the general population, and it is more likely to be bilateral and to occur at a younger age. In most cases, other symptoms of multiple sclerosis preceded trigeminal neuralgia.

A 45-year-old woman presented with sharp and shooting pain on her right face for 2 years. The pain was in the distribution of the third division of right trigeminal nerve and often triggered by light touch. The pain had been initially well controlled with carbamazepine 200 mg bid, but had recently worsened despite increasing the dosage to 300 mg bid. The neurologic examination was normal including facial sensation. The MRI

showed a hyperintense linear lesion along the right intramedullary trigeminal nerve root on T2-weighted image and multiple white matter lesions compatible with dissemination in space of multiple sclerosis. Follow up MRI revealed dissemination in time with new lesions in T2-weighted image. The patient has received interferon beta 1a.

We report a rare case of multiple sclerosis presenting as trigeminal neuralgia and review the MRI findings of trigeminal neuralgia in multiple sclerosis.

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Abstract – WCN 2013

No: 1965

Topic: 8 – Headache

Relapse in acute migraine treatment: Comparison of frovatriptan with other triptans

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Background: Besides abrupt relief from pain, relapse is a very important outcome parameter for treatment of acute migraine attacks. The International Headache Society (IHS) defined relapse as the most important measure of drug efficacy after pain freedom at 2-h.

Objective: The relapse rates of frovatriptan were compared to those of rizatriptan, zolmitriptan, and almotriptan by pooling together data from three double-blind, randomized, controlled, cross-over trials with an identical design.

Methods: Patients aged 18 to 65 years with migraine with or without aura (IHS criteria) and having experienced one to six migraine attacks per month for six months prior to entry into the study, were enrolled. Patients had to treat three consecutive migraine attacks with frovatriptan 2.5 mg and three with a comparator (rizatriptan 10 mg, zolmitriptan 2.5 mg, or almotriptan 12.5 mg).

Results: 346 patients were included in the intention-to-treat analysis. They treated 987 attacks with frovatriptan and 986 with another triptan. Pain-free at 2-h did not differ between frovatriptan (30%) and the other three triptans (34%, OR = 1.20; CI = 0.98–1.46; $p > 0.05$). The rate of 24-hour relapse was lower under frovatriptan than under other triptans: 16.2% vs. 23.2% (OR = 0.64; CI = 0.43–0.95; $p < 0.05$). This was the case also for the 48-hour relapse rate, which was 26.7% under frovatriptan and 39.7% under comparators (OR = 0.55; CI = 0.39–0.77; $p < 0.001$).

Conclusion: Frovatriptan has a lower recurrence rate than the compared triptans and this difference is clinically relevant. The low relapse rate of frovatriptan is likely explained by its specific pharmacological properties with a long half-life time.

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Abstract – WCN 2013

No: 1952

Topic: 8 – Headache

Interictal increase in CGRP levels in peripheral blood as a reliable biomarker for chronic migraine

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Objective: To determine CGRP levels outside migraine attacks in peripheral blood as a potential biomarker for CM.

Methods: Women older than 17 and diagnosed as CM were recruited. Matched healthy women with no headache history and women with EM served as control groups, together with a series of patients with episodic cluster headache in a pain-free period. CGRP levels were determined in blood samples obtained from the right antecubital vein by ELISA outside a migraine attack and having taken no symptomatic medication the day before. Due to ethical reasons, preventatives were not stopped.

Results: We assessed plasma samples from 103 women with CM, 31 matched healthy women, 43 matched women with EM and 14 patients with episodic cluster headache matched for age. CGRP levels were significantly increased in CM (74 · 90 pg/ml) as compared to control healthy women (33 · 74 pg/ml), females with EM (46 · 37 pg/ml) and episodic cluster headache patients (45 · 87 pg/ml). Thresholds of 43 · 45 and 58 · 22 pg/ml optimize the sensitivity and specificity to differentiate CM from healthy controls and episodic migraine, respectively. In the CM group, CGRP levels were significantly increased in women with a history of migraine with aura vs. those only experiencing migraine without aura. Variables such as age, analgesic overuse, depression, fibromyalgia, vascular risk factors, history of triptan consumption or kind of preventative treatment did not significantly influence CGRP levels.

Conclusion: Increased CGRP level measured in peripheral blood outside migraine attacks and in the absence of symptomatic medication seems to be a reliable marker for CM.

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Abstract — WCN 2013

No: 826

Topic: 8 — Headache

Frovatriptan in the treatment of menstrually related migraine: Evidence from late trials and current recommendations

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Menstrually-related migraine (MRM) is a subtype of migraine without aura, with attacks occurring on or between 2 days before the onset of menstruation and the first 3 days of bleeding, in at least two out of three menstrual cycles and additionally at other times of the cycle. Menstrual attacks are generally more severe and disabling, last longer, frequently relapse and are less responsive to symptomatic medication than attacks at other times of the cycle.

MRM treatment is based on acute or prophylactic strategies, though predictability of menstrual attacks offers the opportunity of effective perimenstrual prevention. Triptans, including one of the latest representatives of this class, frovatriptan, are indicated as first line treatment for moderate-severe migraine attacks, including MRM. Frovatriptan is recommended by guidelines as short-term preventive treatment of menstrual attacks that do not respond adequately to acute treatment. Results of post-hoc analyses of randomized, cross-over, direct comparative trials also reinforce the role of frovatriptan in the acute treatment of MRM. In these studies, including 187 women, frovatriptan showed pain free and pain relief rates similar to those of other triptans (23 vs. 30% and 37 vs. 43%, respectively) but significantly ($p < 0.05$) lower relapse rates (15 vs. 26%). Frovatriptan was as safe as the other triptans with a frequency of adverse events of 5% (vs. 4% with comparators).

In conclusion, among the variety of effective strategies available, frovatriptan, owing to its sustained effect and good tolerability profile, offers specific attributes suited to acute and prophylactic management of MRM.

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Abstract — WCN 2013

No: 1982

Topic: 8 — Headache

Alice in Wonderland meets the Beauty and the Beast

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Background: Alice-in-Wonderland syndrome (AIWS), named after the novel by Lewis Carroll was firstly described in 1955 as possible symptom in migraine, epilepsy, intoxication or structural brain damage. Symptoms include pseudo-hallucinating self-(somesesthetic aura) or environmental (metamorphopsia) perception. In migraine aura “cortical spreading depression” is commonly accepted as underlying pathomechanism. In AIWS case reports describe a fronto-parietal hypoperfusion in SPECT and occipital and parietal activation in fMRI, respectively.

Case report: A 37 y.o. right-handed female migraneur with (visual) and without aura reports the keys of the keyboard getting bigger and smaller while she was working on her computer. Another time the head of her boyfriend swelled up and horns grew out of his head similar to those of the beast in Walt Disney's *The Beauty and the Beast*. Additionally not further specified objects flew around in the middle of the room. The symptoms lasted about 15 min and within 20 min her typical migraine headache appeared. At this time frequency of migraine attacks was once to twice per month. She used naratriptane for acute treatment but no prophylactic medication. Family history for migraine was positive, cranial MRI was normal.

Conclusion: Unlike the more commonly described micro- or macro-somatognosia, this case demonstrates a rare variant of migraine aura consisting of a metamorphoptic perception of the environment. Prophylactic therapy seems not to be mandatory, but should be considered in the case of more frequently recurring events, especially in view to the patient's profession as a trauma-surgeon to prevent harmful complications for her patients.

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Abstract — WCN 2013

No: 1974

Topic: 8 — Headache

Hyperexcitability in flash visual evoked potentials of migraine patients

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Background: Pattern-reversal visual evoked potentials (PRVEPs) in migraine showed variable results, but Flash visual evoked potentials (FVEPs) in migraine showed usually in the increased amplitude. We have tried to ascertain whether the amplitude of FVEPs increases and the other abnormality of FVEP present.

Methods: FVEPs were recorded in 25 patients with migraine with aura, 23 patients with migraine without aura during interictal period without specific medications associated with migraine and 48 normal subjects. The migraine groups consisted of 11 men and 14 women in migraine with aura patients aged 20 to 71 years (mean age; 49 years), and 12 men and 11 women in migraine without aura patients aged 27 to 70 years (mean age; 57 years). The normal control group consisted of 23 men and 25 women in healthy volunteers aged 28 to 71 years (mean age; 54 years).

Results: The amplitude of FVEPs in migraine showed significant increase ($p < 0.001$) compared to normal subject, and only A1(N1–P1) amplitude in migraine without aura showed significant increase ($P < 0.05$) compared to migraine with aura.

Conclusion: The increased amplitude of FVEPs compared to PRVEPs results in other study suggests that migraine brain is more excitable in flash stimulation associated Y-system than pattern stimulation associated with X-system. These findings are explained by specific cortical distribution of serotonin and noradrenaline in X,Y-system of the visual cortex.

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Abstract – WCN 2013

No: 1064

Topic: 8 – Headache

Frovatriptan versus other triptans in the acute treatment of migraine with aura attacks: Pooled analysis of double-blind, randomized, cross-over, multicenter, studies

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Background: Approximately 20–30% of migraineurs experience attacks with aura. Such symptoms may cause anxiety and distress, making headache attacks disabling. Presently no data are available on the efficacy of treating migraine attacks while experiencing signs and symptoms of aura.

Objective: To evaluate the efficacy of frovatriptan vs. other triptans administered at the onset of a migraine attack with aura (IHS criteria) through a retrospective, pooled analysis of data from randomized, double-blind, cross-over, multicenter studies.

Methods: Subjects were randomized to frovatriptan 2.5 mg or comparator (rizatriptan 10 mg, zolmitriptan 2.5 mg, almotriptan 12.5 mg). Patients were asked to treat 1 to 3 attacks with each drug in no more than 3 months, before switching to the other treatment. 117 attacks treated during occurrence of aura in 56 subjects of the intention-to-treat analysis (mean age \pm SD: 40 \pm 10 years; 91% females) were considered.

Results: Pain free episodes at 2-h did not significantly differ under frovatriptan and comparators [30% vs. 17%; OR: 0.47 (0.19–1.14), $p = 0.091$]. Rate of pain free at 4-h was similarly distributed between treatments [frovatriptan: 51% vs. comparators: 40%; OR: 0.64 (0.31–1.34), $p = 0.237$]. Conversely, relapse at 48-h was significantly less in frovatriptan-treated attacks [67% vs. 90% comparators; OR: 0.22 (0.08–0.61)]. Additionally, at 48-h from drug intake, headache intensity was reduced significantly more by frovatriptan than by comparators (76% vs. 58%).

Conclusions: Frovatriptan seems to have a similar immediate efficacy, but a more sustained effect and a better tolerability than the other triptans in migraine with aura attacks.

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Abstract – WCN 2013

No: 1947

Topic: 8 – Headache

Orthostatic intolerance in syncopal migraine patients

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Background: Orthostatic intolerance (OI) including postural orthostatic tachycardia syndrome (POTS) and orthostatic hypotension (OH) is known to be associated with migraine.

Objective: To evaluate the association between syncopal migraine and OI.

Patients and methods: Study sample consists of 155 patients which underwent tilt table testing using Westminster protocol, divided in four groups:

Gr. I (N = 65) – syncopal migraine,

Gr. II (N = 38) – migraine,

Gr. III (N = 32) – syncope without migraine and

Gr. IV (N = 20) – healthy controls.

POTS was considered as heart rate increment of ≥ 30 beats/min within 10 min and OH – a reduction of systolic blood pressure of 20 mm Hg or diastolic blood pressure of 10 mm Hg within 3 min of standing or head-up tilt. All the data collected were analyzed using SPSS.

Results: Mean age in the study sample was 35.40 \pm 2.3. In the syncopal migraine group POTS was in 40 % and OH in 26.1%, in the migraine group 2.6% POTS and 10.5% OH, in the syncope group 12.5% POTS and 37.5% OH in the controls 10 % OH. The results were statistically significant for POTS in the Gr. I vs. Gr. II ($P < 0.001$), Gr. I vs. Gr. III ($p < 0.05$) and for OH in the Gr. I vs. Gr. II ($p < 0.05$), Gr. III vs. Gr. I ($p < 0.05$).

Conclusion: Strong association between syncopal migraine with POTS (as a form of orthostatic intolerance) suggests the presence of more pronounced dysfunction of central autonomic regulation mechanisms in the cardiovascular system in these patients.

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Abstract – WCN 2013

No: 1877

Topic: 8 – Headache

Programmable lumboperitoneal shunting (pLPS) in the management of pseudotumor cerebri (PTC)

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Background: Medication-resistant PTC or idiopathic intracranial hypertension (IIH) is typically managed with a cerebrospinal fluid (CSF) diversion procedure in order to prevent severe morbidity such as papilledema-associated blindness or to treat intractable headaches. The recent advent of pLPS devices marked a possible significant improvement in the surgical treatment of PTC.

Objectives: We reviewed our personal cases to evaluate the impact of pLPS on our surgical PTC practice, particularly our operative revision needs.

Methods: Our PTC patient collective was retrospectively reviewed (2002–2013) focusing on revision surgery vs. pLPS adjustment for pressure-related changes.

Results: There were sixteen patients (all female) with a mean age of 40 years (21- to 60-years-old). Follow-up was between 9 months and 8 years. Six patients had a STRATA pLPS (Medtronic, Inc.) placed. Four of these patients required from one to three reprogramming sessions, whereas one patient required numerous reprogramming sessions due to a very variable intractable headache syndrome. Patients with a fixed pressure LPS (N = 10) needed a total of eight revision surgeries, of which three were pressure setting related.

Conclusion: pLPS is a very useful technique for PTC management and avoids pressure-related surgical revisions in a substantial number of patients.

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Abstract – WCN 2013**No: 1834****Topic: 8 – Headache****Botulinum a toxin (Botox), in chronic migraine: Our experience in 100 patients**

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Since Botox was approved to treat chronic migraine in USA in October 2010, we treated 100 patients, who have had chronic migraine and the following.

- (A) Severe migraine for at least 2 years.
- (B) Minimum 15 days of migraine headache per month.
- (C) Headache lasting for 4 or more hours and
- (D) failure of response to migraine medications, prophylactic or active, i.e. Triptans.

We followed the standard protocol for Botox in migraine, i.e. 155 units on face, head and neck. Various muscles were injected according to the protocol and consisted of both corrugators, procerus, both frontalis, temporalis, occipitalis, paraspinals and trapezius.

After the injections patients were required to keep a diary in which they would write their response on a daily basis pertaining to a number of headaches per month, the intensity and the duration of headaches and any complications following the injection. Patients were followed in 1 or 2 months and repeat injections were given in 3 months.

Forty patients had between 50 and 60% improvement in duration, intensity and the number of headaches per month. Forty eight patients had 80% improvement. Twelve patients did not respond twice within 6 months and were considered treatment failure. Ten patients had transient ptosis without diplopia lasting for 1 to 2 weeks. Four patients had worsening of headaches during the week after the injection but then showed a good outcome. Ten patients had minor bleeding on the face.

In conclusion Botox is helpful in refractory chronic migraine patients and should be tried.

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Abstract – WCN 2013**No: 1629****Topic: 8 – Headache****Decreased prevalence of migraine after clipping of saccular intracranial aneurysms (SIA)**

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Background: SIA is associated with an increased prevalence of migraine but the effect of surgical treatment remains unclear [1].

Objective: To compare the prevalence of different types of headaches in patients with SIA before and after clipping of SIA and to identify factors associated with migraine remission.

Patients and methods: Out of 199 patients with SIA, 87 patients with SIA (45 men and 42 women, mean age 39.5) had headache before clipping of SIA. They were interviewed before and 1 year after clipping using a semistructured interview. The remission rate of headache in these patients was compared to the rate in 92 clinic patients after 1 year. Diagnoses of headache were made according to the ICHD-2 criteria.

Results: Migraine prevalence decreased from 58.6% to 14.9% 1 year after clipping ($p < 0.001$). It was significantly more ($p < 0.001$) than in controls where the prevalence decreased from 51.1% to 42.4%. The SIA of internal carotid artery ($p = 0.04$, OR 3.76, 95% CI 1.0–14.1), left sided SIA ($p = 0.04$, OR 3.86, 95% CI 1.0–14.9), and the presence of cerebral infarct after SAH ($p = 0.04$, OR 5.19, 95% CI 0.98–27.4) were associated with the presence of migraine after clipping. Prevalence of other headaches (tension type, cluster and posttraumatic) did not change significantly after clipping.

Conclusion: Migraine prevalence in patients with SIA decreases significantly after clipping of SIA. This supports a role of cerebral arteries or their innervation in migraine.

1. ER Lebedeva, NM Gurary, VP Sakovich, Jes Olesen. Migraine before rupture of intracranial aneurysms. *J. Headache and Pain* 2013, 14:15 <http://dx.doi.org/10.1186/1129-2377-14-15>.

doi:10.1016/j.jns.2013.07.1779

Abstract – WCN 2013**No: 1710****Topic: 8 – Headache****Metabolic syndrome and migraine headache: A case control study**

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Introduction: Evaluating the correlation of metabolic syndrome and migraine headache has shown in some previous studies but there is no study that compared the prevalence of metabolic syndrome in the patients with and without migraine. This study was therefore undertaken to compare prevalence of metabolic syndrome in patients with and without migraine headache.

Method: Two hundred patients with migraine, diagnosed according International Headache Society and 200 people without migraine enrolled in this study. Metabolic syndrome was diagnosed according to ATP III criteria in these two groups and compared with each other.

Result: In the present study 34 (17%) of patients with migraine and 30 (15%) of patients without migraine had metabolic syndrome (P value = 0.58). Body mass index (P value = 0.05) and waist circumference (P value = 0.03) were significantly more frequent metabolic syndrome components in patients with migraine headache in comparison with control group.

Conclusion: Our results demonstrate that metabolic syndrome and migraine headache had not significant correlation, however, higher body mass index and waist circumference as metabolic syndrome components had correlated with migraine headache.

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Abstract – WCN 2013**No: 1726****Topic: 8 – Headache****Comparison of headache characteristics in migrainous with and without metabolic syndrome**

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Objectives: Recent studies show that insulin resistance is more prevalent in patients with migraine. Insulin resistance has a basic role in pathogenesis of metabolic syndrome which is directly

correlated with obesity and other cardiovascular risk factors. We aimed to compare the severity, duration and frequency of migraine headaches in migrainous with and without metabolic syndrome.

Methods: Two hundred migrainous were selected for this study. These patients divided to two groups, with and without metabolic syndrome. Severity, duration and frequency of migraine headaches were compared in two groups in the first visit, 30 and 60 days later.

Results: 34 patients (17%) had metabolic syndrome. They were older (p value < 0.001) and mostly female (p value < 0.01). Duration of migraine headaches was also more in metabolic syndrome group (p value = 0.03). The mean frequency of headache (9.1 versus 10.1, p value = 0.54) and mean severity of headache (7.6 versus 7.3, p value = 0.31) did not have impressive difference in two groups. 30 and 60 days after starting the treatment, severity, frequency and duration of headaches were decreased significantly in both groups, but there were no significant difference in the amount of these reductions in two groups.

Conclusion: Our study showed that metabolic syndrome is present in 17% of patients with migraine. Metabolic syndrome was correlated with the patient's age and duration of their headache attacks, but had no significant relationship with the frequency and severity of headache.

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Abstract – WCN 2013

No: 1762

Topic: 8 – Headache

Do vestibular migraine patients differ from migraine patients without history of vertigo in anxiety?

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Background: Studies showed that anxiety and mood disorder were more common in migraine patients than healthy controls.

Objective: The aim was to compare vestibular migraine patients, migraine patients without history of vertigo (migraine only) and healthy controls according to their anxiety and depression scores.

Patients and methods: 35 definite vestibular migraine patients according to Neuhauser criteria, 30 migraine only patients and 30 healthy controls were included in the study. Hamilton Anxiety Rating Scale (HAMA), The State-Trait Anxiety Inventory (STAI-X1 and STAI-X2), Beck Depression Inventory (BDI), Anxiety Sensitivity Inventory-3 (ASI-3), Panic-Agoraphobic Scale (PAS; assessing panic like symptoms, stress sensitivity, anxious expectation, illness phobia and hypochondriasis, and reassurance seeking) and Penn State Worry Questionnaire (PENN) were used for assessment. ANOVA test was used in between group comparisons and BONFERRONI test was used for post-hoc analysis.

Results: We found that there were significant differences in HAMA ($p = 0.050$), PENN ($p = 0$), 43STAI-X2 ($p = 0.003$) and panic like symptoms ($p = 0.001$), agoraphobia ($p = 0.012$) subscales of PAS scores also general score (0,005) between vestibular migraine and control group. There was also significant difference in HAMA ($p = 0,016$) between migraine only patients and control group. Migraine only and vestibular migraine patients differed in reassurance orientation ($p = 0.04$) and agoraphobia ($p = 0,030$) in subscales of PAS.

Conclusion: Our results showed that migraine only was related to higher levels of anxiety whereas vestibular migraine was related to

both to anxiety and depression symptoms. Psychiatric symptoms may contribute to severity and disability related to migraine only.

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Abstract – WCN 2013

No: 1756

Topic: 8 – Headache

Idiopathic intracranial hypertension; clinical features and prognosis

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Background: Idiopathic intracranial hypertension (IIH) is a disorder that predominantly affects obese women of childbearing age. The most important finding of elevated intracranial pressure (ICP) is papilledema which may lead to visual loss.

Objective: We aim to determine the demographic features, clinical signs and symptoms, radiological findings and investigate if any of initial features and findings are associated with the prognosis.

Patient and methods: In this study we included 59 IIH patients followed up in our neuroophthalmology department. We assessed demographic features, clinical signs and symptoms, radiological findings, and ICP. We also investigate the prognosis.

Results: 52 patients were female (88.1%), 7 were male (11.9%), mean of ages were 30.25 ± 13.12 . The duration of the symptoms was 23.7 ± 32.4 months when they accepted to the department. Reported complaints were headache (78%), visual disturbances (48%), nausea (30.5%), vertigo (18.4%), tinnitus (8.5%), and diplopia (13.2%). Mean visual acuity was 0.78 ± 0.29 . Visual field deficits 69% and papilloedema occurred in 71% of the patients most of which were bilateral (66.1%). Mean ICP was 308.7 ± 100.2 mm H₂O. 19.7% of the patients were obese, 17.2% of patients have endocrinopathy. Empty sella (15.3%), sinus thrombosis (8.5%) and optic hydrops (6.8%) were the most seen pathological MRI findings. The prognosis was good in 64.7% of the patients and 35.3% of patients worsened. Demographic features, initial complaints, mean ICP, pathological MRI findings and neuro-ophthalmological findings except visual field defects were not associated with the prognosis of participants. The prolonged duration of the time passed before patients gets treated and initial visual field defects were associated with the poor prognosis.

Conclusion: The prolonged duration of the time passed before patients get treated and initial visual field defects can be associated with the poor prognosis. Therefore, IIH must be diagnosed immediately, treated appropriately and follow up regularly.

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Abstract – WCN 2013

No: 1359

Topic: 8 – Headache

Post-dural puncture headache after spinal anesthesia and perioperative blood pressure

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Background: The pathophysiology of post-dural puncture headache (PDPH) is believed to be related to low cerebrospinal fluid (CSF) pressure caused by CSF leak through the puncture site of dura. Considering that the blood pressure (BP) is one of major determinants of CSF pressure, the role of BP on the occurrence of PDPH should be elucidated.

Objective: To ascertain the effect of perioperative BP on the development of PDPH in the patients who received surgery under spinal anesthesia.

Patients and methods: We evaluated the presence of PDPH in all consecutive 199 patients (122 males, 77 females, age: 15–76 years) who received elective knee surgery under spinal anesthesia between September 2012 and February 2013. The spinal anesthesia was performed by the same anesthesiologist with 25-G Quincke needle. Data regarding previous history of headache, pre- and post-operative BP, highest and lowest BP during operation as well as demographic features were analysed.

Results: The overall incidence of PDPH was 9.0%. It was higher in female than in male (15.6% vs 4.9%, $p = 0.02$). Age, history of hypertension or recurrent headache was not different between patients with and without PDPH. The duration of operation or spinal anesthesia was not different between the two groups. BP variables were expressed as pre- and post-operative mean arterial pressure (MAP), the highest and the lowest MAP during operation, and their differences were not different either.

Conclusion: PDPH after knee surgery under spinal anesthesia occurred more frequently in female patients, and was not influenced by their perioperative BP states.

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Abstract – WCN 2013

No: 1558

Topic: 8 – Headache

Memory improvement after spreading depression by NMDA blocker as memory destructor

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Spreading depression (SD) is transient neural hyperexcitability followed by depolarization wave, which propagates through the brain and modulate electrical gradient and synaptic activity. Data have shown that SD wave distributes coincidence between neural activity and behavioral activity. Neural activity and electrical potential effect on memory retrieval have been demonstrated. Inhibitory effect of NMDA receptors in SD procedure can control memory impairment caused by SD. However, the negative effect of NMDA receptor blockage on memory has been proven in previous studies. In the present study the effect of NMDA receptors blockage (MK801) used to evaluate its efficiency in subsiding of SD negative influence on memory. Wistar rats (60–80 g) were randomly chosen in 6 groups and (NMDA blocker 0.63–1 mg/kg) were administrated after 3 mol/L KCl injection for induction of repetitive SD in rat. The groups were evaluated by T-maze test and SD groups were compared with control groups, including (NMDA blocker 1–0.63 mg/kg controls) and sham group. T-maze data have showed that repeated SD could significantly alter memory retrieval performance. However, in the second week memory enhancement was induced by SD induction. Repeated SD induction during other weeks indicated impairment in memory. Application of NMDA blocker showed significantly enhanced memory retrieval and could potentially control memory impairment after SD. The studies indicated that NMDA blocker may decrease memory performance, on the other hand the effect of MK801 on inhibition of SD propagation may somehow weaken memory improvement due to its memory destruction effects.

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Abstract – WCN 2013

No: 1647

Topic: 8 – Headache

Prevalence of migraine among medical students in Kuwait University

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Background: Prevalence of migraine among medical students is of particular interest as they are subjected to lots of tests and stresses that may precipitate migraine attacks.

Objectives: To determine the prevalence of migraine among medical students in Kuwait University.

Methods: This cross-sectional and descriptive study, which included students registered to Medical Faculty at Kuwait University in the academic year of 2012–2013. Out of 808 registrants, 621 students accepted to participate in the study. Participants who had two or more headaches in the last 3 months were subjected to two preliminary questions and participants with at least one positive response were asked to perform the validated ID-Migraine™ test. The frequency of headache per month and severity of headache by Numeric Rating Scale (NRS) were reported.

Results: Migraine was detected in 173 subjects (27.9%) based on the ID-Migraine™ test. The mean age of the migraine students was 20.17 ± 2.29 (16–25 years). Thirty-seven were male (21.4%) and 136 were female (78.6%). Migraine was significantly more frequent in the last 2 grades (35.5% and 44%, $p < 0.000$). The frequency and the severity of headache were significantly increased during the last 2 grades (5.55 ± 1.34 and 7.23 ± 1.27 , $p < 0.000$) (6.00 ± 0.76 and 6.68 ± 1.25 , $p < 0.000$) respectively. Stress 43 (24.9%), irregular sleep 36 (20.8%), and much reading 32 (18.5%) were the most common triggering factors.

Conclusion: There is a high prevalence of migraine among medical students in Kuwait University. The frequency and severity of headache increase with years of educations.

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Abstract – WCN 2013

No: 1617

Topic: 8 – Headache

A randomized doubled blinded trial of treatment with diamino-oxidase (DAO) in patients with migraine and deficit of enzyme's activity

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Background: Histamine has been considered as a chemical mediator of migraine. The degradation is done in two different pathways. One of the enzymes that allow this process is the diamino-oxidase (DAO).

Objective: The aim of this study is to identify the prevalence of the deficit in the activity of DAO in patients with migraine, and test the supplementation of this enzyme in a randomized controlled double-blind trial.

Material and methods: This was a randomized parallel-group controlled study. After a 1-month run-in, patients with migraine attacks/month between 4 and 14 were randomized 1:1 to placebo or DAO three times a day during one month. Primary outcome measures were diminution of hours of pain, and the use of antimigraine drugs.

Results: We studied 137 patients with migraine, and find the deficit of DAO activity (< 80 HDU/ml) in 119 (87%).

One hundred patients were randomized and included in the intention-to-treat analysis. Between run-in and first month of treatment, the mean number of hours of pain decreases in both groups but with significant difference in the final control in the group treated with DAO compared with placebo (6,3 vs 5,1: $p < 0.03$).

The use of the acute antimigraine drug was significantly reduced in the DAO but not in placebo group ($p > 0.022$).

There were no adverse events in either group.

Conclusions: Deficit in the activity of DAO is very prevalent in population with migraine.

The supplementation with the enzyme is effective and safe as a preventive therapy for migraine.

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Abstract – WCN 2013

No: 1621

Topic: 8 – Headache

Subdural haematoma as a late complication of spontaneous cerebrospinal fluid hypovolemia (SCH) syndrome:

Two case reports

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Background: SCH syndrome is relatively common, and a CSF leakage can be occasionally demonstrated. Subdural haematoma (SDH) has been reported in patients with SCH, mainly in older men, or these displaying longer time to diagnosis of SCH.

Objectives: We report two cases that developed SDH after apparent resolution of SCH.

Material and methods: A 41 year old man developed severe orthostatic headache and neck stiffness after repeated sneezing. Lumbar puncture showed an opening pressure of 0 mm H₂O. Cisternography demonstrated cervicothoracic CSF leak. A 43 year old man presented sudden headache after sport activity, highly suggestive of SCH, developing unilateral abducens palsy after several days. Cranial tomography (CT) was normal in both cases, with resolution of symptoms after 3 months of conservative therapy.

Results: Three months after the onset, when patients were almost asymptomatic, a control MRI showed subacute bilateral SDH. In both, resolution of the haematomas was verified after some weeks without need of drainage.

Conclusion: SCH is characterized by orthostatic headache, low CSF pressure, and sometimes typical MRI image. The development of SDH, although rare, has been reported. In our cases the late development of SDH is remarkable, perhaps related to the persistence of SCH for a long time. We emphasize the importance of monitoring patients with SCH, and consider conservative measures only in cases with a brief course. Epidural patching or surgical repair may prevent potentially serious complications such as SDH. Moreover, SCH should be excluded as a cause of SDH in young patients without risk factors.

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Abstract – WCN 2013

No: 1298

Topic: 8 – Headache

Analysis of MAP0004 subjects with menstrually related migraine vs. non-menstrually related migraine

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Background: Menstrually related migraine (MRM), as defined by The International Classification of Headache Disorders, 2nd edition, occurs from days –2 to +3 of menstruation in ³2 of 3 menstrual cycles and at other times of the cycle. MRM generally lasts longer and is more severe and difficult to treat than non-MRM. MAP0004 is an investigational product that delivers dihydroergotamine through the lungs via a breath-synchronized metered-dose inhaler.

Objective: This post hoc analysis of phase 3 data evaluated the efficacy and tolerability of MAP0004 in MRM vs non-MRM.

Patients and methods: This analysis included 149 women from a modified intent-to-treat population who treated MRM (n = 45) and non-MRM (n = 104) with MAP0004. The study used the following clinical end points: pain relief and pain free at 2 h and sustained pain relief and sustained pain free at 2–24 h and 2–48 h.

Results: The efficacy of MAP0004 did not differ significantly in MRM vs non-MRM at 2 h (pain relief 62% vs 64%; pain free 36% vs 29%), at 2–24 h (pain relief 53% vs 49%; pain free 31% vs 23%), and at 2–48 h (pain relief 38% vs 38%; pain free 24% vs 15%). No significant differences were found in frequency of adverse events, and no drug-related serious adverse events were reported.

Conclusion: In this post hoc analysis of phase 3 data, MAP0004 was similarly effective and well tolerated in treating both MRM and non-MRM.

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Abstract – WCN 2013

No: 1295

Topic: 8 – Headache

Analysis of the development of allodynia: Correlation between migraine duration and severity

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Background: Allodynia, the perception of pain from non-nociceptive stimuli, is a clinical presentation of central sensitization. Allodynia is reportedly common during migraine attacks. Although factors leading to development of allodynia are not well understood, duration and severity of migraine have been implicated.

Objective: This retrospective analysis evaluated the relationship between allodynia and the duration and severity of migraine to better understand the mechanisms related to migraine-induced central sensitization.

Patients and methods: This analysis included 792 patients from the double-blind period of a phase 3, placebo-controlled, randomized clinical trial of an investigational acute treatment for migraine (MAP0004). Baseline pain levels were recorded by patients using an electronic diary, and baseline allodynia data were obtained using a standard questionnaire. Correlations between percentage of patients reporting allodynia, severity of migraine, and duration of migraine were analyzed by Fisher's exact test or Chi-square test, as indicated.

Results: At baseline, 53% of patients reported allodynia. The presence of allodynia did not change in relation to the duration of the migraine (Chi-square $P = 0.2182$), regardless of migraine severity (moderate pain, Chi-square $P = 0.1807$; severe pain, Chi-square $P = 0.5830$). Patients reporting severe pain experienced significantly more allodynia (58.4%) than patients with moderate pain (48.2%; Fisher's exact test $P = 0.0053$).

Conclusion: The presence and development of allodynia is associated with severity but not duration of migraine. This retrospective analysis suggests that migraine severity is a significant factor in central sensitization.

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Abstract — WCN 2013

No: 1411

Topic: 8 — Headache

Electrophysiological evaluation of headaches associated with temporomandibular dysfunction

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Objective: To study clinical and neurophysiological peculiarities of headaches associated with temporomandibular dysfunction (TMD) and to evaluate involvement of trigemino-cervical system in pathogenesis of TMD headaches.

Methods: 30 patients aged 17–59 years old with headache complaints and TMD, confirmed by clinical and instrumental methods, were included into study. 30 healthy subjects aged 18–54 years old presented control group. Neurophysiological methods included trigeminal evoked potentials (TEP), blink reflex (BR) and EMG of masticatory muscles. Fisher exact test and correlation analysis with Pearson criterion were used for statistical analysis.

Results: The present study showed the trend to TEP latencies decrease, statistically significant latencies decrease of BR components R1 ($p < 0.05$) and R2 ipsi- and contralaterally ($p < 0.001$) compared with healthy controls. The analysis of the results of stimulative EMG revealed M-response asymmetry with amplitude decrease on the side of pain ($p < 0.05$).

Correlation analysis showed strong direct correlation between latencies of TEP and M-response of masticatory muscles ($r = 0.83$) and inverse correlation between latencies of R2 component of BR ipsilaterally and M-response amplitudes ($r = -0.91$).

Conclusion: The obtained results demonstrated increased reflex irritability of trigeminal system and brainstem structures in patients with headaches associated with TMD. Functional state of trigemino-cervical system determines parameters of M-response of masticatory muscles. Increased irritability of trigeminal system is accompanied by increased muscle tone of masticatory muscles and leads to enhancing of pain syndrome. According to the obtained results, complex therapy of TMD headaches is recommended, including anticonvulsants in case of trigeminal system involvement.

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Abstract — WCN 2013

No: 653

Topic: 8 — Headache

The role of plasm nitrites in diagnosing of migraine severity in children

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Objective: Studying of plasma concentration of steady nitrogen oxide metabolite under migraine in children with regard to the type of migraine (with and w/o aura), frequency, rate and duration of a seizure.

Methods: The Study group is comprised of 81 children with migraine (54 boys, 27 girls) at the age from 6 to 18 y.o. Control group is 20 healthy children. Defining of nitrites in blood plasm was being

conducted in accordance with methodology by Karpyuk with co-author (2000). The accuracy of differences: t-criterion of Student at $p < 0.05$.

Results: There's been diagnosed proven increase of plasm nitrite level up to 3.2 ± 0.8 nM/l in the Study group against 2.2 ± 0.09 nM/l in the Control group. The group of patients with frequent seizures (≥ 2 times per month) is comprised of 61 children with seizure frequency at the rate of 3.3 ± 1.5 per month and pain rate per visual analogue scale is 6.5 ± 2.0 points. The group of children with occasional seizures (< 2 times per month) – 20 children with seizure frequency at the rate of 0.25 ± 0.2 per month and pain rate at 7.4 ± 2.0 . Frequent seizure group appeared to have increase of nitrite plasm level up to 3.4 ± 0.9 nM/l against the group with occasional seizures (2.7 ± 0.9 nM/l). The study didn't show any dependence of migraine type, pain rate and seizure duration on the level of nitrogen oxide.

Conclusion: The level of plasm nitrites can serve as a laboratory indicator of disease severity and depends on the frequency of migraine seizure.

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Abstract — WCN 2013

No: 1110

Topic: 8 — Headache

The primary headache subtypes in Nazareth city; co morbidity potential risk factors and disability rate

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Introduction: To investigate the primary headache subtype prevalence and disability rate associated with mood disorders in low socio economic status (LSES) may underlie both diseases.

Methods: I identified 276 headache sufferers, drawn from more than 1254 individuals representative of heterogeneous community of Nazareth city, which were referred to our center by family physicians for various neurological disorders, were subdivided subsequently in tension type headache (TTH) and migraine disorders according to IHS criteria and undergone complete work up profile with therapeutic strategies, secondary headaches were excluded. The socioeconomic status (SES) for every individual was considered and meticulously examined according employment, income and educational gradients. Verbal informed consent was obtained during the first round.

Results: The vast majority of those individuals were belonging to LSES (57.3%) and subdivided according IHS and DSM-IV (for psychiatric illnesses, mood disorders, depressive and anxiety disorders) in 172 (62.3%) sufferers of TTH with 69.3% depressive disorders background and other psychiatric illnesses, 102 (36.96%) migraine (included with/without aura) with 30.7% mood disorders, and finally, 2 (0.72%) sufferers of cluster headache.

Conclusions: The main target of this observational study was to verify the relationship between primary headache disorders and potential co morbidity risk factors. Psychiatric co morbidity was assessed by DSM-IV criteria. The LSES was considered as remarkable co morbidity risk factor for both headache subtypes, migraine and tension type headache.

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Abstract — WCN 2013

No: 894

Topic: 8 — Headache

CaMEO (Chronic Migraine Epidemiology & Outcomes) Study: Design, Methodology, and Baseline Cohort Characteristics

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Background: CaMEO, a prospective cohort study, collects longitudinal and cross-sectional data in migraineurs to characterize migraine clinical course, family burden, barriers to care, endophenotypes, and comorbidities.

Objective: To describe the methodology and characterize the population in CaMEO.

Methods: Migraineurs were recruited from a 2.4 million member web-based panel and demographically matched with the general population. Using ICHD-2 criteria, participants were divided into episodic (EM) or chronic migraine (CM) based on headache days/month. Participants complete surveys every 3 months for 1 year (starting fall 2012) and also recruit spouses/children in the household to assess familial migraine-related burden. Baseline data includes headache features/frequency, comorbid depression/anxiety, medication use, healthcare consultation, disability, and quality of life.

Results: Of 489,537 invitees, 80,783 (16.5%) responded to the screening survey. Eligible respondents (16,789; 3.4%) were invited into the 12-month assessment and were ≥ 18 years old, spent an appropriate amount of time completing the survey (≥ 10 min), and had EM ($n = 15,313$; 91.2%) or CM ($n = 1,476$; 8.8%). Compared with nonrespondents, respondents were older ($P < 0.0001$), more likely to be women ($P < 0.0001$), white ($P < 0.0001$), or married ($P < 0.0001$), and less likely to be employed full/part time ($P < 0.0001$) or have incomes $> \$50,000$ ($P < 0.0001$).

Conclusion: CaMEO characterizes the course of EM and CM over 1 year in a community-based web panel of migraineurs. With repeated assessments, CaMEO data will quantify variations in headache frequency, disability, comorbidities, medication use, and familial impact, and can also be used to define migraine endophenotypes for future genetic studies.

Disclosure: Funded by Allergan, Inc.

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Abstract – WCN 2013

No: 898

Topic: 8 – Headache

Sociodemographic, disability, and employment differences between persons with chronic and episodic migraine: CaMEO (chronic migraine epidemiology & outcomes) study results

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Background: Previous research suggests sociodemographic, headache-related disability, and employment status differences between individuals with chronic migraine (CM) and episodic migraine (EM).

Objective: To characterize sociodemographics, headache-related disability, and employment status among individuals with CM and EM in a large population-based sample.

Methods: CaMEO prospectively recruited migraineurs from a 2.4 million member web-based panel, demographically matched with the general population. 16,789 of 80,783 (20.8%) respondents met ICHD-2-based definitions for CM (migraine diagnosis, ≥ 15 headache days/month) or EM and were eligible for inclusion. We used descriptive and inferential statistics to contrast those with CM and EM based on baseline survey responses.

Results: Of the 16,789 eligible respondents, 1476 (8.8%) had CM and 15,313 (91.2%) had EM; these groups had comparable mean ages (40.6 y vs 41.0 y; $P = 0.32$). Compared with the EM group, the CM group had a greater proportion of women (81.1% vs 73.8%; $P < 0.001$)

and whites (87.5% vs 83.3%, $P < 0.001$), experienced higher headache-related disability (mean MIDAS, 60.5 vs 13.1; $RR = 4.6$, $P < 0.001$), completed fewer years of education (34.9% vs 45.9% had bachelor's degrees or higher; $P < 0.001$), were more likely to have individual and household incomes below the median (69.1% vs 59.1%; $P < 0.001$, and 59.8% vs 49.5%; $P < 0.001$, respectively), and less likely to be employed full- or part-time (56.4% vs 66.0%; $P < 0.001$).

Conclusion: CM confers a greater financial and occupational burden than EM. Ongoing data collection will allow us to characterize the longitudinal course and consequences of CM and EM.

Disclosure: Funded by Allergan, Inc.

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Abstract – WCN 2013

No: 1138

Topic: 8 – Headache

The temporal relationship between daily stress and delayed modulation of chronic headache pain

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Background: Although triggers are known for headache pain, many chronic headache patients do not know what causes episodic pain fluctuations. Previous fibromyalgia and complex regional pain syndrome studies report ten-day delays in pain flare onset following stressful events. Delayed neuropathic pain flares are associated with stress-released thyroxine modulation, which has latent effects due to attachments with serum thyroxine-binding globulins.

Objective: The temporal relationship between reported stress levels and episodic headache pain intensity was investigated to determine if chronic headache shows latent stress-related pain modulation.

Patients and methods: Two female subjects diagnosed with chronic migraine completed pain and stress inventories daily across ten weeks. Pain was assessed using the Visual Analog Pain Scale, McGill Pain Questionnaire Short Form, and modified pain-body diagram. The Visual Analog Stress Scale quantified daily stress.

Results: Serial lag correlations revealed the highest correlation between stress and pain to occur after a ten-day lag in subject 1 ($r = +0.63$, $p < 0.005$). Subject 2 showed a high negative correlation between stress and pain ten and eleven days later ($r = -0.37$ and $r = -0.46$, respectively, $p < 0.05$).

Conclusions: Patients manifested delayed pain changes following stress in diametrically opposed ways, where headache pain ten days later seemed to increase as a function of stress in one yet decreased in the other. This finding is consistent with existing literature on pain modulating effects from stress-related release of thyroxine, whereby some individuals increase thyroxine output as a result of psychogenic stress and others notably decrease thyroxine output.

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Abstract – WCN 2013

No: 1226

Topic: 8 – Headache

Hemicrania epileptica IHS code 7.61

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Chronic headache of over a 3 month duration is a common disorder for which advice from a neurologist is invariably sought. Over 2500 cases of chronic headache seen between March 1998 and 2013 by

men have significant number of cases (600) which had an epileptic background and no seizures.

Headache improved in all cases using antiepileptic drugs (AED) only. Three subgroups were observed from these patients:

- Calcified granuloma – 210 patients 35%.
- Febrile seizures/child hood seizure – 120 patients 20%.
- Family h/o seizures with EEG showing intencinal finding – 270 patients 45%.

Based on this analysis, it is desirable to keep in mind the epileptic background of some of the chronic headache cases and treat appropriately (with AED).

All cases (600) studied did not have seizures – headache is the only symptom of presentation for consultation, in these cases headache is the manifestation of seizures without loss of consciousness and only responding to AED – CT scan of brain, EEG and history are great importance to clinch the diagnosis and good response to AED. Hemispheric epileptics was coined as a distinct clinical entity in mid nineteenth century but in 1984 Isler all documented topographic epileptic form activity in unilateral headache for a 10 year duration in young female patients.

IHS classification does show it as one of the minor code (7.61) secondary headaches. It is the next most common form of headache to migraine in over all incidence of headache in the general population.

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Abstract – WCN 2013

No: 1250

Topic: 8 – Headache

Assessment of neurovascular reactivity at posterior cerebral arteries to visual stimulation in migraine headaches

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Background: The intracranial vessels have an important role in migraine pathophysiology, although neuronal mechanisms have been disclosed as primary point of interest.

Objective: We aimed the evaluation of neurovascular reactivity using transcranial Doppler ultrasonography in patients with migraine with aura and without aura.

Patients and methods: Sixty nine patients with migraine without aura, 58 patients with migraine with aura, and 75 healthy subjects served as controls were enrolled in the study. All patients were studied during headache attack and a headache-free period with transcranial Doppler. Reactivity is defined as relative changes between blood flow velocities measured during stimulus and rest.

Results: Increased reactivity was found at the headache side in all migraine patients during pain-free period comparing to control group (53% and 46% respectively, $p = 0.01$). In addition, reactivity was found significantly higher in migraine with aura (60%) than those of both migraine without aura and control groups ($p < 0.001$) in the headache-free period. Although, all migraine patients showed lower reactivity (headache side %31, non-headache side %32) than that of control group ($p < 0.001$ vs $p = 0.002$, respectively) during headache attack.

Conclusion: Patients with migraine with aura have higher reactivity to visual stimulation than that of control group in attack-free period. This is compatible with increased cortical excitability in patients with migraine with aura. However, reactivity was significantly lower in both migraine groups than control group during headache period. Inflammation and dilatation of dural and meningeal arteries during

headache phase are not able to dilate more to visual stimulation resulting lower degree reactivity.

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Abstract – WCN 2013

No: 1294

Topic: 8 – Headache

Optic coherence tomography for optic disc and clinical evaluation in idiopathic intracranial hypertension

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Idiopathic intracranial hypertension (IIH) is the clinical syndrome of raised intracranial pressure (ICP), in the absence of space-occupying lesions or vascular lesions, without enlargement of the cerebral ventricles. It occurs commonly in overweight women and is encountered by most neurology departments on a regular basis.

Optic coherence tomography (OCT) is a non-invasive imaging technique producing high-resolution cross-sectional images of the retina. Studies indicate that OCT is becoming valuable in various conditions with increased retinal nerve fiber layer (RNLF) but there are very few studies evaluating ability of OCT to identify and quantify papilledema in IIH. Therefore, the aim was to evaluate the applicability of peripapillary OCT evaluation in 57 patients with confirmed IIH according to Modified Dandy Criteria.

Subjects were evaluated by means of direct ophthalmoscopy, peripapillary retinal nerve fiber layer thickness (RNFLT) measurements (OCT 3 Version 4 Carl Zeiss Ophthalmic System Dublin, USA).

Measurements of RNFLT were performed at admission and during the follow up periods. At the time of diagnosis, the mean RNFLT of the right eye was 111 μm whereas the value for left eye was 113.5 μm . After the treatment and during the following examinations the RNFLT reduced to 96 μm for the right eye and 98 for the left eye ($p < 0.001$).

This presents promising findings for the use of OCT for evaluating papilledema in IIH patients during diagnosis and for the further clinical follow-up and management.

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Abstract – WCN 2013

No: 974

Topic: 8 – Headache

Do migraineurs with vertigo/dizziness display some common characteristics? Results from a population-based study

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Background: Migraine and vertigo/dizziness are both common, distressing and interrelated. They occur together 3 times more than by chance, and both impact on quality of life.

Objective: Our aim was to assess the frequency of vertigo/dizziness and to define the differences between clinical characteristics related to vertigo/dizziness in a community-based study.

Patients and methods: We designed a community-based prevalence study in adults, with face-to face interviews by specially trained general practitioners using a structured electronic questionnaire.

Comprehensive interview form included diagnostic questions based on the ICHD-II criteria. The questionnaire assessed all diagnostic headache features, headache related impact, demographics, comorbidities and disability assessed by MIDAS questionnaire. Descriptive statistics were applied and Chi-square test, t test and logistic regression test were used for comparisons.

Results: 5323 participants were reviewed. Vertigo/dizziness was significantly higher in migraineurs when compared to tension type headache sufferers. There were 534 migraineurs with vertigo/dizziness (MwVD) and 337 patients without vertigo/dizziness as controls. MwVD patients established 61.3% of definite migraineurs. MwVD patients had significantly more nausea/vomiting, more headache aggravation with head motions and had more visual aura. Frequency of headaches and pain killers used per month was also significantly high in MwVD patients. MwVD patients reported significantly more motion sickness, allergy, allodynia and osmophobia, interestingly. Furthermore MwVD patients had significantly low quality of life.

Conclusions: Our findings showed that more than half of the migraineurs have vertigo/dizziness. These patients showed many significant differences in clinical characteristics and have low quality of life when compared to other migraineurs.

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Abstract – WCN 2013

No: 844

Topic: 8 – Headache

Age of migraine onset in lateral cerebral ventricles asymmetry patients

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Background: Recent neuroimaging studies reveal that 12–60% of chronic migraine (CM) patients evince a “benign” lateral ventricles asymmetry (LVA). Migraine preventive treatment in these cases seems to be less efficient in decreasing migraine attack frequency as a hint of a worse migraine evolution in such patients.

Material and methods: We studied 124 same age (mean 36.56 ± 1.09 y.o.) CM patients diagnosed according to ICHD criteria (2nd edition, revised 2006) that performed an MRI 3 T examination. The subjects were divided into four groups according to ventricular frontal horns asymmetry index (AI):

Group I (control) – CM + minimal LVA (AI = 1.0–1.24),

Group II – CM + mild LVA (AI = 1.25–1.99),

Group III – CM + moderate LVA (AI = 2.0–2.99) and

Group IV – CM + severe LVA (AI ≥ 3.0).

One of selection criterion for the study was the similar disease duration: 18.11 ± 1.61 y.o. in group I, 18.91 ± 2.39 in group II, ($p > 0.05$), 18.65 ± 2.09 in group III, ($p > 0.05$) and 19.60 ± 1.92 in group IV ($p > 0.05$). All patients underwent a clinical assessment and data were statistically analyzed.

Results: We ascertained a younger disease onset in CM patients associated with significant LVA. So, in moderate LVA CM patients, migraine started at the age of 13.65 ± 1.60 ($p < 0.01$) y.o., in severe LVA CM patients – at 12.35 ± 1.20 y.o. ($p < 0.001$), in comparison to the control group (20.42 ± 1.09 y.o.).

Conclusions: According to our results, significant lateral ventricles asymmetry could be considered as an aggravating factor for the chronic migraine, maybe based on the common underlying serotonin deficiency that possibly fasten migraine onset.

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Abstract – WCN 2013

No: 896

Topic: 8 – Headache

Prospective recordings of the clinical presentation of migraine with typical aura

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Background: About a third of migraine patients have attacks with aura, usually one or more neurological symptoms arising from the cortex or brainstem. The clinical presentation of aura can vary considerably and many descriptions of the clinical characteristics are based on purely retrospective studies.

Objectives: To describe the clinical presentation of migraine with typical aura in a large group of patients enrolled in a clinical trial, and to compare self-reported migraine symptoms to prospective recordings of attacks in the same patients.

Methods: 267 patients with migraine with typical aura were enrolled and provided data on the clinical presentation of migraine aura, headache, and associated symptoms. Baseline characteristics provided by patients on inclusion were compared with migraine symptoms collected prospectively during the study.

Results: Visual aura symptoms were reported by all patients, the most prevalent symptoms being dots or flashing lights (70.4%), wavy or jagged lines (46.8%) and blind spots (42.3%). Non-visual aura was less prevalent, reported by around half the patients. Symptom intensity for migraine symptoms (pain, nausea, photo- and phonophobia) was mainly ($\geq 70\%$) graded moderate to severe. Patient reported data prior to the trial indicated a consistently higher symptom score (more severe phenotype) than in the prospectively collected data, ($P < 0.01$ for all).

Conclusion: Based on this large clinical collection of data exclusively from MA attacks, we show that visual aura symptoms are more prevalent than non-visual aura symptoms. The severity of self-reported migraine symptoms was great than that recorded in the prospective data set.

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Abstract – WCN 2013

No: 897

Topic: 8 – Headache

Distinctive anatomical and physiological features of migraine aura revealed by 20 years of patient recording

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Background: Descriptions of the migraine aura have been presented as prospective self-observations by physicians and other professionals who had migraine with aura or migraine aura without headache.

Objectives: To present detailed characterization of a large number of visual auras recorded in one patient over two decades.

Methods: A patient documented and made detailed drawings of 1000 aura attacks over 20 years. Drawings were made in real time with documentation of the aura wavefront at 1 min time intervals.

Results: Consistent patterns of aura initiation, propagation, and termination were observed in both right and left visual fields. Most aura attacks originated centrally (within 10° eccentricity), but there were also other distinct sites of initiation in the visual field. Auras preferentially propagated first through lower nasal field (69–77% of all auras) before travelling to upper and temporal fields, on both sides. Some auras propagated from peripheral to central regions of the visual field – these typically followed the same path as those

travelling in the opposite direction. The mean velocity of the perceived visual phenomenon did not differ between attacks starting peripherally (17.6 mm/min \pm 1.06) and centrally (16.2 mm/min \pm 0.59), ($P = 0.41$). Based on duration and imaging we estimated the speed of the underlying cortical event to be 2.1–3.1 mm/min.

Conclusion: These results indicate that there can be multiple distinct sites of aura initiation in a given individual, and that there are consistent patterns of aura propagation that indicate non-concentric patterns of spread.

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Abstract – WCN 2013

No: 933

Topic: 8 – Headache

Characteristics of migraines during the postpartum period: A long-term prospective study

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Background: Although there have been many reports regarding the course of migraines during pregnancy, the prevalence and characteristics of migraines during the postpartum period remain unclear.

Objective: To investigate the characteristics of migraines over a long postpartum period.

Patients and methods: We prospectively investigated the course of migraines experienced during the postpartum period by patients in a postnatal ward. The patients were surveyed during the first postpartum week and 1, 3, 6 and 12 months after delivery. The patients were provided a headache diary to assess medication use and migraine attack frequency, severity (the faces pain scale) and duration.

Results: The migraine remission rate was 63%, 83% and 85% during the first, second and third trimesters, respectively. No patient experienced a worsening of headaches during pregnancy. Headache recurrence during the first month after delivery was more frequent in the patients >30 years of age than in those \leq 30 years of age ($p < 0.05$). The percentage of women experiencing recurrence at 1, 3, 6 and 12 months after delivery was 63%, 75%, 78% ($n = 60$) and 87.5% ($n = 40$), respectively. In breastfeeding patients, the rates were 50%, 65.8%, 71.1% and 91.7% and in bottle feeding patients, the rates were 86.4%, 90.9%, 95.5% and 81.3%, respectively.

Conclusion: In our study, we found that 85% of the patients with migraines experience remission during pregnancy and that more than 50% experience recurrence during the first month after delivery. Breastfeeding is associated with a lower recurrence rate than bottle feeding until six months after delivery.

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Abstract – WCN 2013

No: 912

Topic: 8 – Headache

Does pramipexole treatment improve headache in patients with concomitant migraine and restless legs syndrome?

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Background: Recent studies have suggested a link between migraines and restless legs syndrome (RLS). Although these disorders may share a dopaminergic dysfunction in the hypothalamic A11

nucleus that contributes to this association, there have been no clinical studies evaluating the effect of dopaminergic treatment on migraine symptoms in patients with concomitant migraines and RLS.

Objective: To report an illustrative case and to investigate the effects of immediate-release pramipexole (P-IR) treatment on migraine headaches.

Patients and methods: We retrospectively reviewed the medical records of patients who experienced both migraines and RLS.

Results: Ten patients (9 patients from the previously completed single-center study) received P-IR treatment were included in the study. Five out of the 10 patients (50%) reported subjective and objective improvement according to the Clinical Global Impression rating scale for migraine headaches. Of these 5 patients, 4 (80%) reported morning headaches.

Conclusion: Our results suggest that the identification of RLS in migraine patients is important and that dopaminergic treatment may improve both migraines and RLS symptoms.

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Abstract – WCN 2013

No: 753

Topic: 8 – Headache

Cerebellar infarction due to vertebral artery dissection presented with isolated headache in a patient with migraine

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Background: Migraine is a known risk factor for ischemic stroke, especially in young woman. Also, cervical artery dissection is the most common single etiology of stroke in young adults. However the mechanism of migraine-related arterial dissection is still unknown. It is proposed that the repeated attack of migraine would make the involved artery more vulnerable to tearing and lead to dissection.

Patients and methods: (Case) A 34-year-old female was presented with sudden severe headache during 24 h. The patient complained severe headache with nausea and vomiting after awakening. She suffered from migraine for more than 10 years. The throbbing nature headache usually persisted with 1 or 2 days associated with nausea, vomiting, and photophobia. At emergency department, routine laboratory test and brain CT showed normal findings. We started conservative management for decreasing pain. However, headache and vomiting persisted, and dizziness and imbalance were also noted later. After admission, the neurological examination showed dysmetria, gaze-evoked nystagmus, and ataxia. Brain MRI demonstrated a high signal on the diffusion weighted image and low on the apparent diffusion coefficient image in the right cerebellar hemisphere consisted with acute infarction. Cerebral CT angiography revealed the left vertebral artery dissection. The patient was reluctant to use an anticoagulant. Also, topiramate and beta blocker were given to patient for migraine prophylaxis.

Conclusions: The exact mechanism of migraine-related intracranial artery dissection has yet to be proven, we propose that this may be caused by vessel wall edematous changes with repeated migraine attacks resulting in sudden or unusual stretching.

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Abstract – WCN 2013

No: 786

Topic: 8 – Headache

Study on mood and anxiety disorders in patients with tensin-type headache

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Background: Mood and anxiety disorders seem to be closely related to headaches.

Objective: The aim of the study was to evaluate the risk of mood and anxiety disorders in patients with tension-type headache (TTH).

Patients and method: We enrolled in the study 68 patients (55 women and 13 men), aged between 23 and 49 years (mean age 37.5 years) who fulfilled the inclusion criteria: diagnosis of TTH according to the International Headache Society criteria, at least 8 years of education, no previous psychiatric illness. Both mood and anxiety disorders were diagnosed according to DSM IV criteria. The patients were divided into 2 groups:

group A composed of 39 patients (35 women and 4 men) with chronic TTH and

group B composed of 29 patients (20 women and 9 men) with episodic TTH.

All the patients were assessed by MMSE, Hamilton Depression Scale, Hamilton Anxiety Rating Scale, Hospital Anxiety and Depression Scale, and Headache Pain Scale Interpretation. The results were analysed by Student Test and Chi Square Test.

Results: Mood and anxiety disorders appeared in 66% of the patients: 79.4% in group A and 48.2% in group B. The patients had dysthymia (8.8%), unclassified depressive symptoms (26.4%), panic disorders (5.8%), generalized anxiety disorder (8.8%) and association between depression and anxiety (16.1%). None of the patients had major depression. Depressive disorder was found especially in women.

Conclusion: The frequency of depressive disorders was statistically significantly higher in women, but there was no significant difference between groups regarding the type of mood and anxiety disorders.

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Abstract – WCN 2013

No: 483

Topic: 8 – Headache

Gender-specific influence of socioeconomic status on the prevalence of migraine and tension-type headache in Korea

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Background: Primary headache disorders including migraine are famous for gender difference in its prevalence. Socioeconomic status has the important role in pain coping strategy and this influence on primary headache disorder may differ according to their gender.

Objectives: To separately evaluate the influence of socioeconomic variables on prevalence of migraine and tension-type headache according to gender.

Patients and methods: A stratified random population sample of Koreans, over the age of 19, was selected and evaluated using a face-to face interview designed to identify headache type using ICHD-2 criteria. Education levels, the size of district, and household income were evaluated as socioeconomic variables. Age and body-mass index were analyzed as co-variables in multiple regression analysis.

Results: Among 1507 participants, the 1-year prevalence of headache, migraine, and tension-type headache was 70%, 9.2%, and 29.1% in women and was 52.7%, 2.9%, and 32.5% in men. In a multiple regression analysis in women, college level education related to a lower prevalence

of tension-type headache (OR 0.38, 95% confidence interval 0.20–0.73). Interestingly, living in rural areas related to a higher prevalence of migraine (OR 5.30, 95% CI 2.43–11.55) and a lower prevalence of tension-type headache (OR 0.30, 95% CI 0.15–0.61) in women. In a multiple regression analysis in men, all socioeconomic variables had no influence on prevalence of migraine or tension-type headache.

Conclusions: Women are more susceptible to socioeconomic influence on primary headache disorders. Gender-difference in primary headache disorders may be partly related to the gender-specific influence of socioeconomic status.

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Abstract – WCN 2013

No: 329

Topic: 8 – Headache

Cluster-like headache associated with symptomatic Chiari type I malformation: A case presentation

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Background: Several cases of symptomatic cluster-like headaches (CLH) have been reported such as arteriovenous malformations, medullary infarcts, and aneurysms. However, CLH associated with Chiari 1 malformation (CM-1) has been described in only one case.

Objectives: We describe a case considered as CLH associated with symptomatic CM-1.

Case: A 28 year-old female patient visited our outpatient clinic because of recurrent headaches since the age of 20 years. She had severe, recurrent, right frontal and periorbital area pain and headache, and lasted about 20–90 min. She suffered from tearing, rhinorrhea and ptosis during headaches. These pain attacks occurred irregularly two to four times per day. Although attack usually appeared spontaneously, it was precipitated by neck movements. In addition, she also suffered occasional pain in occipital–suboccipital area and vertigo, blackout in the eyes, palpitation, and sweating complaints, all of which would disappear in 5–10 min. Cardiac tests were normal. Cranial/cervical MRI presented CM-1.

Conclusion: While CM-1 and CH are clearly different diseases, determining the actual cause of CH when two pathological processes are present is obviously difficult. CM-1 can be an incidental finding when MRI is conducted for other reasons. In CM-1, variable caudal displacement of cerebellar tonsils occurs into the upper cervical canal. These structural abnormalities in CM-1 may include stretching of cranial nerves or direct compression of brain stem nuclei, and compression of the medulla. The pain of CLH may be associated with dysfunction in an area of the brain stem and/or craniocervical pressure dissociation, stimulating pain-sensitive structures in patients with CM-1.

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Abstract – WCN 2013

No: 433

Topic: 8 – Headache

Role of the insolvent connective tissue in the development of pathological deformation of cerebral vessels in patients with primary headaches

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Aim: Analysis manifestations of connective tissue disease in patients with pathological strain of cerebral vessels.

Methods: The study included 60 patients with primary headaches. Patients were divided into 2 groups: 30 patients with and 30 patients

without abnormalities of the brain. The examination included: clinical neurological research, the study of dysplasia on the questionnaire, MRI, MSCT angiography and duplex scanning.

Results: In the first group of patients with headaches started in childhood with migraine pain, the presence of signs of insolvent connective tissue, whereas in the group without anomalies at puberty or adulthood. Thus, in the first group, patients had increased extensibility, and hyper elastic skin: take the pleated skin was pulled easily by a few centimeters. There was a slight fragility of the skin, even with minimal trauma to heal slowly, leaving colloid scars. In the chronic arthralgia, there were no signs of inflammation in the joints of 13.7% of patients. Frequent complication in patients with failure of the connective tissue is a bundle of extra- or intracranial segments of the vertebral arteries, which were observed in 5 patients with basilar migraine. In the second group, in the group of patients with no signs of abnormalities of connective tissue disease was found.

Conclusion: 1. Revealed diffuse failure of connective tissue in patients with deformities of the brain vessels. 2. It is shown that the systemic connective tissue dysplasia is a risk factor for the development of pathological deformation. 3. To detect pathology of the connective tissue to be included in the survey design of primary headaches screening of connective tissue.

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Abstract – WCN 2013

No: 231

Topic: 8 – Headache

Some features of neuroimaging research methods in the diagnosis of pathological tortuosity of cerebral vessels in patients with primary headaches

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Aim: Analysis of the advantages of MSCT angiography and duplex scanning (DS) of extracranial vessels (ECV) in the diagnosis of pathological tortuosity of cerebral vessels.

Methods: The analysis of MSCT angiography versus DS of ECV in the diagnosis of pathological tortuosity of cerebral vessels.

Results: We observed 100 patients: 60 women and 40 men aged 17–45 years. 81 patients with migraine – 81%, with a cluster headache 4 (4%) and tension-type headache 15 patients (15%). 35 patients were performed MSCT angiography of the brain, which revealed changes observed in 27 patients with migraine and in 1 patient with a cluster headache, in 7 patients with tension headache were detected. The deformation of blood vessels by type kinking and coiling had incidence in women – 53%, compared with men – 47%, in the internal carotid artery (ICA) – 72.3%, in the vertebral arteries (VA) – 28.7%. It was also revealed hypoplasia of the VA: 25.5% of the middle cerebral and in 13.5% of cases in the posterior communicating artery. Pathological tortuosity was presented in the form of C-shaped crimps 12% of patients, S-shaped, 14%, and looped – 8% of cases. Hypoplasias of the ICA and the VA have been observed more in patients with migraine than in patients with cluster headache.

Conclusion: The pathological deformation in cerebrovascular vessels has a high incidence in patients with primary headaches, and in their diagnosis invaluable role to play MSCT angiography of the brain, but with constraints, and in view of the high cost of the study is the method of choice for DS of the ECV.

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Abstract – WCN 2013

No: 380

Topic: 8 – Headache

Clinical analysis of orthostatic headache in Asian patients

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Background: The major cause of orthostatic headaches is cerebrospinal fluid (CSF) leak. But orthostatic headache also occurs in variable diseases.

Objective: We analyzed clinical, radiological, and etiological differences between favorable and non-favorable groups.

Methods: We retrospectively reviewed a group of 44 consecutive patients with orthostatic headache between January 1, 2005 and April 30, 2012. All patients underwent MRI with gadolinium enhancement while clinically symptomatic, and performed conservative therapy with analgesics. Autologous epidural blood patching was carried out in patient who did not respond to conservative therapy.

We divided these patients to two groups, favorable and non-favorable groups. More than two weeks of admission day, two more trials of autologous epidural blood patching, or relapse of orthostatic headache defined as non-favorable group.

Results: 21 (48%) of 44 patients were classified as a favorable group. Favorable group had short hospitalization period (7.5 days vs. 16.0 days, $p = 0.009$) and complain of nausea ($n = 17$ vs. $n = 12$, $p = 0.031$). An unfavorable group had lots of abnormal findings in brain MRI (5 vs. 19, $p < 0.001$). But no significant differences were found between two groups except pachymeningeal enhancement. In unfavorable group, 12 of patients (50%) showed pachymeningeal enhancement in brain MRI (3 patients in favorable group, $p < 0.001$). 16 of favorable group and 7 of non-favorable group were defined as normal brain MRI patients.

Conclusions: Orthostatic headache presenting unfavorable outcome had more brain MRI abnormalities, CSF leakage on upper spinal level. Especially pachymeningeal enhancement could be a sign of unfortunate prognosis.

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Abstract – WCN 2013

No: 322

Topic: 8 – Headache

A self-help program—PROAA improves the quality of life of patients with migraine and/or tension-type headache. A 7-year follow-up

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Background: Headache is a major medical complaint with considerable functional and economic consequences. Psycho-social factors may play a major role in primary headache. Therapeutic strategies controlling these factors may be valuable in headache therapy.

Objective: To evaluate the impact of a self-help program (PROAA) on quality of life (QoL) in patients with refractory migraine and/or tension-type headache.

Patients and methods: Prospective study between 2/01/2003 and 2/01/2010. Patients with unacceptable control of headache-symptoms, according to the treating neurologist, were included.

The PROAA program improves stress-control and anxiety. It is based on the Modeling Development Behaviour (DBM®), physical therapy

and relaxation techniques. For QoL, the SF-12 questionnaire/scale was used before and one month after the program.

Results: 252 patients (222 F/30 M) were included. Median age: 36.6 years. 45% had tension-type headache, 24% migraine and 15.87% both types. For all headache-types, the perceived physical health score (PPHS) before the program was 38.27 ± 9.93 ; after 48.94 ± 7.84 ($p < 0.001$). The perceived mental health score (PMHS) before the program was 31.03 ± 11.68 ; after 47.71 ± 8.68 ($p < 0.001$). For migraine: PPHS before 37.4 ± 9.96 ; after 48.3 ± 8.50 ($p < 0.001$); PMHS before 33.4 ± 12.97 ; after 48.4 ± 9.02 ($p < 0.001$). For tension-type headache: PPHS before 39.1 ± 10.30 ; after 48.9 ± 7.60 ($p < 0.001$); PMHS before 31.2 ± 10.61 ; after 48.2 ± 8.14 ($p < 0.001$).

Conclusion: The self-help program—PROAA significantly improves QoL in patients with migraine and/or tension-type headache and may be useful as non-pharmacological adjunct therapy.

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Abstract – WCN 2013

No: 268

Topic: 8 – Headache

Grey matter in chronic migraine patients

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Introduction: Previous studies have reported a thickening of the somatosensory cortex (SSC) and visual motion processing areas (V3A/MT+) in migraine without aura (MO). However, these changes have not been confirmed. In chronic migraine (CM), atrophy/thinning of amygdala, parietal operculum, middle and inferior frontal gyri, insula and anterior cingulate cortex have been observed.

Objective: To investigate regional cortical thickness/atrophy in CM (ICHD II, 2004) patients (CMP).

Patients and Methods: We compared 23 CMP (age 38.7 ± 11.5 , 20 females) vs 21 controls (age 34.9 ± 4.7 , 13 females). Cortical thickness was measured in 146 regions using FreeSurfer and volumetric T1 weighted images.

Results: Migraineurs presented a mild focal thinning of the grey matter of the right ventral posterior cingulate cortex (vpCC) (6.4%, $p = 0.014$) and left pericallosal sulcus (PS) (4.7%, $p = 0.046$). Thickening of the left occipito-temporal gyrus (12.8%, $p = 0.006$), right occipito-temporal sulcus (5.7%, $p = 0.048$), left inferior temporal sulcus (6.0%, $p = 0.023$), right transversal temporal sulcus (12.4%, $p = 0.016$), belonging to the V3A/MT+ areas, and of the left postcentral gyrus (7.0%, $p = 0.034$), that is part of the SSC was observed.

Conclusion: The thinning of several cortical regions reported in CM was not confirmed; our preliminary result of a cortical thinning of the right vpCC and left PS, not observed previously, needs to be confirmed. Our finding of a thickening in the SSC and V3A/MT+ areas, not reported in the only study performed on CM, but in keeping with previous observations in MO, supports the hypothesis that repetitive attacks could lead to neuroplastic changes in the grey matter.

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Abstract – WCN 2013

No: 515

Topic: 8 – Headache

Electroencephalographic abnormalities in patients with migraine

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Background: Migraine is one of the humanity's most common afflictions.

Objective: The aim of this study is to describe the electroencephalographic (EEG) abnormalities in patients with migraine in comparison with control group.

Methods: This prospective study was conducted in our center from January 2011 to January 2012. We report the interictal EEG of 24 migraineurs, diagnosed according to International Headache Society (IHS) criteria, in comparison with control group. We observed abnormal patterns on EEG in 12 migraineurs with aura, in 12 migraineurs without aura and in 24 healthy persons. EEG was performed between attacks of headaches (interictal phase) three times during one year.

Results: Abnormal EEG was found to be much more frequent in migraineurs than the control group (36% vs. 9%). The most common abnormality was slow high voltage unilateral or bilateral occipital waves, which was observed in 58.8% migraine patients with aura, 35% patients without aura, and 9% in control group. The frontal intermittent rhythmic delta activity (FIRDA) was significantly higher in the migraine group than the control group (26% vs. 6%). The less common finding was focal spikes which detected in 9% in migraineurs with aura, 4.1% in migraineurs without aura and 0% in control group.

Conclusion: EEG abnormalities are reported more common in migraineurs, especially in patients with aura, than in headache-free control subjects. The presence of EEG abnormalities in migraine may help to initiate further researches to determine the pathogenesis of these EEG changes in migraine as well as the management and prognosis.

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Abstract – WCN 2013

No: 488

Topic: 8 – Headache

Ultrasound guided onabotulinum injections in chronic migraine: Analysis of first data of our experience

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Purpose: During the early years of botulinum toxin type A (BoNT-A) treatment for motor conditions, investigators noted a significant benefit of BoNT-A on pain that often exceeded the improvement in muscle contractions. Recently BoNT-A received indication for the treatment of chronic migraine (CM). In this report we analyze how ultrasound-guided techniques, allowing the visualization of tissues, potentially improve the accuracy of the needle placement exemplifying BoNT-A procedures also in the treatment of untreatable CM with onabotulinum toxin A.

Method: Patients with CM and inadequate response or intolerance to previous two prophylactic treatments and considered "untreatable class I CM" patients were treated with pericranial injections of about 150 UI of BoNT-A every 3 months. The dose could be increased up to 200 U in case of no response. Patients reported headache diary, MIDAS score effect of BTA on the frequency of disabling attacks, and consumption of analgesics.

Results: All meet the IHS criteria for analgesic overuse. The number of sessions studied with BTA was 4. Patients achieved reduction >70% in headache frequency and intensity. Those four patients reduced NSAID and analgesic consumption significantly.

Conclusion: US guidance provides clear advantages also in CM patient injections because it permits to visualize the spread of BTA from the needle and to validate procedure's outcome. US guided

BoNT-A injections could be a profitable option both clinically and pharmaco-economically.

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Abstract – WCN 2013

No: 454

Topic: 8 – Headache

Acute administration of 5-HT precursor 5-hydroxytryptophan upregulates constitutive isoform of neuronal nitric oxide synthase activity in the rat brain

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The possibility that endogenous elevation of serotonin is influencing blood–brain barrier (BBB) permeability, neuronal nitric oxide synthase activity was examined in the brain and spinal cord of rats. Elevation of endogenous serotonin level was achieved by administration of its precursor, 5-hydroxytryptophan (150 mg/kg, i.p.). The elevation of serotonin level in the brain and spinal cord was confirmed by immunohistochemical demonstration of 5-HT activity. Four hours after administration of 5-HTP, the animals exhibited hyperthermia and other behavioral symptoms. In these animals, ultrastructural studies revealed breakdown of the BBB permeability at this time period. In 5-HTP pretreated rats, there was a marked increase in neuronal NOS labelling in the several brain regions and in the spinal cord. Morphological examination suggests profound cell injury and edema in the regions associated with NOS upregulation. These observations for the first time demonstrate that endogenous elevation of 5-HT is associated with NOS upregulation. Alternatively, 5-HT mediated cell injury, BBB disruptions are regulated by NOS, not reported earlier.

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Abstract – WCN 2013

No: 246

Topic: 8 – Headache

Recent concept in pathophysiology and treatment of pseudotumor cerebri (PTC)

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Objectives: Study the role of intracranial venous sinuses in the pathophysiology of pseudotumor cerebri.

Materials and methods: Twenty patients diagnosed as PTC according to Dandy diagnostic criteria underwent general and neurological assessment, ophthalmologic assessment, laboratory investigations were done, radiological assessment included CT scan brain ± MRI brain without contrast, and MRV of the intracranial venous system. Lastly all underwent digital subtraction cerebral angiography (DSA) (venous phase) to confirm the validity of filing gaps seen at the level of MRV whether they are true stenosis or not.

Results: MRV brain showed that 14 patients (70%) showed filling gaps suggestive of sinus stenosis. However, digital subtraction cerebral angiography (venous phase) gave different data that only 5 patients (25%) had stenosis in their dural sinuses. MRV showed to be a good screening tool since it had 100% sensitivity and negative predictive value. Therefore, if MRV is normal no further investigations are needed. However, since it has a moderate specificity (62%) with a positive predictive value (PPV) of only 35%, then lesions detected should be confirmed with digital subtraction cerebral

angiography (venous phase) particularly those involving the transverse and sigmoid sinus.

Conclusion: Studying the intracranial venous system in patients with PTC is an important step in understanding the pathophysiology of the disease. Detection of venous sinus stenosis opens the way to a novel therapeutic option for refractory patients like venous sinus.

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Abstract – WCN 2013

No: 439

Topic: 8 – Headache

Total migraine freedom (TMF) for single pulse transcranial magnetic stimulation (sTMS) versus triptans for the early acute treatment of migraine

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Background: sTMS is effective and well tolerated for the acute treatment of migraine with aura. Triptans are the gold-standard for acute treatment of migraine. No head-to-head studies have compared the efficacy of sTMS to triptans. Total migraine freedom (TMF) at two hours (2 h TMF) is a composite endpoint that incorporates freedom from pain, photophobia, phonophobia, and nausea. It is a sensitive, patient-centric, clinically relevant summary measure which more powerfully measures treatment effects than the four co-primary endpoints it incorporates.

Objective: To compare the magnitude of the treatment effect for sTMS versus triptans in randomized placebo-controlled trials using an early treatment design where treatment was administered while pain was mild.

Methods: The literature was systematically reviewed to identify placebo-controlled mild pain trials of acute migraine treatment with sTMS or triptans. We identified one sTMS study and 3 triptan trials. For each study we computed the absolute risk reductions (ARR) for 2 h TMF (2 h TMF for active, –2 h TMF for placebo). Data was summarized including all sites and excluding a site which had a high placebo response.

Results: ARR for 2 h TMF for the sTMS study were 12.2% for all sites and 26.3% excluding a single site. For triptan trials, ARR for 2 h TMF were: rizatriptan 10 mg (27.5%), eletriptan 20 mg (10.4%), eletriptan 40 mg (41.0%), sumatriptan 50 mg, (21.4%), and sumatriptan 100 mg (28.1%).

Conclusions: TMF is a patient-centric, clinically relevant, composite endpoint. sTMS demonstrates efficacy in a range that overlaps with the triptans for this robust treatment outcome measure. Head-to-head comparative studies of triptans with sTMS are recommended.

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Abstract – WCN 2013

No: 16

Topic: 8 – Headache

Concomitant administration of alprazolam and ibuprofen in acute migraine headache

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Background: Migraine is a chronic neurologic disorder which a wide variety of drugs are used for its prevention and treatment. Combination

therapy has shown its efficiency in treating migraine. In this study, we have administered the combination of ibuprofen and alprazolam as a probable efficient compound.

Material and methods: In this clinical trial study, 90 migraine patients (with 2–6 attacks per month) were allocated in 3 groups of 30. Patients were treated with single dose of either ibuprofen 200 mg, ibuprofen 400 mg, or ibuprofen 200 mg plus alprazolam 0.5 mg. Headache severity, functional disability, and associated symptoms were recorded and before and 2 h after taking each regimen; and were scored from 0 to 3. Then three groups were compared statistically.

Results: There were no significant differences regarding age, gender and drug histories among the 3 groups. The severity of the headache was decreased significantly after drug therapy in all 3 groups (36%, 46%, and 74% respectively; $P < 0.001$) and statistically significant decreases in patient's pain, nausea/vomiting, photophobia, and phonophobia scores during migraine attack were observed. On the other hand, an improvement in patients' function was seen in all treatment groups. Ibuprofen 200 mg plus alprazolam was more effective than ibuprofen 200 mg alone.

Conclusion: All three drug regimens (ibuprofen 200 mg, ibuprofen 400 mg, and ibuprofen 200 mg plus alprazolam; in single doses) showed efficacy in migraine treatment that are effective in treating migraine attacks. The analgesic effect of ibuprofen in migraine headache is augmented by alprazolam.

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Abstract – WCN 2013

No: 12

Topic: 8 – Headache

Are the ICHD-II migraine diagnostic criteria scientifically valid?

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Background: The International Headache Society classification of migraine – the ICHD-II – is almost universally accepted by researchers and clinicians. It is highly unlikely that reputable journals will accept submissions for publication if the cohorts have not been selected strictly according the ICHD-II criteria. Likewise, in the clinical setting, treatment is prescribed according to how the patient's headache is classified.

Objective: To examine the validity of the ICHD-II criteria for the diagnosis of migraine.

Material: The data on which the ICHD-II migraine diagnosis is based are examined.

Results: The ICHD-II migraine diagnostic criteria are not based on data.

Conclusion: The ICHD-II migraine diagnostic criteria have no scientific basis, and their scientific validity is questionable. As the ICHD-II migraine diagnostic criteria cannot be scientifically validated, then the results of research based on the ICHD-II also have no scientific validity. The practical implications of this are that the research data on the efficacy of migraine drugs may be compromised. Patients diagnosed according to a classification that is not supported by scientific evidence may also be given the incorrect treatment.

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Abstract – WCN 2013

No: 71

Topic: 8 – Headache

Migraine with prolonged aura (global dysphasia):
A clinical reminder

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Migraine is a very common condition being more prevalent in females. Recent advances in medicine have demonstrated the high complexity in the clinical presentations, variable clinical features and diagnosis of this condition. Many studies have shown the overlap of signs and symptoms between migraine and stroke/transient ischaemic attacks. Here, we describe such a complex case with classical migraine headaches associated with global dysphasia. A 65 year old lady presented with unilateral left sided headache and global dysphasia. There was no convincing history of any vascular disease, head trauma or signs of infections. Collateral history from relatives eluded to sudden onset confusion noticed by the family which led to the patient's attendance at the hospital. There was no history of head trauma, loss of consciousness, decrease in cognition, inattention, slurring of speech, motor weakness or sensory disturbance. Past medical history included previous history of migraine however she was currently not on any medication for migraine. On examination vital signs were stable with no focal neurological deficit except for expressive and receptive dysphasia that started simultaneously with the headache. Fundoscopy was normal. No negative symptoms. Neurological examination did not show any focal neurological deficits apart from global dysphasia. CT head and LP ruled out life threatening conditions. After 18 h, symptoms resolved spontaneously and patient was diagnosed to have migraine with prolonged aura of global dysphasia. We urge a high index of suspicion for atypical presentations of migraine especially with prolonged aura as these may easily be confused with stroke and transient ischaemic attacks.

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Abstract – WCN 2013

No: 261

Topic: 8 – Headache

Anticephalgic photoprotective premedicated mask: A report of a successful study of a treatment for migraine and/or tension headaches

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Objectives: This study was performed to determine the efficacy of an anticephalgic photoprotective mask in conjunction with a topical medication containing *Bryonia* and *Rhus toxicodendron* in the treatment of migraine and/or tension headache.

Background: Many clinicians are seeking headache treatment modalities with improved safety profiles. A premedicated mask would serve not only as a delivery system for benign topical medication, but simultaneously provide photorelief and exert external pressure which may alleviate vascular headaches by collapsing painfully distended extracranial arteries and reducing peripheral sensitization.

Methods: Thirty-three patients were given masks and tubes of topical medication containing the *Bryonia* and *Rhus toxicodendron*. They were instructed to apply the medication to their frontalis and/or temporalis regions in the event they should suffer a headache and apply a photoprotective mask. They subsequently filled out forms rating the degree of relief which they attributed to the topical medication and the mask using a 0–10 scale.

Results: Thirty out of 33 patients stated that the medication and the mask were effective over and above the normal degree of relief they were receiving from their oral and/or parenteral medications. This study demonstrated a significant efficacy rate (91%) in the treatment of migraine and/or tension headache with the anticephalgic mask in conjunction with a topical cream containing *Bryonia* and *Rhus toxicodendron*.

Conclusions: This study demonstrated a significant efficacy rate in the treatment of migraine and/or tension headache with the anticephalgic

mask in conjunction with a topical cream containing *Bryonia* and *Rhus toxicodendron*.

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Abstract – WCN 2013

No: 264

Topic: 8 – Headache

Chronification of migraineurs presenting various cervical abnormalities

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Object and Background: Despite of therapeutic approaches, some migraineurs have progression to chronic course. The chronification of migraine disturbs the quality of patient's life. Chronic migraine headache is an enormous burden.

Generally speaking there are no abnormal findings in laboratory tests in migraineurs according to the second edition of the International Classification of Headache Disorders (ICHD-2). In practice chronic migraineurs show some abnormalities in cervical spine. There are no references that chronic migraineurs have any abnormalities in cervical spine.

Subjects and methods: The procedure was performed during the physical examination from Jan. 2009 to Nov. 2012 at Dept. of Neurology, Pusan National University Hospital. The 200 patients with migraine without aura, who complained of chronic headache for more than 3 years, were included.

The exact location may be a good clue or guide to improve diagnostic accuracy, we applied Korean hand therapy method to confirm migraine headache using the location of tender points on the head and neck.

Cervical spine study was included during routine laboratory tests.

Results: Among 200 chronic migraineurs, the number of female was 148, that of male was 52. Mean age was 48 years old.

The X-ray finding of cervical vertebrae in migraineurs (200 persons) was various.

Conclusion: The chronic migraineurs having chronic progressive course and long periodic cycles for more than 3 years have some abnormalities on the radiological study.

It may be helpful to include the cervical spine study in the chronic migraineurs and to consider the correction of cervical abnormalities for prevention of chronic processes.

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Abstract – WCN 2013

No: 263

Topic: 8 – Headache

The pain location for diagnosis of primary headache is closely related with Gold Meridian System of Korean Hand Therapy

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Background: The diagnosis of primary headache has not been sufficient for appropriate treatment. We mainly depend on the history taking and criteria of International Headache Classification. The location of headache might be crew to diagnosis. We emphasized to manage the patient individually as personalized medicine. In Eastern Asian Medicine, there are meridian and acupuncture points on the body

including on the head. It is well described but it is complicated to use easily. We need simplified meridian and acupuncture point system. We propose to use Gold Meridian and Acupuncture Points of Korean Hand Therapy to determine the location.

Subjects and methods: This procedure was performed during physical examination based on well performed history taking at Department of Neurology, Pusan National University Hospital from March 2011 to Feb. 2012. The 200 primary headache patients were included. We checked pain location on both sides and sites of head using 20 new modified acupuncture points on gallbladder meridian (CM1–12) and urinary bladder (CI1–8) on each side.

Results: The headache points are grouped such as gallbladder and urinary bladder gold meridian system, the migraine headache patients belonged to gallbladder and tension type headache belonged to urinary bladder meridian, mixed type headache belonged to various combined gallbladder and urinary bladder meridians.

Conclusion: The pain location of intractable headache patients presented as mixed form headache. The location of pains of such headache should be considered for Botox injection. We can inject small dosage of Botox injection depending on right or left exact side and sites.

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Abstract – WCN 2013

No: 255

Topic: 8 – Headache

Aquaporin-4 antibodies in Egyptian patients with idiopathic intracranial hypertension

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Background: The aetiology of idiopathic intracranial hypertension (IIH) is not clear. A dysfunction of the water channels could affect the cerebrospinal fluid (CSF) secretion or absorption and potentially contribute to its pathogenesis.

Objective: is to assess the possible role of aquaporin (AQP)-4 antibody in the pathogenesis of IIH.

Subjects and methods: Eighteen symptomatic IIH patients who were diagnosed according to the updated modified Dandy criteria were selected from the Neurology Department of Kasr El-Aini Hospital, Cairo University. Detection of anti-aquaporin antibodies in their CSF samples was measured by ELISA based assay.

Results: The CSF samples of IIH patients were positive for the aquaporin (AQP)-4 antibody in 16 (88.9%) of the patients. The CSF level of AQP-4 antibodies was 15.59 ± 8.03 U/ml.

Conclusions: There are two reports investigating the anti-AQP4 antibodies in IIH. Our study contradicted the results of these reports and indicated that AQP4 antibodies have a role in IIH pathogenesis and provided some support for the contribution of inflammatory mechanisms in IIH.

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Abstract – WCN 2013

No: 236

Topic: 8 – Headache

HaNDL syndrome presenting during pregnancy

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Background: Headache associated with neurological deficits and cerebrospinal fluid lymphocytosis (HaNDL) is a self-limited syndrome characterized by sudden-onset headache with a temporary neurological deficit and cerebrospinal fluid (CSF) lymphocytosis.

Objective: We report a case of HaNDL syndrome presented during pregnancy.

Patient: A 20-year-old female presented with a 5-day history of severe, bilateral throbbing headache accompanied by nausea, vomiting, and phonophobia. Approximately 2 days after the pain developed, she became acutely confused for less than 90 min. There were no symptoms consistent with meningoencephalitis. Her medical and family histories were unremarkable. She was pregnant and at 11 weeks gestation. A fundoscopic examination showed bilateral papilledema with no visual field defects.

The neuroradiological examination was normal. A lumbar puncture was performed. The CSF revealed lymphocytic pleocytosis, mildly elevated protein (57 mg/dL), and increased opening pressure (320 mm H₂O). After excluding all other conditions that cause CSF pleocytosis and severe headache, a diagnosis of HaNDL was made.

Conclusion: HaNDL is considered a secondary headache syndrome. The precise etiology is unknown, although an inflammatory or infectious origin and autoimmune factors have been proposed. Moreover, the risk factors and medical conditions associated with HaNDL are unknown. It is obviously difficult to determine whether the pregnancy was coincidental or associated in this case. We believe that comprehensive studies are needed to clarify the risk factors and medical conditions associated with HaNDL.

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Abstract – WCN 2013

No: 102

Topic: 8 – Headache

Venous discirculation

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Research objective: Assessment of cerebral venous hemodynamics in children and teenagers having cranialgia with updating cause-effect relations of venous discirculation.

According to the data of ultra sonic testing (in C- and PW-modes) 109 patients (at the age from 2 to 18) it was found that discirculation in the system of vertebral veins is linked with apparent extravasal effects on bloodstream in internal jugular vein (with vessel compression on the side of vasoconstriction registration) ($r = +0.67$), spasms of posterior cerebral artery ($r = +0.63$), coiling of internal carotid and vertebral artery ($r = +0.20, +0.32$). In case of discirculation in the system of internal jugular vein, the changes mentioned are interrelated with extravasal compression on the level of internal jugular vein surrounding soft tissues or compression in the bone canal ($r = +0.76$), anterior cerebral artery spasms, hyperperfusion of vertebral and posterior cerebral artery ($p < 0.05$). Intracranial venous discirculation depends on the straightness of vertebral artery in the bone canal ($r = +0.33$), discirculation intensity on the level of vertebral vein (V_{max} right $r = +0.73, p > 0.05$).

Vasoconstriction in the vein of Galen is accompanied by ipsilateral hypersthenia of vertebral, internal carotid and middle cerebral artery

(reflectory changes), and is also interrelated with flexures, sigmoid coiling of internal carotid artery.

The link of “headache syndrome” with accelerated venous blood flow along the veins of Galen turned out to be quite low ($r = +0.22$).

Conclusion: Main causes of children's vasoconstriction are either congenital pathology of cervical spine (with arcuation and tortuosity of bone canal), or “birth injuries with pseudo-luxation of cervical vertebrae”.

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Abstract – WCN 2013

No: 92

Topic: 8 – Headache

Idiopathic hypertrophic cranial pachymeningitis: A rare cause of chronic headache and progressive cranial neuropathies:

A case report

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Background and objective: We report a case of a 75-year-old male with chronic daily headache for more than one year and later on developed deafness, vocal cord paralysis, ophthalmoplegia, and loss of vision. On physical examination, there was ptosis and left gaze palsy, pale optic disc, only light perception on both eyes, weak gag reflex and gross hearing loss.

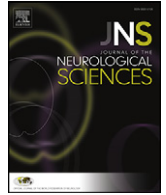
Methods: Work up included cranial MRI that revealed abnormal pachymeningeal enhancement or thickening of the dura in the posterior fossa, cavernous sinus and orbital apex and bilateral mastoiditis. CSF analysis showed elevated CSF protein, negative results for AFB, KOH, VDRL, TB and bacterial culture, and no malignant cells. ANCA, ANA and RF were negative. Search for occult malignancies was negative. There was elevated ESR, consistent with an inflammatory process.

Idiopathic hypertrophic pachymeningitis is a rare chronic fibrosing inflammatory disease characterized by marked diffuse thickening of the cranial dura that causes progressive neurological deficits by compression of anatomic structures by the meninx, thickened by inflammation.

Results: After initiation of pulse corticosteroid therapy, methotrexate and maintenance oral prednisone, there was remarkable improvement on hearing, swallowing, and ocular muscle movement, less headaches and marked regression of pachymeningeal enhancement on repeat MRI. However, profound vision loss remained unchanged.

Conclusion: A high index of suspicion of this rare disease is important because early institution and long-term maintenance of steroid may result to complete or partial remission of the neurologic deficits and may help prevent irreversible neurologic sequelae, especially blindness.

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Topic: 9 - Pain

Abstract – WCN 2013

No: 7

Topic: 9 – Pain

The effects of prenatal morphine exposure on pain response

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Background: Drug abuse during pregnancy is a growing problem in all developed countries of the world. Maternal drug abuse affects the developing system and its long-term effects can persist till adulthood so it can decrease the rate of their maturation. Since endogenous opioid induced analgesia and morphine can interact with it, the present study was designed to determine whether the exposure to the morphine during gestation permanently alters pain response.

Objective: To determine the effects of prenatal morphine exposure on pain response.

Materials and methods: 12 pregnant rats were divided to morphine and control groups. Morphine was administrated (S.C.) to female rats twice a day (08 h and 20 h) on gestational days 11–18, (5 mg/kg morphine for 3 days and 10 mg/kg for 5 days). Analgesic response of pups (P90, n = 6) were tested by formalin test.

Finding: The results of our experiment demonstrated that prenatal morphine exposure rats exhibited significantly lower pain thresholds.

Conclusion: Prenatal morphine exposure impairs pain sensitivity.

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Abstract – WCN 2013

No: 468

Topic: 9 – Pain

Life without pain—A family presenting with HSAN type 1 and dementia

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Hereditary sensory autonomic neuropathy type I (HSAN I) is a neurological disorder characterized mainly by distal sensory loss and autonomic symptoms. This autosomal dominant inherited disorder manifests typically during the 2nd–4th decades of life. This case report presents four family members who among the classic features of HSAN I also developed deafness and dementia. We had the opportunity to examine only one member, while the data for other three members were taken retrospectively as they were dead at that time. The four of them, father and three sons, presented with similar clinical findings: sensory disturbances in lower extremities which began during their twenties followed by painless ulcerations in these

areas. Shortly after, hearing difficulties progressing to deafness appeared and by late thirties their clinical condition deteriorated further with dementia. All patients died before the age of fifty. The absence of sensory nerve action potentials characterized the EMG of the examined patient, while a predominant frontal atrophy was evident on his head MRI.

Dementia is a very rare finding in HSAN type I disease course. Currently, only few cases that have occurred in Japan and USA are described in literature.

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Abstract – WCN 2013

No: 496

Topic: 9 – Pain

Sensory profile and its impact on quality of life in patients with painful diabetic polyneuropathy

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Background: Painful diabetic polyneuropathy (PDN) is common and causes significant disability. Sensory profile in each patient is different and affects quality of life.

Objective: To describe the demographic, details of sensory profile and its impact on quality of life in patients with PDN.

Patients and methods: A cross-sectional survey in patients with PDN who were treated in a university hospital. They were interviewed with standard questionnaires, which included neuropathic pain scale (NPS), short-form McGill Pain Questionnaire (SF-MPQ) and short form-36 quality of life survey (SF-36). Descriptive statistics were used in demographic data. Student's t test was used to analyze continuous data. Multiple comparisons for proportions and correlations were by Pearson Chi-Square test and Pearson's coefficient of correlation, respectively.

Results: Thirty three patients were included in this study. 58% were female. Their average age, duration of diabetes and duration of pain were 60.5, ten and four years respectively. In NPS, sharp pain is most common and itching is least common. However, the unpleasantness was most affected with score 6.9. Mean VAS was 56 mm. In SFMPQ, sensory score, affective score and present pain score were in moderate range. In SF-36, the most affected domain is physical functioning and the least affected domain is social function.

Conclusion: PDN affects to both physical activity and quality of life in these patients. Almost all patients have many types of pain and sharp pain is most common.

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Abstract – WCN 2013**No: 180****Topic: 9 – Pain****Differential endogenous pain modulation in patients with neuropathic pain after spinal cord injury correlates with contact heat evoked potential amplitude**

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Background: Central neuropathic pain (NP) is associated with change in endogenous pain modulation (EPM) evoked by hetero-topic noxious conditioning stimulation (HNCS).

Objective: Characterise EPM disruption in patients with NP following spinal cord injury (SCI) and its relationship with contact heat evoked potentials (CHEPs).

Methods: Ten healthy subjects and 20 patients with SCI (injury level below Th2) were recruited, 10 of which presented non-evoked NP (>5/10 NRS). Cz/Fz-CHEPs with corresponding perception of evoked heat pain (EvHP) from the C6 dermatome were assessed. Test stimulation (TS) was a 30 s tonic heat stimulus with the plateau intensity set individually at the temperature that evoked 3/10 (0–10, NRS, “pain-3”). The HNCS was a 30 s immersion of the contralateral hand in an innocuous (33 °C) or painful (12 °C) water bath. Subjects performed three protocols: non-conditioned TS; innocuous-HNCS TS and noxious-HNCS TS. TS pain intensity was assessed at 10 s (t1), 20 s (t2) and 30 s (t3).

Results: Significant net t1 EPM of TS intensity was evoked in the healthy (-1.0 ± 0.3 ; $p < 0.01$) and SCI-noNP groups (-0.8 ± 0.3 ; $p < 0.05$), but not in the SCI-NP group ($+0.2 \pm 0.3$; $p > 0.05$). Mean net t1–t3 EPM was different between the healthy and SCI-NP groups ($p = 0.04$). Importantly mean net t1–t3 EPM correlated highly with the CHEP amplitude only in patients with SCI-NP ($\rho = 0.8$; $p = 0.015$), suggesting that disruption of EPM following NP may be quantified with this objective biomarker of pain perception.

Conclusion: Endogenous inhibitory pain modulation is lost in subjects with SCI NP and is reflected by an increase in sensory evoked potentials in response to contact heat stimulation.

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Abstract – WCN 2013**No: 549****Topic: 9 – Pain****Palliative care for HIV/AIDS in Kenya**

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Background: Patients with HIV/AIDS on Antiretroviral Therapy (ART) suffer from physical pain, psychological and spiritual problems. Despite international policy explicitly stating that a multidimensional approach such as palliative care should be delivered throughout the disease trajectory and alongside treatment, the effectiveness of this approach has not been tested in ART-experienced populations.

Methods: This mixed methods study uses a Randomised Controlled Trial (RCT) to test the null hypothesis that receipt of palliative care in addition to standard HIV care does not affect pain compared to standard care alone. An additional qualitative component will explore the mechanism of action and participant experience. The sample size is designed to detect a statistically significant decrease in

reported pain, determined by a two tailed test and a p value of ≤ 0.05 .

Results: Recruited patients will be adults on ART for more than one month, who report significant pain or symptoms which have lasted for more than two weeks. The intervention under trial is palliative care delivered by an existing HIV facility nurse trained to a set standard. Following an initial pilot the study will be delivered at Kenya Medical Research Institute, using two parallel independent Phase III clinical RCTs. Qualitative data will be collected from semi structured interviews and documentation from clinical encounters, to explore the experience of receiving palliative care in this context.

Conclusion: The data provided by this study will provide evidence to inform the improvement of outcomes for people living with HIV and on ART in Africa.

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Abstract – WCN 2013**No: 272****Topic: 9 – Pain****Preoperative intradermal acupuncture reduces postoperative pain, nausea and vomiting, analgesic requirement and sympathoadrenal responses**

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Background: In a controlled and double-blind study, preoperative insertion of intradermal needles at acupoints 2.5 cm from the spinal vertebrae (bladder meridian) provides satisfactory postoperative analgesia.

Methods: Enrolled patients scheduled for elective upper and lower abdominal surgery. Before anesthesia, patients undergoing each type of surgery were randomly assigned to one of two groups: acupuncture ($n = 50$ and $n = 39$ for upper and lower abdominal surgery, respectively). In the acupuncture group, intradermal needles were inserted to the left and right of bladder meridian 18–24 and 20–26 in upper and lower abdominal surgery before induction of anesthesia, respectively. Postoperative analgesia was maintained with epidural morphine and bolus doses of intravenous morphine. Incisional pain at rest and during coughing and deep visceral pain were recorded during recovery and for 4 days thereafter on a four-point verbal rating scale.

Results: Starting from the recovery room, intradermal acupuncture increased the fraction of patients with good pain relief as compared with the control ($P < 0.05$). Consumption of supplemental intravenous morphine was reduced 50%, and the incidence of postoperative nausea was reduced 20–30% in the acupuncture patients who had undergone either upper or lower abdominal surgery ($P < 0.01$) and epinephrine concentrations were reduced 30–50% in the acupuncture group during recovery and on the first postoperative day ($P < 0.01$).

Conclusion: Preoperative insertion of intradermal needles reduces postoperative pain, the analgesic requirement, and opioid-related side effects after both upper and lower abdominal surgery.

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Abstract – WCN 2013**No: 680****Topic: 9 – Pain****Latent modulation of neuropathic pain intensity via hypothalamus–pituitary–thyroid axis of psychogenic stress**

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Background: In patients with complex regional pain syndrome (CRPS) delayed pain flares consistently occur ten days following salient

psychogenic stress episodes. Timing of latent flares suggests pain modulation via hypothalamus–pituitary–thyroid (HPT) axis hormones. **Objective:** To determine if thyroxine (T₄) may modulate latent stress-related neuropathic pain flare intensity, temporal relationships between daily stress, serum T₄ levels, and perceived pain intensity in patients with CRPS were investigated.

Patients and methods: Daily, for ten weeks, three patients with type I CRPS and no thyroid pathology Hx provided blood samples for T₄ assay and ratings of stress and pain. Measures included visual analog pain scale, McGill pain questionnaire, and Daily Stress Scale. Blood draws yielded bound T₄ and free thyroxine index (FTI) values using microplate enzyme immunoassay. Each sample was split for blind assay from two independent labs.

Results: Across patients, 14 peak stress episodes and 26 significant pain flares were reported. Each stress episode was followed ten days by a significant pain flare and free T₄ values exceeding normal adult range (2.4 ng/dL). Serial lag correlations were strongest between stress and pain for pain experienced ten days after peak stress episodes ($r = +0.381$, $p < 0.05$). FTI correlated strongest with stress ten days following a stressful episode ($r = +0.454$, $p < 0.001$). Same-day pain and FTI correlated at $r = +0.643$, $p < 0.001$.

Conclusions: Increased pain ten days following stressful events is related to psychogenic HPT activity in CRPS patients. Pain modulation by thyroxine may assist understanding pain fluctuation etiology and suggest new treatment avenues for managing neuropathic pain.

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Abstract – WCN 2013

No: 339

Topic: 9 – Pain

Duloxetine-induced life-threatening long QT syndrome

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Background: There are few reports of cardiovascular adverse effects of DH.

Objective: The present article describes a case of prolonged QTc and life-threatening arrhythmias in patient treated with DH.

Patients and methods: A 52-year-old Slovenian female was admitted in 2012 to hospital, because of resistant major depressive disorder (MDD). Her medications previous to hospitalization included bupropion 300 mg daily (BUP). There was no family history of sudden cardiac death. Treatment with DH titrated from 30 mg to 90 mg daily was introduced in the hospital, which was followed by gradual symptoms improvement (QTc = 388 ms, normal heart rate and blood pressure). After two months of this treatment, palpitations and loss of consciousness were reported with increased heart rate (76 bpm), sinus arrhythmia and prolonged QT (QTc = 460 ms) interval. Clinical pharmacist advised dose reduction to 30 mg of DH daily. After a one-week dose reduction of DH to 30 mg daily, cardiovascular status returned to normal (QTc = 398 ms, 65 bpm). She left hospital 2 weeks later.

Results: These findings were attributed to adverse effect associated with DH use. Symptoms of MDD were treated successfully. CYP2D6 has a major role in the metabolism of BUP, so we believe that pharmacokinetic drug interaction with DH and BUP occurred, which led to toxic concentration of DH.

Conclusion: In conclusion, even though DH is generally known to be a well-tolerated and safe drug, physicians and clinical pharmacists should be aware of the risk of cardiovascular adverse effects.

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Abstract – WCN 2013

No: 736

Topic: 9 – Pain

Refractory chronic pain due to compressive myelopathy:

Case report

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Background: Spondylolisthesis prevalence is reported to increase with age and to be higher among women than men and knowledge about progression of this disease is limited.

Objective: To report a case of refractory chronic pain which investigation showed compressive myelopathy secondary to severe spondylolisthesis and degenerative cervical spondylodiscopathy, without neurological deficit.

Patients and methods/material and methods: Anamnesis, physical examination, magnetic resonance imaging (MRI) and medical literature.

Results: CMM, female, 71 years old, reported auto accident 11 years ago with right humerus fracture and subsequent surgery. Some months later a cervicobrachial, thoracic and lumbar pain described as stab, intermittent and severe started. Ten years ago began treatment for chronic pain, without improvement. On examination global hyperreflexia was found. MRI showed compressive myelopathy with areas of reduced diameter of spinal canal associated with edema and myelomalacia: important anterior axial subluxation of C5 above C6 and degenerative cervical spondylodiscopathy especially from C4 to C7. Surgery is indicated for listhesis larger than 50% and refractory pain, however, given the instability of the cervical spine and age, the treatment was palliative. Diagnosis could be earlier on emergency care after the auto accident with X-ray; spinal pain and age above 50 years old would indicate resonance at the beginning of ambulatorial treatment.

Conclusion: This case report shows some peculiarities. The first one is rarity of myelopathy associated to severe listhesis without neurological deficit. The other one is about the medical intervention: the possibility of early diagnosis and the choice of palliative treatment.

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Abstract – WCN 2013

No: 740

Topic: 9 – Pain

Changes in skin tissue modulate

immobilization-induced hypersensitivity

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Background: Cast immobilization is known to induce pain in humans and experimental animal models. However, the detailed mechanism underlying this pain has not yet been elucidated. Recently, several lines of evidence indicate a complex system of communication between keratinocytes and the sensory afferents innervating the skin, which modulate pain sensitivity.

Objective: In this study, we investigated 1) changes in sensory nerve fibers; 2) whether nerve growth factor (NGF) expression increases in the epidermis; and 3) whether the expression of ion channels, particularly transient receptor potential vanilloid 1 (TRPV1) and purinergic receptor P2X₃, changes in the epidermis in plantar skin of rat hind paw after 1, 2, and 4 weeks of ankle joint immobilization by casting.

Material and methods: The von Frey test and plantar test were performed to examine noxious sensitivity of the skin. Sensory nerve fiber densities were assessed in the basal epidermal layer and expression levels of NGF, TRPV1, and P2X₃ in the epidermis were examined by immunohistological methods.

Results: In the skin of immobilized rats, both myelinated A fibers and unmyelinated C fibers were increased. NGF, TRPV1, and P2X₃ expression levels in the epidermis were also increased. Although, the expression levels of NGF did not show a meaningful change through the immobilization periods, other changes became remarkable, depending on the period of immobilization.

Conclusion: Our results suggest that changes in peripheral nerves, NGF, TRPV1 and P2X₃ in skin might be partly responsible for immobilization-induced pain.

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Abstract – WCN 2013

No: 934

Topic: 9 – Pain

Three dimensional computerized mobilization of the cervical spine for the treatment of chronic neck pain and associated headache

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Background: Physical therapy has been successfully used for the treatment of chronic neck pain (NP).

Objective: Investigate the safety and efficacy of computerized mobilization of the cervical spine in 3-dimensional space for the treatment of chronic NP.

Methods: Pilot, open, non-controlled, trial utilizing the *Occiflex* device (Headway Ltd. Misgav, Israel). We recruited patients with chronic NP due to facet joint disorder, myofascial pain syndrome, and whiplash injury. We excluded patients with myelopathy or radiculopathy. Treatment sessions lasted 20 min, twice a week, for 6 weeks. We evaluated patients with weekly visual analog score (VAS), neck disability index (NDI), neck range of motion (CROM), and joint position error (JPE).

Results: We recruited 7 women and two men, with a mean age of 50.5 ± 13.5 years. Treatment was not associated with any significant adverse effects. Comparing baseline to the last week of the trial (6), pain scores dropped by 2.9 VAS points ($p < 0.01$); CROM increased by 11% on the average of the 6 combined movements ($p = 0.01$); NDI showed significant reduction ($p = 0.03$); JPE significantly decreased at the end of the study ($p < 0.05$). Five out of 7 patients with concomitant headache (tension type headache, migraine, and cervicogenic headache) reported that their headache improved during the trial and 2 weeks following treatment completion.

Conclusions: Preliminary results of this trial show that computerized mobilization is safe and potentially effective for the treatment of chronic neck pain and associated headache. The co-occurrence of neck pain and headache and their response to treatment, suggests shared pathophysiological mechanisms.

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Abstract – WCN 2013

No: 871

Topic: 9 – Pain

Dose-dependent paclitaxel-induced neuropathy in mice

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Paclitaxel is a common anti-neoplastic drug, with benefits counter-balanced by painful sensory neuropathy, resulting in dosage limitation. Animal models are used in research for drugs that can minimise this type of neurotoxicity; however, literature quotes several doses that can be administered in studies involving mice, with no consensus regarding the ideal dose.

The objective of this study was to assess mechanical and thermal allodynia after administering different doses of paclitaxel in a mouse model.

Twenty-four BALB/c mice received i.p. injections with paclitaxel/saline (mg/kg b.w.) for seven consecutive days as follows: group P1 (1 mg), group P2 (2 mg), group P5 (5 mg) and group C (control). The animals' mechanical and thermal thresholds were assessed. Experimental data were compared with baseline and with control group. Statistical analysis was performed.

In group P1, thermal hyperalgesia appeared after the second dose ($p = 0.02$), but was short-lasting. There were no differences in mechanical thresholds throughout the experiment. In group P2, both thermal and mechanical persistent hyperalgesia appeared around the 16th day of the experiment ($p = 0.03$, $p = 0.001$). In group P5, both thermal ($p = 0.005$) and mechanical persistent hyperalgesia appeared around the 7th day post-administration.

Results indicate that a 2 mg/kg b.w. dose is ideal for replicating paclitaxel-induced peripheral neuropathy in mice. A 1 mg/kg b.w. dose not only induced short-lasting modifications but also different time-frames in mechanical and thermal hyperalgesia, whereas the 5 mg/kg b.w. dose induced a decrease in the overall quality of the animals' life.

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Abstract – WCN 2013

No: 830

Topic: 9 – Pain

Analgesic efficacy of fulranumab in patients with painful diabetic peripheral neuropathy in a randomized, placebo-controlled, double-blind study

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Background: Nerve Growth Factor (NGF) antibodies offer a potential treatment option in diabetic peripheral neuropathy (DPN).

Objective: To evaluate analgesic efficacy of fulranumab in patients with painful DPN.

Patients and methods: Phase-2 study with a 12-week double-blind (DB) efficacy-, 40-week DB safety extension-, and 52-week open-label safety extension phase. Patients with moderate-to-severe pain, stratified by concomitant treatment for neuropathic pain (yes/no), randomly (3:2:2:3) received placebo or fulranumab (1 mg-, 3 mg-, or 10 mg; every 4 weeks [Q4wk] subcutaneously).

Results: Due to USFDA clinical hold, 77 of planned 200 patients were enrolled: 62 (81%) patients (55.8% men; 81.8% white; mean age 59 years) completed the DB efficacy phase. Change in 7-day average pain intensity score from baseline to week-12 (primary endpoint) showed a positive dose response ($p = 0.014$, one-sided); the pair-wise comparison between 10 mgQ4wk dose to placebo was significant (nominal $p = 0.04$, two-sided). Least square (LS) means differences (95% CI) in change from baseline in average pain intensity scores vs. placebo were: 1 mgQ4wk, 0.1 (−1.26; 1.36); 3 mgQ4wk, −0.6 (−1.98; 0.74); and 10 mgQ4wk, −1.2 (−2.44; −0.06). 30% responder rates at week-12 were: placebo, 21%; 1 mgQ4wk, 44%; 3 mgQ4wk, 43%; and 10 mgQ4wk, 61%. Secondary efficacy endpoints showed dose-

related improvement trends. Most common (≥ 5 patients) adverse events for total fulranumab-treated patients vs. placebo were: peripheral oedema (11% vs. 0%), arthralgia (11% vs. 13%), and diarrhoea (9% vs. 8%). No cases of joint replacement or deaths were reported.

Conclusions: Despite early study termination, fulranumab treatment resulted in dose-dependent efficacy and was generally tolerable.

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Abstract — WCN 2013

No: 1068

Topic: 9 — Pain

Pregabalin beneficial effects on sleep quality or health-related quality of life are poorly correlated with reduction on pain intensity

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Background: Pregabalin (PGB) has been shown to improve sleep quality and Health-related Quality of Life (HRQoL) as well as pain intensity in neuropathic patients.

Objectives: To explore the magnitude of the correlations between changes in pain intensity, sleep quality and HRQoL after PGB treatment.

Methods: 138 patients suffering from neuropathic pain of any origin and without an adequate response to analgesics received an 8-week treatment course of PGB in an open-labeled fashion. Pain intensity, sleep quality and HRQoL outcomes were evaluated at baseline and at week 8 by means of a numerical rating scale (NRS), the Pittsburgh Sleep Quality Index (PSQI) and the EuroQuol health-state visuoanalogic (EQ-5D VAS) score, respectively.

Results: At week 8, mean PGB dose was 166.7 ± 7.8 mg/day. Pain intensity NRS score, PSQI total score and EQ-5D VAS score were improved by $66.5 \pm 1.9\%$, $40.0 \pm 3.6\%$ and $26.4 \pm 4.7\%$ (all $p < 0.01$) respectively. Correlations between percent change from baseline in pain NRS score and PSQI total score or EQ-5D VAS scores were $r = 0.36$ ($p < 0.01$, $R^2 = 0.11$) and $r = -0.20$ ($p < 0.02$, $R^2 = 0.05$) respectively. A multivariate logistic regression analysis disclosed that PSQI score change below the median (i.e. a better outcome) was related to higher EQ-5D VAS score change (OR = 2.15 [95%CI = 1.09–4.25]) whereas pain intensity NRS score change below the median was not (1.58 [0.78–3.23]).

Conclusion: In our study PGB-related improvements in sleep quality and HRQoL were marginally related to reductions in pain intensity in neuropathic patients. Improvement in sleep quality was a significant predictor of better HRQoL whereas pain intensity reduction was not.

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Abstract — WCN 2013

No: 969

Topic: 9 — Pain

One year follow-up study on pain in patients with Parkinson Disease

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Background: Pain is recognized as a non-motor symptom in Parkinson Disease (PD). There is musculoskeletal, neuritic, radicular or dystonia-associated pain.

Patients and method: We enrolled in our study 72 patients (30 men and 42 women) with idiopathic PD. They were on stages 2 to 4 on Hoehn–Yahr Scale. They had stable dose of l-dopa 3 months prior to study (average dose 875 mg/day) and no other disease responsible for the pain. We assessed the intensity of the pain using the 5-Point Verbal Rating Scale (VRS-5), 6 Point Behavioral Rating Scale (BRS-6) and 11-Point Box Scale (BS-11). Most of our patients suffered from musculoskeletal pain and dystonia-associated pain. We slowly reduced total daily l-dopa dose from average dose of 875 mg/day to average dose of 785 mg and we concomitantly introduced dopamine agonist ropinirole extended release 2 to 16 mg/day (average dose 7.25 mg/day). The intensity of pain was assessed at the beginning of the study after 6 months and one year later. The results were analyzed by Student's Test.

Results: 12% of patients did not present pain, 42% of them had mild pain, 32% discomforting pain and 14% distressing pain. At the beginning of the study we obtained 4.2 BRS6 Score and 7.9 BS11 Score. One year later our PD patients showed 3 BRS6 Score and 4.2 BS 11 Score.

Conclusion: By adding dopamine agonist and reducing l-dopa daily dose the intensity of pain in PD patients can be significantly lowered. This fact leads to the improvement of the quality of PD patient's life.

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Abstract — WCN 2013

No: 832

Topic: 9 — Pain

Clinical characteristics of toxin in Japanese poisonous *Clitocybe acromelalga* mushroom

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Background: Erythromelalgia has been reported in cases of mushroom poisoning with *Clitocybe acromelalga* in Japan and South Korea, and *Clitocybe amoenolens* in France and Italy; however, the detailed clinical features have not been well described in the English-language literature. The responsible toxin remains undetermined, although acromelic acid isolated from both species is regarded as a candidate.

Objective: To find the clinical characteristics of the toxin in *C. acromelalga*.

Patients and methods: We analyzed the clinical course of three patients with the poisoning. Their ingested mushrooms were identified as *C. acromelalga*, with inspection by specialists and mass spectrometric determination detecting acromelic acid.

Results: Intervals between ingestion and onset were 1, 3, and 5 days, respectively. Allodynia was more severe in the fingers and toes, but redness and swelling were absent. Severe arthralgia was provoked by motion in all joints of the extremities, so much so that the patients could hardly move their limbs as if suffering from quadriplegia. Pain occurred also in the neck, auricles, tongue, and pharynx, causing dysphagia in two patients. Three weeks after onset, the allodynia disappeared, but multiple arthralgias persisted, resisting medications in the two patients. Scores on the 100-mm visual analog scale were 52 mm and 73 mm, respectively, 3 months after onset, and then decreased gradually over several months.

Conclusions: The toxin in *C. acromelalga* does not necessarily cause erythema. It can exert its effects several days after being ingested, and affect sensory nervous systems originating from various body parts, characteristically including joints.

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Abstract – WCN 2013**No: 968****Topic: 9 – Pain****Reduction of thalamic tremor with deep brain stimulation performed for post stroke chronic central pain**

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Background: Deep brain stimulation (DBS) for the treatment of medically intractable pain has been in use for nearly 60 years. This experience was supported by later observations that electrical stimulation of the thalamus or thalamotomy could alleviate or arrest limb tremor. Presently DBS of the sensory thalamus and the periventricular/peri-aqueductal grey area complex may be applied for treatment of intractable neuropathic pain syndrome.

Objective: To present the patient who underwent the DBS surgery for chronic central pain (with stimulation of the periventricular/peri-aqueductal grey matter and ventroposterolateral thalamic nucleus) and experienced alleviation of thalamic tremor.

Patients and methods: We present a patient who experienced ischemic stroke within the posterolateral part of left thalamus with subsequent severe burning pain localized in right upper limb, predominantly within the hand and thalamic tremor which occurred 4 months after stroke. After 2 years of ineffective treatment the patient was offered implantation of electrodes to the periventricular grey matter (PVG)/periaqueductal grey matter (PAG) as well as implantation an electrode to ventroposterolateral thalamic nucleus (VPL).

Results: Soon after starting simultaneous PAG/PVG and PVL stimulation we observed significant alleviation of the patient's thalamic tremor in the hand, which persisted over subsequent months. We discuss possible mechanism underlying tremor suppression in our patient, probably at the level of cerebellar outflow pathways.

Conclusions: The study highlights the fact that DBS provide more insight into the functional anatomy of the thalamus, which used to be available from animal studies only.

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Abstract – WCN 2013**No: 1305****Topic: 9 – Pain****Minimally invasive treatment of refractory low back pain caused by degenerative spine disease**

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Background: Puncture techniques—nucleoplasty and radio frequency denervation—have recently been widely used for the treatment of pain syndrome caused by intervertebral disc protrusion.

Objective: The purpose of the study was to assess the effectiveness of nucleoplasty and radio frequency denervation of intervertebral joints as advanced minimally invasive surgical techniques for alleviating pain in the case of intervertebral disc disease, spondylarthrosis and other vertebroneurologic diseases accompanying degenerative disc disease.

Materials and methods: All patients had a history of pain syndrome for not less than 3 months and were followed up after undergoing minimally-invasive procedures for a year. To assess the effectiveness the Visual Analogue Scale and Treatment Satisfaction Scale were used. 81 patients, aged between 30 and 57 years, underwent nucleoplasty

and radio frequency denervation. 70 patients of control group were treated therapeutically.

Results: No complications were noted. At a 12 month follow-up all operated patients showed the decrease of pain syndrome intensity according to the Visual Analogue Scale ($p < 0.0001$), and treatment satisfaction scores were higher compared to patients treated therapeutically ($p < 0.001$).

Conclusions: Minimally invasive electrosurgical techniques appeared to be safe and effective in the treatment of carefully selected patients suffering from low back and leg pains caused by a degenerative-dystrophic process. Nucleoplasty technique is more effective in younger patients. Procedures performed at several levels also result in marked clinical improvement. Primary advantages of these techniques were their safety and efficacy, minimal invasiveness and the absence of a peridural scar. They do not complicate further microsurgical intervention.

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Abstract – WCN 2013**No: 1386****Topic: 9 – Pain****Control of chronic pain by the ubiquitin proteasome system**

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Chronic pain is maintained by long-lasting neuroplastic changes in synapses and several proteins critical for synaptic plasticity are degraded by the ubiquitin–proteasome system (UPS). Here, we show that proteasome inhibitors prevented the development and reversed nerve injury-induced pain behavior. Proteasome inhibitors blocked mechanical allodynia and thermal hyperalgesia in three pain models although they did not modify responses to mechanical stimuli, but partially inhibited responses to thermal stimuli in control rats. The effects of proteasome inhibitors on chronic pain were apparently mediated through several cellular mechanisms indispensable for chronic pain, including those of dynorphin A release and postsynaptic actions, and of CGRP secretion. Levels of several UPS proteins were reduced in animals with neuropathic pain, suggesting that UPS downregulation, like effects of proteasome inhibitors, counteracts the development of chronic pain. The inhibitors did not produce marked or disabling motor disturbances at doses that were used to modify chronic pain. These results suggest that the UPS is a critical intracellular regulator of pathological pain, and that UPS-mediated protein degradation is required for maintenance of chronic pain. Furthermore, in adjuvant-induced arthritis and experimental osteoarthritis models the inhibition of proteasome reversed the pain behavior and also reduced the severity of arthritis and joint destruction. Our findings suggest that nontoxic proteasome inhibitors may represent a novel pharmacotherapy for chronic pain.

Ossipov et al. (2007) *J Neurosci* 27: 8226–37; Ahmed et al. (2010) *Arthritis Rheum* 62: 2160–9; Ahmed et al. (2012) *Pain* 153: 18–6.

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Abstract – WCN 2013**No: 1603****Topic: 9 – Pain****Effects of arterial hypertension on the electromyographic data in patients with injuries of nerves**

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Introduction: The structural changes in vasa nervorum in hypertensive rates are similar to the changes in vessels of the brain that can lead to ischemia of the peripheral nerves (Sabbatini M. et al., 1996).

Methods and materials: Thirty nine patients (31 men and 8 women, average age—48.1 ± 1.7) with trauma of nerve (in the period from 3 months to 1 year after the injury) and neuropathic pain have been surveyed: 23 patients with arterial hypertension (AH), 16 patients without AH. Nerve conduction study of injured nerves and neuropathic pain scale were assessed in all patients.

Results: Intensity of neuropathic pain in the group with AH was 33.0 ± 2.6, in the group without AH—51.0 ± 1.9 ($p < 0.01$). Amplitudes of M-response ($M \pm m$, mV) in patients with and without AH were as follows: n. musculocutaneus—2.1 ± 1.2 and 4.6 ± 1.0, n. suprascapularis—1.1 ± 0.4 and 1.3 ± 1.1, n. axilaris—1.7 ± 1.2 and 4.9 ± 1.2, n. medianus—1.5 ± 0.4 and 1.9 ± 0.5, n. ulnaris—1.9 ± 0.4 and 1.6 ± 0.5, n. radialis—1.4 ± 0.3 and 1.7 ± 0.3. Motor conduction velocities ($M \pm m$, m/s) in patients with and without AH were as follows: n. medianus—28.9 ± 2.8 and 38.7 ± 1.7, n. ulnaris—28.5 ± 2.3 and 36.5 ± 2.9, n. radialis—24.9 ± 4.3 and 39.4 ± 1.7. Motor conduction velocity was slower in the group of patients with AH ($p < 0.01$). The amplitude of M-response did not depend on AH ($p > 0.05$).

Conclusions: Our data suggests that arterial hypertension in patients with traumatic neuropathy can predispose for more severe nerve injuries and more intensive neuropathic pain.

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Abstract — WCN 2013

No: 1077

Topic: 9 — Pain

Dynamic evaluation of the cervical spine in 3T reveals position-dependent changes of pathology in patients with cervical pain syndrome

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Background: Neck pain syndromes account for major costs of medical treatment and non-productive time in developed countries. However, there is poor correlation between pain severity and morphological findings.

Objective: The aim of this study was to examine the cervical spine by means of 3T-MRI in dynamic positions. Pathologic changes were measured and correlated with a pain-questionnaire.

Patients and methods: MR-imaging was performed with a 3T-Scanner (TrioTim, Siemens, Germany). Dynamic examination of the cervical Spine involved neutral position, ante- and retroflexion. For assessment of pain and functional impairment the d-NPAD pain-questionnaire was performed. Statistic evaluation with SPSS.

Results: 50 patients with neck pain syndromes (25m/25f), age ranging from 23 to 77 years (average 47.8 ± 13.3 years) were included. Sagittal T2w-TSE-images in all three positions were obtained within 6:06 min. Processes leading to stenosis of the spinal canal were significantly pronounced in retroflexion. Most pathologic changes were present on the level of C6/7 and C7/Th1. There was no significant correlation of the morphological findings with the reported pain intensity or functional impairment.

Discussion: 3T-dynamic MRI-examination of the cervical spine helps to differentiate pathologic changes of the cervical spine. Bones and soft-tissue structures are depicted in excellent quality. Stenosing degenerative processes of cervical spine are pronounced in retroflexion.

Conclusion: Stenosing degenerative changes of the cervical spine are pronounced in retroflexion. The lack of correlation between dynamic

morphological changes and pain-questionnaire remains a problem of assessment of the complex disease of neck pain syndrome and makes further research necessary.

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Abstract — WCN 2013

No: 1667

Topic: 9 — Pain

Post-operative pain control for perineal surgeries: A comparative study of caudal ketorolac versus lignocaine block among Pakistani population

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Background: Post operative pain is a strong contributing factor towards morbidity and limited mortality. Correct plan management regarding pain control helps in avoiding excessive health expenditure thus reducing financial strain in addition to improving quality of life.

Aim: Comparison was done between the post operative analgesia provided by caudal Ketorolac versus Caudal Lignocaine after perineal surgeries like hemorrhoidectomy, anal fissurectomy and anal fistulectomy.

Methods: This study was conducted at the Anaesthesiology department IIMC-T, Railway Hospital, Rawalpindi, Pakistan over a period of 6 months. A total of 50 patients undergoing perineal surgeries under general anesthesia were studied by conventional sampling. These patients were given injection Nalbuphine 10 mg I/V peroperatively. They were divided into two study groups; A & B. Group A comprised of 25 patients who were given injection Lignocaine 1% through caudal block. Group B also comprised of 25 patients, received Ketorolac 10 mg through caudal route. Pain relief was assessed by using Pakistan Coin Pain Scale (PCS) and Visual Analogue Scale (VAS).

Results: It was observed that caudal lignocaine provided prolonged analgesia (135 ± 20 min versus 90 ± 12 min) postoperatively after perineal surgeries as compared to caudal Ketorolac. Patient mobility was affected less with Ketorolac (18% versus 39%). Bowel and bladder paralysis was not seen in group B patients.

Conclusions: This study shows that caudal lignocaine provides better analgesia, with fewer side effects and it is more economical as compared to Ketorolac since need of adjuvant pain killers was less frequent in this group. Early mobilization of patients and discharge from hospital were seen in Ketorolac group which required frequent adjuvant drugs i.e. injection Nalbuphine adding to the cost of treatment.

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Abstract — WCN 2013

No: 1789

Topic: 9 — Pain

Epileptic seizures of pain (algic seizures) in children

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Background: Predominantly epileptic patients don't feel pain during the seizures. But in some forms of epilepsy patients have special types of simple focal seizures so-called algic seizures accompanying with pain sensations, sometimes very severe.

Objective: The aim of the presented study was analyzing the forms of epilepsy with algic seizures, its etiology and localization of ictal (seizure) patterns on the EEG.

Patients and methods: Among the patients investigated and treated in the Psycho-Neurological Department №2 of RCCH at the period 2006–2012 years were revealed 15 children with algic seizures fixed by video-EEG monitoring.

Results: The group of patient (n = 15) consists of 9 girls and 6 boys (age range 11 month–14 years) suffered with algic seizures: somatosensory algic seizures—6 patient, ictal headache—5, somatosensory + headache—2, headache + vomiting + ictal amaurosis—1 and ictal abdominal pain—1 girl. The etiology of epilepsy was symptomatic in 10 cases (Kozhevnicov–Rasmussen encephalitis—2 cases, case of Kozhevnicov epilepsy, Sturge–Weber syndrome, dysembryoplastic neuroepithelial tumor (DNET), focal cortical dysplasia (FCD), post-traumatic epilepsy, hippocampal sclerosis and diffuse hypoxic-ischemic encephalopathy), cryptogenic in 3 cases and also in 3 patients—idiopathic (Panayiotopoulos syndrome—2, and migraine-epilepsy). Ictogenic brain regions were temporal in 4 cases, occipitoposteriotemporal—4, occipital—3, parieto-temporal—2 and parieto-central—also in 2 patients.

Conclusion: Epileptic seizures of pain are rare but not so casuistic condition. Epileptic cerebral mechanism of pain must be considered by neurologists and other clinicians. This group of patients needs specific therapy with antiepileptic drugs and sometimes in symptomatic cases with pharmacoresistance—epileptic surgery.

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Abstract – WCN 2013

No: 1735

Topic: 9 – Pain

Fibromyalgia: Another clinical feature of generalized pain syndrome?

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Background: Fibromyalgia (FM) is underestimated neurological disorder characterized by widespread pain on axial skeleton and body, with typical symmetrical tender points. Considered as a disease or syndrome, FM is estimated to affect 2–4% of the population, although the vast number of patients with FM is being diagnosed by 5th year of suffering.

Objective: Our aim was to reveal potential comorbidities in patients with FM.

Material and methods: 123 patients with confirmed diagnosis of FM, 70 females and 53 males, age 51 ± 15 years, were evaluated for comorbidities: Hypothyreosis, Tension Type Headaches (TTH), Chronic Fatigue Syndrome (CFS), Irritated Bowel Syndrome (IBS) and Hyperhomocysteinemia.

Results: CFS was found in 96 patients (78%), Hyperhomocysteinemia was found in 68 patients (55%), Hypothyreosis (including Hashimoto type) was found in 42 patients (34%), TTH was established in 82 patients (66%) and IBS in 15 (12%). Most of the patients indicated the serious impact of the pain in their quality of life and everyday activities.

Conclusion: All patients with FM need thoughtful evaluation for possible comorbidities. Although FM has distinct diagnostic criteria, most of the patients with FM were suffering of other discreet comorbidities, including thyroid dysfunction, elevated level of homocysteine and Tension Type Headaches, some had different combination of several. In our opinion, FM with those comorbidities must be considered as part of Generalized Pain Syndrome, and each patient with TTH must be additionally evaluated for FM also. In-time

diagnostics and clinical evaluation for other features of Generalized Pain Syndrome are paramount for successful management.

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Abstract – WCN 2013

No: 1896

Topic: 9 – Pain

Lower trunk brachial plexopathy by lung mass, demonstrated with ultrasound

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The brachial plexus supplies most of the upper extremity. Due to its large size and superficial location, it is vulnerable to trauma, and it can be influenced secondarily by neighboring structures affected by skeletal, pulmonary, or vascular disease. We report a case of brachial plexopathy caused by a lung mass, discovered by ultrasound. A 45-year old man had complained of right scapular area pain over 6 months, and right hand grasp power weakness developed. He had conservative care including physical therapy, injection and acupuncture, but his symptoms aggravated. To rule out cervical radiculopathy, he was referred to our department for electrodiagnostic test, and the test was performed at 5 months after the onset. The findings of the electrodiagnostic test were compatible with injury of lower trunk of brachial plexus. Ultrasound imaging study revealed collapsed right internal jugular vein by a remarkable mass. Shortly afterward, a head& neck angio (CE) 3D MDCT and MRI were done, and a lung tumor was found in the right lung apex and supraclavicular region, sized 4.5 times 4 cm. The lung mass caused the scapular pain and brachial plexopathy.

Ultrasound is an effective and established technique in musculoskeletal imaging, and by using ultrasound in brachial plexus imaging, we could access each individual components of the brachial plexus continuously by shifting the probe back and forth. Therefore, it will be helpful for physical medicine and rehabilitation specialists to be familiar with brachial plexus, using ultrasound.

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Abstract – WCN 2013

No: 1760

Topic: 9 – Pain

The results of anxiolytic treatment in patients with chronic vertebrogenic low back pain syndrome

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Background: Accompanying increase in levels of anxiety and depression in patients with chronic low back pain has negative influence on course of primary disease.

Objective: To study effects of anxiolytic therapy in patients with chronic vertebrogenic low back pain syndrome.

Material and methods: 30 patients with chronic vertebrogenic low back pain syndrome were divided onto 2 groups:

1st group received standard protocol treatment,
2nd—standard protocol treatment and Tenoten.

During the 60 day observation period patients had four visits (days 0, 7, 30, 60). On each visit patients underwent neurological examination;

Hamilton Anxiety Rate Scale (HARS) assessment; completed Brief Pain Inventory (BPI), Beck Depression Inventory (BDI-II) and SF-36 questionnaires.

Results: According to the results of HARS on screening 60% of patients had clinically significant anxiety level. BDI-II assessment revealed that 70% of patients suffer from depression of various severity. On day 60 visit a group of patients receiving Tenoten showed anxiety and depression level reduction (41.8% and 44.8% respectively), control group had increase of mentioned parameters. Influence of pain on quality of life (BPI results) was gradually decreasing during treatment period in group with anxiolytic therapy and reached 3.4 points (5.1–control group). Along with anxiety and depression levels reduction patients had improvement of SF-36 results.

Conclusion:

1. Patients with chronic vertebrogenic low back pain syndrome have increase of anxiety and depression levels which influence primary disease.
2. Anxiolytic therapy in patients with current pathology leads to reduction of pain influence and has positive effect on quality of life.

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Abstract – WCN 2013

No: 1989

Topic: 9 – Pain

Progesterone reduces pain-related behaviour in chronic constriction injury model of neuropathic pain

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Background: Neuropathic pain results from lesions or diseases affecting the somatosensory system and often responds poorly to typical analgesics. Several studies support the crucial role of neuroactive steroids in the modulation of pain. Progesterone and its derivatives as neurosteroids produce antinociception and contribute to sex-based differences in pain sensation. In addition, progesterone reduces neuronal damage and improves functional outcome in animal models of neurological disorders.

Objective: This project carried out to study the effect of progesterone on expression and development of hyperalgesia and allodynia.

Material and methods: The chronic constriction injury (CCI) method was performed on the sciatic nerve. Radiant heat was applied as thermal stimulation for heat hyperalgesia. The cold and mechanical stimulations were applied through acetone and von Frey filament. Progesterone (10 mg/kg i.p.) was administered daily on days 1–14 after surgery. Behavioural tests including allodynia and hyperalgesia phenomena were done on the rats before and 14 days after the surgery.

Results: The CCI model caused significant increase in the behavioural scores of cold and mechano-allodynia and heat hyperalgesia. The chronic administration of progesterone reduced the behavioural scores of neuropathic pain significantly but single injection of progesterone on 14th day post-surgery did not have any effect on behavioural scores of neuropathic pain.

Conclusion: Our data indicate that chronic administration of progesterone prevents the development of neuropathic pain but its acute injection does not change the expression of neuropathic pain. Our results suggest that progesterone could be considered as a new approach for management of neuropathic pain.

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Abstract – WCN 2013

No: 2013

Topic: 9 – Pain

The effect of lithium on painful diabetic neuropathy in streptozotocin-induced diabetic rats

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Background: Lithium is widely used in treatment of bipolar disorder as a mood stabilizer. Recently, increasing body of evidences report the neuroprotective effects of lithium against cell injuries caused by different noxious stimuli in cultured cells and animal model of neurodegenerative diseases.

Objective: The present study was conducted to determine the effect of acute and chronic administration of lithium on pain-related behavior in a rat model of peripheral diabetic neuropathy.

Material and methods: Animal studies were carried out on male Wistar rats. All experiments followed the guidelines on ethical standard for investigation of experimental pain in animals. Diabetic neuropathy was induced by a single injection of 60 mg/kg streptozotocin. From the seventh day after the induction of neuropathy, the neuropathic rats received lithium intraperitoneally (5, 10 and 15 mg/kg/day) for ten days. The behavioral scores of neuropathic pain were measured on the first day before the induction of neuropathy as control and before and after lithium administration. The radiant heat plantar and von Frey filament test were used for the assessment of heat hyperalgesia and mechanical allodynia as behavioral scores of neuropathic pain.

Results: Chronic administration of lithium decreased mechanical allodynia scores with dose of 10 and 15 mg/kg in diabetic rats significantly but the acute administration of lithium did not change the behavioral scores of neuropathic pain.

Conclusion: Our results suggest that lithium reduces the behavioral scores of diabetic neuropathic pain and can be considered as a new therapeutic potential for painful diabetic neuropathy.

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Abstract – WCN 2013

No: 2144

Topic: 9 – Pain

Grey matter changes in chronic pain patients with and without unilateral non-dermatomal sensory deficits (NDSDS)

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Background: About 20–40% of chronic pain patients have widespread sensory deficits often in hemisensory distribution ipsilateral to the site of pain, a phenomenon termed non-dermatomal sensory deficits (NDSDs). Patients with NDSDs show no pathological findings in standard investigations, but decreased activity in the sensory system has been found in PET and fMRI studies.

Objectives: Cerebral grey matter changes were investigated in chronic pain patients with and without NDSDs compared to healthy controls (HC) using voxel-based morphometry.

Patients and methods: High resolution structural MRIs of the brain were acquired in 25 chronic pain patients with unilateral NDSs, 23 chronic pain patients without NDSs (termed “pain-only”), and 29 HC. Mechanical detection thresholds (MDT) were determined in all patients in the face, dorsum of hand and foot.

Results: MDT was significantly lower ipsilaterally to pain only in NDSs, whereas “pain-only” patients had no significant side differences. In patients with NDSs grey matter increases were found in primary sensory cortex, parietal regions and thalamus, as well as in lateral temporal regions. “Pain-only” patients showed a grey matter increase in the posterior insula and less pronounced changes in sensorimotor cortex. In all patients, grey matter in the right middle and inferior temporal gyrus was negatively correlated with MDT in the face.

Conclusion: These findings suggest that the dysfunctional sensory processing in NDSs is associated with structural changes in the somatosensory system. Correlations of temporal cortex grey matter with sensory loss suggest a role of this region in altered sensory perception.

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Abstract – WCN 2013

No: 2217

Topic: 9 – Pain

Genetics and underlying mechanisms associated with hereditary sensory and autonomic neuropathies

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Hereditary sensory and autonomic neuropathies (HSAN) are a group of diseases characterized by a reduced sensitivity to pain, which often results in unnoticed burns and mutilation of the fingers and toes. It is not uncommon that affected individuals have to undergo amputations. HSANII are now classified according to their underlying genetic defect. We are studying two of the three forms of HSANII: HSANII-A (*WNK1/HSN2*) and HSANII-C (*KIF1A*). Both disorders are autosomal recessive and caused by mutations in an alternatively spliced exon of the gene they belong to. In HSANII-A the mutations are in an exon referred to *HSN2* of *WNK1* and in HSANII-C they are in an exon referred to as 25b. A conditional mouse model has been generated for HSANII-A and a second one is in preparation for HSANII-C. While these models should provide novel insights about these neuropathies, cellular and in vitro experiments are also conducted to understand the underlying pathogenic mechanisms at play. In parallel, we are also pursuing more genetic studies using an exome sequencing approach to identify other genes which could explain HSAN cases that are not caused by *WNK1/HSN2* or *KIF1A* (exon 25b).

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Abstract – WCN 2013

No: 2078

Topic: 9 – Pain

The effectiveness of Onabotulinum Toxin A injection in the treatment of severe shoulder pain

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Background: Shoulder pain has three common pain generators: rotator cuff muscles and tendons, bursa and shoulder capsule. Any one of these structures can become a pain generator if they are inflamed or injured. It can be hard to diagnose the pain generator exactly as the symptoms, with these pathologies, are typically very

similar to each other. Most of these pathologies will cause symptoms of pain, weakness, and loss of motion.

Objective: This study aims to evaluate the effectiveness of 200 U of Onabotulinum Toxin A (Botox®) injection in the management of severe shoulder pain.

Patients and methods: Trial was conducted over a period of 9 months at the International Medical Center, KSA. 20 patients with severe shoulder pain and moderate osteoarthritic/inflammatory changes were evaluated, randomly given 200 U of Botox® injection therapy or trigger point injection. Botox® was injected in the trapezius, deltoids, and rotator cuff muscles, as well as intra-articular, in all cases. Inclusive criteria: 8 males, 12 females; ages from 28 to 65, with mean age of 47 years. Exclusive criteria: patients who have infection (where Botox® will be injected), pregnant, patients with neurological disorder (e.g. Myasthenia Gravis), and medicines that may interact with Botox® (e.g. Quinidine).

Results: Average improvement of about 75% was achieved by patients who were treated with 200 U Botox®, and sustained for 8 months.

Conclusion: Botox® injection in 200 U dose is more helpful in addressing severe chronic shoulder pain patients than trigger point injection, and also in establishing long sustained efficacy.

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Abstract – WCN 2013

No: 2158

Topic: 9 – Pain

Topical high-concentration menthol—Reproducibility of a human surrogate model of Neuropathic Pain

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Background: The topical application of high-concentration menthol is used as a surrogate human model to study cold and mechanical hyperalgesia. In this study, the reproducibility of this pain model as a prerequisite for pharmacological trials was investigated.

Methods: High-concentration (40%) menthol was topically applied to ten healthy volunteers' left and right ventral forearm at intervals of four hours in a randomly assigned sequence. The procedure was repeated in a second study period one week later. Selected quantitative sensory testing (QST) parameters were assessed before and after menthol application using the standardized protocol of the German Research Network on Neuropathic Pain (DFNS). Agreement between variables was investigated using the paired t-test, reliability was investigated by intra-class correlation and correlation analysis was performed by using Pearson's product-moment correlation.

Results: A significant reduction of cold pain (CPT) and increase of mechanical pain sensitivity (MPS) occurred after the application of menthol, reflecting cold and mechanical (pinprick) hyperalgesia ($p < 0.001$ resp.). High correlation coefficients ($r = 0.96/0.89$ (CPT); $0.93/0.87$ (MPS)), high reliability and no significant differences (t-test; $p = 0.21-0.74$) were assessed by correlation analysis.

Conclusions: The study demonstrated a high reproducibility of the menthol surrogate pain model within the period of one week. This indicates a suitability for the investigation of the induced symptoms of cold and mechanical hyperalgesia, e.g. in pharmacological trials.

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Abstract – WCN 2013**No: 1937****Topic: 9 – Pain****FabryScan – A screening tool for Fabry disease**

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Background: Fabry disease is an X-linked lipid storage disorder with early onset, morbidity and mortality. As enzyme replacement therapy is available, early detection of Fabry disease is important.

Objective: Our aim was to develop and validate an easy to use questionnaire and a set of bedside-tests distinguishing between Fabry disease and most frequent differential diagnosis.

Methods: A questionnaire inquiring 15 relevant clinical, mainly pain related, characteristics of Fabry disease and three bed-side tests assessing sensory small and large fiber function (temperature discrimination, sensation of light touch, pallesthesia) were compiled by Fabry specialists and pain experts. The provisional version was tested in a prospective multicenter trial with 138 patients suffering from chronic extremity pain due to Fabry disease (n = 55), painful polyneuropathy (n = 40), or rheumatoid arthritis (n = 43).

Results: For all three bed-site test parameters the chi-square test showed statistically significant differences between Fabry patients and controls (p < 0.001). Analysis of variance (ANOVA) showed statistically significant differences for 10 items of the questionnaire between Fabry patients and controls (p < 0.05). Only these 10 questions and the bed-side tests were used for calculation of sensitivity and specificity by multivariate analysis. A high proportion of correctly identified patients (76%) with a sensitivity of 88% and a specificity of 87% was achieved with a cut-off score of 12/33 (corresponding to the number of positive points).

Conclusion: Combining a brief questionnaire and three simple bed-side tests the FabryScan is a screening tool for Fabry disease with good discriminative value.

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Abstract – WCN 2013**No: 2175****Topic: 9 – Pain****The efficacy of using ozone injection for chronic sacroiliac pain**

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Background: Sacroiliac (SI) joint, located between the sacrum (tailbone) and ilium (hip bone) in the pelvis, is a common, but frequently overlooked source of low back pain. Pain which is felt at the very bottom of the lower back, described as a band of pain across the whole width of the back is often caused by sacroiliac dysfunction. Although the concept of SIJ causing lower back pain is now better understood, evaluation and treatment of sacroiliac dysfunctions are

still debatable because of its complex anatomy and movement patterns at the joints and area in general.

Objective: The purpose of the study is to evaluate the effectiveness of using sacroiliac ozone injection in the treatment of chronic sacroiliac pain.

Patients and methods: 60 patients with chronic and radicular pain were evaluated at the Pain & Headache Center, International Medical Center, KSA, and were diagnosed clinically and by imaging, as chronic sacroiliac pain. They all have received sacroiliac injection with ozone 20 cm³, 30%. Inclusive criteria: 38 males, 22 females, ages ranging from 44 to 82; mean age is 63. Exclusive criteria: pregnant women, children, anyone who is allergic to any of the medication ingredients, history of low blood pressure, and patients who have liver or kidney disease.

Results: Average improvement of 68% was appreciated, as per numeric pain scale, within one week and sustained for at least 6 months, with no evidence of major side effects.

Conclusion: Sacroiliac ozone injection helps significantly in the management of chronic sacroiliac pain.

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Abstract – WCN 2013**No: 2350****Topic: 9 – Pain****Spinal cord stimulation for neuropathic pain through an open approach without preliminary test: A clinical series**

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Background: Spinal cord stimulation (SCS) for intractable neuropathic pain is, nowadays, a well-established method of treatment. Strict indications, thorough clinical-imaging-electrophysiological examination and psychological assessment are the best prognostic factors. In this setting, careful surgical placement of multi-polar paddle-shaped leads warrants excellent results even without preliminary testing.

Objective: To demonstrate the efficiency of SCS through a minimally invasive approach without need of preliminary test.

Material and methods: 26 patients aged 41–73 were treated for neuropathic pain due to Failed Spine Surgery Syndrome (back and neck), peripheral nerve lesion and post-herpetic neuralgia refractory to medical treatment. All patients were clinically assessed for predominantly radicular or neuralgic components of pain and submitted to proper radiological imaging for exclusion of residual compression, spinal instability and medullary lesions. Previous responses to gabapentin treatment and/or improvement after TENS were considered mandatory prerequisites. Furthermore, electrophysiological studies (SSEPs) were performed in spinal cases to confirm the integrity of dorsal column fibers and psychological consultation in certain patients. All patients underwent placement of a multi-electrode (2 × 8 or 2 × 4) paddle lead through an open interlaminar approach with intraoperative radiological confirmation of proper position.

Results: 19 patients confirmed >50% pain reduction after a mean follow-up of 2 years with concomitant medication reduction, 24 patients declared satisfied with the procedure and no post-operational complications were recorded.

Conclusion: Based on current bibliographic data and modern technological advances, we assume that after careful patient selection SCS through an open approach is a minimally invasive procedure which guarantees excellent results even without preliminary percutaneous test.

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Abstract – WCN 2013**No: 2366****Topic: 9 – Pain****Diagnostic value of various blink reflex parameters in idiopathic trigeminal neuralgia**I. Mikula. *Neurology, Medikol Polyclinic, Zagreb, Croatia*

We have done a study investigating the value of some less frequently considered Blink reflex parameters for establishing the diagnosis of idiopathic trigeminal neuralgia. The study was done on 50 patients suffering from idiopathic trigeminal neuralgia, diagnosed according to the guidelines of the International Classification of Headache Disorders, with no other apparent illness. We have stimulated the supraorbital nerve at the forehead (foramen n. supraorbitalis) and recorded the reflex response on both mm. orbiculares oculi. Incidence of following findings was determined:

- (1) occurrence of ipsilateral R3 component,
- (2) prolonged duration (>25 ms) of R2 when stimulating the affected side and
- (3) occurrence of R1 component during the stimulation of contralateral supraorbital nerve.

We have compared these findings to those of 50 healthy subjects from the control group (Chi square, $p < 0.05$). Sensitivity, specificity and diagnostic value for individual parameters were determined. All three parameters tested proved to have a significantly higher incidence in the group of subjects. The occurrence of R3 component on the affected side showed the highest diagnostic value.

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Abstract – WCN 2013**No: 2424****Topic: 9 – Pain****Manipulative therapy improves sensorimotor integration in patients with neck pain in combination with thoracic pain**V. Zabarovski, L. Anatskaia, T. Svinkovskaya. *Neurology, Republican Research and Practical Center of Neurology and Neurosurgery, Minsk, Belarus*

Background: Neck and thoracic pain can cause the cortical neuroplasticity changes. Recent research suggests that these changes may be reduced with cervical spinal manipulation and motor-skill training.

Objective: The objective of this study is to evaluate the sensorimotor neurophysiological effects of cervical and thoracic spine manipulations and training therapy course using somatosensory evoked potentials (SEPs) in patients with subacute neck pain in combination with thoracic pain due to cervical or thoracic movement dysfunction.

Patients and methods: Twenty-nine patients (15 women and 14 men aged 35 to 49) with subacute neck pain in combination with thoracic pain due to movement dysfunction were studied clinically. The SEP values of the N9, N13, N20, N9–N13, and N9–N20 were obtained before the first and after the last session of cervical and thoracic spine manipulations and training therapy.

Results: There was a decrease in latency and amplitude of the spinal N13 and the parietal N20 and interpeak values of the N9–N13 and N9–N20 after the last treatment session compared to pre-manipulative SEP values, $p < 0.001$.

Conclusion: Course of cervical and thoracic spine manipulations and training therapy in patients with neck pain in combination with thoracic pain due to cervical or thoracic movement dysfunction not

only helped reduce pain and restore function, but also improved sensorimotor integration at the spinal and parietal cortical levels thus preventing pain chronicity.

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Abstract – WCN 2013**No: 2357****Topic: 9 – Pain****Evaluation of sensory and pain perception and mechanisms of central modulation of pain perception in chronic low back pain**E. Vlckova^{a,b}, I. Okacova^{a,b}, R. Kopacik^{a,b}, M. Hnojckikova^{a,b}, P. Praksova^{a,b}, B. Micankova-Adamova^{a,b}, J. Bednarik^{a,b}. ^aCentral European Institute of Technology, Masaryk University, Brno, Czech Republic, ^bDepartment of Neurology, Masaryk University and University Hospital Brno, Brno, Czech Republic

Background: Chronic low back pain (LBP) is one of the most common pain conditions. The pain intensity in LBP patients is frequently unrelated to degenerative changes of lumbar spinal cord and possible role of abnormal central processing of pain perception may be hypothesized.

Objective: To evaluate the sensory profile and function of central modulation of pain perception in order to reveal underlying mechanisms of chronic LBP, which are still not fully understood.

Patients and methods: A detailed evaluation of sensory and pain perception and of mechanisms of central modulation of pain perception was performed in 28 patients with chronic LBP (16 men, median age 35 years, no symptoms or signs or risk factors for polyneuropathy, no nerve root lesions) and 22 age-matched healthy volunteers (10 men, median age 29 years). Among others, magnitude of conditioned pain modulation (CPM) and temporal summation (TS) using thermal stimuli were assessed.

Results: Seventy-one percent of LBP patients (and 14% of controls) showed at least one abnormality of thermal perception in lower extremities (elevated thermal perception thresholds or decreased thermal-pain thresholds or most frequently so called “paradoxical heat sensation”, i.e., cold stimuli perceived as a burning hot painful sensation). The differences in CPM and TS between the groups were not significant.

Conclusion: Frequent abnormalities of thermal or thermal-pain perception were found in LBP patients compared to controls, while no significant effect of abnormal CPM or TS on the LBP development was disclosed.

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Abstract – WCN 2013**No: 2467****Topic: 9 – Pain****Application of botulinum toxin type A for piriformis syndrome treatment**G.L. Vasques^a, J.P.R.P.T. de Azevedo^a, I.B. Arruda^b, M.V.B. Sasaki^b, F.O. Gomes^b, D.J. da Silva^a. ^aHospital das Clínicas, Universidade Federal de Goiás, Goiânia, Brazil, ^bFaculdade de Medicina, Universidade Federal de Goiás, Goiânia, Brazil

Background: The piriformis syndrome (PS) is an important cause of pain in gluteal region, often accompanied by sciatica. It is currently described as a form of entrapment of the sciatic nerve in the piriformis muscle topography, causing pain from the buttock to the radicular distribution of the nerve. There are no specific diagnostic

tests for this syndrome, becoming an underdiagnosed disease. Traditional treatments include the use of anti-inflammatories, corticosteroids, opioids, local anesthetic injections and, in extreme cases, surgery. Treatment with botulinum toxin is not common, but has proven effective.

Objective: This paper reports a diagnosed case of SP treated with botulinum toxin.

Patient and methods: Patient of 25 years old, female, started a progressive growing pain in the right buttock, not associated with trauma, radiating to the ipsilateral member and disabling evolution. The pain symptoms worsened and were refractory to common analgesics, NSAIDs and opioids. The patient was unable to walk due to the movement restriction of the affected limb. She presented positive Lasègue, Freiberg and Pace sign, also pain in the path of the sciatic nerve. Lumbar MRI and buttock US were normal. The treatment began with sessions of botulinum toxin type A injection in the topography of the piriformis muscle, at intervals of 3 months between sessions, concomitant with physiotherapy sessions.

Results: After the first session, a reduction of pain and the full mobility regain of the right leg were observed.

Conclusion: The application of botulinum toxin is an excellent therapy for PS and its use is becoming more frequent.

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Abstract – WCN 2013

No: 2521

Topic: 9 – Pain

The role of sodium channel Nav1.9 in the activation of visceral afferents by histamine and its contribution to IBD

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Introduction: The aim of this study was to investigate the contribution of the sodium channel Nav1.9 to the afferent sensitivity to histamine in the small intestine of mice. Such study could elucidate a potential role of Nav1.9 in the visceral hypersensitivity that has been linked to pain in inflammatory bowel disorders and irritable bowel syndrome.

Materials and methods: An in vitro preparation was used to make electrophysiological whole-nerve recordings of multiunit activity in mesenteric afferents supplying segments of the small intestine in Nav1.9 wild-type (WT), heterozygote (hets) and knock out (KO) mice.

Results: In WT preparations, histamine elicited a dose-dependent increase in afferent nerve discharge that was typically biphasic following higher doses of the mediator. Histamine perfused into the tissue bath evoked a robust, and in some cases a biphasic increase in afferent nerve discharge activity. The afferent responses to histamine in hets and KO mice were not significantly different ($p > 0.05$) from that of WT mice. Although the preliminary findings suggest that Nav1.9 may not have a role in mediating the afferent responses to histamine, further experiments that will utilize higher n values with more thorough investigations are needed.

Conclusion: Future experiments will look to establish a more superior analysis of the histamine response profile in WT mice in order to carry out an adequate, multifaceted comparison with the response in Nav1.9 deficient mice. In the anticipation of such experiments, the contribution of Nav1.9 cannot be as yet ruled out entirely.

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Abstract – WCN 2013

No: 2604

Topic: 9 – Pain

Clinical and neurophysiological analysis of pain in vibration-induced hand disorders

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For patients with Hand-Arm Vibration Syndrome (HAVS) the most common complaints are pain, tingling or numbness of fingers and heightened sensibility to cold or fingers of hands albication. The perception of pain is always subjective. The research of the character and the neurophysiological mechanisms of pain syndrome, including occupational diseases, is an actual problem nowadays.

The aim of the present study was to investigate correlation between electrophysiological methods and pain questionnaires.

26 working miners (48.3 ± 0.9 years old) with upper limb disorders filled in the screening questionnaires for neuropathic pain: DN4 and Pain Detect (PD) and were examined with electroneuromyography (ENMG), quantitative sensory testing (QST).

High values of DN4 and PD point to a high probability of presence of neuropathic pain component. The results of ENMG which detects symptoms of axonal-demyelization process (polyneuropathy) and QST (changing of temperature thresholds of four submodalities and the vibration sensitivity) indicates the presence of signs of injury somatosensory nervous system.

Statistically significant Spearman correlation was revealed between questionnaires VAS and PD; questionnaires (PD) and QST (CS) or (WS); questionnaires (DN 4) and ENMG (SNAP). Between ENMG (sensory NCV) and QST (WS) $r = -0.719^*$, $p = 0.045$; ENMG (sensory NCV) and QST (VT) $r = -0.409^*$, $p = 0.038$; ENMG (SNAP) and QST (CP) $r = -0.400^*$, $p = 0.043$; ENMG (SNAP) and QST (HP) $r = -0.401^*$, $p = 0.042$.

During the analysis of QST between ENMG data we have received correlation dependencies. It may indicate injury of C-warm-fiber, C- or A-delta nociceptors and A-beta fibers. Complex usage of pain questionnaires and QST can contribute to optimization of therapeutic tactics.

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Abstract – WCN 2013

No: 2271

Topic: 9 – Pain

Pattern of abnormalities of quantitative sudomotor axon reflex test in complex regional pain syndrome

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Background: Complex regional pain syndrome (CRPS) is complex to diagnose, since it is based on the clinical symptoms and signs without specific laboratory or radiologic findings. Though, quantitative sudomotor axon reflex test (QSART) is introduced to examine the sweating disturbance objectively, its pattern and characteristics in the CRPS patients have been seldom reported. We aimed to know if there was any correlation with the severity of QSART abnormality and disease duration.

Methods: We analyzed the records of the CRPS patients who met the Budapest criteria and underwent QSART during a year period. QSART was regarded abnormal when total sweat volume was reduced or it showed ultrashort latency or persistent sweat activity. We scaled the CRPS grade from 0 to 3 by the number of limbs showing abnormal QSART findings.

Results: Forty-nine CRPS patients were included in this study. Thirty of them were male and median age was 44 (from the age of 20 to 72 years). Median duration of symptom was 16 months, ranged from 2 months to 120 months. All the patients showed abnormal QSART findings regardless of the clinical symptoms and disease duration. Most common abnormality was absent or reduced sweat volume. There was no correlation between QSART abnormal degree and morbidity period or age.

Conclusion: QSART abnormal degree, referred to as the spatial progression of sudomotor dysfunction, did not reflect the morbidity duration or severity of CRPS. QSART alone should not be interpreted as an objective diagnostic tool for accessing the severity and disease progression of the CRPS.

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Abstract – WCN 2013

No: 2592

Topic: 9 – Pain

Involvement of the middle short gyrus of the insula in pain processing: Intracerebral electric stimulation of epileptic patients

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Objectives: The data of this study suggests the involvement of the upper middle short gyrus in the procedures of pain.

Materials and methods: For this study, we included 25 patients suffering from severe drug refractory partial epilepsy investigated by stereo-electroencephalography (SEEG). At least one electrode was used to explore the insular cortex using an oblique approach (trans-frontal or trans-parietal). 313 stimulations were performed in 27 insula. 83 responses induced by insular electrical stimulation, eight (9.6%) were reported by five patients as painful sensations. The stereotactic approach allows us to identify the stimulation sites within the insular cortex in terms of its gyri and sulci. Also, the stimulation sites were anatomically localized via image fusion between pre-implantation 3D MRI and post-implantation 3D CT scans revealing the electrode contacts.

Results: We could obtain pain responses to direct ES of a small anatomical region. We discuss our results in terms of anatomical localization in gyral substructures of the insular cortex. The findings suggest that middle short gyrus is involved in the processing of pain producing stimuli. These sensations were evoked ipsilaterally or bilaterally to stimulation. The data provide evidence of a highly restricted area of the insular cortex inducing painful sensation in response to direct ES. The region of the head is more posterior than the trunk and limbs.

Conclusion: Our results are the first to described painful responses evoked by electrical stimulation of the human insular cortex classified in terms of its gyri and sulci.

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Abstract – WCN 2013

No: 1267

Topic: 9 – Pain

A case with generalized anhidrosis and limb pain

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A forty one yr old man has been suffering cholinergic urticaria. He has been treated with systemic steroid. But this symptom didn't recovered fully. 2 months later, he also noted some change of sense and severe lancinating pain in both legs with loss of sweating. In dermatologic evaluation, face showed dry erythematous patches, and in both axillae and periumbilical area, erythematous patches noted. Under the impression polyneuropathy, we have studied large and small fibers with nerve conduction study (NCS), sympathetic skin response (SSR), heart rate variability test with deep breathing, Valsalva maneuver (HRT), Quantitative axon reflex test (QSART) and thermoregulatory sweat test (TST). Blood chemistry, serologic evaluations, and skeletal surveys were done.

The skeletal survey, blood chemistry (glucose, HbA1c, liver, kidney function tests), serologic evaluations, and α -galactosidase were normal. IgE elevation was noted without significance.

NCS with F waves was normal but SSR showed reduced amplitude in hands and absent response in feet. HRTs were normal. There were generalized loss of sweating on whole body area, and small sweating was noted on both toes and fingers only in TST. QSART revealed generalized absence of response to acetylcholine, compatible with postganglionic sympathetic dysfunction.

For evaluation on the loss of sweat skin biopsy was done on upper back (non-sweating area) and scalp (sweating area). Biopsy result was perivascular and pterygoid ductal mild lymphocytic infiltration without significance.

He has been diagnosed with non-length dependent pattern, small fiber neuropathy clinically AIGA.

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Abstract – WCN 2013

No: 2853

Topic: 9 – Pain

Single versus repetitive injection of lidocaine HCl in management of carpal tunnel syndrome

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Objective: The aim of the present study is to determine the efficacy of single versus repetitive injection of lidocaine HCl into carpal tunnel in management of carpal tunnel syndrome (CTS).

Methods: This prospective, randomized trial included 42 patients (42 median nerves) with clinical and electrophysiological evidence of CTS. The 42 patients were randomly assigned to 1 of 2 groups: group 1 was injected with 4 cm³ 1% lidocaine HCl for once and group 2 was injected 4 cm³ 1% lidocaine HCl twice a week for two weeks. Clinical and electrophysiological evaluations were performed at the study onset, 6 and 12 weeks post treatment.

Results: At the study onset significant differences were not observed between the groups with respect to DML (distal motor latency), CMAP (compound motor action potential) amplitude, CSAP (compound sensory action potential) amplitude, SNCV (sensory nerve conduction velocity), and VAS (visual analog scale); however all the parameters in group 2 improved 6 weeks after treatment (all P < 0.05), and these parameters persisted at 12 weeks after treatment (all P < 0.05). VAS scores improved significantly in group 1 at 6 weeks after treatment and this improvement persisted at 12 weeks after treatment. Group 2 had better scores in CMAP amplitude and VAS at 6 and 12 weeks after treatment (all P < 0.05).

Conclusion: Repetitive local lidocaine HCl injection was effective in reducing the symptoms of CTS and improving electrophysiologic findings.

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Abstract – WCN 2013

No: 2902

Topic: 9 – Pain

Modulation of oxido-nitrosative stress induced neuro-inflammatory cascade & monoaminergic pathway by resveratrol in reserpine induced pain depression dyad in rats

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Background: The emerging concept for pain-depression pathogenesis includes dysfunction of biogenic amine-mediated CNS pain control and the possible involvement of oxido-nitrosative stress-induced neurogenic inflammation. Thus the aim of the present study was to investigate the effect of resveratrol on the basis of its monoamine replenisher and antioxidant potential in reserpine-induced pain-depression in rats.

Methods and results: Administration of reserpine (1 mg/kg; sc) for three consecutive days in male Wistar rats led to a significant decrease in nociceptive threshold as evident from reduced paw withdrawal threshold in Randall Sellitto and von-Frey hair test and a marked increase in immobility time. This behavioral deficit was integrated with decrease in the biogenic amine (dopamine, norepinephrine and serotonin) levels along with increased substance-P concentration, nerve growth factor, oxido-nitrosative stress, inflammatory cytokines (TNF- α & IL-1 β), NF- κ B and caspase-3 levels in different brain regions of the reserpinised rats.

Conclusion: These results reveal that common mechanisms appear plausible as co-activating factors which result in neurophysiological overlap between pain and depression. Furthermore, the study demonstrates the effectiveness of resveratrol in ameliorating the behavioral deficits associated with pain and depression by restoring behavioral, biochemical, neurochemical and molecular alterations against reserpine-induced pain depression dyad.

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Abstract – WCN 2013

No: 3020

Topic: 9 – Pain

Radiotherapy in symptomatic vertebral hemangiomas

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Objective: Vertebral hemangiomas are common benign vascular lesions. They are often asymptomatic. Pain is the most common symptom. Treatment options are: surgery, intralesional injection of alcohol, vertebroplasty with methyl methacrylate, embolisation, and radiotherapy. We present the results of radiotherapy in symptomatic vertebral hemangiomas treated in our clinic.

Methods: 34 patients (24 female, 10 male; mean age 52; range 30 to 65 years) with symptomatic vertebral hemangiomas were treated with external radiotherapy at a dose of 24–40 Gy. All patients had pain and one patient had paraplegia at presentation. Results in terms of pain and pain relief were assessed before and after therapy and during follow-up.

Results: After radiotherapy, 6 patients had less than 50% response to treatment, 10 patients had more than 50%, and 18 patients had a complete response with a median follow-up of 48 months.

Conclusion: Radiotherapy is a non-invasive, safe, and effective treatment option for symptomatic vertebral hemangiomas, especially those cases in which pain is the main symptom.

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Abstract – WCN 2013

No: 2922

Topic: 9 – Pain

Leukocytoclastic vasculitis due to duloxetine

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We report on a 54-year-old woman, previously in good health, who had syncope, loss of consciousness and neuropathic pain hospitalized for one day and discharged with pregabalin 150 mg/day and duloxetine 60 mg/day. After 2 weeks of this treatment, she developed multiple palpable purpura lesions on her feet and exacerbated in 5 days in lower extremities. The skin biopsy revealed leukocytoclastic vasculitis and direct immunofluorescent was negative for fibrinogen, IgA and C3. No other organ involvement was diagnosed and ANA, ANCA, Anti-CCP were negative. Duloxetine treatment was stopped and she was started on oral prednisone 30 mg daily. In 3 weeks she had resolution of the vasculitic skin lesions. Pregabalin is still the only treatment for today and the patient has no skin lesions in one year follow-up after duloxetine treatment.

According to our knowledge, the case that we presented is the first report of leukocytoclastic vasculitis due to duloxetine and neurologists should be aware of this rare but potentially serious, possible adverse effect of duloxetine.

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Abstract – WCN 2013

No: 2935

Topic: 9 – Pain

Long-term prognostic factors for microvascular decompression for trigeminal neuralgia

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The purpose of this retrospective study was to identify preoperative imaging characteristics and surgical findings that predict pain relief after microvascular decompression (MVD) for trigeminal neuralgia (TN). This study included 141 patients with follow-up ranging from 6 months to 10 years (mean follow-up = 26.3 months). Preoperative images were assessed in 90 patients who were evaluated with constructive interference in steady-state (CISS) MRI in the last 6 years. These findings were compared with the severity of neurovascular conflict (NVC) found at operation to identify imaging findings useful for prognosis. Using Kaplan–Meier analysis, we found that the success rate of MVD was 91.1 ± 2.5% at 1 year and was 76.3 ± 7.5% after 5 years. A higher degree of NVC at operation ($p = 0.000$), no vein compression ($p = 0.049$) and single vessel compression ($p = 0.000$) were good prognostic factors for pain relief. Two meaningful positive MRI findings, specifically, the “cerebrospinal fluid rim sign” and the “deviation sign” were statistically significantly associated with the severity of NVC at operation and MVD success ($p = 0.000$). In this study, 34 patients (24.1%) complained of facial numbness postoperatively, and the oral herpes simplex virus was

reactivated in 19 patients (13.4%). The involvement of a single arterial offender in NVC is the most important prognostic factor for MVD in TN, and the positive MRI findings described in this report may be helpful in selecting patients for MVD.

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Abstract – WCN 2013

No: 2844

Topic: 9 – Pain

Effects of transcranial magnetic stimulation in the treatment of phantom limb pain in landmine victims: A randomized clinical trial

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The rTMS on motor cortex (M1) has efficacy of 52–88% in some refractory neurogenic pain by descending modulation of corticothalamic tract. The aim is to evaluate the efficacy of rTMS in the treatment of PLP in victims of anti-personnel landmines.

We performed a randomized, double-blind placebo-controlled trial in 49 patients with unilateral amputation, 26 patients received real rTMS over the hand area of M1 contralateral to amputated leg and 23 patients received sham. A Visual Analogue Scale (VAS) was administered at baseline, after 15 and 30 days of last session.

There were no significant differences when the percentage change of VAS at 15 days (-53.2 ± 54.2 vs -28.2 ± 58.4 , $p = 0.11$) or at 30 days (-43.6 ± 48.6 vs -21.5 ± 53.4 , $p = 0.14$) was analyzed comparing both groups, however, a significant difference in the proportion of subjects who presented a decrease greater than 30% in VAS at 15 days in the active group vs placebo (69.2% vs 39.1%, $p = 0.03$) and a tendency at 30 days (58.3% vs 33.3%, $p = 0.09$) was observed. The analysis of a subgroup of subjects with less than 10 years since amputation ($n = 38$) showed differences between the percentage change at 15 days (-51.2 ± 59.5 vs 55.7 , $p = 0.05$), and at 30 days (-49.4 ± 48.9 vs -9.9 ± 46.7 , $p = 0.02$). There were clear differences in the proportion of subjects with a decrease in more than 30% of pain at 15 days (68.4 vs 31.6%, $p = 0.02$) and at 30 days (61.1 vs 23.5, $p = 0.02$).

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Abstract – WCN 2013

No: 3061

Topic: 9 – Pain

Effect of manipulative therapy in combination with kinesiio taping on cervicothoracic pain in highly qualified athletes

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Background: The most effective method for treatment of cervicothoracic pain in highly qualified athletes is manipulative therapy (MT). Kinesio taping (KIT) may be an additional treatment option to MT creating favorable conditions for the activation of local and regional microcirculation in the connective tissue, improving lymphatic flow, encouraging the tissue healing process.

Objective: The aim of the study was to determine the effects of MT in combination with KIT on neck and thoracic pain and skin

microhemodynamics (SMHD) in highly skilled athletes with cervicothoracic pain.

Patients and methods: 30 athletes with moderate cervicothoracic pain were treated with MT daily in combination with KIT by intervals of 3 days. Response to treatment was evaluated with a 100-mm visual analog scale and speckle-optical investigation of skin blood flow over a pair of antagonist muscles in a state of imbalance. SMHD was evaluated with average fluctuation frequency ($\langle F \rangle$), the spectral power (SP) and asymmetry factors (As).

Results: The statistically significant pain severity decrease was determined after the course of combined treatment of MT and KIT, $p < 0.05$. The SMHD values of $\langle F \rangle$ and SP were higher as compared with data before treatment by 11% ($p < 0.05$) and 27% ($p < 0.05$) respectively indicating the increase of skin blood flow velocity.

Conclusion: MT in combination with KIT has been found to be an effective treatment for highly skilled athletes with cervicothoracic pain due to local or regional perfusion improving by increasing blood flow and accelerated elimination of pain neurotransmitters and nociceptor stabilization.

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Abstract – WCN 2013

No: 3152

Topic: 9 – Pain

Botulinum toxin type (A) in severe diabetic neuropathy: Double blind cross study

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Background: Managing patients with painful diabetic neuropathy is a great challenge, and many lines of treatment are available but there are many patients that cannot tolerate large doses of these medications or not responding to them. Recent experimental evidence suggests that botulinum toxin type A may have a role in the management of neuropathic pain.

Methods: A total of 42 patients with severe painful diabetic neuropathy were enrolled in this study (22 patients injected with botulinum toxin and 20 patients injected with saline) and evaluated the effectiveness using visual analogue scale (VAS) for pain and Pittsburgh Sleep Quality Index (PSQI) for sleep quality.

Results: Patients show significant reduction in VAS at 1, 4 and 12 weeks post injection with botulinum toxin type (A) compared to patients injected with saline (first week .04, 4th week .001 and 12th week .000) ($p < .05$).

Conclusion: Botulinum toxin type A is effective in managing patients with severe diabetic neuropathic pain, but large studies are required for more confirmation.

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Abstract – WCN 2013

No: 3142

Topic: 9 – Pain

Pharmacokinetics of DS-5565, a novel $\alpha_2\delta$ ligand, in rats and monkeys and assessment of DDI risk

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DS-5565 is an oral analgesic drug that binds to the $\alpha_2\delta$ subunit of voltage-dependent Ca^{2+} channels. We elucidated the pharmacokinetic profiles of DS-5565 in animals and drug interaction risk as a perpetrator.

DS-5565 is the salt, and the free form of DS-5565 is active moiety. Plasma concentration of the active moiety was determined by a validated LC–MS/MS method. The profiles were investigated in F344 rats, streptozotocin-induced diabetic BN rats as the neuropathic pain model, and cynomolgus monkeys. Distribution was assessed by whole body autoradioluminography following an oral administration of ^{14}C -labeled active moiety in F344 rats.

The plasma exposure increased proportionally with the investigated dose in both strain and species. The bioavailability was higher than 85% in both species. The radioactivity was detected in most of the tissues at 30 min but was primarily detectable in limited organs at 24 h post-dose. The plasma protein binding in rats, monkeys and humans in vitro was low. A few metabolites, which have no pharmacological activity, were detected qualitatively at low levels in plasma in both after oral administration. The primary excretion route of the radioactivity was urine; $\geq 87\%$ of the dose was recovered within 7 days after an oral administration of the ^{14}C -labeled compound in rats and monkeys. DS-5565 did not induce CYP1A2/3A4 in human hepatocytes, and did not inhibit various CYP isoforms in human liver microsomes and drug transporters in overexpressing cells.

Pharmacokinetics of DS-5565 in animals was favorable. DS-5565 has low potential to be a perpetrator in drug–drug interaction.

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Abstract – WCN 2013

No: 3140

Topic: 9 – Pain

Pharmacological, pharmacokinetics and safety profiles of DS-5565, a novel $\alpha_2\delta$ ligand

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DS-5565 is an analgesic drug that binds to the $\alpha_2\delta$ subunit ($\alpha_2\delta$ -1 and $\alpha_2\delta$ -2) of voltage-dependent Ca^{2+} channels. The $\alpha_2\delta$ -1 is the main target for the analgesic effect of $\alpha_2\delta$ ligands. The contribution of the $\alpha_2\delta$ -2 to the CNS side effects of $\alpha_2\delta$ ligands still remains to be elucidated. To clarify the characteristics of DS-5565, we conducted the experiments using pregabalin (PGB) as a reference. The binding affinity and dissociation rate were investigated with rat $\alpha_2\delta$ -1 and $\alpha_2\delta$ -2 transfected cells. The analgesic effect was investigated with a von Frey test in streptozotocin (STZ)-induced diabetic rats. The CNS side effects were investigated with rota-rod performance (RR) and locomotor activity (LA) in rats. The plasma drug concentration was measured by LC–MS/MS. The binding affinities of DS-5565 for $\alpha_2\delta$ -1 and $\alpha_2\delta$ -2 were comparable to those of PGB. Interestingly DS-5565 showed a slower dissociation rate from $\alpha_2\delta$ -1 than $\alpha_2\delta$ -2, in particular $\alpha_2\delta$ -1 compared to PGB. DS-5565 showed potent and sustained analgesic effects and the ED_{50} was ca 2.5 mg/kg (ED_{50} for PGB: 29.3 mg/kg). The plasma concentration of DS-5565 in the STZ rats, however, was about 65-fold less than PGB. DS-5565 inhibited RR (ID_{50} : 9.4 mg/kg) and LA (ID_{50} : 43.9 mg/kg) and the ratios $\text{ID}_{50}/\text{ED}_{50}$ (CNS safety margin) were ca 3.8 in RR and ca 18 in LA. The ratios for PGB were 0.4 and 3.9, respectively. DS-5565 has superior analgesic effects with a wider CNS safety margin relative to

PGB. These profiles of DS-5565 are possibly due to its unique binding characteristics to $\alpha_2\delta$ -1 and $\alpha_2\delta$ -2.

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Abstract – WCN 2013

No: 3192

Topic: 9 – Pain

Neuropathic pain and neuromodulation techniques

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Neuropathic pain is a common chronic intractable pain syndrome in clinical setting. It arises as a consequence of a lesion or disease affecting the somatosensory system [1] and may be associated with abnormal sensations called dysesthesia, pain produced by normally non-painful stimuli called allodynia and spontaneous pain. The mechanism of neuropathic pain remains unclear so far, and the management of patients with chronic neuropathic pain is challenging [2–6]. In this paper, I will discuss in brief the etiology, physiopathology and clinical management of neuropathic pain.

First, in etiology and physiopathology, I will discuss about craniocerebral injury, spinal cord injury, peripheral nerve injury, and come to a conclusion that a correct understanding of the neuropathic pain, especially its etiology and pathology, is extremely important to the prevention and treatment of it. Second, in clinical manifestations, I will discuss about relationship between injury and pain, pain duration, features of pain, thalamic pain, nutritional disorders, and motor dysfunction, which shows that more than 25% of patients may suffer from central pain within 1 week to 1 month after central nervous system damage. But in some other patients, central pain may occur after a few months or even years, and Pain occurs in somatic sensory impairments or deletion regions. Thalamic pain is a typical central pain known as hypothalamic syndrome. Third, in treatment, I will discuss about pharmacological therapy, Reducing sympathetic nervous stimulation, neuromodulation techniques, and get the result for spinal cord stimulation (SCS) (the National Pain Management & Research Center firstly performed SCS in China). From 2003, we have performed 202 SCS procedures (Fig. 1) and there are over 80% of patients who obtained pain relief successfully.

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Abstract – WCN 2013

No: 3145

Topic: 9 – Pain

Is an easy and reliable diagnosis of localized neuropathic pain (LNP) possible in general practice? Development of a screening tool

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Background: Neuropathic pain (NP) affects 26 million people worldwide [1]. 60% of NP patients present with localized symptoms [2]. Screening tools (DN4 [3]/painDETECT/LANSS [4]) are used depending on the countries, but NP remains frequently under-

diagnosed [5], an easy reliable tool for screening localized NP (LNP) patients is lacking.

Methods: A screening tool for diagnosing LNP proposed by international experts was tested by GPs (Badalona-Municipal-Hospital/Spain; Nov '12–Mar '13), and validated against pain specialist diagnosis as reference. Grading system principles proposed for NP [6] by IASP were basis for development, focusing on medical history and distribution of painful symptoms and sensory signs. Positive predictive value (PPV) and negative predictive value (NPV) were calculated.

Results: 2079 patients with chronic pain were consecutively screened by 31 GPs (Age: 60.7 ± 11.1 years; female = 69.9%). LNP was diagnosed in 394 patients. Screening, including patient examination, took 7mn (mean). GPs rated the tool as useful (24/31) or very useful (7/31) and facilitating practice (30/31). PPV/NPV was 41%/89%.

Discussion: An easy reliable tool for screening LNP by GPs is proposed, that could facilitate targeted treatment. The screening tool narrowly adheres to the NP algorithm proposed by IASP, being the first attempt to validate it in a general population of chronic pain patients on GP level. In contrast, established screening tools used strict inclusion criteria which eliminated unclear pain conditions. Thus, spectrum of disease, most relevant bias in diagnostic studies, is not biased in our real-life study, e.g. “gray zone” is not excluded. Hence, diagnostic accuracy of the tool is realistic. Meeting and study were sponsored by Grünenthal.

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Abstract – WCN 2013

No: 3201

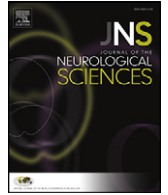
Topic: 9 – Pain

The mechanism of occurrence of the endoneural edema induced by histamine

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The effect of intravenous injection of histamine hydrochloride (10^{-4} M) on endoneural microvessels of white rat sciatic nerve was investigated at the light and ultrastructural levels. Endoneural edema characteristics appeared after 5 min of histamine injection. The presence of pores or transendothelial channels in endothelial layer of endoneural microvessels has not been revealed. The occurrence of edema liquid and the absence of convective ways of transport through the endothelial layer of microvessels indicate the increase of hydraulic permeability of their walls. The lumen expansion of all types of endoneural microvessels, especially arteriole and precapillaries, was observed after analyzing the occurrence of the endoneural edema after injection of histamine. The lumen expansion ostensibly reduces peripheral vascular resistance, and promotes blood intake with elevated hydrostatic pressure in the lumen of endoneural capillaries and postcapillary venule. This explains to be the cause of the appearance of edematous fluid in the endoneural space of the sciatic nerve. Thus, the increase of endoneural liquids stretches the perineural sheath and affects the lumen of venules allowing transit passage through it. Finally, the drastic increase of postcapillary resistance strengthens the filtration through the wall not only in capillaries, but also in arteriolar microvessels, and in extreme cases leads to infringement of the perineural sheath integrity. Based on collected data, the cause of degenerative changes of nervous fibers of peripheral nerves not only affects the increase of pressure of endoneural liquid but also the infringement of perineural sheath integrity. The role of shear stress in the expansion of lumens of endoneural microvessels is being considered.

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Topic: 10-Neurorehabilitation

Abstract—WCN 2013

No: 3212

Topic: 10—Neurorehabilitation

Prevention of cognitive impairment through a cognitive stimulation and rehabilitation program mediated by computers and internet

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Background: Neurodegenerative diseases present high prevalence over 50 years and geometrical progression over 60 years. Evidence shows cognitive impairment is preventable by cognitive stimulation. “Remembrance Workshop” is a cognitive stimulation and rehabilitation program based on computers and internet use.

Objective: The objective of this study is to determine whether a cognitive stimulation and rehabilitation program based in computers and internet use can prevent cognitive impairment measured by MMSE.

Material and methods: Intervention controlled cohort study, quasi experimental, was approved by Brazilian National Health Council. All participants were 50 or older, had memory complaints and had normal MMSE ≥ 25 . Analysis was performed by logistic regression after univariate and bivariate analyses, $p < 0.05$. Outcome was MMSE < 25 after the second interview. Intervention group had program plus medical follow-up and control group had only medical follow-up. Intervention: 20 biweekly workshops, 1.5 h each.

Results: Intervention group 112, 99 (88.4%) remained having no cognitive impairment and 13 (11.6%) showed having cognitive worsening. Control group, 96, 70 (72.9%) had no cognitive impairment and 26 (27.1%) had cognitive worsening. Control variables: gender, age, education, marital status, social status, physical activity, obesity, depression, cardiovascular disease, cerebrovascular disease, hypothyroidism, polypharmacy, dyslipidemia, smoking, use of benzodiazepines, diabetes, functional capacity, time between the first and second interviews, and initial MMSE. Hypertension was significant only in the bivariate analysis ($p = 0.031$, RR = 2.18 95% CI 1.02 to 4.65). Intervention was the only significant variable with an independent protective factor of 0.42 (95% CI 0.35–0.90, $p < 0.029$), NNT = 6.5.

Conclusion: The intervention was associated with prevention of cognitive impairment.

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Abstract—WCN 2013

No: 3183

Topic: 10—Neurorehabilitation

Spinal cord abscess due to congenital dermal sinus

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An intramedullary spinal cord abscess is rare and many cases are related with congenital dermal sinus in children. We report two cases of intramedullary abscess that needed rehabilitative treatment. A 34-month-old boy presented with fever, progressive quadriplegia, and respiratory difficulty and was diagnosed with an intramedullary spinal cord abscess extending to the medulla secondary to a dermal sinus infection. A 2-month-old boy presented with decreased movement of both lower extremities and irritability during diaper changing for several days. He had a fever 2 weeks earlier, and a macular lesion with dimpling was noted over the coccyx. Magnetic resonance images revealed intramedullary and extramedullary abscesses in the thoracic and lumbosacral spinal canal. Both children suffered from paraplegia, and neurogenic bladder and bowel despite surgery, antibiotic therapy, and rehabilitative care. An intramedullary spinal cord abscess is uncommon but it can cause persistent neurologic sequelae, which are more common with a delayed diagnosis. Because the condition is rare, a high index of suspicion is important for an early diagnosis.

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Abstract—WCN 2013

No: 3188

Topic: 10—Neurorehabilitation

Questionnaires underestimate sleep disorders and daytime complaints in neurorehabilitation

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Background: Sleep disorders, mainly SRBD and PLMS typically lead to complaints. Questionnaires like the Epworth Sleepiness Scale (ESS) and the Pittsburgh Sleep Quality Index (PSQI) normally are sensitive selection criteria for sleep screening.

Objective: Disturbed sleep will delay the rehabilitation progress, especially poststroke. The predictive value of ESS and PSQI in such patients was evaluated.

Patients and methods: 378 patients, 213 of them after stroke, underwent sleep polygraphy with an ambulatory device (SOMNOscreen, Fa. SOMNomedics GE). Inclusion criteria were normal speech and communication and sufficient mobility. In the afternoon of the screening the patients filled themselves the ESS and PSQI. The answers of the ESS and PSQI were evaluated thereafter. Independent raters scored the questionnaires and screenings.

Results: ESS was not completed in 36% females and 35% males after stroke without correlation with SRBD (AHI) or PLMS. ESS > 10 was found in 31% females but only in 17% males poststroke. AHI $> 10/h$ was found in 38% females and 79% males poststroke. ESS, PSQI, BMI and age did not correlate with AHI or PLMI in individuals poststroke nor in the other 165 patients (MS, other neurological disorders, orthopedic disease).

Discussion: ESS and PSQI do not correlate with type or amount of sleep disorders in rehabilitation patients. ESS and PSQI are not predictive and therefore should not be used as selection criteria for sleep screening or treatment indication. Nevertheless ESS and PSQI remain important tools for individualized patient education.

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Abstract—WCN 2013

No: 1162

Topic: 10—Neurorehabilitation

The relationship between students' bonding to school and multiple health risk behaviors among high school students in south-east of Iran

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Background: School is the first social institution that affects adolescents' lives and determines their opportunities, life qualities and behaviors. Thus, the aim of this study is to determine the relationship between bonding to school and multiple (problem) behaviors among high school students in Kerman.

Methods: The present study is a descriptive cross-sectional research carried out on students of all levels in high schools in Kerman during the 2010–2011 academic year. The research sample included 1024 students (588 females and 436 males) aged 15 to 19 years. The Multi-stage Cluster sampling method was used to collect data. Using A CTC (Communities That Care Youth Survey) questionnaire, which was designed by Using an standard questioner, the profile of risk behaviors of students were collected.

Results: In the final multivariate logistic regression, two variables including age ($OR_a = 1.15$, $p = 0.02$) and gender ($OR_a = 2.14$, $p = 0.001$) had a significant positive association with Multiple Health Risk Behaviors (MHRB). School commitment ($OR_a = 0.38$, p value = 0.001) and school rewards for involvement ($OR_a = 0.80$, p value = 0.21) had significant negative association with MHRB.

Conclusion: This research also demonstrated that students who were strongly influenced by school commitment and school reward for involvement are less likely to engage in risky behavior. The role of gender and age in these behaviors must also be taken highly into consideration.

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Abstract—WCN 2013

No: 2983

Topic: 10—Neurorehabilitation

Interval vacuum therapy in the rehabilitation of patients after microsurgical removal foraminal stenosis with the installation of the interspinous stabilization

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Introduction: Intermittent vacuum therapy is the widely method of treatment.

The aim of our research was to estimate the effectiveness of usage of "VACUMED" device for intermittent vacuum therapy in rehabilitation of patients after microsurgical removal foraminal stenosis with the installation of the interspinous stabilization.

Material and methods: Clinical and neurophysiologic examination of 82 patients (67% male, 33% female) with peripheral paresis of lower extremities in early postoperative period (middle time after

operation was 4 ± 1 days) after elimination of foraminal stenosis with the installation of the interspinous stabilization at L4–L5 and L5–S1 levels was analyzed in our research. The cohort was divided into two groups: the 1st group (38 patients) had the standard rehabilitation program applying system "VACUMED", the 2nd control group (44 patients) had only standard rehabilitation treatment. We used six-mark scale of muscle strength, miotonometer and electro-neuromyographic examination on apparatus Viking-Quest (Nicolet, USA) to estimate neurological status at the beginning of course and on the 14th day of rehabilitation.

Results: The decreasing of paresis degree was fixed in both groups of the patients: in the 1st group from 3.05 ± 0.17 to 4.72 ± 0.22 marks ($p < 0,05$); in the 2nd group from 3.12 ± 0.19 to 3.48 ± 0.18 marks.

Conclusions: Thus, the use of interval vacuum therapy in patients with interspinous spinal stabilization system in the early postoperative period is safe and effective compared to traditional rehabilitation.

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Abstract—WCN 2013

No: 2986

Topic: 10—Neurorehabilitation

Functional electrical stimulation in combination with robotic mechanic in patients with acute ischemic stroke

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Objective: The aim of our research was to evaluate the efficacy and safety of functional electrical stimulation, combined with a robotic mechanical therapy in patients with central hemiparesis in acute ischemic stroke.

Material and methods: 104 patients with acute ischemic stroke in middle cerebral artery were divided into 2 groups. Group I included 58 patients who received a course of rehabilitation therapy with the inclusion of training on robotic system Erigo and synchronized with functional electrical stimulation. Patients with group II (46) received treatment without FES. To assess the rehabilitation measures, we used a 6-point scale paresis and electrophysiological examination (impedance cardiography, Doppler ultrasound of the affected middle cerebral artery, evoked potentials) before, during and after the sessions of robotic mechanotherapy.

Results: In all patients, there was a decrease in the degree of paresis of 1.4 points in group 1 and at 0.54 in group 2. Hemodynamic abnormalities during robotic therapy in combination with FES were not revealed. At 20 days of rehabilitation, there was a decrease of the segmental latency response in both groups of patients, which was more pronounced in patients of group 1.

Conclusion: It was found that the use of functional electrical stimulation is safe in acute stroke, and the effect of the combination of FES with robotized mechanical therapy is higher than in conventional kinezioterapii.

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Abstract—WCN 2013

No: 2990

Topic: 10—Neurorehabilitation

Robotic technology in rehabilitation of patients after surgical revascularization of brain

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Objective: The aim of our study was to evaluate the effect of robotic mechano motoring deficiency in patients after surgical revascularization of the brain on the symptomatic stenosis of the internal carotid artery.

Material and methods: The patients were examined and divided into 2 groups: Group 1 – 95 patients after carotid endarterectomy and Group 2 – 39 patients after balloon angioplasty with stenting. Age stroke was from 17 to 523 days; prescription transactions from 1 to 3 months; and the degree of paresis of the lower extremity from 3 to 4 points. To assess the rehabilitation measures we used: a 6-point scale paresis, Stroke Scale NIHSS, Barthel ADL Index, Hauser Gate Index and electrophysiological examination. Patients received Lokomat training once a day for 6 days a week for 3 weeks. Discharge weight was on average no more than 25% of body weight, and treadmill speed was 1.5–2.5 km/h.

In two groups of patients there was a significant ($p < 0.05$) decrease in the degree of paresis in the lower limb and a significant increase in absolute and percentage figures in index walk Hauser. Barthel ADL Index increased in group 1 – with up to 46.0 ± 1.41 75.6 ± 1.2 ($p < 0.001$), and in group 2 – from 47.0 ± 1.4 to 74.4 ± 0.7 ($p < 0.001$) points. In this case, significant differences between the groups were not registered.

Conclusions: It was found that locomotor training with the help of Lokomat improves motor function and quality of life and that it doesn't depend on the type of surgery.

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Abstract—WCN 2013

No: 3148

Topic: 10—Neurorehabilitation

Transcranial direct current stimulation (tDCS) and vision restoration training (VRT) in post-acute stroke rehabilitation – Implications for further investigations

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Background: Posterior-cerebral-artery stroke usually results in visual field deficits varying in size and extent of the blind field and areas of residual vision (ARV). Vision restoration training (VRT) may reduce visual field impairment. Combined application of transcranial direct current stimulation (tDCS) together with VRT indicates that tDCS may accelerate VRT-effects in the post-acute stage. Feasibility of tDCS-VRT treatment in post-acute stroke was not yet investigated.

Objective: In order to plan a controlled efficacy study of tDCS-VRT in post-acute stroke, we investigated two patients with lesion ages of 2 and 5 months after posterior cerebral artery stroke with different lesion sizes.

Patients and methods: One patient with a small scotoma and ARV and one patient with homonymous clear-cut hemianopia without ARV were treated with 5 tDCS-sessions (2 mA, 15–20 min, cathode: Cz, anode: O1) combined with VRT. Perimetric detection thresholds at post were compared to baseline.

Results: After conducting the same stimulation and training protocol with both patients, we found significant improvement of detection accuracy in both cases. However there was a higher effect size in the patient with presence of ARV.

Conclusion: Our data support the theory that neural plasticity after stroke in the visual cortex can be facilitated by tDCS if there is residual functionality. Further studies investigating the efficacy of tDCS in rehabilitation of stroke should consider the presence of ARV as a predictor for a clinically relevant treatment effect.

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Abstract—WCN 2013

No: 3052

Topic: 10—Neurorehabilitation

Neurorehabilitation for amelioration of the quality of life of multiple sclerosis patients

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Aim: Structuration of a neurorehabilitation algorithm in MS and clinical approval of our physiotherapy programme and comparative evaluation of the effect of the application of different rehabilitation complexes – by a synergic combination of natural and preformed physical modalities: physiotherapy, kryotherapy, ergotherapy, pulsed magnetic field, electrostimulations, education of MS patients in activities of daily living.

Material and methods: During the period 1996–2012 in three Bulgarian Neurorehabilitation Clinics we effectuate observations on an experimental group of 189 patients with cerebro-spinal form of MS, in remission phase, with a clinically manifested quadripyramidal syndrome with hemiparesis or inferior paraparesis, discoordination syndrome, urinary incontinence, emotional disbalance. We divided experimental group in six therapeutic groups with rehabilitation complexes, including physiotherapeutic programme and different pre-formed modalities. The control group was formed by 49 in-patients, with standard rehabilitation programme. We observed MS patients – before and after physical therapy and one month later. All patients signed informed consent for rehabilitation and investigations. For statistical analysis we used the statistic package SPSS – options t-test (analysis of variances ANOVA) and Wilcoxon rank test (non-parametrical distribution analysis).

Results: During prospective observations we noted a significant effect on spasticity, balance, stability of gait, autonomy in activities of daily living – self service (movement in the bed, toilet, dressing, mobility, eating), test of 20 m walking, depressivity, and autonomy in ADL (ICF). The effect is stable – with duration of one month after the end of rehabilitation.

In conclusion we underline the favourable effect of neurorehabilitation on patient's quality of life.

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Abstract—WCN 2013

No: 3053

Topic: 10—Neurorehabilitation

Potential of neurorehabilitation in post stroke hemiparesis

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Introduction: In the context of the significant increase of the frequency of stroke, the authors propose some concrete algorithms for assessment of functional deficit and for management of major physical, cognitive and behavioral impairments resulting to functionality of hemiparetic patients with hemiparetic shoulder.

Goal: Evaluation of the efficacy of different physical-therapeutic and rehabilitation programs with the objective to construct neuro-rehabilitation complexes, adapted to the concrete clinical signs – for functional recovery and improvement of the independence in

activities of daily living of patients suffering from central post-stroke hemiparesis and hemiparetic shoulder.

Material and methods: A total of 388 post-stroke in-patients were observed (1997–2012). The rehabilitation complex includes kryokinesitherapy in all cases; additionally – electrostimulations of hand extensors (in group 2), and occupational therapy (in group 3). The patients were investigated before and after therapy and one month later, according a protocol with all results (including neurological and functional status, evaluation according to international classification of functioning, disability and health – ICF). Statistical evaluation was effectuated with t-test (analysis of variances ANOVA) and Wilcoxon rank test (non-parametrical correlation analysis). Comparative analysis of results proved a statistically significant favorable effect ($p < 0.001$) on some important parameters: muscle weakness, range of motion and functional capacity of the hand (including grip); stabilization of the balance and locomotion (tests of Brunnstrom, Barthel-index, Michel's test); reduction of spasticity; autonomy in activities of daily living (ICF). The stability of the results one month after the end of neurorehabilitation is satisfactory. In conclusion we underline clinical efficacy of complex neurorehabilitation program.

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Abstract—WCN 2013

No: 2104

Topic: 10—Neurorehabilitation

Assessment of the functional states of patients after stroke who underwent early rehabilitation

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Background: The main objective of the undertaken rehabilitation is a complete function playback or the compensation of the functions directly lost.

Objective: The aim of this study was to assess the functional status of stroke patients eligible for the early neurological rehabilitation.

Patients and methods: The research involved 37 patients, meeting the criteria for inclusion in the research. The assessment of functional status was performed on day 1 of hospitalization and 3 months after rehabilitation. To measure the functional status of a patient we used Lovett's scale, Brunnstrom's scale, Jebsen's test, Rankin's scale, ADL Barthel, and MMSE.

Results: The quality of the functional status of patients in the period from day 1 to 3 months after stroke improved according to Barthel's ADL scale by an average of 6 points. In the assessment of muscle strength improvement was seen in more than half of the patients; in 75.67% of them there was an improvement on the lower limb (LL) by an average of 1.1 points, in 48.6% of patients the improvement in the upper limb (UL) was by 2.1 points in the Lovett's scale, which also resulted in improving the functionality in the Rankin's scale in 59.4% of patients. The profile of changes in patients' functional status depended on severity of stroke symptoms.

Conclusions: In the first three months post-stroke functional improvement was achieved in two thirds of patients, the rest of them received no gait. The detailed assessment of the functional status allows for setting the objectives of early taken rehabilitation.

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Abstract—WCN 2013

No: 3091

Topic: 10—Neurorehabilitation

Shoulder pain after stroke: Experience in the University Hospital of Casablanca

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Background: The prevalence of shoulder pain after hemiplegic stroke is around 70%. A shoulder-hand syndrome, adhesive capsulitis may be the cause, favored by a glenohumeral subluxation or significant spasticity. The etiological diagnosis is mainly clinical. The treatment is mainly based on preventative tapping, correct positioning in bed, prudent mobilization and electrostimulation.

Objective: The objective of this study is to recall the different clinical, therapeutic and prognostic elements and to evaluate our results based on data from the literature.

Material and methods: Prospective study on the management of shoulder pain in 52 hemiplegic patients seen between October 2008 and March 2013.

Results: Age: 61 ± 14 years; post-stroke time: 2 to 29 weeks; sex ratio: 33 F/19 H; hemiplegic side: 41 right/11 left; pain assessed by VAS (initial: 8 ± 1 ; after treatment: 3 ± 1); etiologies: 46 subluxations/6 complex regional syndrome type I.

Treatment: All patients were treated with strapping, sling immobilization when standing or sitting, analgesic medication, TENS, and local infiltration of corticosteroids in cases of complex regional syndrome type I.

Conclusion: Shoulder pain is particularly high in hemiplegic particular patients, requiring particular therapeutic attention warranting research. The diagnosis of upper limb pain in the hemiplegic remains a subject of controversy. Its origin is multifactorial dominated by glenohumeral subluxation, complex regional syndrome type I, capsulitis, and spasticity. We must stress the importance of the prevention of decoaptation of glenohumeral by proper installation of the patient, proper diagnosis and initiation of early treatment.

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Abstract—WCN 2013

No: 2906

Topic: 10—Neurorehabilitation

Whole body vibration training improved balance and coordination of individuals with spinocerebellar ataxia

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Background: Whole body vibration, a newly developed strengthening paradigm, has been reported to have instant effect on regulating the strength of reciprocal inhibition in individuals with spinocerebellar ataxia (SCA). The purpose of this research was to investigate the long term effect of whole body vibration on balance and coordination in individuals with SCA.

Methods: Thirteen individuals with SCA were randomly assigned into training and control groups. Subjects in the training group received whole body vibration training in standing position for 3 sessions per week for four weeks. The control group received no training. The international cooperative ataxia rating scale (ICARS), Berg balance scale

(BBS), and one-leg standing time were used to evaluate coordination and balance before and after four weeks.

Results: After four weeks, the grade of ICARS ($p < 0.05$), BBS ($p < 0.05$) and one leg standing time ($p < 0.05$) improved in the training group. The control groups did not show changes.

Conclusion: Individuals with SCA can benefit from strength training by whole body vibration. The long-term training effect can be translated to the functional improvement.

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Abstract—WCN 2013

No: 2714

Topic: 10—Neurorehabilitation

Impact of early versus late treatment on goal areas in patients treated with botulinum-toxin A (BoNT-A) for post-stroke upper-limb-spasticity (ULIS-2-study)

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Objective: The objective of this study is to investigate primary goal achievement in patients with ULS based on time between stroke and first BoNT-A treatment for ULS.

Design: Subgroup analysis of 454 adults with post-stroke ULS within an observational, prospective cohort study on current practice in an international, multi-center design in 84 centers in 22 countries (ULIS-2-study).

Interventions: One cycle of BoNT-A treatment and concomitant therapies in accordance with routine local clinical practice.

Outcome measures: Primary goal achievement in relation to time after stroke.

Results: 180 patients were treated ≤ 1 year after stroke (early treatment, ET) and 274 > 1 year after stroke (late treatment, LT).

If ULS treatment starts ≤ 1 year after stroke, a higher proportion of goals related to pain (88.5%, 95%CI 69.8%, 97.6% ET versus 80.0%, 95%CI 63.1%, 91.6% LT), pROM (81.8%, 95%CI 64.5%, 93.0% ET versus 76.4%, 95%CI 64.9%, 85.6% LT) and active function (76.6%, 95%CI 62.0%, 87.7% ET versus 68.4%, 95%CI 54.8%, 80.1% LT) were achieved.

A higher proportion of goals related to ease of care (81.6%, 95%CI 68.0%, 91.2% ET versus 88.0%, 95%CI 79.0%, 94.1% LT) and reduction of involuntary movements (68.4%; 95%CI 43.4%, 87.4% ET versus 85.7%; 95%CI 63.7%, 97.0% LT) were achieved in the long-term.

Conclusions: Further long-term studies should examine patients characteristics and goal areas to develop recommendations on situations demanding an early treatment and a long-term ULS-management in terms of maintaining/improving functioning.

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Abstract—WCN 2013

No: 2952

Topic: 10—Neurorehabilitation

Therapeutic effects for human umbilical cord blood-derived mesenchymal stem cells with methylprednisolone treatment in the contused rat spinal cord

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Methylprednisolone (MP), a glucocorticoid steroid, has an anti-inflammatory action and seems to inhibit the formation of oxygen

free radicals produced during lipid peroxidation in a spinal cord injury (SCI). Currently MP is the standard therapy after acute SCI on reported neurological improvements. The combination therapeutic effect of human umbilical cord blood-derived mesenchymal stem cells (hUCB-MSCs) for transplantation time (1 d, 7 d, and 30 d) after MP treatment on the axonal regeneration and on the behavioral improvement in SCI was studied in the rat. The spinal cord was injured by contusion using a weight-drop at the level of T9 and MP (30 mg/kg, i.m., 10 min and 4 h) was acute administered after injury. hUCB-MSCs were labeled GFP and our study was performed the efficacy for transplantation time (1 d, 7 d, and 30 d) of hUCB-MSCs into the boundary zone of injured site. Efficacy was determined by histology, anterograde and retrograde tracing, and behavioral test. We found that hUCB-MSCs with MP treatment exerted a significant beneficial effect by neuroprotection and reducing cavity volume. Also the transplantation of hUCB-MSCs with MP treatment was significantly improved functional recovery. Combined transplantation at 7 d after SCI provided significantly greater efficiency than combined transplantation at 1 d and 30 d. These results suggest that transplantation time window of the hUCB-MSCs with MP treatment give rise to an earlier neuron protection strategy and effect of cell grafting in SCI. Thus our study may be considered as a therapeutic modality for SCI.

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Abstract—WCN 2013

No: 2975

Topic: 10—Neurorehabilitation

Experience of physical medicine and rehabilitation department: Treatment of spasticity by botulinum toxin injection, about 30 cases

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Introduction: The therapeutic use of botulinum toxin has been an important development in movement disorders.

Materials and methods: This is a retrospective study from 1 March 2012 to 30 February 2012 in the Physical Medicine and Rehabilitation department for the treatment of spasticity by botulinum toxin injection, which is indicated for the local treatment of spasticity or dystonia which awkward movement or health of the patient through a central nervous system, an assessment of the severity of the disorder by the Ashworth scale and functional evaluation using generic scales (FIM and Barthel).

Results: 30 patients have benefited from the injection of TB. The average was 27 years (3–75 years). Etiologies were vascular accident (51%), cerebral motor infirmity (29%), head injury (13%), tuberculous myelitis (4%), spinal cord injury (4%) and parkinsonism syndrome with dystonia (4%). The severity of spasticity was between 2 and 4 on the Ashworth scale, the injection objectives were variables: functional improvement of the members and hygiene of life and the prevention of joint deformations. Clinical and functional results were satisfied with a reduction in spasticity and an improvement in functional scores.

Discussion and conclusion: The injection of botulinum toxin is the treatment of choice for focal spasticity, and several studies confirm the very satisfied results and significant improvement in quality of life of patients and their functional recovery.

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Abstract—WCN 2013**No: 2980****Topic: 10—Neurorehabilitation****Cerebellar ataxia: Contribution of rehabilitation: 15 cases**

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Introduction: Cerebellar ataxia is a disorder that affects motor complex planning and execution of movements and reduces the precision and coordination.

Purpose: The purpose of this study is to assess the impact of rehabilitation on cerebellar ataxia in CHU Ibn Rochd Casablanca.

Method: A retrospective study of 15 cases of cerebellar ataxia received a rehabilitation program.

Results: The majority of patients were men (60%), the average age is 19 years (4–54), and etiologies found were dominated by multiple sclerosis (40%), and head trauma (33.5%). Evaluation of balance and walking according to Tinetti showed 73.5% of patients with high risk of falling, the Barthel scale evaluation found 13% had severe dependence, 53, 5% had moderate dependence and 33.5% had a slight dependence. Rehabilitation has improved complete autonomy in activities of daily life in 6.5% of patients and improved level of Tinetti scale in 26.7%.

Discussion and conclusion: Cerebellar ataxia is one of the most characteristics of the cerebellum damage. The beneficial effect of rehabilitation on neurological signs has been proved by several authors; intensive training coordination improves motor performance and reduces ataxia symptoms. Others believe that there is no valid information on the actual value of the physical rehabilitation and psychological support as a treatment for cerebellar ataxia, and that there is a tendency to spontaneous regression of cerebellar syndromes, but it can be slow, incomplete, extending over months or years.

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Abstract—WCN 2013**No: 2942****Topic: 10—Neurorehabilitation****Combining electric brain stimulation and source-based brain-machine interface (BMI) training in neurorehabilitation of chronic stroke**

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Over the last years, clinical trials indicated that brain-computer or brain-machine interface (BCI/BMI) training coupled with behavioral physiotherapy can facilitate motor recovery after severe stroke. However, results were heterogeneous and training required weeks of daily sessions. Here we introduce a novel BMI-based rehabilitation regime that promises to improve specificity and effectiveness of such training by incorporating the latest methodological advancements, such as area-specific (source-based) real-time reconstruction of brain activity and simultaneous electric brain stimulation during BMI-training to facilitate learning.

Methods: A 48 yo female patient unable to move her right fingers who suffered a subcortical stroke at the age of 25 affecting the left capsula interna, putamen and nucleus caudatus was invited to three consecutive BMI-training sessions. A 275-sensor whole-head magnetoencephalograph (MEG) was used to localize and reconstruct task-related neuromagnetic activity in the left motor cortex (M1) during the attempt to move the affected fingers. Source-based activity was assessed using a LCMV beamformer and translated into

online feedback delivered through a hand-exoskeleton opening and closing the paralyzed hand. During the BMI-training, an anodal DC-electric current of 1 mA was delivered to the left M1.

Results: Area-specific real-time reconstruction of motor-related brain activity was reliably possible during electric brain stimulation. The patient achieved good BMI-control across all sessions. Brain stimulation was well tolerated. This is the first report on successful real-time combination of source-based BMI-training and electric brain stimulation in a stroke patient, building the grounds for larger clinical studies further investigating efficacy of such approach in the rehabilitation of severe stroke.

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Abstract—WCN 2013**No: 3027****Topic: 10—Neurorehabilitation****A new onset of Parinaud's syndrome in a rehabilitation setting**

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Background: Parinaud's syndrome involves supra nuclear vertical gaze palsy due to pathology in dorsal midbrain. There is no specific rehabilitation strategy in literature.

Objective: Through a case story, we assess the clinical and service implications.

Patients and methods: A 35-year old right handed male was diagnosed with cerebellar pilocystic astrocytoma presented with headache and ataxia for 4 months. Post operative complications include hydrocephalus, repeated shunt infections. He was transferred to Neurorehabilitation after clinical stabilisation.

Due to substantial visual impairment, he was re-referred to Neurologists who found severe diplopia in all directions but worse horizontally, sluggish pupil with ptosis, absent pupillary reflexes with no near light response, conjugate vertical gaze paralysis with bilateral internuclear ophthalmoplegia, absent vertical and horizontal optic kinetic reflexes. Vertical and Horizontal nystagmus was present with preserved corneal reflex. Eye movements demonstrated convergent nystagmus.

In summary there was nuclear 3rd and 6th nerve palsies evident bilaterally, convergent nystagmus and absent near light near reflex with clinical signs in keeping with a Parinaud's syndrome except eye lid retraction. Repeat neuro-imaging showed subdural bleed but no dorsal midbrain lesion. We speculated that the likely cause of these new onset Parinaud's stemming from raised intracranial pressure i.e. Pre-tectal syndrome.

He was re-referred to Neurosurgeons.

Conclusion: Apart from the curious clinical presentation, this case raised important issues relating to service reconfigurations e.g. continued engagement with neurologists are needed to guide the rehabilitation programme, prognostication and active interventions. A careful consideration of budgetary implication of neuro-rehabilitation department is needed.

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Abstract—WCN 2013**No: 3047****Topic: 10—Neurorehabilitation****Botulinum toxin (BT) therapy for adult spasticity — A service improvement projects over 9 years**

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Royal College of Physicians (UK) produced Guidelines (2003 & 2009) detailing the Standards of multidisciplinary (MDT) rehabilitation of adult spasticity using BT. In 2006, an audit was conducted ($n = 100$) against that Standard 1 and the identified action points were implemented.

Methodology: A re-audit in 2009 analysed data of 70 consecutive patients (2006–9) against the RCP Standards. A repeat-re-audit was done on a small sample (15) in 2012.

Result and discussion: Demography and case mix remain similar to 1st audit. We noted substantial improvements in defining treatment goals with patients and carers 94% (Cf 68% in the 1st audit). The follow up of patients by therapists 2 weeks & 6 weeks were 54% & 46%. Standardised outcome measurement tools used in 49% cases. Written communication between community physiotherapists and hospital team showed no improvements. Specific gaps identified in the initial audit showed improvement e.g. documentation of aggravating factors (100%), dilution of BT improved (62%), named therapist was identified pre-injection in 96% (cf 26%) in 1st audit. However, 2012 data showed substantial deterioration.

Conclusion: Such complex service requires co-ordinated care plan by MDT from different organisations within their context. The key gap is ensuring timely post-injection therapy. Identifying a named therapist improved the service. Communication between community & the hospital team deteriorated. This project highlights the value of the iterative process of re-auditing since different challenges will emerge for improvement and sustainability.

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Abstract – WCN 2013

No: 2164

Topic: 10 – Neurorehabilitation

Quality of life and functional outcome in stroke survivors

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Background: Modern treatment and rehabilitation strategies reduce the risk of death or dependency after stroke. However, better functional outcome may not always be translated to better quality of life (QoL). The aim of the study was to assess QoL in stroke survivors in relation to their functional outcome.

Material and methods: We questioned 142 consecutive patients one year after stroke, who underwent early post-stroke neurorehabilitation in our department. To assess their QoL we used the Stroke-Specific Quality of Life Scale developed by Williams. Functional outcome was assessed with modified Rankin scale (mRS).

Results: 19 (13%) patients had died within one year and only 50 patients (mean age 68.5 years; 25 men) delivered answers to all questions from the query. The most affected areas of life, where patients scored <40% of the total score in the selected sub-scale were 'work' (54% patients), 'energy' (36%) and 'personality' (33%). Functional improvement by at least 1 point in the mRS was observed in 72% of patients. There were no differences in any assessed QoL domains between patients with good and unfavorable functional outcome. With univariable logistic regression we did not find any association of functional improvement with the change in any of individual QoL domains. In multivariable stepwise backward analysis 'work' and 'family roles' were the most associated with functional outcome (model: $p = 0.036$, $\rho^2 = 0.208$).

Conclusions: Functional improvement after stroke does not guarantee better quality of life. Individualization of post-stroke rehabilitation programs should include patients' families or caregivers.

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Abstract – WCN 2013

No: 3012

Topic: 10 – Neurorehabilitation

Fluoxetine for post stroke depression: Experience of rehabilitation unit in Ibn Rochd University hospital of Casablanca

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Background: Depression after stroke is an obstacle to the rehabilitation which imposes early antidepressant treatment.

Objective: The objective of this study is to evaluate the efficacy of fluoxetine in depression after stroke and its effect on patient response to rehabilitative treatment.

Material and methods: Prospective study over 17 months from September 2011 to February 2013 effecting 41 depressed stroke patients followed in the rehabilitation unit. Onset of stroke had to be less than 30 days, patients with severe aphasia were excluded. The diagnosis of depression after stroke was as defined by the DSM IV, the severity of depression was assessed using the MADRS, and the effectiveness of depression was assessed by the PASS. Treatment with fluoxetine was introduced to all patients at a rate of 20 mg/d. The MADRS and the PASS were reassessed at 1 and 2 months.

Results: Of 56 patients with hemiparesis, 41 patients developed post-stroke depression (73.2%); the mean age was 54.3 years ranging from 31 to 85 years, the sex ratio was 1.7, ischemic stroke accounted for 76.9%, 30 patients (73.1%) had minor depression and 11 patients (26.8%) severe depression. PASS averaged 17.2/36 in early assessment and was 26.9/36 to 2 months of treatment. Fluoxetine was well tolerated, no adverse effects were noted.

Conclusion: In patients with depression after stroke early prescription fluoxetine should be coupled with rehabilitation; this treatment has a dual effect, on depression and on motor recovery as demonstrated by authors reporting the effect of fluoxetine on brain neuroplasticity after stroke.

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Abstract – WCN 2013

No: 3002

Topic: 10 – Neurorehabilitation

Vestibular rehabilitation improves postural stability in the degenerative spinocerebellar ataxia patient (DSA): A case report

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Background: DSA is genetically associated with postural instability. The DSA patients usually have gait ataxia, fear of falls and decrease balance due to limited activity in daily life. The rehabilitation program of these patients is long term. So, it should be designed for clinical use and home program.

Propose: To determine the effectiveness in increasing the stability after vestibular rehabilitation (VR) in DSA patient and to create the new concept of treatment ataxia patients.

Method: A case report. A 63 years old male with 6 years of DSA symptom who can walk by rolling wheel walker with supervision.

He received the treatment program and the home program that consist of

1. Cawthorne and Cooksey exercise.
2. substitution exercise.
3. balance training.

The subject was treatment at the physical therapy clinic 1 day/week and home program 3 day/week in 6 weeks. The stability test used Time-up and Go test (TUG) before and after treatment at the clinic.

Result: After 6 weeks, the TUG score before and after rehabilitation program was decrease from 35.17 ± 15.39 s to 29.17 ± 8.7 s ($P = 0.001$) and the difference score in each week of treatment program decrease 17, 3, 10, -3.2 and 7 s. The average TUG score before and after program = 6 ± 6.99 s (95%CI = -1.33 to 13.33).

Conclusion: Although, the VR can improve the stability of DSA patient, the DSA is a degenerative disease. The patient should keep continuous exercise to maintain the body function, balance and strength.

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Abstract – WCN 2013

No: 2991

Topic: 10 – Neurorehabilitation

Neuromodulation for intractable primary headaches

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Objectives: This review discusses the role of neuromodulation in the management of intractable headaches.

Discussion: Although most patients benefit from a range of acute and preventive drugs, a subgroup of patients remain refractory to treatment. For these patients, invasive techniques should be considered. Despite good efficacy in some forms of primary headache, nerve blocks with either local anaesthetics or steroids or combinations thereof, provide only temporary relief in most patients.

Neuromodulatory approaches can be pragmatically classified according to implantation site and invasiveness. Given that complex networks of pain processing structures are activated and deactivated upon stimulation of a single target¹, neuromodulation which infers activation only. The mode of action differs substantially between different approaches.

Conclusion: In clinical practice, delivery of the appropriate neuromodulatory approach remains a challenge. Therefore, an interdisciplinary approach involving a team with expertise in both diagnosis and treatment of headaches and the implantation of invasive neuromodulatory devices are crucial for the success of this promising addition to our armamentarium against intractable primary headaches.

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Abstract – WCN 2013

No: 2031

Topic: 10 – Neurorehabilitation

Reach-to-grasp coordination by avoiding obstacle collision at first and after twelve-months post-stroke

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Background: Substantial controversy regarding reach-to-grasp (RTG) coordination and planning deficit was found during the first twelve-month post-stroke. One of the important factors leading to the conflicting results can be the different ways of analyzing parameters that have been used to detect the coordination.

Objective: To investigate RTG coordination in the paretic hand of individuals with stroke, performing RTG actions avoiding obstacle collision.

Material and methods: Right handed individuals during the first twelve-month post-stroke and after the twelve-month post-stroke having mild severity of upper extremity impairment were recruited with an equal number of non-disabled adults. Kinematic RTG movements with obstacle avoidance at a fast speed were assessed. RTG coordination was quantified using cross-correlation analysis. The correlation coefficient (spatial aspect) and time lag (temporal aspect) were reported. Kinematic variables of RTG actions were also analyzed.

Results: Both post-stroke groups showed a significant disturbance in RTG coordination only with respect to time lag compared to non-disabled adults. The percentage of time to maximum aperture reflecting planning of grasping showed a deficit only in the first twelve-month post-stroke period. In contrast, we did not find a planning deficit of reaching, quantified by the percentage of time to maximum transport velocity following the stroke.

Conclusion: Individuals with mild stroke showed impairment in RTG coordination. The coordination was disturbed in the temporal aspect but preserved in the spatial aspect. The findings indicate that the planning of grasping showed a deficit in the first twelve-months and recovered after twelve-months post-stroke, whereas planning of reaching remained intact.

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Abstract – WCN 2013

No: 2892

Topic: 10 – Neurorehabilitation

A 22-year follow-up of the incidence of traumatic spinal cord injuries in Western Denmark

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Background: Retrospective population-based epidemiological study.

Objective: To assess temporal trends in the incidence of traumatic spinal cord injuries (TSCI) and demographic and clinical characteristics of a geographically defined cohort in the period 1990–2011.

Methods: Patients were identified from hospital records. Crude rates and age-adjusted rates were calculated for each year. The multivariate relationship between cause of injury, age at injury, time of injury and gender was examined using a Poisson regression model.

Results: A total of 669 patients were admitted to the Spinal Cord Injury Centre of Western Denmark. The annual incidence varied between 5.9 and 13.7 per million; men 9.1–23.3 and women 0.7–7.4 per million. Mean age on admission for whole period was 38.6 years (men: 37.9 years and women: 41.6 years). The incidence of MVA-related injuries increased during the observation period, especially among men < 30 years. The incidence of TSCI increased among men < 60 years.

Motor vehicle accidents (MVA) was the most common cause of injury (50.2%), followed by sports and leisure activities (24.5%). More women were injured during sports and leisure activities (29.8% vs. 23.4%) and more men were injured in MVA (50.4% vs. 47.9%). The lesion level was cervical in 48.6%, and clinically incomplete in 57.2%.

The incidence of incomplete tetraplegia decreased among men <30 years, and increased among men >60 years.

Conclusion: The incidence of TSCI has remained stable during the past 22 years. The most frequent causes of TSCI, MVA and sports and leisure activities, are potentially preventable causes.

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Abstract – WCN 2013

No: 2830

Topic: 10 – Neurorehabilitation

The burden of paediatric cerebral palsy on caregiver's mental health

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Background: Paediatric Cerebral Palsy (PCP) is the leading cause of childhood disability affecting function and development with huge social burden because of disease both costs of treatment and number of caregivers.

It's well known that PCP affects not only the child, but also their immediate caregivers (usually mothers).

Aim: To rule out the level of depression in mothers with PCP child.

Design: 96 mothers with PCP kids were evaluated during their children rehabilitation in the city Jermuk, Armenia. Beck Depression Inventory (BDI) was assessed. The level of education was also included.

Results: 75 (78.1%) of mothers with kids with PCP had depression. Distribution of depression was 23 (24%) mild, 47 (49%) moderate, and 5 (5.2%) severe. We found strong correlation between severity of depression and education. Mothers with university or higher degree level show most severe level of depression. 40 (88.9%) of mothers with university or higher degree level had depression. Distribution of depression among mothers with university or higher degree level was 11(24.4%) mild, 25(55.6%) moderate, and 4(8.9%) severe.

Conclusion: PCP must be considered as disease with huge impact on caregiver's mental health. Mothers with kids with PCP must be mandatorily evaluated for depression, for a successful management of PCP.

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Abstract – WCN 2013

No: 2828

Topic: 10 – Neurorehabilitation

Low level laser therapy improves neurological deficits after embolic strokes in rats

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Background: Stroke remains the third leading cause of death. The sequel to stroke leads to disability in daily living activity performance and great economic burden.

Objective: Low-level laser therapy (LLLT) has been used a wide range in rehabilitation field. However, few researches have mentioned the

effects on stroke. The purpose of this study is to reveal the effects of LLLT on stroke recovery and possible mechanisms involved.

Material and methods: Seventy two (300–350 g) Sprague-Dawley rats were divided into the sham (n = 6) and studied groups. The studied rats were applied with a transient occlusion of the middle cerebral artery through internal carotid artery (tMCAO). The studied rats were further divided into 8 subgroups and 6 rats each were irradiated to 0J and 8J of LLLT for 1, 4, 7 and 14 days after tMCAO. Histological studies were performed with triphenyl tetrazolium chloride (TTC) staining to demonstrate the ischemic volume. Behavior patterns could reflect the motor recovery from the impairment. Molecular assays were used to assessed the mechanism. **Results:** After LLLT on tMCAO tissues, the ischemic volume decreased. Rat movements including front limb symmetry, cylinder test, and swimming task showed much improvement. The mRNA of growth-associated protein-43(GAP-43) increased and tumor necrosis factor (TNF- α) and interleukin (IL-1) significantly decreased after LLLT.

Conclusion: LLLT could improve rat neural regeneration in the tMCAO tissue and promote motor recovery. LLLT also regulates the nerve related gene expression GAP-43, TNF- α and IL-1. LLLT could offer a non-invasive treatment adjuvant for stroke.

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Abstract – WCN 2013

No: 2787

Topic: 10 – Neurorehabilitation

Functional outcomes after surgery for heterotopic ossifications: 22 cases

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Introduction: Heterotopic ossification is a frequent complication in head injury patients. The functional impairment may be severe. The goal of surgery is to improve function.

The aim is to determine functional outcomes of patients undergoing surgery for heterotopic ossifications.

Material and methods: This was a prospective study of patients who underwent surgery for heterotopic ossifications followed by intensive rehabilitation care in our department from January 2009 to March 2013. The evaluation included a joint assessment and a functional assessment for each affected joint.

Results: There were 22 patients (29 operated joints). Predominantly male (82%) and the average age 31.6 years (19–41 years). Operated joints were: knee (n = 11, 38%), elbows (n = 11, 38%), and hips (n = 7, 24%). All patients received physical therapy based mainly on continuous passive mobilization of the elbow or knee in addition to functional work. For operated hips, the Postel Merle d'Aubigné (PMA) score improved from 6.5 to 8. In patients who had knee surgery, the functional status improved: one patient recovered the ability to walk and the others good sitting position. For patients who had elbow surgery, the functional assessment revealed improved possibilities for global nutrition (hand–mouth), hygiene (hand–face) and grooming (hand–neck).

Discussion and conclusion: The main objective of surgery for heterotopic ossifications is to restore joint mobility and function. Appropriate rehabilitation in an experienced PRM unit greatly contributes to improved functional capacities.

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Abstract – WCN 2013**No: 2812****Topic: 10 – Neurorehabilitation****Impact of soft-tissue-shortening on goal achievement in patients treated with botulinum-toxin a (BONT-A) for post-stroke upper-limb-spasticity (ULIS-2 study)**

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Background: Soft-tissue-shortening (STS) is a common long-term sequel of upper-limb spasticity (ULS) affecting different levels of functioning if not treated appropriately.

Objective: To describe the impact of STS on goal achievement in different goal areas in patients after BoNT-A treatment of ULS.

Patients and methods: Analysis of primary goal achievement in relation to STS in adults with post-stroke-ULS within an observational, prospective cohort study on current practice in an international, multi-center design (84 centers in 22 countries: ULIS-2).

Intervention included one cycle of BoNT-A treatment and concomitant therapy in accordance with routine local clinical practice.

The presence of soft tissue shortening was defined as severe restriction of passive range of motion (pROM) in at least one segment (shoulder, elbow, wrist, hand).

Results: 340 (74.5%) patients had no STS and 116 (25.5%) presented STS in at least one segment.

If STS was present, goals related to pain control (87.5% with STS versus 81.1% without STS) and ease-of-care (87.5% with STS versus 84.8% without STS) were more often achieved.

A higher proportion of patients without STS achieved goals in active function (73.9% without STS versus 58.3% with STS), maintaining/improving pROM (83.3% without STS versus 63.0% with STS) and reduction of involuntary movements (84.4% without STS versus 55.6% with STS).

Conclusions: Goals related to pain control and ease-of-care were more often achieved in patients with STS, whereas a higher proportion of those without STS achieved goals in active function, improved pROM and reduction of unwanted involuntary movements.

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Abstract – WCN 2013**No: 2779****Topic: 10 – Neurorehabilitation****Functional diagnosis of brain function in patients with severe chronic disorders of consciousness**

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Background: Recent research showed that some patients with severe chronic disorders of consciousness (SC-DOC) have partially higher brain functions and therefore a certain level of residual consciousness, which cannot be assessed by clinical examination. Functional MRI was discovered as a possible additional tool to the clinical examination.

Objective: The aim of this study is to investigate, if clinical testing evaluations using the Coma Recovery Scale-Revised (CRS-R) and the

Wessex Head Injury Matrix (WHIM) and a so-called “Own-name-paradigm” have similar results in differentiating between these two states of severe chronic disorders of consciousness, namely the minimally conscious state (MCS) and the unresponsive wakefulness state (UWS).

Material and methods: Twenty-six patients with SC-DOC were assigned into the 2 states according to detailed clinical examination and by CRS-R and WHIM. Using an event-related fMRI paradigm, the brain activity during a sentence (for example “Martin, hello Martin”) with the own name or another name was investigated. Afterwards in 7 previously defined regions of interest (ROI) the results of the patients were compared with the activation in healthy subjects.

Results: According to the clinical examination and the testing, 19 UWS and 7 MCS patients were diagnosed. In 17/19 UWS patients and in 5/7 MCS patients activations similar to these of healthy subjects were found. In both groups only 2 patients showed no activation in the 7 ROIs.

Conclusion: It could be demonstrated that there is a higher brain function in diagnosed UWS patients. We believe that the fMRI is an important tool to reconsider the diagnosis.

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Abstract – WCN 2013**No: 2784****Topic: 10 – Neurorehabilitation****Neuromodulation of the sensorimotor cortex by vibration stimulation of the whole-hand**

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Background: It has been recently shown that 20 min of mechanical flutter stimulation induces lasting motor cortical excitability changes, as assessed by transcranial magnetic stimulation in relaxed hand muscles.

Objective: The present functional magnetic resonance imaging (fMRI) study aims to examine if such neuromodulatory changes are reflected in the BOLD signal during a motor test.

Materials and methods: Two groups were recruited: one group receiving whole-hand flutter stimulation with a frequency of 25 Hz (FSTIM group, n = 22) and a second group receiving no stimulation (NOSTIM group, n 22). As motor test finger-to-thumb tapping was performed to activate a wide sensorimotor network during the fMRI measurements. Three fMRI measurements were obtained with this test: before stimulation (PRE), after stimulation (POST1), and 1 h after stimulation (POST2). Three regions of interest (ROIs) were defined: primary motor area (M1), primary somatosensory area (S1), and supplementary motor area. In the absence of baseline differences between both groups, the FSTIM group showed increased movement-related brain activations compared with the NOSTIM group, both at POST1 and POST2.

Results: ROI analysis revealed increased blood-oxygenation-level-dependent (BOLD) responses within contralateral S1 (p20%) and M1 (p25%) at POST1, which lasted until POST2. These poststimulatory effects within S1 and M1 obviously reflect neuroplastic changes associated with augmented cortical excitability.

Conclusion: We could demonstrate neuromodulation of the sensorimotor cortex by vibration stimulation of the whole-hand that can be

applied as a pre-conditioning of the sensorimotor cortex for a consecutive motor therapy to improve outcome in neurorehabilitation.

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Abstract — WCN 2013

No: 2735

Topic: 10 — Neurorehabilitation

Modulation of cortical plasticity by whole-hand electrical stimulation in attempt to improve hand motor functions after stroke

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Background: Peripheral electrical stimulation has been proved to modulate cortical plasticity in healthy and in patients. Such effects also occur after application of electrical mesh-glove stimulation (MGS) in our previous studies on healthy subjects.

Objective: The effect of whole-hand electrical stimulation on motor recovery in stroke patients at the subacute stage is examined.

Materials and methods: Patients with cortico-subcortical stroke and predominantly motor hemiparesis of the upper extremity were recruited. MGS was applied on the paretic hand daily for 60 min before the standard rehabilitation training over three weeks. Hand motor and sensory functions were evaluated with Wolf Motor Function test, Fugl-Meyer Assessment score, Nine-hole-peg Test, and Semmes-Weinstein monofilaments. Single and paired-pulse transcranial magnetic stimulation (TMS) were applied to evaluate the corticospinal excitability changes over the treatment period. Further, functional magnetic resonance imaging (fMRI) was conducted to assess the cortical brain reorganization changes after the treatment. Effects of MGS were compared to a control group receiving sham stimulation.

Results: Patients from both groups showed significant functional improvement as assessed with the motor functional tests. The improvement for the MGS group was increased compared to the control group. These functional effects correlated with neuroplastic changes within the sensorimotor area as revealed by TMS and fMRI.

Conclusion: Electrical stimulation applied before a physiotherapeutic training raises the motor cortical excitability in the lesioned cortex so that the subsequent training becomes more effective. The obtained results provide better understanding how modulation of central motor controlling structures by somatosensory stimulation correlates with functional motor recovery.

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Abstract — WCN 2013

No: 2767

Topic: 10 — Neurorehabilitation

Neurological bladders disorders: Clinical and urodynamic aspects

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Introduction: Therapeutic of neurological bladder need an Urodynamic exploration. It's prevented urinary complications.

Objectives: Our objective was to determine the Urodynamic exploration aspects to patients with neurological bladder.

Patients and methods: We conducted a retrospective study in Rehabilitation department of teacher hospital of Ibn Rochd (Morocco). Patients with neurological bladder who benefited a complete Urodynamic exploration between October–March 2013 were included. Urinary disorders which can be explicated for organic urinary lesions were excluded. Biographic, urinary symptoms, results of neurological exams and Urodynamic exploration were noted.

Results: We included 55 folders of patients. Patient aged under 15 year at 54, 5% of case. Women represented 56, 4%. Urinary disorders were low voiding jet (30.9%), urinary hesitation (20%), incomplete draining (18.2%), enuresis (7.3%), and urinary incontinence (50, 9%). Other urinary disorders were urge incontinence (18.2%), pollakiuria (5.5%). Myelitis (11%), spinal cord tumors (7.3%), multiples sclerosis (5.5%), encephalitis (3.6%), traumatic spinal cord (3, 6%), and spina bifida (3, 6%) were the principal etiologic. Unknown etiologic represented (56.4%). Urodynamic exploration found dysuria (36.4%), vesicospincter dyssynergia (25.5%), high bladder activity (44.9%), low compliance (18.4%), level basic tone (18.4%), high compliance (16.3%), sphincter deficiency (22.4%), and sphincter hypertension (8.2%).

Conclusion: Urodynamic exploration aspects in neurological bladders are multiple. Adequate therapeutic depends on results of this exploration.

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Abstract — WCN 2013

No: 2745

Topic: 10 — Neurorehabilitation

The outcome of stroke patients at 1 year after discharge

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Background: Stroke is common condition leading to disability and decreasing quality of life. Therefore, continuing home rehabilitation is the important way to maintain or improve patient's ability and to prevent further complication.

Objective: To study outcome of stroke patients at 1 year after discharge.

Material and methods: Chronic stroke patients who were admitted to rehabilitation ward, Srinagarind hospital were recruited. All participants were asked to complete the questionnaire which consisted of 6 items: personal data, stroke severity scale, basic Activity of Daily Living (ADLs), instrumental Activity of Daily Livings (IADLs) and mental health.

Results: Of these 54 stroke patients, most (74%) were male, with mean age 60 (SD11) years old. Regarding ADLs, 44% were physically independence, 40% had mild to moderate disability, and 16% had severe disability. Most of them (74%) continued self rehabilitation at home. The major problem of self rehabilitation was easily fatigue and spasticity (30%), distress (17%) and insufficient family support (15%). About half had poor mental health when compared with general population. Although they could do self home rehabilitation, they still need the home rehabilitation by health professional. Surprisingly, 80% of patients received alternative medicine; Thai massage, acupuncture and herbs.

Conclusion: Most stroke patients had been continuing home rehabilitation, however home visit by health professional still be necessary.

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Abstract — WCN 2013

No: 2747

Topic: 10 — Neurorehabilitation

The people empowering people (pep) program for stroke patients in Srinagarind hospital

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Background: Stroke patients have impairments and disability. The empowerment is important to enhance the patient wellness.

Material and methods: The 115 stroke patients and 115 caregivers who visited rehabilitation clinic during October 2012–March 2013 were recruited in the program. The PEP program was conducted in small group fashion (5–6 patients/group). The participants were encouraged to express their own issues and discussion. Moreover, essential skills and knowledge for self care activity, and counseling using BATHE technique were instructed by the health professionals.

Results: Most of patients (80%) were able to do self care activity independently. About 30% had complications; joint stiffness, pressure ulcer, aspiration pneumonia and urinary tract infection. All of participants had good knowledge in stroke disease and risk factor. The individual issue mainly focused on emotional problem (depression, anxiety, anger, emotionalism) after stroke, lack of family support, inappropriate environment and being unable to walk.

Conclusion: The PEP program provided an opportunity for patients and their caregiver to share experience with other patients who have had the same experience and health professional as well. This program would be an effective intervention to stimulate patient's well being.

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Abstract – WCN 2013

No: 2765

Topic: 10 – Neurorehabilitation

Multifocal tuberculosis revealed by cerebellar ataxia: One case

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Introduction: Multifocal tuberculosis is uncommon and is defined by the achievement of two extra-pulmonary sites with or without pulmonary involvement. Multifocal tuberculosis is grave and responsible for 16–25% cases mortality. It is mainly associated with immunocompromised person.

Purpose: To describe the characteristics of multifocal tuberculosis in an immunocompetent.

Case report: 38-year-old woman with history of epilepsy consulted for difficulty in walking, ataxia and weight loss of 10 kg, physical examination found a dyspnea, a normal conscious, a normal voluntary control, a vivacious deep tendon reflexes, balance disorder, enlargement of support polygon, drunken gait, all in a context of apyrexia and impaired general condition. The chest X-ray objectified a miliary tuberculosis, chest CT scan was in favor of pulmonary tuberculosis and lymph nodes, cerebral MRI showed nodular lesions in the left region of occipital lobe with triventricular hydrocephalus in favor of neuromeningeal tuberculosis. The diagnosis of multifocal tuberculosis (pulmonary, cerebral and lymphoid) was retained. Patient received her BCG vaccination without any immunocompromised therapy. Immunocompromised disease was not found. Management was 12 month of antibiaccilar treatment with rehabilitation. Clinical, laboratory studies and radiological evolution were good.

Discussion: Multifocal tuberculosis is a severe form which can affect even immunocompetent patient particularly in the Moroccan context, hence the need for a systematic dissemination checkup assessment for better management. This treatment should be instituted as soon as possible to avoid the sequelae.

Conclusion: Multifocal tuberculosis is severe, to improve its prognosis, we emphasize the importance of early diagnosis and therefore the treatment.

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Abstract – WCN 2013

No: 2757

Topic: 10 – Neurorehabilitation

Atypical evolution of Guillain Barré syndrome: One case

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Introduction: The Guillain Barré syndrome is an acute polyradiculoneuritis affection. Involvement of the peripheral nervous system with bladder dysfunction is rare.

Purpose: We describe a case report of uncommon manifestation of GBS with bladder dysfunctions and literature study.

Case report: Mme R.A 37 years old, without particular past history, presented an acute polyradiculoneuritis GBS in acute phase with flask tetraparesis, areflexia, facial diplegia, oropharyngeal dysphagia, respiratory difficulty, constipation and urinary retention. The EMG showed a heterogeneous distal and proximal sensory motor function loss with demyelinating stigma, an important axonal injury without any signs of active denervation indicating a severe case. Lumbar puncture revealed an albuminocytologic dissociation. Urodynamic investigations objectified a dysuria with important post-void residual urines, detrusor was hypoactive, hyposensitive and hypercompliant.

Evolution was a total recuperation of motor function and persistence of urinary retention after 2 months contrary to what has been said in literature. Urinary disorders were managed by an intermittent auto urinary drainage.

Discussion: Bladder dysfunction may be the first manifestation of GBS and urinary retention is the most frequent of these disorders like in the case of our study and is likely secondary to sacral parasympathetic nerve dysfunction, but a detrusor over activity can be observed whose mechanism is unclear.

It is important to be aware of these disorders which lead to uro nephrologic complications and need a treatment as soon as possible until complete recovery.

Conclusion: Bladder dysfunction prognostic in GBS was good. A total disappearance of symptoms is constant but the period differs in the literature.

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Abstract – WCN 2013

No: 1875

Topic: 10 – Neurorehabilitation

Neuromuscular electrical stimulation applied to upper limb flexor muscles of tetraplegics: Interlimb reflex response assessed by electromyography – Pilot study

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Background: Some researchers report that if lower limbs are stimulated, there is a reflex response in the upper limbs, but little research has been done on the inverse response in tetraplegic subjects.

Objective: To observe if there are responses in tetraplegic individuals, analyzing reflex activities when applying neuromuscular electrical stimulation.

Patients and methods: Five patients with chronic tetraplegia were selected and divided in to two groups (Group I; Group II). Electromyographic signals were recorded from surface electrodes placed on the contralateral lower limb muscles: *rectus femoris*, *vastus lateralis*, *vastus medialis*, *tibialis anterior*, *gastrocnemius medialis*, *soleus*, *fibularis longus* and *extensor digitorum longus* when neuromuscular electrical stimulation was applied to the *flexor digitorum profundus* and *opponens pollicis*.

This stimulation was repeated five times in order to calculate Root Mean Square (RMS) averages.

Results: The groups were composed of, AIS-B ($n = 2$) and patients, AIS-A ($n = 3$). Group I did not present any signals and group II showed signal in the lower limb muscles. When the muscles were stimulated, in group II, the first subject presented responses in muscles: RF (mean RMS = $93.58\text{mv} \pm \text{SD} = 5.1$), TA (164.36 ± 6.1) and S (100.55 ± 1.89). The second patient presented in all muscles: RF (90.64 ± 3.37), VM (63.91 ± 15.27), VL (136.66 ± 2.61), TA (106.89 ± 6.2), GM (87.06 ± 6.2), S (88.27 ± 3.06) FL (129.38 ± 5.67) and EDL (91.16 ± 3.59).

Conclusion: The presence of electromyographic signals suggests a possible interaction between upper and lower limbs when applying neuromuscular electrical stimulation in upper limbs. However more studies are needed to confirm this theory.

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Abstract – WCN 2013

No: 1769

Topic: 10 – Neurorehabilitation

Conceptualizing war-induced neurological trauma through personalized psychosocial approach

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Background: Traumatic brain injury (TBI) is a major war-related health problem that leads to deficits in executive function, a term used for the cognitive operations responsible for decision making and goal management. This, along with emotional and behavioral deficits that follow TBI, contributes to injured Veterans' reduced ability to return to work or school and to regain satisfactory personal lives.

Objective: The COMPASS (Community Participation through Self-Efficacy Skills Development) program aims at testing a patient-centered intervention framework that can be utilized as a platform for VA community re-integration comparative effectiveness research.

Patients and methods: The COMPASS intervention integrates principles of goal self-management, emphasizing its positive effect on self-efficacy mediating a neurologic trauma consequences. Veterans with mild TBI will be randomized into two groups: the COMPASS group and the supported discharge group.

Results: It is expected that personalized conceptualization of the RCT approach based on *two consecutive months treatment phase for the Veterans enrolled in the intervention group will result in their greater social re-integration into the society.*

Conclusion: Goal-setting and goal management concepts are part of the “natural language” of rehabilitation. It might be argued that re-training returning Veterans with TBI how to self-manage their goals, with appropriate help and support, would essentially treat deficits in executive function. A structured approach to goal self-management would foster greater independence and self-efficacy, help clients to gain insight into goals that are realistic for them at a given time, and help clinicians and clients to work more effectively as true collaborators.

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Abstract—WCN 2013

No: 2084

Topic: 10—Neurorehabilitation

Evaluating efficacy and user's expectations of a virtual reality training system: A multi-centre randomised controlled trial using mixed methods

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Background and objective: Virtual reality (VR), widely used in gaming, has now found its way into neurorehabilitation. The aim of this study is the efficacy evaluation of a VR training system including two nested qualitative evaluations of patients' and therapists' perspectives.

Methods: Sixty stroke patients will be randomly allocated to an experimental VR (EG) or a control group using conventional physiotherapy or occupational therapy (16 sessions, 45 min each). Using custom data gloves, patients' finger and arm movements are displayed in real-time on a monitor and move and manipulate objects in various virtual environments. A blinded assessor will test patients' motor and cognitive performance twice before, once during and twice after the intervention using the Box and Block Test, Chedoke–McMaster stroke assessments (subscale hand, arm shoulder pain), Chedoke–McMaster Arm and Hand Activity Inventory, Line Bisection Text, Stroke Impact Scale, Mini-Mental-State Examination, and Extended Barthel Index. Semi-structured interviews will be conducted with patients in EG after intervention finalisation focussing on the expectations and experiences regarding the VR training. Therapists' perspective on VR training will be reviewed in focus groups.

Results: Therapists in all centres completed the study training. Until now, seven patients were recruited (one female, age 66.6 ± 5.3 y). One patient finished the study and took part in a semi-structured interview. After verbatim transcription the data coding and categorisation based on a phenomenological approach have started.

Conclusion: The study protocol including the patients' interviews seem to be feasible and patients enjoy the VR training. The study is registered with clinicaltrials.gov: NCT01774669.

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Abstract—WCN 2013

No: 2700

Topic: 10—Neurorehabilitation

Outcomes related to mobility in patients treated with botulinum toxin a (BoNT-A) for post-stroke upper-limb spasticity (ULIS-2-Study)

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Background: The impact of upper-limb spasticity (ULS) on mobility is debated.

Objective: To describe the characteristics of patients with mobility-related primary goals compared to those with primary goals in other

areas (e.g. passive/active function, pain, impairment). To compare mobility related goal achievement with achievements in these other areas.

Patients and methods: Analysis of 456 adults with post-stroke ULS within an observational, prospective cohort study conducted in 84 centers in 22 countries on current practice of ULS treatment (ULIS-2-Study).

Interventions: One cycle of BoNT-A treatment and concomitant therapy in accordance with routine local clinical practice.

Outcome measures: Primary goal achievement; the Neurological Impairment Scale (adapted for spasticity), composite Modified Ashworth Scale (MAS).

Results: Of 456 adults with post-stroke ULS presenting for treatment with BoNT-A, 45 (9.9%) had a primary goal related to mobility and 411 patients had primary goals in other areas.

Mobility goals were related to involuntary movements during walking (n = 41), balance (n = 3) and transfers (n = 1). Sub-classification of mobility goals also related to speed of walking (n = 3), arm swing (n = 3), independence (n = 1), walking efficiency (n = 4), cosmetic appearance (n = 3) and dressing (n = 1). Baseline demographics and total BoNT-A dose were similar for the two groups.

33/45(73%) achieved their mobility goal compared with 330/411(80%) in the group with other goals (Chi-squared p = 0.33).

Conclusions: Treatment of ULS with BoNT-A can lead to improved mobility by reducing involuntary movements. Improved understanding of the impact of ULS on mobility might lead to a higher rate of goal achievement.

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Abstract—WCN 2013

No: 2188

Topic: 10—Neurorehabilitation

“REVIS”: Restoration of vision after stroke using non-invasive alternating and direct current stimulation

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Non-invasive brain stimulation using alternating current stimulation (ACS) has recently been shown to improve vision after optic nerve damage. It is assumed that transorbital ACS induces neuronal networks to propagate synchronous firing probably activating residual areas surviving the injury. The aim of the ‘Restoration of Vision after Stroke’ (REVIS) network is to determine if bilateral transcranial direct current stimulation (tDCS) and transorbital ACS may also have a positive impact on vision restoration in patients with post-chiasmatic visual pathway lesions. In particular, the potential of non-invasive current stimulation in ameliorating vision impairment following stroke (hemianopia) is the key issue addressed by the REVIS study group.

Within the REVIS network a number of small sample, randomized, controlled, clinical studies including patients with post-chiasmatic lesions due to monohemispheric, ischemic or hemorrhagic stroke were initiated by clinical partners from Germany (Institute of Medical Psychology and Department Biomedical Magnetic Resonance, University

of Magdeburg), Italy (Department of Neurology, Catholic University of Rome), and Finland (Department of Neurology, Helsinki University Central Hospital).

The network also includes a basic neuroscience project that studies neuronal network reorganization in the cat visual system after stroke (Department of Neurophysiology, Nencki Institute, Warsaw). Beyond the European network, a collaboration with the Institute of Automation at the Chinese Academy of Science was established.

This contribution presents the overall aims of the network activity, the functional, physiological and imaging parameters to be studied, and the rationale of the clinical endpoint selection.

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Abstract—WCN 2013

No: 2322

Topic: 10—Neurorehabilitation

Spectral brain mapping in children with cerebral palsy treated by the Masgutova Neurosensorimotor Reflex Integration method

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Background: Rehabilitation in children with neuromotor impairment disorders can be carried out with the use of effective therapeutic methods. The new one is the Masgutova Neurosensorimotor Reflex Integration (MNRI®) method.

Objectives: The aim of this study was to determine the efficiency of rehabilitation carried out with the use of MNRI® in children with cerebral palsy using objective electrophysiological tools.

Patients and methods: The study involved a group of 15 children with cerebral palsy treated by MNRI®. We measured their brain activity by multichannel EEG and analysed the obtained signals with use of the spectral brain mapping.

Results: By analysis of the spectral maps we documented the changes in electric brain activity as an effect of neurorehabilitation.

Conclusion: The rehabilitation carried out by the MNRI® method in children with cerebral palsy causes the reorganisation of the electrical activity of central nervous system network. It may be responsible for positive treatment effect of this therapeutic method.

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Abstract—WCN 2013

No: 2127

Topic: 10—Neurorehabilitation

Disability due to traumatic brain injury in Ukraine (6 years experience)

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Traumatic brain injury (TBI) is one of leading causes of mortality and disability worldwide. In spite of it, no TBI official statistical disability data were collected in Ukraine since 1987.

Methods: 6 years (2007–2012) dynamics of TBI disability incidence rates for adult population (per 10,000) was investigated. Disability

grade (“invalidity” status) in Ukraine is set according to the Ministry of Health Order (no. 561, August 05, 2011) based on ICF (2001) principles. We collected primary disability data structure due to TBI and consequences (ICD X codes S00–S04, S06–S09, S06.0 and T90.5) in Ukrainian regions.

Results: Panukrainian TBI (S00–S09) disability incidence was decreased: 0.72–0.72–0.64–0.59—accordingly, as well as the index for TBI consequences (T90.5): 0.99–0.97–0.91–0.84–0.47–0.58. Unexpectedly, a disability data for codes S00–S04 were revealed (total rates 0.17–0.13–0.13–0.07–0.05–0.05). Another peculiarity was role of mild TBI (S06.0) in intracranial TBI statistics (incidence rate 0.23–0.24–0.18–0.24–0.15, stake ranged 42.1–40.4–35.9–46.0–42.1–30.0%).

Conclusions: Revealed data are the subject of further disability epidemiological investigation of posttraumatic syndromes diagnostic correctness followed disability determination and proper expert-rehabilitation diagnostics.

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Abstract—WCN 2013

No: 1238

Topic: 10—Neurorehabilitation

Effects of virtual reality-augmented training on cortical activation in incomplete spinal cord injury patients: An fMRI study

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Background: Interactive, multimodal sensory stimuli and training using virtual reality (VR) allow the investigation of unique neuroscientific questions and clinical applications.

Objective: In this study we examined the neural correlates of imagination, imitation and interactive execution (playing) of goal-directed lower limb movements and the effects of a 4-week active therapist-supervised training paradigm on these cortical activation patterns.

Patients and methods: Brain activation of incomplete spinal cord injury patients (N = 8) was measured before and after the training period using 1.5 T fMRI. The stimuli were presented in blocks of 30 s. In each block the subject viewed a video (1st-person perspective) of ball juggling with the feet and was instructed to imagine performing the action, imitate the action or play the game. The baseline view was a scrambled version of the videos. The training paradigm consisted of 16–20 interactive training sessions of 45 min each supervised by a therapist.

Results: The playing condition produced broader and stronger activation in prefrontal and subcortical areas than imitation and imagination. The activation patterns of the patients were generally reduced in magnitude after the training, suggesting improved efficiency in recruiting cortical areas to control lower limb movements.

Conclusion: These results may be of significance for VR-augmented lower limb neurorehabilitation and motor therapy. The fact that the cortical changes resulted from active patient VR interactions, with the therapist in a coaching role, suggests that it may be also possible to achieve similar effects with minimal therapist involvement.

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Abstract—WCN 2013

No: 2335

Topic: 10—Neurorehabilitation

The prognostic value of cognitive domain deficits on functional recovery in stroke patients after acute physical rehabilitation

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Background: Few studies investigated what factors may affect functional recovery in post-acute stroke patients treated with a physical rehabilitation program.

Objective: To assess the frequency of domain-specific cognitive deficits and their prognostic value, along with neurological, clinical and demographic predictors, for functional recovery in post-acute stroke patients after discharge from a Neurorehabilitation Department.

Patients and methods: This outcome research study included 226 stroke patients (M = 121, F = 105; mean age 75.9 ± 10.7 yrs; education 8.1 ± 4.1 yrs). Patients were assessed at admission and at discharge using the Functional Independent Measure (FIM), and through a comprehensive neurological and neuropsychological evaluation (i.e. reasoning ability, attention and executive functions, memory, visuo-spatial abilities, and language). The main outcome considered is functional recovery, defined as change in FIM score from admission to discharge.

Results: Total FIM score at baseline was 62.65 ± 25.3, and FIM cognitive and motor sub-scores were respectively 23.89 ± 8.35 and 38.74 ± 19.21. At the time of discharge FIM score and FIM motor and cognitive sub-scores were respectively 84.58 ± 31, 26.67 ± 8.33 and 57.88 ± 24.53. A linear regression model, considering FIM at discharge as the dependent variable, and baseline motor and cognitive FIM sub-scores, premorbid functional independence, and reasoning ability as main independent variables, was used to assess if neurological, clinical and cognitive domains independently affect functional recovery. The regression model produced an R square value of 67%.

Conclusions: Baseline motor and cognitive status, premorbid functional independence, and reasoning ability account for 67% of the total FIM score variance at the time of discharge in patients admitted for post-stroke rehabilitation.

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Abstract—WCN 2013

No: 979

Topic: 10—Neurorehabilitation

Impact of therapeutic input on goal achievement in patients treated with botulinum-toxin a (BoNT-A) for post-stroke upper-limb-spasticity (ULIS-2 study)

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Background: Current guidelines on treating upper-limb spasticity (ULS) with BoNT-A emphasize a combined approach (BoNT-A plus concomitant therapies (CT)) as best practice, although type and intensity of CT remains open.

Objective: To compare the impact on goal achievement in patients with lower versus higher therapeutic input (TI) after BoNT-A treatment of ULS.

Design: Subgroup analysis of 451 adults with post-stroke-ULS within an observational, prospective cohort study on current practice in an international, multi-center design in 84 centers in 22 countries (ULIS-2).
Interventions: One cycle of BoNT-A treatment and CT in accordance with routine local clinical practice.

Higher TI was defined as more than 10 sessions of associated physiotherapy and/or occupational therapy between BoNT-A treatment and follow-up.

Outcome measures: Primary goal achievement in relation to higher versus lower TI.

Results: The median (range) follow-up time was 14(2.6–32.3) weeks. Of 451 adults with post-stroke upper limb spasticity presenting for BoNT-A treatment, 238(52.8%) received higher TI and 213(47.2%) received lower TI.

Goal achievement in the higher TI group was 83.6% (95%CI:78.3%, 88.1%) and in the lower TI group 74.6% (95%CI:68.3%, 80.3%).

Conclusions: BoNT-A treatment combined with higher TI led to a higher rate of goal achievement. However, a substantial proportion of those with low TI still achieve their goals indicating the high pharmacological impact of BoNT-A treatment in ULS.

Further research is required to examine the characteristics of patients and goal areas, which demand higher TI. Type and intensity/frequency of TI has to be reviewed to develop an algorithm for appropriate CT following BoNT-A injection.

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Abstract—WCN 2013

No: 2289

Topic: 10—Neurorehabilitation

The modified modified ashworth scale (MMAS) in the assessment of muscle spasticity: Intra-rater reliability

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Background: The MMAS is a new clinical scale for measurement of muscle spasticity. There is little information regarding the intra-rater reliability of MMAS.

Objective: The purpose of this study was to determine the intra-rater reliability of the MMAS in the evaluation of upper limb muscle spasticity in adult patients with stroke.

Material and methods: In this study with a clinical measurement design, one rater randomly graded the spasticity of shoulder adductors, elbow flexors, and wrist flexors in the affected upper limb of 24 patients with hemiparesis twice with at least a 1 week interval to establish intra-rater reliability. The intra-rater quadratic weighted kappa was calculated.

Results: The quadratic weighted kappa was very good for elbow flexors and wrist flexors, and good for shoulder adductors.

Conclusion: The intra-rater reliability of the MMAS was acceptable for measuring muscle spasticity in adult patients with stroke.

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Abstract—WCN 2013

No: 2215

Topic: 10—Neurorehabilitation

The use of biomechanical visualisation in neurorehabilitation and its effect on ankle-foot orthosis (AFO) tuning in stroke

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Background: Difficulties interpreting the biomechanical data captured by 3D gait analysis (3DGA) systems mean that it is rarely used in routine clinical practice for gait rehabilitation of stroke patients. Biomechanical visualisation software has been designed to make data clinically useful in the context of AFO fitting and tuning for stroke patients. Therapists can use objective quantitative data to assist clinical decision-making. Participants receive an improved understanding of their condition/treatment, and it allows progress tracking.

Objective: Test the hypothesis that stroke patients will receive improved outcomes when biomechanics visualisation is used in the AFO fitting and tuning process.

Patients and methods/material and methods: An RCT is being used (ISRCTN52126764). The intervention arm receive AFO fitting and tuning using 3DGA and visualisation, and the non-intervention arm receive an AFO by standard care (clinicians using observation). Walking velocity, 3D kinematics and kinetics, step length, gait symmetry, mRMI and EuroQol (EQ-5L-5D) are measured at four time points (baseline, AFO provision, three months and six months). Ten participants, 5.7(6) weeks post-stroke, with an average age of 56.4(17) years have been recruited.

Results: Walking velocity improvement (before/after AFO provision) data for the intervention group (n = 5) was 22(21) cm/s versus 1.6(6.4) cm/s for the non-intervention group (n = 5). The difference is significant (p < 0.05, Mann–Whitney U Test). More extensive results will be available at the time of presentation.

Conclusion: Early data indicate that the visualisation of biomechanical data appears to assist the AFO tuning process, providing stroke patients with better immediate improvements in walking velocity.

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Abstract—WCN 2013

No: 2246

Topic: 10—Neurorehabilitation

Assessment of heart rate variability during complex stimulation in patients with disorders of consciousness: Preliminary results

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Background: Autonomic changes can be induced by different stimuli in patients with disorders of consciousness (DOC) and provide clues on their responsiveness.

Aim: To assess the changes in heart rate variability (HRV) induced by auditory/musical stimuli in patients with DOC.

Materials and methods: The HRV of 5 patients with DOC (3 males, age 35.8 ± 15, LCF = 2.4 ± 0.55) was assessed 9 ± 2 months after severe brain injury (SBI). Patients were assessed while they rested in a wheel chair and listened to a neutral story (NN), a series of autobiographical data read with neutral tone (NA), the story of an episode emotionally important for them (EE), him/her favorite song played live (CdC) and the description of the patient written by his/her relatives (DDL) with musical enrichment. The stimulation was preceded by a baseline of 10 min during which the patient heard

only the sound of a stream (baseline). The patients were assessed twice: the first time, the order of stimuli followed an increasing emotional level and complexity; the second time, the order of stimuli was random.

HRV was derived by a 3 lead ECG using autoregressive (AR) algorithm.

Results: LFnu, HFnu and LF/HF ratio did not differ during baseline, NN, NA, EE, and CdC, irrespectively of the order of stimuli administration. The LF/HF ratio tended to increase during increasing emotional level and complexity stimuli, but was statistically different only during DDL compared to baseline ($p = 0.028$).

Conclusion: Enriched stimulation in an immersive environment increased the sympatho-vagal balance of DOC patients.

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Abstract—WCN 2013

No: 1195

Topic: 10—Neurorehabilitation

Clinical efficacy of non-invasive transorbital alternating current stimulation in optic neuropathy: A double-blind, randomized, sham-controlled multi-center study

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Introduction: Non-invasive brain stimulation enhances neuronal plasticity in the visual system in normals and patients with visual field loss (Antal et al., RNN 2012; Sabel et al., RNN 2011) with one of the proposed mechanisms being the neuromodulation of oscillatory brain activity (Schmidt et al., Brain Stimul 2013).

Objectives: In order to improve visual functions after pre-chiasmatic damage to the visual pathway due to various reasons including glaucoma and post-neuritic optic atrophy the efficacy of repetitive transorbital alternating current stimulation (rtACS) was evaluated in a randomized, multi-center clinical trial.

Methods: Ninety-eight patients were randomized into rtACS and sham-group both treated with 10-daily sessions (20–40 min). Outcome measures included visual fields and resting-EEG obtained before and after stimulation, and at 2-months follow-up.

Results: Eighty-two patients completed the study. While rtACS-treated patients ($n = 45$) showed improvements in visual detection rates (Hodges-Lehmann-estimator median increase 6.4%, 95%-CI(2.9%,11.6%); $p < 0.001$), no change was observed after sham-stimulation ($n = 37$, 1.1%,95%-CI(−2.0%;4.3%); $p = 0.256$). The primary endpoint, percentage change of detection, was significantly better in rtACS compared to sham ($p = 0.011$; one-sided U-test). Secondary analysis revealed pronounced visual field improvement in the central 5° visual field. Visual field improvements were significant at 2-months follow-up. EEG power changes were observed in the alpha band.

Conclusions: RtACS led to visual field improvement in patients with optic neuropathies. Together with EEG powerspectra change this finding indicates that residual vision activation is associated with plastic alterations in neuronal brain networks and that rtACS is a valid method to partially restore vision loss after optic nerve damage.

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Abstract—WCN 2013

No: 2179

Topic: 10—Neurorehabilitation

MR diffusion tensor imaging and fiber tracking in spinal cord lesions

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Background: Magnetic resonance diffusion tensor imaging (MR DTI) can be used to visualize neural pathways within white matter (tractography or fiber tracking). While fiber tracking is of increasing importance in the assessment of brain lesions like space-occupying tumors, experience with spinal cord lesions is limited primarily due to technical limitations.

Objective: To evaluate the feasibility of MR fiber tracking to detect spinal cord lesions.

Patients and methods: In four men (58 +/− 17 years) with spinal cord lesions (cervical spinal cord infarction—ASIA D; compressive cervical myelopathy due to epidural abscess—ASIA D; thoracic herpes zoster myelitis—ASIA C; compressive thoracic myelopathy due to disk herniation—ASIA C) and with pathological somatosensory and motor evoked potentials MR-DTI was performed on a Philips Achieva 3T MRI scanner. Furthermore, a 21 year old man with suspected psychogenic paraparesis who had normal MRI of the spinal cord and normal somatosensory and motor evoked potentials was examined.

Results: In one patient DTI data could not be analyzed due to movement artifacts. In the other three patients fibertracking showed rarification or disconnection of fibers at the level of injury. No discontinuity of fibers was found in the patient with psychogenic paraparesis.

Conclusion: MR fiber tracking is capable to detect lesions of neural tracts in spinal white matter. Quantification of lesion extent and correlation with clinical and electrophysiological data is part of ongoing studies.

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Abstract—WCN 2013

No: 2199

Topic: 10—Neurorehabilitation

Stroke and aphasia quality of life scale (saqol-39). Evaluation of acceptability, reliability and validity of Chilean version

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Background: 21 to 38% of stroke has aphasia. Quality of life became a useful indicator in rehabilitation in medicine. There are few instruments to evaluate psychosocial aspect in aphasic patients.

Objective: To evaluate acceptability, reliability and validity of SAQOL-39 to Chilean aphasic population.

Method: The cross-cultural adaptation of the SAQOL-39 into Chilean Spanish was making following translation and back translation method. 31 patients were tested with SAQOL-39 using Boston Aphasia test (BAT), General Head questionnaires (GHQ) and Barthel index. Acceptability was evaluated by examining the floor/ceiling effects and the missing data. The reliability was assessed by Cronbach's alpha and intraclass correlation coefficients.

Results: 61.5 % male, 70 % of the patient must be retired after stroke. 42% has memory loss. They are not difficult to translate in the English version. The median Boston scale was 3, Barthel index 95 and GHQ-12 = 4. The median SAQOL-39 was 3.6. There good acceptability demonstrated by 2.4% of missing data, 66.7% of ceiling effects and

12.8 % of floor effect. Test re-tests reliability for overall score and the sub scales scores was 0.95(0.76 to 0.95). The Internal consistency analysis by Cronbach's was 0.947.

Conclusion: The Chilean version of SAQOL-39 seems appropriate for clinical use with adequate acceptability, reliability and validity.

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Abstract—WCN 2013

No: 2221

Topic: 10—Neurorehabilitation

Task specific, robot-assisted training in patients with impaired upper limb motor functions—A clinical investigation

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Background: Literature showed that robot-assisted arm therapy (RAT) is a safe treatment method that improves upper limb motor function in patients after stroke. There is a need to evaluate the importance of therapy structure, content, and individual adaptation for clinical implementation. There is insufficient evidence for the application of RAT for further pathologies, e.g. Parkinson's disease and multiple sclerosis.

Objective: To conduct arm therapy using the ARMin III robot technology in a clinical setting with patients with various motor impairment diseases. Evaluation of objective motor function and subjective experiences reported by patients and therapists.

Patients and methods: Explorative study investigating RAT in patients with various diagnoses (myopathy, neuropathy, multiple sclerosis, cerebral palsy, Parkinson's disease, amyotrophic lateral sclerosis, status after stroke, traumatic brain injury, neuro-surgical intervention, spinal cord injury, or fractures of upper limbs after consolidation. RAT intervention applied 1 to 3 ×/week for 30–45 min. Pre- and posttesting include objective and subjective assessments depending on diagnosis. Data analysis will focus on therapy parameters.

Results: Study was approved by the responsible ethics committee. Patient recruitment is ongoing. Occupational therapists are being trained to apply RAT receiving comprehensive training sessions. Three pilot treatments were conducted with patients. Additionally, one patient (70 years old, male) was treated once during a training session; one male (58 years old) received 9 RAT, one female (33 years old) received 10 treatments.

Conclusion: Intensive training of therapists is necessary. Clinical implementation seems feasible. Patients enjoyed RAT. Active range of motion and increased muscle tonus seems to be positively influenced by RAT.

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Abstract—WCN 2013

No: 2138

Topic: 10—Neurorehabilitation

Errors in tool use reflecting insufficient recall of the target object for tools

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Background: There has been reported several types of errors in the patients with impairment in tool use. Some types of errors seem to indicate the underlying brain dysfunction. For example, content errors may stem from the conceptual degradation of tools. However, there still remain various errors which cannot be accounted for.

Objective: The purpose of the study is to identify the new type of errors of tool use.

Patients and methods: We administered the tasks of single tool use and sequential plural tool use to three patients who showed impairment of tool use due to the left parieto-temporal lesion, and analyzed the error patterns.

Results: All the patients did not show any sequential nor content errors, however demonstrated a peculiar pattern of error. They could reach and grasp the tool, and the movements for tool use were properly performed without any problems. The only unusual point of the performances was mistaking the objects the tool gives function for the proper ones. For example, “correctly striking a match on incorrect side of the matchbox.” As a result, they could not have accomplished these tasks.

Conclusion: This error pattern in tool use can be deduced to be ascribed to insufficient recall of the target object. This is a new type of errors in impairment of tool use suggesting that there are the mechanisms of retrieval of concept not only the tool itself but also the concept of the target object the tool will act on.

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Abstract—WCN 2013

No: 2135

Topic: 10—Neurorehabilitation

Islands of consciousness in vegetative state or functional locked-in syndrome?

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Background: In the last five years residual cortical processing in the absence of behavioural signs of consciousness has been demonstrated in some non-communicative severely brain damaged patients, by means of functional neuroimaging and electrophysiology studies. The term “functional locked-in syndrome” has been proposed for patients with a dissociation between extreme motor dysfunction and preserved higher cortical functions, only identified by functional imaging techniques.

Objective: Patients clinically diagnosed as being in the vegetative state who are able to perform mental imagery tasks are still diagnosed as in the vegetative state with preserved islands of consciousness, not as having functional locked-in syndrome. Our proposal is that functional neuroimaging and electrophysiology should be used in doubtful cases of disorder of consciousness (DOC) to rule out tests for the diagnosis of vegetative state in the presence of residual cognitive functions, at least when performance of complex mental imagery tasks and intentional communication ability has been demonstrated.

Material and methods: Some video recordings of cases during recovery from vegetative state who present with eyes-based communication, together with fMRI data during resting state and after emotional stimulation will be discussed.

Conclusion: The scientific community should finally clearly state that clinically “vegetative patients”, in whom complex mental imagery tasks and intentional communication ability have been demonstrated by means of fMRI or neurophysiology, are not vegetative or unresponsive,

but subjects with total functional locked-in syndrome, clinically misdiagnosed as vegetative state.

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Abstract—WCN 2013

No: 2148

Topic: 10—Neurorehabilitation

Psychiatric and cognitive disorders in neurological patients

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Introduction: Apart from a multitude of physical complaints, neurological disorders can also lead to various kinds of psychiatric and cognitive disorders like mental and cognitive changes or reduced Quality of Life (QoL). In such cases, mental health, mood, attention, vigilance and/or memory may be affected as well as the actual mental and/or cognitive processes themselves. Important factors in this can be the severity of the disorder on one hand, and the duration of prior therapy on other hand.

Methods: The study was carried out involving two groups of randomly selected persons, neurology patients and healthy participants. All patients were selected according to their clinical diagnosis (ICD-10).

So far, data have been gathered more than 50 healthy persons (42 male; 9 female) and around 150 neurological patients (101 male; 47 female) (with various neurological clinical pictures) using different tests to research the psychiatric/mental and cognitive status as well as the QoL.

Findings: Testing of psychiatric, cognitive and QoL achievements revealed highly significant differences between healthy persons and neurological patients (all parameters: $p < .001$). Analysis of the degree of severity showed for neurology patients no significant differences between mild and severe status ($p > .050$).

Discussion: The study revealed that patients with neurological diseases (strokes, cerebrovascular diseases, brain traumas, brain tumors, etc.) show problems, deficits and disorders concerning in different areas of psychiatric/mental and cognitive achievements as well as in multidimensional QoL. In contrast, the degree of severity of the disorders (neurology patients) was not relevant.

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Abstract—WCN 2013

No: 2075

Topic: 10—Neurorehabilitation

The effect of short-term rehabilitation for patients with spinocerebellar degeneration

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Background: The effect of rehabilitation has not been well analyzed in patient with spinocerebellar degeneration (SCD).

Objectives: The aim of this study is to clarify the effect of short-term intervention of rehabilitation for patients with SCD.

Patients and methods: We enrolled 10 SCD patients (Multiple System Atrophy (MSA) with predominant cerebellar ataxia 8 case, MSA with predominant parkinsonism 1 case, Spinocerebellar ataxia 1 case). The ages at onset and duration of disease were 58.6 ± 11.6 years old

and 4.9 ± 1.9 years, respectively. Rehabilitation was carried out for 12.7 ± 4.3 days. The rehabilitation program was composed of multidimensional exercises such as coordination exercise, proprioceptive neuromuscular facilitation, and mental practice. To evaluate the effect of our program, we carried out measurement of the following four scales before and after the program; 1) Scale for the Assessment and Rating of Ataxia (SARA), 2) Functional balance scale (FBS), 3) Ten meter walk test, and 4) MSA quality of life (MSAQOL). 4) Center of mass \times direction (COMx) trajectory.

Results: The score of SARA (12.7 ± 5.7 to 10.4 ± 4.2 , $p < 0.05$) and FBS (39.3 ± 8.6 to 45.1 ± 6.7 , $p < 0.05$) and gait velocity (40.0 ± 17.6 to 48.8 ± 13.7 m/min, $p < 0.05$) and MSAQOL (43.6 ± 26.8 to 35.8 ± 25.3) were improved after rehabilitation. COMx trajectory was improved from 109.5 ± 39.1 to 92.7 ± 41.1 mm ($p < 0.05$).

Conclusion: Short-term rehabilitation shows beneficial effects in motor function and QoL of patients with SCD.

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Abstract—WCN 2013

No: 2091

Topic: 10—Neurorehabilitation

A case report: Neurorehabilitation results in a patient suffering from a “lock-in” syndrome

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Background: The 44 year old female patient was hospitalized and treated of disseminating encephalomyelitis. Because of the area affected (pons and crus cerebri) she developed a rarely seen syndrome that included difficulty speaking, swallowing, quadriplegia and incontinence while maintaining full time awareness—the “lock-in” syndrome (LIS).

Objective: The goal was to rehabilitate the patient in a way to establish as much as her previous somatic state as possible.

Material and methods: A patient is a 44 year old white female whose therapy consisted of analgetics, antibiotics, antimicrotics, gastroprotectives, antiedematose, anticoagulant and antidepressant therapy as well as intravenous potassium replacement, nutrition supplement, a one time blood transfusion and every-day physical rehabilitation therapy. Diagnostics included lumbar puncture, spinal tap and hemoculture analysis, heart and lungs Rtg scan, brain MRI, cervical and thoracic spine, transcranial ultrasound, PCR method and hereditary thrombophilia testing. The nature of illness also requested consultation of other medical experts.

Results: Hospitalization lasted three months during which the patient regained speech and communication skills and after additional neurorehabilitation was able to fully take care of herself.

Conclusion: One of the problems with LIS is the lack of literature concerning its course and prognosis. However, the survival rate for LIS found in Doble et al. (2003) study was 83% for 5 and 10 years and 40% for a 20 year survival rate. So far our patient is in the first 83% and we hope she will continue enduring in order to reach the 20 year survival group.

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Abstract—WCN 2013

No: 1990

Topic: 10—Neurorehabilitation

Biomarkers for prediction of hand function after stroke — The ProHand study protocol

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Improved prediction of recovery after stroke has broad implications for clinical decisions on the type, duration and goals of rehabilitation. Increased understanding of prognostic factors may offer new targets for specific interventions. Almost half of surviving stroke patients are left with impaired function of the hand but individual profiles of weakness, spasticity, sensory and bimanual function vary widely. Combining behavioural and imaging measures can improve prediction of motor recovery (Stinear et al, 2012). In this study we use novel quantitative behavioural measures together with structural and functional MRI measures to understand hand function recovery. We hypothesize that the structural and functional brain connectivity in the early phase after stroke can predict (i) the hand function profile in the late phase and (ii) degree of recovery.

One hundred stroke patients with early signs of upper limb hemiparesis will be recruited at our neurorehabilitation clinic. Imaging and behavioural assessments will be performed at 4 weeks, 3 months and 6 months after stroke. MRI includes anatomical sequences for lesion mapping and diffusion tensor imaging and resting-state functional MRI for structural and functional connectivity analyses, respectively. Clinical hand function measures will be combined with novel validated methods to quantify spasticity, grip force control, sensory and bimanual function. The study protocol along with preliminary data will be presented. ProHand will provide insights into neural mechanisms related to recovery of hand function after stroke. Imaging and behavioural markers important for recovery will be identified which should enable enhanced prediction of prognosis and development of targeted interventions.

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Abstract—WCN 2013

No: 2066

Topic: 10—Neurorehabilitation

MoVo-LISA – implementing a short movement coaching programme to establish a physical active life style: A feasibility study

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Background and objectives: For patients after a disease of the central nervous system (CNS) it is difficult to regain an active life style. The short movement coaching programme MoVo-LISA (Motivation Volition-life style integrated sporting activity) has shown beneficial short and long-term effects in patients with osteoarthritis and obesity. The aim of the study was to implement and evaluate MoVo-LISA in patients with a disease of the CNS.

Methods: A case series with a pre-post design was conducted. Before and six weeks after the intervention patients were assessed with the MoVo-LISA-specific questionnaire including subscales for: current complaints, activities of daily living (ADL), positive and negative consequence expectations, self-efficacy expectation (SEE), goal intention (GI), planning (P), perceived barriers, action and relapse strategies (AS + RS), and social support. The MoVo-LISA-programme included three sessions:

- (1) group session (60 min) to summarise health goals and movement ideas,

- (2) individual session (10 min) to refine the activity plan,
- (3) group session (90 min) to discuss barriers and establish an activity protocol for the upcoming six weeks.

After three and six weeks patients received a letter and a telephone call reminder.

Results: Totally, 13 patients (age $57.9 \pm 15.4y$, 9 females, 5 stroke patients) were included. ADL reduced about 79 min/week, sporting activity increased about 204 min/week. MoVo-LISA-questionnaire results revealed positive trends regarding SEE, GI, P, AS.

Conclusion: The MoVo-LISA-programme was feasible in groups of five or less. Patients needed help to fill in the questionnaires. It might be considered including information on training options and shift the intervention start to 4 weeks after entry.

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Abstract—WCN 2013

No: 2008

Topic: 10—Neurorehabilitation

Functional connectivity network breakdown and restoration in blindness

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Background: Loss of vision after brain damage is thought to be caused mainly by the primary tissue loss. Little is known how the damage affects interactions in widely distributed brain networks and how this, in turn, contributes to vision loss.

Objective: We now studied functional connectivity in the brain of partially blind subjects to delineate the role of wide range neuronal networks in blindness.

Methods: Resting state eyes-closed EEG activity was recorded in patients with partial optic nerve damage ($n = 15$) and uninjured controls ($n = 13$). Power density and functional connectivity (coherence, Granger Causality) were analyzed, the latter as (i) between-areal coupling strength and (ii) individually thresholded binary graphs. Functional connectivity was then modulated by non-invasive repetitive transorbital alternating current stimulation for 10-days (rtACS for 40 min daily; $n = 7$; sham, $n = 8$) to study how this would affect connectivity networks and perception.

Results: Blind patients had lower EEG spectral power ($p = 0.005$) and decreased short- ($p = 0.015$) and long-range ($p = 0.033$) coherence in the high-alpha EEG band (11–14 Hz) and less densely clustered coherence networks ($p = 0.025$). rtACS strengthened short- and long-range coherence again which correlated with recovery of detection ability ($r = 0.57$, $p = 0.035$) and processing speed ($r = 0.56$, $p = 0.049$).

Conclusion: Vision loss in the blind is not only caused by primary tissue damage but also by a synchronization breakdown of short- and long-range connections in brain networks. Because re-synchronization of alpha band coherence is associated with visual field improvements, brain connectivity is a key component in restoration of vision in partial blindness.

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Abstract—WCN 2013**No: 2025****Topic: 10—Neurorehabilitation****The second face of blindness: Processing speed deficits in the intact visual field after pre- and post-chiasmatic lesions**M. Bola, C. Gall, B.A. Sabel. *Institute of Medical Psychology, Otto von Guericke University of Magdeburg, Magdeburg, Germany*

Background: Damage along the visual pathway results in a visual field defect (scotoma) retinotopically corresponding to the damaged tissue. But other parts of the visual field, processed by the uninjured tissue and believed to be intact, suffer from perceptual deficits as well.

Objective: We studied features of the visual field and scotoma in patients to elucidate factors predicting intact field deficits.

Methods: Patients with pre- (n = 53) or post-chiasmatic lesions (n = 98) were tested with high resolution perimetry – a method used to map visual fields with supra-threshold light stimuli. Reaction time (RT) of detections in the intact visual field was then analyzed as an indicator of processing speed and correlated with features of the visual field defect.

Results: Patients from both groups exhibited processing speed deficits in their presumably “intact” field when compared to a normative sample ($p < 0.001$). Further, processing speed was found to be a function of two factors. Firstly, a spatially restricted (retinotopic) influence of the scotoma was seen in longer RT when stimuli were presented in intact field sectors close to the defect. Secondly, patients with larger scotomata had on average longer RT in their intact field indicating a more general (non-retinotopic) influence of the scotoma.

Conclusions: Visual system lesions have more widespread consequences on perception than previously thought. Because dysfunctions of the seeing field are expected to contribute to subjective vision, including visual tests of the presumed “intact” field may help to better understand vision loss and to improve methods of vision rehabilitation.

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Abstract—WCN 2013**No: 584****Topic: 10—Neurorehabilitation****Care assistant support following specialist rehabilitation for people with Parkinson's and carers in the community: Findings from the SPIRIT RCT**H. Gage^a, L.D. Grainger^a, S. Ting^a, P. Williams^b, C. Chorley^a, G. Carey^a, N. Borg^a, K. Bryan^c, B. Castleton^d, P. Trend^e, J. Kaye^f, S. Khan^g, D. Wade^h. ^a*School of Economics, University of Surrey, Guildford, UK;* ^b*Department of Mathematics, University of Surrey, Guildford, UK;* ^c*School of Health and Social Care, University of Surrey, Guildford, UK;* ^d*The Runnymede Hospital, Chertsey, UK;* ^e*Department of Neurology, Royal Surrey County Hospital, UK;* ^f*GPDRG, University of Surrey, Guildford, UK;* ^g*Surrey Clinical Research Centre, University of Surrey, Guildford, UK;* ^h*Oxford Centre for Enablement, Oxford, UK*

Background: Multidisciplinary rehabilitation gives short term benefits in Parkinson's but these are not sustained.

Objectives: To assess (1) if ongoing support from trained Parkinson's care assistants (PCA) is effective at sustaining benefit from time-limited rehabilitation; (2) costs; and (3) acceptability.

Patients and methods: Pragmatic three parallel group randomised controlled trial, Surrey, England; people with Parkinson's (all stages), and live-in carers. Groups A and B received domiciliary specialist rehabilitation from a multidisciplinary team (MDT), tailored to individual needs (about 9 h per patient). In addition, Group B received ongoing support for a further four months from a PCA, embedded in the MDT (1 h per week). Control group (C) receives care as usual (no coordinated MDT care or ongoing support).

Results: 269 people with Parkinson's (155 live-in carers) were analysed at baseline [A, n = 88 (52); B, n = 88 (50); C, n = 93 (53)]. Significant improvements for patients arose at the end of the MDT intervention in disability, non-motor symptoms, health related quality of life, psychological wellbeing, and speech. Over 6 months, sustained marginal improvements were recorded in mobility (Group A vs. C, $p = .063$), psychological wellbeing and speech (Group B vs. Groups A and C, $p = .085, .047$ respectively). Carers in B recorded reduced strain (B vs. A, $p = .065$; B vs. C, $p = .041$), during the PCA intervention. Cost per patient: £833 (MDT); £600 (PCA). Participants welcomed the intervention and reported knowledge gains.

Conclusions: Support from PCA benefits carers. Further research on sustaining benefits from rehabilitation is needed.

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Abstract—WCN 2013**No: 2043****Topic: 10—Neurorehabilitation****Importance of intervention program for the promotion of physical activity in the perception of illness in people with multiple**L. Pedro^a, J.L. Pais Ribeiro^b, J. Pascoa Pinheiro^c. ^a*Rehabilitation, Polytechnic of Lisbon, ESTESL, Lisbon, Portugal;* ^b*Psychology, University of Porto, Porto, Portugal;* ^c*Rehabilitation, University of Coimbra, Coimbra, Portugal*

Multiple sclerosis (MS) is a chronic neurological disease. The promotion of physical activity can be an important factor on the perception illness of these patients. The aim of this study is to examine the perception of disease of an implementation of a program of physical activity, with patients with MS.

Methods: This is a prospective study. We use the question “Please classify the severity of your disease” The study includes 24 patients with MS diagnosis according to relevant medical criteria, between 20 and 58 years, mean age 44 years, 58.3% women, 37.5% currently married, 67% retired, mean school level of 12.5 years, being diagnosed at least 1 year ago, and EDSS score under 7. Intervention followed the recommendations of Helsinki Declaration.

The program consists of an intervention to promote physical activity in a group of eight people, in once a week sessions of 90 min. The program was held for seven weeks. We use the program statistic SPSS version 18, the General Linear Model, repeated measure.

Results: Comparing the results obtained from the assessment before the intervention program (time A) to promote physical activity and the end of the program (time B), we find that (Time A): $\underline{M} = 5.46$, $\underline{DP} = 2.13$ (Time B) $\underline{M} = 5.04$, $\underline{DP} = 2.01$ ($F = 8.01$; $p < 0.009$).

Conclusions: There are statistically significant differences between the two moments of assessment, which suggest that physical intervention programs for promotion of physical activity can play an important role in the perception of disease severity, in EM.

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Abstract—WCN 2013**No: 1951****Topic: 10—Neurorehabilitation****Changes in intrathecal baclofen dosage during long-term treatment in patients with spasticity due to traumatic spinal cord injuries**B. Skoog^a, L. Hylliemark^b, B. Hedman^b, L. Rutberg^a, A.-K. Karlsson^a. ^a*Institute of Clinical Neuroscience and Physiology, Gothenburg, Sweden;* ^b*Clinical Neuroscience, Karolinska Institutet, Stockholm, Sweden*

Background: Intrathecal baclofen is an effective treatment for severe leg spasticity but tolerance was reported during short term treatment. **Objective:** To investigate dosage changes during long-term treatment. **Methods:** The 24 patients included in the study had spinal cord injuries and had been treated with an intrathecal baclofen pump (Syncromed) because of severe leg spasticity for at least 7 years. Seven patients were excluded from the study due to factors which aggravated spasticity. Of the remaining 17 patients 13 had cervical and 4 had thoracic spinal cord injury ASIA A (n = 6), ASIA B (n = 5), ASIA C (n = 3) and ASIA D (n = 3). The effect was estimated by patient/doctor dialogue, usually 4–10 times per year, in order to reduce leg spasticity and at the same time avoid an increase in leg/trunk weakness.

Results: One year after the start of intrathecal baclofen the patients needed a median dose of 264 µg baclofen per 24 h (range 30–383) in order to reach an optimal effect. After an additional year the mean dose had not changed (95% CI +/- 6%). Seven years after the start of baclofen the dose had increased by 9% compared to the first year dose (95% CI +/- 12%), after ten years by 17% (95% CI +/- 15%, n = 14) and after fifteen years by 34% (95% CI +/- 25%, n = 6).

Conclusion: The dose which needed to reach an optimal effect on leg spasticity was not increased significantly after seven years, while it was slightly increased beyond this time.

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Abstract—WCN 2013

No: 1939

Topic: 10—Neurorehabilitation

A standardised motor imagery introduction program (MIIP) for patients with sensorimotor impairments: Development and evaluation

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Background and objective: A solid motor imagery (MI) introduction is crucial to understand and use MI to improve motor performance. The study's aim was to develop and evaluate a standardised MI group introduction program (MIIP) for patients after stroke, multiple sclerosis (MS), Parkinson's disease (PD), and traumatic brain injury (TBI).

Methods:

- Phase 1: Development of a MIIP comprising MI theory (definition, type, mode, perspective, planning) and MI practice (performance, control).
Phase 2: Development of a 27-item self-administered MI questionnaire to evaluate MIIP: MI knowledge (12 items), MI ability self-evaluation (5 items), MIIP satisfaction (10 items).
Phase 3: Evaluation of MIIP and MI questionnaire by 2 independent MI experts based on predefined criteria and 2 patients using semi-structured interviews.
Phase 4: Case series with a pre-post design to evaluate MIIP (3 × 30 min) using the MI questionnaire, Imaprax (IMA_{vis}), Kinaesthetic and Visual Imagery Questionnaire (KVIQ_{vis + kin}), and Mental Chronometry (MC).

Results: Eleven patients (5 females; age 62.3 ± 14.1 years; Mini-Mental State Examination score 28.4 ± 1.5; 5 stroke, 4 MS, 1 PD, 1 TBI) were included. MI knowledge improved significantly from 5.4 ± 2.2 to 8.8 ± 2.9 (p = 0.01). Self-evaluated MI ability changed from 16.2 ± 4.3 to 18.5 ± 2.4 (p = 0.47). Patients demonstrated high satisfaction with MIIP (40.3 ± 5.6). MI ability remained high: IMA_{vis} 33.3 ± 3.3 to 33.6 ± 3.5 (p = 0.8), KVIQ_{vis} 36.2 ± 10.5 to

37.0 ± 6.2 (p = 0.2), KVIQ_{kin} 33.6 ± 9.7 to 29.7 ± 8.2 (p = 0.1). MC revealed ratios between 0.5 to 1.9 pre-test and from 0.5 to 2 (p = 0.9) post-test.

Conclusion: The MIIP seems to be valid and feasible for patients with sensorimotor impairments resulting in improved MI knowledge. MIIP sessions can be held in groups of four or less.

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Abstract—WCN 2013

No: 1966

Topic: 10—Neurorehabilitation

Neuroprotective role of docosahexaenoic acid and gamma-linolenic acid in lead induced neurological deficit in Swiss albino mice

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Lead is virtually toxic to every organ of the body including central nervous system where it may manifest as encephalopathies and neuropathies, but also various behavioral changes indicative of cerebral dysfunction like; periodic convulsions, irritability, hyperactivity, retardation of normal development, emotional instability, behavioral disorders, low attention span, impaired motor development, and antisocial behavior. The purpose of present study was to characterize the putative neuroprotective role of docosahexaenoic acid and gamma-linolenic acid. PRO-PL (British Biologicals, Bangalore) dietary supplement containing docosahexaenoic acid and gamma-linolenic acid was fed in diet to study neuroprotective role using rota rod apparatus. A total number of 48 adult Swiss albino mice of either sex were included in the study consisting of equal numbers (six each) in Standard, Control, Control + Dietary Supplement and Experimental, and Experimental + Dietary Supplement Groups. Experimental groups received 4.5% and 5% Lead Nitrate and Lead Acetate Trihydrate orally alone and with Dietary Supplement for a period of 3 weeks. Diazepam (1 mg/kg i.p.) was used as the standard drug. All the experimental work was approved by the Institutional Animal Ethics Committee (Ref. No. IAEC/257). In rota rod apparatus, there was significant increase in time spent by the animals on revolving rod whereas; in elevated-plus maze there was significant increase in time spent and number of entries into the open arms. Dietary supplement containing docosahexaenoic acid and gamma-linolenic acid showed prominent neuroprotection in lead induced Swiss albino mice.

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Abstract—WCN 2013

No: 1841

Topic: 10—Neurorehabilitation

Targeting sensory and awareness problems in upper limb rehabilitation in chronic stroke. A report of three cases

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Background: Regaining normal function of the upper limb after stroke occurs in around 10% of cases. Sensory problems such as proprioceptive deficits, learned non-use, and manifest neglect are among the causes of disability.

Objective: Targeting these-frequently-overlooked symptoms may boost functional recovery.

Methods: We suggest a treatment protocol consisting of simultaneous bihemispheric TDCS + TENS + mirror therapy followed by ADLs training.

Case 1: 78 years old, had severe left proprioceptive deficit 3 years after her second stroke with intact motor power. She had pseudoathetosis in her left upper limb. Her left hand would get in her way with eyes open. After 24 sessions; enjoys knitting and other bimanual activities.

Case 2: 68 years old, had learned non-use and moderate right hand weakness 6 years after second stroke. He had shortening in long flexors, weak finger/wrist extensors 1–2/5. Amputated distal phalanges in the right hand. Needed help with all ADLs. He presented to us after his family left him. After 30 sessions; managed to live alone.

Case 3: (managing expectations): 65 years old, presented 1.5 years after stroke with inability to move his left arm. He had a fixed flexion deformity in his arm that was painful with manipulation and had averted him from therapy. He had USN causing and visuospatial problems (could not locate or draw the exit door of the room). After 2 months, safe ambulation at home. He declined a tenotomy for his arm.

Conclusions: Targeting sensory and awareness problems in chronic stroke with multimodal sensory stimulation modalities may augment recovery in selected cases.

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Abstract—WCN 2013

No: 1847

Topic: 10—Neurorehabilitation

Postural profile of individuals with HAM/TSP

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Background: HTLV-1 also has high prevalence in Brazil, especially in the city of Salvador. HTLV-1-associated myelopathy or Tropical Spastic Paraparesis (HAM/TSP) is characterized by a demyelinating, slowly progressive disease, associated with weakness of lower limbs, sphincteric dysfunction, and sensitivity impairment. A computerized postural assessment may help to systematize the clinical practice of physical therapy in the treatment of patients with HAM/TSP and to monitor the results of specific interventions.

Objective: This study sought to delineate the postural profile of these individuals by comparing them with a group of healthy subjects.

Patients and methods: This was cross-sectional research conducted at the HTLV Center of "Escola Bahiana de Medicina e Saúde Pública, Bahia", Brazil. Subjects who were diagnosed with HTLV-1 by ELISA and confirmed by Western Blot Test, and manifested clinical signs compatible with HAM/TSP were included in the study. Patients suffering from other disorders that compromised the definition of postural profile, patients unable to remain in orthostasis without assistance and those who had cognitive impairment that could compromise the postural assessment, as well as those who were undergoing physiotherapy were excluded from the study. 30 volunteers with HAM/TSP were paired with 30 healthy subjects, who underwent a postural evaluation by means of a Postural Assessment Software (SAPO®).

Results: A trend characterized by postural projection of the trunk forward or backward, forward displacement of the body, bending the knees and ankle angle reduction was noted.

Conclusion: This study demonstrates that there is a posture typical of the HAM/TSP patient which manifests itself with changes in the sagittal plane.

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Abstract—WCN 2013

No: 1541

Topic: 10—Neurorehabilitation

Effects of sitting training on a tilting platform in patients with acute stroke: A randomized control trial

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Background: It is important to improve lateral trunk control after stroke. However, the effects of sitting training on a tilting platform are still poorly understood.

Objective: To investigate the effects of sitting training on a tilting platform after stroke.

Patients and methods: Thirty stroke patients participated in this study after providing their written informed consent. Patients were randomly allocated to either an experimental group (n = 15) or a control group (n = 15).

Intervention: The experimental group patients sat on a platform tilted 10° to the paretic side in the frontal plane. The control group patients also sat on a horizontal platform. Both groups moved their trunks laterally to the non-paretic side 60 times. The intervention was in addition to conventional therapy, and was performed for 1 week.

Assessment: Subjects sat on a horizontal platform and moved their trunk laterally to the left or right as much as they could. During this task, the trunk angle was recorded by using the 2-dimensional movement analysis system. As a clinical measurement, patients were evaluated with the Stroke Impairment Assessment Set (SIAS) and the Trunk Control Test (TCT).

Results: The experimental group showed a greater increase in lateral bending to the un-affected side than that of the controls. Although SIAS and TCT significantly improved in both groups, we found a significant interaction in TCT of the experimental group that had a higher score.

Conclusion: These results indicate that tilting platform sitting training can improve lateral trunk control and trunk performance of acute hemiparetic patients.

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Abstract—WCN 2013

No: 1873

Topic: 10—Neurorehabilitation

Efficacy of conventional neurorehabilitative therapy using Tc-99m HMPAO single photon emission computed tomography in patients with subacute infarction

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Background: To objectively evaluate the efficacy of neurodevelopmental therapy on rehabilitative unit was difficulty.

Objective: The purpose of this study was to objectively assess the efficacy of neurodevelopmental therapy on rehabilitative unit using Tc-99m HMPAO single photon emission computed tomography (SPECT) in patients with subacute left MCA infarction.

Method: A total of seven subacute stage patients with left MCA infarction were included in this study. These patients received conventional neurorehabilitative therapy including physical and occupational therapy, twice per day for 3–5 weeks on admitted condition. Brain perfusion SPECT images obtained at pre-Tx and post-Tx were reconstructed using statistical parametric mapping in these patients. A voxel with an uncorrected p-value of less than 0.01 was considered to be statistically significant.

Result: First SPECT scans were performed at 31 days \pm 22 after stroke onset and second SPECT scans were performed at 27 days \pm 9 after first study. The overall regional cerebral blood flow (rCBF) in the left cerebral hemisphere was increased on pre-Tx SPECT images in comparison to that observed on the post-Tx SPECT images. The rCBF in the left temporal, parietal, and frontal cortices was increased on the post-Tx SPECT images in comparison to the pre-Tx SPECT images. These findings indicate that mild improvement in the rCBF can be expected after conventional neurorehabilitative therapy.

Conclusion: Improvements in rCBF following neurodevelopmental therapy were objectively demonstrated in the patient group through SPM analysis of brain perfusion SPECT images, which also correlated well with the clinical symptoms in these patients.

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Abstract—WCN 2013

No: 1706

Topic: 10—Neurorehabilitation

Four periodical botulinum toxin type A injections, followed by home-based functional training in poststroke patients with severe upper limb hemiparesis

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Background and objective: Botulinum toxin type A (BoNT-A) has been reported to be an effective treatment for limb spasticity after stroke. However, few studies have reported improvements in the active motor function in poststroke patients with spastic upper limb hemiparesis. The aim of this study was to assess the clinical influence of four periodical BoNT-A injections, followed by special instructions for a home-based functional training on not only passive but also active motor functions in poststroke patients with severe upper limb hemiparesis with spasticity.

Patients and methods: Of the 295 patients, 41 patients with spastic upper limb hemiparesis were studied. The severity of hemiparesis was categorized as Brunnstrom stage of 3 for hand-fingers in all patients. BoNT-A (maximum dose of 240 U) was injected into the target muscles of the affected upper limb after a clinical evaluation using the modified Ashworth scale, Fugl-Meyer Assessment. Following the injection, occupational therapists provided home-based functional training for each patient on a one-to-one basis. The follow-up evaluation was performed 4 weeks after the four periodical injection.

Results: The changes in the Fugl-Meyer Assessment and the modified Ashworth scale indicated a significant improvement in the active motor function of the affected upper limb, compared evaluation after four periodical injection with before injection.

Conclusion: Our proposed protocol of the periodical BoNT-A injection, followed by home-based functional training seems to have the potential to improve the active motor function of the affected upper limb after stroke, although the efficacy should be confirmed in a randomized-controlled trial.

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Abstract—WCN 2013

No: 1668

Topic: 10—Neurorehabilitation

The importance of extracranial carotid stenosis detection after stroke

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Introduction: Stroke is the primary cause of permanent disability. It is estimated that 25 to 50% of all strokes were caused by these changes.

Patients and methods: The study included patients that suffered a stroke (first or recurrent), 90 days of occurrence, regardless of the degree of neurological deficit. Both carotids were examined by Doppler ultrasound (DUS), the findings were grouped in multiple groups: up to 50% stenosis, 50–75%, 75–99% stenosis and occlusion. The NIHSS score was used to estimate the severity of stroke while the main risk factors monitored were blood fats, high blood pressure and smoking.

Results: During the study, 93 patients underwent DUS screening for carotid stenosis, the mean age of patients was 65.4 years (MD 8.4), 51 (54.83%) were male. 79% of all strokes were ischemic. There was a mean NIHSS score of 7.62, mean total cholesterol 6.48, triglycerides 1.99. 29 patients (31.2%) had normal findings in carotid, 26 had (27.9%) stenosis of 50% (19 on symptomatic, 7 on asymptomatic side), 21 (22.5%) had stenosis of 50–75% (16 on symptomatic, 5 on asymptomatic side), 11 had severe stenoses of 75–99% (9 on symptomatic and 2 on asymptomatic side) and 6 occlusions. There was no statistically significant connection of certain categories of carotid stenosis with NIHSS score and recorded risk factors.

Conclusion: Due to prevalence of severe carotid stenoses, the increased availability of DUS examination, different medications and surgical treatments, the DUS should be a mandatory screening method for high-risk categories of patients.

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Abstract—WCN 2013

No: 1677

Topic: 10—Neurorehabilitation

The effect of transcutaneous electrical nerve stimulation (TENS) combined with Bobath on post stroke spasticity. A randomized controlled study

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Objective: The aim of this study was to evaluate the effectiveness of combined Bobath-TENS therapy in relation to simple Bobath among patients who suffered from stroke and are in the rehabilitation phase of their disease presenting with spastic paralysis.

Methods: A randomized controlled clinical trial study was conducted at the rehabilitation department of Tongji Medical Hospital over a period of 2 years. Thirty stroke patients with ankle plantar flexor spasticity were recruited. The mean age was 55.2 years (37 to 70 years) and mean duration of stroke was 4.73 months (3 to 8 months). Patients were randomly assigned to two groups; 15 each were assigned to the treatment group and the control group. Patients in the control group received 15 min of inhibitory Bobath techniques and a combination of 30 min of TENS on lower leg acupuncture points and inhibitory Bobath techniques was applied to another group for 4 weeks, 5 days a week. MAS score of the affected ankle, passive ankle joint dorsiflexion range of motion, dorsiflexion strength test, gait speed, and the Brunnstrom stages of motor recovery were noted.

Results: The combined therapy group 1 with TENS-Bobath showed extremely successful results in terms of selected parameters. All the parameters turned out to be statistically significant ($P < 0.05$). Ankle passive dorsiflexion movement improved massively followed by 10 meter walk test in the combination therapy group.

Conclusion: 20 sessions of TENS combined with Bobath decreased plantar flexor spasticity, improved dorsiflexors strength, and increased gait velocity significantly more than Bobath alone.

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Abstract—WCN 2013**No: 1620****Topic: 10—Neurorehabilitation****Efficiency of visual biofeedback training for balance correction in neurorehabilitation of the patients with the central hemiparesis syndrome after stroke**

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Background: It's well known that the balance and gait are being changed by the central hemiparesis syndrome.

Aim: To prove expediency of the use of visual biofeedback training for balance correction in neurorehabilitation of the patients with the central hemiparesis syndrome.

Methods: Research was performed with application of the complex MBN "Biomekhanika" (Moscow) and Smart Equitest Balance Manager (USA). The 45 male and 51 female patients were aged between 42 and 70 years with post-stroke central hemiparesis syndrome which had passed 8–10 trainings during 20–30 min. Visual biofeedback trainings included the gradual displacement of the center of gravity towards the hemiparesis of the body side.

The estimation of balance condition was performed before and after the trainings with the help of: Berg Balance Scale, Dynamic Gait Index, and also computerized stabilometry, sensory organization test and motor control test.

Results: After the trainings we can observe the progressive displacement of the center of pressure towards the hemiparesis side and the approaching of the actual center of pressure towards "the ideal". The balance scale indices also had the positive dynamics.

Conclusion: These findings let us to conclude the expediency of use of visual biofeedback programs for balance correction in neurorehabilitation of the patients with the post-stroke central hemiparesis syndrome.

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Abstract—WCN 2013**No: 1655****Topic: 10—Neurorehabilitation****CDP-choline (ceraxon) treatment with hypoxia in the newborn infant**

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Long-term observations suggest that infants who have had a severe hypoxia, lead to a wide range of somatic and neuropsychiatric disorders in the future.

See the effectiveness of therapy in the rehabilitation period Ceraxon infants with perinatal brain lesions, controlled neurosonography doplerometriy with cerebral blood flow, ophthalmoscopy fundus.

Study group were 543 infants who suffered severe hypoxia: 247 full-term and 296 preterm infants who were seen after treatment in catamnesis Ceraxon. Catamnesis duration ranged from 3 to 12 months. According to neurosonography after initiation of treatment Ceraxon, intraventricular hemorrhage in pseudocyst formed in 42%, complete lysis in 22%, posthypoxic changes were revealed in 26%, and 10% of ventriculomegaly. By the end of the first week of treatment 82% of children had normalized indices in pools of anterior and middle cerebral arteries and blood flow in the vein of Galen. Just at the end of the first week physical activity significantly improved in 46%, muscle tone in 64%, and reflex activity in 62% of children.

The analysis of the fundus newborns had diffuse opacities, limited foci of hemorrhage and pigmentation. After treatment Ceraxon on

the second week after the initiation of treatment 32% of children improved picture fundus.

It was thus established that the drug Ceraxon has a positive effect on cerebral hemodynamics of patients: there is rapid correction of neurological disorders by the end of the first week of treatment. This is a positive effect both on the timing, and the final outcome of treatment.

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Abstract—WCN 2013**No: 1622****Topic: 10—Neurorehabilitation****Gait assessment among randomly selected group of patients after stroke using the treadmill with biofeedback**

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Objective: The most important needs of stroke patients are to recover the independent, the efficient and safe gait. The aim of the study is the gait assessment using a treadmill with biofeedback among the patients in the late period after stroke.

Material and method: The study included 50 ischemic stroke patients who were treated after six months from the stroke. The mean age of the patients was 69.1 and mean time from the stroke was 27 months. The patients were randomized into an experimental or control group. The experimental group completed training program on the treadmill with biofeedback. The control group practiced on the treadmill without biofeedback. Gait velocity in the 10-Meter Walking Test, 2-minute walk test, functional evaluation by Barthel index and balance in timed 'up and go' test were tested. Spatio-temporal and kinematic gait parameters were also evaluated. Physiotherapy program lasted two weeks.

Results: At baseline, the average walking speed did not differ significantly between the groups ($p = 0.7878$). The average walking speed in both groups significantly increased after the program of rehabilitation but there was no significant difference between groups. The number of steps per minute significantly increased from 68 to 76 steps, step width decreased but there was no significant difference between the groups.

Conclusions: Among stroke patients in the late period the exercises on a treadmill have an impact on a significant gait improvement. Use of biofeedback did not significantly affect the results.

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Abstract—WCN 2013**No: 291****Topic: 10—Neurorehabilitation****Intracortical GABA is decreased after stroke and further suppressed with successful rehabilitation**

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Background: In healthy subjects, decreasing GABA facilitates motor learning. Recent studies, using PET or TMS, have pointed indirectly to a decrease in neuronal inhibitory activity after stroke. Therefore, we hypothesize that a suppression of GABA levels post stroke might be beneficial to motor recovery.

Objective: To relate GABA changes to motor relearning after stroke through the use of j-difference edited Magnetic Resonance Spectroscopy (MRS).

Methods: 21 patients (3–12 months post stroke) and 21 healthy, age-matched subjects were recruited. Patients had mild to moderate hand impairment, and fulfilled the criteria for Constraint-Induced Movement Therapy (CIMT). Patients completed two weeks of CIMT, and were scanned before and after training. For MRS a $2 \times 2 \times 2$ cm voxel was placed on the “hand knob” in the affected hemisphere of the patients and in the dominant hemisphere of the healthy subjects. GABA was expressed as a ratio to Creatine (Cr). Motor function was measured using the Wolf Motor Function Test (WMFT).

Results: GABA/Cr was significantly lower ($p < 0.001$) in patients (0.33) at baseline compared to healthy subjects (0.42). After therapy, patients showed a significant improvement in hand function ($p < 0.001$), which was negatively correlated with GABA/Cr changes ($R = -0.57$, $p = 0.015$) – larger improvements in patients were associated with greater reductions in GABA/Cr. Results were also significant after correcting for changes in intracortical grey matter volume.

Conclusion: A decrease in GABA levels appears to facilitate motor recovery after stroke. GABA, as measured non-invasively with MRS, could be a biomarker for neuronal plasticity during recovery and guide rehabilitation interventions.

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Abstract—WCN 2013

No: 1595

Topic: 10—Neurorehabilitation

Amantadine treatment of aphasia after stroke

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Background: Amantadine hydrochloride has been used as a central stimulating pharmacological agent for treatment of disordered consciousness in patients with traumatic injury, but not for stroke patients suffering from aphasia and other cognitive deficits in the domains of execution, attention and processing speed.

Objective: To evaluate the effect of short term amantadine treatment on aphasia in subacute stroke patients suffering from aphasia and other cognitive deficits in the domains of execution, attention and processing speed.

Materials and methods: Nine subacute stroke patients (age 30–69 years, time since onset 32–150 days) suffering from aphasia and various other stroke sequelae, receiving inpatient neuro-rehabilitation, completed the cohort study. No blinding was attempted and the intervention was not placebo controlled. Participants were treated with amantadine (50 mg \times 2 on days 1–2 and 100 mg \times 2 on days 3–7) in order to ameliorate cognitive deficits. Participants were evaluated for changes in the severity of aphasia by application of the Western Aphasia Battery (WAB), the WAB Aphasia Quotient (WAB-AQ) being the primary outcome measure.

Results: Amantadine treatment improved the verbal performance as measured by the WAB-AQ by 11 points ($CI_{95\%}$: 6.7; 15.3), $p < 0.0001$. The verbal performance after the subsequent three days of amantadine wash-out was decreased by 5.1 (0.6; 9.6), $p = 0.028$ on the WAB-AQ.

Conclusion: Treatment with amantadine seems to improve verbal performance in patients suffering from aphasia after stroke. This is a preliminary pilot trial and interpretation of the results must be made with caution until further studies are performed.

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Abstract—WCN 2013

No: 1528

Topic: 10—Neurorehabilitation

The role of energocorrection of asthenoneurotic syndrome in different neurologic disorders

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Objective: To evaluate the prevalence of asthenic syndrome in patients with different general medical conditions and neurologic disorders.

Patients and methods: 171 patients with different peripheral nervous system disorders receiving inpatient treatment at hospitals were included (mean age 36.9 ± 8.1 years). Screening revealed the presence of asthenoneurotic syndrome (ANS) of different severity in 120 patients (70.6%) aged 25–56 years. To evaluate the dynamics of ANS complex monitoring was performed on days 1 and 25, including analysis of complaints, assessment of physical and neurologic status, and electroencephalography (EEG-neurokartograf MBN-20). The intervention group received conventional therapy plus energocorrector and antioxidant antihypoxant cytoflavin ($n = 63$) 2 tabs twice a day for 25 days. The control group received conventional therapy plus placebo ($n = 108$) according to the same regimen.

Results: 28.1% of the patients with ANS demonstrated marked personality accentuation with anxiety and excitable traits. At day 25 in the intervention group 44.4% of the patients demonstrated no asthenic symptoms, 31.8%—only mild symptoms, 15.9%—moderate symptoms and 7.9%—severe symptoms ($p < 0.001$). The control group showed an insignificant improvement ($p = 0.257$). There was a positive trend in the subscales “anxiety” and “depression” with a strong correlation with asthenia reduction in the intervention group ($r > 0.5$; $p < 0.05$). There was also an improvement in EEG indices—an increase in α -rhythm from 23% to 46% at day 25 in the intervention group ($p < 0.05$). Overall improvement was registered in 88.9% of the patients, in the control group—27.8% ($p < 0.05$).

Conclusion: Energocorrection therapy in ANS results in a significant decrease in asthenic and neurotic symptoms.

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Abstract—WCN 2013

No: 1486

Topic: 10—Neurorehabilitation

Power effective electrical stimulation of frog's tibial-gastrocnemius preparation for 200-s continuous maximum contraction

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Background: Electrical nerve stimulation (ENS) is not widely adopted in clinical therapy due to a lack of standard and verification of efficacy. A previous study shows that ENS is beneficial to prevent atrophy. Nevertheless, nerve fibrosis might occur if the pulse frequency exceeds 40 Hz. For effective standing, various muscles need to maximally contract for more than 200 s. Therefore, an ENS protocol should be developed for this purpose.

Materials and methods: The contraction force (CF) of the gastrocnemius of frogs was measured and digitized for processing. An adjustable pulse was used to achieve a 200-s continuous maximum contraction (CMC) with minimal power delivery. The maximum contraction force was obtained by the interpolated twitch technique.

Results and discussion: Our data show that the best ENS protocol for 200-s CMC is 8 Hz, 120 mV, and 35% duty cycle. Compared with

previous papers, although the frequency used is much lower, the gastrocnemius still reaches maximum contraction. It implies that the slow muscle in the gastrocnemius accumulates power to reach maximum contraction by multiple stimulation. That means the power could be distributed to the whole stimulating span and drastically reduced, and thus, safer for possible clinical application.

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Abstract—WCN 2013

No: 1521

Topic: 10—Neurorehabilitation

The short-term effects of plantar vibration therapy on the balance of patients after stroke

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Background: Balance disability is common in patients following stroke. The vibration is one of the physiotherapy modalities used in neurological rehabilitation. The effects of local vibration on balance impairment poststroke have not been investigated.

Objective: The aim of the present study was to investigate the short-term effects of plantar vibration on the balance of patients with stroke.

Material and methods: In this pretest–posttest clinical trial, eighteen patients with stroke participated. Outcome measures were Timed Up and Go test (TUG), Functional Reach test (FR), and posturography to assess balance changes; and the Modified Modified Ashworth Scale (MMAS) for evaluation of ankle plantar flexor spasticity. Vibration with a frequency of 100 Hz was applied to the sole of the affected foot for 5 min. All measurements were taken before and after vibration therapy.

Results: The vibration therapy significantly improved the TUG test ($p = 0.03$) and the ankle plantar flexor muscle spasticity ($p = 0.008$). The changes of posturography measures and FR test have not been improved significantly ($p > 0.05$).

Conclusion: Plantar vibration therapy may have beneficial effects on the balance disability of the patients with stroke.

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Abstract—WCN 2013

No: 1374

Topic: 10—Neurorehabilitation

Analysis of independence in older subjects and assessment of the impact of physiotherapy intervention on functional independence and autonomy

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Background: Reduction of locomotion due to neurological disease reduces the quality of life of older people and places increased demands on health and social care.

Aim of the study was to analyze the rate of self-sufficiency of geriatric patients and assessment of the impact of physiotherapy intervention on their functional independence and autonomy.

Patients and methods: The study consisted of 43 males (35.54%), mean age 74.48 and 78 females (64.46%), 78.82 (SD ± 7.26) who underwent rehabilitation 30 min a day 5 times a week. Length of hospital stay was 21 days. Bobath and Kabat neurorehabilitation techniques with sensorimotor stimulation were used. Effect of rehabilitation was tested using FIM for each patient before and after treatment.

Results: Throughout the set an improvement of 10.45%, $Z = 8.69$, $p = 0.000$ was achieved. Input mean of FIM test in males was 97.12 points (SD ± 25.15). After rehabilitation it was 106.70 points (SD ± 20.30), with a difference of 9.58 points (8.91%). Input mean of FIM test in females was 98.15 points (SD ± 22.99) which after rehabilitation was improved by an average of 11.16 points (11.32%) to an average of 109.27 points (SD ± 20.54). The assumption that patients will be improved in the FIM based on gender after completion of the rehabilitation program is not statistically confirmed $Z = -2.5$, $p = 0.409$. Dependency rate of improvement from age is not statistically significant $U = 10$, $p = 0.631$ but there was a statistically significant correlation of diagnoses $Z = -0.212$, $p = 0.000$.

Conclusion: The rate of improvement in FIM after rehabilitation is not dependent on age and gender. Differences of improvement after rehabilitation depend on the patient's diagnosis.

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Abstract—WCN 2013

No: 1385

Topic: 10—Neurorehabilitation

The effects of mirror therapy on arm and hand function in subacute stroke inpatients

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Aim: The aim of this study was to evaluate the effect of mirror therapy on arm and hand function in subacute stroke inpatients.

Material and methods: The study included 60 hemiparetic right-handed patients, eight to ten weeks after ischemic stroke. They underwent stationary comprehensive rehabilitation in the Rehabilitation Centre. There were two study groups: mirror ($n = 30$) and control ($n = 30$) and two subgroups in each: one for people with right arm paresis ($n = 15$) and the other for those with left arm paresis ($n = 15$). The mirror group received an additional intervention: 20 min of exercise program with mirror therapy (twice a day for five and a half days a week for 21 days). The control group ($n = 30$) underwent a conventional rehabilitation program without mirror therapy. To evaluate self-care in performing activities of daily living the Functional Index “Repty” was used. To evaluate hand and arm function the Frenchay Arm Test (FAT) and the Motor Status Score (MSS) were used. Measurements were performed twice: before and after 21 days of applied rehabilitation.

Results: Significant improvement in ability to perform activities of daily living (ADL) in the right arm paresis subgroup of the mirror group has been observed. No significant improvement of hand and arm function in both subgroups in FAT and MSS scales has been observed.

Conclusions: Mirror therapy for arm paresis after stroke is effective in improving independency in activities of daily living.

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Abstract—WCN 2013

No: 1317

Topic: 10—Neurorehabilitation

Comparison of influence of EEG biofeedback and classical rehabilitation on motor function of children

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Background: The aim of the study was to measure the effect of EEG biofeedback therapy on motor and cognitive deficits in children with ADHD or ADD and compare this effect with classical rehabilitation.

Patients and methods: The study included 65 children with mild central motor disorders with ADHD, divided into two subgroups: the first group was composed of 35 children (mean age 9) who underwent EEG biofeedback, and the second group was composed of 30 children (mean age 8.50) who underwent classic neurorehabilitation, with each group undergoing 30 sessions, 2–3 times/week/30–45 min with PANESS/CIT tests before/after procedures.

Results: The mean achieved score of EEG biofeedback was lower ($M = 29$) compared to the score before the procedure ($M = 60$). The second group with classic rehabilitation had a total score of $M = 41.90$ before and $M = 45.83$ after. Test of gaits/hopping/station showed a statistically significant better score for EEG biofeedback compared to classical neurorehabilitation ($Mdn = 55.00$, $p < 0.01$, $Z = -4.89$, $r = -0.63$). For CIT results, mean achieved age of impulsivity after EEG biofeedback was higher ($M = 9.83$) compared to before therapy ($M = 8$) and mean achieved age of attention after the procedure was higher ($M = 10.94$) compared to before the procedure ($M = 9.20$). Changes in overall time showed that EEG biofeedback achieved lower overall time ($M = 94.46$) after the procedure compared to before the procedure ($M = 109.5$). In classical rehabilitation after the procedure there was a lower overall time ($M = 110.58$) compared to before therapy ($M = 132.79$). Both procedures documented an improvement in overall time but absolute values are better in the neurofeedback group. After EEG biofeedback therapy good correlation was shown in the overall score of PANESS and Attention $r = 0.389$, $p = 0.021$.

Conclusion: Our study documents that EEG biofeedback is significantly more effective in a selected group of children with mild to moderate motor and neuropsychological deficits.

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Abstract—WCN 2013

No: 654

Topic: 10—Neurorehabilitation

OnabotulinumtoxinA in lower limb spasticity: Safety results from a pooled analysis

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Objective: Evaluate the safety of onabotulinumtoxinA (BOTOX®) in treating adult post-stroke lower limb spasticity (LLS).

Background: Approximately 80% of post-stroke spasticity patients have lower limb involvement and 66% have involvement of ankle joint muscles, the most commonly affected muscle group.

Patients and methods: Results from eight studies in adult patients with LLS were integrated to evaluate the safety profile of 700 patients. Overall safety population was summarized across all onabotulinumtoxinA doses for double-blind placebo-controlled (DBPC) exposure (onabotulinumtoxinA = 415; placebo = 265) and any onabotulinumtoxinA exposure ($N = 625$; median dose = 300 U).

Results: Overall adverse event (AE) rates during DBPC exposure were 62.2% in the all-onabotulinumtoxinA group, 55.1% in the placebo group, and 68.0% for any onabotulinumtoxinA exposure. During DBPC exposure, AEs $\geq 1\%$ higher in the all-onabotulinumtoxinA group than placebo were fall (7.2%), peripheral edema (4.1%), urinary tract infection (3.9%), headache (3.4%), arthralgia (2.9%), and joint sprain (2.2%). Treatment-related musculoskeletal and administration site AEs were consistent with known onabotulinumtoxinA tolerability. Incidence of serious AEs (SAEs) during DBPC exposure was 11.1% and

8.7% for all-onabotulinumtoxinA placebo groups, respectively. Discontinuation due to AEs was 1.4% in the all-onabotulinumtoxinA group (none in placebo) during DBPC exposure. SAEs and discontinuations due to AEs represented co-morbidities and underlying post-stroke conditions. No reported events were associated with distant spread of toxin and no clinically relevant changes were noted in laboratory tests and physical examinations.

Conclusion: OnabotulinumtoxinA treatment in LLS was well tolerated. The majority of AEs were considered not related to onabotulinumtoxinA, and treatment-related AEs were consistent with the known mechanism of action of onabotulinumtoxinA.

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Abstract—WCN 2013

No: 1354

Topic: 10—Neurorehabilitation

Electromyographic assessment of the tetraplegic upper limb during functional movement

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Background: Tetraplegia compromises the upper limb motor function, thus affecting reach, grasp and handling movements.

Objective: To assess and compare the dominant upper limb muscle recruitment of tetraplegic individuals during reach and grasp movements.

Patients and methods: Four subjects with chronic tetraplegia (motor level: C5–C7) were recruited and divided in two groups (Group I: complete injury, AIS A; Group II: incomplete injury, AIS B). Electromyographic signals were recorded at 4000 Hz sampling rate and 15 to 1000 Hz bandpass-filtering from surface electrodes placed on the biceps brachii, triceps brachii, extensor digitorum communis (EDC) and flexor digitorum superficialis (FDS) muscles. The proposed movement was to reach and pick up a cylindrical object (Phase I) and bringing it to the mouth afterwards (Phase II). This cycle was repeated five times in order to calculate phase duration and root mean square (RMS) averages.

Results: Both variables were analyzed for each phase and compared between groups. Data was presented as group I (mean/SD) and group II (mean/SD), respectively. Phase I—duration (s): (1.83/0.50), (1.93/0.89). RMS (mv): biceps (13.74/2.26), (25.09/7.67); triceps (11.55/1.99), (43.21/4.92); EDC (37.17/11.74), (118.09/63.63); FDS (18.51/11.91), (126.99/60.77). Phase II—duration (s): (1.59/0.46), (2.03/1.08). RMS (mv): biceps (18.64/9.83), (34.47/4.60); EDC (26.73/10.95), (74.05/5.32); FDS (9.67/2.14), (no response in the case of group II).

Conclusion: Individuals with incomplete injuries show greater muscle recruitment in both phases compared to those with complete injuries. Furthermore, participants developed rather unique strategies to accomplish all tasks.

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Abstract—WCN 2013

No: 1346

Topic: 10—Neurorehabilitation

Anatomic localization of motor points in flexor carpi radialis and flexor carpi ulnaris muscles

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Upper limb spasticity usually involves the flexor muscle groups. Flexor carpi radialis (FCR) and flexor carpi ulnaris (FCU) are commonly targeted muscles during neuromuscular blockade in the management of spastic wrist flexors. Precise localization of the motor endplate may enable maximization of the paralytic effect of chemodenervation with botulinum toxin.

The objective of this study was to determine the location of the motor points and intramuscular branches of the FCR and FCU muscles in relation to bony landmarks.

This study was performed on 16 adult cadaveric arms with intramuscular innervation of the flexor carpi radialis and flexor carpi ulnaris muscles.

The site of entry of the motor nerve into the muscle belly was recorded as the motor point in FCR and FCU. We expressed the three parameters (motor points, proximal limit point, distal limit point) in transverse and vertical coordinates in relation to the reference lines.

The mean length of the FCR reference line was 27.2 ± 1.5 cm and that of the FCU reference line was 25.3 ± 1.6 cm. Single motor points in 16 limbs were located for the flexor carpi radialis muscle. On the other hand, one motor point in 6 limbs and two motor points in 10 limbs were identified for the flexor carpi ulnaris muscle.

Based upon these data, we conclude that the optimal area for injection is at a quarter point, for the flexor carpi radialis muscle, and at one-third, for the flexor carpi ulnaris muscle, along each reference line from the medial epicondyle.

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Abstract—WCN 2013

No: 522

Topic: 10—Neurorehabilitation

OnabotulinumtoxinA efficacy in lower limb spasticity:

Results of three studies

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Objective: Evaluate the efficacy of onabotulinumtoxinA (BOTOX®) in the treatment of adult post-stroke lower limb spasticity (LLS).

Background: Approximately 66% of post-stroke patients with LLS have involvement of ankle-joint muscles contributing to functional disturbances.

Patients and methods: Three placebo-controlled studies (Study 1, N = 120; Study 2, N = 85; Study 3, N = 78) utilizing similar criteria for patient selection and injection paradigm were included in this evaluation of onabotulinumtoxinA (median dose = 300 U) administered to ankle plantar-flexors in adults with LLS with baseline modified Ashworth score (MAS) or Ashworth score (AS) ≥ 3 .

Results: Greater AS ankle score improvements and higher response rates for patients receiving onabotulinumtoxinA versus placebo were reported in all studies. In Study 1, reductions in MAS ankle score were significantly greater with onabotulinumtoxinA versus placebo ($p < 0.001$). Proportion of responders (≥ 1 point reduction from baseline muscle tone ankle score) was greater with onabotulinumtoxinA than placebo ($p < 0.001$). Study 2 demonstrated a significant difference between onabotulinumtoxinA 300 U (but not 200 U) and placebo in AS ($p = 0.011$). Study 3 demonstrated clinically and statistically significant improvements with onabotulinumtoxinA versus placebo in functioning measured by Goal Attainment Scale. In all studies, Physician

Global Assessment of Response improved with onabotulinumtoxinA, and significant correlations were observed between muscle tone and study-specific global measure by physician at relevant time-points ($p \leq 0.008$). OnabotulinumtoxinA was well tolerated. Most frequently-reported AEs were myalgia, injection site pain, fall and urinary tract infection.

Conclusion: OnabotulinumtoxinA 300 U significantly decreased ankle flexor tone in adults with post-stroke LLS. This was considered clinically meaningful, as shown by significant correlations with study-specific investigator global assessments.

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Abstract—WCN 2013

No: 1457

Topic: 10—Neurorehabilitation

Methods of recovery of patients with vestibular problems

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Introduction: Vertigo is present in 5–10% of patients who consult general practitioners while 10 to 20% of these patients see the specialists of Otolaryngology/Neurology.

Material and methods: The study was conducted in Constanta County Hospital, Romania, from 01.01.2012 to 01.06.2012, with a total of 26 patients with peripheral and central vestibular syndrome, 13 of whom received medical treatment only and 13 treated with medication and vestibular rehabilitation. Scales were used DHI. Of the 26 patients 16 had severe vertigo attacks of which 10 were diagnosed with peripheral vertigo and 6 with central vertigo.

Results and discussion: The studied groups were clearly observed on the scales made from the important role of neuromotor re-education versus those who received only medications. Complex treatment of patients with vestibular syndrome pursues a number of objectives because we want to improve balance in standing and walking. The effect of functions and kinetic protocol focuses on the main objectives of the study.

Conclusions: These results demonstrate efficacy kinetic rehabilitation focused on body balance and gait, which is particularly important in preventing falls. We believe that physical therapy programs implemented early in the treatment of people with vestibular syndrome improves quality of life.

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Abstract—WCN 2013

No: 1462

Topic: 10—Neurorehabilitation

Virtual reality game-based therapy for restoring postural and coordination abnormalities in patients with TBI

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Background: A traumatic brain injury (TBI) disrupts the central and executive mechanisms of motor coordination, involving abnormal postural control, arm(s) and postural (trunk and legs) interaction, difficulties with arms coupling, lack of agility, and movement precision. To address these issues, we developed the virtual reality (VR) therapy, which instead of regular exercises includes a series

of immersive VR games and scenarios, delivered with low-cost equipment. The therapy replicates the conventional exercise and activity sequence recommended for restoring postural and coordination abnormalities after TBI, and can be delivered either in a supervised clinical setting or in a patient's home.

Objective: Therapy efficacy was tested in 15 participants with moderate manifestations of TBI-related postural and coordination deficits, in the framework of a phase II clinical trial. Therapy included 15 sessions, delivered in-clinic, each approximately 50–55 min in duration, scheduled 2–4 times a week over 5 consecutive weeks. Each participant was evaluated with a battery of clinical tests and movement performance parameters at baseline, immediately after the therapy; and 1 month after the completion of training.

Results: Upon completion of the therapy most participants improved their postural stability, gait, and upper extremity movements. The effects were maintained fully or partially over the retention interval. Safety criteria were established for practicing in-home.

Conclusion: Results will be used to refine the current version of the therapy into a cost-effective, highly-accessible approach, which can be delivered remotely via telerehabilitation.

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Abstract—WCN 2013

No: 1293

Topic: 10—Neurorehabilitation

Safety criteria of verticalization in acute stroke

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Background: Traditionally, to assess the safety verticalization of patients with stroke there are tests used with half-orthostasis and hypercapnia. But the majority of these patients are not able to perform them adequately.

Objective: To assess the safety of verticalization based on indicators of cerebral blood flow (CBF), and to determine simple functional tests to predict the risk of verticalization.

Patients and methods: Fifty patients (63.7 ± 9.3 years old) underwent a series of head-up tilts (HUT) from the 1st to the 5th day after stroke onset. CBF by transcranial Doppler (TCD) and blood pressure (BP) were measured every 3 min during 30 min HUT. Before verticalization was started the overshoot coefficient was determined on the affected side. Outcomes were assessed by NIHSS scale. Patients were divided into two groups: Group 1 (26) with the full recovery on the 14th day according to NIHSS (Δ6.0 ± 3.2), and Group 2 (24) with lower recovery (Δ4.0 ± 2.1).

Results: In Group 1 the overshoot coefficient was higher on the first day (6.4 ± 2.5%) compared to Group 2 (2.1 ± 0.8%, $p = 0.034$), and it increased at the 14th day by 8.1% in Group 1 and by 6.8% in Group 2.

Conclusions: TCD can be used to estimate the risk of complications of early verticalization, thus providing a personalized and intensified approach. Before verticalization a test on reactivity should be performed to determine the overshoot. The higher overshoot coefficient at the start of treatment (>6%) is associated with better recovery.

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Abstract—WCN 2013

No: 1147

Topic: 10—Neurorehabilitation

Influence of plantar somatosensory input on calorically induced and pathologic vertigo

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Background: Active plantar somatosensory stimulation (PSS) via direct pressure has been reported to reduce aspects of induced vertigo in normals. Questions remain whether passive PSS applications also reduce vertigo symptoms and whether the phenomenon has viability for treating patients with pathologic vertigo.

Objective: To compare the effects on vertigo of two PSS methods, direct plantar surface pressure versus plantar TENS, and determine if PSS methods are effective for treating episodic symptoms in patients with benign paroxysmal positional vertigo (BPPV).

Patients and methods: Caloric vertigo was induced with 5 °C water irrigation to external acoustic meatus in 21 asymptomatic adults. Using within-subjects design, each participant received direct plantar pressure, plantar TENS, and sham “subsensory” stimulation control treatments. Visual-analog intensity ratings and electrooculographic measures of nystagmus intensity and duration assessed influence on vertigo. Seventeen patients with BPPV positionally induced vertigo and were treated with a random presentation of the three treatment conditions across six sessions. Repeated measures ANOVA with multiple contrast analyses compared treatment responses.

Results: While not significantly different from each other, the two PSS conditions yielded significant reductions in vertigo intensity compared to control ($p = 0.019$) in normals. No significant differences in vertigo duration were found between the groups ($p = 0.424$). Patients with BPPV similarly showed PSS reductions in vertigo intensity ($p = 0.037$) for treatments over control.

Conclusions: Plantar somatosensory stimulation attenuates intensity of vertigo, whether via direct plantar pressure or TENS, for both induced normals and patients with BPPV. Both methods of providing plantar pressure appear similarly efficacious.

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Abstract—WCN 2013

No: 1245

Topic: 10—Neurorehabilitation

Lateralized response in dynorphin A peptide levels after traumatic brain injury

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The dynorphins, endogenous opioid peptides have been implicated in secondary CNS injury and neurodegeneration. To gain better insight into the pathophysiological role of dynorphins in the CNS response to trauma, we analyzed short term (1 day) and long term (7 day) changes in dynorphin A (Dyn A) levels in the frontal cortex, hippocampus and striatum induced by unilateral left-side or right-side cortical TBI in mice. TBI effects were significantly different from those of sham operation (Sham), while the Sham also produced noticeable effects. Both Sham and TBI induced short-lasting changes in peptide levels in the striatum, and long-term changes in all three regions. Two types of responses were generally observed. In the hippocampus, Dyn A levels were predominantly altered ipsilateral to the injury. In the striatum and frontal cortex, injury to the right (R) hemisphere affected Dyn A levels to a greater extent than on the left side (L). The R-Sham but not L-Sham reduced the

peptide levels in the striatum at the 24 h time point, and R-TBI but not L-TBI produced Dyn A changes in the striatum and frontal cortex at 7 days after the injury. Injury to the R-side induced changes that were similar in both left and right hemispheres. Thus, trauma may uncover an existing lateralization in the mechanism mediating the response of Dyn A-expressing neuronal networks in the brain. These networks may differentially mediate effects of left and right brain injury on the lateralized brain functions.

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Abstract—WCN 2013

No: 1263

Topic: 10—Neurorehabilitation

Repetitive sensory stimulation in stroke rehabilitation—Perspectives and potential

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Background: Rehabilitation employs task specific training and massed practice to drive neuroplasticity mechanisms. However, since many patients suffer from restricted mobility and sensory loss, development of additional and alternative approaches that could supplement, enhance, or even replace conventional training procedures would be advantageous.

Objective: We report effectiveness and feasibility of repetitive sensory stimulation for rehabilitation. The rationale is to target plasticity processes within and around those brain areas that became dysfunctional.

Patients and methods/material and methods: We summarize findings from several subacute and chronic stroke patient studies with left or right cerebral artery thromboembolic infarction. We use high-frequency intermittent electrical stimulation of all fingers of the affected hand applied via custom-made stimulation gloves. Treatment time was 2 weeks in subacute, and several months in chronic patients. For the healthy and the affected limb we assessed performance for touch perception and discrimination, haptic exploration, proprioception, dexterity and grip force.

Results: Repetitive stimulation induced substantial improvement of tactile and sensorimotor performance long lastingly when applied over weeks. Furthermore, positive effects of sensory, motor and proprioceptive functions emerged after months of stimulation in long-term chronic patients.

Conclusion: This effectiveness together with the advantage of usage under everyday conditions by laypeople at their homes, make repetitive stimulation-based principles prime candidates for intervention. A particular advantage of repetitive stimulation is its passive nature, which does not require active participation or attention. Therefore, it can be applied in parallel to other occupations, making it easier to implement and more acceptable to the individual.

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Abstract—WCN 2013

No: 1241

Topic: 10—Neurorehabilitation

Rehabilitation of the patients with deep brain stimulation for consciousness recovery—Our results

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Aim: To compare the condition of patients with severe consciousness disorder (persistent vegetative state or minimally conscious state) as a result of brain trauma or global hypoxia/ischemia and with implanted electrodes for deep brain stimulation for consciousness recovery, at the beginning of rehabilitation and at discharge.

Methods: We obtained data from 9 patients on stationary neurological rehabilitation, 22% were women. Average age was 23.7 ± 10.1 years. Hypoxic–ischemic brain injury was present in 66%, and traumatic brain injury in 34%. There was an average interval of 197.1 ± 137.9 days between injury and DBS implementation, with one of the patients with bilaterally implanted electrodes (on two occasions). Glasgow Coma Scale (GCS) after injury and Functional Independence Measure (FIM) index on admission to rehabilitation and GCS, FIM and Glasgow Outcome Scale (GOS) at discharge were determined. Patients spent an average of 163.0 ± 105.8 days on stationary rehabilitation.

Results: Recovery of consciousness was observed measured by GCS (4.2 ± 2.7 vs. 10.9 ± 1.8 , $p < 0.0001$), but not at FIM index (19.7 ± 5.0 vs 24.3 ± 17.7 , $p = 0.4$). In the majority of the patients (56%) GOS at discharge was SD (severe disability), 33% was CV (coma vigilie) and one patient was GR (good recovery).

Conclusion: The number of patients is too small to draw conclusions about the effect of deep brain stimulation on the recovery of patients with severe brain injury. There has been a recovery of consciousness (GCS), but no significant functional recovery (FIM, GOS), except for sporadic cases. A thorough neurophysiological assessment of candidates for DBS and a larger number of patients is needed.

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Abstract—WCN 2013

No: 1146

Topic: 10—Neurorehabilitation

Posture influence on the pendulum test of spasticity in spinal cord injured patients

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Background: The pendulum test has been used for the assessment of spasticity in patients with different diseases, including spinal cord injury (SCI). However, there is not a standard position for the test.

Objective: To assess whether different patient postures do interfere on spasticity results.

Material and methods: Three individuals with tetraplegia and two with paraplegia went through the pendulum test in three different positions: supine, semi-supine at an angle of 30° and sitting up at an angle of 60° . An electrogoniometer was attached to the right leg for measurement of knee joint angles. All situations were performed five times on different days and the averages were taken for analysis.

Results: Relaxion index (RI), test duration in seconds, initial flexion angle (Fang) and resting angle (Rest ang) were analyzed at three different positions. Supine: RI (mean/sd) 0.93/0.32; test duration 6.9/1.7; Fang 72.6/22.6; Rest ang 55.7/14.2. Semi-supine: RI 0.97/0.39; test duration 8.9/2.7; Fang 75.7/17.8; Rest ang 63.3/5.3. Sitting up: RI 0.62/0.08; test duration 13.9/4.1; Fang 87.9/13; Rest ang 54.8/12.8. Differences are shown for sitting up position.

Conclusion: Spasticity reduction and an increase in quadriceps femoris relaxation were observed in the sitting up position due to muscles shortening in that position, when compared to the others. Furthermore, in supine and semi-supine positions autonomic dysreflexia symptoms (i.e. arterial hypertension) were observed during the tests.

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Abstract—WCN 2013**No: 1107****Topic: 10—Neurorehabilitation****Quantitative MRI indexes in neuromuscular and muscle diseases**C. Angelini^a, M. Fanin^b, E. Peterle^b. ^aIRCSS San Camillo, Venice, Italy;^bUniversity of Padova, Padova, Italy

Objectives: We propose two new types of quantitative measurement to evaluate muscle atrophy by imaging: the quadriceps index and the left vastus lateralis index and compare it in muscle biopsy.

Methods: We have used T1 sequences on thigh muscle MRI, at about 15 cm from the head of the femur (second slide of MRI in lower extremities). In these sequences we measured the muscle area of the left quadriceps femoris and of the left vastus lateralis. These measurements were carried out in 11 patients with various types of neuromuscular diseases and myopathies, i.e. 2 lipid storage myopathies (LSM), 1 ALS, 1 FSHD, 1 myofibrillar myopathy, 2 LGMD2A, 1 LGMD1F, 1 myositis ossificans, and 2 aspecific myopathies. We investigated by morphometry and molecular markers of atrophy or autophagy, i.e. MURF, LC3, and muscle biopsies.

Results: By MRI the measurement of the muscle area of the quadriceps femoris in 11 patients, resulted in an average of 3711 mm² ± SD 792, and the muscle area of vastus lateralis in 11 patients had an average of 963 mm² ± 303. In the most atrophic sub-group the values ranged from 400 to 900 mm (mean 658.7) and these included cases of LGMD2A, 1F, LSM and ALS.

Conclusion: In ALS and metabolic myopathies a high degree of atrophy was found and similar changes were present in calpainopathy and LGMD1F, due to trasportinopathy. The correlation of imaging indexes with fiber atrophy parameters obtained in muscle biopsy is promising to evaluate rehabilitation in neuromuscular cases.

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Abstract — WCN 2013**No: 1191****Topic: 10 — Neurorehabilitation****New approaches to the use of transcranial magnetic stimulation in rehabilitation of patients suffering from cerebral insult**A.V. Musayev^a, S.Q. Huseynova^b, F.K. Balakishiyeva^c. ^aResearch Institute of Medical Rehabilitation, Azerbaijan; ^bClinically-Neurophysiological Laboratory, Azerbaijan; ^cNeurology, Research Institute of Medical Rehabilitation, Baku, Azerbaijan

One of the most perspective aspects of increasing the efficacy of restorative therapy in patients with cerebral insult (CI) is the use of transcranial magnetic stimulation (TMS).

The aim of this research is to assess the TMS method (with the influence on cerebral motor structures).

Material and methods: We recruited 57 patients with a diagnosis of CI. To study therapy efficacy, all the patients underwent neurologic investigation, the assessment of a patient's daily activities (according to the Barthel scale), functional state of central neuromotor system as well as cognitive functions (according to MMSE and FAB scales). All patients received TMS on motor centers of both hemispheres within 10–15 min, 10–12 procedures per course of therapy.

Results: The patients with CI showed regression of paresis rate (39.4%), spasticity (38.2%), and gait improvement (35.9%). The clinical improvement was confirmed by positive dynamics of the Barthel index which had been increased ($p < 0.01$) and also by the improved functional state of the central neuromotor system (by increased amplitude of evoked motor response and reduced central motor conduction time, $p < 0.05$). The treatment improved the state of main cognitive functions — memory, attention, orientation, etc. (44.8%).

Conclusions: The method developed by us of a patient's therapy with a diagnosis of CI enables the regression of motor deficit, improvement of daily activities, a patient's "quality of life" and correction of cognitive disorders as well. Of course, the functional changes in cerebral cortex, activation of collateral tracts are the underlying basis of its therapy effect.

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Abstract — WCN 2013**No: 978****Topic: 10 — Neurorehabilitation****Priming at the sub-acute stage after stroke accelerates recovery of upper limb function**W.D. Byblow^{a,b}, C.M. Stinear^{a,c}, M.A. Petoe^{a,c}, S. Anwar^{a,d}, P.A. Barber^{a,c}. ^aCentre for Brain Research, The University of Auckland, Auckland, New Zealand; ^bDepartment of Sport & Exercise Science, The University of Auckland, Auckland, New Zealand; ^cDepartment of Medicine, The University of Auckland, Auckland, New Zealand; ^dRehab Plus, Auckland, New Zealand

Background: Upper limb impairment is common after stroke and recovery of upper limb function reaches a plateau within 6 months for most patients.

Objective: We hypothesised that bilateral priming before upper limb therapy would accelerate upper limb recovery.

Patients and methods: We enrolled 57 patients with first-ever monohemispheric ischaemic stroke in this randomised double-blinded trial. For bilateral priming, patients actively flexed and extended their non-paretic wrist in a mechanical device, which moved their passive parietic wrist in a mirror-symmetric manner. Control priming consisted of 15 min of weak cutaneous electrical stimulation. Priming was done for 15 min immediately before 20 min of upper limb therapy, 5 days/week for 4 weeks, starting 2 weeks after stroke. Corticospinal tract integrity was assessed with TMS and DW-MRI, and BDNF genotyping performed. Upper limb function was assessed with the ARAT at 2, 6, 12 and 26 weeks. The primary endpoint was recovery of upper limb function at 12 weeks, using ITT and PP analyses.

Results: Primed patients were 3x more likely than control patients to reach 75% of their maximum recovery within 12 weeks (ITT OR 2.7, 90% CI 1.1–7.0; PP OR 3.4, 90% CI 1.2–9.9). Corticomotor excitability was normalised to a greater extent after bilateral priming than control ($F_{3,129} = 2.91$, $p = 0.037$).

Conclusion: Bilateral priming accelerated recovery of upper limb function and normalised motor cortex excitability. This is a simple, inexpensive way of enhancing the efficiency of upper limb rehabilitation after stroke.

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Abstract — WCN 2013**No: 1126****Topic: 10 — Neurorehabilitation****The use of choline donators in the cognitive rehabilitation after severe brain injury**

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Background: Constant growth of frequency of neurotrauma with consequent cognitive impairment that worsens the prognosis of rehabilitation after traumatic brain injury, a high degree of disability, makes the problem of diagnosis and treatment of cognitive dysfunction socially significant.

Objective: To assess the results of use of the donors of choline during neurorehabilitation of patients with cognitive impairment as the consequences of severe traumatic brain injury.

Patients and methods/material and methods: We identified some particular features of the dynamics of cognitive impairment in the late period of severe traumatic brain injury in three groups of patients: isolated traumatic hematoma, focal contusions of the brain, and a combination of intracranial hematoma with focal contusions. In our study we had enrolled 205 patients with severe consequences of brain injury (the main group 133, the control group 72 observations). We supervised and assessed the dynamics of cognitive impairment after the combined treatment with using the donor of choline (Gliatilin) and acetylcholinesterase inhibitors (Reminyl, Exelon). We used the following scales – MMSE, Barthel, and Rankin, the observation period was 48 months.

Results: The best results were obtained in patients in the combined treatment that included Gliatilin. The combination of Gliatilin with acetylcholinesterase inhibitors in patients with severe mental disorders improves the long-term quality of life.

Conclusion: The analysis of the effects of treatment showed that the inclusion of donor of choline is a promising direction for both prevention and treatment of mental disorders after severe traumatic brain injury.

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Abstract – WCN 2013

No: 906

Topic: 10 – Neurorehabilitation

Task-specific interactive game-based virtual reality rehabilitation system for stroke patients: A usability test and two clinical experiments

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Background: Virtual reality is not commonly used in the clinical rehabilitation setting and commercial VR gaming system may provide mixed effects to stroke patients.

Objectives: To develop a task-specific interactive game-based VR system for stroke rehabilitation, RehabMaster™, and to assess usability and clinical efficacy for rehabilitation.

Methods: A participatory design and usability tests were carried out for development of RehabMaster with representative user groups. Also, two clinical trials were carried out. As a first trial for seven chronic stroke patients, we conducted an observational study, in which they received 30 min of RehabMaster interventions for two weeks. Secondly, as a randomized controlled trial, 16 acute and subacute stroke patients were recruited. They were assigned to conventional occupational therapies (OT-only group) or conventional occupational therapy plus 20 min of RehabMaster intervention (RehabMaster + OT group) for 10 sessions. Fugl-Meyer assessment (FMA), modified Barthel Index (MBI), adverse effects and the drop-out rate were recorded.

Results: The requirements of the VR system for stroke rehabilitation were established and reflected on RehabMaster. Reported advantages from usability tests were improved attention, immersive flow experience, and individualized intervention. The first clinical trial showed that FMA and MBI differed between evaluation times ($P < .05$). The second trial revealed that RehabMaster + OT group improved better than OT-only group ($P = .07$) in terms of FMA, without effects in MBI. One chronic patient discontinued the trial and no adverse effects were reported.

Conclusion: The RehabMaster is a feasible and safe VR system to enhance upper extremity functions for stroke patients.

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Abstract – WCN 2013

No: 798

Topic: 10 – Neurorehabilitation

The effect of anodal transcranial direct current stimulation to the sensorimotor cortex on somatosensory evoked magnetic fields

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Background: Several studies have revealed that tDCS modulated the human motor cortex. However, there are few studies to investigate the change of SEFs using tDCS.

Objective: The aim of the present study is to investigate the effect of anodal tDCS on the sensorimotor cortex under the influence of SEFs.

Material and methods: Seven healthy subjects (24.9 ± 3.8 years) participated in this study. All subjects gave their written informed consent. tDCS was delivered using a direct current stimulator through a pair of saline-soaked surface sponge electrodes (35 cm^2). The anode was placed on the left scalp over the area representing the right thenar muscle, and the other electrode was placed above the contralateral orbit. Anodal tDCS was applied for 15 min to the left motor cortex at 1 mA. We recorded SEFs following a 0.5-Hz electrical stimulation of the right median nerve at the wrist using a 306-channel whole-head MEG system both before and after tDCS. The intensity of this stimulation was fixed at 1.2 times that of the motor threshold.

Results: The current strength of N20m was unchanged after tDCS, whereas that of P35m and P60m increased after tDCS (P35-pre: $24.7 \pm 12.7 \text{ nAm}$, post: $33.5 \pm 11.6 \text{ nAm}$; P60-pre: $30.6 \pm 23.3 \text{ nAm}$, post: $36.9 \pm 26.5 \text{ nAm}$). The ECD locations of P35m and P60m were unchanged before and after tDCS.

Conclusion: Our study revealed that anodal tDCS of the sensorimotor cortex affects the P35m and P60m components of the SEF.

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Abstract – WCN 2013

No: 988

Topic: 10 – Neurorehabilitation

Use of functional independence measure instrument at a neurological step-down unit

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Introduction: The Functional Independence Measure (FIM) is an outcome measure of the severity of physical and cognitive disabilities for an inpatient rehabilitation setting. The severity of disability changes during rehabilitation treatment, making changes in the FIM scale an indicator of treatment benefits and its results. The FIM is a seven level scale representing patient's independence and dependence. So far there is no evidence of the functional profile of patients followed by a physiotherapist at a Neurological Step-Down Unit (NSDU).

Objective: To evaluate the functional profile of patients followed by physiotherapists at the NSDU.

Methods: A cohort study, evaluating the FIM scale of patients admitted to NSDU at the Albert Einstein Jewish Hospital, older than 18 years old, during an eight month period, and that were followed by physiotherapists. FIM scale was applied by trained and certified physiotherapists at day one (when patient was alert and responsive) and at NSDU discharge. The FIM scale rates from 18 to 126 points, evaluating: self care, sphincter control, transfer and mobility, deambulation, communication and social cognition.

Results: During the study period, 137 patients were evaluated, with a median age of 73 years (range of 18–98). There was a significant functional improvement at NSDU discharge when compared to day one (median of 59, range of 18–126 vs. median of 84, range of 18–126, respectively; $p < 0.001$).

Conclusions: The FIM scale represents applicability during clinical assistance, showing improvement in patients' physical function at NSDU discharge. This improvement may represent the quality of physiotherapy assistance.

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Abstract – WCN 2013

No: 990

Topic: 10 – Neurorehabilitation

Profile of patients followed by physiotherapy at a neurological step-down unit

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Introduction: The Neurological Step-Down Unit (NSDU) is a specific unit for neurological patients, with moderate complexity, that requires intensive care due to high dependence and continuous monitoring. The physiotherapy treatment at the NSDU may vary due to cultural and specific characteristics of each service, region or country, focusing on patients' respiratory and motor problems. So far there is scarce evidence of patients' characteristics followed by physiotherapists and demand of this speciality at the NSDU.

Objective: To describe patients' profile followed by physiotherapy at the NSDU.

Methods: A prospective, observational study, during an 8 month period after the inauguration and physiotherapy service at our NSDU at the Albert Einstein Jewish Hospital, Sao Paulo, Brazil. Demographic data, number of patients' admissions, number of physiotherapy prescriptions, admission diagnosis, and prevalence of noninvasive ventilation (NIV) use.

Results: During the study period, a total of 297 admissions were registered, of these 137 (47%) had physiotherapy prescription. The median age was 73 years (range of 18–98). Of patients with physiotherapy prescription, 45% ($n = 62$) had been diagnosed of stroke, followed by 8% ($n = 11$) of brain tumor resection, 6% ($n = 8$) of spine arthrodesis, 5% ($n = 7$) of subarachnoid hemorrhage, and 4% ($n = 6$) of convulsive crisis. NIV was used in 18% ($n = 25$) of patients. The median duration of physiotherapy treatment at the NSDU was 2 days (range of 1–34).

Conclusion: Physiotherapy service treats almost half of patients admitted to the NSDU, most of them with stroke diagnoses, low prevalence of NIV and few days of treatment.

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Abstract – WCN 2013

No: 1085

Topic: 10 – Neurorehabilitation

Smooth pursuit eye movement training accelerates recovery from auditory/visual neglect and reduces disability and unawareness: 2 randomized controlled trials

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Background: No randomized controlled trials (RCT) evaluated treatments for auditory neglect and unawareness, and few reported effects on disability/functional measures.

Objective: RCT1 evaluated the effects of 5 sessions Smooth Pursuit Eye Movement Therapy (SPT) versus Visual Scanning Therapy (VST) on auditory/visual neglect, and compared their efficacy separately for mild vs. severe neglect. RCT2 evaluated the effects of 20 sessions SPT versus VST – at the bedside – on disability, unawareness, rehabilitation status and functional (in) dependence.

Patients/methods: RCT1: $n = 50$ patients with right-sided stroke, 3 months post-lesion, RCT2: $n = 30$ patients with right-sided stroke, 1 month post-lesion. In both trials, SPT therapy entailed smooth pursuit eye movements to the neglected side using computer-generated, leftward moving dots on a computer screen. VST patients viewed the same but static stimuli and had to scan these with saccades.

Results: SPT and VST groups in both trials did not differ in clinical/demographic and neglect measures at pre-therapy. RCT1: SPT-patients showed large improvements in auditory and visual neglect; no significant effects of VST were observed. Effect sizes (Cohen's d) were fivefold higher after SPT versus VST, both for mild and severe neglect. Treatment effects were maintained at 2-week follow-up. RCT2: SPT-patients improved significantly more in unawareness and functional neglect (gaze orientation, search objects on tray, find pictures, bisection) after treatment; effects remained stable at 2-week follow-up.

Conclusions: Repetitive, contralesional SPT induces superior, multimodal and lasting therapeutic effects in mild and severe neglect, and reduces unawareness and functional neglect in the activities of daily living. Moreover, SPT is effective as bedside-therapy.

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Abstract – WCN 2013

No: 1035

Topic: 10 – Neurorehabilitation

Role of vision in gait stabilization: Local dynamic stability in treadmill walking while blindfolded

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Background: Vision plays a fundamental role in human locomotion, especially in orientation and obstacle avoidance tasks. On the other hand, it is known that the stepping pattern is mainly regulated by proprioceptive feedbacks and spinal processing.

Objective: To assess the lateral stability of gait in the absence of vision using a nonlinear stability index.

Methods: 100 healthy individuals (20 to 69 years, mean 44 years) performed three walking trials of a 5 min duration on a motorized treadmill:

- 1) at preferred walking speed, eyes open (EO);
- 2) at new preferred (slow) walking speed with eyes closed (EC), and
- 3) at the same speed as in 2 with EO.

Attached onto the sternum, a triaxial accelerometer recorded the trunk accelerations. The local dynamic stability (LDS) of gait in the mediolateral direction was assessed by estimating the maximal Lyapunov exponents from short-term divergence between neighbor trajectories in a reconstructed state space that reflected the gait dynamics.

Results: The participants decreased their preferred walking speed when walking EC (condition 1) vs. condition 2 (–17%). They also exhibited slightly shorter steps (–3%) and higher cadence (+3%) in EC condition (i.e. 2 vs. 3). LDS was not substantially modified in EC condition (2 vs. 3 +1%, not significant).

Conclusion: In the absence of visual feedback, healthy individuals adopt a more cautious walking pattern (lower speed, shorter steps). However, the local dynamic stability in the frontal plane seems not affected. It is therefore likely that the somatosensory and vestibular feedbacks are sufficient to maintain an optimal gait stability.

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Abstract – WCN 2013

No: 879

Topic: 10 – Neurorehabilitation

Neural substrates underlying early motor skill learning in chronic stroke patients: A fMRI study

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Background: Motor skill learning plays a central role in post-stroke motor recovery but little is known about the neural substrates underlying motor skill learning in stroke patients. Recently, we used functional magnetic resonance imaging (fMRI) in healthy volunteers while they learned a visuomotor skill. The most efficient volunteers improved their speed/accuracy trade-off (shifters), while the other ones did not (fitters). The supplementary motor area (SMA) was the key area underlying early successful motor skill learning, i.e. shift of the speed/accuracy trade-off.

Objective: To explore the neural substrates underlying early motor skill learning in chronic stroke patients, according to their motor skill learning pattern (shift/fit).

Patients and methods: Sixteen chronic stroke patients learned the visuomotor skill with their paretic upper limb during fMRI acquisition. The skill consisted in moving a cursor across a circuit using a fMRI-compatible mouse, as quickly and as accurately as possible.

Results: In the shifter stroke patients (n = 8), the network underlying motor skill learning involved the SMA, bilateral primary motor, premotor areas & somatosensory areas, the posterior parietal cortex in the damaged hemisphere, and the anterior cerebellum. In the fitters (n = 8), the network encompassed the SMA, bilateral posterior parietal cortex, the anterior cerebellum, and primary motor area & dorsolateral prefrontal cortex in the damaged hemisphere.

Conclusion: In chronic stroke patients, early motor skill learning relied on a reorganised network compared to healthy volunteers. In the efficient stroke patients (shifters), the fMRI pattern encompassed a premotor–motor–parietal network, whereas higher-order attentional areas were recruited in the less efficient fitters.

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Abstract – WCN 2013

No: 568

Topic: 10 – Neurorehabilitation

Effect of non-robotic equipment on improvement of movements of the upper limb in children with hemiparesis syndrome

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Background: Pilot study tested improvement of upper limb movements in children with hemiparesis syndrome.

Objective: The aim of the study was to determine the effect of Armeo® therapy system on movement and the ability to grip of upper limb in patients with hemiparesis syndrome.

Patient and methods: The object of investigation consisted of five patients 10 to 16 years old with impaired upper limbs. They all have taken twenty therapies in the Armeo® system, wherein one therapy lasted 45 min of active exercise and frequency was minimal twice a week. Patients were tested before and after completion of therapy using goniometric investigation and by testing grip of paretic hand (cylindrical, spherical, hook...).

Results: After rehabilitation by Armeo® device the patients achieve greater range of motions in the upper limb which resulted in a higher average output score (M = 615, SD ± 28.723) than the input score (M = 515, SD ± 21.622), $t(5) = -9.00$, $p = 0.001$. Significantly better results demonstrate the improvement in hand grip which resulted in higher average output score (M = 34.20 SD ± 18.78) compared with the input score (M = 29.80 ± SD 18.70), $Z(5) = -2.023$, $p = 0.043$.

Conclusion: Significant positive results were achieved due to Armeo® therapy system in improving the range of motion in the hemiparetic upper extremity and similar significant results have shown improvement in grip ability of paretic hand.

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Abstract – WIN 2013

No: 941

Topic: 10 – Neurorehabilitation

Mirror therapy for patients with severe arm paresis after stroke – A randomized controlled trial

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Objective: To evaluate the effects of individual or group mirror therapy in patients with severe arm paresis after stroke.

Methods: In this randomized controlled single blinded trial 60 patients with severe arm paresis who underwent an inpatient rehabilitation within three months after stroke were included. Patients were randomized into one of three groups:

- (1) individual mirror therapy,
- (2) group mirror therapy and
- (3) control intervention with restricted view on the affected arm.

We evaluated motor function on impairment (Fugl–Meyer Test) and activity level (Action Research Arm Test), independence in activities

of daily living (Barthel Index), quality of life (Stroke Impact Scale) and visuospatial neglect (Star Cancellation Test).

Results: After five weeks, no significant group differences for motor function were found ($P > 0.05$). Pre-post differences for the Action Research Arm Test and Fugl–Meyer Test: individual mirror therapy: 3.4 (7.1) and 3.2 (3.8), mirror group therapy: 1.1 (3.1) and 5.1 (10.0) and control therapy: 2.8 (6.7) and 5.2 (8.7). However, a significant effect on visuospatial neglect for the individual mirror therapy compared to control group could be shown ($P < 0.01$). Furthermore, it was possible to integrate a mirror therapy group intervention for severely affected patients after stroke.

Conclusion: This study showed no effect on sensorimotor function of the arm, activities of daily living and quality of life of mirror therapy compared to a control intervention after stroke. However, a positive effect on visuospatial neglect was indicated.

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Abstract – WCN 2013

No: 749

Topic: 10 – Neurorehabilitation

Standardization of motor free visual perception test (MVPT-3) in Korean adults

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Background: The motor free visual perception test (MVPT) is most widely used in stroke rehabilitation in Korea. But MVPT was not adapted and standardized in Korean, thus test application and interpretation depend on an arbitrary decision. MVPT-3 is the latest version and consisted of 65 items including visual discrimination, form consistency, visual short term memory, visual closure, spatial orientation and figure ground.

Purpose: This study was performed to establish reference values of MVPT-3 in Korean adults.

Method: The test instructions of the MVPT-3 were translated and adapted to Korean and back-translation was also performed by a proficient bilingual. The original stimuli of MVPT-3 were not changed. Healthy volunteers who aged over 20 years were recruited. People who have a previous history of stroke, neuropsychiatric illness, severe visual impairment, and decrement of K-MMSE scores of more than 1 SD as compared to the same age group.

Results: Three hundred twenty one adults participated in this study and mean age and education years were 50.96 ± 21.03 and 11.64 ± 5.31 years, respectively. Total mean MVPT-3 score is 51.45 ± 8.32 and the score significantly correlated with age and education years. Post-hoc analysis classified age and education years into 5 groups (age; 20–59, 60–79, and over 80 years, education; 0, 1–9, over 9 years).

Conclusion: Visual perception is significantly influenced by age and education. Reference values provided in this study will help to apply and interpret the MVPT-3 in Korean and evaluation of visual perceptual dysfunction in stroke patients.

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Abstract – WCN 2013

No: 698

Topic: 10 – Neurorehabilitation

Diabetic polyneuropathy prevention method

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Introduction: In the Republic of Belarus diabetes affects 3900 of every 100,000 people. Diabetic distal sensory polyneuropathy (DDSPN) is one of the most common diabetes complications. 5 years after diabetes manifests 12.5–14.5% of patients are diagnosed with DDSPN (FIELD Study, 2012).

Goal: Develop methods of DDSPN prevention.

Methods: From 2010 to 2012, there were 249 patients examined with type 2 (82%) and type 1 (18%) diabetes without neurologic disorders (male: 179, female: 70, average age 38 ± 13). Vibrational, tactile, temperature, and pain sensitivity of legs were defined and electro-neuromyography (ENMG) was used. There a group of patients (186 people) was picked without manifestations of DDSPN, but with ENMG criteria typical for initial manifestations of this pathology (San Antonio single ENMG program, 1988). They were divided into 2 groups: A. (96 people) – glycemic control, diet and iontophoresis applied to the lower limbs were prescribed; B. (90 patients) – glycemic control and diet were prescribed.

Results: After the treatment ended, 64 people (61%) from group A showed the growth of amplitude and velocity indexes of n. peroneus, n. tibialis, and n. suralis to normal ($p < 0.001$), which lasted for 6 months. In group B the improvements were for 7 patients (6%). Dropping of above indexes of the examined nerves and clinical manifestations of DDSPN were revealed for 2 (2%) patients in group A, and for 30 (27%) patients in B ($p < 0.001$).

Conclusion: Delivery of anticholinesterase drugs to lower limbs by iontophoresis is a method of DDSPN prevention on the clinical stage.

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Abstract – WCN 2013

No: 699

Topic: 10 – Neurorehabilitation

Benefits of inpatient multidisciplinary rehabilitation in multiple sclerosis

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Background: Rehabilitation is often recommended to MS-patients but data on its efficacy is limited.

Objective: To evaluate the benefit of inpatient multidisciplinary rehabilitation.

Methods: A rater-blinded, randomized, waiting list controlled exploratory study. 19 participants completed the study with ten allocated to the intervention group and nine to the waiting list group. Assessment of outcome-parameters was done at baseline and after 3 months. Time walking tests (TWTs) and a 9 hole peg test were used to objectively assess the level of activity, Functional-Assessment-in-MS and MS-Self Efficacy-Scale to assess participation and quality of life and Expanded Disability Status Scale (EDSS) to assess bodily function. Additionally, Rivermead Mobility Index, Berg Balance Scale, Tinetti-Test, MS-Functional Composite and a rater-blinded evaluation of a video-analysis on walking performance were done.

Results: Mean change scores of timed 50 m walk ($p = 0.014$), walking speed ($p = 0.034$), 2- ($p = 0.204$) and 6-min walk ($p = 0.034$) indicated an improvement favoring inpatient multidisciplinary rehabilitation. We could not demonstrate a benefit for upper limb function and some improvement was seen in other outcome parameters without reaching statistical significance. EDSS remained unchanged.

Conclusion: Inpatient multidisciplinary rehabilitation is effective in MS patients with positive effects on the level of activity as measured by TWTs covering both short and long distance ambulation.

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Abstract – WCN 2013

No: 825

Topic: 10 – Neurorehabilitation

Mirror therapy for improving motor function after stroke

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Background: Mirror therapy is used to improve motor function after stroke. During mirror therapy, a mirror is placed in the patient's midsagittal plane, thus reflecting movements of the non-paretic side as if it were the affected side.

Methods: We conducted a systematic Cochrane review to summarise the effectiveness of mirror therapy after stroke. We electronically searched relevant databases and handsearched conference proceedings, research registers and reference lists. We included randomised controlled trials (RCTs) and randomised cross-over trials. Two authors independently selected trials based on the inclusion criteria, and documented the methodological quality of studies and extracted data. We analysed the results as standardised mean differences (SMDs) for continuous variables.

Results: We included 14 studies with a total of 567 participants. When compared with all other interventions, mirror therapy may have a significant effect on motor function (post-intervention data: SMD 0.61; 95% confidence interval (CI) 0.22 to 1.0; $P = 0.002$; change scores: SMD 1.04; 95% CI 0.57 to 1.51; $P < 0.0001$). Additionally, mirror therapy may improve activities of daily living. We found a significant positive effect on pain which is influenced by patient population, and limited evidence for improving visuospatial neglect. The effects on motor function were stable at follow-up assessment after six months.

Conclusion: With some limitations, the results indicate evidence for the effectiveness of mirror therapy for improving upper extremity motor function, activities of daily living and pain, at least as an adjunct to normal rehabilitation for patients after stroke.

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Abstract – WCN 2013

No: 768

Topic: 10 – Neurorehabilitation

Influence of virtual reality environment in robotic-assisted treadmill training on motor functions in children with cerebral palsy

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Background: In the past decade there has been a striking increase in the use of robotic therapy, especially in patients with strokes, cerebrospinal trauma and also in children with cerebral palsy. Virtual

reality (VR) environments were developed to enhance robotic-assisted treadmill training (RATT).

Objective: To examine the influence of VR environment in robotic-assisted treadmill training on gross motor functions in children with cerebral palsy (CP).

Material and methods: 42 children (25 boys) with bilateral spastic CP, aged 4.3–12.9 years underwent 20 sessions of RATT during a 4–5 week period with a frequency of 3 to 5 times a week using driven gait orthosis Lokomat. Patients were randomly allocated into two groups according to whether they trained in a VR environment (VR-LOKO, $n = 26$) or not (LOKO, $n = 16$). Outcome measures were dimension A (lying, rolling), B (sitting), C (crawling, kneeling), D (standing), and E (walking, running, jumping) of the Gross Motor Function Measure (GMFM-88).

Results: After completing 20 sessions, patients of both groups demonstrated highly statistically significant improvement ($p < 0.001$) in all dimensions of the GMFM. Comparing the average improvement in outcome parameters in both groups (LOKO vs. VR-LOKO), we documented a statistically significant difference ($p < 0.05$) in the dimension of A, C, and overall improvement in the GMFM in favour of patients in VR-LOKO group.

Conclusion: Results of our study indicate that the environment of VR implemented during RATT affects the amount of the achieved improvement of motor functions in CP patients. VR-based training conditions represent a valuable approach to enhance active participation during RATT in children with CP.

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Abstract – WCN 2013

No: 782

Topic: 10 – Neurorehabilitation

Active very early rehabilitation safety and effectiveness following acute stroke

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Introduction: Active very early rehabilitation (AVER) emphasizes on early mobilization for target acute stroke patients at ≤ 24 h admission to ASU, whose physiological parameters are within the set limits.

Aims:

1. To evaluate the AVER on its feasibility, and primary & secondary outcomes; and
2. To determine the LOS in ASU with associated factors.

Methodology: A descriptive, quasi-experimental design with pre- and post-tests was employed.

Results: 103 patients were screened and 64.4% ($n = 65$) were recruited for AVER. Mean time to first mobilization from ASU admission was 17.0 h (SD 4.9). 80% ($n = 52$) were ischemic stroke. After AVER, the **primary daily living and ambulatory independency outcomes** were significantly improved, and included Modified Barthel Index 100 (BI-100), Modified Rankin Scale (MRS) and Modified Functional Ambulatory Categories (MFAC) (paired t test, $p < 0.001$). 70.8% of patients achieved daily living independency ($MRS \leq 2$) & 69.2% achieved satisfactory ambulatory independency ($MFAC \geq 5$). The **secondary safety outcomes** like mortality rate and post-stroke complications were low (12.3%, $n = 8$). They occurred in severe stroke (mean NIHSS 9.75, SD 8.50). The mean length of stay (LOS) was 5.38 days (SD 2.76). The stepwise multivariate regression analysis showed that LOS was associated with NIHSS and MRS post-AVER ($p < .001$). This model correctly predicted 46.1%. Each additional point on the LOS would increase by approximately. 193 N_1 where N_1 is the NIHSS and 795 N_2 where N_2 is the MRS post-AVER.

Conclusion: AVER in ASU is feasible, safe & effective. Further research to review the AVER beneficiaries is recommended for future development.

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Abstract – WCN 2013

No: 759

Topic: 10 – Neurorehabilitation

Assessment of cerebral activation patterns while operating a driving simulator with TBI survivors: A functional near-infrared spectroscopy study

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Background and objective: Driving is an important activity of daily living. However, the ability to drive is often affected after traumatic brain injury (TBI).

The aim of this study was to investigate the fundamental region of neural activity in healthy subjects, which is believed to be necessary for driving ability and to examine the effect of brain injury on driving safety.

Patients and methods: Experimental studies were performed on 8 healthy right-handed adults and 8 patients with moderate to severe TBI (5; resumed driving, 3; did not resume driving). The participants were asked to drive in the driving simulator. During driving, changes in oxy-Hb levels were measured using functional near-infrared spectroscopy at 34 sites including both hemispheres.

Results: The areas that showed significant activity in healthy subjects spanned from the frontal region to the temporal and parietal regions, and were more prominent in the right cerebral hemisphere than in the left. Patients who resumed driving showed similar patterns as healthy subjects; cortical activations near damaged regions were retained as seen on CT or MRI. However, the patients who could not resume driving showed no cortical activations near the lesions.

Conclusion: Our results provide further evidence that driving a car is a complex cognitive skill, and we believe that it is important to check the site of brain injury and the brain region where the damage has occurred before providing sufficient real car driving practice to patients with brain injury.

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Abstract – WCN 2013

No: 611

Topic: 10 – Neurorehabilitation

Effect of 8-week rebound therapy exercise on dynamic and static balance of male students affected by Down syndrome

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Objective: Balance is a complex motor skill that describes dynamic body posture that prevents the possibility of falling. Down syndrome is a genetic disorder in which chromosome 21 or part of it is tripled. Balance is not perfect in patients with Down syndrome. Rehabilitation may be effective in enhancing balance in these patients.

Rebound therapy is a rehabilitation program made possible by the therapeutic use of trampoline.

Methods: 30 students were enrolled in groups. They were matched with mean age of 18.54 years, and mean height of 164/2 cm. 15 persons as the test group voluntarily took part in the experiment (i.e. rebound therapy) with mean age of 19/36 years, mean weight of 74/35 kg, and mean height of 165/7 cm. To measure the static balance, BERG standard test was used and TGUG test was used for dynamic balance. The balance exercise program was performed by means of a trampoline which lasted 8 weeks, for 3 sessions of 20 minutes each week. For descriptive statistics, mean and standard deviation were used. KS test was employed for the group normalization, and in the section inferential statistics, t-test (dependent and independent) at $\alpha = 0.05$ was performed and ANOVA test for extra group comparison.

Results: Difference of the variables in posttest measure in the rebound therapy group is found significant, but it isn't significant in the control group.

Conclusion: It can be concluded that rebound therapy exercise is effective for improvement of static and dynamic balance in the male high school students affected with Down syndrome.

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Abstract – WCN 2013

No: 689

Topic: 10 – Neurorehabilitation

Gait rehabilitation in subacute hemiparetic stroke: Robot-assisted gait training versus conventional physical therapy

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Background: Despite improved management of patients after stroke, the majority of surviving patients are disabled with impaired walking function. Robot-assisted gait training was introduced as a potential strategy for gait rehabilitation in the severely disabled.

Objective: To compare the effect of robot-assisted gait training and conventional physical therapy on ambulation in subacute hemiparetic stroke patients.

Material and methods: Sixty non-ambulatory stroke subjects were randomly assigned to an experimental or control group. Both groups received a similar type and intensity of rehabilitation, except the gait training session. The experimental group received 30-minute robotic-assisted gait training plus a 30-minute conventional physical therapy while the control group received a 60-minute conventional physical therapy on every working day for four consecutive weeks. Gait function and disability were assessed before, one month after treatment, and 3 months follow-up, using the Functional Ambulation Category, Barthel Index, Berg Balance Scale, Resistance to Passive Movement scale, lower extremity part (REPAS), 10-meter walk test, and 6-minute walk test by a blinded assessor.

Results: The intention to treat analysis revealed significant higher scores of all measures, except the REPAS, in the experimental group compared with the control group at the end of the first and the third month post treatment ($p < 0.05$).

Conclusion: The robot-assisted gait training plus conventional physical therapy is significantly better than conventional physical therapy alone in subacute stroke patients as regards the ability of ambulation, activity of daily living, balance, gait speed, step length and endurance.

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Abstract – WCN 2013**No: 35****Topic: 10 – Neurorehabilitation****Brain electrical activity in stroke patients under transcranial magnetic stimulation therapy**G.E. Vinajera. *Neurophysiology EEG Department, Institute of Neurology and Neurosurgery, La Habana, Cuba*

Objective: A double-blind prospective and longitudinal study was carried out to assess electrical brain activity and to evaluate clinical evolution in a sample of 9 subjects with chronic stroke after rehabilitation and application of rTMS (1 Hz).

Methods: The sample in the study was randomly divided into two groups: 5 patients received sham rTMS and 4 patients received real rTMS both with daily sessions for 20 days. Electroencephalograms (EEGs) were recorded before and after rTMS. The neurophysiological measures used were the resting EEG power spectrum, Delta/Alpha ratio (DAR), the spike-frequency and the spike-amplitude. Clinical characterization was assessed using Scandinavian (SS) and Barthel Index (BI) scales.

Results: 1 Hz rTMS caused a tendency to increase ($p = 0.06$) in the Alpha band power spectra in both brain hemispheres. There was also a decreasing tendency of the Delta band power spectra in both brain hemispheres. DAR diminished 23% more in the 1 Hz rTMS group than in the sham rTMS group, and the spike-frequency also increased in the 1 Hz rTMS group after stimulations. Clinical scales after the rTMS showed a greater tendency to increase punctuations in the SS ($p = 0.06$) in the 1 Hz rTMS group.

Conclusions: Stroke patients who received 1 Hz rTMS sessions experienced modifications on resting EEG, suggesting a propensity to the cortical activation in both brain hemispheres and an increment in cortical excitability. The 1 Hz rTMS group had better clinical recovery and brain electrical activity, reflected in the modifications of the SS and DAR.

doi:10.1016/j.jns.2013.07.2010

Abstract-WCN 2013**No: 304****Topic: 10-Neurorehabilitation****Feasibility of speed of processing training in middle-aged and older adults with HIV: A pilot study**D.E. Vance^a, P.L. Fazeli^b. ^a*School of Nursing, University of Alabama at Birmingham, Birmingham, AL, USA;* ^b*Center for Nursing Excellence, University of Alabama at Birmingham, Birmingham, AL, USA*

Background: Nearly 50% of adults with HIV experience some form of cognitive deficit which can be observed in memory, attention, executive functioning, and speed of processing. In fact, as people age with this disease, this may predispose them for developing even more severe cognitive deficits that impact everyday functioning such as driving. Therefore, it is necessary to develop way to augment such cognitive abilities in lieu of such neurological manifestations.

Objective: The purpose of this study is to examine if a computerized home-based cognitive remediation training program is effective in improving cognitive functioning (i.e., useful field of view, a visual speed of processing measure important for safe automobile driving).

Patients and materials: In this single-group pre-post design experiment, middle-aged (40+) and older participants ($N = 20$) with HIV received a brief neuropsychological assessment at baseline, given a computerized cognitive remediation training program to be taken home and played for 10 hours, and then reassessed again approximately 6 weeks later.

Results: The attrition rate was 25%. On average, participants spent 8.20 ($SD = 3.43$) hours engaged with the cognitive training program.

From the remaining 15 participants, using an intention to treat approach, participants improved their visual speed of processing ($t(14) = 2.80, p = .014$).

Conclusion: These results are encouraging in that they demonstrate that adults with HIV vulnerable of developing cognitive deficits can benefit from cognitive training in the comfort and privacy of their homes. In addition, this particular cognitive ability is very important for driving and everyday functioning.

doi:10.1016/j.jns.2013.07.2011

Abstract – WCN 2013**No: 324****Topic: 10 – Neurorehabilitation****Neuropsychiatric evaluation of traumatic brain injury**A. Jain^a, R. Mittal^b, A. Sharma^b. ^a*Psychiatry, ESIC Model Hospital, India;* ^b*Neurosurgery, SMS Medical College, Jaipur, India*

Introduction: Assessment and treatment of TBI typically focus on physical and cognitive impairments, yet psychological impairments represent significant causes of disability and poor quality of life. Depression and anxiety may be the most common and disabling psychiatric condition in individuals with TBI.

Aims and objective: To investigate the prevalence and determinants of depression and anxiety in patients of TBI and its relationship with injury characteristics. To understand the relationship of depression and anxiety to quality-of-life outcome in TBI.

Material and method: 156 Mild to moderate TBI patients of 18 years or older age were included in this study after applying appropriate inclusion and exclusion criteria. Apart from recording their sociodemographic profile and injury history, all the participants were assessed with PHQ-9, GAD-7, WHOQOL-BREF and GCS to assess depression, anxiety, quality of life and severity of trauma respectively. All the patients were subjected to neuroradiology to localize sites of injury.

Results: Depression and anxiety was found in 46 vs 29% cases respectively with majority (60%) of depressed cases qualifying for moderate severity as compared to anxiety. First three months after TBI was associated with maximum occurrence of depression and anxiety (68% vs 54%). Strong correlation was found between severity of trauma and severity of depression and anxiety. Left temporal and frontal lobe involvement predominantly caused depression. Depressed and anxious patients had poor quality of life.

Conclusion: TBI patients must be evaluated for concomitant moods and anxiety disorders. Timely treatment for these disorders may improve overall outcome and quality of life.

doi:10.1016/j.jns.2013.07.2012

Abstract – WCN 2013**No: 336****Topic: 10 – Neurorehabilitation****Cortical endogenic neural regeneration of adult rat after traumatic brain injury**X. Yi, G. Jin, X. Zhang, W. Mao, H. Li, J. Qin, J. Shi. *Nantong University, Nantong, China*

Background: Focal and diffuse neuronal loss happened after traumatic brain injury (TBI). With little in the way of effective repair, recent interest has focused on endogenic neural progenitor cells (NPCs) as a potential method for regeneration.

Objective: To determine whether endogenic neural regeneration happens in the cortex of adult rats after traumatic brain injury.

Material and methods: Adult Sprague–Dawley rat models of medium TBI were induced by controlled cortical impact and were divided into sham group and TBI group. Rats were injected with bromodeoxyuridine (BrdU) at 1–7 days post-injury (dpi) to allow identification of differentiated neurons and sacrificed at 1, 3, 7, 14 and 28 dpi for nestin/sox-2, GFAP/sox-2, DCX/BrdU and NeuN/BrdU immunofluorescence.

Results: Nestin/sox-2 NPCs or GFAP/sox-2 double positive radial glial-like cells emerged in injured cortices and peaked at 3 dpi. The number of GFAP/sox-2 double positive cells was less than nestin/sox-2 double positive cells. Nestin/sox-2 double positive cells from posterior periventricle (pPV) immigrating into injured cortices through the corpus callosum were found. DCX/BrdU double positive neuronal progenitors in peri-injured cortex were found at 3, 7, 14 dpi and peaked at 7 dpi. NeuN/BrdU double positive mature neurons were not found in injured cortices at 1, 3, 7, 14 and 28 dpi.

Conclusion: NPCs from pPV and reactive radial glial-like cells emerged in peri-injured cortices of adult rats after TBI and differentiated into neuronal progenitors, but the latter failed to grow up to mature neurons.

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Abstract – WCN 2013

No: 296

Topic: 10 – Neurorehabilitation

Rho kinase inhibitor fasudil regulates the polarization and function of bv-2 immortalized microglial cell line

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Background: Increasing data demonstrate that macrophage/microglia have different phenotypic and functional states, and may exert toxic or protective effects in neurological disorders depending on microenvironment. M1 polarization is associated with inflammatory responses, while M2 results in anti-inflammatory effects.

Objective: In this study, we try to explore characteristics of microglia polarization and investigate whether Fasudil can influence the polarization and function of microglia.

Material and methods: A maintained BV-2 immortalized microglial cell line was treated with LPS and/or Fasudil. A BV-2 conditioned medium treated with LPS and/or Fasudil was co-cultured with PC12 cells.

Results: M1 markers iNOS and IL-12 of BV-2 were elevated after LPS stimulation, but significantly declined after Fasudil treatment. M2 marker CD23 was significantly decreased after LPS stimulation ($p < 0.001$), and elevated after Fasudil treatment ($p < 0.05$). Simultaneously, LPS stimulated iNOS up-regulation ($p < 0.05$) and NO production ($p < 0.001$), but did not influence Arg-1 expression. Fasudil significantly inhibited iNOS expression ($p < 0.05$) and NO production, enhanced Arg-1 expression ($p < 0.05$). In LPS-stimulated mouse brain, LPS also induced iNOS expression on CD11b + microglia, while Fasudil inhibited its expression. In contrast, Arg-1 expression on CD11b + microglia was enhanced by Fasudil treatment. Fasudil inhibited NF- κ B activity and inflammatory cytokines IL-1b, IL-6 and TNF- α , but increased anti-inflammatory cytokine IL-10, leading to neuroprotective effects in cultured PC12 neurons by a Fasudil-conditioned medium.

Conclusion: These results clearly indicate that Fasudil can convert M1 microglia to an M2 phenotype, inhibiting inflammatory response by controlling a specific polarization in microglia/macrophages. This study was supported by the National Natural Science Foundation of China (81272163).

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Abstract – WCN 2013

No: 535

Topic: 10 – Neurorehabilitation

Impact of two training packages for caregivers on quality of care of neurosurgery patients – A randomized controlled trial

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Introduction: Family caregivers of neurosurgery patients function as informal extensions of the health system but they are untrained and unprepared for their new role. It has been felt that their problems related to care provision can be resolved by appropriate training.

Objectives: To compare the impact of two training packages on knowledge and practices of family caregivers of operated neurosurgery patients.

Materials and methods: A randomized controlled trial was done among the operated neurosurgery patients and their caregiver dyads ($n = 90$). They were randomly allocated to receive training package 1 (TP1 = self instruction manual & one to one training) or training package 2 (TP2 = self instruction manual only). Sequentially numbered sealed envelopes were used for allocation concealment. Primary outcome measure was knowledge gain of the caregivers. A variety of secondary outcome measures was also assessed in three monthly follow ups.

Results: The attrition rate was 15.5%. Intention to treat analysis was followed. Caregivers in the TP1 group had significant knowledge gain (95% CI of mean difference = 9.4,14.5, Bonferroni corrected p value < 0.05). The number of caregivers who followed correct care giving practices was significantly greater in TP1 group. Life satisfaction of the caregivers in the TP1 group improved significantly. We could not find any significant improvement in the degree of patients' disability and in the caregivers' strain between the groups.

Conclusions: Training of caregivers by providing information along with one to one training is an effective strategy for improving the knowledge and skills of caregivers regarding care provision to the operated neurosurgery patients.

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Abstract – WCN 2013

No: 471

Topic: 10 – Neurorehabilitation

Electro-echocardiographic correlation in chronic dialyzed patients and short rehabilitations in ischemic stroke

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Background: The AURORA study analyses the occurrence of stroke and cardiac events in patients on haemodialysis treated with Rosuvastatin.

Objective: Our aim was to evaluate the efficacy of the different electrocardiographic (ECG) indexes in detecting left ventricular hypertrophy (LVH, concentric or eccentric) among patients with chronic renal failure under dialysis and carotid ischemic stroke as a prognostic factor.

Subjects and method: We assessed 59 dialyzed-patients (median age of 55 +/- 2 years, 30 men) between 2010 and 2012, and followed up for 1 month after the ischemic stroke.

The neurological evaluation was performed using the NIHSS scale and brain MRI. For diagnosis of the ECG-LVH, the following indexes were considered: Sokolov > 35 mm, Gubner > 22 mm, Romhilt > 5 points, Cornell > 28 mm in men and > 20 mm in women and Lewis > 17 mm.

Results: ECHO-LVH was detectable in 70% of all the patients, 29% with concentric hypertrophy (CH) and 22% with eccentric hypertrophy (EH). Considering LV geometry, the Romhilt index also demonstrated superiority in the detection of both CH (SE > 26%) and EH (SE > 36%). All patients presented major hemispheric ischemic stroke syndrome, defined an NIHSS >= 12 in the territory of the middle cerebral artery (MCA) or internal carotid artery (ICA) and a GCS score < 14. The MRI showed hypersignal T2, lesions primarily in the large MCA territory, in all cases.

Conclusions: 30 patients who were dialyzed, and had ECHO-LVH, and Sokolov > 35 mm died in the first 3 weeks after stroke. The dialysis patients with cardiac hypertrophy and ischemic stroke have a greater mortality, due to cardiac disease. Early rehabilitation have poor outcomes in patients.

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Abstract — WCN 2013

No: 473

Topic: 10 — Neurorehabilitation

Aortic plaque thickness and diabetes as a risk predictor for ischemic stroke rehabilitation

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Background: An increased aortic plaque thickness in diabetic patients with ischemic stroke is demonstrated, but data regarding the value as a prognostic risk factor for the cognitive stroke outcome is limited.

Objective: To identify aortic plaque thickness as a risk predictor in patients with diabetes and acute ischemic stroke.

Patients and method: We assessed 95 patients, mean age = 69 ± 12 years, (51 male) with first-ever acute stroke confirmed by brain MRI and an initial post-stroke MMSE score > 15 (mean interval between stroke and examination, 6.5 ± 3 days). All present type 2 diabetes mellitus (T2DM), and insulin therapy was initiated during hospitalization. Carotid plaques were classified as simple or complex on the basis of thickness >= 4 mm, ulceration, or mobility. The NIHSS scale was performed in days 4–7.

Results: The mean concentration (HbA(1c)) was 0.9% lower for the patients with intensive treatment. The number of patients with the aortic plaque thickness mean of 3.4 mm (2.3 to 3.6 mm range) was significantly higher (p > 0.047), and the median NIHSS score in patients with thickness >= 4 mm was 14 (range 3 to 38), and scores differed depending on the arteriographic findings (P < 0.001). The MMSE score at 7 days was 18.6, and 25.6 at 3 months with major difficulties in attention, short-term memory impairment and aphasia.

Conclusions: Carotid plaque thickness is a simple and noninvasive marker of subclinical atherosclerosis and it is often associated with acute ischemic stroke in patients with type 2 diabetes. Hyperglycemia

and MMSE score could not predict deterioration or improvement in cognitive outcome.

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Abstract — WCN 2013

No: 17

Topic: 10 — Neurorehabilitation

Neuroprotective activity of combination of progesterone and curcumin on sub-acute phase changes induced by partial global cerebral ischemia in mice

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Background: There is need to search for neuroprotective agents in stroke to contain morbidity and mortality.

Objective: Progesterone, a neurosteroid and curcumin, a phytophenol were evaluated in combination as neuroprotective agents in a bilateral carotid artery occlusion (BCAO) model in mice.

Material and methods: 48 albino Swiss mice were divided into 4 groups: sham operated, DMSO (0.2 ml)- , Progesterone (15 mg/kg)- and progesterone (5 mg/kg) + curcumin (100 mg/kg)-treated BCAO groups. BCAO was produced for 10 min with aneurysmal clips. The animals were subjected to behavioral, brain biochemical and histopathological studies on day 15 while drug or vehicle treatment was given for 14 days following BCAO. Drugs were administered intraperitoneally. The protocol was approved by the Institutional Animal Ethics Committee.

Results: Compared to the sham group, the BCAO (DMSO treated group) resulted in significant impairment of exploratory behavior (hole-board test) and motor co-ordination (rota rod test) while the seizure susceptibility to kainic acid and anxiety level (elevated plus maze test) were significantly increased; BCAO caused a significant increase in histopathological scores and brain levels of malondialdehyde and TNF- α ; and a significant decrease in the levels for superoxide dimutase, catalase and glutathione peroxidase. Progesterone-alone caused significant reversal of the values for all the parameters as observed for the DMSO treated group following BCAO. The combination of progesterone and curcumin showed a significant synergistic improvement for all the parameters as compared to those observed with DMSO and progesterone-alone treated groups following BCAO.

Conclusion: The combination of progesterone and curcumin possesses a translational value as potential synergistic neuroprotective agents in stroke.

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Abstract — WCN 2013

No: 49

Topic: 10 — Neurorehabilitation

Complications of intrathecal baclofen pump: Prevention and cure

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Increasingly, spasticity is managed with surgically implanted Intrathecal Baclofen pumps. Intrathecal Baclofen pump revision surgery unrelated to programmable pumps during end-of-life is not uncommon, requiring special attention during pre-, intra-, and post-operative management. We aimed to identify and describe complications of

Intrathecal Baclofen pump as well as to report avoidance and management of complications.

Methods and materials: Through 2002–2006, at the department of neurosurgery, Henry Ford and Oakwood Health Systems, Intrathecal Baclofen pumps were implanted in 44 patients; 24 children versus 20 adults; 30 «primary-implant-patients»; 14 «revision-only patients». We evaluated reasons for revision surgeries and diagnostic work-up requirements.

Results: Eight out of 30 «primary-implant-patients» required 14 revisions and 7 of 14 «revision-only-patients» needed 13 procedures. Seven patients with slowly increasing baclofen-resistant spasticity had either:

- i) Unsuspected pump–catheter connector defects [N = 4] with a subcutaneously dislocated intrathecal catheter (N = 1)
- ii) An X-ray-documented pump–catheter connector defect [N = 1]
- iii) An X-ray-demonstrated fractured catheter with intrathecal fragment [N = 2] requiring laminectomy (N = 1). Injection studies revealed intrathecal peri-catheter arachnoiditis, managed without laminectomy (N = 1), and connector-related dye leakage [N = 3]. Implant infections occurred in 4 cases [3 were pre-operated]. Scintigraphy revealed occult CSF leakage [N = 1]. Intrinsic pump failure [N = 1].

Conclusions: Intrathecal Baclofen pumps, although very gratifying, have a high, technique-related complication incidence during implant life. Meticulous technique, high clinical suspicion, appropriate work-up, and timely surgical management can reduce surgical complications of Intrathecal Baclofen pump implantation.

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Abstract – WCN 2013

No: 144

Topic: 10 – Neurorehabilitation

Impact of mild traumatic brain injury on language skills:

Pre- and post-injury functioning

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Objective: Pre- and post-injury language performance scores following mild traumatic brain injury (mTBI) in a child were investigated through a replication and subsequent extension of a previous case study where a trend for declining language scores was described based on pre- and post-injury data.

Patient and methods: The case child was a control participant in a longitudinal language study being undertaken at the University of Queensland, Australia. He was 8 years and 7 months of age at his initial language assessment. Twelve months after his initial assessment, he sustained a head injury following a fall from a non-motorised scooter.

His Glasgow Coma Scale and Emergency Medicine documentation met the American Congress of Rehabilitation Medicine's classification criteria of mTBI. The child returned to the University of Queensland for subsequent language assessment at age 10 years and 7 months. Test norms were used to descriptively analyse the child's pre- and post-morbid language performance and performance score changes. Score changes were subsequently statistically analysed using data obtained from a group of age-matched non-brain-injured control children.

Results: Descriptive analysis identified a trend for declining skill on general and high-level language skills in the child. Statistical analysis failed to differentiate the child with mTBI from the control group on language score changes.

Conclusions: While supporting the earlier identified trends in language decline following mTBI, subsequent statistical analysis undertaken to extend the earlier findings failed to detect statistically significant changes in language outcomes following mild injury. The need for monitoring of yet-to-emerge higher-level adolescent language skills, however, is highlighted.

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Abstract – WCN 2013

No: 178

Topic: 10 – Neurorehabilitation

The efficacy of neuroaid in stroke recovery: A meta-analysis

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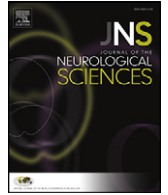
Background: Stroke is the third leading cause of death worldwide and is a major cause of morbidity. To date, there is no effective treatment in reducing chronic disability secondary to stroke. NeuroAid is a new treatment option under investigation in stroke rehabilitation research. The Fugl Meyer Assessment (FMA) Scale is the gold standard in measuring sensorimotor function in hemiplegic post-stroke patients against which all other scales are assessed.

Objectives: To determine the efficacy of NeuroAid on motor recovery using the FMA scale in patients with ischemic stroke by meta-analysis.

Results: The meta-analysis included 2 double-blind placebo-controlled studies on NeuroAid. A random effects model and a 95% confidence interval was used which showed no statistical significance with a *p* value of 0.331 at 4 weeks post-treatment and a *p* value of 0.088 at 8 weeks post-treatment.

Conclusion: This meta-analysis did not significantly demonstrate the efficacy of NeuroAid in the motor recovery of patients with ischemic stroke when compared to placebo.

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Topic: 36 - Other Topic

Abstract – WCN 2013

No: 184

Topic: 36 – Other Topic

Immunological mechanisms of pathogenesis of chronic brain ischemia

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Background: Immunological mechanisms have significant value in the pathogenesis of chronic brain ischemia (CBI).

Purpose: The purpose of investigation was to study immunological status and efficiency of cortixin in the treatment of the patients with CBI.

Material and methods: We examined 187 patients with CBI. The patients were divided into 2 groups: I group included 98 patients who were treated with cortixin, group II consisted of 89 patients receiving therapy without cortixin. The levels of IL1 β , TNF α and antibodies to myelin basic protein (MBP) in blood serum were determined by immunoenzymatic analysis. Cortixin was injected in a dose of 10 mg during 10 days.

Results: The level IL1 β in patients of group I before treatment was 18.4 ± 0.8 pg/ml, after 14.0 ± 0.5 pg/ml ($P < 0.05$). In patients of group II the level of IL1 β before treatment was 17.9 ± 0.6 pg/ml, after 16.8 ± 0.5 pg/ml. The level of TNF α in patients of group I before treatment was 13.2 ± 0.7 pg/ml, after decreased in 1.5 times. In group II before treatment the level of TNF α was 12.8 ± 0.7 pg/ml, after 11.9 ± 0.6 pg/ml. At the first day of study patients observed high levels of antibodies to MBP ($0.202 \text{ ng} \pm 0.011 \text{ un.opt.dens.}$). The levels of antibodies to MBP at the 10th day of treatment showed that in patients of group I this parameter was $0.152 \text{ ng} \pm 0.02 \text{ un.opt.dens.}$ In group II the levels of antibodies to MBP were decreased by 18.6%.

Conclusion: Cortixin resulted in a decrease of the levels of proinflammatory cytokines that specify its ability to reduce activity of immune inflammation.

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Abstract – WCN 2013

No: 145

Topic: 36 – Other Topic

Childhood intrathecal chemotherapy: Language outcomes 11 years on

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Background and objective: Studies suggest that major language indices do not differentiate children treated for acute lymphoblastic

leukaemia (ALL) with intrathecal chemotherapy (ITC) from control children matched on age, gender and educational level. This case-control study used a sibling as a control to investigate language outcomes in a male child 11 years after the administration of ITC for ALL.

Method: The Index case was a male aged 13 years 8 months at the time of his involvement in the study. At age 2 years 3 months he was diagnosed with standard risk ALL and was treated with ITC. For the current study, the Index case underwent behavioural assessment of language skills and neurophysiological assessment of language processing efficiency, with performance measures being descriptively compared to performance by his younger sibling.

Results: Behavioural language testing failed to differentiate the siblings on current language performance, but neurophysiological assessment revealed that the ITC-treated child required more time to process information during a picture-word matching task.

Conclusions: The study's findings offer pilot data of language outcomes following ITC beyond the early stage of survivorship. Information processing speed is important for skill and knowledge acquisition in normal development and it has been proposed that a slowed rate of information processing may be the first indicator of emerging neurocognitive deficits in children following CNS-directed treatments. Thus the Index case may be at risk of "growing into deficit" as he develops adolescent language skills. The benefits and limitations of using siblings in research investigating cognitive outcomes are discussed.

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Abstract – WCN 2013

No: 164

Topic: 36 – Other Topic

Management of post-hemorrhagic hydrocephalus in preterm infants with germinal matrix hemorrhage

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Study design: A prospective study over the period from Jan 2008, to June 2011, by the Department of Neurosurgery, in Mansoura University Hospitals. All preterm babies admitted to neonatal care unit who developed symptoms suggestive of neurological insult were investigated to detect the cause of this insult.

Objectives: Neonatal intraventricular hemorrhage in preterm infants resulting from germinal matrix hemorrhage with subsequent post-hemorrhagic ventricular dilatation and hydrocephalus (PHH) should be managed adequately to minimize neurological morbidity and mortality.

Patients and methods: All preterm infants who are admitted to neonatal unit in Mansoura Children Hospital were eligible to our study. Once we suspect a germinal matrix hemorrhage (GMH), cranial

ultrasound, and then CT scan brain were done for +ve cases to establish the diagnosis and exclude grades 1 and 4 according to papile, a grading system, only patients with grades 2 and 3 are involved in our study.

Results: 33 patients with GMH were involved in our study 21 case (grade 2) and 12 case (grade 3). Ventriculoperitoneal shunts were inserted only for 17 cases who developed post-hemorrhagic hydrocephalus as proved by clinical and radiological follow up.

Conclusion: Management of PHH in preterm infants is a debatable issue that needs a careful selection of the treatment modality. Ventriculoperitoneal shunts are needed in a significant proportion of preterm infants with grade II, and grade III GMH who survived and developed PHH.

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Abstract – WCN 2013

No: 192

Topic: 36 – Other Topic

Diagnostic and clinical peculiarities of PANDAS

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Last years was advanced supposition on connection different choreiform hyperkinetic disorders, tics, myoclonias and neurosis like fived states in children with B-hemolytic streptococcus group A (BHSGA).

Aim of study: To research peculiarities of course for PANDAS.

Materials and methods of study: Examined 17 children at the age from 7 to 15 years with complaints on hyperactivity, chorea form and tic like hyperkinetic disorder, including generalized tic emotional lability, absent-mindedness, disorder of attention, problem with sleeping, and obsessions.

Results of study: Acute beginning and wave shaped course with the following full or partial remission were in all 17 patients, and, it has chrome logical connection with acute chronic tonsillitis, and an increase of temperature. In the smears from the fauces children revealed BSHGA, moderately positive were acute phase indices (CPB, cryoglobulins and raise of antistreptolysin-o titers). Symptoms of dysgenia for connective tissue (hypermobile syndrome, dysplastic cardiopathy) were present in 9 children. In 5 people from 11 being examined at MRT-angiography pathologic convolution of carotid and vertebral arteries was revealed. In 9 from 17 patients at record of sleep diagnosis of dysplastic or metabolic cardiopathy was made. By the date of EchoCg Prolapse of mitral valve of the first level was revealed. Confirming criteria for rheumatism were not revealed.

Conclusion: Today neurologists together with cardiologists have all bases to review ordinary imaginations on the treatment of neurosis like, choreiform, hyperkinetic disorders in children. From this contingent it is necessary to divide the patients with PANDAS, and, to use new tactics in therapy.

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Abstract – WCN 2013

No: 177

Topic: 36 – Other Topic

Atypical neurological manifestations of lymphomatous malignancy in immunocompetent patients: Case series of four patients

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Background: Lymphomatous malignancies in immunocompetent patients may masquerade as infective and inflammatory disorders of the central or peripheral nervous system which may delay the diagnosis. We discuss how high index of suspicion with supporting CSF, imaging, PET and pathology may aid in the diagnosis.

Objective: To describe the atypical clinical features of patients of lymphomatous malignancy presenting with neurological findings.

Patients and methods/material and methods: We present the clinical features of four different patients with varied clinical syndromes; all of them were immunocompetent, relatively young and mimicking infectious and autoimmune diseases.

Results: The first patient had febrile illness with acute disseminated encephalomyelitis (ADEM) like presentation with a fulminant course. Repeated CSF revealed Burkitt's lymphoma. The second patient presented with seizures with speech and swallowing difficulty with imaging suggestive of vasculitis but biopsy revealing Non-Hodgkin's lymphoma. The third patient presented with a febrile episode followed by catatonia and mutism. PET brain was suggestive of primary CNS lymphoma. The fourth patient presented with multiple radiculopathies and cranial neuropathy with CSF showing abnormal cells. All these patients were immunocompetent.

Conclusion: Lymphoid malignancies can present with fever of unknown origin especially in young patients. Biopsy and repeated CSF cytology can help to cliché the diagnosis in various suspected cases. Neuroimaging features of lymphoid malignancies may also mimic infectious, inflammatory, demyelinating and vasculitis. Biopsy is warranted to establish the diagnosis. PET scan may give a helpful lead in establishing the diagnosis where biopsy may not be possible.

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Abstract – WCN 2013

No: 208

Topic: 36 – Other Topic

Bilirubin enhances neuronal excitability by increasing glutamatergic transmission in the rat lateral superior olive

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Hyperbilirubinemia is one of the most common clinical phenomena observed in human newborns. To achieve effective therapeutic treatment, numerous studies have been done to determine the molecular mechanisms of bilirubin-induced neuronal excitotoxicity. However, there is no conclusive evidence for the involvement of glutamatergic synaptic transmission in bilirubin-induced neuronal hyperexcitation and excitotoxicity. In the present study, using gramicidin-perforated patch-clamp techniques, spontaneous excitatory postsynaptic currents (sEPSCs) were recorded from lateral superior olive (LSO) neurons isolated from postnatal 11–14-day-old (P11–14) rats. The application of 3 μ M bilirubin increased the frequency, but not the amplitude, of sEPSCs. The action of bilirubin was tetrodotoxin (TTX)-insensitive, as bilirubin also increased the frequency, but not the amplitude, of mEPSCs. The amplitudes of GABA-activated (I_{GABA}) and glutamate-activated (I_{glu}) currents were not affected by bilirubin. Under current-clamp conditions, no spontaneous action potentials were observed in control solution. However, the application of 3 μ M bilirubin for 4–6 min evoked a considerable rate of action-potential firing. The evoked firing was partially occluded by d,l-2-amino-5-phosphonovaleric acid (APV), an NMDA receptor antagonist, but completely inhibited by a combination of APV and 2,3-dihydroxy-6-nitro-7-sulfamoyl-benzo[f]quinoxaline-2,3-dione (NBQX), an AMPA receptor antagonist. These results indicate that bilirubin facilitates presynaptic glutamate release, enhances glutamatergic synaptic transmission by activating postsynaptic AMPA and NMDA receptors, and

leads to neuronal hyperexcitation. This study provides a better understanding of the mechanism of bilirubin-induced excitotoxicity and determines for the first time that both AMPA and NMDA receptors are likely involved in the excitotoxicity produced by bilirubin.

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Abstract – WCN 2013

No: 195

Topic: 36 – Other Topic

Clinical and neurophysiological features of 79 children with developmental and benign movement disorders

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Background: Developmental and benign movement disorders (DBMD) are characterized by the absence of associated neurological manifestations and by their favorable outcome.

Objective: To analyze clinical and neurophysiological characteristics of the DBMD cases evaluated in our center.

Patients and methods: We recruited patients from our center during the past two years, including children with normal neurologic and psychomotor development. Children with abnormal interictal or ictal EEG, abnormal MRI findings were excluded. Neurological examination, detailed history taking, EEG, and video monitoring were performed.

Results: 79 patients (38 male) were diagnosed with DBMD. The ages at the time of admission ranged between 1 month and 14 years. 52 cases (66%) started within the first year of age versus only 34% of the cases with onset after 12 months. The diagnosis showed a wide spectrum of DBMD including stereotypies 15/79 (19%); benign myoclonus of early infancy 9/79 (11.4%); benign jitteriness of newborns 9/79 (11.4%); transient dystonia of infancy 8/79 (10%); gratification behavior in early childhood 7/79 (8.9%); sleep-related rhythmic movement disorders 6/79 (7.6%); benign paroxysmal torticollis 6/79 (7.6%); benign neonatal sleep myoclonus 5/79 (6.3%); mirror movements 5/79 (6.3%), shuddering attacks 4/79 (5.1%); spasmus nutans 2/79 (2.5%); paroxysmal tonic upgaze 2/79 (2.5%); and Sandifer's syndrome 1/79 (1.3%).

Conclusion: Recognition and management of DBMD depend from careful neurological examination, detailed history taking, and home video recordings. We revealed predominance of stereotypies, benign myoclonus in structure of DBMD. Proper identification of DBMD is crucial, as it avoids unnecessary concern, unnecessary and costly investigations, and ineffective and potentially toxic treatments.

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Abstract–WCN 2013

No: 201

Topic: 36–Other Topic

Case observation of polyneuropathy as a predicative factor of possible neoplastic process

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Background: According to the recent medical studies a paraneoplastic polyneuropathy that becomes clinically evident before

neoplasm diagnosing or metastasis verification is observed in almost 50% of cancer suffering patients.

Objective: To detect the probable interdependence between polyneuropathy development process and subsistence of earlier undiagnosed neoplasm development.

Material and methods: 38 patients with confirmed neoplastic diagnosis (blood disease, lung cancer, ovarian carcinoma, urinary bladder cancer, breast cancer, stomach cancer) were observed in Angioneurology Department of Institute of Urgent and Recovery Surgery n.a. V.K. Gusak presented the first study group. The second group numbered 24 patients with primary diagnosed polyneuropathy. All 62 patients went through the careful neurologic observation. NSS (Neuropathy Symptom Score) and Dyck P.J. (1988) scale determined the polyneuropathy severity grade. Supplementary tests consisted of general clinical examinations and oncomarker analysis (beta-2-microglobulin, Ca 15-3, Ca-125, Ca 19-9, CEA, HCG, Ca 15-3, PSA). EMG with using 4-channel electromyography "Reporter" (Biomedica, Italy) was the principal method.

Results: 49% of patients with active neoplasm got such polyneuropathy EMG-markers as NCV (Nerve Conduction Velocity) lowering to 22–30 m/s as well as reducing amplitude of M-answer by 2–2.5 times. In 8 patients (37%) of the second group a detailed clinical observation and oncomarker analysis showed subclinical neoplastic disease.

Conclusion: An active neoplastic process is very often accompanied by polyneuropathy. The lowering of nerve conductance velocity by EMG and general polyneuritic clinical scene of a patient needs to be examined by somatic detailed test that sometimes allows finding out a neoplastic progress much earlier.

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Abstract – WCN 2013

No: 244

Topic: 36 – Other Topic

Neurological and neuroradiological manifestations of acute dengue fever infection – a single centre study from tropical country

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Introduction: Dengue virus is generally considered as nonneurotropic, apart from case reports there is paucity of data available on neurological manifestation of acute dengue viral infection.

Methods: A prospective single centre study done at KIMS Hyderabad, during Nov 2007 to Jun2011. Acute dengue virus infection defined as positive serology for IgM antibodies or PCR positive for dengue. CT/MRI brain, EEG and other necessary lab investigations were done. Patients admitted with acute dengue virus infection studied for neurological and radiological manifestations of acute dengue virus infection and followed for next 6 months. Detailed statistical analysis done with SPSS ver 17 software.

Results: Out of 281 patients with acute dengue fever/DHF who were admitted in our hospital during the study period, 31 (11.08%) had neurological manifestations. Age was 14 to 68 years with mean age of 36.6 years with M:F ratio 2:1. Mean GCS on admission was 11.8. Diffuse slowing to be the commonest finding and there was no correlation of EEG findings on hospital stay or outcome [p = 0.459]. On MRI Pontine T2 hyperintensity and blooming in the thalamus were significantly associated with hospital stay [p < 0.005]. GCS at admission has significant correlation with hospital stay, neurodeficits or condition at discharge & outcome measures [p < 0.005].

Discussion: Neurological manifestations of dengue present in wide range as encephalitis at one end and GBS or myositis at other end. Dengue associated encephalopathy should be considered as differential diagnosis in patients having fever, thrombocytopenia and recent onset neurological deterioration or in patients having acute encephalitis/encephalopathy of unknown cause. MRI is a useful aid in diagnosis and GCS on admission is an important prognostic factor.

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Abstract – WCN 2013

No: 227

Topic: 36 – Other Topic

Neuropsychological deficits in ADHD: Development of a classification system

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Background: Recent experimental literature on ADHD has identified unique underlying cognitive dysfunction, specific to ADHD. Therefore, there is a need to incorporate information on cognitive mechanisms underlying ADHD and inculcate such information in the diagnostic system, which would provide a more sensitive as well as specific tool in differential diagnosis of ADHD.

Objective: The present study evaluated the diagnostic capabilities of certain measures based on cognitive-motivational tests that were chosen with respect to the specific cognitive deficits in Attention Deficit Hyperactivity Disorder (ADHD).

Methods: A total of 240 children (120 with ADHD and 120 healthy controls) in the age range of 6–9 years and 32 children with Oppositional Defiant Disorder (ODD) (aged 9 years) participated in the study. Stop-signal, attentional-disengagement, attentional-network, and choice-delay test were administered to all the participants. A total of nineteen parameters were derived from the four tests. Receiver operating characteristic (ROC) analysis and multinomial logistic regression (MLR) was performed to examine the diagnostic efficiency of each parameter.

Results: MLR was performed for combining parameters across tests. Data fusion produced improved overall diagnostic accuracy and a combination of stop-signal reaction time, post-error-slowness, mean delay, switch costs, and %LDR produced overall classification accuracy of 97.8% and with internal validation, the overall accuracy was 92.2%.

Conclusions: Combining performance from different tests enabled an accurate classification of ADHD children from healthy controls and those with ODD. The study has theoretical and clinical implications for the theories of executive control mechanisms that underlie the pathology of ADHD.

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Abstract – WCN 2013

No: 60

Topic: 36 – Other Topic

Distinctive behaviors of SK-N-MC cells towards menadione- and hydrogen peroxide-induced toxicities

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Background: There is an increasing body of evidence indicating that oxidative stress plays a vital role in the pathogenesis of neurodegenerative diseases. Nerve cells are incessantly exposed to environmental

stresses leading to overproduction of some harmful species like reactive oxygen species (ROS). ROS including hydrogen peroxide and superoxide anion are potent inducers of various signaling pathways encompassing MAPKs and JAK-STAT pathways. JNK/p38 MAPKs, JAK2 and STAT3 are deemed stress-responsive factors involved in oxidative stress-induced cell death.

Objectives: We scrutinized the effects of hydrogen peroxide and superoxide anion on SK-N-MC neuroblastoma cells to elucidate the mechanism by which each oxidant modulated above-mentioned pathways leading to SK-N-MC cell death.

Methods: SK-N-MC cells were exposed to hydrogen peroxide and the superoxide anion donor, menadione. Phosphorylation of MAPKs, JAK2 and STAT3 and cell death were investigated.

Results: Hydrogen peroxide and superoxide anion induced distinct responses in SK-N-MC cells as we showed that unlike JAK2 which was activated by both oxidants, STAT3 and p38 were activated in response to hydrogen peroxide and not superoxide radicals in SK-N-MC cells and menadione induced JNK-dependent p53 expression and apoptotic cell death which were not detected in H₂O₂-induced JNK activation.

Conclusion: ROS type has a key role in selective instigation of JNK/p38 MAPKs and JAK2-STAT3 pathways in SK-N-MC cells. Identifying these differential behaviors and mechanism of function illuminates therapeutic targets in the prevention or treatment of ROS-induced neurodegenerative diseases such as Alzheimer's disease.

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Abstract – WCN 2013

No: 233

Topic: 36 – Other Topic

Brain wave symmetry in depressive patient's electroencephalography

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Background: One of the most frequent diagnosis of mental disorder is depression. Depression is a common disease among people that need help for psychological problems. Advances in neuroimaging and in quantitative electroencephalography (QEEG) have made it possible to examine features of brain activity that are associated with response. Some of studies suggest that depressed persons in compare of healthy subjects have different brain wave activity. The aim of this study was to find out the probability of hemispheric asymmetry in depressive patients through EEG.

Material & methods: This is a descriptive-comparative study in which 15 depressive persons from among psychiatric clinic clients were selected and studied. The artifact free EEG epoch was analyzed by fast Fourier transformation (FFT) and relative power values of particular frequency bands (delta, theta, alpha and beta) were observed on the brain regions. The results were analyzed by MANOVA analysis.

Results & conclusion: The results of investigation indicated, that depressive patients have hemispheric asymmetry in alpha activity of frontal, temporal and parietal regions. This means that depressive persons have greater alpha activity in right rather than left hemisphere. In addition there is a decrease in theta activity in central and occipital area compared with healthy subjects. These results suggest that the disorder changes the brain wave activity.

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Abstract – WCN 2013**No: 234****Topic: 36 – Other Topic****Comparison of brain wave patterns in aggressive and non aggressive subjects**

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Background and objectives: Human aggression is an important social and clinical problem. There are many studies in determining the criteria for diagnosis and assessment of aggression. The aim of this study was to study aggressive subject's brain wave activity compared with healthy subjects by qEEG.

Materials and methods: The research method was contrastive–descriptive over which some 30 people of aggressive persons compared and contrasted with some 30 nonaggressive persons were demographically similar to aggressive group. The artifact free EEG epoch was analyzed by fast Fourier transformation (FFT) and power values of particular frequency bands (delta, theta, alpha and beta) were observed on the brain regions.

Results and conclusion: Results of variance analysis showed that aggressive subjects' theta activity in frontal, central, parietal and temporal areas is lower than normal subjects. In addition there is an increase in beta activity in frontal, central, parietal and temporal areas compared with nonaggressive subjects. These results suggest that the disorder changes the brain wave activity.

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Abstract – WCN 2013**No: 258****Topic: 36 – Other Topic****Tongue somatosensory evoked potentials in microvascular decompression treated trigeminal neuralgia**

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Background: Somatosensory evoked potentials of the tongue (tSSEP) provide useful information about trigeminal afferent pathway. The aim of this study was to evaluate tSSEP in trigeminal neuralgia treatment with microvascular decompression.

Methods: Two patients with trigeminal neuralgia refractory to conservative treatment underwent microvascular decompression of the trigeminal nerve. tSSEP was performed a month prior to surgery and in the month after the surgery in both patients. Pain frequency and tSSEP were analyzed before and after surgery.

Results: In both patients a complete resolution of pain occurred. In patient 1, tSSEP latencies became shorter than before surgery and wave N1 appeared. The intensity of stimulation necessary to reach the threshold was 4 mA before the surgery and 1 mA after the surgery. A complete recovery of tSSEP after the operation was achieved in patient 2.

Conclusions: The results of present study demonstrate the value of tSSEP in the evaluation of trigeminal conduction recovery after microvascular decompression.

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Abstract – WCN 2013**No: 271****Topic: 36 – Other Topic****Reduce the risk in vulnerability in HIV**

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Background: Before the people can reduce the risk and vulnerability to HIV, individuals and communities must understand the urgency to the epidemic. They must be given basic facts about HIV/aids, taught set of protective skills and offered access to appropriate services and products.

Methods: A cross sectional community based survey has been conducted between January 2011 and April 2011 by interview of 419 heads of the households regarding the knowledge of HIV transmission means in Accra, Ghana.

Results: Out of the interviewed 419 household heads, 287 (68.5%) were females. 36%, 28% and 34.3% were in the age group of 40+, 31–40 and 15–30 respectively. The findings revealed that only 21 (5%) of study participants mentioned four ways of HIV transmission (unprotected sex, mother to child, sharp materials and blood transfusion). On the other hand, 63 (15%) mentioned any three of the above route of transmission, whereas the majority, 209 (50%) and 86 (20.5%) mentioned two and one means of HIV respectively. On contrary about 40 (10%) of interviewed heads of household mentioned hardly any of the transmission means. Males were about 2.4 times more likely to mention unprotected sex as one means of transmission than females.

Conclusion: In general, knowledge of residents of Medina in the capital town of Ghana about HIV transmission and prevention means was low. Appropriate HIV/Aids education means ought to be tailored to residents.

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Abstract – WCN 2013**No: 280****Topic: 36 – Other Topic****Ineffectiveness of topiramate and levetiracetam in infantile spasms non-responsive to steroids**

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Objectives: To compare the effectiveness of 2 novel antiepileptic drugs Topiramate and Levetiracetam as a second line treatment for infantile spasm when oral steroids fail.

Methods: Forty infants within 2 years of life with clinically- and EEG-proven infantile spasms that did not respond to prednisone (2 mg/kg/day in 2 divided doses) were recruited and randomized into 2 groups. They were randomly assigned to either Topiramate (group 1; 1 mg/kg/day for 3 days then increased by 1 mg/kg/day every 3rd day up to 6 mg/kg/day) or Levetiracetam, (group 2; 10 mg/kg/day for 5 days and then increased by 10 mg/kg/day every 5 days up to 60 mg/kg/day).

Results: Of the 20 patients included in the final data analysis, 11 (55%) were administered with Topiramate and 9 (45%), Levetiracetam. Eighteen patients did not respond to the first drug and subsequently to the other drug when crossed-over, if the first drug proved ineffective. Two patients with infantile spasm responded to either one drug respectively without crossover. Their EEGs improved with time. In accordance with Helsinki Declaration and Belmont Report Ethical codes, we stopped recruitment at 20 patients due to the ineffectiveness of both drugs. Thus, only 20 children were included in final data analysis.

Conclusion: The present study demonstrated the ineffectiveness of Topiramate and Levetiracetam suggesting that current treatment modalities are grossly inadequate underscoring the urgent need for more research efforts to overcome current deficiencies. Two patients with cryptogenic infantile spasm responded to treatment suggesting

the potential for treatment of such patients with these two drugs which merits further multicenter investigation.

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Abstract – WCN 2013

No: 281

Topic: 36 – Other Topic

Limbic encephalitis in a child with an atypical presentation

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Background: Limbic encephalitis (LE) is a rare disorder that generally has a subacute onset evolving over days to weeks. Patients present with a variable combination of memory loss, seizures, and psychiatric disturbance, and it is not rare for patients to be initially misdiagnosed.

Patient: We report a previously healthy twelve-year-old boy who developed his first episode of seizures at eight years of age. He had a total of eight prolonged focal seizure episodes, and each episode was followed by a month of behavioral changes and short term memory loss. There was no family history of seizures or other neurological disorders, and he had an otherwise unremarkable neonatal and past medical history.

Results: Magnetic resonance imaging during each episode of seizures showed alternating unilateral brain hemispheric involvement consistent with LE that was followed by resolution for a total of six times. Despite a negative laboratory evaluation for a large panel of paraneoplastic antibodies, the clinical scenario and exclusion of other possible disorders made recurrent LE the most likely diagnosis.

Conclusion: Limbic encephalitis is a rare disorder diagnosed primarily on clinical criteria and often associated with the presence of a paraneoplastic antibody. However, the lack of a positive paraneoplastic antibody in a patient with a triad of seizure, behavioral changes and short memory loss does not exclude the diagnosis. The unique presentation in a seronegative patient like ours may indicate a previously unrecognized antibody. To the best of our knowledge, such a case has never been reported.

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Abstract – WCN 2013

No: 46

Topic: 36 – Other Topic

Homeostatic plasticity and human visual system

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Background and objective: The ability of cortical networks to regulate neuronal activity within a useful dynamic range, called homeostatic plasticity, has received growing attention. The threshold and direction of excitability changes primed by low- and high-frequency rTMS in the primary motor cortex (M1) can be reverted by a preceding session of tDCS. Whether there is a similar mechanism in human visual pathways is a matter of debate.

Materials and methods: In 15 healthy subjects we evaluated changes in visual evoked potential (VEP) amplitude at two different contrasts

(K90% and K20%) by applying anodal or cathodal tDCS (20', 1.5 mA) to occipital cortex, followed by low- or high-frequency rTMS (0.5 for 20' and 5 Hz for 60', respectively).

Results: Anodal tDCS improved the amplitude of VEPs and this effect was reverted by applying high-frequency rTMS (two-way ANOVA, $p < 0.001$). Concurrently, cathodal tDCS led to a decrease in VEP amplitude, which is reverted in its turn by a subsequent application of 0.5 Hz rTMS (two-way ANOVA $p < 0.01$). There are no significant changes in RMT values over time ($p > 0.5$), confirming the spatial selectivity of our protocol.

Conclusion: Our data show that preconditioning excitability with tDCS over the V1 can modulate the direction of plasticity induced by subsequent application of 1 or 5 Hz rTMS. This pattern was observed for both N1 and P1 components; since they appear to be generated in striate and extrastriate cortex, respectively, these findings indicate that homeostatic mechanisms could operate in both primary and higher-order visual areas.

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Abstract - WCN 2013

No: 53

Topic: 36 - Other Topic

Tongue's motor evoked potentials in the differential diagnosis between amyotrophic and primary lateral sclerosis: A preliminary report

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Introduction: Primary lateral sclerosis (PLS) is a neurodegenerative disorder due to a selective loss of precentral pyramidal neurons. Our purpose was to evaluate preferential impairment of pyramidal tract to bulbar muscles in patients with PLS and identify a reliable electrophysiological method to help clinicians in the differential diagnosis from amyotrophic lateral sclerosis (ALS).

Materials and methods: We recorded motor evoked potentials (MEPs) from tongues and anterior tibialis muscles in six patients with PLS and compared the results, in terms of central motor conduction time (CMCT), amplitude and duration of contralateral silent period (cSP), with those obtained both from age-matched healthy volunteers and patients affected by ALS.

Results: For lower limbs, CMCT resulted significantly increased in PLS and ALS compared with healthy subjects ($p < 0.01$); we did not disclose differences between ALS and PLS groups ($p = 0.417$). For tongue's recordings, CMCT, absolute amplitude of MEPs and cSP were significantly altered in PLS patients towards both ALS patients ($p < 0.05$) and healthy volunteers ($p < 0.01$).

Discussion: We showed that tongue's MEPs are selectively impaired in PLS compared with other motorneuron diseases. Our results strengthen the hypothesis of a proximal axonal degeneration as a pathological marker of PLS, probably reflecting a greater loss of corticomotoneuronal connections rather than in ALS patients. That could be confirmed by the shortening of cSP in cranial nerves, since for bulbar muscles the spinal contribution for the generation of cSP is ruled out.

Tongue's MEPs could represent an interesting electrodiagnostic test, potentially useful for the early diagnosis of PLS.

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Abstract - WCN 2013**No: 63****Topic: 36 - Other Topic****Head ultrasound findings in healthy preterm infants: Their correlation with gestational age and mode of delivery**Y. Yueniwati, D.R. Erawati. *Radiology, University of Brawijaya, Malang, Indonesia*

Background: Brain injury is a major complication of preterm birth. Some perinatal risk factors and demography could increase the morbidity rate in preterm infant. Head ultrasound becomes important diagnostic tool in neonates. Healthy preterm infants could have brain abnormality visualizing on ultrasound examination.

Objective: To analyze the correlation between head ultrasound findings in healthy preterm infants with gestational age and mode of delivery.

Patients and methods: Observational-analytic study using cross sections took place in Saiful Anwar Hospital Malang – Indonesia between July and September 2012. Healthy preterm infants underwent head ultrasound examination within the first four days of life. Fischer Exact test was used to analyze the correlation between head ultrasound findings with gestational age and mode of delivery.

Results: About 89.5% of 38 samples was ≥ 32 weeks gestational age; and 52.6% of samples had sectio caesaria as mode of delivery. There were three abnormal findings in head ultrasound: increasing periventricular echogenicity (5.3%), increasing deep parenchym echogenicity (5.3%) and obscuration of white-grey matter differentiation (5.3%). There is no significant correlation between ultrasound findings with gestational age and mode of delivery.

Conclusion: There were abnormal head ultrasound findings in some healthy preterm infants although there is no significant correlation between ultrasound findings with gestational age and mode of delivery. Head ultrasound in preterm infant could become screening tool for early detection of brain injury.

doi: [10.1016/j.jns.2013.07.2042](https://doi.org/10.1016/j.jns.2013.07.2042)**Abstract - WCN 2013****No: 69****Topic: 36 - Other Topic****The effect of epilepsy comorbidity in children with attention deficit hyperactivity disorder**G.-Y. Sim^a, W.S. Kim^b. ^a*Pediatrics, St. Mary's Hospital, Chengju, Republic of Korea*; ^b*Chungbuk National University, Cheongju, Republic of Korea*

Purpose: The rate of attention deficit hyperactivity disorder (ADHD) is higher in children with epilepsy than in the general population. The 31–40% of ADHD is accompanied with epilepsy. There are not many studies in ADHD with epilepsy. So we checked the effect of epilepsy comorbidity in children with ADHD.

Methods: This study retrospectively examined the effect of epilepsy comorbidity in children with ADHD. We studied 34 ADHD children with epilepsy at Chungbuk National University Hospital and 38 ADHD children without epilepsy at Cheonju St. Mary's Hospital because of ADHD with epilepsy or ADHD from January 2005 to June 2010.

Result: In ADHD with epilepsy, there were 12 cases (35.2%) of partial seizure, 11 cases (32.2%) of generalized seizure, 11 other cases (32.2%). Among the abnormal sites found in EEG there were 15 cases in the frontal lobe, 7 cases in the central lobe, 6 cases in the temporal lobe, and 3 cases in the occipital lobe. In ADHD with epilepsy, the combined type is 76.4% and in ADHD without epilepsy, the inattentive type is 50.5% ($P = 0.004$). There is more learning disability in ADHD with epilepsy than in ADHD without epilepsy ($P = 0.01$).

Conclusion: In ADHD with epilepsy, the combined type showed 76.4%. There is more learning disability in ADHD with epilepsy than in ADHD without epilepsy. This study showed the effect of epilepsy comorbidity in children with ADHD.

doi: [10.1016/j.jns.2013.07.2043](https://doi.org/10.1016/j.jns.2013.07.2043)**Abstract - WCN 2013****No: 14****Topic: 36 - Other Topic****Primary central nervous system lymphoma or vanishing tumor in a patient presenting a Parinaud's syndrome**G. Piccirillo^a, L. Lavorgna^a, A. Tessitore^a, S. Bonavita^a, M. Cirillo^b, F. Tortora^b, M.R. Monsurrò^a, G. Tedeschi^a. ^a*Department of Neurological Sciences, II Division of Neurology, Second University of Naples, Naples, Italy*; ^b*Department of Neuroradiology, Second University of Naples, Naples, Italy*

A 24-year-old man presented a one month palsy in the upward gaze. MRI, immediately after the onset, showed a single lesion located in the posterior midbrain with a maximum diameter of 2 cm, and perilesional edema, involving the posterior commissure: hyperintense in T2, hypointense in T1 and homogeneously enhanced after gadolinium. Once admitted the patient showed signs of Parinaud's syndrome. CSF showed 1 lymphocyte/ μL and was negative for infection and demyelinating disease. The patient underwent a pulse of methylprednisolone of 1 g/die for five days with no symptom remission. One month later MRI showed: decreased volume of the lesion with unchanged signal characteristics including homogeneous contrast enhancement. HIV serology, serological tests for autoimmune autoantibodies, PET and CT total resulted negative. A diagnosis of primary central nervous system lymphoma was considered and to attempt an histological diagnosis we halted the steroid treatment for two months. At this point, once again the MRI showed a lesion whose size and signal were comparable to the first MRI. Forty days later, as the patient was scheduled for a stereotactic biopsy, we performed another MRI and we found that the lesion had almost completely disappeared: biopsy was no longer feasible. As the lesion had almost regressed, and no other possible explanation could be envisioned by the clinical and instrumental characteristics, we suggested a diagnosis of vanishing tumor: a case in which there is a strong suspicion of brain tumor from the clinical history and in which a gadolinium enhancing lesion spontaneously disappear or decrease, within 3 months, to less than 70% of the initial volume. Most vanishing tumors in the central nervous system might eventually be diagnosed as PCNSL.

doi: [10.1016/j.jns.2013.07.2044](https://doi.org/10.1016/j.jns.2013.07.2044)**Abstract - WCN 2013****No: 10****Topic: 36 - Other Topic****Air- and bone-conducted cervical and ocular vestibular-evoked myogenic potentials in patients with superior vestibular neuritis**S.-Y. Oh^a, J.S. Kim^b, B.-S. Shin^a, T.-H. Yang^a, S.-K. Jeong^a. ^a*Department of Neurology, Chonbuk National University, Jeonju, Republic of Korea*; ^b*Neurology, Seoul National University Bundang Hospital, Seongnam, Republic of Korea*

Objective: To determine the characteristic changes in cervical (cVEMP) and ocular (oVEMP) vestibular evoked myogenic potentials in superior vestibular neuritis (SVN), we recorded cVEMPs and oVEMPs in patients with SVN ($n = 33$) and normal controls ($n = 45$) to air-conducted sound (ACS) and bone-conducted vibration (BCV).

Results: ACS induced cVEMPs revealed mostly normal (81.9%) whereas results of ACS induced oVEMPs showed high rates of abnormalities (82%). oVEMP by BCV at the mid-frontal (Fz) revealed high rates of abnormalities (62.5%) especially on the contralateral eye and none of them showed abnormality on the ipsilateral eye. cVEMPs and oVEMPs induced by BCV at the mastoid process revealed dissociated results. cVEMP to mastoid BCV showed low rates of abnormalities (31.2%) whereas oVEMP to mastoid BCV revealed high rates of abnormalities (75%).

Conclusions: The oVEMP in response to ACS may be mediated by the superior vestibular nerve, probably due to an activation of the utricular receptors. The high rates of abnormalities in Fz BCV oVEMP also suggest a dependence on the intactness of the superior vestibular nerve afferents. The dissociations in the abnormalities of cVEMP and oVEMP induced by BCV at the mastoid in patients with VN selectively involving the superior vestibular nerve suggest that the origin of the vestibular nerve afferents of oVEMP to mastoid BCV differ from those of cVEMP to mastoid BCV. Therefore oVEMP to mastoid BCV also reflects functions of the superior vestibular nerve afferents and most likely the utricular function.

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Abstract - WCN 2013

No: 6

Topic: 36 - Other Topic

Sleep apnea in patients with diabetic neuropathy

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Background: Diabetes mellitus (DM) is a major chronic disease with high morbidity, mortality and economic burden. Sleep apnea is an increasingly recognized medical problem with its important role in metabolic, vascular and behavioral aspects. The association between sleep apnea and DM is likely to be complex.

Objectives: To detect prevalence, types and severity of sleep apnea in diabetic patients, and to detect any relation between sleep apnea and diabetic state, diabetic peripheral and autonomic neuropathy and phrenic nerve affection.

Patients and methods: Forty four diabetic patients were included in the study. Another 44 age and sex matched healthy subjects representing the control group were included. All the subjects were evaluated for diabetic state, peripheral neuropathy, autonomic neuropathy and phrenic nerve affection. Sleep disorders were assessed by overnight polysomnography recording.

Results: Sleep apnea was significantly more frequent in diabetic patients (84.1%) than in non-diabetics (18.2%). Obstructive sleep apnea was the most frequent type (65.9%) followed by mixed (13.7%) and lastly central type (4.5%) in the diabetics. All the grades of sleep apneas (mild, moderate and severe) were significantly more frequent in diabetics (45.4%, 18.2%, 20.5% respectively). There was significant relation between sleep apnea and age of the patient, male sex, uncontrolled DM, duration of DM, diabetic autonomic neuropathy and phrenic nerve affection. There was no relation detected between sleep apnea and diabetic peripheral neuropathy.

Conclusion: Sleep apneas (especially obstructive type) are more frequent in diabetic patients with autonomic neuropathy and phrenic nerve affection.

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Abstract - WCN 2013

No: 81

Topic: 36 - Other Topic

LAT software based comparative clinical trial of alcohol addicts in Serbia and Austria

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Background: Serbia is lacking marketing approval of few essential anticraving medications to treat alcohol addiction. Comparative research was undertaken to provide deeper insight and grounds for reimbursement decisions.

Objective: Comparison of two European samples of alcoholic addicts processed by LAT software to detect major differences related to clinical evolution and treatment.

Patients and methods: 127 (116 males) Serbian and 136 (78 males) Austrian alcohol addicts were clinically followed from January 2011 to March 2012. The patients were assessed using the Lesch alcoholism typology instrument (LAT).

Results: Age of onset was slightly higher in the Austrian sample (28.5 vs 30.0; $p < 0.005$). The Serbian sample has significantly higher rate of anxiety disorders than in the Austrian sample (89.8% vs 26.5%, $p < 0.001$). Suicidal tendencies, independent of alcohol intake or withdrawal syndrome, are significantly higher in the Austrian sample (1.6% vs 13.2% $p < 0.001$). There was no difference between the two samples in Lesch Type IV (26 vs 28), but there was only a slight increase in the Serbian sample in Type I (15 vs 10). In Austria, significantly more Type II patients (32 vs 52) had been included, while the Serbian sample comprised significantly more Type III patients.

Conclusion: Austrian and Serbian patients are quite similar, and that there is no inhibiting factor that would further delay an introduction and reimbursement of the modern anticraving medications in Serbia. The differences in anxiety disorders might be due to the 1990s war, and should be further investigated.

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Abstract - WCN 2013

No: 89

Topic: 36 - Other Topic

Slug, twist, and E-cadherin as immunohistochemical biomarkers in meningeal tumors

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Introduction: The overexpression of twist and slug and subsequent down-regulation of E-cadherin facilitate the acquisition of invasive growth properties in cancer cells. It is unclear which of these molecules are expressed in mesenchymal tumors in the central nervous system.

Methods: In the present study, we investigated 10 cases each of hemangiopericytoma, solitary fibrous tumor, meningothelial, fibrous, angiomatous, and atypical meningiomas, and 5 cases of anaplastic meningioma for slug, twist, e-cadherin, and n-cadherin immunoreexpression.

Results: Nuclear slug expression was observed in 9/10 (90%) hemangiopericytomas and 5/10 (50%) solitary fibrous tumors, but not in any meningiomas, except for 1 case. Similarly, nuclear twist expression was

more extensive in hemangiopericytomas and solitary fibrous tumors than meningiomas. In contrast to slug and twist, the positive expression of E-cadherin was observed in 39/45 (87%) meningiomas, but not in any hemangiopericytomas or solitary fibrous tumors ($P < 0.0001$). The fraction of tumor cells expressing E-cadherin in meningeal tumors was negatively correlated to those of twist ($P = 0.004$) and slug ($P < 0.0001$).

Conclusion: The overexpression of slug and twist with down-regulation of E-cadherin was characteristic findings in hemangiopericytomas and solitary fibrous tumors, but not in meningiomas. The immunohistochemical profiles of the two tumor groups may be useful as diagnostic markers in cases that present a differential diagnosis challenge, and be associated with aggressive behavior in meningeal tumors.

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Abstract - WCN 2013

No: 94

Topic: 36 - Other Topic

Cannabis sativa induces sedation in mice

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Cannabis sativa (Marijuana) is a tobacco-like substance. It is a dry shredded green and brown mix of flowers, stems, seeds and leaves of *Cannabis sativa* plant. It is used for medicinal purposes; as a recreational drug and for obtaining fiber. In this study, the effect of acute administration of *Cannabis sativa* on sedation was evaluated using elevated plus maze and staircase test in mice. Intraperitoneal administration of the cannabis at doses of 250–1000 mg/kg significantly ($P < 0.05$) and dose-dependently decreased the mean open and close arms entry; with no significance on the open and close latencies except the close arm latency of the 500 mg/kg animal group when compared to the control group. On the other hand, there was a significant ($P < 0.05$) and dose-dependent decrease in the number of stairs climbed and number of rearing on the staircase test as compared to the control. These results therefore suggested that acute *Cannabis sativa* administration induces sedation in mice.

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Abstract - WCN 2013

No: 109

Topic: 36 - Other Topic

The case of prenatal diagnostics of Dandy–Walker syndrome

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Congenital malformations of the central nervous system (CNS) is one of the first places in mortality and one of the causes of serious neurological pathology and early childhood disability.

Dandy–Walker syndrome (DWS) is a rare and severe congenital anomaly of the CNS (the frequency is 1 on 25,000–35,000) with difficulties of prenatal diagnostics. This usually becomes possible after 18–20 weeks of pregnancy. Further, a case is presented with the prenatal diagnostics of DWS by an ultrasound examination and in utero MR imaging. Life history and medical history of pregnant women – without peculiarities. Ultrasound examination at 18 weeks of gestation – without visualization of pathology. Ultrasound examination at 24 weeks of pregnancy – unclear visualization of worm of cerebellum, hypoplasia

of the cerebellar hemispheres as a “banana”, difficult visualization of the corpus callosum. To clarify the diagnosis and differential diagnosis in utero MR imaging was performed.

Conclusion: Dandy–Walker syndrome. In utero MR imaging is a modern non-invasive and exact method for CNS abnormality imaging of the fetus. MR imaging may improve significantly the quality of prenatal diagnostics with optimization of the obstetric tactics and subsequent treatment of the newborn. Improvement of methods of diagnostics, screening for pregnant women with determination of congenital pathology of the fetus, and the introduction of modern perinatal technologies are the main directions for reduction of the perinatal mortality and early childhood disability.

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Abstract - WCN 2013

No: 101

Topic: 36 - Other Topic

We need a paradigm shift in research on reading and dyslexia: Fundamental problems with fMRI studies of written language processing

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Background: Over the past two decades, functional MRI has become a widely accepted tool to study reading. fMRI-based research claims that:

- 1) specific brain sites exist where visual letters are converted to oral language phonemes;
- 2) these sites are impaired in dyslexic readers; and
- 3) intensive remediation on letter–sound correspondences (phonics) can literally “repair” these sites.

Psychological models of reading, however, call these claims into question.

Objective: To demonstrate that

- 1) fMRI is an inappropriate tool to study reading and dyslexia; and
- 2) the neuroscientific study of reading and dyslexia needs to incorporate the psycholinguistic nature of meaning construction and its neuroanatomic foundation in cortical–subcortical circuitry.

Cortical–subcortical circuitry must be reconceptualized as a feed-forward, not feed-back system.

Materials and methods: Literature reviews are conducted of the formal linguistics of letter–sound mapping, saccades and fixations during reading, cortical–subcortical circuitry, and the psycholinguistics of reading.

Results: Letter–sound mappings differ significantly from what is assumed in fMRI-based research. Eye movement studies reveal that proficient readers do not fixate on fully one third of the words in a visual text display. Cortical–subcortical tracts constitute the feed-forward component of executive functioning. Meaning-construction relies on psycholinguistic strategies far more efficient and effective than letter–sound conversion. The spatial and temporal resolution capacity of fMRI is adequate for identifying letter–sound brain regions, but not for identifying meaning-construction neuroanatomy. **Conclusion:** Reading must be described within a meaning-construction psychological paradigm. It is an executive process beyond the technical resolution capacity of fMRI. Its neuroanatomic basis lies in feed-forward cortical–subcortical tracts.

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Abstract - WCN 2013**No: 136****Topic: 36 - Other Topic****Cardiovascular autonomic function in healthy offsprings with parental history of type 2 diabetes mellitus**

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Background: It has been shown that patients with type 2 diabetes mellitus (T2DM) exhibit altered autonomic activity of cardiovascular system.

Objective: The current study aimed to compare cardiovascular autonomic function between the healthy offsprings with and without parental history of T2DM.

Material and methods: Heart rate variability (HRV) and cardiovascular autonomic reactivity of 80 healthy male subjects, between the age group of 18 and 30 years were assessed. The study group consisted of 40 subjects with parental history of T2DM and control group included 40 subjects without parental history of diabetes. HRV was assessed by fast Fourier transformation of supine 5 min ECG epoch recorded at spontaneous respiratory rates. Cardiovascular autonomic reactivity was assessed by deep breathing, Valsalva maneuver, handgrip and head up tilt.

Results: In HRV, all the time domain measures which are the markers of parasympathetic activity were comparable. In frequency domain measures, low frequency (LF) which is the marker of sympathetic activity and high frequency (HF) which is the marker of parasympathetic activity as well as LF/HF which is the sympathovagal balance were comparable. Among autonomic reactivity variables, deep breathing and Valsalva ratio which are the markers of parasympathetic reactivity were comparable. Similarly, rise in diastolic blood pressure during handgrip and change in systolic blood pressure in head up tilt which are the markers of sympathetic reactivity were also comparable.

Conclusion: This study concludes that the parental history of diabetes does not have an impact on cardiovascular autonomic function in offsprings between the age group of 18 and 30 years.

doi:10.1016/j.jns.2013.07.2052

Abstract - WCN 2013**No: 113****Topic: 36 - Other Topic****Cepheid GeneXpert® MTB/RIF in CSF diagnosis of tuberculous meningitis among HIV-infected patients in Africa: 3 case reports from Kenya**

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Background: Tuberculous meningitis (TBM) is one of the most aggressive forms of meningitis with high mortality outcomes if not diagnosed and treated early, especially in immunosuppressed individuals. Few case reports and studies have been documented on the use of Cepheid GeneXpert® MTB/RIF in CSF. The aim was to highlight the use of Cepheid GeneXpert® testing for the presence of *Mycobacterium tuberculosis*.

Methods: The clinical features of 3 new HIV patients diagnosed with TBM using Cepheid GeneXpert® MTB/RIF are described and collated with other reported cases in a systematic literature review.

Results: Case I involved a 39 year old HIV-infected female with headache, lower back pain, productive cough, and dizziness, had positive CSF evidence of *Cryptococcus neoformans*, high CSF protein (118.3 mg/dL), and positive evidence of *Mycobacterium tuberculosis* on GeneXpert® analysis. Case II involved a 28 year old HIV-infected female

with headache and altered mental status, negative CSF evidence of *Cryptococcus neoformans*, high CSF protein (386.2 mg/dL), and positive evidence of *Mycobacterium tuberculosis* on GeneXpert® analysis. Case III involved a 28 year old HIV-infected female who presented with headache and reduction in loss of consciousness, had negative CSF evidence of *Cryptococcus neoformans*, high CSF protein levels (315 mg/dL), and positive evidence of *Mycobacterium tuberculosis* on GeneXpert® analysis.

Conclusion: Despite the paucity of data on its use, Cepheid GeneXpert® PCR technology can be used relatively cheaply within resource-limited settings in the timely diagnosis of TBM so as to improve the prognosis of HIV-infected patients who present with clinical suspicion of TBM.

doi:10.1016/j.jns.2013.07.2053

Abstract - WCN 2013**No: 115****Topic: 36 - Other Topic****A case of autoimmune encephalitis caused by antibodies to GABA-B receptor**

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Objective: To describe a case of autoimmune encephalitis caused by antibody to GABA_B receptor.

Background: Autoimmune encephalitis resulting from antibodies to GABA_B receptor is uncommon. These patients usually present with early or prominent seizures. About one third of these cases can have an underlying tumor, usually small cell lung cancer. Immunotherapy and treatment directed at the underlying tumor may cause resolution of the condition.

Method: Case report.

Results: We report a case of a 29-year old male presenting with 2 weeks of history of cognitive and behavior dysfunction and new onset generalized tonic-clonic seizures. There were no focal neurological deficits on examination. EEG showed diffusely slow background. MRI of the brain and was essentially normal except for a subtle questionable leptomeningeal enhancement in bilateral occipitoparietal sulci but with no corresponding FLAIR abnormality. CSF analysis demonstrated a lymphocytic pleocytosis and mildly elevated protein. Viral studies were negative. High titers of antibody against the GABA_B receptor was detected in both the serum and the CSF confirming the diagnosis of GABA_B receptor autoimmune encephalitis. CT chest, abdomen and pelvis and a testicular ultrasound were all normal. He was treated with steroids, intravenous immunoglobulin and rituximab but remains encephalopathic three months after the treatment.

Conclusions: GABA-B receptor autoimmune encephalitis is uncommon. Recognition of this condition as well as the other forms of autoimmune encephalitis is important as immunotherapy and treatment of the underlying tumor can be effective.

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Abstract - WCN 2013**No: 121****Topic: 36 - Other Topic****Thalamic lesions profile by MRI as predictors for outcome in cases of acute disseminated encephalomyelitis**

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Background: Acute disseminated encephalomyelitis (ADEM) is a febrile episode associated with disturbed consciousness, seizures and

neurological dysfunctions. The aim is to correlate between thalamic MRI findings and outcome and to determine if brain imaging can predict sequelae or not.

Patients and methods: Thirty two cases of ADEM were recruited in the acute condition. Patients were hospitalized and given pulse methylprednisolone and IVIG. Cranial or cranio-spinal neurological dysfunction was assessed clinically. Brain MRI, digital EEG and lumbar puncture were done for all cases.

Results: Mean age at presentation was 4.6 ± 2.4 years. Twenty were males and 12 females. Patients presented with motor weakness (100%), fever (80%), disturbed consciousness and seizure (73%). MRI brain showed multifocal punctate to large flocculent high T2 and FLAIR signal intensity involving subcortical white/grey matter junction (86%), periventricular white matter (80%), brain stem (40%), cerebellum (31%) and bilateral thalami (25%). The calloseseptal interface was not involved in any of them. Twenty three patients recovered without neurological deficits while 9 showed sequelae. All patients having bilateral thalamic involvement had sequelae. The thalamic lesions were in 2 cases in the form of hemorrhagic foci and necrosis. These two cases had severe neurological deficits in the form of spasticity with speech regression.

Conclusion: Thalamic lesions in ADEM are associated with neurological sequelae in the form of spasticity, squint and hemiplegia. MRI findings that may predict severe neurological deficits are hemorrhagic foci, necrosis or edema in thalamus.

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Abstract - WCN 2013

No: 122

Topic: 36 - Other Topic

Posterior cranial fossa tumors in children at a referral hospital in Sub-Saharan Africa

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Background: Posterior fossa tumours in children results in morbidity & mortality worldwide. In Sub-Saharan Africa delayed diagnostic modalities and lack of awareness of the symptoms and signs is common.

Objective: To review the management of posterior fossa tumours in children.

Design: A retrospective analysis of children treated for posterior fossa tumours between 2009 and 2012.

Setting: Neurosurgery unit, Kenyatta National Teaching and Referral Hospital. This is the only specialized unit among the public hospitals in Kenya where such patients are referred.

Results: 54 children were treated for posterior fossa tumours between 2009 and 2012. 30 males and 24 females were attended to giving a M:F ratio of 1.25:1. The age varied between 1 and 16 years with a mean of 7.4 years. Cerebellar symptoms were the most common mode of presentation (32%) followed by headaches and vomiting. 30% of our patients were blind at presentation probably due to chronic effects of raised intracranial pressure. Out of 18 patients with histological diagnosis of medulloblastomas, over 90% were females and only 2 were males. Astrocytomas were evenly distributed at 7 males and 9 females. The mean duration of symptoms was 5.4 months while it took 6 weeks between time of diagnosis and treatment.

Conclusion: Posterior fossa tumours in our set-up are more common in males than in females, M:F ratio of 1.25:1. Over 90% of medulloblastomas are found in female children. The delay in diagnosis and definitive management is noted probably due to lack of awareness and expensive management modalities.

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Abstract - WCN 2013

No: 141

Topic: 36 - Other Topic

Clinical outcome after microsurgical resection of spinal canal meningiomas: Multidisciplinary team experience

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Background: Meningiomas are primary and the second most common type of benign tumors of the central nervous system derived from meningeothelial cells.

Objective: To evaluate the clinical outcome of posterior approach microsurgical resection of spinal meningiomas.

Patients and methods: 32 patients underwent microsurgical resection via the posterior approach; visual analogue scale (VAS) and medical research council (MRC) were used for clinical evaluation. All patients underwent clinical evaluation, MRI pre- and post-operation. Follow up duration was about 1 year after surgery.

Results: The patients enrolled comprise 26 females and 6 males, with a median age of 48 years old (range 22–74). 26 cases experienced clinical improvement following operations. The extent of tumor resection was Simpson Grade I in 5 patients, Grade II in 20, and Grade III in 7. No recurrence could be detected after one year of follow up. There were two cases of postoperative CSF leakage and two cases of worsened neurological status due to operative morbidity. No immediate postoperative mortality.

Conclusion: Gross total resection is associated with no recurrence and favorable clinical outcome when applied, regardless of dural origin control.

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Abstract - WCN 2013

No: 484

Topic: 36 - Other Topic

Clinical characteristics in association with neurodegenerative diseases and cancer

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Background: Patients with neurodegenerative diseases sometimes suffer from malignancies.

Objective: The purpose of our study was to examine whether there are clinical features in Japanese patients with both neurodegenerative diseases and cancers.

Patients and methods: We retrospectively analyzed the clinical characteristics of consecutive Japanese patients with neurodegenerative diseases during the past five years, including amyotrophic lateral sclerosis (ALS), Parkinson's disease (PD), dementia with Lewy bodies, progressive supranuclear palsy (PSP), corticobasal degeneration (CBD), and multiple system atrophy (MSA).

Results: Out of 292 patients, the following 39 patients had cancers, including a past history: ALS, 16 (16.0%); PD, 8 (9.3%); PSP, 7 (19.4%); CBD, 1 (6.7%); and MSA, 7 (15.9%). About 10% of patients with ALS, PD, PSP or MSA developed cancer after onset of the disease; about 30% of patients with ALS, PD or PSP occurring with cancers died of cancer. Gastric cancer was the most common before onset of ALS (62.5%), but did not develop after onset of ALS. Conversely, PD patients frequently developed gastric cancers after the onset of neurological signs (60.0%) in spite of no cancer before onset of PD. The proportion of breast cancer in MSA (45.5%) was

significantly higher than in other neurodegenerative diseases (2.7%).

Conclusion: ALS, PD or MSA patients with cancer showed clinical characteristics unique to each neurodegenerative disease in Japan, compared to other countries. It is necessary to be aware of cancer in patients with ALS, PD or PSP because one-third of their deaths is caused by cancer.

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Abstract – WCN 2013

No: 467

Topic: 36 – Other topic

Subacute sclerosing panencephalitis: A familial occurrence of two cases in Kosovo

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Subacute sclerosing panencephalitis is a progressive neurodegenerative disease of children and adolescents. It arises as a late complication of primary measles infection. SSPE is an active brain infection with a defective measles virus. We report two cases of SSPE that developed in two unvaccinated first cousins that come from a rural part of Kosovo. Familial occurrence of SSPE is already reported, but the predisposing genetic mechanisms that may be inherited are yet to be proven. Atypical SSPE presentation is found in one of the cases, while the classic one in the other. SSPE is an important disease to be excluded in the differential diagnosis in children manifesting neurodegenerative diseases, especially in developing countries such as Kosovo.

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Abstract – WCN 2013

No: 457

Topic: 36 – Other topic

Opioid receptors influence blood–brain barrier permeability, cerebral blood flow and serotonin levels following forced swimming exercise in young rats

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Effect of forced swimming (FS) exercise on cerebral blood flow, blood brain barrier permeability and regional brain serotonin level was examined in young normotensive rats. Subjection of rats to FS for 30 min resulted in marked increase in the regional blood–brain barrier permeability in 14 brain regions. The regional cerebral blood flow (CBF) exhibited significant decrease in 6 brain regions whereas, no changes in 4 brain regions were observed. In another 4 regions there was a mild increase in the regional CBF. Measurement of regional brain serotonin content exhibited increase in few regions while in many other regions the serotonin level was decreased significantly.

Pretreatment with naloxone, an opiate receptor antagonist significantly attenuated the breakdown of the BBB permeability and reduced cerebral blood flow changes. The serotonin level also did not alter significantly. This effect was more pronounced in rats treated with high dose (10 mg/kg, i.p.) of the compound, compared to lower (1 mg or 5 mg/kg, i.p.) dosages. These observations suggest that opioids

significantly influence FS induced changes in BBB permeability, CBF and serotonin levels, not reported earlier.

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Abstract – WCN 2013

No: 429

Topic: 36 – Other topic

Prognosis of benign paroxysmal positioning vertigo: Long term outcome and its prognostic factors

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Background: Even though Benign Paroxysmal Positional vertigo (BPPV) is most common cause of episodic vertigo, its prognosis is not well defined. In this study we aimed to identify long-term prognosis of BPPV and prognostic factors associated with its recurrence: sex, age, affected canal type, and etiology of BPPV, previous history of BPPV, and time period for cure.

Methods: A total of 1123 consecutive patients were diagnosed with BPPV through 10 years from January, 2000. Using Kaplan–Meier survival analysis, we investigated the cumulative recurrence rate of BPPV and calculated hazard ratio of the variables associated with recurrences.

Result: After excluding intractable cases or follow-up loss before cure, 106,955 patients (mean age: 56.7 ± 13.4 years old, female: 72.97%) were analyzed in this study. The 30 day, 3 month, 6 month, 1 year, 2 year, 3 year, 5 year, 7 year and 10 year cumulative recurrence rates of total subject were 3.8%, 10.9%, 16.4%, 21.8%, 27.6%, 30.8%, 34.1%, 36.0%, 3.8%, 10.9%, 16.4%, 21.7%, 27.4%, 30.6%, 33.9%, 35.8%, and 41.8% respectively. Recurrence rates of female and time period for cure (TPC) more than 1 day were 46.7% and 54.10% respectively, which were higher than those of male (30.9%) and TPC ≤ 1 day (38.4%) with statistical significance in the survival analysis and those were independently raised hazard of recurrence.

Conclusion: We observed that substantial proportion of patients with BPPV recurred. Gender and time period for cure independently raised the risk of recurrence.

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Abstract – WCN 2013

No: 441

Topic: 36 – Other topic

Recurrent tetanus in diagnosed breast cancer patients of the Philippine General Hospital

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This is the first report of recurrent tetanus in two cases of breast cancer. The first case is of a 40-year old female with intraductal carcinoma of the right breast who was admitted for tetanus with breast mass ulcers as presumptive focus. She was given anti-tetanus vaccines and appropriate antibiotics. No wound debridement was done for fear of tumor seeding. She was discharged clinically cured of tetanus. Despite adequate treatment, the patient had recurrence of the tetanus 40 days after discharge. The second case is of a 56-year old female diagnosed with intraductal carcinoma of the left breast who developed tetanus after accidentally injuring the breast mass with a door nail. Similarly, she was given anti-tetanus vaccines and appropriate antibiotics. There was no consent to any intervention on the breast mass. She was discharged cured, but returned after 32 days with recurrent tetanus. The recurrence of tetanus in these patients could be due to incomplete elimination of the infection,

presence of chronic wound, and inability to mount sufficient immunity. Tetanus is a preventable and treatable condition with high mortality. Gathering of local data on recurrent tetanus and vaccine efficacy may be subject of future studies. It is recommended that guidelines on the management of tetanus specifically in immunocompromised patients with untreated chronic wounds as well as policies on routine vaccination of cancer patients be formed.

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Abstract – WCN 2013

No: 442

Topic: 36 – Other topic

Diagnostic challenge and steroid-dependence in a HIV patient with cryptococcal immune reconstitution inflammatory syndrome

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Background: Cryptococcal immune reconstitution inflammatory syndrome (IRIS) developing in human immunodeficiency virus (HIV) patients has been well described following immune recovery with highly active antiretroviral therapy (HAART). Prompt diagnosis of cryptococcal IRIS and treatment often lead to better outcomes. Diagnosing IRIS may be challenging. Optimal duration of treatment for cryptococcal IRIS is unknown.

Objective: To describe the challenges in diagnosis and treatment of cryptococcal IRIS in a HIV patient.

Patients and methods: Patient followed up in Departments of Neurology and Infectious Disease from 2007 to 2013.

Results: A 43-year-old male HIV patient with multiple opportunistic infections (pneumocystis carinii pneumonia, cytomegalovirus retinitis, cryptococcal meningitis and disseminated mycobacterium avium complex infection) was treated with HAART from Dec 2007. Initial CD4 count of 21 gradually improved to 82 in May 2009. He subsequently developed recurrent episodes of right hemiparesis, numbness, dysarthria, dizziness and seizures from Oct 2008 to Jun 2009. Serial brain MRI scans showed progressive T2-weighted white matter hyperintensities and leptomeningeal enhancement, predominantly in the left parietal region. Cerebrospinal fluid showed lymphocytic pleocytosis and negative for bacterial, viral and fungal etiologies. He was initially empirically treated with antituberculosis medications and prednisolone, but his seizures would recur whenever steroids were tapered. Brain biopsy in Jul 2009 eventually showed cryptococcal meningitis and cerebritis. He remained well with anticonvulsants and oral steroids, but could not be weaned off steroids (currently 5 mg daily) as his seizures would recur.

Conclusion: Cryptococcal IRIS diagnosis can be challenging. Duration of steroid treatment for cryptococcal IRIS may be prolonged.

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Abstract – WCN 2013

No: 451

Topic: 36 – Other topic

Management preferences and prognosis for primary intracranial melanocytic neoplasms

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Background: Contrary to relatively frequent metastatic melanoma, primary melanocytic CNS neoplasms are rare, coming up from

melanocytes situated in the leptomeninges. These lesions incorporate diffuse meningeal melanomatosis, melanin-pigmented meningioma, well-differentiated benign melanocytoma, and primary malignant intracranial and/or spinal melanoma.

Objective: The authors report a case of a 59-year man with a primary intracranial malignant melanoma which was successfully operated on. The management preferences and prognosis of such a rare disease are discussed, based on the clinical characteristics, neuroradiological, and pathohistological diagnostics, and a literature review.

Material and methods: Brain computed tomography and magnetic resonance imaging are performed to disclose a solitary contrast enhanced intraparenchymal parasagittal mass lesion of the left occipital lobe, mimicking a falx meningioma. The tumor manifested with the right side homonymous hemianopsia due to compression of the adjacent brain. The patient underwent the left side occipital osteoplastic craniotomy and the tumor was completely microsurgically removed. Pathohistological and immunohistochemical analysis of the tumor tissue samples was carried out to make a distinction between melanocytoma, as well as other pigmented brain tumors, and malignant melanoma. Postoperatively, the additional whole brain irradiation was applied.

Results: At a one year follow-up, the patient clinical and neurological recovery was comprehensive, while neuroradiology found no tumor remained or re-emergence of the disease.

Conclusion: The biological behavior of primary melanocytic neoplasms is uncertain and these lesions may recur as malignant melanoma. Hence, a thorough radiological diagnostics, meticulous pathohistological examinations, and complete tumor surgical resection followed by irradiation therapy may be prognostically beneficial.

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Abstract – WCN 2013

No: 444

Topic: 36 – Other topic

Prevalence of Alzheimer's and Parkinson attending physiotherapy services

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Introduction: Alzheimer and Parkinson diseases have been increasing in its presentation by the phenomena of aging, and to determine their prevalence is important for its implications for movement and functional performance.

Objective: To establish the prevalence of cases of Alzheimer's and Parkinson attended in Physiotherapy services.

Materials and methods: Cross-sectional study in which statistical records was taken care of patients over 45 years old seen during 2012 in the health care practices of Physical Therapy Program at the University Manuela Beltran.

Results: During 2012 they responded to a total of 3063 people over 45 years old. Alzheimer's disease reached a prevalence of 6.82% (n = 209, standard error = 0.01), whereas 2.12% of treated patients had a diagnosis of Parkinson's disease. The 77.51% (n = 162, standard error = 0.01) of those who were treated for Alzheimer were female in gender ($\chi^2 = 30,146$ p < 0.05, OR = 2.47, 95% CI 1.80–3.35), while in the case Parkinsons 52.30% of those affected were male (n = 34, standard error = 0.05) ($\chi^2 = 3.85$ p < 0.05, OR = 1.62, 95% CI 1.05–2.48). In the observation period were treated 344 people with chronic degenerative neurological conditions among which Alzheimer's disease represent the 60.75% of cases ranking first (n = 209, standard error = 0.02).

Conclusions: Alzheimer's disease has a higher prevalence of occurrence relative to Parkinson and it occurs most frequently among persons of female gender.

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Abstract – WCN 2013

No: 445

Topic: 36 – Other topic

Prevalence of chronic degenerative diseases and traumatic nervous system in adolescents and adults

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Introduction: The demographic, social and lifestyle of the population factors have led to the increase in cases of chronic degenerative and traumatic neurological diseases that have significant impacts on the processes of life for people.

Objective: To establish the prevalence of chronic degenerative and traumatic diseases of the nervous system in adolescents and adults.

Materials and methods: Cross-sectional study in which statistical records were taken care of patients over 15 years old seen in 2012 in the health care practices of Physical Therapy Program at the University Manuela Beltran.

Results: During 2012 they attended a total of 4122 people over 15 years old. 25.76% (n = 1062, standard error = 0.00) of these people were intercepted with neurological problems. 17.98% (n = 191, standard error = 0.02) of people with neurological diseases were due to trauma, and 32.39% (n = 344, standard error = 0.02) had chronic degenerative diseases. 44.5% (n = 85, standard error = 0.03) of people with traumatic disease had brain injury, while among those with chronic degenerative diseases the 60.75% (n = 209, standard error = 0.02) reported disease cases, e.g. Alzheimer. 72.94% of people with traumatic brain injury were male (n = 62, standard error = 0.02), and 77.51% (n = 162, standard error = 0.01) of people with Alzheimer's disease were women.

Conclusions: Chronic degenerative diseases tend to be more prevalent diseases in the nervous system than traumatic diseases, the first disease chronic degenerative as Alzheimer, and in traumatic disease as brain injury.

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Abstract – WCN 2013

No: 125

Topic: 36 – Other topic

Diffusion-weighted cranial MR imaging in Wernicke's encephalopathy associated with enterocutaneous fistula

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Wernicke's encephalopathy is a rare neurological disease due to thiamin deficiency and characterized by ophthalmoparesis, disturbed consciousness and ataxia. It is known that periventricular white matter, medial thalamic nuclei, massa intermedia, and mammillary bodies are affected in Wernicke's encephalopathy. Chronic alcoholism, anorexia, peritoneal dialysis, hyperemesis gravidarum and total parenteral nutrition are the most common causes of Wernicke's encephalopathy. We presented three patients who had Wernicke's encephalopathy due to total parenteral nutrition and thiamin deficiency.

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Abstract – WCN 2013

No: 525

Topic: 36 – Other topic

Association between DBH gene polymorphisms and attention deficit hyperactivity disorder in Korean children

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Attention deficit hyperactivity disorder (ADHD) is a common disorder of the school-age population. ADHD is familial and genetic studies estimate heritability at 80–90%. The aim of the present study was to investigate the association between the genetic type and alleles for DBH gene in Korean children with ADHD. The sample consisted of 142 ADHD children and 139 control children. We diagnosed ADHD according to DSM-IV. ADHD symptoms were evaluated with Conners' Parent Rating Scales and Dupaul Parent ADHD Rating Scales. Blood samples were taken from the 281 subjects, DNA was extracted from blood lymphocytes, and PCR was performed for DBH polymorphism. Alleles and genotype frequencies were compared using the Chi-square test. We compared the allele and genotype frequencies of DBH gene polymorphism in the ADHD and control groups. This study showed that there was a significant correlation among the frequencies of the rs1611115 (OR = 0.64, 95% CI = 0.42–0.97, p = 0.034) of alleles of DBH, but the final conclusions are not definite. Follow-up studies with larger patient or pure subgroups are expected. These results suggested that DBH might be related to ADHD symptoms.

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Abstract – WCN 2013

No: 527

Topic: 36 – Other topic

Neurological complications after neonatal bacteremia: The clinical characteristics, risk factors and outcomes

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Background: Neonates with bacteremia are at risk of neurological complications, but the relevant information is understudied.

Objective: To characterize the clinical features, risk factors, and outcomes of neonatal bacteremia-related neurological complications (BNCs).

Methods and materials: A retrospective cohort study of neonates with BNCs in a tertiary-level neonatal intensive care unit was conducted. Systemic chart review was performed to identify clinical characteristics and outcomes. A cohort of related conditions was constructed as the control group, and logistic regression was used to identify independent risk factors for BNC.

Results: Of 1075 episodes of bacteremia, 35 neonates had BNCs. Three-fifth (21/35) of BNCs were due to meningitis, six were presumed meningitis and eight occurred after hypoxia and septic shock. The most common pathogen to cause BNC is Group B streptococcus (GBS) (15/35, 42.9%) and *Escherichia coli* (6/35, 17.1%). The major BNCs consisted of seizure (27), hydrocephalus (19), encephalomalacia (11), cerebral infarction (7), subdural empyema (6), and post-infectious encephalopathy (3). 8 (22.8%) neonates died, and 7 neonates were discharged at critical condition after the family requested and withdrew all treatment. Among 20 survivors, 10 had neurological sequelae when discharged. In the cohort of neonates with meningitis and septic shock, extremely preterm (gestational age <28 weeks) and meningitis caused by GBS were independent risk factors for development of BNC.

Conclusion: Neonatal bacteremia with meningitis is associated with high risk of neurological complications, especially caused by GBS. Aggressive treatment is worth consideration because bacteremia-related neurological complications potentially result in mortality and morbidities.

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Abstract – WCN 2013

No: 511

Topic: 36 – Other topic

Vestibular evoked myogenic potentials (VEMPs) from upper limb muscles

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Background: Vestibular evoked myogenic potentials (VEMPs) are a relatively new method of recording function from the vestibular nervous system. It relies on the fact that the saccule has retained its sensitivity to sound and can be stimulated by such. Recordings are frequently performed from the sternocleidomastoid muscle (cervical VEMPs or cVEMPs) and more recently from the inferior oblique muscle (ocular VEMPs or oVEMPs). Recording from the limbs is rarer and has so far been done in only a few selected muscles such as the gastrocnemius and triceps. A systematic study of all limb muscles within and between individuals has not so far been done.

Objective: To systematically study the characteristics of VEMPs from upper limb muscles.

Material and methods: Fourteen physiologically normal volunteers were recruited (age range 17–25 years, 4 males and 10 females). Together with the cVEMPs and oVEMPs, attempts to record VEMPs in response to moderate intensity (120 dB pSPL) tone (500 Hz) stimulation were done from the deltoid, biceps brachii, triceps brachii and flexor carpi radialis muscles.

Results: Reproducible responses against a background of tonic contraction were obtained from all muscles, with mean initial peak times of 15 ms (cVEMP), 9.9 ms (oVEMP), 11.5 ms (deltoid), 12.3 ms (biceps), 12.12 ms (triceps) and 16.0 ms (FCR).

Conclusion: It is possible to record VEMPs from various upper limb muscles with different mean latencies. This should prove useful in the future to evaluate lesions in the anterior funiculus of the spinal cord at different levels.

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Abstract – WCN 2013

No: 514

Topic: 36 – Other topic

Neural correlates of inspection time task performance: A developmental study

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Background: Inspection Time Task (ITT) is used as an index of individual differences in perceptual discrimination speed. It is a reliable predictor of the relation between processing speed and psychometric intelligence. Converging findings showed that people with higher IQ identify a briefly presented stimulus more quickly and with greater accuracy than those of a lower IQ.

Objective: To examine the neural correlates of developing intelligence by contrasting the behavioral performance as well as the event-related potentials (ERPs) of two groups of children, one with high IQ scores and one with low. To our knowledge, speed of processing as neural correlate of intelligence has not been studied systematically during school age and adolescence.

Material and methods: This study involved two age-matched samples of 158 children aged 7 through 18 years, selected on the basis of their scores on Wechsler Abbreviated Scales of Intelligence. Children performed the ITT while their electro-cortical activity was registered using a high-density 128-channel electroencephalography acquisition system.

Results: Differences were found in the amplitude course of ERPs between 'high IQ' and 'low IQ' individuals which are consistent with their differences in inspection time. 'High IQ' individuals were faster, less prone to err, and demonstrated significantly larger N1 responses. N1 latency and other ERP components did not differentiate between the two IQ groups.

Conclusion: Given the specificity of ERP group differences to the N1, the results of the present study suggested that allocation of attentional resources is more difficult for individuals with lower IQ scores.

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Abstract – WCN 2013

No: 505

Topic: 36 – Other topic

Hyperactivity and impulsivity in children with allergic rhinitis

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Background: Allergic rhinitis (AR) is the most common chronic allergic disease in school-age children. In addition to physical complications, sleep disturbances, poor school performance, and hyperactivity are all mental complications seen in many children related to their nasal allergies. Increased prevalence and odds ratio of attention-deficit/hyperactivity disorder (ADHD) in AR patients were also noted by analyzing the Taiwan's National Health Insurance Research Database. Inattention and hyperactivity in AR children have not been investigated by objective and scientific measurements.

Objective: The goal of this study was to see if there was a difference in inattention and hyperactivity between AR children and age-matched control children by questionnaires and software.

Patients and methods: In this study, we applied the AR symptom score (T5SS), the ADHD symptom and severity scale (SNAP-IV) and the computerized continuous performance test (CPT) to study the attention and the impulsivity in AR children and age-matched control children (aged 6 to 15 years old).

Results: Fifty-nine AR children and seventeen control children were enrolled. There were no differences of age and gender between two groups. Hyperactivity subscales of SNAP-IV from both parents and teachers scored significantly higher in AR children ($p < 0.01$). CPT revealed higher commission error rate, shorter reaction time, and higher d-prime ($p < 0.01$).

Conclusion: AR children were more hyperactive and impulsive than control children. For AR children, behavior and emotion problems in family and at school need close attention and further investigation.

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Abstract – WCN 2013**No: 498****Topic: 36 – Other topic****Electrical vestibular stimulation after vestibular deafferentation and in vestibular schwannoma**

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Background: Vestibular reflexes evoked by human electrical vestibular stimulation (EVS) are used to investigate vestibular function and its pathways. Vestibular schwannoma causes vestibulo-cochlear nerve compression in the internal auditory meatus interrupting the signal conduction along the vestibular pathway.

Objective: We investigated the electrically-evoked vestibulo-ocular reflex (eVOR) after vestibular deafferentation and in vestibular schwannoma.

Patients and methods: EVORs were recorded as binocular 3D eye rotations evoked by bipolar or unipolar, 100 ms current-step at 0.9–10.0 mA with dual-search coils from 4 vestibular schwannoma patients confirmed by MRI, 12 unilateral vestibular deafferentation (UVD), 5 bilateral vestibular deafferentation (BVD), 2 unilateral labyrinthectomy (UL) patients and 17 normal subjects.

Results: After BVD, bipolar EVS elicited no eVOR. After UVD, bipolar EVS of one functioning ear elicited biphasic excitatory eVOR to cathodal EVS with 9 ms latency and inhibitory eVOR to anodal EVS, opposite in direction, at half the amplitude with 12 ms latency, exhibiting excitatory–inhibitory asymmetries. Both excitatory and inhibitory eVORs were maintained with EVS durations and graded with EVS intensities. The eVOR from vestibular schwannoma was consistent with eVOR from UVD demonstrating the functional loss on the lesion side. We found that unipolar EVS activates both ears due to current spread. Unipolar EVS of the UVD ear produced one-third of the bipolar eVOR, instead of the expect absent response.

Conclusion: We showed that eVOR from one functioning vestibular labyrinth was biphasic with a latency difference between the normal and lesion side. This EVS test may be utilized to investigate unilateral vestibular lesion such as vestibular schwannoma.

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Abstract – WCN 2013**No: 499****Topic: 36 – Other topic****Vestibular and saccadic abnormalities in Gaucher's disease**

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Background: Gaucher's disease (GD) is a hereditary lysosomal storage disease characterized by abnormal deposition of glucocerebroside due to the enzyme glucocerebrosidase deficiency, resulting in multi-organ pathology. GD type III has a progressive neurological involvement.

Objective: To determine vestibular and saccadic abnormalities in GD type III.

Patients and methods: The vestibular and saccadic responses of two siblings with genetically-identified GD Type III on enzyme replacement therapy were evaluated. Vestibular functions were assessed with the head impulse test (HIT), vestibular evoked myogenic potentials (VEMPs) and electrical vestibular stimulation (EVS). Saccadic functions were investigated with volitional horizontal and

vertical saccades to $\pm 20^\circ$. Three-dimensional head and eye movements were recorded with dual-search coils and VEMP with surface electrodes.

Results: Individual semicircular canal functions were impaired with angular vestibulo-ocular reflex (VOR) gains to HIT halved and absent horizontal refixation saccades. Ocular and cervical VEMPs to air-conducted clicks were absent in the older sibling and only cervical VEMP was present in the younger sibling indicating otolithic dysfunction. EVS showed prolonged onset latency and attenuated tonic and phasic responses suggesting impaired neural conduction and vestibular function. Horizontal saccadic velocity was miniscule ($<30^\circ/s$) and multiple back-to-back saccades with saccade–vergence interaction were utilized to minimize eye position error in the older sibling. Vertical saccades, vergence and smooth pursuit were normal in both siblings.

Conclusion: Our findings suggest that GD affects the vestibular nuclei in addition to paramedian pontine reticular formation. These vestibular and saccadic abnormalities may have implications for activities of daily living that requires visual–vestibular interaction.

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Abstract – WCN 2013**No: 495****Topic: 36 – Other topic****Syphilitic polyradiculoneuropathy in an immunocompetent patient**

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Background: Although the incidence of neurosyphilis has decreased, it is still prevalent. Its most common clinical presentation includes tabes dorsalis and general paresis. In contrast, polyradiculoneuritis has rarely been reported during syphilis. The authors report a case of acute polyradiculoneuritis caused by *Treponema pallidum* in an immunocompetent patient.

Objective: This case reports the interest to evoke a syphilitic origin in polyradiculoneuropathy.

Case report: A 45 year old man without medical past history was referred to neurology outpatient for acute leg weakness and heaviness, walking difficulties. Clinical examination revealed bilateral leg weakness. Deep tendon reflexes were absent at the knees and the ankles, cutaneous hypoesthesia on the legs. There were no pyramidal signs, no cranial nerves abnormalities and no incontinence. Electroneuromyograms showed sensorimotor demyelination and axonal loss evoking polyradiculoneuritis. Lumbar punctures showed an albumino-cytological dissociation (hyperproteinorachy at 2.2 g/l, normal cell count and glucose level). Serum VDRL and TPHA were positive (VDRL: 1/16, TPHA: 1/1280). The VDRL and TPHA were positive in the CSF (VDRL: 1/6, TPHA1/640). Serodiagnostic tests (HIV, Borrelia, hepatitis C and B) were negative. The patient received 30 million units of intravenous penicillin G daily for 10 days, four times with delay of 3 months. Muscular leg weakness disappeared and his clinical condition improved.

Conclusion: Syphilitic polyradiculoneuropathy has rarely been reported. Infection due to *T. pallidum* is a curable cause of polyradiculoneuritis which may occur in HIV-negative patients; appropriate tests in both serum and CSF should be performed in patients with polyradiculoneuritis.

doi:10.1016/j.jns.2013.07.2075

Abstract – WCN 2013**No: 435****Topic: 36 – Other topic****Sleep problem of premature infants: Clinical utility of the modified Chinese version of the brief infant sleep questionnaire (CBISQ)**S.-M. Chu, Y.-S. Huang, M.H. Tsai. *Chang Gung Memorial Hospital, Taoyuan, Taiwan, ROC*

Background: Previous studies have suggested differences in sleep quality between preterm and term born infants, and premature infants are at higher risk for sleep-disordered breathing.

Objective: We aimed to evaluate sleep problems of preterm infants and compare the results with full-term infants based on the modified Chinese version brief infant sleep questionnaire (CBISQ).

Methods and materials: To validate the modified CBISQ, we collected 69 6-months old full term infants and 177 premature infants born at <37 weeks of gestation. All parents completed a modified CBISQ. First we examined the test–retest reliability of the modified CBISQ. Then, we compared the questionnaire-responses with other subjective (sleep diary) and objective (actigraphy) measures of sleep. Finally, we evaluated the validity and potential usage of the questionnaire for diagnostic purpose by comparing results to those obtained through nocturnal polysomnography (PSG).

Results: Test–retest reliability of CBISQ was acceptable. The nocturnal sleep duration, number of night waking, daytime sleep duration, time spent with mouth breathing and loud-noisy breathing had significant correlation with sleep diary, actigraphy and polysomnography. The CBISQ showed that the sleep on infants' side, nocturnal sleep duration, being held to fall asleep, number of night waking, daytime sleep duration, subjective consideration of sleep problem, loud-noisy breathing and time spent with crying during night were significantly different between premature infants and full term infants.

Conclusion: The CBISQ is a reliable and valid tool for the measurement of sleep problems in infants. Premature infants have more sleep problems compared to term born infants.

doi:10.1016/j.jns.2013.07.2076

Abstract – WCN 2013**No: 387****Topic: 36 – Other Topic****Coronal oblique orientation offers improved visualization of neuroforamina in cervical spine magnetic resonance imaging**W. Freund^a, S. Klessinger^b, M. Mueller^a, M.-E. Halatsch^c, G. Hoepner^a, B.L. Schmitz^d. ^aDiagnostic and Interventional Radiology, University Hospitals Ulm, Ulm, Germany; ^bNeurosurgery, Nova Clinic, Biberach, University Hospitals Ulm, Ulm, Germany; ^cNeurosurgery, University Hospitals Ulm, Ulm, Germany; ^dNeuroradiology, University Hospitals Ulm, Ulm, Germany

Background: Angulated projections are standard in conventional radiography of the cervical spine but rarely used in magnetic resonance imaging (MRI). Improved visualization of neuroforaminal pathology is necessary especially preoperatively to discern the degree of neuroforaminal stenosis and the cause (osseous or soft).

Patients and methods: In a retrospective setting, 25 consecutive patients with cervical monoradiculopathy were identified. T2-weighted sagittal, coronal oblique and transversal slices were anonymized, generating 75 “cases” consisting of only one spatial orientation. 2 radiologists and 2 neurosurgeons blinded to the diagnosis individually read the cases. One of each pair was experienced with the coronal oblique slices; the other received a 10-minute training. Criteria were site, cause and grading of the neuroforaminal stenosis and the level of

confidence on a 100-point visual analogue scale (VAS). We computed interrater agreement and t-tests, taking $p < .05$ as statistically significant.

Results: The sensitivity to detect the relevant neuroforaminal stenosis was .45 for transversal, .63 for sagittal and .61 for coronal oblique scans. The combination of three planes increased sensitivity to .77 to detect the relevant lesion. The readers felt significantly more confident in attributing the cause of pathology on coronal oblique planes (76.4 VAS points, $p = .006$) vs. 68.2 (sagittal) vs. 68.6 (transversal).

Interrater agreement was significantly better for experienced (kappa .48) than inexperienced readers (.32, $p = .02$).

Conclusion: Coronal oblique planes in cervical spine MRI increase sensitivity and confidence in attributing the cause of neuroforaminal obstruction. They are easy to interpret; however, experience with the sequence increases interrater agreement.

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Abstract – WCN 2013**No: 377****Topic: 36 – Other Topic****Adult-onset of mitochondrial encephalomyopathy lactic acidosis and stroke-like episodes (MELAS) syndrome mimicking herpes encephalitis: A case report and review of literature**S. Collorone, S. Pontecorvo, A. Francia. *'Sapienza' University of Rome, Rome, Italy*

Background: Mitochondrial encephalomyopathy lactic acidosis and stroke-like episodes (MELAS) syndrome is a multisystem disorder with a widespread phenotypic variability.

Objective: The objective of this work is to report an unusual presentation of MELAS mimicking herpes encephalitis compared to cases described in literature.

Description: A 47 year-old man presented acute onset of confusion, agitation and productive aphasia. He had a history of spina bifida, allergy to acetylsalicylic acid, sensorineural hypoacusis and migraine-like headache. There was no family history of neurological disease. A cerebral magnetic resonance imaging (MRI) showed large bilateral lesions of temporal lobes, hyperintensity in T2-FLAIR and apparent reduction in diffusion coefficient (ADC). Empirical treatment with antiviral and corticosteroids was begun for presumed herpes simplex encephalitis, with improvement in his clinical conditions and reduction of temporal lesions. After one month he developed balance disorder and dizziness so he was hospitalized in our neurology department. Cerebral MRI showed new large lesions (temporal lobe, parietal lobe and calcarine cortex) with same MRI features of first episode. Magnetic resonance spectroscopy (MRS) showed increase in lactate in lesions and liquor confirmed by the lumbar puncture. MELAS was suspected and diagnosis was confirmed by detection of nucleotide 3243 A → G mutation in mitochondrial DNA.

Discussion: Six MELAS cases mistaken for viral encephalitis are described in medical literature: correct diagnosis was achieved after a second stroke-like episode or after ineffective treatment.

Conclusion: MELAS should be included in differential diagnoses of herpes encephalitis on the basis of detailed history, which provides early diagnostic clues, and cerebral MRI features. Our case brings out the importance of MRS: detecting lactate increase in liquor and lesions, MRS could be a crucial aid for early diagnosis.

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Abstract – WCN 2013**No: 409****Topic: 36 – Other Topic****Contribution of major amyotrophic lateral sclerosis related genes to the etiology of the disease in Chinese**Z.-Y. Zou, X.-G. Li, M.-S. Liu, L.-Y. Cui. *Peking Union Medical College Hospital, Beijing, China***Background:** Mutations in SOD1, ANG, TARDBP, FUS, VCP, C9ORF72, and PFN1 genes have been identified in ALS patients.**Objective:** The objective of this work is to determine the mutations in major ALS-related genes in a large cohort of Chinese familial (FALS) and sporadic (SALS) patients and the genotype–phenotype associations.**Methods:** Screening for mutations of SOD1, ANG, TARDBP, FUS, VCP, C9ORF72, and PFN1 genes was consecutively carried out in 20 index FALS patients, 324 SALS patients, and 245 healthy controls.**Results:** Overall, mutations were detected in 35.0% (7/20, 95%CI = 14.1–55.9%) and 4.0% (13/324, 95%CI = 1.9–6.1%) of FALS and SALS patients, respectively. SOD1 (5/20, 25.0%) and FUS (2/20, 10.0%) account for all mutations in FALS patients, whereas FUS (6/324, 1.9%) was the most frequently mutated gene in SALS patients, followed by SOD1 (3/324, 0.9%), TARDBP (3/324, 0.9%), and ANG (1/324, 0.3%). No mutations were detected in VCP, C9orf72, and PFN1 gene. Patients with FUS mutations were younger at onset ($P < 0.01$) and had shorter lifespan ($P < 0.01$), compared with those without the mutations.**Conclusion:** Mutations in major ALS-related genes were present in approximately 35% and 4% of Chinese FALS and SALS patients, respectively. SOD1 and FUS are the most frequently mutated genes both in FALS and SALS patients in Chinese. It appears to be different from the profiles reported in Caucasian ALS patients, in which C9orf72 and SOD1 are the most common mutated genes. Moreover, FUS mutations are associated with an early onset and poor prognosis. The results suggest that there is an ethnic difference in the genetic background of ALS.

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Abstract – WCN 2013**No: 400****Topic: 36 – Other Topic****Hashimoto's encephalopathy associated with an elevated intrathecal IGG4 level**Y. Hosoi^a, S. Kono^a, T. Terada^b, T. Konishi^a, H. Miyajima^a. ^aFirst Department of Medicine, Hamamatsu University School of Medicine, Hamamatsu, Japan; ^bLaboratory of Human Brain Imaging Research, Molecular Imaging Frontier Research Center, Hamamatsu University School of Medicine, Hamamatsu, Japan**Background:** Hashimoto's encephalopathy is a steroid-responsive neurological syndrome associated with the existence of anti-thyroid antibodies, which is characterized by various neuropsychiatric symptoms. The autoimmune mechanisms are thought to play a pathogenic role in this disorder. Recently, some reports showed an elevated serum IgG4 level in patients with Hashimoto's thyroiditis.**Objective:** The pathogenic role of IgG4 in a patient with Hashimoto's encephalopathy is investigated.**Patient and method:** The patient was a 60-year-old Japanese male who presented with a 6-month history of progressive gait disturbance. He developed memory disturbance and involuntary movements of his face and neck 2 months after presentation. The IgG4 concentrations in the serum and cerebrospinal fluid, MRI scans of brain, and response of corticosteroid therapy were investigated.**Results:** A neurological examination revealed cerebellar ataxia and grimacing face with cervical dystonia. The MMSE score and FAB score were within normal range. Laboratory tests revealed a high titer of

anti-thyroglobulin antibodies in the serum. The serum IgG4 concentration was elevated (298 mg/dl; normal: 4.8–105). A CSF analysis revealed an elevated level of IgG4 (3.5 mg/dl; normal: not detected). There was no clinical finding suggesting complications of IgG4-related disease. A brain MRI revealed symmetrical periventricular high signal intensity on T2-weighted images and FLAIR images. Corticosteroid therapy improved the neurological symptoms, intrathecal IgG4 level, and abnormal MRI findings.

Conclusion: An elevated IgG4 level in the cerebrospinal fluid, as well as in the serum, may provide helpful clues about the possible autoimmune mechanism involved in Hashimoto's encephalopathy.

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Abstract – WCN 2013**No: 404****Topic: 36 – Other Topic****BMI impact on neurological diseases**J. Xhaxho^a, I. Alimehmeti^b, S. Xhaxho^c, J. Kruja^a, D. Dobi^c. ^aDepartment of Neurology, Tirana, Albania; ^bDepartment of Endocrinology, Tirana, Albania; ^cCUHT Albania, Tirana, Albania**Introduction:** In neurological practice there were more patients with obesity-related diseases such as follows: stroke, headache, tunnel carpal syndrome, intracranial hypertension, etc.**Aim:** The aim of this study is to investigate the impact of BMI in some neurological diseases.**Methods:** We included in this study 263 persons. 154 of them were hospitalized in the Clinic of Neurology, in University Hospital Centre "Mother Teresa", Tirana and 109 were control group (They are random persons with the same age-group with the admitted persons). For all of them we fulfilled a form with general data and specific data on risk factors for several neurological diseases.

We classified them in 4 groups. For every person we calculate the BMI. We compared BMI of control group with BMI of persons with neurological diseases.

Results: We found that the BMI was 1 kg/m² greater in stroke patients compared with control group and that there was a difference of 0.6 kg/m² in patients with other neurological diagnosis compared with control group ($z < 0.01$).BMI was 0.5 kg/m² greater in control group with arterial hypertension compared with patients with arterial hypertension with neurological diseases.BMI was 1.25 kg/m² greater in control group with mellitus diabetes compared with patients with mellitus diabetes with neurological diseases.BMI was 2 kg/m² greater in smoker patients with neurologic diseases compared with control group.BMI was 2 kg/m² greater in alcoholic persons with neurologic disease than control group.**Conclusion:** Obesity is a significant risk factor for neurological diseases.

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Abstract – WCN 2013**No: 374****Topic: 36 – Other Topic****Neurological diseases in pregnancy**D. Ndoja, A. Kuqo, J. Naska, A. Rroji, L. Stefanidhi, L. Buda, J. Kruja. *Department of Neurology, UHC Mother Theresa, Service of Neurology and Radiology, Tirana, Albania***Objective and materials:** Eleven pregnant women aged 28.72 years old (DS +/- 7.18; mean age: 16–37 years) admitted in our

department for 6 months in 2012 (one at the first trimester, three at the second or the last trimester respectively) were prospectively studied.

Results: Three were at their 1st pregnancy, 4 were post-partum while others from 2nd to 4th pregnancies. Some ATCD reported were: miscarriages in 4, trauma during pregnancy, venous thrombosis of the inferior extremities and appendectomy in one respectively. The etiology reported were: epilepsy (E) in 5 while 6 showed DVT, posterior ischemic stroke, and headache (primary, post-PL orthostatic hypotension from the anesthesia during SC) in two patients respectively, and reversible vasoconstriction syndrome in one. Among the epileptic pts; one had its first seizure (S) before pregnancy, and others; S heralding DVT, epilepsy from right MCD, post-traumatic S during pregnancy, and post-partum S in one consecutive case. Two presented CPS and 3 PM secondary generalized S. MRI was normal in 5; DVT and ischemic stroke in 2 patients respectively; multi lacunar P bilateral stroke and right P schizencephaly in two patients. EEG on E cases displayed anterior bilateral left sided dominance SHW and SW. The therapy useful was the small MW anticoagulants in stroke and DVT, and AEDs (CBZ, LTG) in E cases. All patients delivered in term after the neurological improvement except one, which interrupted its 4th pregnancy.

Conclusion: Epilepsy followed by DVT, stroke and headache were the most frequent neurological diseases that occurred during pregnancy.

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Abstract – WCN 2013

No: 379

Topic: 36 – Other Topic

Vascular burden in acute stroke: An MRI study

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Background: Intracranial atherosclerotic disease (ICAD) is the most common mechanism of ischemic stroke worldwide, particularly among Asians. Magnetic resonance angiography (MRA) is a reliable modality for diagnosis. Utilizing magnetic resonance imaging (MRI) as the primary neuroimaging tool for all patients with suspected acute stroke, we prospectively evaluated the association of acute stroke with background ICAD, chronic microvascular ischemic changes (CMI) and microhemorrhages (MH).

Methods: 468 consecutive patients presenting over a three-month period had MRI with axial diffusion weighted imaging, T2W, gradient sequences and 3D TOF MRA performed; 5 patients were too restless for 3D TOF MRA. Fisher's exact test was used to analyze the data.

Results: Acute stroke with restricted diffusion was reported in 260 (55.6%) patients. ICAD was present in 339 (73.2%); 321 (68.6%) revealed CMI while 168 (35.9%) showed MH. Underlying ICAD, CMI and MB were significantly present in patients with acute stroke ($p < 0.0001$, $p < 0.0001$ and $p = 0.0154$ respectively). In 139 patients, both MH and CMI were present, demonstrating the strong association between them ($p < 0.0001$).

Conclusion: ICAD is very common in our Asian population comprising largely Chinese, Indian and Malay ethnicities. MRI in acute stroke demonstrated a strong association with underlying ICAD, CMI and MH.

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Abstract – WCN 2013

No: 386

Topic: 36 – Other Topic

Evolution of regionally accentuated reversible brain grey matter reduction in ultra marathon runners detected by voxel based morphometry

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Background: During the 4487 km ultra marathon TransEurope-FootRace 2009 (TEFR09), runners showed catabolism with considerable reduction of body mass. We hypothesized that the athletes have acquired changes to grey matter (GM) brain morphology due to continuous high volume training and will show additional changes during the race.

Material and methods: Prior to the start of the race 13 runners volunteered to participate in this study of planned brain scans before, twice during, and 8 months after the race. A group of matched controls was recruited for comparison.

Scanning was performed with identical 1.5 T Siemens Avanto scanners, in a mobile MRI unit escorting the runners. A volumetric 3D dataset was imaged using a MPRAGE sequence. A level of $p < 0.05$, family-wise corrected for multiple comparisons was set.

Results: Comparison of TEFR09 participants and controls revealed no significant differences regarding the GM brain volume. During the race however, voxelwise morphometry revealed that GM concentration decreases in regionally distributed brain regions. These included the bilateral posterior temporal and occipitoparietal cortices as well as anterior cingulate and caudate nucleus. After eight months, regional GM differences came back to baseline.

Conclusion: We did not observe baseline differences between TEFR09 athletes and rather sedentary controls. However, during the race GM concentration decreased in brain regions normally associated with higher visuospatial and language tasks. Also the possible reduction of the energy intensive default mode network as a means to conserve energy during catabolism is discussed. These changes are reversible after 8 months and seem to be adaptive.

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Abstract – WCN 2013

No: 359

Topic: 36 – Other Topic

A leprosy patient presenting with ejaculatory dysfunction

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Leprosy is a chronic granulomatous disease of the skin and nerves caused by the bacillus *Mycobacterium leprae* (*M. leprae*). *M. leprae* has a high affinity for Schwann cells. This tissue tropism is the basis of the characteristic nerve damage associated with leprosy, often leading to peripheral neuropathy. The ulnar nerve is most commonly involved. The patients predominantly present with mononeuritis multiplex. Sometimes, but not very frequently, involvement of the pudendal nerve is also observed. In this case report, a patient, with a diagnosis of leprosy and presented with ejaculatory dysfunction is followed up.

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Abstract – WCN 2013**No: 346****Topic: 36 – Other Topic****Voltage-gated potassium channel antibodies (VGKC-AB) associated with limbic encephalitis: Borderline positive levels in Asian patients?**

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Background: VGKC-Ab has been described in patients with features of limbic encephalitis (LE). In contrast with paraneoplastic LE which typically heralds a poor prognosis, VGKC-Ab-related LE has potential for reversibility. This has thus far been under-documented in Asian patients.

Objectives: The objective of this study is to describe the clinical, serological and neuroimaging features of eight Chinese patients with encephalitis associated with borderline positive titres (100–400 pM) of VGKC-Ab.

Patients and methods: Eight Chinese patients (four males, four females; aged 23–87 years) were identified in a tertiary centre over a 3-year period. Clinical data was acquired from a retrospective review of medical records.

Results: Memory impairment and psychiatric symptoms were prominent in five older patients. Three experienced disorientation and agitation. Three reported sleep disturbances or hallucinations. Two younger patients had seizures. One suffered quadriparesis. All eight had elevated VGKC-Ab (ranging 113–352 pM). Paraneoplastic antibodies were negative in all six tested. Autoantibody testing revealed thyroperoxidase, thyroglobulin, antinuclear, NMDA or NMO antibodies in five patients. CSF studies of five patients showed leucocytosis with lymphocytic predominance in three. Cranial MRI demonstrated hippocampal abnormalities in four patients. Seven patients received intravenous immunoglobulin, methylprednisolone or plasmapheresis. Six patients demonstrated good clinical response to treatment, with four achieving pre-morbid function and two undergoing active rehabilitation. Two exhibited persistent psychiatric deficits.

Conclusions: Clinical features at presentation and response to treatment differ based on age – older patients present with cognitive and behavioural changes while younger patients present with seizures and have better outcome. Concomitant autoantibodies may be a feature in Asian patients with borderline positive VGKC-Ab.

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Abstract – WCN 2013**No: 309****Topic: 36 – Other Topic****The pharmacological effect of *Artemisia absinthium* extract in protecting adult rats against lead neurotoxicity**

W. Sansar, H. Gamrani. *University Cadi Ayyad, Faculty of Sciences Semlalia, Marrakech, Morocco*

Lead (Pb) is a heavy metal with no apparent biological function. It is recognized as a dangerous neurotoxin, since it induces morphological and functional abnormalities in the brain. Several studies reported, in the rat brain, the antioxidant and anti-inflammatory effects of aqueous extracts of *Artemisia absinthium*. Here, we investigated the potential role of *A. absinthium* aqueous extract in protecting brain against the effect of chronic lead exposure, especially on the dopaminergic neurons together with the glial system. Using immunohistochemistry of tyrosine hydroxylase (TH), the number of dopaminergic neurons in substantia nigra pars compacta (SNpc) was found to decrease by 50% in lead-treated group, while the glial fibrillary acidic protein (GFAP) immunohistochemistry shows hypertrophic immunoreactive astrocytes in the frontal cortex. The quantification of immunolabelled astrocytes shows an increase by 48% in

comparison with controls. Pharmacological treatment with *A. absinthium* aqueous extract (200 mg/l) during 4 weeks restores most of the changes in the glial and dopaminergic systems which occur in lead intoxicated rats. Our findings suggest that *A. absinthium* might be beneficial for the treatment of the glial and neuronal alterations observed during chronic lead intoxication in adult rat.

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Abstract – WCN 2013**No: 290****Topic: 36 – Other Topic****The comparisons of adrenergic and vagal baroreflex sensitivity between patients with diabetic neuropathy and control group**

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Background: The baroreflex is responsible for maintaining stable blood pressure despite of positional change. Measurement of baroreflex sensitivity (BRS) using beat-to-beat blood pressure monitoring can calculate the adrenergic and vagal components separately. Several indexes during Valsalva maneuver (VM) have recently been introduced and an index of the adrenergic components of the BRS has been validated. Autonomic neuropathy in diabetic patients is relatively common and bears high mortality and morbidity.

Objective: The purpose of this study is to analyze if the adrenergic and vagal components of BRS could selectively be damaged in diabetic autonomic neuropathy (DAN) and if it would be correlated with Composite Autonomic Symptom Scale (COMPASS).

Patients and methods: We prospectively collected patients with diabetic neuropathy and control group. We performed Korean version of COMPASS, and VM using Finapress® in both groups. During VM, four phases from phase 1 to phase 4 were defined and three variables including pressure recovery time (PRT), adrenergic BRS (BR_{Sa}), and vagal BRS (BR_{Sv}) were calculated.

Results: The statistical consequences of COMPASS, PRT, BR_{Sa}, and BR_{Sv} in DAN (n = 37) were 37.1 points (± 16.9), 2.90 s (1.880–7.250), 5.50 mm Hg/s (2.20–9.50), and 0.27 min/mm Hg (± 0.27). Control group (n = 62) was checked as 32.13 points (± 18.4), 2.0 s (1.200–3.175), 12.00 mm Hg/s (7.65–26.25), and 0.42 min/mm Hg (± 0.29) respectively.

Conclusions: Our study showed that BR_{Sa}, BR_{Sv}, and PRT in DM patients were lower and longer than in control group. However, there was no meaningful difference in COMPASS between two groups. We concluded that especially, adrenergic component of baroreflex sensitivity (BR_{Sa}, PRT) would be more predictable for the severity in DAN.

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Abstract – WCN 2013**No: 690****Topic: 36 – Other Topic****Neurosyphilis in a young patient presented as rapid cognitive decline: Case report**

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We present the case of a 38 year old man who was admitted for a rapid cognitive decline and headache. CT and MRI studies revealed a normal pressure hydrocephalus. He was assessed for surgery and a ventriculo-peritoneal shunt was realized. Neurologic examination

revealed mild spastic tetraparesis, right Argyll–Robertson pupil, gait disturbance, severe cognitive decline with amnesic aphasia and asymmetric myoclonic jerks predominantly in the lower limbs. The CSF showed normal pressure, 12 lymphocytes/mm³, 0.55 g/L proteins, and positive TPHA test. Serum TPHA and VDRL were intense positive at small dilutions with negative HIV serology. EEG showed a theta rhythm with intermittent generalized triphasic discharges. Psychiatric and neuropsychological examinations showed a MMSE test of 12 with anterograde amnesia, nominal aphasia, and ideomotor apraxia. The final diagnosis was neurosyphilis with moderate–severe dementia and the patient received ceftriaxone 4 g intravenous daily. Mild improvement of the symptoms was observed, but the patient must be reassessed periodically.

Syphilis is a sexually transmitted infection caused by *Treponema pallidum*, known to have three distinct phases. The central nervous system involvement may occur as early as the second phase generally with syphilitic meningitis, but neurosyphilis always occurs in the tertiary phase with meningeal, meningovascular or parenchymal manifestations. Even with a constant reduction of the incidence worldwide, syphilis should always be considered in young patients with rapid cognitive decline and other neurologic manifestations.

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Abstract – WCN 2013

No: 658

Topic: 36 – Other Topic

Chronic demyelinating polyradiculoneuropathy-like disorders with and without diabetes mellitus: Clinical and electrodiagnostic study

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Background: Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is characterized by relapsing or progressive dysfunction of the motor and/or sensory fibers of the peripheral nervous system due to inflammatory demyelination. CIDP is important to recognize as it is potentially treatable.

Objective: The objective of this study is to study the clinical and electrophysiological characteristics of patients with CIDP and compare between CIDP patients with and without diabetes mellitus.

Methods: Prospective study of patients referred for electrodiagnostic studies in Benghazi who fulfilled the diagnostic criteria of the European Federation of Neurological Societies/Peripheral Nerve Society for CIDP. The patients were studied between August 2003 and 2009.

Results: A total of 85 patients were studied. Their mean age was 57 years (range 27–80) and 48 were males. Fifty-six patients had DM type-II. The mean duration of diabetes was 7 years (ranged 1–20 years). The main duration of illness was 10 months (ranged between 2 and 60 months). Weakness and paraesthesia of the lower limbs were the most common symptoms. Electrophysiological studies were taken on all patients and demonstrated marked slowing and more frequent unrecordable motor and sensory as well as F-wave responses in patient with CIDP and diabetes (CIDP-D). The frequencies of conduction block and electromyographical (EMG) evidence of denervation and reinnervation were similar between idiopathic CIDP (I-CIDP) and CIDP-D.

Conclusion: CIDP associated with diabetes mellitus (CIDP-D) had more severe disease and was associated with more significant and higher frequency of electrophysiological findings that indicated a demyelinating neuropathy compared with idiopathic CIDP (I-CIDP).

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Abstract – WCN 2013

No: 627

Topic: 36 – Other Topic

The frequency of functional disorders in new neurology-outpatients: An audit study

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Background: A few studies have investigated the incidence of functional disorders among neurological patients. They demonstrate around 16% of patients present with functional symptoms that neurologists are unable to explain organically.

Objective: Information on the nature and relative frequency of diagnoses made in referrals to outpatient neurology clinics is essential to target specific services tailored for patients' needs. The aim of this study is to address the frequency of functional symptoms among neurology outpatient referrals to The Walton Centre NHS Trust (Liverpool, UK).

Method: 503 medical notes of newly referred outpatients attending neurology clinics at The Walton Centre in June 2011 were collected and screened. For each patient, the following information was gathered: age; main presenting symptom; whether the diagnosis was organic or functional; was the organic/functional diagnosis changed in 1 year time; was a second opinion requested and eventual diagnosis formulated.

Results: 15% of the referrals examined exhibited functional symptoms (n = 77). Of these, 65% (50) were females and 35% (27) were males. Functional disorders were the second most common diagnostic group, with the most common being headache. The most common functional symptoms were, in order: sensory disturbance, blackouts/funny turns/dizziness, seizures. Functional patients had a younger average age of 42.5 years, compared with 48 for all patients. Results confirm those of previous studies.

Conclusions: The percentage of functional symptoms is quantitatively relevant and cannot pass unnoticed, requiring special attention from both the clinicians and the local health service. Different professional approaches must be integrated in a unique multidimensional treatment.

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Abstract – WCN 2013

No: 646

Topic: 36 – Other Topic

Susac's syndrome: Three cases with predominant branch retinal artery occlusion at first presentation

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Background: Susac's syndrome encompasses the triad of encephalopathy, branch retinal artery occlusion and hearing loss. At first presentation, this clinical spectrum is usually incomplete which often delays diagnosis and appropriate treatment.

Patients: We report three cases of Susac's syndrome with predominant branch retinal artery occlusion at first presentation.

Results: In two cases, diagnosis was made early in the disease course and aggressive immunosuppression could eventually prevent persistent visual field deficits. In the third patient, diagnosis and initiation of corticosteroid treatment were delayed and persistent visual field deficits developed. Retinal artery fluorescein angiography showed leakage of arteries at first presentation in all three patients.

Pathognomonic corpus callosum hyperintensities on magnetic resonance imaging were found in two of our patients at presentation and helped in establishing the diagnosis early.

Conclusion: Only one of our patients showed the full triad of Susac's syndrome including hearing loss. Retinal artery fluorescein angiography revealed to be crucial for early diagnosis and useful in treatment monitoring.

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Abstract – WCN 2013

No: 540

Topic: 36 – Other Topic

Anaplastic ependymoma with ependymoblastic multilayered rosettes

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Background: Anaplastic ependymoma, WHO grade III, is a malignant glioma with ependymal differentiation characterized by high mitotic activity often accompanied by microvascular proliferation and necrosis, where, generally, much fewer ependymal rosettes are found than in ependymoma, WHO grade II. Ependymal rosettes, forming a single layer of tumor cells, differ from ependymoblastic multilayered rosettes which are characteristic histological features of ependymoblastoma, a variant of central nervous system primitive neuroectodermal tumor.

Patient history: A 22-year-old woman had a headache and nausea, and was admitted with a generalized convulsion one month later. Diagnostic imaging revealed a mass lesion of the left frontal lobe. A partial resection of the mass was then performed and the pathological diagnosis was “malignant ependymoma.” The patient deteriorated gradually with local progression and died 23 months after the surgery, at the age of 24. An autopsy was performed.

Methods and Results: Microscopic examination showed the aggregation of true rosettes with multilayering of tumor cells resembling the ependymoblastoma histology. Single-layered ependymal rosettes and perivascular radial arrangements of the tumor cells were also observed. FISH analysis and differential PCR revealed the absence of 19q13.42 amplification, a specific molecular hallmark of ependymoblastoma and embryonal tumor with abundant neuropil and true rosettes, supporting the diagnosis of anaplastic ependymoma.

Conclusion: Anaplastic ependymoma can quite rarely show ependymoblastic multilayered rosettes. Although differentiating this type of ependymoma from ependymoblastoma is challenging, it is expected that the incorporation of molecular and cytogenetic analyses would facilitate the diagnosis.

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Abstract – WCN 2013

No: 631

Topic: 36 – Other Topic

Amyloid β_{42} administration impairs energy metabolism *in vivo* and *in vitro*

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Background: Amyloid β_{42} ($A\beta_{42}$) impairs neuronal metabolism in Alzheimer's Disease (AD). However, the effects of circulating $A\beta_{42}$ on whole body metabolism are unknown.

Objectives: The objectives of this study were to determine the effects of $A\beta_{42}$ on 1) glucose metabolism in insulin sensitive cells *in vitro*; and on 2) whole body metabolism *in vivo*.

Materials and methods: 3T3-L1 adipocytes and FAO hepatocytes were treated with monomeric $A\beta_{42}$ ($mA\beta_{42}$) or aggregated $A\beta_{42}$ ($aA\beta_{42}$) for 48 h, with respective controls being monomeric or aggregated scrambled $A\beta_{42}$ ($scrA\beta_{42}$). Basal and insulin stimulated glucose uptake and glucose production were measured in 3T3-L1 adipocytes and FAO hepatocytes respectively.

In vivo studies: 8 week old, male C57Bl/6J mice ($n = 10/\text{group}$) were treated with $mA\beta_{42}$ and $scrA\beta_{42}$ (control) for two weeks ($1 \mu\text{g}/\text{day}$; I.P. injections). Bodyweight and food intake were monitored daily. Indirect calorimetry was performed after 14 days of treatment and oxygen consumption, respiratory quotient and substrate oxidation determined.

Results: In FAO hepatocytes, $mA\beta_{42}$ increased glucose production, while $aA\beta_{42}$ had no effect. Similarly, $mA\beta_{42}$ impaired glucose uptake in 3T3-L1 adipocytes, while $aA\beta_{42}$ had no effect.

Administration of $mA\beta_{42}$ to mice had no effect on bodyweight or food intake compared with control mice. However, administration of $mA\beta_{42}$ reduced oxygen consumption and total carbohydrate oxidation compared with control animals ($p \leq 0.05$).

Conclusion: This data shows that $mA\beta_{42}$ impairs glucose metabolism while $aA\beta_{42}$ had no effect. Thus, $A\beta_{42}$ may be involved in the dysregulation of metabolism in obesity and type 2 diabetes, where circulating $A\beta_{42}$ levels are elevated.

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Abstract - WCN 2013

No: 636

Topic: 36 - Other Topic

Analysis of background activity in ventilated preterm infants with hypoxic ischemic encephalopathy

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Introduction: The electroencephalogram (EEG) is used as a diagnostic tool for investigation of dysfunction activity and maturity of preterm baby. [I. Alvarado-Guerrero, et al, 2011]. Study of EEG recording of ventilated preterm infants can indicate changes of cerebral activity and diagnose the degree of hypoxic ischemic encephalopathy (HIE).

Method and study groups: EEG recordings were obtained with 13 electrodes by using the international 10–20 system placement and investigation of background activity. All EEG recordings was obtained for 80 preterm baby in age corrected with term newborn. Of those, we enrolled preterm ventilated infants with HIE-50 babies and preterm not ventilated infants without HIE-30 babies.

Results: All ventilated infants ($50/98 \pm 4\%$) have a dominating EEG frequency which is delta activity and not in ventilated infants ($19/63 \pm 17\%$). The background depression (amplitude $< 25 \text{ mV}$) was observed only for ventilated babies ($24/48 \pm 13\%$).

Conclusions: Chronic-stage abnormalities of background EEG were diagnosed in ventilated preterm babies with HIE. These changes include immaturity patterns – slow waves activity (delta frequencies) and depression delta activity.

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Abstract - WCN 2013**No: 260****Topic: 36 - Other Topic****Resistant hypertension de novo after aneurysmal subarachnoid haemorrhage**

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Background and purpose: From the studied variables in subarachnoid hemorrhage (SAH) risk, hypertension is probably the most controllable one. The aim of this study was to determine whether confirmed hypertension, prospectively diagnosed with strict criteria, is an independent risk factor for aneurysmal SAH.

Methods: A case-control study was conducted in 2 Colombian cities between July 2004 and June 2005. There were 163 new cases of SAH (mean age 51 years; 107 were women) with 2 controls per case: 1 hospital and 1 community control. Hypertension was defined according to cardiovascular criteria, based on target organ damage. In addition to hypertension, other variables were studied: present smoking, recent alcohol consumption, alcohol dependency, coffee consumption, cocaine use, and body mass index. A multivariate logistic regression model was used to determine whether hypertension was an independent risk factor.

Results: Among the studied variables, including confirmed hypertension, only present smoking became an independent risk factor for SAH.

Conclusions: Confirmed hypertension is not an independent risk factor for aneurysmal SAH.

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Abstract - WCN 2013**No: 589****Topic: 36 - Other Topic****Dynamics of neurological symptoms in acute period of leptospirosis**

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Background: Leptospirosis is a zoonosis, that, when developing in human, may affect central nervous system. Krasnodar region is endemic for leptospirosis, the morbidity varies from 7.75 up to 29.6 per 100 thousand.

Objective: To investigate the neurological state's dynamics in patients with leptospirosis.

Patients and methods: 66 patients (average age 49.5 ± 4.2 years old) with an ikchetero-hemorrhagic form of leptospirosis were observed.

Results: By the first disease week end 57 (86.4%) patients developed acute encephalopathic syndrome with confusion of consciousness as a depression predominance 36 (54.5%), or psychomotor agitation 24 (36.4%). Focal neurological symptoms appeared at the second week of the disease and were presented by extrapyramidal 6 (9.1%), cerebellar 14 (21.2%) patients, and pyramidal signs with central palsy of limbs 9 (13.7%) and Seizure syndrome 17 (25.8%). Focal neurological deficiency was associated with cerebral edema in 32 (48.5%) cases, serous meningitis 12 (18.2%), serous meningoencephalitis 1 (1.5%) subarachnoid hemorrhage, 1 (1.5%). 68.6% patients developed distal para-infectious polyneuropathy.

Conclusion: When affecting central nervous system, leptospirosis begins with diffuse neurological symptoms, changed by focal neurological deficiency till the second week of the disease end. Depression of consciousness and generation of diffuse and/or focal

neurological symptoms is the objective marker of severe clinical course of ikchetero-hemorrhagic form of leptospirosis.

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Abstract - WCN 2013**No: 557****Topic: 36 - Other Topic****HHV-6 and VZV dual infection**

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Background: In a recent study we found HHV-6 specific IgG oligoclonal bands (OCBs) in many patients with demyelinate symptoms. In a further study we also found VZV specificity. Three patients had both specificities at the same time.

Objective: To describe clinical findings in patients with OCB specificity to both HHV-6 and VZV.

Patients and methods: 79 patients with OCBs in CSF were found when the serum and CSF samples from patients with demyelinate symptoms were analyzed.

OCBs were detected with isoelectric focusing and immunofixation with immunoperoxidase staining. HHV-6 and VZV specific OCBs were detected using affinity driven immunoblot.

Results: Three patients had separately visualized HHV-6 and VZV specific OCBs in the CSF. All were young females under 30 years. All had increased WBC level, IgG index and IgG concentration in the CSF. CSF protein and albumin levels were normal. All fulfilled Barkhof criteria (3–4/4) in MRI of the head. All were diagnosed with MS after CSF analysis.

HHV-6 and VZV specificity in OCB was already present in the initial CSF sample obtained 0–3 months after the episode. All received immunomodulative medication after the MS diagnose. No new episodes appeared during the follow-up time of two years.

Conclusion: HHV-6 and VZV specific OCBs can both be found in patients with MS. The presence of these bands already in the early phase suggests a role for these viruses in the pathogenesis of MS.

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Abstract - WCN 2013**No: 603****Topic: 36 - Other Topic****Isolated cerebral schistosomiasis presenting as seizure in a 32 year old female: A case report**

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Schistosomiasis, or Bilharziasis, is a disease caused by parasitic trematodes. It is considered by the WHO as one of the Neglected Tropical Diseases. In Asia, *Schistosoma japonicum* is more commonly found. Approximately 2.5 million are directly exposed in our country alone. They usually migrate to the gastrointestinal or the genito-urinary veins of the body, to be shed along with the feces and urine, completing their lifecycle outside the body to their corresponding snail hosts. A cerebral manifestation is a rare presentation of the disease. There should be a high index of suspicion, especially in endemic countries, and differentials can include Malaria, Toxoplasmosis or Tuberculoma.

Diagnostics include using the Kato-Katz technique or a biopsy of the infected tissue. We report a case of a 32 year old female, with an isolated cerebral infection due to *Schistosoma* sp, who presented with new-onset seizure, negative diagnostic tests, and necessitated brain tissue biopsy. Patient was subsequently treated with praziquantel 600 mg/tab twice a day.

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Abstract - WCN 2013

No: 552

Topic: 36 - Other Topic

Pseudobulbar affect (PBA) in multiple sclerosis (MS), Parkinson's disease (PD), stroke and amyotrophic lateral sclerosis (ALS)

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Background: PBA is an affective disinhibition group of signs and symptoms characterized by uncontrollable episodes of laughing or crying. Its prevalence in ALS is 34%, in MS 8%, in PD 16% and in stroke 15%.

There is currently no conclusive information about the involvement of the pyramidal or extrapyramidal tract in its pathogenesis.

Objective: to diagnose PBA in ALS, MS, PD and non acute stroke.

Materials and Methods: 10 patients with PD, non acute stroke (more than three months since the diagnosis), MS remission recurrence type and ALS that concurred spontaneously to a specialist follow-up between April and June 2012 were evaluated with the CNS-LS and PLACS PBA scales.

Results: There were 23 men and 17 women. The mean age was 54 (± 13.31) years for ALS, 37 (± 12.14) for MS, 71 (± 16.59) for stroke and 61 (± 9.92) for PD. Sixteen patients were diagnosed of PBA with de CNS-LS (31.2% PD, 25% ALS, 25% Stroke and 18.8% MS). 9 were diagnosed with PLACS (33.3% Stroke, 33.3% ALS, 22.2% PD y 11.1% MS). Of them all, 75% had pyramidal tract dysfunction, 22.5% extrapyramidal and 5% none of the above. Between those diagnosed by CNS-LS, 75% had pyramidal signs and 25% extrapyramidal. With PLACS the percentages were 88.9 and 11.1 respectively.

Conclusion: PD was the disease with most PBA diagnoses with CNS-LS. With PLACS were ALS and Stroke. MS had the lowest PBA diagnoses with both scales. PBA patients had the same frequency of pyramidal and extrapyramidal dysfunction as the general population interviewed.

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Abstract - WCN 2013

No: 567

Topic: 36 - Other Topic

Safety of bevacizumab with or without anticoagulant treatment in neuro-oncological patients: A systematic review

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Background: Neoangiogenesis has recently become a major target for the development of new antineoplastic drugs. The most serious adverse events linked to angiogenesis inhibitors are venous/arterial thromboembolism and hemorrhage. Thus, there is a need to define with more certainty the impact of these new drugs in terms of adverse effects in neurological patients.

Objective: the aim of the study is to assess the risk of venous-thromboembolism (VTE) and bleeding in neurooncological patients

treated with bevacizumab with or without concomitant anticoagulant therapy.

Material and methods: a review of published literature was performed in Medline, from which we identified 476 records. We assessed for eligibility 27 full-text articles including retrospective analyses, retrospective reviews, and open label trials. The investigated drugs included bevacizumab alone, bevacizumab plus chemotherapy with/without concomitant radiation therapy; only two articles dealt with bevacizumab in association with anticoagulant treatment.

Results: a total of 2208 neurooncological patients were identified and included in the analysis. Data confirmed that patients receiving bevacizumab had a major risk of developing VTE that increased when bevacizumab is associated with radio-chemotherapy (4.27% vs. 7.46%). Regarding bleeding, data showed that patients treated with anticoagulant had a significantly increased risk of severe intracranial bleeding compared to patients not receiving anticoagulant therapy (0.6% vs 8.2%).

Conclusion: the use of bevacizumab combined with chemoradiotherapy is associated with a higher risk for VTE compared to patients receiving antiangiogenic therapy alone. The associated use of anticoagulants and bevacizumab far increases the risk of developing intra-extracranial bleeding higher than grade 3, compared to patients receiving bevacizumab alone.

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Abstract - WCN 2013

No: 586

Topic: 36 - Other Topic

Alexander von Humboldt and his medical research

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Background: Medical research in the second half of the 18th c was characterised by a remarkable dynamism in theory construction and a turn towards hard science. For a long time Humboldt's (AvH) contribution to this was overshadowed by his other achievements.

Objective: Inspired by Galvani's publication of Animal Electricity in 1791, AvH conducted numerous neurophysiologic experiments between 1792 and 1797. He published his results in "Experiments on stimulated muscular and nervous tissue" in 1797. How significant were his findings?

Material and methods: In nearly 4000 stimulation tests by means of electrical power he experimented on 3000 different animals and also on himself. Having perused his publication and analysed his ideas, methods and results, we demonstrate his procedures and achievements with the help of examples.

Results: AvH often drew precise and far-reaching conclusions of which an essential one was that there was no stable nerve excitability, but that it depended on numerous factors. However, perplexing observations are also discussed, such as the additive nature of his experiments and the absence of a systematic approach. AvH conducted his experiments with utmost skill strictly following the procedure of Observation – Experiment – Formal application of mathematics – Conclusion, which was by no means a common approach in physiology at the time.

Conclusion: AvH's work must be seen in the context of contemporaneous schools of medical thought. Being influential in a number of areas, his major achievement was that he pioneered the exact scientific research methodology of the 19th century. His work reflects 19th c medicine in an enlightening way.

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Abstract - WCN 2013**No: 789****Topic: 36 - Other Topic****Case of acute ischemic neuropathy, caused by the obliterating atherosclerosis of arteries of the bottom extremities***Y. Sorokin, Department of Neurology, Lugansk State Medical University, Lugansk, Ukraine*

Background: The atherosclerosis of arteries of the legs causes development of the chronic ischemia, however more expressed neurologic disorders are observed in case of sudden termination of blood flow into the extremity.

Material and methods: We surveyed the man of 49 years old who had sudden pains, numbness and weakness in the legs. Neurologic survey has revealed 2-sided neuropathy of fibular and tibial nerves with expressed paresis of foot. Ultrasonic methods and electro-neuromyography are applied to blood flow estimation in the main arteries and a condition of peripheral nerves of the legs.

Results: Duplex scanning has revealed dense and soft circular atherosclerotic plaques in the abdominal aorta, signs of 2-sided occlusion of the general and external iliac arteries, stenosis of 50% of the lumen of both general femoral arteries and considerable decrease in the speed of blood flow in the right deep and left superficial femoral arteries. Operative intervention was made – thrombectomy from distal aorta, endarterectomy from superficial femoral artery of the left hip, bilateral aorto-femoral shunting by a synthetic artificial limb, cross-section sympathectomy of the left side. After the operation the blood flow in the legs and the function of fibular and tibial nerves were practically completely restored. However, electroneuromyography showed the evident nature of axonal affection of fibular and tibial nerves from both sides of the foot area, that reflects depth of damage of nervous structures.

Conclusion: On-time prevention of atherosclerotic affection of the main vessels of extremities is necessary for prevention of ischemic neuropathies and persistent incapacity.

doi:10.1016/j.jns.2013.07.2103

Abstract - WCN 2013**No: 778****Topic: 36 - Other Topic****Investigating the role of poly-(ADP-Ribose)-polymerase as a therapeutic target in pediatric high grade glioma***Y. Chornenkyv^a, S. Agnihotri^b, P. Buczkowicz^a, P. Rakopoulos^a, O. Becher^c, C. Hawkins^a. ^aLaboratory Medicine and Pathobiology, University of Toronto, Canada; ^bArthur and Sonia Labatt Brain Tumour Research Centre, Hospital for Sick Children Research Institute, Toronto, ON, Canada; ^cPreston Robert Tisch Brain Tumor Center, Duke University Medical Center, Durham, NC, USA*

Pediatric supratentorial high-grade astrocytomas (pHGAs) and diffuse intrinsic pontine gliomas (DIPG) are devastating pediatric malignancies for which no effective therapies exist. Poly-(ADP-Ribose)-Polymerase (PARP) protein expression is found in ~60% of DIPGs suggesting PARP may be a potential therapeutic target.

PARP1/2 were characterized by Western-blotting in normal human astrocytes (NHA), pHGA cell lines (SJG2, SF-188), DIPG cell lines (DIPG-M, DIPG58), and one murine brainstem glioma cell line (mBSG). Cell viability in response to different dosages of Olaparib, Veliparib, or Niraparib was determined using the MTT Assay. Levels of PARP activity, apoptosis, and DNA damage were determined by Western blotting against PAR, cleaved PARP, and phosphorylated γH2AX, respectively. Western-blotting demonstrated that, compared with NHAs, PARP1 were highly expressed in SJG2, DIPG-M, and DIPG-58 cells. PARP2 expression was only detected in SJG2 cells. All PARP inhibitors

reduced PARP activity as indicated by reduced PAR levels. Olaparib reduced SJG2, mBSG, DIPG58 and DIPGM cell viability at concentrations of 5 or 10 μM (P < 0.05), Whereas Niraparib induced cytotoxicity at concentrations of 2 μM and above (P < 0.05). Olaparib and Niraparib induced DNA damage and apoptosis in SJG2 at doses of 5, 10 μM and 2, 5, 10 μM, respectively.

Our data provides *in vitro* evidence that PARP inhibition may be an effective therapeutic avenue for treatment of pHGA and DIPG. Furthermore while all PARP inhibitors suppress PARP activity not all PARP inhibitors reduce cell viability. Thus not all PARP inhibitors can be expected to be equally efficacious in a clinical trial setting.

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Abstract - WCN 2013**No: 809****Topic: 36 - Other Topic****Head-shaking aids in diagnosis of acute audiovestibular loss due to anterior inferior cerebellar artery infarction***Y.E. Huh^a, J.-W. Koo^b, H. Lee^c, J.-S. Kim^a. ^aDepartment of Neurology, Seoul National University Bundang Hospital, Seongnam-si, Republic of Korea; ^bDepartment of Otolaryngology, Seoul National University Bundang Hospital, Seongnam-si, Republic of Korea; ^cDepartment of Neurology, Keimyung University School of Medicine, Daegu-si, Republic of Korea*

Background and objective: To determine patterns and diagnostic value of head-shaking nystagmus (HSN) in patients with acute audiovestibular loss.

Patients and method: Eighteen patients underwent evaluation of spontaneous nystagmus, gaze-evoked nystagmus, HSN, head impulse test, ocular tilt reaction, subjective visual vertical, bithermal caloric tests, and pure tone audiogram. The findings were compared with those of 21 patients with labyrinthitis.

Results: Fifteen patients (83%) exhibited HSN, and the horizontal HSN usually beat contralesionally (10/14, 71%). However, 9 (50%) patients also showed patterns of central HSN that included perverted HSN (n = 7), HSN in the opposite direction of spontaneous nystagmus (n = 4), and HSN beating towards unilateral canal paresis or abnormal head impulse testing (n = 3). Overall, central HSN, gaze-evoked nystagmus, and normal head impulse testing were specific for AICA infarction. Moreover, central HSN was the only sign that indicated stroke in one of our patients with isolated audiovestibular syndrome. Lesion subtraction analyses revealed that damage to the flocculus was relatively frequent in patients with perverted HSN.

Conclusions: In AICA infarction, HSN was common with both peripheral and central patterns. Careful evaluation of HSN may provide clues for AICA infarction in patients with acute audiovestibular loss.

doi:10.1016/j.jns.2013.07.2105

Abstract - WCN 2013**No: 810****Topic: 36 - Other Topic****PPAR-γ agonist decreased CA1 hippocampal neuronal density loss on an ethanol induced neurodegenerative model in rats***D. Muñoz-Jiménez^a, G. Espinosa-Villanueva^a, G. Muñoz-Jiménez^b, L.J. Camargo-Pérez^c. ^aLaboratorio de Investigación, Facultad Mexicana de Medicina, Universidad La Salle, Mexico; ^bHospital General Dr. Manuel Gea González, Mexico; ^cBiomedical Research and Development, Aedes Biomedical and Biotechnological Research, Mexico, Mexico*

Background: Cellular waste induced oxidative stress is one of the leading pathways in the physiopathology of neurodegenerative diseases

as in normal degenerative aging process; this neuron decay is observable by the assessment of mean neuronal density of several hippocampal regions which strongly correlates with cognitive impairment symptoms. PPAR- γ agonists (thiazolidinediones), had shown to diminish oxidative stress as they activate oxidative metabolic waste deposition and, therefore, had been used as a novel therapeutic strategy against a wide spectrum of oxidative stress related diseases.

Objective: To assess the possible effects of pioglitazone and rosiglitazone on the mean neuronal density decay on hippocampal regions on an ethanol induced neurodegeneration model.

Methods: Male Wistar Rats received 20% ethanol as unique hydration source while Pioglitazone or Rosiglitazone were administered in powdered food during six months. Thereafter, hippocampal regions were systematically extracted and mounted for optical and electron microphotography. Mean neuronal density was assessed by stereological unbiased software-aided methods.

Results: All 20% ethanol groups showed a significant decrease in CA1 mean neuronal density, whereas the rosiglitazone+ethanol group did show a significant higher neuronal density compared to ethanol-control group. Pioglitazone did not show a significant difference with ethanol-control group.

Conclusion: Rosiglitazone but pioglitazone showed a significant decrease in neuronal density loss suggesting a novel therapeutic strategy for neurogenerative oxidative stress related diseases where hippocampal neuronal loss are relevant for clinical outcome.

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Abstract - WCN 2013

No: 797

Topic: 36 - Other Topic

Central nervous system manifestations of lassa fever in Nigeria and the effect on mortality

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Introduction: Lassa fever (LF) is endemic in Nigeria and causes severe multisystem disease in some infected individuals. The central nervous system (CNS) may be involved in the later stages of the course of the illness resulting in an increase in mortality. There is a paucity of data on the CNS manifestations of LF in Nigeria and the effect on mortality.

Objective: To determine the CNS manifestations of LF and the effect on mortality.

Method: 162 patients (85 males and 77 females) who were RT-PCR confirmed for LF and admitted to our hospital between January 2008 and December 2010 were studied. They were grouped into severe CNS (SCNS), non-severe CNS (NSCNS) and no CNS (NCNS) features, respectively.

Results: 30.9% (50 patients: 24 males; 25 males) had SCNS features (seizure, coma, meningitis, restlessness, ataxia, deafness, disorientation, confusion, tremors, irritability, irrational behaviour); 11.1% (18 patients: 10 males; 8 females) had NSCNS features (miningism, dizziness, vertigo, drowsiness); 58% (94 patients 51 males; 43 females) had NCNS features. Case fatality rates (CFR) were 74% (37/50) for SCNS, 27.8% (5/18) for NSCNS and 13.8% (13/94) for NCNS groups respectively.

($\chi^2 = 50.04, 2df; p < 0.0000$).

Conclusion: A wide spectrum of CNS features was seen in a significant number of our patients. 73.5% (50/68) of patients with CNS features had SCNS manifestations with consequent high CFR.

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Abstract - WCN 2013

No: 710

Topic: 36 - Other Topic

New diagnostic technology in the prediction and evaluation of effective treatment of newborns with hypoxic brain lesions

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Background and aims: In connection with high rate of perinatal brain lesions in newborns and their consequences to the present there are no absolute markers posthypoxic brain lesions.

This has been our aim to develop new methods for predicting, diagnosing, correcting and evaluating the effectiveness of treatment of the effects of neurological disorders.

Methods: A comprehensive examination 169 newborn infants with conducting neurological examination, Doppler ultrasound of the brain, determining the number of neuropeptide S-100 protein and factor neyrotoroficheskogo brain BDNF levels.

It is established that the development and transformation of neurological disorders during postnatal neuroontogenesis occurs in correlation with a change BDNF production and protein S-100, disorders autonomic regulation and motor development.

It is found that the rate of brain-derived neurotrophic factor in serum in children 3 months of age has prognostic significance for the further development of the engine, based on which a new method for predicting violations of motor development.

Results: The study shows that the inclusion of neuropeptide preparation in complex rehabilitation therapy helps to reduce the production of protein S-100, has a positive effect on the autonomic regulation, cerebral hemodynamics, and optimizes the correction of neurological disorders in children.

Such a way a result of the research has developed a system of activities for children in the first year of life with the consequences of perinatal hypoxic brain damage, which includes new technology forecasting, diagnosis, correction and evaluation of the treatment of neurological disorders.

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Abstract - WCN 2013

No: 383

Topic: 36 - Other Topic

Listerial encephalitis with abscess formation and cervical myelitis in an immunocompetent adult

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Background: Brain abscesses and myelitis are very rare manifestations of listerial infection, especially in immunocompetent adults.

Objective: To highlight that early recognition and treatment are crucial to favourable outcome.

Method: Case Study. A 49-year-old otherwise healthy man was admitted to our Neurology department in October 2011 with a history of headache, vomiting and low-grade pyrexia three weeks earlier, followed, 6 days before admission, by right-sided lower motor neurone facial palsy and rapidly progressive left-sided pyramidal limb weakness. Upon arrival, he was alert and apyrexial, with an unremarkable general physical examination.

Results: MRI revealed 2 large, ring-enhancing intracranial lesions, one in the right basal ganglia/corona radiata and the other in the left cerebellar hemisphere, and an extensive cervical cord lesion with oedema. CSF analysis showed WBC < 1, elevated protein of 1.88 g/l and normal glucose, with negative Gram stain, AFB stain, culture, viral PCR study, cryptococcal antigen and oligoclonal bands. ESR and CRP were moderately elevated for the first 2 days only. Whole body CT scan and extensive haematological and immunological blood tests

were all normal. The first out of 3 blood cultures grew *Listeria monocytogenes*. The patient was treated with IV gentamycin for 2 weeks and a 6-week course of high dose IV amoxicillin and cotrimoxazole. He made steady progress, being left with mild left hemiparesis, and five-week interval MRI confirmed considerable improvement.

Conclusion: Listerial infection should be considered in the differential diagnosis of acute myelitis and/or encephalitis with abscess formation, even in immunocompetent patients.

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Abstract - WCN 2013

No: 738

Topic: 36 - Other Topic

Viral meningitis triggers anti-NMDA receptor encephalitis

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Background: Anti-NMDA receptor encephalitis is recognized increasingly in the past few years. Like other autoimmune diseases, environmental factors such as infection can precipitate autoimmune disease.

Objective: To review clinical course of the patient who had viral meningitis with subsequently developed anti-NMDA receptor encephalitis.

Patients and methods: A case report of twenty-seven year old priest who was admitted at Prasat Neurological Institute during 12th November to 14th December 2012.

Results: A 27 year-old priest developed fever and nuchal rigidity for 2 weeks before arrival. Neurological examination showed neck stiffness, but fully consciousness. CSF analysis demonstrated pleocytosis 380 cells (lymphocytic predominate) with slightly elevated protein, but normal glucose. PCR for herpes was positive for Human herpesvirus type 7. During the course of intravenous acyclovir treatment, he was improved. But later on he developed acute psychosis, body stiffening with limb dystonia, dysautonomia and altered mental status respectively. MRI brain revealed abnormal signal at left posterior thalamus and left amygdala. EEG showed moderately severe diffuse encephalopathy. NMDA-receptor antibody was detected in CSF. The intravenous methylprednisolone and plasma exchange were started concurrently. Oral prednisolone and symptomatic treatment were continued. He was discharged after one month and his clinical full recovery after 1 month follow up.

Conclusion: Human herpesvirus type 7 caused viral meningitis and trigger anti-NMDA receptor encephalitis.

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Abstract - WCN 2013

No: 747

Topic: 36 - Other Topic

Oculographic analysis of nystagmus in orthostatic vertigo

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Background and objective: To elucidate objective vestibular dysfunction and the mechanisms of orthostatic vertigo.

Materials and methods: Thirty-three consecutive patients with orthostatic dizziness/vertigo and profound orthostatic hypotension (OH) were recruited. Profound OH was defined that systolic BP falls of ≥ 30 mm Hg occurs within 3 min after standing and is sustained for at least 2 min during head-up tilt test. Eye movements were recorded during two orthostatic challenging test, the Schellong and Squatting–standing test.

Results: All patients developed vertigo during orthostatic challenging test. Associated symptoms included blurred vision (45%), fainting (42%), and tinnitus (15%). Ten of 33 (30%) patients developed nystagmus during orthostatic challenging test with a latency of several seconds. Five of them showed mixed downbeat and horizontal nystagmus with or without torsional component, while the remaining four had pure downbeat nystagmus. In one patient, pure horizontal nystagmus and prominent downbeat nystagmus mixed with horizontal and torsional component appeared alternatively at a few second intervals. There were no significant differences in the mean drop of the BP between patients with or without nystagmus, but the patients with nystagmus had less longer duration of orthostatic vertigo than in patients without nystagmus (1.1 ± 1.6 vs 11.0 ± 9.7 min, $p < 0.001$).

Conclusions: The present study demonstrates that OH can give rise to true vestibular vertigo, which may be due to transient ischemia of the peripheral labyrinth or the central cerebellum based on the objective eye movement recording. The nystagmus in longstanding OH might have disappeared due to central adaptation by repetitive orthostatic vertigo.

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Abstract - WCN 2013

No: 741

Topic: 36 - Other Topic

A novel KRAS gene mutation of Noonan syndrome with severe peripheral nerve hypertrophy

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We reported a female case of Noonan syndrome (NS) with the significant peripheral nerve hypertrophy. Because her gait disturbance gradually deteriorated and the nerve conduction velocity was obviously reduced, we firstly thought she had Charcot–Marie–Tooth disease. But, she did not have the mutation of *PMP22* gene. The NS with peripheral neuropathy was comparatively reported as the merger with neurofibromatosis (NF). Cafe au lait spots and skin tumors, which are characteristic in NF cases, were not seen in our case.

The RAS/MAPK syndrome including NS, a Cardio-Facio-Cutaneous (CFC) syndrome, Costello syndrome and so on is proposed as a comprehensive concept of the congenital malformation diseases by the molecular abnormality of RAS/MAPK pathway in recent years. A novel *KRAS* gene mutation (c.211T>G, p.Y71D) was identified in our case. There was a previous report the *KRAS* gene mutation (c.211T>C, p.Y71H) in CFC syndrome case with peripheral nerve disorder.

We think that the *KRAS* mutation “p.Y71D”, “p.Y71H” may be related to peripheral nerve disorder closely. In addition, the peripheral nerve disorder is the phenotype of NS, CFC syndrome, and furthermore RAS/MAPK syndrome.

doi:10.1016/j.jns.2013.07.2112

Abstract – WCN 2013**No: 943****Topic: 36 – Other topics****Transient global aphasia after cerebral arteriography: A possible reversible contrast medium neurotoxicity mimicking stroke**Y. Handok, S. Biguzzi, C. Bompreszi, S. Morresi, M.G. Passarin.
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Background: Cerebral arteriography (CA) is the gold standard in the diagnosis of many cerebral lesions. However it is an invasive procedure with potential complications. Transient neurologic deficits presumably caused by temporary disruption of the blood–brain barrier (BBB) due to nonionic radiographic contrast medium (CM) have been reported in literature.

Case report: A 69-year-old woman with a previous history of subarachnoid haemorrhage following basilar aneurysm rupture, underwent coiling in May 2010, was admitted in September 2011 in our unit for a routine CA. During CA she developed transient global aphasia with a complete recovery in a few minutes. Three hours after CA she presented aphasia again with total loss of the ability to speech and severe impairment of comprehension. An urgent magnetic resonance imaging (MRI) was performed. MRI diffusion did not reveal acute infarction and MRI angiography did not show any stenosis, spasm or occlusion at the cerebral vessels. The electroencephalogram (EEG) demonstrated a slow nonpileptiform activity in the left derivations. The day after another brain MRI was also negative. Neurologic signs and EEG abnormalities improved completely after 3 days.

Discussion: CM is reported to disrupt the BBB temporarily causing an encephalopathy that is usually self-limiting. This case report is atypical because no neuroradiological abnormalities due to a CM encephalopathy were found.

Conclusion: CM injection could be the possible causative factor of transient neurologic deterioration, despite the fact that the mechanism still remains speculative. EEG examination was more sensitive than MRI in detecting the brain injury in these patients.

doi:10.1016/j.jns.2013.07.2113

Abstract – WCN 2013**No: 733****Topic: 36 – Other topics****Structural integrity of medial temporal lobes of patients with acute mild traumatic brain injury**T.M. Luoto^a, K. Holli-Helenius^b, A. Brander^b, M. Wäljas^a, G.L. Iverson^c, J. Öhman^a. ^aDepartment of Neurosciences and Rehabilitation, Tampere University Hospital, Tampere, Finland; ^bMedical Imaging Centre and Hospital Pharmacy, Department of Radiology, Tampere University Hospital, Tampere, Finland; ^cDepartment of Physical Medicine and Rehabilitation, Harvard Medical School and Red Sox Foundation and Massachusetts General Hospital Home Base Program, Boston, MA, USA

Background: The duration of post-traumatic amnesia (PTA) is commonly used to estimate the severity of brain injury. Lesions in temporal lobes, especially hippocampal regions, are thought to be connected to memory loss. However, conventional neuroimaging has not revealed neuropathological correlates of PTA in MTBI. Texture analysis (TA) is an image analysis technique that quantifies the minor MRI signal changes among image pixels and thus the variations in intensity patterns within the image.

Objective: The objective was to apply the TA technique to MR images of MTBI patients and control subjects and to assess the microstructural damage in medial temporal lobes of patients with MTBI with definite PTA.

Patients and methods: TA was performed for FLAIR images of 50 MTBI patients and 50 age- and gender-matched controls in the

regions of amygdala, hippocampus, and thalamus. It was hypothesized that

- (i) there would be statistically significant differences in TA parameters between patients with MTBIs and controls, and
- (ii) the duration of PTA would be related to TA parameters in patients with MTBI.

Results: No significant textural differences were observed between patients and controls in the regions of interest. No textural features were observed to correlate with the duration of PTA. Subgroup analyses were conducted on patients with PTA > 1 h (n = 33) and compared the four TA parameters to the controls (n = 33). The findings were similar.

Conclusion: This study did not reveal significant textural changes in medial temporal structures that could be related to the duration of PTA.

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Abstract – WCN 2013**No: 903****Topic: 36 – Other topics****Immunological profiles of the CSF from anti-NMDA receptor encephalitis patients**K. Tanaka^a, M. Tanaka^b, M. Fujita^a, M. Matsui^a, T. Takegami^c. ^aNeurology, Kanazawa Medical University, Uchinada-Machi, Japan; ^bNeurology, Utano National Hospital, Kyoto, Japan; ^cLife Science, Medical Research Institute, Kanazawa Medical University, Uchinada-Machi, Japan

Background: Anti-N-methyl-D-aspartate receptor autoantibodies (NMDAR-Abs) are thought to relate closely to the pathogenesis of NMDAR encephalitis. The antibody-titre parallels with the disease activities and antibody-deprivation treatments are effective. This disease is thought to be immune-mediated; however, studies have not been done extensively from the immunological aspects.

Objective: To study the immunological features of the patients with NMDAR encephalitis.

Design/methods:

- 1) Chronological changes of lymphocyte populations in the peripheral blood (PB) and CSF were studied with flow cytometry.
- 2) Cytokine concentrations in the CSF and sera were analysed with the cytokine antibody arrays.
- 3) The antibody-effects on the cells expressing NMDARs were studied under an incubation microscope.

Results:

- 1) In the early stage, CD4⁺CD29⁺ T/CD4⁺CD45RO⁺ T cells were increased in the patients' CSF but not in the PB that gradually decreased parallel to the recovery.
- 2) There was no significant difference in each cytokine level between the patients' CSF and the control CSF.
- 3) The NMDAR-expressed HEK cells that were incubated with patients' CSF showed a fragmented staining pattern of NMDAR clusters inside the cells, while the cells incubated with the control CSF showed cell surface staining.

Conclusions: The lymphocyte-population profiles in the patients' CSF suggested the active antibody production in the central nervous system without inflammatory cytokine production. The patients' CSF stimulated endocytosis of the NMDARs suggesting protective effects of the antibodies for cells damaged from the extreme glutamate stimulation that resulted for the good outcome of this disease.

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Abstract – WCN 2013**No: 907****Topic: 36 – Other topics****Compressed sensing technology could accelerate MRI at 7 T**

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Background: Compressed sensing is an innovative theory of signal acquisition and processing based on the areas of applied mathematics. It works by using the mathematical algorithm to make an appropriate domain transformation for the collected signals and changing them into sparse or compressible signals. Afterwards, gathering the compressed signals directly to reconstruct the original signals at speedy, fine quality by the method of reconstruction algorithm. Due to its excellent temporal resolution advantages and satisfactory temporal resolution, compressed sensing has become a research hotspot in the field of medical imaging.

Objective: Based on the compressed sensing image reconstruction algorithm, we aim to use less data to obtain high quality reconstructed images which is good to accelerate medical imaging speed and reduce the radiation.

Material and methods: We firstly make a sparse representation of the image scanned at 7 T via a multiscale wavelet transform by using MATLAB. After being compressed, the retained energy is 99.29%, and the number of zeros is 93.73%. That is, by wavelet transformation, most coefficients are small though nearly all the image pixels have nonzero values. Then we use the histogram method of the original image and synthesized image and make sure that the step of compression is significant. Finally, we reconstruct the image by the way of sequential minimal optimization.

Conclusion: This method will be significant to achieve rapid high quality imaging in MRI at 7 T and it can reduce the cost during processing, storage and transmission.

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Abstract – WCN 2013**No: 888****Topic: 36 – Other topics****Blink reflex excitability in patients with fibromyalgia syndrome**

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Objective: We aimed to investigate the blink reflex excitability in fibromyalgia syndrome.

Subjects and methods: A total of 23 women with FMS and 14 healthy female controls were studied. Blink reflexes were elicited by electrical stimulation of the supra-orbital nerve. The stimuli were applied at first single stimulus and then paired stimulus by using ISIs by 100–600 ms. Electromyography signals were recorded from the right orbicularis oculi muscle.

Results: There are significant differences between FMS and controls in responses obtained by paired stimulus at 200–400 ms. Responses of R2 obtained by ISI at 100–400 ms were smaller than those obtained by single stimulus in controls. This inhibition disappeared in response to paired stimulus by 500, 600 ISI in controls. Yet, in FMS patients, this expected inhibition only appeared for iR2 and cR2 obtained at 100 ms ISI and then did not appear at the paired stimulus at 200–400 ms ISIs.

Conclusions: Lack of BR R2 response inhibition to paired stimulus by ISIs might be an electrophysiologic evidence that indicates central sensitization in FMS.

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Abstract – WCN 2013**No: 881****Topic: 36 – Other topics****Semantic abilities in children with dyslexia**

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The goal of this research was to deduct the frequency of dyslexia and achievement of children with dyslexia on the Test of Short-term Verbal Memory, Test of Dominant Lateralization, Triage Articulation Test, Semantic Test, and Test of Voice Analysis and Synthesis.

Methodology: From a total of 280 children which were tested at the end of the second, third and fourth grade, with the analysis of handwriting (dictation, explanation of what was happening, transcription), 38 children (13.57%) were singled out with dysgraphia, from which 57.9% with dyslexia and 42.1% with motor dysgraphia. They are all tested with the Tridimensional Reading Test (Helen Sax), and for this sample it singled out 25 (8.9%) children with dyslexia. From the total number of dyslexic children, 88% have dyslexic dysgraphia, and the remaining 12% have graphomotor problems. Children with intellectual dispairment, bilingual children, as also socio-educationally ignored children, are excluded from the research.

Results: The best results were shown on the test of metonyms wherein 80% of children gave all the answers, and 20% were partially successful, which means, there were no unsuccessful children. There were no incompletely unsuccessful children on the test of homonyms, but 60% of children have given just 2 of the expected 4 answers. On the test of antonyms there were 44% unsuccessful children. The test of synonyms has especially bad results, because 48% of children didn't give any answer, and 24% of children have given just one answer, meaning, only 28% have done well in this part of the test.

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Abstract – WCN 2013**No: 873****Topic: 36 – Other topics****Determination of expression pattern of big-H3 gene in the process of regeneration in a model of zebrafish eye cryoinjury**

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Introduction: Humans have a rather limited capacity to regenerate after injury, while other vertebrates such as the zebrafish are capable of regenerating many anatomical structures. One of them is transforming growth factor beta induced regeneration (initially called BIGH3, BIG-H3), which signals have emerged as one of the components of the inductive process. Our investigation focused on the role of BIGH3 in the process of retinal regeneration in zebrafish after cryoinjury, wherein it was important to establish its pattern of expression.

Materials and methods: ABTL wild type zebrafish strains of both sexes 10–14 months of age were bred in standard fish facility conditions. In total, 45 zebrafish were used for the investigation of the process of regeneration in the retina. 40 of them were subjected to cryoinjury. Postinjury measurements were grouped into the following periods: 1 day postinjury (dpi), 2 dpi, 3 dpi, 5 dpi, 7 dpi, 15 dpi, 21 dpi and 28 dpi. RNA from retina was obtained by using the TRIZOL reagent. RNA concentrations and purity were measured using UV-spectrophotometry. Equal amounts of RNA were reverse transcribed into cDNA using StrataScript reverse transcriptase. Quantitative RT-PCR was performed for each sample in duplicate, using β -actin primers as control.

Results: Pronounced upregulation of the expression was observed at 5 dpi. At 7 dpi, BIGH3 expression reached the highest point as

compared to other days. At 21 dpi, expression declined to the baseline level.

Conclusion: The data on the performed research indicated that BIGH3 is an important protein in retinal regeneration.

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Abstract – WCN 2013

No: 834

Topic: 36 – Other topics

Sleep disturbances and risk of arterial hypertension and stroke in the 25–64 year old female population in Russia: WHO program “Monica–Psychosocial” study

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Objective: To explore the influence of sleep disturbances (SDs) on relative risk of arterial hypertension (AH) and stroke in the 25–64 year old female population in Russia over 16 years of follow-up.

Material and methods: Under the third screening of the WHO “MONICA–psychosocial” program random representative samples of women aged 25–64 years (n = 870) were surveyed in Novosibirsk. The “Awareness and Attitude Towards the Health” questionnaire was used to estimate quality of sleep. From 1995 to 2010 women were followed for incidence of AH, stroke. Cox regression model was used for relative risk assessment (HR) of stroke, AH.

Results: The prevalence of SD in women aged 25–64 years was 64.9%. AH was developed in 33.4% of women, and stroke in 5.1% of women. HR of AH in women with SD was 4.35-fold higher (95.0% CI:1.29–14.59; p < 0.05) and it was 2.69-fold higher (95.0% CI:1.01–7.15; p < 0.05) compared to those with good sleep for the first 5 and 10 years of follow-up, respectively. HR of stroke in women with SD was 1.95-fold higher (95.0% CI:1.01–3.79; p < 0.05) than in those without SD for over 16-years of study. Rates of AH incidence were significantly higher in middle and first-line managers and physical labor workers with SD (p < 0.05). There were tendencies of growth of stroke rates in married women with SD having college degrees.

Conclusions: The prevalence of SD in women aged 25–64 years is more than 60%. Women with SD had significantly higher relative risk of AH and stroke. Rates of AH development were more likely in managers and physical laborers.

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Abstract – WCN 2013

No: 54

Topic: 36 – Other topics

Callosal regulation of contrast gain control machinery in the human visual system

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Background: Visual cortical areas in the two hemispheres interact via the corpus callosum, but the precise role of the callosal pathway in visual processing remains controversial. As emerged from animal studies, the callosal pathway appears to transfer preferentially

information related to high contrasts and low temporal/spatial frequencies.

Objective: Here, we have investigated the function of transcallosal projections in the human primary visual cortex (V1).

Materials and methods: Visual evoked potentials (VEPs) triggered by grating stimuli of different contrasts were recorded before and after functional inactivation of the occipital cortex of one hemisphere via off-line low-frequency repetitive transcranial magnetic stimulation (rTMS; 0.5 Hz stimulation for 20 min). VEPs were recorded in V1 before (T₀), immediately after (T₁) and 45' following the completion of rTMS (T₂).

Results: Low-frequency rTMS has an inhibitory effect on VEP amplitudes at all contrasts in the treated side (p < 0.05). Remarkably, reduction of VEP amplitudes in the inhibited hemisphere at T1 was accompanied by an increase in VEP amplitudes in the contralateral side only at mid-high contrasts (50–90%: two-way repeated-measures ANOVA, p < 0.01). This disinhibitory effect was observed with both central and hemifield stimulation (p < 0.01). No changes in VEP amplitudes were observed when rTMS was applied to a cortical site more anterior with respect to V1 (p > 0.05).

Conclusions: These data provide the first evidence that a mechanism of transcallosal inhibition dampens neural responses at high contrasts in the human visual cortex.

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Abstract – WCN 2013

No: 706

Topic: 36 – Other topics

Magnetic resonance imaging findings in a case of vertebral arteriovenous fistulas in a patient of neurofibromatosis type one

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Background: Vascular abnormalities associated with neurofibromatosis type one (NF1) include occlusive or stenotic diseases, aneurysms, arteriovenous malformations (AVMs) and, rarely, arteriovenous fistulas (AVFs). We report a case of vertebral arteriovenous fistulas (VAVFs) presenting with neurological disturbance in a patient with known NF1 and review the literature.

Case report: A 30-year-old woman with a history of NF1 presented with right hand weakness 2 weeks after a shoulder trauma. There was initial bilateral hand numbness and neck pain radiating to the right forearm with subsequent weakness of right hand grasp. MRI of cervical spine revealed large flow voids, representing a right VAVF. Endovascular treatment using coils resulted in successful occlusion of fistula.

Conclusions: In VAVFs associated with NF-1, there are two possible mechanisms by which a VAVF may arise. First, dysplastic smooth muscle or neurofibromatosis proliferation in the vessel wall may lead to rupture into adjacent veins. The second possibility would be that the AVF arises congenitally as a manifestation of mesodermal dysplasia. In our patient, it is possible that the fistula and adjacent veins were initially small, but enlarged secondary to tissue disruption with mild trauma, leading to expansion and compression of the nerves. Diagnosis of spinal VAVF can be established by MR imaging, angiography or CT. Confusion of the VAVF with a neurofibroma may occur on MRI because of high vascularity of some neurofibromas and similar presentation as dumbbell-shaped mass with bone scalloping. However, a flow-related signal void usually indicates the greater likelihood of the VAVF.

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Abstract – WCN 2013**No: 479****Topic: 36 – Other topics****Rabies virus infection is associated with mitochondrial dysfunction**

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Studies in an experimental model of rabies have shown major structural changes in the brain involving neuronal processes that are associated with severe clinical disease. Cultured adult mouse dorsal root ganglion (DRG) neurons are permissive to infection with the CVS strain of rabies virus (RABV). RABV-infected DRG neurons show axonal swellings and 4-hydroxy-2-nonenal protein adduct immunostaining, indicating evidence of oxidative stress, and also reduced axonal growth versus mock-infected DRG neurons. We hypothesized that RABV infection induces mitochondrial dysfunction leading to oxidative stress. We investigated the effects of RABV infection on mitochondrial parameters. In RABV infection there were increased activities of electron transport chain Complexes I and IV versus mock infection, whereas Complexes II–III, citrate synthase, and malate dehydrogenase activities were unchanged. Increases in Complex I activity, but not Complex IV activity, correlated with cellular susceptibility to RABV infection. Mitochondrial respiration studies showed that RABV infection increased maximal uncoupled respiration and Complex IV respiration, whereas coupled respiration and the rate of proton leak were unchanged. A high mitochondrial membrane potential was generated. RABV infection reduced the intracellular ATP level and altered the cellular redox state with a high NADH/NAD⁺ ratio. In RABV-infected neurons, basal production of reactive oxygen species (ROS) was unaffected, but a higher rate of ROS generation occurred with mitochondrial substrates and inhibitors. We conclude that RABV infection induces mitochondrial dysfunction leading to ROS overgeneration and oxidative stress. We postulate that the fundamental abnormality is due to the interaction of the RABV phosphoprotein with Complex I leading to its increased activity.

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Abstract – WCN 2013**No: 1029****Topic: 36 – Other topics****Epidemiology of tuberculosis of central nervous system in patients with HIV-infection in the Irkutsk Region of Russia**

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Background: Increasing prevalence of tuberculosis infection in the Irkutsk Region of Russia remains one of the most common problems. **Objective:** This study presents results of epidemiological investigation devoted to the comorbidity of tuberculosis and HIV-infection in the Irkutsk Region.

Patients and methods: 240 patients (184 males and 56 females) aged from 20 to 56 have been observed.

1. Group of patients with tuberculosis of central nervous system and HIV-infection (196 patients).

2. Group of patients with tuberculosis of central nervous system without HIV-infection (44 patients).

Results:

1st group:

Clinical forms of tuberculosis of the central nervous system: meningitis – 124 (63.3 ± 3.4%); meningo-encephalitis – 66 (33.7 ± 3.4%); meningo-myelitis – 1 (0.5% ± 0.5%); meningo-encephalo-myelitis – 4 (2.0 ± 1.0%); tuberculoma of the brain – 1 (0.5% ± 0.5%). HIV-associated diseases (n = 181): candidosis of the gastro-intestinal system – 178 (90.8%), pneumocystic pneumonia – 3 (1.5%). Social characteristics: drug abused (injecting drug users) – 158 (80.6 ± 2.8%), alcohol abused – 14 (7.1 ± 1.8%).

2nd group:

Clinical forms of tuberculosis of central nervous system: meningitis – 38 (86.4 ± 5.0%); meningo-encephalitis – 2 (4.6 ± 3.0%); meningo-myelitis – 3 (6.8% ± 4.0%); meningo-encephalo-myelitis – 1 (2.3 ± 2.0%); tuberculoma of the brain – 0. Social characteristics: drug abused (injecting drug users) – 3 (6.8 ± 4.0%), alcohol abused – 7 (16.0 ± 3.0%).

Conclusion: Obtained data show severe epidemiological situation of tuberculosis in patients with HIV-infection in the Irkutsk Region of Russia. Frequent combination of central nervous system tuberculosis and HIV-infection was revealed. There was comorbidity with meningo-encephalitis with generalized tuberculosis infection combined with candidosis, and hepatitis in drug abused patients with HIV-infection.

doi:10.1016/j.jns.2013.07.2124

Abstract – WCN 2013**No: 1023****Topic: 36 – Other topics****The value of preoperative fMRI in drawing attention to possible unexpected language area representations in a patient with anaplastic astrocytoma**

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This is a case study of a right handed patient with a grade III anaplastic astrocytoma in the superior frontal cortex with intra-axial extensions to the corpus callosum. Presurgical functional MR imaging (fMRI) was done to visualize language network distribution. Intraoperative direct cortical stimulation (DCS) was accordingly done to prevent eloquent functional regions from resection. fMRI results showed a distributed language network including the left inferior frontal gyrus (pars opercularis), the left and right frontal orbital cortices, the left insular cortex, the left superior and middle temporal gyri, the left precentral and postcentral gyri, and the supplementary area. Additionally, statistically stable but unusual activation was found in the left superior frontal gyrus (SFG). During surgery, DCS demonstrated that the stimulation of the SFG – involved in language function according to the fMRI – led to anomie disturbances. These deficits in word finding were persistently reproducible when stimulating the same area. Afterwards, tumor removal was performed under navigation and fluorescence guidance as well as subcortical electrostimulation, thus preserving eloquent motor and language areas and tracts. Regarding the results of the fMRI and DCS in this patient, it can be speculated that the altered distribution of eloquent language areas could be the result of

tumor-induced reorganization processes. This case study demonstrates that presurgical fMRI was able to identify a localized unusual language network distribution, which was validated with direct cortical mapping during awake surgery. It underlines the importance of careful presurgical planning, keeping in mind the limitations of all used methods.

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Abstract – WCN 2013

No: 1096

Topic: 36 – Other topics

The role of Omega-3 fatty acids in patients with mild cognitive impairment by means of the PHOTOTEST

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Background: Omega-3 fatty acids have been shown to improve cognitive function in patients with cognitive impairment. The PHOTOTEST is a short cognitive test indicated for the detection and monitoring of patients with cognitive impairment. It is applicable to illiterate patients and it produces results that are not influenced by educational level.

Objective: To assess, by means of the PHOTOTEST, the benefit of an Omega-3 food supplement in patients with mild cognitive impairment.

Patients and method: A total of 30 patients diagnosed with mild cognitive impairment were randomly divided into two groups: no Omega-3 group with normal treatment; and Omega-3 group with normal treatment plus the Omega-3 food supplement (2 ACUTIL capsules per day: 500 mg DHA + 80 EPA per day). Assessment of cognitive function was done by means of the PHOTOTEST at visit 0 (day 0), visit 1 (3 months) and visit 2 (6 months).

Results: 26 patients (mean age of 72.5 years) completed the study: 13 in the no Omega-3 group and 13 in the Omega-3 group. PHOTOTEST results:

- Positive but not significant trend ($p = 0.15$) in the Omega-3 group at 3 months.
- Significant difference ($p = 0.003$) between the groups at 6 months in favour of the Omega-3 group.

Conclusion: After 6 months with the Omega-3 supplement, the PHOTOTEST results show a significant cognitive improvement in the group that received the Omega-3 supplement compared to the group not receiving any supplement. Omega-3 tolerance was good.

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Abstract – WCN 2013

No: 1067

Topic: 36 – Other topics

Autosomal dominant cerebellar ataxias: A systematic review of clinical features

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Background: Precise frequency and distinction of diverse clinical features of spinocerebellar ataxias (SCAs) are lacking.

Objective: To assess distinctive or common clinical signs of autosomal dominant cerebellar ataxias (ADCAs), genetically known as SCAs, through a systematic review.

Methods: A structured search was conducted by two independent reviewers in electronic databases up to September 2012. Publications containing proportions or descriptions of ADCA clinical features and written in several languages were included. Gray literature was also included and retrieved publication reference lists were back-searched. Clinical findings and demographic data from genetically confirmed patients were extracted. Data was analyzed by Chi-square test controlling for alpha-error inflation by Holm's step-down procedure.

Results: One thousand sixty-two publications containing 12,141 patients (52% male) from 29 SCAs were analyzed. Mean age at onset was 35 ± 11 years. Onset symptoms in 3945 patients revealed gait ataxia as the most frequent sign (68%), whereas overall non-ataxia symptom frequency was 50%. Some ADCAs began frequently with non-ataxia symptoms, like SCA7 (visual impairment), SCA14 (myoclonus), SCA13 (developmental delay), and SCA17 (parkinsonism, psychiatric alterations and cognitive impairment). During the overall disease course, dysarthria (90%) and saccadic eye movement alterations (69%) were the most prevalent non-ataxia findings. Some ADCAs were clinically restricted to cerebellar dysfunction and others had frequently other features, allowing categorization into two groups: pure and mixed.

Conclusions: ADCAs encompass a broad spectrum of clinical features with an elevated frequency of non-ataxia symptoms. Some features distinguished several genetic subtypes from each other. An updated Harding's classification is proposed.

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Abstract – WCN 2013

No: 1075

Topic: 36 – Other topics

Characteristics of water channel protein aquaporin expression in Parkinson's disease

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We have previously reported marked changes in the expression of aquaporin 1 (AQP1) and AQP4 in relationship to amyloid β peptide ($A\beta$) deposition in human Alzheimer disease (AD) brains (Hoshi A et al, JNEN, 2012). In the present study, we investigated the AQP expression in autopsied brains affected by Parkinson's disease (PD). Expression of α -synuclein (α -syn), GFAP, AQP1, and AQP4 in the temporal lobes of 11 patients with PD, 5 patients with AD, and 5 age-matched controls was examined immunohistochemically. In accordance with the DLB consensus criteria, the PD patients were classified to neocortical ($n = 4$), limbic (5), and brainstem (2) groups. In the neocortical group, AQP4 immunoreactivity appeared more intense and the number of AQP1-positive astrocytes was larger than those observed in the other groups except AD group. On the other hand, in the limbic/brainstem groups both AQP1 and AQP4 expressions were not changed in comparison with those of the controls. In the neocortical group, there were negative correlations between expression levels of AQP1 and α -syn, and also between those of AQP1 and $A\beta_{42}$. Thus, in PD brains AQP expression seems altered in association with pathological processes involving the deposition of α -syn and $A\beta$.

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Abstract – WCN 2013**No: 1063****Topic: 36 – Other topics****Clinical and physiologic evaluation of breathing patterns of premature children with neurological complications of HIV infection**

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Newborn respiratory distress-syndrome is the main factor determining the severity of very preterm infants.

The aim: To investigate and estimate the breathing pattern of premature infants with neurological complications of HIV infection in the Aral Sea region of Uzbekistan.

Materials and methods: The project included 27 infants with a gestational age of 27–35 weeks. All of the children were subjected to clinical examination with assessment of imaging studies, showing indicators during bronchography. Data were processed with program “Statistics-6”.

Results: In 21 (74%) cases a complicated course of newborn respiratory distress-syndrome was observed. Development of broncho-pulmonary dysplasia was observed in 7 children, pneumonia in 13, and ventilation was applied gently to 17 children in the first hours after birth, and seizures of apnea were detected in 11 premature children. During auscultation there is broken crepitus, moreover a weakened irregular breathing. While comparing the signs shown by the broncho-phonogram we identified functional abnormalities in the lungs and obtained a regularity of increasing indexes of acoustic breathing equivalent (integral characteristic of energy costs by the broncho-pulmonary system to excite the acoustic signal).

The study of children with broncho-pulmonary dysplasia revealed the presence of airflow obstruction impaired by restrictive type because of the formation of lung parenchyma fibrosis. While assessing, breathing patterns showed statistically significant changes of broncho-phonogram indicators. Retention of changes on a broncho-phonogram on high spectrum indicates the presence of latent bronchial obstruction and requires basic therapy.

Conclusions: The use of modern diagnostic approaches can develop algorithms to optimize the research and basic therapy.

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Abstract – WCN 2013**No: 994****Topic: 36 – Other topics****Acute mild traumatic brain injury is not associated with white matter change on whole brain diffusion tensor imaging**

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Background: Mild traumatic brain injury (MTBI) is associated with microstructural changes in white matter assessed with diffusion tensor imaging (DTI). However, methodological limitations, such as small sample sizes and failure to control for pre-injury morbidity, can confound the results.

Objectives: To determine if MTBI is associated with microstructural changes in white matter in a large, homogenous, sample that was carefully screened for pre-injury medical, psychiatric, or neurological problems.

Patients and methods: Participants were 75 patients with MTBI and 40 controls. Nine exclusion criteria were used to rule out pre-existing medical conditions or other confounding factors. The WHO criteria for MTBI were used. Whole-brain DTI (3T-MRI, mean imaging time post-injury 5.8 days, IQR 4.1–7.3) analysis was done with tract-based spatial statistics (TBSS) and the DTI parameters included: (i) fractional anisotropy (FA), (ii) apparent diffusion coefficient (ADC), (iii) radial diffusivity (RD) and, (iv) axial diffusivity (AD).

Results: All MTBI patients and controls were compared using age and gender as covariates and in age- and gender-matched subgroups (n = 40 versus n = 40). None of the DTI parameters (FA, ADC, RD, or AD) showed significant differences between patients and controls (p > 0.01). A subgroup of MTBI patients with more serious injuries by conventional severity criteria (at least one of the following: LOC > 1 min, PTA > 3 h, traumatic lesion on conventional MRI, GCS = 14, total subgroup n = 28) was compared to age- and gender-matched controls. No significant DTI abnormalities were detected in this subgroup (p > 0.01).

Conclusion: In this large homogeneous, premorbidly healthy sample, MTBI was not associated with DTI abnormalities detectable with TBSS.

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Abstract – WCN 2013**No: 997****Topic: 36 – Other topic****Cardiovascular and metabolic complications of spinal cord injury: Findings from a national population health study**

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Background: Metabolic and cardiovascular complications are a growing concern among individuals with spinal cord injury (SCI).

Objective: The objective of this study was to evaluate the association between Type 2 diabetes and SCI, as well as cardiovascular disease (CVD) and SCI, in a large, representative sample.

Patients and methods: Data were obtained on 60,678 respondents to the Statistics Canada 2010 Cycle of the cross-sectional Canadian Community Health Survey (CCHS). Multivariable logistic regression, incorporating adjustment for confounders and probability weights to account for the CCHS sampling method, was conducted to quantify this association.

Results: After adjustment for both sex and age, SCI was associated with a significant increased odds of Type 2 diabetes (Adjusted odds ratio = 1.66, 95% Confidence Interval [1.16, 2.36]), a significant increased odds of heart disease (Adjusted odds ratio = 2.72, 95% Confidence Interval [1.94, 3.82]), and a significant increased odds of stroke (Adjusted OR = 3.72, 95% Confidence Interval [2.22, 6.23]).

Conclusion: These heightened odds highlight the need for future cohort or case-control studies examining a causal relationship between SCI and these secondary complications, which may ultimately result in treatment and prevention strategies targeted towards individuals with SCI.

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Abstract – WCN 2013**No: 652****Topic: 36 – Other topic****Another cause of reversible splenial lesion:****Hepatic encephalopathy**

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Background and purpose: Reversible splenial lesion has been reported to be associated with encephalitis due to different pathogens, mostly viruses, methotrexate toxicity, systemic lupus erythematosus, reversible vasoconstriction syndrome and migraine. But it has not been reported yet under imaging findings of hepatic encephalopathy.

Case: 58 year-old, male patient was admitted to our inpatient clinic with the complaints of confusion, persisting headache, fatigue and difficulty in balance for one week. He was known to have cirrhosis and portal hypertension for 2 years and type 2 diabetes mellitus for 20 years in his medical history. He was not on any medication and had been lost to follow-up for 3–4 months. There was not any precipitating factor for this attack.

Neurologic examination revealed confusion, disorientation to time and place, bilateral dysmetria which was more evident on the right side, bilateral flapping tremor and ataxia.

On MRI, T2 and flair images showed symmetric hyperintensities at the level of posterior pons, bilateral cerebral peduncles, posterior limbs of internal capsule, splenium of corpus callosum, posterior regions of the centrum semiovale and periventricular white matter. These areas showed mild hyperintensity on diffusion weighted images, which suggested vasogenic edema around the corticospinal tract. Gradient echo images revealed hypointensity in bilateral globus pallidi suggesting heavy metal deposition.

The patient got better both clinically and radiologically under appropriate treatment.

Conclusion: MRI findings may differ according to the type of liver failure in hepatic encephalopathy. It is the first time, reversible splenial lesion is found to be associated with hepatic encephalopathy.

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Abstract – WCN 2013**No: 895****Topic: 36 – Other topic****Spinal cord myelitis after zoster infection**

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Background: Varicella zoster virus (VZV) can have significant neurologic complications. Myelitis is a relatively rare neurological complication of VZV and may develop in the absence of rash.

Objective: To report main clinical, biological and MRI features of zoster myelitis.

Patients and methods: Five patients were followed-up (1992–2012) for zoster myelitis. All patients had lumbar puncture and medullar MRI.

Results: Our observations concern two women and three men of 17 to 78 years of age. Four patients had history of intercostal rash. After 3 weeks (4–45 days), patients presented paraparesis/plegia, sensory loss, and sphincter dysfunction. Cerebrospinal fluid analysis revealed pleocytosis in two cases, an elevated protein level in all patients. Medullar MRI demonstrated T2 hyperintense lesions with occasional swelling and enhancement. The diagnoses in all 5 cases were supported by laboratory evidence, including increasing titer of anti-VZV antibodies in the CSF. All

patients received a 3-week course of intravenous (IV) acyclovir (1500 mg/m²/day) and IV methylprednisolone in 2 cases. Most patients fully recover within 2 months but sphincter dysfunction may persist.

Conclusion: As the pathogenic mechanism of herpes zoster myelitis is considered to be direct viral invasion of the spinal cord with subsequent necrosis or post-infectious, early initiation of acyclovir treatment is necessary for the recovery.

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Abstract – WCN 2013**No: 972****Topic: 36 – Other topic****Analysis of sleep microstructure in Parkinson's disease: A case-control study**

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Objective: Sleep in Parkinson's disease (PD) is frequently disrupted leading to altered sleep macrostructure and awakenings. Nevertheless, more subtle sleep alterations involving its microstructure may play an important role in sleep fragmentation. We evaluated macrostructure and microstructure of sleep (cyclic alternating patterns or CAP, micro-arousals) in a group of parkinsonian patients, compared with age-matched non-parkinsonian subjects.

Material and methods: 31 PD patients (mean age 59.5 ± 12.4 years; mean Hoehn–Yahr (H-Y) score: 3.4 ± 1.8) were recruited for standard polysomnography. Standard sleep stage scoring and CAP scoring rules were used. The Parkinson's disease sleep scale (PDSS) was used to quantify sleep problems in PD, while daily motor performances were evaluated using the Unified Parkinson's disease Rating Scale (UPDRS).

Results: Compared to controls, polysomnography analysis in PD showed significant increase of sleep onset latency ($p = 0.01$) and decrease of both sleep efficiency ($p = 0.04$) and slow wave sleep ($p < 0.01$). Alteration of these parameters correlated with PDSS score, UPDRS score, H-Y score and duration of the disease. A subgroup of 14 patients with H-Y score < 3 presented sleep macrostructure parameters not significantly different from controls. Nevertheless, in this group CAP rate was significantly higher than controls ($p = 0.02$) and correlated only with disease duration ($r = 0.67$, $p = 0.04$).

Conclusion: Our study showed alterations of NREM sleep microstructure in PD, even at an early stage of the disease and independently from motor symptoms, so that an involvement of brainstem nuclei contributing to the building up and maintenance of NREM sleep could be hypothesized in this disease.

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Abstract – WCN 2013**No: 981****Topic: 36 – Other topic****The relationship between engagement in amateur sports and individual psychological characteristics of university students**

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Physical activity contributes to a healthy lifestyle, helps to prevent numerous chronic diseases (such as cardiovascular diseases,

hypertension, etc.), and plays a therapeutic role in addressing a number of cognitive and psychological disorders (such as depression, low self-esteem etc.). The aim of our investigation was to detect the relationship between engagement in amateur sports and typological and personal differences of university students. 67 students (34 women and 33 men, age 17–23 years, mean age 20.5 years) of National Taras Shevchenko University of Kyiv, Ukraine participated in this study. The control group included students (220 women and 61 men) with no experience playing sports. It was shown that the group of amateur sportsmen had a higher level of arousal and mobility of neural processes, and a low level of neuroticism, lower level of neurotization, anxiety, psychic tension and depression. It was established that amateurs had pronounced outward euphoric and less pronounced outward dysphoric reactions to external stimuli and more inclined to direct physical aggression. We found that amateur sportsmen have more pronounced general and spatial anticipation and lesser intensity of burnout and its symptoms and phases. They provided prerequisites for the inclination to sports (physical activity) and allow the achievement of success. Our data indicate that sports prevents the development of burnout and reduces the negative impact of emotional stress, gives socially acceptable ways of aggressiveness, and teaches athletes to cope with difficulties.

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Abstract – WCN 2013

No: 1122

Topic: 36 – Other topic

Ethical problems in neurology

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Background: Increase in the role of ethical component in the assessment of the effectiveness of the professional work of neurologist.

Objective: Assess the motivation of medical students and practicing neurologists towards studying biomedical ethics.

Material and methods: 25-year experience of teaching biomedical ethics to lower and upper level students in Moscow medical schools, as well as teaching continuous education courses on biomedical ethics to graduate medical students and practicing physicians neurologists.

Methods: Sociological surveys, self-administered questionnaires, standardized and non-standardized interviews. The motivation index (MI) was calculated in the proportion of 1 to 10 per every 10 persons.

Results: Professional interest towards bioethics among the medical students increases, as they approach graduation and become more involved in clinical disciplines: the lowest interest was demonstrated by first-year students (on average – .08), the highest – by the 6th year students (.7). Of particular interest was material on professional ethics; here the MI among the 6th year students reached .85. Graduate medical students and doctors practicing neurology and epileptology demonstrate a high level of interest in ethical problems (.8–.9). At the same time they admitted that there is a significant deficit in knowledge in the field of bioethics.

Conclusion: In order to radically improve the situation with bioethics education in medical schools of Russia, the bioethics classes should be moved to the curricula of upper level students and included in the system of the continuous medical education.

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Abstract – WCN 2013

No: 1106

Topic: 36 – Other topic

Six-year survival of progressive multifocal leukoencephalopathy associated with follicular lymphoma

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Background: Progressive multifocal leukoencephalopathy (PML) occurs mainly in immunocompromised individuals as well as associated with immunomodulatory therapies including rituximab. The disease course of PML is usually progressive and fatal. The median survival of patients without HIV infection is only three months. Long-term remission is rare, but rather occurs in HIV-positive individuals.

Objective: To present a long-term remission case of PML associated with follicular lymphoma.

Patient and methods: A 57-year-old male was presented with rapidly progressing neurological signs 6 years ago (modified Rankin-scale was 5). PML was diagnosed by detailed neuroradiological and histopathological evaluations of brain biopsy specimen. The presence of JC virus was demonstrated by immunohistochemistry and PCR examination from the diseased brain tissue. An inflammatory response with abundant CD8 T-cells was prominent. The first appearance of PML preceded the diagnosis of follicular lymphoma and rituximab therapy. Clinical and neuroradiological improvement followed splenectomy, high-dose corticosteroids and later immunotherapy including rituximab.

Results: After the termination of chemotherapy the patient was rehabilitated and has been in a stable neurological condition (modified Rankin-scale is 3). Clinical improvement correlated with the neuroradiological result.

Conclusion: The inflammatory form of PML extends the spectrum of JC-virus related brain disease and there is a chance of clinical improvement following immune reconstitution.

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Abstract – WCN 2013

No: 1108

Topic: 36 – Other topic

Evolutionary ideas in ancient and medieval neuroscience

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Background: Evolutionary ideas have been essential to understanding the nervous system since antiquity. This lecture will examine the close relationship between the emergence of innovative doctrines and social developments in ancient and medieval times.

Objective: Evolutionary concepts of the pre-modern era are identified and linked to their cultural context.

Material and methods: Important primary sources such as the writings of physicians and philosophers as well as secondary materials are reviewed.

Results: Various interrelations between neurological and political doctrines are demonstrated in detail. F. e., the concept that the brain governs the body corresponds to Plato's idea of a philosopher-king who

rules over the state. Plato's student Aristotle took the opposing view and described the heart as the seat of perception, motion, and sensation, thus relating this organ to the "acropolis" of a contemporary Greek city. Meanwhile Hippocratic authors shaped the doctrine of the four humors, whose harmonious mixture symbolized the democratic interaction of various groups in a Greek community; diseases including epilepsy and apoplexy were believed to occur if and when the excess of one humor provoked a noxious "tyranny" amongst the inner parts of the body. In 2nd century Rome, the physician Galen was able to synthesize various traditions of neurological thinking, acting like the ruling emperor who integrated various territories into the Roman Empire.

Conclusions: A thorough analysis demonstrates that pre-modern evolutionary ideas can be viewed as a direct response to cultural and political circumstances. It would be worth examining later epochs on the same lines.

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Abstract – WCN 2013

No: 868

Topic: 36 – Other topic

Low tryptophan uptake in contrast-enhancing lesions predicts long-term survival in patients with a previously treated glioblastoma: A pet study

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Background: MRI has limited prognostic value in patients with a previously treated glioblastoma. Positron emission tomography (PET) imaging with alpha[¹¹C]methyl-L-tryptophan (AMT), a radiotracer tracking tryptophan transport and metabolism via the immunosuppressive kynurenine pathway, can effectively differentiate glioma recurrence from radiation injury.

Objective: To evaluate the prognostic value of AMT uptake measured on post-treatment PET in patients with glioblastoma treated with surgery followed by chemoradiation.

Patients and methods: AMT-PET was performed in 32 patients (mean age: 60 years) with a previously treated glioblastoma. The PET scans were done 6–62 months after initial surgery after MRI demonstrated a contrast-enhancing lesion suspicious for tumor progression. AMT uptake values were measured (on summed activity images 30–55 min after tracer injection) in contrast-enhancing regions and contralateral cortex, and correlated with survival after PET. A receiver operating characteristic analysis was performed to determine the optimal cutoff threshold to predict 1-year survival.

Results: At the end of follow-up, 23 patients were deceased and 9 alive (for up to 83 months). A cutoff threshold of 1.65 for lesion/cortex AMT uptake ratios provided 82% sensitivity/specificity for 1-year survival after PET. Age-corrected Cox regression analysis showed a strong association between above-threshold AMT uptake ratios and shorter survival (odds-ratio: 4.3 [1.5–12.5], $p = 0.005$). Below-threshold AMT uptake ratios ($n = 13$) were associated with an additional year median survival compared to patients with high ratios (618 days vs. 257 days).

Conclusion: Low AMT uptake on PET, measured in contrast-enhancing lesions suspicious for tumor progression, predicts long-term survival in patients with a previously treated glioblastoma.

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Abstract – WCN 2013

No: 1131

Topic: 36 – Other topic

Sleep curtailment is associated with altered autonomic tonus in normal-weight individuals

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Introduction: Increased sympathetic tonus has been suggested as a mechanism linking short sleep duration to overweight and hypertension. The present study was proposed to assess autonomic tonus in relation to sleep duration in a representative sample of eutrophic individuals from the general adult population.

Methods: A representative sample of the city of Sao Paulo was selected (20–80 years), including 1024 individuals, from which 224 females and 193 males were eutrophic (body mass index ≤ 25). They underwent full polysomnography with ECG recording. Heart Rate Variability (HRV) was analyzed for the whole night using a polysomnography ECG lead (D2 modified). Time and Frequency domains variables were calculated for individuals who slept more (controls) and less than 5 h (experimental group) per night, assessed by objective measures. One-way ANOVA was performed considering insomnia syndrome and $AHI > 5$ as covariates.

Results: Sleep curtailment was significantly associated with reduced SDNN ($p = 0.02$) and SDNNINDEX ($p < 0.01$) which are related to a reduction in HRV. RMSS was also reduced ($p < 0.01$) indicating reduction in parasympathetic tonus, and LF/HF ratio increased suggesting high sympathetic tonus.

Conclusion: Sleep shortage was associated with reduced HRV and parasympathetic tonus, and increased sympathetic tonus in eutrophic individuals. These findings support the concept that altered autonomic tonus is associated to sleep curtailment independently from the presence of obesity. This alteration could mediate the link between short sleep, obesity and cardiovascular risk. Longitudinal studies are needed to support this hypothesis.

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Abstract – WCN 2013

No: 597

Topic: 36 – Other topic

Modelling of mismatch negativity indicates age related impairment of cortical dynamics in frontal regions

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Background: Auditory mismatch negativity (MMN) is an electroencephalographic (EEG) response strongly correlated with attention control. MMN is known to attenuate with age.

Objective: In this study we investigated the cortical networks underpinning attention control in the ageing brain.

Methods: MMN and dynamic causal modelling was used to study connectivity networks of attention control in healthy young and healthy old subjects. An auditory odd-ball paradigm was used to elicit MMN registered with EEG in two groups of healthy subjects with mean age 74 ($n = 30$) and 26 ($n = 26$). Networks with different hierarchical complexity with up to five cortical nodes (right and left primary auditory cortex, right and left superior temporal gyrus, and right inferior frontal gyrus) were analysed using dynamic causal modelling.

Results: Bayesian model comparison showed that the five node model with full modulation of all network connections was the most

likely of all the twelve inverted models. Model inversion of the five node model showed that older subjects had significantly increased inhibition of pyramidal cells ($p < 0.05$) with reduced modulation of this activity within the right inferior frontal gyrus ($p < 0.02$).

Conclusion: Deterioration in frontal-based control mechanisms caused attenuation of MMN with age. Impairment of cortical dynamics in the right inferior frontal gyrus on stimulus change provided a neurobiological mechanistic explanation for the reduction of MMN and attention control in the ageing brain.

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Abstract – WCN 2013

No: 1127

Topic: 36 – Other Topic

Polysomnographic sleep parameters in an aging population

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Introduction: Although many studies have shown the evolution of sleep parameters across the lifespan, not many have included a representative sample of the general population. The objective of this study was to describe changes in sleep patterns in the general population of Sao Paulo throughout adulthood.

Methods: We selected a representative sample of the city of Sao Paulo, Brazil that included both genders and an age range of 20–80 yrs. This sample included 1024 individuals that were submitted to polysomnography (PSG) and structured interviews. We sub-divided our sample into 5-year age groups.

Results: Total sleep time, sleep efficiency, percentage of REM sleep and slow wave sleep showed a significant age-related decrease ($p < 0.05$). WASO, arousal index, sleep latency, REM sleep latency, and the percentage of stages 1 and 2 showed a significant increase ($p < 0.05$). Furthermore, AHI increased and oxygen saturation decreased with age. The reduction in the percentage of REM sleep significantly correlated with age in women, while the reduction in the percentage of slow wave sleep correlated with age in men. The PLM index increased with age in men and women.

Conclusions: Sleep structure and duration underwent significant alterations throughout the aging process in the general population. There was an important effect of age on sleep respiratory parameters and PLM index. In addition, men and women showed similar trends but with different effect sizes.

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Abstract – WCN 2013

No: 1129

Topic: 36 – Other topic

Incidence and mechanisms of traumatic brain injury and the relationship with psychological health among Ontario adolescents

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Background: Limited population based data is available on the prevalence of adolescent traumatic brain injury (TBI) and its adverse psychological correlates.

Objective: To describe lifetime prevalence, the mechanisms of injury, and links between self-reported TBI and mental health, suicidality, bullying and other conduct behaviours among a population based sample of adolescents.

Methods: Data were derived from 4735 questionnaires administered to 7th and 12th graders as part of the 2011 Ontario Student Drug Use and Health Survey.

Results: In total, 19.5% of adolescents reported a TBI in their lifetime. Sports injuries were the most likely mechanism for mTBI among all reported causes (55.8%, 95% CI: 46.9, 4.2). When holding constant sex and grade, adolescents with TBI had significantly greater odds for reported elevated psychological distress (OR = 1.51), attempting suicide (OR = 3.39), seeking counselling through a crisis help line (OR = 2.10), and being prescribed medication for anxiety, depression or both (OR = 2.43). Moreover, they had higher odds of being victimized through bullying at school (OR = 1.70), being cyber-bullied (OR = 2.05) or being threatened with a weapon at school (OR = 2.90), compared with adolescents who never had a TBI. Adolescents with TBI had also higher odds of victimizing others and engaging in numerous violent and non-violent conduct behaviours.

Conclusion: High rates of TBI among adolescents in the general population, its high association with team-sports and adverse psychological effects highlight the need for urgent prioritization of prevention efforts.

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Abstract – WCN 2013

No: 992

Topic: 36 – Other topic

Unselected brain imaging in suspected meningitis delays lumbar puncture, can prolong hospitalisation and may increase antibiotic costs - a pilot study

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Background: Antibiotics reduce mortality in bacterial meningitis; a lumbar puncture (LP) will demonstrate that many patients with suspected meningitis do not need them; but delays reduce chances of culture, particularly if > 8 h. Guidelines advise a LP without brain imaging unless specific features are present.

Objective: We assessed the duration of hospitalisation and inpatient costs incurred with delays in LP in a Northwest of England teaching hospital.

Methods: We screened the cerebrospinal fluid (CSF) database to identify patients with suspected meningitis over 3 months (07–09/2010). Data were recorded from clinical case notes; costs were calculated with established datasets and the British national formulary.

Results: 142 patients were screened; 35 had a suspected CNS infection; 10 had a CNS infection proven: 3 bacterial meningitis; 4 aseptic meningitis and 3 viral encephalitis.

Brain imaging delayed the LP for 19 (54%), (11.08 vs 5.29 hrs, $p = 0.10$); ten (53%) did not need imaging. 11 (42%) of those given antibiotics before the LP were delayed > 8 h.

For patients with aseptic meningitis and those who had a CNS infection excluded, without prior antibiotics, the delay in LP increased duration of hospitalisation ($r = 0.94$, $p = 0.02$ and $r = 0.96$, $p = 0.01$ respectively).

Overall there was no trend with LP delays and antibiotic cost. However, 4 patients had antibiotics continued despite negative

cultures; 2 of whom had the LP delayed >8 h, with an antibiotic cost of £215.96.

Conclusion: Inappropriate brain imaging often delayed the LP; in patients who did not need antibiotics this led to longer hospitalisation.

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Abstract – WCN 2013

No: 1141

Topic: 36 – Other topic

Worldwide record of REM sleep time in a patient with pedunculopontine nucleus area (PPNa) stimulation

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Background: PPNa stimulation improves gait disorders and mildly increases REM sleep and alertness in previous series.

Objective: To evaluate nighttime sleep with and without PPNa stimulation.

Patients and methods: Four patients (3 female, 1 male, 46–69 y old) with Parkinson's disease for 9–22 years had PPNa stimulation for gait disorders. Underwent sleep monitoring after 2 months with and without deep brain stimulation (DBS), in a double blind randomized cross over manner. After 9 months under chronic DBS, male patient had 4 additional sleep monitoring with stimulation on again (n = 2), and 1 and 6 nights after stimulation was stopped. Active contacts were located via normalized MRI.

Results: In the male patient, who had the clearest benefit on gait, REM sleep percentage was 49% (REM sleep time: 210 min; normal values: 17–23%) of total sleep time with stimulation, and 0% without. REM sleep was still increased 9 months later with continuous PPNa stimulation (41% of REM sleep on stimulation). It decreased to 30% after one night and to 9% after 6 nights without stimulation. In the 3 other patients REM sleep was 4–22% without stimulation and 0–21 % with stimulation. The active contacts had similar localization in the 4 patients.

Conclusion: This dramatic, supranormal (twice higher than the higher bound in normal adults), durable and reproducible increase of REM sleep time with PPNa stimulation, followed by a progressive return to low values when stimulation is stopped, suggests that the PPNa stimulates 'REM sleep on' systems (possibly in the subcoeruleus nucleus).

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Abstract – WCN 2013

No: 513

Topic: 36 – Other topic

Neurological lesions in children of KYIV, born on the year of catastrophe in chernobyl

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Object: To study influence of small-scale irradiation on nervous state of growing organism in children.

102 children born in 1986–1997 years have been examined in dynamics.

At age of 6–7 years in 62% of children autonomous disorders on subclinical level have been observed: 33% with sympaticotonic type and 33% with parasympaticotonic type. Children were irritated and tired quickly, concentration of attention was bad.

With sympaticotonic direction of vegetative reactions index Kerdo increased essentially (up to $42.3 \pm 3\%$). Considerable disbalance of cardiovascular tests was noted. In subgroup of these children 23% had congenital orthopedic pathology of spine and lower extremities were marked. With vagoinsular direction of vegetative reaction in children of this age group congenital defects of locomotor and support were less often (14% of cases).

On other hand chronic disorders of bronchopulmonary system and gastroenteric tract (made up 38% of cases) prevailed. With secondary examination of children at the age of 16–17 years it was noted that vegetative dysfunction (mainly with sympaticotonic type) was observed only in 27% of examined children but they had more often disorders connected with locomotor an support, namely: chondrosis of vertebrosacral spine, dysplasia of vertebrae and large joint of limbs, flatness of anterior part of feet, congenital deformities, syndrome of congenital dysplasia of connective tissue on subclinical level. When patients are at the age of 26–27 years dysfunction of autonomous nervous system has reached only 12%. Sympaticotonic and vagoinsular were almost equally divided among them.

Small-scale irradiation affect mainly autonomous nervous system on subclinical level.

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Abstract – WCN 2013

No: 1204

Topic: 36 – Other topic

Lewy neurite-like structures are rarely associated with Lewy body pathology at the brainstem of multiple system atrophy (MSA)

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Background: Two major differences of synuclein pathology between multiple system atrophy (MSA) and Lewy body disease (LBD) are the location of abnormal α -synuclein aggregation, mainly glial or neuronal, and existence of the peripheral organs involvement. We found a case of MSA with numerous Lewy neurite-like swollen structures (MSA neurites). However, it is unclear whether these swollen neurite of MSA are related with Lewy body pathology or not and provide some insights of the relationship between MSA and Lewy body pathology.

Objectives and methods: To clarify the characteristics of the swollen neurites of MSA, we examined 160 autopsy confirmed cases of MSA at our institute. Patient characteristics: age of death 67.3 ± 8.3 years; disease duration to death 7.4 ± 4.1 years. Clinical phenotype was MSA-C (predominant cerebellar involvement) in 84, MSA-P (predominant parkinsonism) in 71. Tissue blocks were taken from the brainstem, and HE staining and immunohistochemical staining for phosphorylated alpha-synuclein were used. We examined for the presence of MSA neurites and Lewy bodies in the brainstem.

Results: Ten cases (6.3%) showed Lewy bodies: 4 cases in the IIIrd nucleus, 7 cases in the locus coeruleus. Twenty two cases (14%) showed MSA neurites; 20 cases in the IIIrd nucleus, no cases in substantia nigra. Only one case with MSA neurites had Lewy body pathology.

Conclusion: Lewy neurite-like swollen neurites might be one of the characteristic findings of MSA rarely combined with Lewy body-related pathology.

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Abstract – WCN 2013**No: 1186****Topic: 36 – Other topic****Air pollution impacts on pregnant women and low birth weight in Kenya**

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The increasing number of vehicles in Kenya roads and growth of industries could see a rise in underweight babies. Pregnant women exposed to higher levels of air pollution from industrial/car fumes are likely to have underweight babies.

Methods: We studied data from 3 million births at 14 Research Centers in rapidly industrializing cities by focusing on air pollutants which are produced from combustion of fuels in vehicles and industrial machines, burning firewood and dust particles.

Results: The more pollutants, the higher the risk/chance of infant being underweight, with average weight down by 3 g.

A direct consequence is the rise in the rate of infant mortality and childhood diseases as well as later health risks in neurocognitive diseases in life, meaning child mortality rate would go up from 55 in every 1000 live births. Although air pollutants alone could not be associated with mean birth weight, we consider health care systems, premature births, social economic status of demographics and months in which low births rates were observed.

Conclusions: Findings were worse for expectant women staying in polluted environments and who smoked. According to WHO, a child is underweight when it tips the scales at less than 2.5 kg. Air particles could lead to LBW due to their impact on body functions. Most cars pollutants consist of dangerous gases like CO₂, lead, sulphur, nitrogenous gases, dust and inhaling them may reduce oxygen uptake. Lead particles in petrol would inhibit certain enzymes and cause damage to the kidneys, brains and nervous systems.

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Abstract – WCN 2013**No: 1190****Topic: 36 – Other topic****Is education related to cognitive ageing? Results of the interdisciplinary long-term study of adulthood and ageing (ILSE)**

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Objectives: The relationship between educational attainment and cognitive status in the elderly is well known. But studies regarding whether education positively affects the cognitive changes show highly inconsistent results. The aim of our study is to examine the effects of education on trajectories of cognitive abilities at age 63 to age 74.

Methods: Data taken from the Interdisciplinary Long-Term Study of Adulthood and Ageing (ILSE) concerned 321 individuals without dementia (mini-mental-status-test \geq 25, Folstein 1990) who were born between 1930 and 1932. Cognitive functioning was assessed on two measurement points (aged 63 at baseline and 74 years at follow-up). Participants completed 12 subtests in the domains 'memory', 'crystalline intelligence', 'fluid intelligence', and 'speed of processing'. Education was dichotomized to reflect high or low attainment. General linear models for analysis of repeated measures were used.

Results: In our sample we found cognitive declines in 9 subtests, stability in 2 subtests and improvements in one subtest. Levels of education were unrelated to trajectories of 10 cognitive subtests. In two subtests high educational attainment attenuates the decline. Adjusted for baseline performance analysis shows a benefit of education in 10 of 12 subtests.

Conclusion: The findings support the active cognitive reserve hypothesis. High levels of education promote more efficient cognitive processing, which results in smaller declines, more stability and improvements.

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Abstract – WCN 2013**No: 1015****Topic: 36 – Other topic****Influence of smoking on regional cerebral blood flow and brain atrophy**

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Background: It has been reported that chronic smoking is related to decreased cerebral blood flow, global atrophy and neurocognitive decline.

Objective: This study investigated the regional cerebral blood flow (rCBF), brain atrophy, and cognition in patients who smoke with no neurological or psychological symptoms. In order to obtain the precise results, we measured rCBF with three-dimensional stereotaxic ROI template (3DSRT) and fine SRT, and brain atrophy with the voxel-based, specific regional analysis system for Alzheimer's disease (VSRAD).

Patients and methods: rCBF, parahippocampal gyrus atrophy, MMSE, WAIS-R, and WMS-R were studied in 16 patients with no neurological or psychological symptoms. Their findings were compared with those of 16 age-matched normal controls. SPECT was performed using the ^{99m}Tc-ECD Patlak Plot method, and 3DSRT and fine SRT developed by Takeuchi was used to evaluate rCBF. VSRAD was used to determine the degree of parahippocampal gyrus atrophy in patients who smoke.

Results: CBF of the angular, thalamus, lenticular nucleus, cerebellar hemisphere, and pericallosal in the smoking group were decreased as compared with those in the non-smoking group, whereas CBF of the orbital and rectal in the smoking group were increased as compared with those in the non-smoking group. Significant differences in parahippocampal gyrus atrophy and cognition tests were not observed in the smoking and non-smoking groups.

Conclusions: It could be that clinically intact smoking patients with no neurological or psychological symptoms had a decreased CBF in the angular, lenticular nucleus, cerebellar hemisphere, and pericallosal and an increased CBF in the orbital and rectal.

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Abstract – WCN 2013**No: 1194****Topic: 36 – Other topic****Optical coherence tomography in patients with optic disc oedema**

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Background: On examination optic disc oedema (ODE) can have similar fundus appearance with optic nerve head drusen (ONHD). Previously, optical coherence tomography (OCT) has been used to image the optic disc and peripapillary retinal nerve fiber layer (RNFL) for differential diagnosis between these conditions.

Objective: In this study we used OCT to investigate differences of the macular structure in ODE and ONHD for the first time.

Methods: High-resolution spectral domain OCT was used to image the macula and peripapillary RNFL in 36 ODE, 76 ONHD patients and 57 healthy participants. Retinal thickness and macular and peripapillary RNFL thickness were measured in inner and outer annuli divided into quadrants.

Results: Peripapillary RNFL was significantly thicker in all areas in ODE patients compared to ONHD and healthy controls ($p < 0.001$ for all quadrants for both). RNFL thickness in nasal area was significantly different for all groups ($p < 0.05$).

ODE patients had thicker retina in outer temporal and inferior segments ($p < 0.05$ for both) compared to other two groups. Macular RNFL was significantly thicker in ODE patients compared to healthy controls in all areas except inner temporal and outer nasal and superior segments ($p < 0.05$) and thinner in ONHD patients.

Conclusion: This study shows that nasal peripapillary RNFL is a sensitive marker for differential diagnosis for ODE and ONHD. For the first time the findings indicate macula involvement with RNFL thickening in ODE and thinning in ONHD compared to controls which should be taken into consideration in clinical assessment of these patients.

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Abstract – WCN 2013

No: 1168

Topic: 36 – Other topic

Changes in presynaptic inhibition during movement restriction of unilateral lower limb in a hemiparetic patient and healthy individuals

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Background: We previously reported that movement restriction of the non-paretic lower limb improved gait patterns in hemiparetic patients.

Objective: We assessed presynaptic inhibition (PI) to determine if movement restriction of the non-paretic lower limb is regulated by upper motor neuron mechanisms in hemiparetic patients compared with healthy individuals.

Patients and methods: This study included one hemiparetic patient and 16 young healthy individuals. Participants walked with a knee brace on the non-paretic/non-dominant side. The subjects walked normally for about 10 min and then with the brace for another 10 min (gait training period). PI was measured before and after every 5 min in both conditions. PI was assessed in the soleus muscle on the paretic/dominant side, and conditioning stimulation of the common peroneal nerve was used prior to soleus H-reflex measurement.

Results: The single limb support time for the paretic/dominant side was longer after gait training in both the hemiparetic and healthy individuals. Two-way ANOVA revealed a significant interaction for time and restriction effects in healthy individuals. After gait training, PI was greater than during normal walking conditions in healthy individuals. In the hemiparetic patient, only slight PI was observed before gait training, but PI appeared to the same degree as in healthy individuals after gait training.

Conclusion: Our results suggest that gait training with a knee brace may be effective not only for improvement of gait patterns, but also for regulation of upper motor neuron systems caused by increasing weight bearing ratio in paretic lower limbs.

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Abstract – WCN 2013

No: 1165

Topic: 36 – Other topic

Comparison of reposition rate between headshaking maneuver and vibration maneuver in ageotropic horizontal BPPV

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Background: Benign paroxysmal positional vertigo (BPPV) is one of the most common disorders causing dizziness. Posterior Canal(PC)-BPPV has been said to account for 60–90% of all BPPV cases, and Horizontal Canal(HC)-BPPV for 5–30% of the cases. However, HC-BPPV now appears to be more prevalent than was previously thought. The ageotropic type is usually persistent, resistant to reposition maneuver, and its pathophysiology is cupulolithiasis in majority. We compare reposition rate between headshaking with modified Semont maneuver and vibration with modified Semont maneuver.

Method: The subject were 64 patients with ageotropic type of HC-BPPV who were diagnosed by the supine roll test using videonystagmography. We perform modified Semont maneuver after headshaking on 33 patients. We performed modified Semont maneuver after vibration on 31 patients. The patients with remaining vertigo after maneuver were retreated with the same method. After 1 week, We interviewed all patients using call whether vertigo has subsided. 4 patients had vertigo but they didn't come to the hospital because of private reason (3 in headshaking group and 1 in vibration group). We count the number of performance and compare with headshaking group and vibration group.

Result: Fifty twice maneuvers were performed in headshaking group and fiftieth maneuvers were performed in vibration group ($P > 0.05$). The more vertigo is severe, the less maneuver was performed in both groups.

Conclusion: There is no significant difference in recurrence rate between headshaking group and vibration group.

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Abstract – WCN 2013

No: 1163

Topic: 36 – Other topic

Symbiosis of neuro-interventionists: From parallel practice to interdisciplinary patient care

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Backgrounds: In centers with endovascular service established by radiologists, neurosurgeon, cardiologists and/or vascular surgeons, budding interventional neurologists may struggle to portray their role.

Objective: In a university teaching hospital, we delivered neuro-interventional treatment by a team-oriented approach consisting of radiologists, neurosurgeons and interventional neurologists. We aimed to evaluate this multidisciplinary patient care model through a quantitative framework construct survey.

Methods: Responded on a voluntary basis, doctors, nurses, and radiographers defined the existing mode of collaboration over a continuum with parallel practice and non-hierarchical integration at both ends, and gave opinions on 4 major aspects: philosophy (believes in benefits of interprofessional collaboration and knowledge of each other's treatment approach), trust, process (knowledge exchange, physician centrality and conflicts associated with inter-professional collaboration) and outcomes (job satisfaction, personal growth, intention to leave in the following year, and research output).

Results: Thirty-eight team-members (38/53, 71.7%) completed the questionnaire.

Twenty-four responders (63.2%) described the mode of collaboration as multidisciplinary or integrative. Majority of doctors (66.7%) and radiographers (62.5%) expressed a strong belief in benefits of interprofessional collaboration but only 28.6% nurses agreed. Compared with nurses and radiographers, doctors showed a stronger belief that the collaboration would enable a better clinical outcome after endovascular intervention ($p = 0.013$). While most doctors enjoyed a high degree of autonomy and more opportunities for personal growth and research through the collaboration, most nurses and radiographers had less autonomy and described the collaboration as physician oriented (all $p < 0.05$).

Conclusions: An interdisciplinary model for neuro-interventional service is feasible.

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Abstract – WCN 2013

No: 1155

Topic: 36 – Other topic

9.4T MRS characterization of human umbilical cord mesenchymal stem cells underwent death metabolism

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Objective: To explore the metabolite profile of mesenchymal stem cells (MSCs) underwent death using 9.4T high resolution NMR Spectroscopy.

Methods: Human umbilical cord mesenchymal stem cells were cultured and collected. MSCs were treated for 0, 6, 12 and 24 h in a stimulated condition which include hypoxia, serum deprivation and changes of microenvironment. Cell death and the mortality rate was detected by light microscopy, Hoechst staining and flow cytometry analyses. Cell metabolite extraction was prepared by methanol–chloroform (M/C) method and analyzed on a 9.4T NMR device. ¹H-NMR Spectroscopy was obtained and the metabolite concentration of each time point was calculated.

Results: Necrosis is the major form of cell death in the built model. In the early stage of cell death (6 and 12 h), the fatty acid metabolite concentration increased with statistical significance ($p < 0.05$), while in the last stage of cell death, the fatty acid peak decreased ($p < 0.05$).

Conclusions: There are some specific characteristics on MRS of MSCs that underwent death, and the fatty acid peak may serve as a biomarker for cell death.

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Abstract – WCN 2013

No: 1157

Topic: 36 – Other topic

Clinical analysis of longstanding subacute myelo-optico-neuropathy (SMON), a clioquinol intoxication

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Background: SMON is a disease caused by clioquinol intoxication. Nearly 10,000 patients of SMON were observed throughout Japan by the end of 1970.

Objective: To investigate current state of SMON patients.

Patients and methods: The subjects were 730 SMON patients (218 males, 512 females; mean age \pm SD, 78.0 ± 8.7 years; age at onset 37.6 ± 9.8 years; duration of illness 45.3 ± 4.0 years) who were examined in 2012 by the SMON Research Committee, supported by the Ministry of Health, Labor, and Welfare of Japan.

Results: At onset, 60.2% of patients were unable to walk, and 5.1% were in complete blindness. At present time, about 46% of patients were still difficult to walk independently, including 8.6% of complete loss of locomotion. 1.6% of patients were in complete blindness and 7.6% had severe visual impairment. The majority of patients exhibited sensory disturbances including superficial and vibratory sensations and dysesthesia. Dysautonomia was observed as leg hypothermia in 70.0%, urinary incontinence 58.5% and bowel disturbance in 76.3%. 98.6% complicated with physical diseases such as, hypertension (52.6%), cardiac diseases (24.1%), vertebral disease (40.4%), limb articular disease (35.5%), and cataract (62.7%). The prevalence rate of dementia in those SMON patients ages over 65 was 16.6%, which is almost the same as the general population in Japan.

Conclusion: The physical states of SMON patient are still severe situations by sequelae of clioquinol intoxication and also by gerontological complications.

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Abstract – WCN 2013

No: 1257

Topic: 36 – Other topic

Long-term cognitive deficits after treated Lyme neuroborreliosis – Neuropsychological profile and demographical characteristics

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Background: Lyme neuroborreliosis (LNB) is mostly successfully treated with antibiotics, but 10–50% of the patients report persisting complaints years after treatment. We have previously reported cognitive deficits in LNB patients 30 months after treatment compared to controls.

Objective: To compare the neuropsychological test-profile characterizing persons who did not recover to the profile of persons who did recover after LNB treatment.

Patients and methods: 50 persons were treated for LNB with ceftriaxone/doxycycline. After 30 months the following neuropsychological tests were performed: Trail Making test (TMT), Stroop, California verbal learning test (CVLT) and digit symbol.

Clinical recovery was defined as a score of 0 or 1 on a clinical composite score combining subjective complaints and neurological deficits (range 0–52, 0 best), non-recovery > 1 . The neuropsychological test-results and demographic data of the recovery ($n = 19$) were compared to the non-recovery ($n = 31$) group.

Results: The non-recovery group scored lower on TMT2 ($p = 0.022$), TMT5 ($p = 0.003$), Stroop1 ($p = 0.001$), Stroop2 ($p = 0.032$), Stroop3 ($p = 0.012$), Stroop 4 ($p = 0.011$), and digit symbol raw score ($p = 0.009$), but CVLT revealed no differences between the two groups. Patients in the non-recovery group were older, ($p = 0.001$), had more somatic comorbidity ($p = 0.09$) and a lower educational level ($p = 0.003$). None of the patients who recovered had pre-treatment symptoms > 6 weeks.

Conclusion: Persons who did not recover after treatment for LNB score were worse on the neuropsychological tests TMT2,5, Stroop1–4 and digit symbol raw score, but not on CVLT. This might reflect a

neuropsychological profile indicating deficits related to attention/executive functions/processing speed and less verbal memory and learning problems.

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Abstract – WCN 2013

No: 1251

Topic: 36 – Other topic

The relation between benign paroxysmal positional vertigo and motion sickness

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Objective: To investigate the clinical signification of BPPV about posterior semicircular canal in patients with motion sickness (MS) and without MS.

Background: The hypothesis of MS is a condition in which a disagreement exists between vision and balance, visually perceived movement and the vestibular system's sense of movement. We speculated that MS could be in relation of the degrees of nystagmus.

Methods: This is a prospective study of 72 patients from 2011 to 2012. When the Dix–Hallpike maneuver was indicative for BPPV using vestibular nystagmography, a repositioning maneuver was performed. Degrees of nystagmus divided into 3 groups (mild: 1–10, moderate: 11–30, severe: >31° of nystagmus). Motion sickness were identified by history. Patients were followed for 3 months for the resolution and recurrence of symptoms.

Results: MS was noted in 42% of patients with BPPV. It showed a tendency of increasing prevalence with an increasing degree of nystagmus ($\rho = 0.424$, $p < 0.001$).

But there is no relation between reduction and presence of MS ($\rho = 0.129$, $p > 0.05$).

Conclusions: These results suggest that MS had considerable factor to predict the severity of BPPV in posterior semicircular canal.

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Abstract – WCN 2013

No: 306

Topic: 36 – Other topic

What factors influence the relationship between feedback on neuropsychological performance and subsequent driving self-regulation?

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Background: A recent study indicates that providing feedback about neuropsychological abilities is important to safe automobile driving (based on Useful Field of View test performance, a visual speed of processing measure) and may lead to changes in driving self-regulation. However, 42% of participants who received negative feedback did not report any subsequent increase in driving self-regulation.

Objective: This study extended those findings to investigate potential moderators of the relationship between feedback regarding neuropsychological abilities and driving self-regulation.

Patients and methods: Between 2006 and 2008, 129 insured older (75+) drivers were administered an initial neuropsychological battery and then a telephone follow-up interview after 2 to 4 months after receiving feedback.

Results: Significant interactions included feedback by age, feedback by number of eye conditions, and a trend towards significance for feedback by baseline driving exposure. Follow-up regression results showed that older participants (80–94; $n = 39$) who received negative feedback significantly increased subsequent avoidance of challenging driving conditions relative to baseline ($p = .045$). Participants with no reported eye conditions ($n = 36$) who received negative neuropsychological feedback significantly increased subsequent driving avoidance relative to baseline ($p = .007$), and participants with low (below median) baseline driving exposure ($n = 66$) tended to increase subsequent driving avoidance relative to baseline ($p < .009$).

Conclusion: Older adults with minimal driving exposure, those with no reported eye conditions, and those over 80 years old are more likely to modify their driving habits after receiving negative feedback regarding neuropsychological skills related to driving safety.

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Abstract – WCN 2013

No: 1230

Topic: 36 – Other topic

A comparison of co-morbidities between the postural orthostatic tachycardia syndrome and generalized dysautonomias

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Background: Postural Orthostatic Tachycardia Syndrome (POTS) includes a spectrum of disorders which are also present in generalized dysautonomias. A pathophysiological link of POTS to the Autonomic Nervous System (ANS) is therefore suspected. Comparing co-morbidities of both conditions could shed further light onto that link.

Objective: Compare the prevalence of common disorders in POTS with that of disorders associated with diseases of the ANS.

Patients and methods: We reviewed 74 patients with POTS and retrieved all their associated diagnoses, retaining the 11 most common. We repeated the same process with 109 patients diagnosed with autonomic neuropathy (AN) who did not meet the criteria for POTS.

Results: Ten out of the 11 most common conditions associated with POTS were also associated with AN but to different degrees. Migraine was the most common co-morbidity in POTS (55%). Other common co-morbidities included Chronic Fatigue Syndrome (CFS), Fibromyalgia and Irritable Bowel Syndrome (IBS). In AN, the most common co-morbidities were hypertension (56%), hypercholesterolemia (46%), migraine (40%) and diabetes (30%).

Conclusions:

1. Common co-morbidities between POTS and AN could reflect a common pathophysiology and a predominant role of the ANS in POTS.
2. Hemodynamic and metabolic disorders are more prevalent in AN than in POTS, whereas conditions like Fibromyalgia, CFS and IBS are more common in POTS, which suggests that POTS is a syndromic rather than a disease-based dysautonomia.
3. The high prevalence of disorders such as Fibromyalgia and CFS in both POTS and AN, may point to a role of the ANS in these conditions.

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Abstract – WCN 2013**No: 1213****Topic: 36 – Other topic****The alpha attenuation test: Assessment of alpha blockade efficiency in narcolepsy**M. Lakocevic^a, N. Rajsic^b, Z. Sundric^c, P. Simonovic^d, J. Marinkovic^e.^aClinics for Endocrinology, Clinical Centre of Serbia, Belgrade University, School of Medicine, Belgrade, Serbia; ^bClinic for Neurology, Military Medical Academy, Belgrade University, School of Medicine, Belgrade, Serbia; ^cInstitute of Aviation Medicine, Belgrade University, School of Medicine, Belgrade, Serbia; ^dInstitute of Mental Health, Belgrade University, School of Medicine, Belgrade, Serbia; ^eInstitute of Medical Statistics and Informatics, Belgrade University, School of Medicine, Belgrade, Serbia**Background:** The alpha attenuation test (AAT) is based on the fact that power spectrum in alpha frequency band of the EEG (8–12 Hz) increases with eyes open, but tends to decrease with eyes closed.**Objective:** To assess value of AAT for predicting coming sleep in individuals with narcolepsy and idiopathic hypersomnia.**Patients and methods:** Seventy one participants (28 with narcolepsy, 24 with idiopathic hypersomnia, and the rest of the 19 has healthy controls matched to gender and age) underwent AAT which considered modified EEG recording with alternately six times eyes open and eyes closed procedures within the time interval of 12 min. Parts of EEG signals were extracted at the same points within the whole sample, and digital filtering with frequency between five and 32 Hz was applied to channels O2-A1 and O1-A2. The parametric statistical analysis of logarithmic transformed alpha attenuation coefficients (AACs) and alpha powers was performed.**Results:** Our results show that patients with narcolepsy have significant lower absolute AACs and greater absolute powers during eyes open condition in comparison to patients with idiopathic hypersomnia and healthy controls. The mechanism of alpha blockade appears to be less effective in patients with narcolepsy, and thus responsible for both lower AACs and greater absolute powers with eyes open.**Conclusion:** AAT may be a useful screening method for assessment of transitory momentary sleepiness. This test can reveal preclinical sign existence of a condition that can lead a subject to fall asleep.

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Abstract – WCN 2013**No: 1062****Topic: 36 – Other topic****Are there benefits of ultra-high field MR for clinical language localization?**R. Beisteiner^{a,b,c}, M. Wurnig^{a,b,c}, F. Fischmeister^{a,b,c}, E. Matt^{a,b,c}, E. Knosp^d, M. Feucht^e, E. Auff^b, S. Trattnig^{c,f}, S. Robinson^{c,f}, A. Geissler^{a,b,c}.^aDepartment of Neurology, Study Group Clinical fMRI, Medical University of Vienna, Vienna, Austria; ^bDepartment of Neurology, Medical University of Vienna, Vienna, Austria; ^cHigh Field MR Center of Excellence, Medical University of Vienna, Vienna, Austria; ^dDepartment of Neurosurgery, Medical University of Vienna, Vienna, Austria; ^eDepartment of Pediatrics, Medical University of Vienna, Vienna, Austria; ^fDepartment of Radiology, Medical University of Vienna, Vienna, Austria**Background:** A first sensorimotor study (Beisteiner et al. 2011) indicated increase in functional sensitivity in primary sensorimotor cortex with ultra-high field MR systems (7T). However, the primary sensorimotor cortex typically shows low artifacts. Here we investigate the functional sensitivity of ultra-high field MR systems in artifact-prone brain areas such as the inferior frontal lobe, by comparing 3T and 7T clinical fMRI results with an overt speech task.**Methods:** Twenty four right-handed patients performed identical overt speech tasks at 3T and 7T. Functional Broca- and Wernicke ROIs were defined separately for 3T and 7T data using individual 5 mm spheres, centered on the individual peak voxel. Within these Broca/Wernicke spheres, six activation measures were compared:

- (1) voxel count (number of supra-threshold voxels at FWE < 0.05),
- (2) mean t-value,
- (3) peak spmT-value,
- (4) percentage signal change,
- (5) contrast to noise ratio (CNR), and
- (6) peak CNR.

Results: In the Wernicke area, all 6 measures showed significant improvement for 7T. In the Broca area, no measure showed a significant 7T benefit, and some measures even indicated a non-significant decrease. Evaluation of the artifact situation indicated that this was due to significantly more inferior frontal artifacts at 7T.**Conclusion:** Our results indicate that the ability to minimize the increase in artifacts with increasing field strength determines the benefit of ultra-high field MR systems for clinical fMRI.**Acknowledgement:** This study was funded by the Vienna Spots of Excellence Program of the Center of Innovation and Technology, City of Vienna (ZIT), Austria.

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Abstract – WCN 2013**No: 1240****Topic: 36 – Other topic****The era of HIV/AIDS and its neurological complications in the world**B.A. Bukusi^{a,b}, R.N. Mbugua^{c,d,e}. ^aClinical Research, Infectious Diseases Control (Kenya Medical Research Institute), Kenya; ^bObstetrics and Gynaecology Clinical Research, Centre for Diseases Control (Kenya Medical Research Institute), Kenya; ^cResearch, Kenya Network of Women Living with HIV/AIDS, Kenya; ^dClinical Research/Psychiatry, Kenya Neurological Society, Kenya; ^eResearch, Kenya Medical Research Institute, Nairobi, Kenya**Background:** The central nervous system (CNS) is often affected by HIV infection. Over 40% of AIDS cases present with neurological symptoms and CNS lesion are detected by anatomical and pathological studies in 80 to 90% of AIDS cases. There may be infections and tumours secondary to the immunodeficiency and pathologies occur directly due to the neurotropism of the virus.**Methods:** Neurological problems associated with HIV infection include encephalopathies, myelopathies, neuropathies and myopathies. HIV induced encephalopathy may develop at any stages of HIV infection and affects all risk groups equally.

The frequencies reported differ between studies due to differences in sampling methods, geographical factors, diagnostic criteria and investigative methods used. The involvement of HIV infected macrophages and microglial cells has been demonstrated. Indirect mechanisms such as release of lymphokines (tumour necrosis factor-TNF alpha- and interleukin-1) and neurotoxicity of the HIV envelope protein, gp 120, have been suggested.

Results: This disorder is known as cognitive and motor syndrome. It presents clinically as a form of sub-cortical dementia with cognitive problems, motor deficits and behavioural disorders depending on the type and stage of infection. The diagnosis can be made after all other infections and tumours common in HIV patients have been ruled out by appropriate investigations like cerebrospinal fluid analysis, cerebral scan and magnetic resonance imaging.

Conclusions: Neuropsychological studies could be of value in diagnosis and in assessing the response to anti-retroviral treatment. The use of new nucleoside analogue drugs in combination with existing drugs may provide new approaches to managing these patients.

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Abstract – WCN 2013

No: 1252

Topic: 36 – Other topic

Optimum gradient artifact removal from EEG-data using facet

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Introduction: Throughout recent years simultaneous recordings of EEG and fMRI has turned into a viable tool for clinical applications. However, elimination of scanner-induced artifacts is still challenging. Here we present an optimum multi-step algorithm which combines various correction steps for the removal of gradient artifacts.

Methods: FACET is a modular toolbox for fast and flexible correction and evaluation of imaging artifacts. Within this toolbox an optimum algorithm (OptGAR) was constructed by combining various pre-processing steps – like slice onset detection and sub-sample alignment – with an adaptive template generation approach to optimally describe the gradient artifact. Residual artifacts are removed using PCA and ANC. This algorithm was evaluated clinically and by automatic comparison to existing algorithms (Allen 2000 and Niazy 2005) via stepwise evaluation.

Results: Across all measures OptGAR outperformed the other algorithms. The median imaging artifact shows a decrease from 69.9 μ V for Allen to 64.9 μ V for Niazy and 54.7 μ V for OptGAR, the RMS ratio an increase from 84.2 (Allen) up to 109.3. Finally, the median residual activity in the theta band (4–8 Hz) shows a decrease from 20.6% (Allen) to 14.3%. For the beta band (12–24 Hz) a decrease from 35.8% (Allen) via 23.2% to only 8.6% (OptGAR) was achieved.

Conclusions: FACET was used to generate a new artifact removal algorithm. This algorithm not only outperforms existing tools but shows a unique performance in reducing the content of slice frequency and its harmonics. OptGAR together with FACET provides a valuable tool for the removal of imaging artifacts from concurrently recorded EEG.

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Abstract – WCN 2013

No: 1270

Topic: 36 – Other topic

A review of scientific literature on optimal glioblastoma treatment in elderly patients

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Introduction: Glioblastoma (GBM) is the most common primary brain tumor in adults and the incidence steadily increases with age.

The diagnostic rate doubles in the age group between 65–84 years and survival is significantly reduced than younger group. Moreover there isn't an established standard of care: the role of surgery and radio-chemotherapy remain controversial. We assess the scientific literature to define the optimal treatment for older patients.

Material and methods: The recent phase III randomized controlled trials, and five prospective-retrospective uncontrolled phase II trials were identified. A total of 1187 GBM elderly patients, median age 72.4, were enrolled in order to receive: standard radiotherapy (RT) vs hypofractionated RT vs 6 cycles of chemotherapy with temozolomide (TMZ) or standard postsurgical involved-field RT vs 100 mg/m² TMZ on days 1–7, 1 week on–1 week off or supportive care alone vs supportive-care alone in combination with RT. The primary end point was overall survival. Phase II studies compared chemotherapy with TMZ in two cases, radio-chemotherapy by Stupp in two cases, and last compared radio-chemotherapy vs RT alone.

Results and conclusion: Our review did not show a statistically significant superiority of a particular treatment, but demonstrates that all therapeutic decision, both chemo and/or radiotherapy, could improve overall survival of elderly patients with good performance status, rather than palliative care (6.7 months-vs-3.8 months). In particular hypofractionated radiotherapy plus temozolomide chemotherapy may be the best way to treat patients with Karnofsky-Performance-Status (KPS) \geq 70%. In patients with a poor KPS, RT could be a reasonable choice. A meta-analysis on collected data will be performed.

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Abstract – WCN 2013

No: 1474

Topic: 36 – Other topic

Analysis of spina bifida prevalence in Brazil in the period from 2001 to 2010

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Background: Spina bifida is a significant cause of perinatal mortality and a cause of disability in surviving children. At the same time, there is a scarcity of research about its prevalence in Brazil.

Objective: Analyse the presence of spina bifida among liveborn infants in Brazil between the years 2001 and 2010.

Method: The data about liveborn infants and the cases of spina bifida has been retrieved from the system of information about liveborn infants of the Ministry of Health, the Department of the National Health Service. The data collection resulted in a descriptive and comparative analysis of the prevalence of spina bifida among 1000 liveborns in different regions of Brazil.

Results: An increase of 17.28% in the presence of spina bifida was observed in Brazil in 2001–2010. The year of 2004 presented a peak in spina bifida prevalence (0209 cases/1000 liveborns). This was followed by a decrease and another increase of 0190 in 2010. Regional differences apply in the prevalence of the disease, with the highest rates in the southern region (0216) in 2001–2010 and the lowest rates in the Centre-western region (0115) in the same period.

Conclusion: There was a growing tendency of the prevalence of spina bifida in Brazil indicating that the incentive of folic acid intake in pregnant women is desirable as a preventive measure. Prenatal care leading to early diagnosis and reduction in complications is also recommended.

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Abstract – WCN 2013**No: 860****Topic: 36 – Other topic****Cryptococcus neoformans neoformans central nervous system infection in a patient with idiopathic CD4 lymphocytopenia mimicking progressive multifocal leucoencephalopathy (PML)**

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Background: Cryptococcal brain infection is a life-threatening disease, mostly associated with varied causes of immunosuppression.

Objective: To report a case of an HIV negative patient who presented with idiopathic CD4 lymphocytopenia and neurocryptococcosis mimicking PML.

Patient and method: We report the case of an immunodeficient patient with brain *Cryptococcus neoformans neoformans* mimicking PML.

Results: A 46-year-old male patient was admitted due to chronic headache, with visual impairment and right-sided tremor. *Cryptococcus neoformans* var *neoformans* was identified in CSF and he underwent standard treatment. Repeated ELISA serologic HIV testing was negative. Five months after discharge he presented with an intracranial hypertension syndrome. A brain CAT scan disclosed bilateral white matter hypointense lesions with mass effect. Another treatment course for cryptococcosis was performed. Laboratory testing revealed a CD4⁺ count of 131 cells/mm³. Three months later, both his visual and neurological deficits progressed, as well as the brain lesions on CAT scan. After another two months, he presented with right-sided motor seizures, right hemiparesis and intracranial hypertension. Serologic *Cryptococcus* testing in the CSF was positive. Extensive investigation for immunodeficiency causes or organ dysfunction was negative. Brain MRI showed bilateral white matter lesions suggestive of PML. A brain biopsy disclosed infection by *Cryptococcus neoformans neoformans*. After another course of Amphotericin B, his motor deficit and brain MRI lesions resolved completely.

Conclusion: This patient with idiopathic immunosuppression showed clinical and MRI findings consistent with PML but pathology revealed an infection with mass effect by *Cryptococcus neoformans neoformans*, with improvement following etiological treatment.

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Abstract – WCN 2013**No: 694****Topic: 36 – Other topic****Study on euronol metabolic characterization in peritumoral area of C6 rat glioma using 1 h MRS at 7 T**

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Background: Glioma has a tendency to infiltrate the surrounding brain tissue for several centimeters from the core of tumor. The insensitive detection of the invasive scope outside the visible tumor border limits the use of structural magnetic resonance (MR) sequences.

Objective: This study aimed to establish a repeatable rat glioma model and investigate the validity of spectroscopy, a functional MR method, in the assessment of the peritumoral area of glioma.

Methods: The C6 glioma cells were stereotaxically implanted into the right basal ganglia region of twenty SD rats. The SD rats

sequentially underwent MR spectroscopy (MRS) at 7 T MR scanner 7 days after operation. Euronol metabolites were measured within the tumor center, peripheral solid parts, adjacent normal-appearing tissue, and contralateral white-matter. All spectra were then quantified by LCModel.

Results: This C6 rat glioma model had a high degree of repeatability. A significant gradual increased concentration of NAA, Cr, Glu + Gln, macromolecule, and a decreased of Lac, Ala from the tumor center to the contralateral white matter were observed. Moreover, the highest level of Cho and Lip were also found in the tumor peripheral solid parts and tumor center, respectively. There were no significant differences in Ins and Tau peak among the four areas.

Conclusion: Our study provides a repeatable animal model for investigating biochemical specificity of glioma. The acquired MRS data supply additional information about the location of glioma potential border.

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Abstract – WCN 2013**No: 1490****Topic: 36 – Other topic****Facial palsy as an initial symptom of Lyme neuroborreliosis in an Austrian endemic area**

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Background: In areas endemic for *Borrelia burgdorferi*, facial palsy is a possible manifestation of neuroborreliosis. According to this all adult patients admitted with facial palsy to our hospital, are investigated for Lyme neuroborreliosis including CSF analysis.

Methods: In this retrospective analysis from 2007 until 2012, we identified 278 consecutive patients who were admitted with facial palsy. All 278 patients underwent clinical neurological examination. 249 of those patients underwent CSF analysis. Demographic and clinical data were analysed according to a protocol.

Results: In 14 out of 278 patients (5%), a definite diagnosis of Lyme neuroborreliosis according to the DGN criteria was established. Four patients were classified as possible Lyme neuroborreliosis. Median age of all patients (n = 18) with facial palsy and Lyme neuroborreliosis was 49 (m = 14/f = 4). CSF analysis revealed pleocytosis (median cell count 240/μl), elevated protein (median = 128 mg/dl), and glucose (median 56 mg/dl).

Conclusion: In a typical area with endemic *Borrelia*, the frequency of Lyme neuroborreliosis in patients admitted due to facial palsy is only 5%. The stringent diagnostic workup including also CSF analysis in almost all patients with facial palsy makes this dataset of Lyme neuroborreliosis meaningful. Although as a consequence of this study, general CSF testing in patients with facial palsy can be omitted.

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Abstract – WCN 2013**No: 1493****Topic: 36 – Other Topic****Incidence of potential drug–drug interactions and its associated factors in neurology wards of two teaching hospitals in Shiraz, Iran**

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Objectives: Drug–drug interactions (DDIs) are one of the most important problems related to optimized drug therapy. This study

estimates the rate and factors associated with potential drug interactions in prescriptions in neurology wards of two teaching hospitals in Shiraz, Iran.

Methods: Data was retrieved from neurology wards of two teaching hospitals handwritten prescription, every day during a period of 6 months in 2012. DDIs were identified using Lexi-Comp 2012 version 1.9.1. Clinical and demographic characteristics of patients were recorded from their files. After assessment of the detected DDIs for clinical relevance, interventions took place through physicians or nurses for type D and X drug interactions. Patient's age and gender, number of medications and orders, length of hospitalization and type of neurological disorder were examined as associated factors to potential DDIs (pDDIs). Outcome of interventions and any probable adverse drug events were documented. Statistical analysis was performed by SPSS 15.

Results: This study included 589 patients. 4942 drug orders and 3874 medications were prescribed and 4539 DDIs were detected, the overall frequency of DDIs was 35.5%. Among neurological agents phenytoin was the most prescribed and account for 76 (12.70%) of patients. Factors having the greatest influence on pDDIs incidences included increased number of medications and seizure. 74.24% of interventions were accepted by physicians and nurses.

Conclusions: Clinically relevant DDIs are common among patients in neurology ward. Pharmacists along with other health care professionals can play an essential role in management of medication therapy in hospitalized patients.

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Abstract – WCN 2013

No: 587

Topic: 36 – Other topic

Dystonia in the Lights from the North Exhibition of German renaissance drawings and prints from the Musée Du Louvre

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Objective: To describe two medieval depictions of dystonia in the Baron Edmond de Rothschild's Collection from the Musée du Louvre.

Background: Medicine and pictorial art often intermingle. Although traditional portraits of some medical conditions are somewhat common, the representation of movement disorders in medieval prints is somewhat unexpected.

Method: By careful analysis of two prints of the Lights from the North Exhibition (German Renaissance Drawings and Prints in Baron Edmond de Rothschild's Collection from the Musée de Louvre), the authors propose that characters were depicted with different forms of dystonia and make some considerations with the historical context.

Results: In the Lights from the North Exhibition (German Renaissance Drawings and Prints in Baron Edmond de Rothschild's Collection from the Musée du Louvre) presented at the MASP (Museu de Arte de São Paulo), two prints by Jean de Cologne, depicted characters with what could be described as different forms of dystonia. In "Christ Arrested", one of the peasants in the foreground appears to have a left lower limb dystonia, while in "Ecce Homo or Christ presented to the people", one character engaged in conversation seems to have a more complex, generalized form of dystonia. We discuss briefly the historical context and compare it with the traditional historical association with Saint Vitu's Dance.

Conclusions: We briefly described two depictions of dystonia in medieval prints of the German Renaissance series of drawings and prints in Baron Edmond de Rothschild's Collection from the Musée du Louvre and made some comments about the historical setting.

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Abstract – WCN 2013

No: 1450

Topic: 36 – Other topic

Darwinian and Jacksonian evolution

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Background: The appearance of evolutionary ideas in the nineteenth century had a pivotal impact on clinical neurology. Darwinism evoked a strong reaction but ultimately became the most important idea in nineteenth century biology. Jacksonian evolutionary neurophysiology, while not Darwinian, produced a consistent bedside science.

Objective: To examine Darwinian and Jacksonian evolutionary thought.

Methods: Historical research.

Results: Jean-Baptiste Lamarck promoted the inheritance of acquired characteristics in a comprehensive evolutionary theory. Exploiting the explosion of literacy in 1840s Britain, the volume *Vestiges of the Natural History of Creation* made the transmutation of species a popular concept. Biology exploded when Alfred Russel Wallace and Charles Darwin independently proposed the evolution of species by natural selection.

Seeing the idea of progressive development as the key to existence, the Herbert Spencer applied evolutionary theory to the phylogeny of the nervous system. John Hughlings Jackson adapted Spencerian evolution to the physiology and pathology of the nervous system, thereby constructing a system of bedside cerebral localization. William Gowers synthesized Jacksonian evolutionary neurophysiology in a usable form in his *Manual of Diseases of the Nervous System*. Konstantin von Monakow and Henry Head brought Jacksonian evolutionary analysis to twentieth century neurology.

Conclusion: The application of evolutionary theory to neurology has been called "the most successful application of the theory of evolution in medicine." The medical use of evolutionary theory allowed Hughlings Jackson to escape the religious approbation that enveloped Darwin. This shows that the world will accept medical ideas when the same ideas evoke controversy when applied generally.

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Abstract – WCN 2013

No: 1458

Topic: 36 – Other topic

Rubella encephalitis and status epilepticus in a young adult male

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Introduction: Rubella is a childhood disease that was historically widespread. Rubella virus (RV) neurological complications are rare. Viral encephalitis refers to an acute inflammatory process of the brain parenchyma due to direct viral infection.

Objective: We report here a case of encephalitis complicated by status epilepticus in a young man from whom it was possible to isolate RV from the cerebrospinal fluid (CSF).

Case report: A 24-year-old man was admitted for generalized tonic-clonic seizure that lasted for more than 15 min. The management of status epilepticus required endotracheal intubation and mechanical ventilation in an intensive care unit. Status epilepticus resolved after 1 mg intravenous clonazepam followed by phenobarbital (dose 10 mg/kg). Physical examination revealed erythematous maculopapular rash on the face, the trunk and extremities. The body temperature was 38, 2 °C. The neurological examination found that the Glasgow coma score was 10/15, meningeal syndrome and a right hypogloss nerve damage. Cerebral magnetic resonance imaging was normal. Electroencephalogram was normal. CSF analysis showed 260

blood cells/mm³ (90% lymphocytes), glucose 4, 3 mmol/L (serum glucose level: 8, 9 mmol/L), protein 1.77 g/L (normal < 0.4 g/L). Serum and CSF anti-RV immunoglobulin M antibodies were positive. Acyclovir (10 mg/kg × 3/day) was given intravenously for two weeks associated with Phenobarbital 3 mg/Kg/day. He remained asymptomatic after 3 years.

Conclusion: RV infection causes a benign disease, and neurological complications remain rare. Among the latter, encephalitis and status epilepticus are extremely uncommon complications of rubella.

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Abstract – WCN 2013

No: 1418

Topic: 36 – Other topic

Encephalic complications of sinusitis about 10 cases

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Background: Sinusitis can cause multiple encephalic complications, which although they are exceptional, can be life-threatening and functional.

Observations: We report 10 cases of adolescents aged 13 to 19 years with sinusitis and not followed-up, which showed multiple cerebral abscesses in 4 cases, brain empyema in 3 cases, 2 cases of encephalitis and 1 case of cerebral thrombophlebitis.

The germ highlighted in four cases was *Haemophilus influenzae*. For other cases, cultures were negative (probably caused by antibiotic earlier).

All patients were HIV negative, and patients who had suppurative collections received treatment with neurosurgery, favorable for 5 of them while 2 patients died. Patients with encephalitis and cerebral thrombophlebitis received antibiotics for 45 days with favorable outcome.

Discussion: The Intracranial suppurations of otorhinolaryngology origin are due in 87% of cases of sinusitis, they are certainly due to the late arrival of patients in our hospitals because they often come at the stage of complications, our patients had an average of 14 days after the onset of symptoms, which is consistent with the literature where some come after 1 month.

Conclusion: Sinusitis is a disease requiring treatment early, thus avoiding serious complications which are frequent in our country because of the late arrival of patients in hospitals.

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Abstract – WCN 2013

No: 1442

Topic: 36 – Other topic

Selective language aphasia for Turkish language from herpes simplex encephalitis (HSE). (Report of one case)

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Herpes simplex encephalitis (HSE) is the most common cause of sporadic fatal encephalitis. Early recognition and diagnosis is very important for treatment with Acyclovir because it is single most effective treatment. Two antigenic types of HSV are distinguished by serologic testing. Type 1 strains (HSV-1) are responsible for almost all cases of HSV encephalitis.

Patient: Patient was a 54 year old, rural Turkish man, who lived at small village near Urmia City in west Azarbayjan, Iran. He is right-handed and his principal language is Turkish. Although he learned Farsi (Persian language) several years ago, he never spoke routinely using Farsi. He arrived at the emergency ward with fever, neck stiffness, disorientation, headache, seizures, and hemiparesis of the right side and Wernicke aphasia. Early brain CT was normal. On LP, there was pleocytosis of 245 WBCs with 75% lymphocytes, sugar 48 mg/dL and protein 140 mg%. CSF HSV-PCR was positive. Later brain MRI shows bitemporal hypersignal lesions consistent of HSE. Treatment with acyclovir 750 mg q8 h i.v. begun immediately on the first hour of admission plus phenytoin. After the second day of treatment, consciousness was better and seizures were controlled. On the 3rd day, he developed total aphasia for Turkish but began speaking using Farsi language. Patient's son reported that his father never has talked Farsi before.

Conclusion: This case shows relative rareness of this selective language aphasia in HSE. Now, the patient speaks both Turkish and Farsi (bilingual) with some dysphasia dominant for the Turkish language.

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Abstract – WCN 2013

No: 993

Topic: 36 – Other topic

Development of clickclinica: A novel smartphone application to generate real-time global neurological disease surveillance data

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Background: In the case of severe but rare neurological infections such as herpes simplex virus (HSV) encephalitis, few centres have sufficient cases for the studies, that are desperately needed, to be conducted. Nevertheless, globally the combined burden of these rare neurological infections is very significant.

The first step in undertaking such studies is to better understand the epidemiology.

There are barriers to obtaining these data; for example, current surveillance mechanisms make it difficult for the busy doctor to know which diseases to notify, to whom and how, and are also time consuming. Consequently, many cases go un-notified.

Objectives: To use novel technology to increase the identification of major neurological infections.

Methods: We developed a free clinical guidelines smartphone application, collecting data as the doctor reads.

Results: Within the first few months there were >3000 users and >2000 disease notifications, from all across the globe, including resource poor countries.

Some important information was gathered, including: new cases of TB meningitis and rabies; also important clinical information, such as where patients with HSV encephalitis are, and are not, receiving acyclovir within the recommended timeframe.

This is a potential resource for public health institutions and for recruitment into studies; automatically putting doctors notifying a case in contact with the local research nurse.

Conclusions: Parallel studies are needed to assess the utility of such applications.

Nevertheless, current surveillance mechanisms miss many cases.

Applications, such as this, are a novel approach with the potential to generate real-time disease surveillance data to augment current methods.

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Abstract – WCN 2013**No: 1433****Topic: 36 – Other topic****Hippocampal changes and associations with psychological symptoms in subjects suffering from anorexia nervosa**

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Background: Anorexia nervosa (AN) is one of the most and severe diseases with the highest mortality rate of all mental disorders. Individuals with AN show certain personality traits, which precede the onset of the disease, e.g. anxiety, obsessive behaviour, negative emotionality, inflexibility, and perfectionism. Studies found a reduction of total hippocampus–amygdala formation volume, a reduced volume in the parahippocampal gyrus bilaterally, and a hypoperfusion in the amygdala–hippocampus complex.

Objective: Since this region plays a crucial role regarding stress and emotions, the aim of this study was to analyze the volume, thickness, and connectivity of hippocampal subfields, and its relation to specific symptoms.

Patients and methods: Twenty women who were currently suffering from AN, and 20 healthy age-matched control women (CW) were tested. The mean age of the subjects was 21.8 years of age (range 14 to 33), and the mean duration of the illness in AN was 5.7 years. Demographic, personal and behavioural data as well as data regarding cortical volume, thickness, and anatomical connectivity was collected from a 3 T magnetic resonance scanner (MRI). Comparisons between groups were performed using paired t-tests.

Results: Since analyses are currently conducted only preliminary results can be reported in this abstract. The results showed a significant reduction of the hippocampal fissure as well as the fimbria in AN. Correlations with psychological aspects (eating disorder symptoms, personality, and coping) were calculated.

Conclusions: The results can help to better understand biologic mechanisms which might lead to psychological symptoms. Therefore, psychotherapeutic treatments can be improved.

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Abstract – WCN 2013**No: 1427****Topic: 36 – Other topic****Brain lateralization of ERP n400 in children with ADHD and their sibs and first cousins**

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Xia et al. (Psych Res: Neuroimaging 2012; 204:161–167) have noticed regional hypotrophy in ventral anterior, medial dorsal and pulvinar nuclei of left thalamus of children with Attention Deficit/Hyperactivity Disorder. Efferents of pulvinar complex terminate in cortical regions in prefrontal, parietal, occipital, and temporal lobes, and in limbic regions, including the hippocampus. The pulvinar–cortical and pulvinar–limbic connections subserves arousal, attention, learning and memory functions and orientation to visual and auditory stimuli. Thus, structural abnormalities associated with the pulvinar complex in children with ADHD could contribute to disrupted attention. Taking into consideration the genetic components of this disorder, linguistic semantic

abilities were studied analyzing ERP N400 activity of 10 patients with ADHD (7M/3F; avg. age, 10.85; avg. IQ, 99) and 11 of their sibs or first cousins without overt signs of the disorder (5M/6F; avg. age, 11.12; avg. IQ, 96) using high-density EEG (256 channels, EGI, Eugene, OR, USA). Pairs of words with either semantic relatedness (automobile vs. ambulance) or lack of it (automobile vs. animal) appeared on a computer monitor and subjects had to respond on a keyboard whether the words were related or not, while their cortical activity was being measured. Results evidenced diminished N400 activity in left-hemisphere regions corresponding to anterior cingulum ($p = 0.029$), pars opercularis ($p = 0.001$), pars triangularis ($p = 0.000$) and supramarginal/angular areas ($p = 0.000$) in patients, and to anterior cingulum ($p = 0.021$), pars opercularis ($p = 0.046$) and pars triangularis ($p = 0.039$) in their sibs and first cousins.

doi:10.1016/j.jns.2013.07.2178

Abstract – WCN 2013**No: 1424****Topic: 36 – Other Topic****Interobserver agreement and validity of bedside “positive signs” for functional weakness, sensory and gait disorders in conversion disorder**

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Background: Experts in the field of Conversion Disorder (CD) have suggested for the upcoming DSM-V edition to put more weight on clinical findings. To date, neurologists rely on the presence of bedside “positive signs” suggestive of functional disorder to establish such diagnosis. Most of these signs have a strong historical background but no robust data exist concerning their reliability and validity.

Objective: To look at the interrater reliability and validity of common “positive signs” of functional weakness, sensory and gait disorders.

Patients and methods: Standardized video-recorded neurological examination was performed in 20 consecutive CD patients (DSM-IV) and 20 organic controls. Thirty-seven clinical signs (1-general, 17-motor, 5-sensory, 14-gait signs) were systematically rated by two independent blinded neurologists on the video-recordings. Signs showing a significant difference between CD and organic patients (Fisher's $p < 0.05$) and a good to excellent interrater reliability (Cohen's kappa > 0.60) were identified.

Results: Signs which discriminated CD from organic controls ($p < 0.05$) were:

Motor: sterno–cleido–mastoid test (kappa:0.83), drift without pronation (0.78), co-contraction (0.77), give-way weakness (0.67), concavity of the palm (0.65) and irregular drift in Mingazzini (0.60);

Sensory: splitting of vibration (0.66) and splitting the midline (0.63);

Gait: falls towards support (0.83), excessive hesitation (0.66) and

General: suffering behaviour (0.62).

Conclusion: Our study identified 11 “positive signs” (out of 37) with a good to excellent interrater reliability that were significantly more frequent in CD patients than organic controls. The use of these signs should be recommended when diagnosing functional weakness, sensory and gait disorders in CD while further studies will aim at validating their specificity and sensitivity in larger samples.

doi:10.1016/j.jns.2013.07.2179

Abstract – WCN 2013**No: 1429****Topic: 36 – Other topic****Brain activity in response to different taste stimuli in subjects suffering from anorexia nervosa in comparison to healthy controls**

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Background: In the Western countries, anorexia nervosa (AN) is one of the most and severe diseases. Patients with AN have an alteration in cortical regions which are responsible for the regulation of feeding behaviour when taste stimuli are administered.

Objective: Therefore, the aim of this study was to analyze the brain activity in response to three different tastes in patients with AN.

Patients and methods: Twenty women who were currently suffering from AN, and 20 healthy age-matched control women (CW) were tested. Demographic, personal and behavioural data as well as imaging data from a 3 Tesla magnetic resonance scanner (MRI) was collected, while four different stimuli were presented to the participants (sucrose, citric acid, umami, and artificial saliva). Functional MRI (fMRI) data was analyzed for regions of interest (insula, orbitofrontal cortex, amygdala and anterior cingulate cortex) using the General Linear Model. Additionally data was correlated with pleasantness ratings of the stimuli.

Results: Since analyses are currently conducted only preliminary results can be reported in this abstract. The mean age of the subjects was 21.8 years of age (range 14 to 33), and the mean duration of the illness in AN was 5.7 years. Pleasantness ratings did not differ between the two.

Conclusions: Since fMRI imaging analyses can provide new insights into the neurobiology of ED, the results can help to better understand biologic mechanisms which lead to normal and pathologic feeding behaviour. Therefore, new and more specific treatments can be developed.

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Abstract – WCN 2013**No: 1422****Topic: 36 – Other topic****The history of WFN**

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Objective: To follow up with World Federation of Neurology (WFN) 1957–2007 and chronicle the history, initiatives and achievements of the WFN from the year 2007 till present day.

Background: In 1957, during the sixth international congress in Brussels, Ludo van Bogaert's proposal for an international federation of neurological societies was accepted and the birth of WFN took place. Over the years, WFN has established multiple Research Groups, and the Continuing Education Committee to meet the goals of the organization. Furthermore, an official newsletter of WFN, World Neurology, has been made available in hard copy and digital formats. Lastly, WFN has partnered with the World Health Organization (WHO) to improve the position of neurology in the world, draw attention to neurological diseases, and promote facet to the science of neurology.

Methods: Information was gathered through literature review of online journals.

Results: The WFN has been actively promoting education and research in neurology for prevention and treatment of disorders of the nervous system since the year 2007 by forming objective specific

committees and teaming up with other health organizations to provide education and care in impoverished regions of the world.

Conclusions: Ever since WFN was formed, various neurological issues were addressed. Starting 2007 up to now is considered to be the golden age of WFN as it has taken up numerous initiatives to promote the dissemination of information and scientific knowledge in the field of neurology globally particularly in the underserved countries of the world. However, there is lot to be accomplished.

doi:10.1016/j.jns.2013.07.2181

Abstract – WCN 2013**No: 1408****Topic: 36 – Other topic****Recurrent Wernicke's encephalopathy in a 16 year-old girl with atypical clinical and radiological features**

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Background: Wernicke's encephalopathy (WE) is a clinical diagnosis with serious neurological consequences. Classic triad of ocular signs, altered sensorium and ataxia is only found in one-third of patients. Its occurrence is underestimated in non-alcoholics and is uncommon in children. We aim to draw attention to a rare case, which had additional clinical and radiological features.

Case: A 16 year-old girl, who underwent intestinal corrective surgeries at 2 months of age for congenital ileo-jejunal atresia, presented with 3-week history of vomiting secondary to intestinal obstruction. She developed diplopia after hospitalization. Neurological evaluation revealed restriction of bilateral lateral rectus with horizontal nystagmus, bilateral limb dysmetria and depressed deep tendon reflexes. Brain MRI was normal. She had prompt improvement to thiamine. Four months later, she presented with two-week history of headache, bilateral severe deafness and tinnitus. Clinically, she had severe sensorineural hearing loss, bilateral lateral rectus palsy and gait ataxia. CT head showed bilateral caudate nucleus hypo-densities. MRI brain revealed symmetrical T2 and FLAIR hyper-intense signals of caudate nuclei and gadolinium enhancement of mamillary bodies and vermis. She had significant improvement after IV Thiamine. Headache completely resolved while the ocular movements, hearing and tinnitus improved partially in 72 h.

Conclusions: Recurrent WE in children is uncommon. Headache, tinnitus and deafness are rare additional clinical features. Although MRI study at second presentation showed typical features of WE, the presence of bilateral caudate nuclei hypo-densities on CT scan is uncommon. Prompt treatment with thiamine is warranted in suspected cases to prevent permanent neurological sequelae.

doi:10.1016/j.jns.2013.07.2182

Abstract – WCN 2013**No: 1402****Topic: 36 – Other topic****The tongue twister: Hypoglossal nerve neurilemmoma/neurinoma/Schwannoma: A rare cause of cranial XII palsy**

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Background: To present a case report study of CN XII Schwannoma in a 59 year old female.

Objective: Schwannoma is a benign neoplasm of the peripheral sensorimotor nervous system comprising 8–10% of all intracranial tumors. Forty-two cases of CN XII Schwannoma have been described by Ocura et.al (1994) and Piccirilli et.al (2007) found 105 cases in the

English literature. Lee et.al (2007) reported only one case in Taiwan and marks the 70th reported case in Medline. In our institution, this is the only case documented.

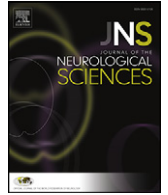
Patient/method: A 59 year-old female was admitted due to tongue numbness and right-sided tongue deviation for ten months. Physical findings revealed all intact except for right sided tongue deviation. The plain cranial CT scan revealed $4 \times 3.2 \times 3.5$ cm left cerebellopontine angle mass extending into the ipsilateral internal auditory canal with a consideration of acoustic or facial nerve Schwannoma; contrast cranial CT scan revealed primary consideration of Schwannoma, meningioma, and cavernous angioma with non-communicating hydrocephalus. Cranial MRI MRA (Magnetic Resonance Angiography) revealed a non-visualization of the left vertebral artery considering hypoplasia or

compression by the ipsilateral cerebellopontine angle mass. Interventricular obstructive (non-communicating) hydrocephalus with MR evidence of increase interventricular pressure was noted.

Result: She underwent suboccipital craniectomy and tumor excision at the left cerebellopontine area; discharged-improved after 20th hospital day. Histopathological findings showed Antoni A and Antoni B bodies typical of Schwannoma. Eight months post-operative, the patient is functioning well.

Conclusion: Because of their scarcity, early diagnosis is intricate. Histopathological result is the best way to confirm the diagnosis.

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Topic: 36 - Other Topic

Abstract – WCN 2013

No: 1356

Topic: 36 Other Topic

Can valerian improve menopausal sleep quality?

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Menopausal women may have sleep problems, which can lead to low quality of life and can use non-pharmacologic methods for its treatment.

Aim: To evaluate effects of valerian on sleep quality in 50–60 years menopausal women.

Material and methods: This is a randomized triple-blind controlled trial. 100 healthy 50–60 years menopause women with insomnia, had been selected from 250 volunteers, whom had been visited in public clinics of west of Tehran. Tools had two main parts of Personal Characteristics and Pittsburg Sleep Quality Index (PSQI). Samples were randomly divided to two groups. Each group received twice a day for four weeks 530 mg of valerian extract or 50 mg starch as placebo. Data analyzed by (T-test). All ethical points were considered by Ethics Committee of Tehran University of Medical Sciences (TUMS).

Results: Equality of personal characteristics and sleep quality before intervention were checked and there were no differences in two groups. Valerian led to significant decrease in average of PSQI in compared to placebo (valerian group before intervention: 9.8 ± 3.6 , after one month intervention: 6.02 ± 2.6) (placebo group before intervention: 11.14 ± 4.3 , after one month intervention: 9.4 ± 3.9) ($p = 0.000$). Also 30% in valerian group and 4% in placebo group had five scores decreased. There was a significant difference between sleep square in two groups ($p = 0.000$).

Conclusion: Findings from this study add to scientific evidence that support the use of valerian in the clinical management of insomnia in menopause.

Acknowledgments: Supported by TUMS and registered in IRCT, year 2010–2011.

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Abstract – WCN 2013

No: 1352

Topic: 36 – Other Topic

Behavioral and neurochemical alterations in C57BL/6 mice exposed to cuprizone: An in vivo ¹H-MRS STUDY at 7.0 T

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Objectives: Recent animal and human studies have suggested that the cuprizone (CPZ) fed C57BL/6 mouse may be used as an animal model of schizophrenia. To date, few studies have used proton magnetic resonance spectroscopy (¹H-MRS) to assess the effects of cuprizone on mouse. In the present study we aim to assess and quantify the regional neurochemical alterations of CPZ-induced mouse model by 7 T ¹H-MRS.

Methods: C57BL/6 mice were given 0.2% CPZ-containing diet for 6 weeks while controls ate the same diet without CPZ. The animals were subjected to behavioral tests and ¹H-MRS scan. An ultra short echo stimulated echo acquisition localization sequence (TR/TM/TE = 5000/10/2.2 ms) was used to measure in vivo proton spectra from the left striatum and thalamus of C57BL/6 mice at 7.0 T and acquired proton spectra post-processed offline with LCModel.

Results: CPZ-fed mice showed significant decrease of spontaneous alternation in the Y-maze test. The concentrations of NAA and NAA + NAAG in the left striatum and thalamus were significantly reduced in CPZ-fed mice. In addition, the concentration of Glu + Gln in the left thalamus of CPZ-fed mice was significantly higher than control mice.

Conclusion: Lower NAA and NAA + NAAG levels may reflect an overall reduction of cellular processes in the caudate of CPZ-fed mice, which may be related to decreases in cell density, or synaptic over pruning. CPZ-fed mice show deficit in working memory as indicated by Y-maze test and have a higher Glx level in their thalamus. These features suggest that the CPZ model is a novel animal model of schizophrenia.

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Abstract – WCN 2013

No: 1339

Topic: 36 – Other Topic

The essence of informed consent

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According to Beauchamp and Childress, the term “informed consent” did not appear until a decade after the Nuremberg trials, almost 70 years ago. There are three critical elements to the concept, as explicated by Bernat: adequate information, presented in terminology the patient can understand; competence on the part of the patient, to understand and accept or refuse what is proposed; and free choice, without coercion. But what is the essence of this concept for a neurologist, in this day and age of information explosion; limited time to spend with individual patients; and increasing pressure to “get the job done”, most efficiently and in the shortest time possible? A controlled clinical trial of therapeutic plasma exchange in 200 consecutive patients with chronic progressive multiple sclerosis afforded the opportunity to explore the meaning of this concept

in practice. In addition, the author's clinical experience in several different cultures (e.g., Denmark; the Republic of Ireland; and India) has validated the conclusions reached in the controlled clinical trial. Differences between the contexts of research and the care of patients with neurologic disease; the impact of practice guidelines, electronic health records, and other modern "improvements" in the practice of medicine in general, and of neurology in particular, will be discussed. The author's conclusion is that the essence of informed consent is a matter of trust – and the creation and affirmation of trust takes time. The implications of such conclusions for a neurologist in today's climate – academic and clinical – will be expounded upon.

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Abstract – WCN 2013

No: 1343

Topic: 36 – Other Topic

Post-ictal EEG findings in a patient diagnosed with alternating hemiplegia of childhood (AHC)

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Background: AHC is a rare disease which hemiplegic physiopathology remains unclear and EEG reports scarce.

Objective: To report and discuss a case of AHC and its EEG findings.

Patients and methods: This patient was evaluated at the Clinical Hospital of Paraná Federal University, Brazil.

Results: An 8-month-old boy was admitted due to a left hemiconvulsion–hemiplegia episode. His first seizure occurred at 3 months of age, a prolonged tonic–clonic fever related episode. He was previously healthy and had unremarkable psychomotor development. Phenobarbital was prescribed but seizures recurred with the same semiologic pattern. After five episodes, convulsions became fever-free and shortly after they evolved into dimidiated tonic–clonic jerks plus ocular deviation to the same side. Post-ictal was then characterized by ipsilateral hypotonic hemiplegia lasting 1 to 3 h outdoing post-ictal somnolence. Seizures increased in frequency and were often prolonged. Laboratory and MRI results were normal. Electroencephalography showed slow activity on the right brain hemisphere and no epileptiform activity during left hemiplegia. On few hours he spontaneously recovered from hemiplegia and at discharge his neurologic exam was normal.

He was diagnosed with AHC and topiramate was prescribed. One month after he had no hemiconvulsion–hemiplegic events.

Conclusion: Although neither basal neurologic abnormalities nor quadriplegic episodes were present yet; hemiconvulsion–hemiplegic syndrome was excluded and he was diagnosed with AHC due to multiple alternating episodes that spontaneously recovered few hours after onset. We present, therefore, an atypical case of AHC which post-ictal EEG and clinical abnormalities are in accordance, suggesting a cortical origin to AHC hemiplegia.

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Abstract – WCN 2013

No: 520

Topic: 36 – Other Topic

Relationship of oxyhaemoglobin signal with blood pressure or skin blood flow during cycle exercise at different intensities: An NIRS study

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Background: Near-infrared spectroscopy (NIRS) is a widely used non-invasive method to measure human brain activation on the basis of the cerebral haemodynamic response and has allowed researchers to visualize cortical activation patterns during locomotor recovery after stroke. However, NIRS has the limitation that systemic changes influence the measured signals.

Objective: This study aimed to determine the relationship between NIRS signals and skin blood flow (SBF) or blood pressure during dynamic movements.

Subjects and methods: Nine healthy volunteers (21.1 ± 0.8 years; 6 women) participated in this study. Each subject received oral and written explanations of the objectives, measurement techniques, and risks and benefits of the investigation. The study was approved by the Ethics Committee of Niigata University of Health and Welfare and was conducted in accordance with the Declaration of Helsinki. The oxyhaemoglobin signal (O₂Hb), SBF, and mean artery pressure (MAP) were measured while the subjects performed multi-step incremental exercise on a cycle ergometer at workloads corresponding to 30%, 50%, and 70% VO₂ peak for 5 min.

Results: Pearson's correlation coefficient between O₂Hb signal and MAP was 0.947 (P < 0.01) at 50% VO₂ peak. The coefficients between O₂Hb and SBF at 30%, 50% and 70% VO₂ peaks were 0.454 (P < 0.05), 0.877 (P < 0.01), and –0.707 (P < 0.01), respectively.

Conclusion: Systemic cardiovascular state changes positively influence O₂Hb signals during moderate-intensity cycle exercise. However, during high-intensity exercise, a negative relationship was observed. To detect cortical activation, clinicians should pay attention to changes in both SBF and MAP.

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Abstract – WCN 2013

No: 956

Topic: 36 – Other Topic

Neonatal onset of a Canavan disease in a Libyan non-Jewish Ashkenazi patient

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Canavan disease is a genetic neurodegenerative disease caused by mutations in the *ASPA* gene. Important clinical features are macrocephaly, hypotonia, head lag and developmental delay. Patients show elevated urinary concentrations of NAA.

We reported here a case of an eight month and a half year old girl, born to related Libyan non-Jewish parents, from complicated pregnancy (eclampsia) and without perinatal risks, brought to our department for the etiologic diagnosis of severe developmental delay.

The neurological exam shows total absence of psychomotor development with central hypotonic syndrome and slight spasticity of the lower extremities with bilateral Rossolimo sign and important macrocephaly. Epileptic seizures were not yet expressed in the infant.

MRI findings show a hyperintense T2 signal interesting bilaterally and symmetrical all subcortical white matter, and periventricular regions with U fibres involvement and a bilateral signal abnormalities in thalamus and palladium.

Proton magnetic resonance spectroscopy of the brain shows an increase in the concentration of N-acetylaspartic acid (NAA).

Although it is a panethnic disease, information on affected individuals in populations of non-Ashkenazi Jewish origin is rather limited. Ongoing research aims at a better understanding of Canavan disease and underlying mechanisms as a basis for new therapeutic approaches.

doi:10.1016/j.jns.2013.07.2190

Abstract – WCN 2013**No: 1307****Topic: 36 – Other Topic****A new clinical maneuver for detection of cerebellar lesions in humans based on cognitive functions of the cerebellum**

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Traditionally the cerebellum has been related almost exclusively to motor functions, in spite of the growing evidence that relates the cerebellum to cognitive functions. Until now, no clinical tests have been previously designed to explore the cerebellar cognitive functions. We describe a clinical test designed to explore the cognitive functions of patients with exclusive cerebellar lesions comparing them to normal subjects. The maneuver consists in the application of 4 neuropsychological standardized tests to the three study groups, in two different clinical conditions. During the first clinical condition, these tests were applied to the subjects in the sitting position. During the second clinical condition, equivalent standardized tests were applied in the Romberg's position. The narration test was the most important test in our study, and the three remaining tests (mental control, category evocation and learning) were applied to evaluate at that same moment, the attention and memory of the volunteers in order to rule out the possibility that an abnormal result in the narration test would be the consequence of having the volunteer's attention focused on the maintenance of the balance during the Romberg's position. When applying the described maneuver and observing the phenomenon we describe as "fluent agrammatism", the risk of having a cerebellum lesion increases 28.6 times.

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Abstract – WCN 2013**No: 532****Topic: 36 – Other Topic****Reduced cerebrovascular reactivity is associated with cortical thinning in children with sickle cell disease**

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Background: Children with sickle cell disease (SCD) suffer from cognitive deficits even with normal appearing conventional MRI. Cerebrovascular reactivity (CVR), defined as the percent change in cerebral blood flow in response to a CO₂ stimulus, is a method for gauging cerebrovascular health and can aid with the clinical assessment of SCD patients. In non-SCD patients, brain regions with reduced CVR have previously been correlated to cortical thinning. However, it is unknown whether compromised CVR in SCD has a direct physiological impact on cortical integrity and could serve as a biomarker of cognitive decline.

Objective: The purpose of this study was to assess if reduced CVR is associated with cortical thinning in children with SCD.

Patients and methods: 20 SCD patients (12–18 years) and 15 controls were imaged on a 3 T MRI. Anatomical and CVR data were acquired. For CVR, a blood-oxygen level-dependent (BOLD) sequence was used during a computer-controlled CO₂ stimulus. CVR maps were computed by correlating the voxel-wise BOLD signal changes to the end-tidal CO₂ waveform, and then coregistered to the anatomical

space. Cortical thickness was computed from the anatomical data and a correlation analysis between thickness and CVR was performed on a region-by-region basis.

Results: Group analysis between patients and controls revealed multiple regions with significant reduction in both cortical thickness and CVR in SCD patients.

Conclusions: The findings indicate that certain cortical areas are increasingly susceptible to poor blood flow regulation, which could potentially lead to cognitive decline. Further studies are needed to investigate this link.

doi:10.1016/j.jns.2013.07.2192

Abstract – WCN 2013**No: 1391****Topic: 36 – Other Topic****Frequency of mutations in the SOD1 gene in ALS patients in Russian population**

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ALS is a fatal neurodegenerative disease characterized by progressive loss of upper and lower motor neurons and death from respiratory and bulbar failure. About 10% of ALS cases are caused by mutations in several genes. The most important is Cu/Zn superoxide dismutase (SOD1) gene. Frequency of mutations in the SOD1 varies from 20 to 35% in familial ALS and from 3 to 7% in sporadic form in different populations.

Patients and methods: First in Russian population, 208 ALS patients (99 females and 109 males) were examined, including 9 patients from 8 unrelated families with familial ALS. Sequence analysis was performed. Molecular modeling analysis was used to demonstrate the pathogenetic role of mutations.

Results: SOD1 mutations were detected in 50% of familial cases and 4.5% of sporadic cases of the disease. Gly16Ala, His48Arg, Leu84Val, Ans86Ser, Asp90Ala, Ser105Leu, Glu133Gly, and Leu144Phe were detected in coding region. All of them are present in ALSod database. For the first time intronic mutations c.-46C>T and c.169+50delAACAGTA (in two unrelated patients) were detected in sporadic ALS cases and weren't present in 385 controls. All 8 revealed coding point mutations of the gene led to moderate or significant changes of the SOD1 protein energy.

Conclusion: The spectrum of SOD1 mutations was revealed in Russian patients. High frequency of SOD1 mutations in familial ALS was detected. The results of in silico analysis of the SOD1 gene mutations confirm staying of ALS within the class of diseases characterize by forming of cytotoxic insoluble protein inclusions in neurons.

doi:10.1016/j.jns.2013.07.2193

Abstract – WCN 2013**No: 880****Topic: 36 – Other Topic****Prospective study of the effectiveness of DOTs regimen in comparison with conventional daily regimen of ATT in tuberculous meningitis**

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Neurotuberculosis comprises around 10% of extrapulmonary tuberculosis. Neurotuberculosis continues to have a high mortality and morbidity.

The standard treatment for tuberculous meningitis (TBM) has undergone considerable change in the recent past. Neurologists all over India

are reluctant to accept Directly Observed Treatment Short Course (DOTS) intermittent regimen for TBM since its introduction. There are no randomized trials to compare daily regular regimen with Revised National TB Control Programme (RNTCP-DOTS) regimen.

Aim: A prospective study to compare the effectiveness in two groups of TBM patients who were assigned to conventional daily and DOTS intermittent regimen.

Material and methods: Patients diagnosed of having TBM according to the diagnostic criteria laid by consensus case definition for TBM at the international TBM workshop were included during the period from September, 2010 to March, 2012. Group I patients received DOTS regimen and Group II patients received conventional daily regimen and they were followed until they completed the course of ATT.

Results: A total of sixty eight patients were included in the study of which 36 were in the DOTS group and 32 in the conventional group. Baseline characteristics, clinical features and laboratory parameters between the two groups were similar. There was no statistically significant difference in terms of mortality, morbidity, number of patients who lost to follow up and number of patients who developed drug induced hepatitis between the two groups.

Conclusion: DOTS and conventional regimen are equally effective in tuberculous meningitis.

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Abstract – WCN 2013

No: 1399

Topic: 36 – Other Topic

Quantitative assessment of the autonomic nervous system in young patients with vertebra-basilar dyscirculation

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Heart rate variability (HRV) has proven to be a sensitive clinical test for early detection of autonomic nervous system (ANS) dysfunction in various neurological disorders.

The aim of the investigation is to estimate dependency of ANS dysfunction from the changes of circulation in vertebra-basilar system in young patients.

89 patients aged from 18 to 44 years with clinical picture of spondylogenic vertebra-basilar insufficiency (SVBD) have been under investigation. 3 groups were formed: 1 – 39 patients with angiodystonic stage of SVBD, group 2 – 26 patients with angiodystonic-ischemic stage of SVBD, and group 3 – 24 patients with ischemic stage of SVBD. For blood supply assessment ultrasound dopplerography and duplex scanning were performed; to diagnose the cervical part of the vertebral column the X-ray examination and MRI were done. To evaluate the changes in ANS, the HRV was performed (Cardiolab, Khai Medica). The investigation was done in the rest and after De Kleyn Test. The study of the HRV, obtained after De Kleyn Test was characterized by increasing power of spectrum in the area of very lower frequency (especially in group 1), that reflects activating of sympathetic part of the ANS, providing realization of ergotropic activity. Overstrain of trophotropic activity in group 2 was characterized by increasing power of spectrum in the area of high frequency, that reflects activating of compensatory mechanism of ANS, directed to the normalization of autonomic disbalance.

These data may be used as a marker of involving of autonomic levels of regulation during performing provocative functional probes.

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Abstract – WCN 2013

No: 1397

Topic: 36 – Other Topic

Clinico-pathological correlates in cerebellar ataxia with neuropathy and vestibular areflexia syndrome (CANVAS)

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Background: CANVAS is a novel vestibulo-cerebellar ataxia with characteristic clinical features, atrophy of the cerebellum and cranial and spinal nerve sensory ganglia. CANVAS is comprised of the clinical triad of cerebellar ataxia, bilateral vestibular hypofunction and a somatic sensory deficit, and is associated with an abnormal visually enhanced vestibulo-ocular reflex (VVOR).

Objective: To further elucidate the pathology underlying CANVAS.

Patients and methods: Brain and spinal neuropathology in two CANVAS patient, together with brain and otopathology in another patient was correlated with clinical, neurophysiological and MRI data on 51 CANVAS patients.

Results: A marked spinal dorsal root and vestibular, trigeminal and facial neuropathy (ganglionopathy) was seen on post-mortem histopathology. Brain pathology revealed selective cerebellar cortical atrophy with marked Purkinje cell loss. All patients had brain MRI and 46/51 had evidence of cerebellar atrophy involving anterior and dorsal vermis, and the hemispheric crus I.

Conclusion: We have been able to advance the clinical understanding and investigation of CANVAS patients by utilizing recently available post-mortem pathological material, and show that the underlying sensory pathology in CANVAS is in the sensory ganglia, including the spinal dorsal root ganglia, as well as those of cranial nerves 5, 7 and 8. Other ataxias which may present with a combination of cerebellar impairment, a bilateral vestibulopathy and a somatic sensory deficit include both spinocerebellar ataxia type 3 (SCA3 or Machado-Joseph disease) and Friedreich's ataxia. The CANVAS patients that we have identified include members of 7 kindreds and identification of the pathological gene is under investigation.

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Abstract – WCN 2013

No: 1384

Topic: 36 – Other Topic

Hereditary spastic paraplegia in children

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Background: A large number of children present with developmental delay and spasticity of the limbs. The causes may vary from perinatal events to the structural anomalies of the brain. In a significant group of children no underlying cause is identified. Hereditary spastic paraplegia is uncommon in children. This has to be considered particularly if family history of a sibling is present.

Objectives: The aim of the study was to find out spectrum of the hereditary spastic paraplegia in children in Oman.

Methods: This is a retrospective study from January 1994 to December 2012 on children with delayed development, gait disorder and motor handicap with signs of symmetrical pyramidal tract involvement. A detailed perinatal and family history including age of onset of symptoms was recorded. The children were labeled as pure or complicated form on the established diagnostic criteria. In families with more than one affected child, all other siblings and parents were also examined.

Results: Eighty five children, from 36 families were diagnosed to have hereditary spastic paraplegia. Parental consanguinity was seen in 91%. Onset of the disease under one year was in 49 (57.6%). Complicated hereditary spastic paraplegia was the most common type in 81%. Speech involvement, mental retardation and epilepsy were the most common associated abnormalities. Nonspecific white matter changes and corpus callosum abnormalities were noted in 21 (24.7%) on magnetic resonance imaging.

Conclusions: The study describes clinical features of 85 children with hereditary spastic paraplegia. Autosomal recessive complicated hereditary spastic paraplegia was seen in 81%.

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Abstract – WCN 2013

No: 1285

Topic: 36 – Other Topic

Can deep brain microstimulation be used to study the thalamocortical connection of the tactile sensation?

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Background: The medial lemniscus is crucial for somatosensory signal transmission from peripheral to telencephalon. In the past, applying adequate stimuli in peripheral was essential for investigating sensory functions. An electrical microstimulation (ES) method was developed here to investigate the possibility of bypassing the peripheral receptors to study connections within this pathway.

Objective: To establish a method of microstimulation in rat thalamus that induces sensory cortex response to mimic the tactile response induced in peripheral.

Material and methods: Male Wistar rat was adequately anesthetized and craniotomy was performed. Tactile stimulation (TS) was applied to the hindpaw, and the contralateral responses were recorded by two glass microelectrodes in thalamus and cortex respectively. Then, ES was applied to the thalamus electrode. The cortical action potentials (APs) obtained by ES were carefully compared with those obtained by TS.

Results: The ES strength and duration for inducing APs were between 40 to 90 V and 1 to 2 ms in a total of 5 rats. The APs of ES responses were similar to those of TS. The time latencies of ES APs were compatible to the time lag between thalamus and cortex of the TS APs (7.93 ± 1.22 ms vs. 9.21 ± 1.13 ms).

Conclusion: The similar latency and waveforms of these APs suggested that ES in thalamus can elicit similar responses in cortex as tactile stimuli do in peripheral. This may shed light on the study of a connection within the entire tertiary tactile pathway in a more precise and controllable way.

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Abstract – WCN 2013

No: 1514

Topic: 36 – Other Topic

Clinical profile of anti-NMDAR encephalitis in patients of China

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Background: Anti-NMDA-receptor (NMDAR) encephalitis is a severe disorder that occurs in association with antibodies to the NR1 subunit of the NMDAR and results in a characteristic syndrome.

Objective: To describe the clinical profile of anti-NMDAR encephalitis in patients of China.

Methods: More than two hundred serum and CSF samples of patients were examined for NMDAR antibodies. 45 of them had NMDAR antibodies.

Results: More than two hundred serum and CSF samples of patients were examined for NMDAR antibodies. 24 patients (53%) were male. 26 (58%) of all 45 patients were both serum and CSF anti-NMDA receptor IgG positive, 17 patients were only CSF IgG positive and 2 patients were only serum IgG positive. There is no difference in CSF anti-NMDA receptor IgG titers between 15 head MRI positive and 30 negative patients. 11 patients also had anti-NMDA receptor IgA in CSF with 3 of them serum IgA positive. Six patients (13%) were serum anti-thyroid antibodies positive (2 anti-TG, 2 anti-TPO, 2 anti-TG and anti-TPO both positive each). 8 patients (18%) were serum ANA positive (titer > 1:100). 2 patients were weak serum Ma2 positive and 1 patient weak serum Yo positive by immunoblotting. CSF examinations: mimic viral encephalitis pleocytosis in 30 patients (75%). 4 female patients had cystic lesions in the pelvis. One patient died after 2 months. Abnormal EEG changes in 40 patients (89%).

Conclusion: Anti-NMDAR encephalitis is highly predictable, recognizable on clinical grounds and can be confirmed with the demonstration of NMDAR antibodies. And early diagnosis and treatment should be emphasized.

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Abstract – WCN 2013

No: 1464

Topic: 36 – Other topic

Sleep disorders in neurological diseases: A preliminary study of the adapted Arabic version of the Pittsburgh Sleep Quality Index scale

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Introduction: Sleep disorders in patients with neurological diseases are often underdiagnosed, although their prevalence and their impact on life quality are significant.

We aimed to translate the Pittsburgh Sleep Quality Index (PSQI) scale in an Arabic version and to assess the reliability of the translated version in patients with neurological disorders.

Methods: Two independent translators converted the French original version into Arabic version simultaneously. The consensual version was converted back into French by two other independent translators. A comparison between the original and the translated-back versions was performed by an independent expert. A study was conducted among 40 patients to assess the understanding practically. Life quality, depression and anxiety were evaluated respectively by SF36, Beck and Hamilton scale.

Results: Mean age of patients was 53 years. Mean PSQI results were 10.52 (range: 1–18). 82% of patients had an impairment of sleep quality. Depression was diagnosed in 73% and anxiety in 42%. Life quality was impaired in 87.5%. Understanding of the scale was unremarkable. Results of different items were homogenous (alpha Cronbach coefficient: 0.78). A positive correlation was found between PSQI results and age, gender, unemployment, smoking, comorbidities and the age of onset. However, no relationship was noted between the type of the disease, period of progress, depression, anxiety and life quality.

Conclusion: The elaborated Arabic version of the PSQI scale presents the same structural hallmarks. It is a simple and reliable tool to

evaluate the occurrence and the severity of sleep disorders in patients with neurological diseases.

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Abstract – WCN 2013

No: 1505

Topic: 36 – Other topic

Paraneoplastic motor neuron disease

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Introduction: Paraneoplastic neurologic syndrome involves neurologic symptoms resulting from damage of the nervous system that are not due to the effects of the primary tumor mass, metastatic disease or other cancer related complications. Paraneoplastic motor neuron disease is rare.

Objective: To describe the clinical and biological features in patients with paraneoplastic motor neuron disease.

Methods: We analyzed, through a retrospective study, the files of patients followed over a four-year (2008–2012) and diagnosed as paraneoplastic motor neuron disease. Diagnosis was made according to Euronet group and Escorial criteria. Electrophysiological examination was performed in all patients. The detection of well-characterized onconeural antibodies and the research of the primitive tumor were made.

Results: We included 6 patients. Mean age was 53 years. All of them had definite Amyotrophic Lateral Sclerosis. Onconeural antibodies were identified in all patients. All patients had definite paraneoplastic syndrome. Breast cancer was found in one patient and lung cancer in another patient.

Conclusion: A motor neuron disease may occur rarely as a paraneoplastic syndrome. It can be isolated or associated with other neurological syndromes. The rapid course of the disorder and the alteration of general state in patients suggest a paraneoplastic syndrome. The identification of the disorder is important because it may lead to the early detection and treatment of the tumor.

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Abstract – WCN 2013

No: 1593

Topic: 36 – Other topic

The prevalence of abnormalities of the main vessels of the head in the open population of the city of Ulyanovsk

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Background: Every year 18 million people die from cerebrovascular diseases (Estrol C., 2011). The priority direction of neurology is to improve the quality of early diagnostics of cerebral vascular diseases (Suslina Z.A., and etc., 2010).

Aim: To study the ultrasonic signs of abnormalities of the main head arteries in the course of chronic cerebrovascular diseases (CCD).

Materials and methods: 236 persons, 40–59 years old, divided into groups, according to clinical manifestations of CCD. 120 subjects didn't have any circulatory system disorders, a group with CCD included 116 people. We evaluated the thickness of intima-media complex, the presence of atherosclerosis plaques, stenosis and vessel tortuosity.

Results: The thickening of intima-media complex was visualized in 150 (63.5%) people, atherosclerosis of cervical arteries with stenosis

was detected in 75 (31.7%) patients, tortuous cervical arteries found in 75 (31.8%) and tortuous vertebral arteries – in 112 (47.6%) people. The thickening of intima-media complex was more common for the CCD group (75%), than for healthy subjects (43.5%), $p < 0.05$. Tortuosity of brachiocephalic arteries (35%) and vertebral arteries (64%) in CCD patients was seen more often, than in the group without circulation disorders (17.4% and 26.1%, respectively), $p < 0.05$. Both groups demonstrated approximately the same frequency of atherosclerosis and stenosis.

Conclusions: The most frequent ultrasound findings in the observed patients are thickening of intima-media complex and tortuosity of vertebral arteries, which are signs of hypertensive angiopathy. Although atherosclerosis with stenosis occurs equally often both in patients with clinical evidence of CCD or without it.

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Abstract – WCN 2013

No: 1580

Topic: 36 – Other topic

Psychometric diagnostic of cognitive functions and motor skills – Findings from a neuropsychological test battery on Niemann–Pick type C-patients

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Niemann–Pick type C (NPC), a rare genetic lysosomal metabolic disorder caused by a mutation in the NPC1 or NPC2 gene, can affect people at all age. The onset, its symptoms and its progression are very inhomogeneous distributed over all patients.

Neuropsychological testings examine areas in the brain which are responsible for cognitive functions like vigilance, concentration, learning, memory, language skills, etc. In addition motor skills can be measured by tests to see how the patient moves in various situations and manages different tasks. Concerning this context it is challenging to have a test battery being able to measure those functions in patients with different stages of disease. In this study we tried to tie on a former study to attain the aim to optimize a neuropsychological test battery for adult NPC-patients.

All in all we tested 22 NPC-patients from three countries with 11 tests including the TMT A/B, PPVT, CRFT, Grooved Pegboard as well as German-speaking tests containing suitable norms of age.

Results show that in some cases tests had been too challenging for some patients in advanced phases of their disease also some did not distinguish enough in lower performance ranges. Nevertheless a few showed specific results of the sample's cognitive functions whereas intraindividual comparisons over time would be interesting. In addition one should think the alternative to relate results of NPC-patients with either older people or in opposite with developmental age of children when norms do not exist in context of particular tests.

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Abstract – WCN 2013

No: 179

Topic: 36 – Other topic

Peripheral neuropathies in childhood, a case report of Churg–Strauss syndrome in a Czech patient

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Churg–Strauss syndrome (CSS) is a rare form of small-vessel vasculitis. We describe a 17-year-old Czech girl, characterized predominantly by peripheral neuropathy, the presence of cardiac and pulmonary involvement, hypereosinophilia, asthma and sinusitis led to the diagnosis of CSS.

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Abstract – WCN 2013

No: 1577

Topic: 36 – Other topic

Clinical characteristics and cerebrospinal fluid adenosine deaminase activity in patients with meningitis caused by Varicella-Zoster virus

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Background: Varicella-zoster virus (VZV) is an unusual cause of meningitis in the immunocompetent patient. Most cases of VZV-associated aseptic meningitis are associated with the skin rash of primary varicella, localized herpes zoster, or disseminated zoster. The diagnostic value of adenosine deaminase (ADA) activity in cerebrospinal fluid (CSF) of tuberculous meningitis has been well documented. VZV meningitis without zoster rash but having high CSF ADA activity tend to be misdiagnosed as tuberculous meningitis.

Methods: We evaluated patients who were admitted due to meningitis from 2010 to 2012 with VZV PCR-positive CSF. According to existence of zoster rash, patients were divided into two groups. We investigated headache intensity, clinical course, concurrent neurological symptoms, CSF profile, the effect of analgesics, and clinical prognosis.

Results: 13 patients (19–55 years of age) had VZV PCR-positive CSF. Only 3 patients (23%) had a concomitant zoster rash. All two groups had acute onset, no other neurological deficit except for neck stiffness, no hypoglycorrhachia and good prognosis. However, patients without rash had more severe headache symptoms, longer headache duration, poorer effect of analgesics, and higher CSF cell, protein, and ADA level than those with rash. (CSF WBC 478.9 vs. 77.7, CSF protein 158.7 mg/dL vs. 56.6 mg/dL, CSF ADA 10.6 IU/L vs. 5.3 IU/L).

Conclusion: The patients with VZV meningitis without accompanying zoster rash tend to present more severe headache and higher CSF ADA activity.

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Abstract – WCN 2013

No: 1549

Topic: 36 – Other topic

Lyme disease presenting with bilateral facial palsy

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Objective: The bilateral facial palsy is a rare form of facial palsy, which usually may be a serious diagnostic problem.

Patients and method: We have presented the clinical case of a 59-year old woman with consecutive affection of the muscles of both facial sides, as well as the diagnostic options for the patient. We have carried out many clinical and paraclinical tests – full clinical examination, laboratory methods, X-ray examination, computer tomography, magnetic resonance tomography, electromyography, immunology. The patient refused a lumbar puncture to be made.

Results: Following the tests that were described above and the continuous monitoring of the patient, we have established that this is

a Lyme disease. The respective etiological treatment was carried out, which leads to a significant improvement in the patient's condition.

Conclusion: The bilateral facial palsy is a condition, which requires a thorough examination of the patient. One of the common reasons for it may be the Lyme disease, which requires timely diagnosis and early etiological treatment.

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Abstract – WCN 2013

No: 1551

Topic: 36 – Other topic

Methionine PET and DTI tractography contribution in differential diagnosis of LETM and mimicking disorders: 3 clinical cases presentation

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LETM – spinal cord disorder with MRI presentation of lesion extended over 3 vertebral segments. Differential diagnosis includes demyelination, autoimmune, infectious process, and tumors. Routine diagnostic procedures include conventional MRI, blood, and CSF tests.

In brain pathology [11C]-methionine-PET and DTI-tractography are used to differentiate neoplastic, inflammatory or ischemic lesions. It is limited data on PET and DTI diagnostic value in spinal cord disorders. 3 clinical cases are presented to demonstrate the significance of these neuroimaging techniques in clinical practice.

59 y.o. female developed subacute progressive cervical myelopathy requiring differential diagnosis between neoplastic, ischemic or demyelinating disorder. Normal [11C]-methionine-PET and brain MRI, spinal cord tracts interruption, negative serology for systemic disorders and AQP4 antibodies, positive CSF-OB complied with the diagnosis of LETM associated with OB-IgG.

21 y.o. female with 2 years history of progressive “autoimmune” myelopathy was treated with corticosteroids, IVIG, immunomodulating and immunosuppressive agents. Positive spinal cord [11C]-methionine-PET and distorted spinal cord tracts on DTI matched the intramedullary neoplastic process. Diagnosis of glioblastoma was confirmed by biopsy. 45 y.o. male presented with rapid progression of paraplegia, and bladder dysfunction. Thoracic spinal cord MRI revealed lesion consistent with LETM. Patient was treated with steroids, and azathioprine without effect. Negative [11C]-methionine-PET confirmed no tumor. MRI revealed diffuse T2-hyperintensive and thickened non-enhancing thoracic spinal cord and enlarged tortuous dorsal spinal vein draining from Th8 to 9 dural arteriovenous fistula. Longitudinal spinal tracts were intact which was considered of possible prognostic value for planned neurosurgery.

It is advisable to incorporate [11C]-methionine-PET and DTI-tractography to optimize neuroimaging protocols in LETM and mimicking disorders.

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Abstract – WCN 2013

No: 1478

Topic: 36 – Other topic

Genome-wide analysis reveals genes associated with structural changes in the human brain

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The human brain structure and susceptibility to pathology are mainly determined by the genetic constitution of the individual. Here we studied relationship between genomic map and brain structural changes in a large cohort of non-demented individuals using neuroimaging-genetic network linkage analysis.

Structural MRI and genomic data were acquired from the Alzheimer's Disease Neuroimaging Initiative (phase ADNI-1/GO/2). T1 images (n = 1083) were nonlinearly registered to form a study-specific template using SyN software. The voxel wise Jacobian map was log-transformed and decomposed into independent spatial maps using Independent Component Analysis. The modulation profiles of each component across subjects with available SNP microarray data – normal (n = 301) and MCI (n = 537) – was used as a quantitative phenotype encoding regional brain expansion/shrinkage and was fitted to a GLM model at each SNP controlled for age, gender, disease status and three axes of genetic stratification reflecting ethnic variation.

ICA showed clusters corresponded with anatomically coherent brain regions (e.g. hippocampus). Volumetric changes in these regions correlated with several genetic polymorphisms which passed genome-wide significance at numerous loci. Functional clustering of these significant genes unraveled several groups encompassing axon guidance (e.g. UNC5C, rs115361061, p-value = 4.3E-12), cell cycle (e.g. COBRA1, rs145030268, p-value = 3.4E-58), cytoskeleton (e.g. WASF3, rs180897335, p-value = 5.74E-09), cell junction (e.g. TJP3, rs148832035, p-value = 2.06E-61), neural differentiation (e.g. NRG2 = rs77901652, p-value = 7.53E-10), neurotransmission (e.g. GRIA4, rs1562223, p-value = 1.32E-10) and the ubiquitin system (e.g. USP13, rs115163683, p-value = 2.04E-08).

Network of genes are associated with distinct regional changes in brain structure that underlies neural development and aging. This network may determine disease susceptibility in neurodegenerative disorders.

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Abstract – WCN 2013

No: 1327

Topic: 36 – Other topic

Internal jugular vein thrombosis secondary to pulmonary tuberculosis: A case of Lemierre syndrome

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Background: Internal jugular vein (IJV) thrombosis (Lemierre syndrome), associated with suppurative infection of the upper aero-digestive tract was first described a century ago. The complications associated with IJV thrombosis are potentially lethal and it is therefore important to recognize and treat this condition early. Herein we describe a case of infective IJV thrombosis secondary to pulmonary tuberculosis.

Case: A 33 year old male presented with blurred vision and bifrontal headache. He had a 3 month history of pulmonary tuberculosis and right transverse sinus thrombosis which were started to be treated. He had bilateral papilledema, visual acuity was decreased and he had high intracranial pressure. Cranial MRI an MR venography revealed irregularities in the right transverse sinus consistent with chronic sequel thrombosis. Bilateral carotid and vertebra artery Doppler USG and angiography revealed right internal jugular venous thrombosis.

Ultrasonography of neck showed lymphadenopathies in both cervical and submandibular lymph nodes. Aggressive antituberculous treatment along with anticoagulation was continued with nearly full recovery of vision.

Conclusion: We want to report this case as we feel, that with the advent of the antibiotic era, this syndrome has become rare; and so “quite forgotten” or overlooked, by many physicians.

When recognized early and treated with appropriate aggressive medical and surgical therapy, death from jugular vein thrombosis is uncommon today.

By being aware of jugular vein thrombosis, the physician can be more vigilant for potential complications and diagnose and treat them earlier.

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Abstract – WCN 2013

No: 1625

Topic: 36 – Other topic

The applications of 7T H-MRS in detecting GLU metabolism during the depression mechanism

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Objective: 7T MRS is used to observe the correlative change about GLU metabolism, and uncover the effects of the metabolic disorders of GLU in depression.

Method: SD rats were used in the maternal deprivation (MD) model, 20 adult mice (10 MD) were used to test their behavior and MRS analysis. The correlation behavior tested includes: sugar water test, open-field and water maze test. 7T MRS, surface coil and an ultra short echo time stimulated echo acquisition (STEAM) pulse sequence were used to test the correlative change about GLU metabolism (GLU, Gln, GABA and so on). The quantitative determination of metabolite was observed by LC Model and the SNR was improved by tuning experimental parameters. Meanwhile, body coil (T2WI) was used to test if the volume of hippocampus changes.

Results: The adult mice exhibit more depression, less pleasant sensation (sugar water test) and worse spatial memory (water maze) compared with the controls. In addition, the MR spectrums of their prefrontal lobes and hippocampus show that they have minor hippocampus volume compared to the controls.

Conclusions: The outcomes of MR spectrum studies about early life stress are not uniform. MRS, as a non-invasive measurement, can detect the metabolite changes in the brain. During the experiment, it can be demonstrated that GLU, the excitatory neurotransmitter in central nervous system, plays an important role in neuronal activities, such as memory formation and information processing. Furthermore, possible mechanisms of antidepressant drugs' effects are revealed by testing GLU content in the prefrontal lobes and hippocampus quantitatively.

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Abstract – WCN 2013

No: 973

Topic: 36 – Other topic

Evaluation of intellectual development in children with congenital heart disease

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Background: Nervous system in congenital heart diseases (CHD) has always attracted the attention of pediatricians, surgeons and neurologists. According to most researchers with cyanotic CHD type shows more diverse changes CNS than the evils of “white” type.

Objective: To study the cognitive function in children with cyanotic heart disease before and after surgery in recovery.

Material and methods: A total number of 97 children aged between 4 and 14 years with cyanotic congenital heart disease.

To assess the level of intellectual development, D. Wechsler's children version was used, which consist of tests adapted for preschool (WPPSI) and school children (WISC) for the Uzbek-speaking children.

Results: The study of cognitive function before surgery showed reduced performance on non-verbal intelligence subtests (65.6%), than on verbal subtests (82.4%). Children experienced some difficulty in carrying out the following tests “Labyrinth”, “Coding”, “Folding the figures”, “Finding similarities”, and “Sentences”.

Thus, suffered more memory, attention, spatial analysis, and thought process that manifest with difficulty in performance of test “Cubes of Kohs” (59.4%). It should be noted that the decline of intelligence is prevalent among patients with seizures (44.9%).

Conclusion: Hemodynamic findings in the brains of children with CHD lead to cognitive impairment, as a reduction in attention and memory, spatial analysis and cognitive functions. In the postoperative period after 1 month period was significant cognitive decline as a result of prolonged cardiopulmonary bypass and the impact of drugs for anesthesia.

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Abstract – WCN 2013

No: 1652

Topic: 36 – Other topic

Primary mitochondrial DNA mutations and probable variants in Leber's hereditary optic neuropathy

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Introduction: Leber's hereditary optic neuropathy (LHON) is one of the most common mitochondrial disorders, which mainly leads to vision loss in young males.

Objective: To describe the clinical manifestations of Leber's hereditary optic neuropathy (LHON), and to determine the genetic mutations responsible for these phenotypes.

Materials and methods: We report the cases of three patients suffering from progressive visual loss. In these patients we have performed genetic study searching for three primary mutations in mitochondrial DNA (m.11778G.A, m.3460G.A and m.14484 T.C).

Results: Two men and one woman have been addressed for a visual loss with optic atrophy. The age varied between 29 and 32 years. The neurological examination was normal except visual disturbances. Visual evoked potentials showed a prolongation of P100 wave. Brain MRI was normal in all patients. We found the point mutation (m.14484 T.C) in a single patient.

Discussion: One of three patients had a mutation. The other two may have other mitochondrial genes that are mutated and only reported in a minority of families.

Conclusion: mtDNA is highly polymorphic in nature with very high mutation rate, 10–17 fold higher as compared to nuclear genome. Identification of new mtDNA sequence variations is necessary to establish a clean link with human disease.

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Abstract – WCN 2013

No: 1517

Topic: 36 – Other topic

A case report: 53-year old Filipino with polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (poems) or Crow-Fukase syndrome

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Background: POEMS syndrome or Crow-Fukase syndrome is a rare multi-systemic disease caused by overproduction of vascular endothelial growth factor (VEGF), probably mediated by monoclonal proliferation of plasma cells. Most patients with POEMS syndrome commonly present with extremity weakness. The syndrome is also associated with monoclonal plasma proliferative disorder, along with organomegaly, endocrinopathy, and skin changes. The Mayo Clinic Experience identified only 170 patients to have POEMS syndrome.

Clinical presentation: A 53 year-old man who complained of 18 months duration of progressive weakness and numbness of both upper and lower extremities associated with hyperpigmentation of the skin, hypertrichosis, and bipedal edema. Predominant distal muscle weakness and atrophy was noted. Multiple large non-tender, cervical lymph nodes were noted. EMG-NCV showed polyneuropathy, sensory and motor diffuse, axonopathic and demyelinating, with signs of acute denervation and chronic denervation changes. The distal sensory and motor nerves of both upper and lower limbs were severely axonopathic. Hypothyroidism and impaired fasting glucose was noted and $\beta 2$ microglobulin determination was also elevated. MRI of the spine showed patchy areas of sclerosis involving the vertebral bodies of C3–C5, T2–T5 and T11.

After treatment with cyclophosphamide and prednisone improvement of motor function and neuropathic pain was noted. On follow-up he was already ambulatory with assistance and less dependent for his activities of daily living.

Recommendations: To our knowledge, there is only one published case of POEMS syndrome in the Philippines. Early recognition of POEMS syndrome is important to limit its morbidity, even if not all the features are present initially.

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Abstract – WCN 2013

No: 1653

Topic: 36 – Other topic

Curcumin nanoparticulate based neuroprotective approach for management of cerebral malaria

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Background: Curcumin is a polyphenol derived from the dietary spice turmeric and possesses diverse biological and pharmacological activities.

Objective: To investigate the potential of nanostructured lipid carriers (NLCs) loaded curcumin for management of cerebral malaria.

Materials and methods: Curcumin loaded NLCs (C-NLC) were prepared using microemulsion technique. They were characterized for surface morphology, particle size, zeta potential and *ex vivo* release studies in sheep nasal mucosa. Toxicity assessment was carried out by SK-N-SH cell lines, hemolytic activity and histopathological studies. Antimalarial efficacy was carried out in *Plasmodium berghei* ANKA murine model of cerebral malaria.

Results: The NLCs showed average particle size of 18–30 nm with zeta potential of -33.2 ± 0.75 mV. Transmission electron microscopy studies confirmed that NLCs were spherical in shape. *In vitro* hemolytic

activity and cytotoxicity value of C-NLC in SK-N-SH cell lines revealed that NLCs were non toxic by SRB assay. The *ex vivo* results indicated that release rate was biphasic and faster from drug suspension as compared to NLCs in simulated nasal fluid. The nasal ciliotoxicity studies were carried out using isolated bovine nasal mucosa which was further subjected to histopathological analysis. Results revealed that nanoparticles formulation had no harmful effect on the microscopic structures of nasal mucosa. The developed C-NLC showed significant antimalarial efficacy.

Conclusion: C-NLCs could be used as a potential treatment for cerebral malaria.

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Abstract – WCN 2013

No: 1607

Topic: 36 – Other Topic

Neuropsychological presentation of Ganser symptoms in a case of Chinese speaking corticobasal syndrome patient

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'Ganser syndrome' refers to a condition of cognitive decline that is characterized by 'near miss' answers to questions. It has been described in both psychiatric and organic brain diseases. However, distinct brain behavior correlation has yet to be defined. We describe a case of corticobasal syndrome with neuropsychological profile in keeping with episode of Ganser symptoms, and the characteristic of Chinese 'alexia' and 'agraphia' seen in this clinical entity. This case illustrates the importance of recognizing neuropsychological characteristic of Ganser symptoms, reemphasizes its association with neurodegenerative diseases, and raises question of role of basal ganglia and frontal-subcortical network dysfunction in producing Ganser symptoms.

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Abstract – WCN 2013

No: 1604

Topic: 36 – Other Topic

Fatigue in neurosarcoidosis

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Sarcoidosis is idiopathic, autoimmune, multisystemic disorder characterised by the presence of noncaseating epithelioid cell granulomas which can affect any organ. About 5–15% of patients with sarcoidosis have nervous system involvement – leptomeningeal and intraparenchymal infiltration of granulomas that lead most frequently to cranial nerve palsies, basal meningitis and endocrine dysfunction. Fatigue is experienced almost universally by autoimmune disease sufferers.

Fatigue can be also one of the symptoms of neurosarcoidosis. It is a disabling problem which decreases the ability of patients to perform previous routine activities and disrupts quality of life. The aim the study was to investigate fatigue in sarcoidosis. 50 sarcoidosis patients referred to the Neurology Outpatients' Clinic with symptoms indicating nervous system involvement were studied. All completed the Fatigue Assessment Scale (FAS). The majority (31–62%) reported fatigue (FAS ≥ 21). We evaluated whether fatigue can be connected with prednisolone usage, abnormal neurological signs, NMR, and pulmonary subtype.

The aim the study was to investigate fatigue in sarcoidosis.

50 sarcoidosis patients referred to the Neurology Outpatients' Clinic with symptoms indicating nervous system involvement were studied. All completed the Fatigue Assessment Scale (FAS). The majority (31–62%) reported fatigue (FAS ≥ 21). We evaluated whether fatigue can be connected with prednisolone usage, abnormal neurological signs, NMR, and pulmonary subtype.

10% of sarcoidosis patients with fatigue had abnormal neurological signs – abnormal sensation, positive Romberg's test, decreased hearing and nystagmus; 10 % had changes in the brain NMR scans – thickening of the dura mater – 3 or disseminates changes – 2, and 20% were treated with prednisolone. In order to determine whether there were any associations between fatigue and neurological examinations, NMR, prednisolone therapy and pulmonary sarcoidosis subtype statistical tests were used.

Our studies indicate that there is no statistical correlation between fatigue, abnormalities in neurological examination, NMR, prednisone therapy and pulmonary sarcoidosis subtypes. Fatigue is a common symptom in sarcoidosis patient which changes the quality of their life. Our study will be continued.

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Abstract – WCN 2013

No: 1684

Topic: 36 – Other Topic

An unusual case of neuro-Behçet disease with coma

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Background: neuro-Behçet disease (NBD) is reported between 5 and 15 % among patients with Behçet disease in different large series. CNS manifestations are usually classified as primary neural parenchymal lesions or non-parenchymal (secondary) lesions. MRI is the best method in demonstrating the CNS lesions. Mesodiencephalic junction, pontobulber brain stem, thalamus and basal ganglia are the most effected parenchymal regions. Diffuse involvement of all of these regions with hemispheric extension is a very rare condition. We present a case of NBD with widespread MRI lesions presented with coma.

Case: 35 years old male patient was admitted to the emergency clinic with loss of consciousness. In his medical history, he had diagnosed as having Behçet disease 3 years before admission with frequent oral ulcerations following uveitis in the right eye and arthritis. He had been hospitalized previously and diagnosed as having neuro-Behçet disease with left third cranial nerve palsy and mild quadriparesia. He was treated with steroids following with azothiopurine 100 mg/day. On his neurologic examination, he was in stupor and could not follow orders. He had no neck stiffness and with positive oculocephalic reflexes. On motor examination he had brisk tendon reflexes with bilateral Babinski sign. His MRI showed diffuse hyperintensity lesions extending from tegmental part of the pons to the bilateral midbrain, thalami, lenticular nucleus and right temporal lobe. The patient could not make recovery and died in his 30th day.

Conclusion: Exacerbation or "acute attack" of neuro-Behçet disease may involve diffuse parenchymal CNS lesion which may lead to coma.

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Abstract – WCN 2013

No: 1669

Topic: 36 – Other Topic

Radiological profile of patients of tubercular meningitis (Tbm) and its correlation with severity: A prospective study

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Background: Prospective data describing serial MRI changes and radiological predictors of clinical outcome have not been studied extensively.

Objectives: To access baseline MRI characteristics, serial MRI changes and their correlation with clinical profile in patients with TBM.

Method: 76 patients of untreated TBM were enrolled in 18 months. Detailed CSF examination and baseline MRI were done in all. Imaging was repeated after 3 and 9 months.

Results: Glasgow coma scale, TBM stage and high ADA level (>10) at initial presentation and hydrocephalus had significant association with final outcome. Use of VP shunt in patients with hydrocephalus improved the survival. Radiologically hydrocephalus, exudates and infarcts had significant association with the final outcome. Evidence of radiological improvement on final MRI correlated significantly with good clinical outcome. Tuberculomas, miliary mottling and border zone encephalitis had no influence on outcome.

Conclusions: Presence of hydrocephalus, exudates and infarcts in the baseline MRI are predictors of poor outcome, despite optimum anti tubercular therapy. Patients with hydrocephalus undergoing VP shunt have favourable outcome. Sizable proportion of patients may show initial worsening, however, evidence of improvement in final MRI is a predictor of good clinical improvement. Thus serial MRI studies of brain can aid in prognostication of illness in TBM.

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Abstract – WCN 2013

No: 1716

Topic: 36 – Other Topic

Myelitis and H1n1 vaccination

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Introduction: The influenza H1N1 vaccine has been used all over the world since the 2009s pandemics and got a significant public health value since then.⁸ There are several side-effects reported and it's also known that vaccination is related to auto-immune reactions as transversal myelitis.^{1,2,3,6,7} We report 2 cases of previously healthy young males with transverse myelitis following Influenza-H1N1 vaccination. Both cases had normal brain MRI but images of acute myelitis with T2 high signal and contrast enhancement. The patients were submitted to a 3–5-day pulse of methylprednisolone and evolved impressively well.

Discussion: The World Health Organization estimates 10 to 100 cases of post vaccinal acute adverse effects by 100,000 influenza H1N1 vaccinations. The most common severe neurological cause is Guillain-Barre syndrome. Transversal myelitis is a rare disorder.^{3,5,8} The important fact is that H1N1 vaccines produce an autoimmune mechanism and several cases of transversal myelitis have been presented. This may lead to a new pathway of investigation in autoimmune triggering.

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Abstract – WCN 2013

No: 1690

Topic: 36 – Other Topic

Anxiety as a consequence of hypokinesia and its correction by citicoline

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Citicoline (cytidine-5-diphosphocholine), a natural endogenous nucleoside in the biosynthesis of phosphatidylcholine, has shown beneficial effects in brain damage, caused by ischemic disorders. As restricted

movement activity – hypokinesia (HK) is an important risk factor for the development of mentioned disorders; the aim of present investigation was an evaluation of citicoline effects on neurobehavioral consequences after 15-days of movement restriction in rats.

White, inbred male rats have been used. As a model of HK, all experimental animals were kept individually in narrow cages for 15 days. Animal's behaviour was evaluated in "Elevated plus maze" test (EPM) after preliminary division of rats with high and low locomotor activity by 'Open field' stress exposure. Rats with low locomotor activity, which is characterized by general locomotor activity less than 10, were used in the experiments. Citicoline was administered at a dose of 12.5 mg/kg, i/p, twice a day.

The data obtained showed that after 15-days of movement restriction, main parameters characterized control group of rat's behaviour in EPM-test were decreased: open time (OT) for 44.6%, open entries (OE) for 46.9%, and central time (CT) for 64.5%. According to experimental data citicoline injection prevents and even increases OT more than twice, OE for 20%, CT for 12.2% in comparison with control.

Thus, our results evidence that citicoline could be used as an effective agent of healthy human pharmacology for the prevention of psychoneurological consequences of movement restriction – important problem of modern life.

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Abstract – WCN 2013

No: 831

Topic: 36 – Other Topic

White matter in aphasia: A historical review of the Dejerines' studies

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Background: Joseph Jules Dejerine (1849–1917) and his wife, Augusta Dejerine-Klumpke (1859–1927), were both involved in aphasia research. He is best remembered for his description of alexia without agraphia. She is also best remembered for her lecture at the aphasiology meeting in Paris in 1908. It remains unknown how their language processing theory was influenced by the combination of neuroanatomy and functional data from aphasia patients.

Objective: The objective was to describe the contributions of Joseph Jules Dejerine and his wife Augusta Dejerine-Klumpke to our understanding of cerebral association fiber tracts and language processing.

Material and methods: Systematic review of primary and secondary literature.

Results: The Dejerines (and not Constantin von Monakow) were the first to describe the superior longitudinal fasciculus/arcuate fasciculus (SLF/AF) as an association fiber tract uniting the Broca's area, Wernicke's area, and a visual image center in the angular gyrus of a left hemispheric language zone. They were also the first to attribute language-related functions to the fasciculi occipito-frontalis (FOF) and the inferior longitudinal fasciculus (ILF) after describing aphasia patients with degeneration of the SLF/AF, ILF, uncinata fasciculus (UF), and FOF. These fasciculi belong to a functional network known as the Dejerines' language zone, which exceeds the borders of the classically defined cortical language centers.

Conclusion: The Dejerines provided the first descriptions of the anatomical pillars of present-day language models (such as the SLF/AF). Their anatomical descriptions of fasciculi in aphasia patients provided a foundation for our modern concept of the dorsal and ventral streams in language processing.

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Abstract – WCN 2013**No: 1712****Topic: 36 – Other Topic****Somatosensory evoked potentials and brainstem auditory evoked potentials in assessment of brainstem dysfunction in multiple sclerosis. Comparison with MRI**

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Background: Brainstem lesions are common in multiple sclerosis (MS) and often lead to manifestation of neurological symptoms. According to some studies both Somatosensory Evoked Potentials (SSEP) and Brainstem Auditory Evoked Potentials (BAEP) give further valuable information about specific brainstem structures, in particular pathways of deep sensation and auditory pathways.

Objective: To examine the relationship between abnormalities in both SSEP and BAEP in MS and to study whether their impairment correlates with MRI brainstem lesions.

Material and methods: 40 controls and 30 patients with clinically definite MS were examined by both SSEP from median nerve stimulation and BAEP. All patients underwent brain MRI and EDSS assessment. Interpeak latencies (IPL), N9–N13 and P14 latencies were analyzed in SSEP. IPL I–III, III–V, I–V and amplitude ratios (AR) I/III, I/V, III/V, and also BAEP score were assessed in BAEP. Correlation analysis (Pearson) and forward logistic regression were used as statistical methods.

Results: Prolonged P14 latency (SSEP) correlated significantly with prolonged IPL N9–N13 (SSEP), prolonged IPLs and impaired ARs in BAEP ($p < 0.005$). We found statistically important relationship between MRI brainstem lesions and BAEP abnormalities – prolonged IPLs and impaired ARs ($p < 0.005$). No statistically significant correlation was found between MRI brainstem lesions and SSEP abnormalities. Higher EDSS score correlated with P14 and AR III/V abnormalities. Statistically significant parameters for predicting brainstem lesions availability were: BAEP score, IPL III–V, AR III/V, AR I/III and EDSS.

Conclusion: Performance of both SSEP and BAEP provides valuable information about brainstem function in MS. SSEP helps to confirm brainstem impairment in MS.

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Abstract – WCN 2013**No: 1725****Topic: 36 – Other Topic****Post-infectious syringomyelia as a consequence of neurobrucellosis**

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Introduction: The neurobrucellosis manifestations occur in about 5% of all brucellosis cases. They tend to be chronic, protean and a high degree of suspicion must be kept in mind for an early and accurate diagnosis.

Case report: A 60 year-old woman was observed with a two year history of anorexia and weight loss. She complained of cervical pain and progressive bilateral hearing loss and there was increasing difficulty in walking with diminished sensation on the four extremities. Constipation and urinary retention, albeit a common occurrence, were progressively worse. The neurological examination revealed bilateral hypoacusis, paraparesis with discrete spasticity, glove and stocking hypoesthesia, diminished reflexes except heightened knees reflexes and bilateral Babinski. The CSF examination disclosed elevated proteins (478 mg/dL), low glucose (24.0 mg/dL) and 160 leukocytes (75% mononuclear), positive Rosa Bengala and Wright (1:320) reactions and positive culture

of Brucella in the CSF. The brain and spine MRI were initially normal but displayed 4 months later, supratentorial white substance non-specific hypersignals and diffuse hypersignal from C2 to D9 with medullary enlargement on all extent of the lesion. The electromyography showed signs of polyradiculoneuropathy. The patient was treated with rifampicine, doxycycline and cotrimoxazol with a favorable outcome. Follow-up at 2 years kept showing the same medullary lesions in spite of good recovery, suggesting a post-infection syringomyelia.

Conclusion: The case described distinguishes himself by the unusual gathering of neurological symptoms with imagiological findings that persisted two years later, despite the favorable outcome of the patient.

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Abstract – WCN 2013**No: 1753****Topic: 36 – Other Topic****Sonographic evaluation of neuromas in patients with limb amputation**

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Background: Neuromas after limb amputation are common. Though high frequency ultrasonography (US) is very useful in the evaluation of peripheral nerves, there are a few reports about neuromas after limb amputation.

Objective: To investigate the utility of US in patients after limb amputation.

Patients and methods: We retrospectively analyzed three patients with limb amputation and evaluated clinical features and US findings, especially maximum cross sectional area (CSA) of nerves. For the US evaluation we used US systems equipped high-frequency (10–18 MHz) broadband probes.

Results: Patients were three males. The each sites of amputation were left shoulder, left forearm and right distal third leg. Clinically all patients showed phantom limb syndrome and one had sever stump neuralgia. In US findings, all demonstrated neuromas, maximum CSA of neuromas in patient with left shoulder amputation demonstrated 42 mm² at C4 and 80 mm² at C5 root in the upper part of brachial plexus, neuromas with right leg amputation showed 38 mm² at superficial peroneal nerve and 18 mm² at sural nerve near the stump, neuromas with left forearm amputation revealed 132 mm² at median and 59 mm² at ulnar nerve.

Conclusion: All patients with limb amputation revealed neuromas. US is very useful tool in the evaluation of neuroma after limb amputation. This information may be very useful for therapeutic approach.

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Abstract – WCN 2013**No: 1709****Topic: 36 – Other Topic****Time related changes in electrophysiologic findings of two patients with pons infarctions: Auditory startle reflex and transcranial magnetic stimulation parameters**

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Background: Enhancement of auditory startle reflex (ASR) was previously reported in cerebrovascular disease (CVD). In CVD exaggerated ASR may be due to disruption of corticoreticular

inhibitory fibers. Exaggeration of ASR offers an important potential in clinical rehabilitation of stroke patients since it may be useful for augmenting voluntary movements.

Objective: In this study we present two patients with pons infarctions whose various ASR and motor evoked potential (MEP) parameters were examined at the time of stroke onset and 3 months later.

Patients and methods: Two male patients, aged 40 and 34 with clinically and radiologically established pons infarctions underwent neurophysiological examinations on the acute stage of CVD and 3 months later on follow-up. ASR parameters were recorded bilaterally in orbicularis oculi, sternocleidomastoid, biceps brachii and abductor pollicis brevis muscles and MEP and cortical silent period (CSP) characteristics were noted.

Results: We have observed that in our stroke patients, characteristics of ASR, MEP and CSP showed changes over time. Patients with right and left pons infarctions had fairly similar electrophysiological results at disease onset and follow-up. On chronic stage in both patients total ASR probabilities were increased, muscles that had no reflex response in acute period showed marked reflex responses, MEP and CSP became apparent.

Conclusion: Our results suggest that enhancement of startle reflex and changes in MEP and CSP might be an indication of bulbar and cortical reorganisation and plasticity after brain injury.

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Abstract – WCN 2013

No: 1787

Topic: 36 – Other Topic

Plasmapheresis in Susac syndrome

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Background: Susac syndrome is a rare auto-immune endotheliopathy with a classic triad of encephalopathy, branch retinal artery occlusions and hearing loss. Consensus about the optimal treatment choice is lacking. Immunosuppressive treatment options include steroids, methotrexate, mycophenylate, cyclosporine, and intravenous immunoglobulines.

Objective: Evaluation of plasmapheresis treatment in Susac syndrome.

Patients and methods: Case report of 2 female patients (36 and 55 years old) with a follow-up of 24 and 7 months.

Results: Patients A and B suffered from encephalopathy, branch retinal artery occlusions, hearing loss (A) and gait disturbances (B). In the acute phase, patient A received high pulsed dose (1000 mg) IV methylprednisolone during 5 days with slow tapering. Patient B received oral methylprednisolone (64 mg OD) with slow tapering. Because of a lack of any beneficial effect, both patients subsequently received plasmapheresis for 2 weeks, both with significant clinical improvement, followed by a maintenance treatment with mycophenylate (1000 mg BID). Because of bipolar psychiatric symptoms in patient A 15 months after disease onset, mycophenylate was changed to plasmapheresis. Two months after symptom-onset patient B had a relapse of gait problems and encephalopathy and plasmapheresis was also restarted, again with beneficial outcome.

Conclusion: Optimal treatment choice in Susac syndrome is controversial because of the low prevalence of the disease and a lack of clinical trials and observational studies. Compared to monotherapy with steroids and mycophenylate mofetil, plasmapheresis showed better control of symptoms in our patients, both in the acute phase and as a rescue therapy in the chronic phase.

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Abstract – WCN 2013

No: 1704

Topic: 36 – Other Topic

A novel Csf1r mutation in hereditary diffuse leukoencephalopathy with spheroids

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Background: The differential diagnosis of adult-onset leukoencephalopathy is very broad. A brain biopsy is often needed when no definite diagnosis can be made based on a biochemical analysis and imaging. Hereditary diffuse leukoencephalopathy with spheroids (HDLS) is a rare autosomal-dominant leukoencephalopathy with heterogeneous phenotype and variable age of onset caused by a mutation in the colony stimulating factor 1 receptor (CSF1R) gene.

Objective: To present the clinical picture and a novel CSF1R mutation in a new case of HDLS.

Material and methods: Case report.

Results: A 45-year-old woman presented with a pain syndrome over the left hemicorpus. Clinical examination showed hypertonia of the left arm and leg with pyramidal signs. Subsequently, a severe cognitive decline and a painful dystonia of the left hand developed and she became bedridden. Brain MRI showed extensive confluent periventricular and subcortical white matter lesions without contrast-enhancement. After an extensive diagnostic work-out a definite diagnosis could not be made and a brain biopsy was performed. Pathological examination showed leukoencephalopathy and giant neuroaxonal swellings (spheroids), compatible with a diagnosis of HDLS. Molecular genetic analysis revealed a novel mutation in the CSF1-R gene (c.2466G>A, p.Met822Ile). As a curative treatment is lacking at this moment, our patient deteriorated progressively and eventually died less than 3 years after the onset of the disease.

Conclusion: HDLS is a rare autosomal-dominant inherited neurodegenerative disorder with leukoencephalopathy. Brain biopsy and genetic analysis are needed to confirm the diagnosis. To date only mutations in the CSF1R gene have been linked to this disorder.

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Abstract – WCN 2013

No: 1215

Topic: 36 – Other Topic

Current status and scope of international electives in neurology training programs: A survey of U.S. and Canadian Academic Program Directors

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Objective: To ascertain the number, pattern, and uptake of opportunities for international medical electives by residents and fellows in U.S. and Canadian postgraduate neurology programs.

Background: International medical exchange programs have multiple benefits to neurology trainees and their home institutions including the provision of humanitarian aid, new learning and teaching opportunities, clinical independence, reputation, and a better-educated global health workforce.

Methods: A survey was electronically distributed to all program directors in the U.S. and Canada by the American Academy of Neurology (Oct. 2012–Feb. 2013) to assess the training opportunities, institutional partnerships, and support available to postgraduate neurology trainees to participate in electives in countries outside of Western Europe, Australia, Canada, and the U.S.

Results: A total of 143/234 (61%) program directors responded. Most (53%) programs allow global health electives; however, just 33% provide financial support and 55% state that less than 10% of trainees actually go abroad. Among programs that do not allow global health electives, 86% cited a lack of funding, 55% stated there were no formal programs or partnerships with international sites, and 31% perceived no interest by residents. Only 12% of programs solicit philanthropic donations. If funding were not an issue, 93% of program directors believed their residents would have time to participate in global health training.

Conclusions: In spite of high perceived interest, only half of U.S. and Canadian neurology training programs currently permit global health electives, and the number of trainees venturing abroad remains a minority of all trainees in all programs.

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Abstract – WCN 2013

No: 1782

Topic: 36 – Other Topic

Medication use by individuals with motor neuron disease

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Background: Motor neuron disease (MND) is a chronic condition of unknown etiology characterized by a progressive degeneration of motor neurons leading to weakness, skeletal muscle wasting, and respiratory failure.

Objective: The objective of this study was to quantify to the use of prescription medications by MND patients.

Methods: The Province of Alberta maintains a publicly funded, universally available health care system. A number of administrative databases are maintained and include all physician visits and virtually (>95%) all prescriptions dispensed. A case was defined as individuals receiving three or more services, within a 12 month period, with an ICD-9 code of 335.2. Prescription drugs dispensed were extracted for MND cases for the period January 1 to December 31, 2011. Drugs were grouped using the Anatomical Therapeutic Chemical (ATC) coding system.

Results: 426 MND cases (147 females; 269 males) were included in the study and 39.4% died in 2011. The mean age was 62.2 years (std = 14.6), while the mean duration of illness was 3.7 years (range: < 1 to 18.5 years) with no difference between the sexes for either. Within 2011, the median number of prescriptions filled was 20, and the median number of medication classes was 7. 13% of patients were using Riluzole. The most common drug classes used were analgesics, psycholeptics, psychoanaleptics, antiepileptics, angiotensin inhibitors, and ulcer therapeutics.

Conclusion: Individuals with MND are prescribed a larger number of medications. An understanding of medication use can help

to guide practice to reduce the risk of unintended medication interactions.

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Abstract – WCN 2013

No: 1758

Topic: 36 – Other Topic

Maybe all is not so Rose in cephalic tetanus

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Introduction: Cephalic tetanus is a rare localized form of tetanus with facial palsy the most of the time follows a wound to the face.

Observation: We report the case of a patient with facial palsy but with wound in the foot.

Discussion: The diagnosis of this localization of tetanus is based on some clinical criteria like trismus and a entrance door in face.

But what will happen if you have a patient who is not vaccinated with foot's wound then facial palsy?

Conclusion: Needs new guidelines revised in light of new knowledges in the field of neuroscience for tropical neurological diseases.

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Abstract – WCN 2013

No: 1254

Topic: 36 – Other Topic

Toxoplasma encephalitis in an immunocompetent diabetic patient

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Background: Toxoplasmosis may have severe consequences in immunocompromised patients but cerebral toxoplasmosis is very rare in immunocompetent patients. Herein we present a case of toxoplasma encephalitis in an immunocompetent diabetic patient.

Case: A 54 year old female was admitted with abrupt onset visual loss, weakness evolving to quadriplegia in two weeks, lethargy and high fever. She was diabetic. Neurologic examination revealed a stuporous patient with poor cooperation. Fundi were bilateral hemorrhagic. In cranial MRI DWI revealed diffusion restriction in the anterior part of the right lateral ventricle. Her antitoxo Ig G and Ig M antibodies were positive. A second MRI revealed hyperintense lesions in periventricular frontal areas, posterior part of the left cerebral hemisphere and corpus callosum. An ependymal lesion was seen in right lateral ventricle. Hyperintense lesions were observed in FLAIR sequences consistent with hemorrhage in posterior parts of the lateral ventricles as well as linear hyperintensities in the sulci. The second CT showed hemorrhage in the posterior horns of the lateral ventricles. CSF pressure was high and CSF was hemorrhagic. She was considered as toxoplasma encephalitis and immediate treatment was started, but the patient was unresponsive to treatment and died at the 6th hospital day.

Conclusion: Toxoplasma encephalitis is a very rare condition and the absence of classical abscess formation in MRI in immune competent patients makes the diagnosis very difficult. Toxoplasmosis encephalitis must be kept in mind in the differential diagnosis diabetic patients with encephalitic signs and symptoms.

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Abstract – WCN 2013**No: 1530****Topic: 36 – Other Topic****New gene mutations identified in recessive Charcot–Marie–Tooth Tunisian families**

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Background: Charcot–Marie–Tooth (CMT) disease is the most common hereditary neuropathy resulting from mutations in more than 69 genes, such as MPZ, GDAP1 and SH3TC2 genes, with a wide spectrum of phenotypic manifestations.

Objective: To describe the clinical spectrum and molecular characteristics of new mutations in MPZ, GDAP1 and SH3TC2 genes associated with CMT.

Patients and methods: We included CMT Tunisian families with new mutations. Clinical features and electrophysiological findings were studied.

Results: Three families with 7 affected members presenting with motor and sensory polyneuropathy were investigated. Phenotype in all affected individuals was characterized by an early-onset (2.6 years). Consanguineous union was found in two families. Three new mutations occurring in the MPZ, GDAP1 and SH3TC2 genes were identified, they were associated with autosomal recessive inheritance.

A novel MPZ gene mutation including non sense c.310311>G/p.Asp104GlyfsX18 mutation in a Déjèrine-Sottas Disease was found. Homozygous missense c.117+3A>G mutation in GDAP1 gene in twins leads to axonal neuropathy with pyramidal signs. Homozygous missense c.1969G>A/p.Glu657Lys mutation in SH3TC2 gene was responsible for demyelinating neuropathy with severe scoliosis.

Conclusion: The identification of new mutations associated with different clinical and electrophysiological presentations stresses the broad phenotypic spectrum with MPZ, GDAP1 and SH3TC2 mutations.

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Abstract – WCN 2013**No: 1214****Topic: 36 – Other Topic****Predictors of endothelial dysfunction in obstructive sleep apnea syndrome**

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Introduction: Obstructive sleep apnea syndrome (OSA) is a highly prevalent disorder, which is associated with increased cardiovascular morbidity. One of the most important mechanisms responsible for development of vascular diseases in patients with OSA could be atherosclerosis. Endothelial dysfunction is an initial process and a key component of atherogenesis. Using peripheral arterial tonometry (PAT) we investigated the relationship between reperfusion hyperaemia index (RHI), which is regarded as being representative of endothelial function, and potential vascular risk factors in population with suspected OSA.

Methods: We enrolled 31 patients (42.1 ± 11.7 years) with suspected OSA, without history of regular medication or history of cardiovascular disease. Besides gaining general clinical characteristics, we performed polysomnography, assessment of PAT, assessment of baroreflex sensitivity and blood tests.

Results: Significant inverse correlation was found between RHI and apnea–hypopnea index (AHI) ($r = -0.550$, $p = 0.001$), and between RHI and desaturation index ($r = -0.533$, $p = 0.002$). Positive correlation was found between RHI and minimal nocturnal saturation of blood with oxygen ($r = 0.394$, $p = 0.028$). In stepwise multiple linear regression analysis AHI was the only significant variable to determine RHI ($\beta = -0.522$, $p = 0.003$). We found no significant correlation between RHI and other sleep, clinical or biochemical characteristics. RHI in population with severe OSA (AHI above 30) was significantly lower than RHI in the rest of the population ($p = 0.012$).

Conclusion: AHI was the only significant independent predictor of impaired endothelial function expressed by RHI determined using PAT. RHI was decreased significantly in population with severe OSA.

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Abstract – WCN 2013**No: 1751****Topic: 36 – Other Topic****Effect of albendazole therapy in patients with seizure and radiological lesion consistent with neurocysticercosis: An open labeled randomized placebo controlled trial**

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Background: There are controversies in using albendazole in treatment of solitary cerebral cysticercosis.

Objective: To evaluate the effectiveness in resolution of solitary cerebral neurocysticercosis and seizure free status after one week of albendazole therapy.

Methodology: Randomized placebo controlled trial was done in sixty consecutive patients of solitary cerebral cysticercal lesions attending the Neurology Department at a tertiary hospital in Nepal during the time period November 2009 to December 2010. Treatment group was given oral albendazole and similar looking tablets were given to placebo group along with a short course of steroid and antiepileptics. Follow-up head CT scans were done at end of one, three and six months to look for resolution of lesion. They were followed up for any seizure recurrence during treatment.

Results: Out of a total of 60 patients (treatment $n = 28$ and control group $n = 32$), at one month follow-up complete resolution of cyst was observed in 25% ($n = 7$) of patients in treatment as compared to 15.6% ($n = 5$) in the placebo which was statistically not significant ($p > 0.05$). Resolution rate at 3 and 6 months follow-up was also not significant. There was also insignificant difference in seizure recurrence rate on follow-ups.

Conclusion: The present study showed that patients treated with albendazole therapy did not differ statistically in both the outcome measures in terms of complete resolution of the lesion and seizure free status.

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Abstract – WCN 2013**No: 1754****Topic: 36 – Other Topic****Blood flow velocity changes in anterior cerebral arteries during cognitive tasks performance**

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Objective: Transcranial Doppler sonography (TCD) enables monitoring of blood flow velocity (BFV) in basal cerebral arteries during different cognitive tasks performance with great temporal resolution. BFV changes during mental activity were monitored primarily in middle cerebral arteries (MCAs) and little is known about these changes in anterior cerebral arteries (ACAs).

Aim: To determine the effect of different cognitive tasks performance on mean BFV (MBFV) changes with hemispheric dominance assessment and to define the most suitable activation test for monitoring of MBFVs in ACAs.

Methods: 14 right-handed and 14 left-handed healthy subjects aged 20 to 26 were included in the study. BFVs in both ACAs were recorded simultaneously during performance of cognitive tasks designed to activate frontal lobes: phonemic Verbal Fluency Test (pVFT), Stroop tests (Stroop test with neutral, congruent and incongruent stimuli) and Trail Making tests (TMTA and TMTB) that were presented on a computer screen. The paradigm lasted approximately 65 min.

Results: A statistically significant MBFV increase was recorded in both ACAs during performance of all cognitive tasks. Statistically significant right ACA dominance was found during performance of pVFT and TMTB in both left- and right-handers. The most significant MBFV changes in both groups were obtained during performance of TMTB.

Conclusion: Our result addressed cognitive tests with great activation potential for monitoring of ACAs that might be used in distinguishing of healthy individuals and patients with neurovascular or neurodegenerative diseases.

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Abstract – WCN 2013

No: 1887

Topic: 36 – Other Topic

A clinical study of diabetes mellitus in dementia, Parkinsonism and cerebrovascular diseases

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Background: Diabetes Mellitus (DM) is a risk factor for vascular disease and Alzheimer disease (AD), but liking for sweets is a factor of good controlled Parkinson disease (PD).

Objective: We reviewed the epidemiology of diabetes in patients with dementia, Parkinsonism and cerebrovascular diseases (CVD).

Patients and methods: We identified a series of 3816 patients with or without DM, AD, Vascular Dementia (VaD), other neurodegenerative dementia except AD, PD, Dementia with Lewy disease (DLB) including Parkinson–dementia, non-demented CVD, AD with non-demented CVD at the Department of General Internal Medicine, Diabetes and Clinical Nutrition, and Neurology of Hirakata Kohsai Hospital.

Results: Diabetic incidence of the patients without dementia, with AD, with VaD, and with other neurodegenerative dementia was 13.8%, 33.3%, 41.0%, and 22.1%, respectively. Diabetic incidence of the patients without AD or Parkinsonism, with PD, and with DLB was 14.3%, 10.3%, and 16.7%. Diabetic incidence of the patients without CVD, with non-demented CVD, and AD plus non-demented CVD was 10.8%, 45.9%, and 46.3%.

Conclusions: Incidence of diabetes was not higher among the patients with PD and DLB.

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Abstract – WCN 2013

No: 1879

Topic: 36 – Other Topic

Atypical Bickerstaff's brainstem encephalitis: Clinical vigilance ensures favourable prognosis

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Background: Bickerstaff's brainstem encephalitis (BBE) is a rare autoimmune inflammatory disorder of the central and peripheral nervous system, often sharing common features with Miller-Fisher's and Guillain-Barré syndrome. It typically presents following an upper respiratory tract infection, with reduced consciousness, ataxia, symmetrical ophthalmoplegia, tetraparesis, and electrophysiological evidence of motor axon degeneration and central involvement.

Objective: To stress the importance of high clinical suspicion in regard to atypical BBE cases.

Patients and methods: A 19-year-old, previously healthy male presented to the Emergency Room with a 12-hour history of drowsiness, unsteadiness, and upper extremity tremor. 10 days before, he was diagnosed and treated for GI tract infection. Physical examination revealed severe truncal ataxia, lower-extremity areflexia, right abductor ocular palsy, horizontal nystagmus, dysarthria and hoarse voice. He then developed psychomotor agitation and verbal aggression. Repeated laboratory (serum and CSF), radiological (MRI) and electrophysiological (EMG-magnetic stimulation) tests were negative. However, anti-GM1 and anri-CQ1b antibodies were strongly positive. The patient was readily treated with a 5-day course of intravenous immunoglobulin, displaying prompt and complete remission of his clinical picture.

Conclusion: BBE is a rare neurological entity, of which the diagnosis relies on high clinical suspicion, exclusion of alternative diagnoses, and supportive laboratory evidence. Atypical symptoms, and/or absence of electrophysiological and radiological findings should not rule out the diagnosis. Hence, a great clinical vigilance is required, as prompt delivery of treatment ensures the favourable prognosis of the disease.

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Abstract – WCN 2013

No: 1892

Topic: 36 – Other Topic

Patterns of autonomic nervous system impairment following concussion

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Background: The presence of significant autonomic nervous system (ANS) impairment following concussion has only recently been recognized. We sought to describe the pattern of ANS dysfunction in patients with persistent symptoms following concussion.

Objective: To define the patterns of ANS dysfunction on standard autonomic testing, and to determine whether these patterns correlate with clinical features in patients following concussion.

Methods: We reviewed the clinical features, laboratory studies, and findings on autonomic testing in a series of 22 consecutive patients suffering from concussion.

Results: All patients suffered from headache, postural lightheadedness was present in 11 of 19 patients queried, and 3 patients reported a history of syncope. Autonomic testing was abnormal in all patients, with nearly all patients having an excessive heart rate increment on head-up tilt (mean 54), and excessive oscillations in blood pressure.

Sudomotor function was impaired in 2 patients, cardiovagal function abnormal in 4, and orthostatic hypotension was noted in 1 patient. Baroreflex adrenergic sensitivity was abnormal in all but 1 patient tested and was severely reduced in 9, and excessive in 3. The autonomic findings did not correlate with headache severity, number of concussions, time from concussion to evaluation, or postural catecholamine measurements.

Conclusion: Autonomic testing in patients with persistent symptoms following concussion revealed ANS impairment in all, with features suggesting adrenergic insufficiency in most patients studied and excessive adrenergic ANS activation in a minority of patients. This pattern of impairment did not correlate with clinical features in this cohort of patients.

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Abstract – WCN 2013

No: 1897

Topic: 36 – Other Topic

Comparative neuronal uptake and cytotoxicity of anti-Hu and anti-Ri antibodies in rat cerebellar and hippocampal slice cultures

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Background: Anti-Hu and anti-Ri are paraneoplastic autoantibodies recognizing intracellular antigens present in essentially all neurons. At autopsy, brains of patients with anti-Hu antibody show neuronal destruction. In contrast, anti-Ri antibody is less clearly associated with neuronal death, and patients with anti-Ri antibodies may respond to treatment. We have demonstrated that anti-Yo antibodies, associated with paraneoplastic cerebellar degeneration, accumulate intracellularly in cerebellar Purkinje cells in slice cultures of rat cerebellum and that antibody accumulation is followed by cell death. The present study was conducted to determine whether anti-Hu and anti-Ri antibodies are also taken up by neurons and whether uptake of either antibody is cytotoxic.

Objective: To evaluate neuronal uptake and cytotoxicity of anti-Hu and anti-Ri antibodies in slice cultures of rat cerebellum and hippocampus.

Materials and methods: Rat cerebellar and hippocampal slice cultures were incubated with anti-Hu or anti-Ri antibodies and evaluated over time for antibody uptake and for cell death. Specificity of anti-Hu cytotoxicity was confirmed by adsorbing anti-Hu IgG with recombinant HuD protein.

Results: Anti-Hu and anti-Ri antibodies accumulated in cerebellar and hippocampal neurons. Anti-Hu antibodies produced cell death which was significantly reduced by adsorption of anti-Hu IgG with recombinant HuD protein. In contrast, neurons accumulating anti-Ri antibodies showed no evidence of cell death as compared to controls.

Conclusions: Anti-Hu and anti-Ri antibodies entered and accumulated in cerebellar and hippocampal neurons. Anti-Hu antibody associated neuronal death involved reactivity with HuD protein. Anti-Ri antibody did not affect neuronal viability and may cause neuronal dysfunction rather than cell death.

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Abstract – WCN 2013

No: 1902

Topic: 36 – Other Topic

An evaluation of the risks involved in ischemic encephalopathy among Mongolian children

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Background: Ischemic encephalopathy is one of the emerging issues among third world countries. Hypoxia seems to be the main cause of this problem, but many risk factors are associated in causation of hypoxic ischemic encephalopathy (HIE). The aim of our study was to evaluate these risk factors among Mongolian children.

Methods: This study was carried out at 3 children's hospitals, 85 neonates diagnosed with HIE over a year. Efforts were made to evaluate the questionnaire including details on parity, pre and postnatal histories, and specially the events of birth from labour till delivery of placenta.

Results: There was a remarkable decreased antenatal hospital visits and almost 52% reported that they visited just once or twice during the whole pregnancy period. In 21%, there was history of increased mother age above 40 years while 62% reported that some incidents of hypoxia occurred at the time of birth. 20% used drugs or medication during the course of pregnancy without the consultation of the physician. 31% had a history of prolonged 2nd stage of labour. 49% were delivered by unskilled birth attendants.

Conclusion: The data clearly showed that there are risks that can be avoided easily and a leading cause of mental retardation can be prevented. Lack of health awareness and decreased consultation from physicians on time seems to be the bulk of the problem in addition to lack of trained birth staff and health facilities. Efforts should be made to educate the mothers of child bearing ages, and counsel them to make the antenatal hospital visits more frequent as possible.

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Abstract – WCN 2013

No: 1907

Topic: 36 – Other Topic

Different emotions and strong stimuli: How do we make choices?

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Our understanding of the way the brain makes a decision is still incomplete. We decided to analyze the relationships between color and different emotions and sound and different emotions and their influences to make a choice. Forty-eight undergraduate students of marketing courses were interviewed. We presented 7 objects with different colors while they were listening to a “sad” song and again while listening to a “happy” song. In each case they had to choose one of them. Then we presented six identical objects associated with different sounds while listening to those songs. Forty-six percent of the students selected the red color during the sad moment (SM) and 40% during the happy moment (HM). None of them chose white during SM (however 10% selected white during HM). During HM the “forgotten” color was orange (2% of the students). Twenty-nine percent of the volunteers chose the object associated with the sound of a bomb during SM (the same sound was selected only by 12% during HM). Only 2% selected the sound of the wind (during SM or HM). Our results suggest that strong stimuli (visual or auditory) activate many neural circuits, making someone choose a red object or one associated with the sound of a bomb. Nevertheless a so called

strong color and a strong sound do not have the same significant influence when the person is happy (strong emotion). Therefore a positive activation of the limbic system (happiness) may inhibit visual and auditory hyperexcitability. This is important to make a decision.

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Abstract – WCN 2013

No: 1860

Topic: 36 – Other Topic

A fast method for determination of the parameters of pulsed-CEST MRI

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Objective: The object of the research is to demonstrate a fast method for simultaneous determination of labile proton fraction ratio and exchange rate and other parameters of pulsed chemical exchange saturation transfer (pulsed-CEST) MRI.

Methods: Mathematical models have been developed to describe pulsed-CEST contrast. We simulated pulsed-CEST MRI in MatLab (MathWorks, Natick, MA) for Z-spectra and asymmetry spectra using matrix exponential algorithm. The number of repetition of the sequence is 256. Each Gaussian-shaped pulse was divided into 101 steps, and the evolution within each step was modeled assuming a constant amplitude. We also calculated the above spectra by using the fourth/fifth-order RKF(ode45 in MatLab; MathWorks, Natick, MA, USA) with an absolute error tolerance for each integration step of 10^{-6} , and compared with the results obtained by our method. Calculations were performed using MatLab on Pentium® Dual-Core CPU (2.2 GHz) with 2-GB RAM. There was a good agreement between them, and this method was much faster than the RKF method (by a factor of approximately 1000 in this study). We simulated that the error of the labile proton fraction ratio, exchange rate and other parameters less than 5%.

Results: This method will be useful for determination of labile proton fraction ratio and exchange rate and other parameters, and for analyzing the pulsed-CEST contrast mechanism and/or investigating the optimal RF pulse parameters such as the RF pulse duration or pulse flip angle.

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Abstract – WCN 2013

No: 1859

Topic: 36 – Other Topic

Association between obstructive pulmonary function abnormalities and brain cortical thickness

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Background: Many epidemiological studies have suggested that chronic obstructive pulmonary disease is associated with cognitive dysfunction, mainly focusing on neuropsychological tests. A few brain imaging studies have been conducted, especially on cortical

thickness, in individuals with obstructive pulmonary function abnormalities.

Objective: We assessed the association between obstructive pattern of pulmonary function abnormality and brain cortical thickness using Magnetic Resonance Imaging (MRI).

Materials and methods: Among the individuals who participated in the medical examination at the Samsung Medical Center, 392 subjects who underwent T1-weighted MRI were included in the analyses after excluding those with technical errors, anatomical lesions on MRI and missing values. The cortical thickness (mm) was measured using Euclidean distance. Pulmonary function was categorized as normal, obstructive, restrictive, or mixed pattern, and those with restrictive and mixed patterns were excluded from the analysis. We performed multiple linear regression analysis adjusting for age, gender, the duration of education (years), smoking history (pack years), drinking status (never, former, and current), the history of hypertension, diabetes, and hyperlipidemia, and intracranial volume (10^{-3} mL).

Results: There was a significant difference in frontal cortical thickness between those with obstructive pattern of pulmonary function abnormality ($n = 14$; Mean, 3.05; SD, 0.10) and those with normal pulmonary function ($n = 378$; Mean, 3.12; SD, 0.11) ($p = 0.02$). Those with obstructive pattern had thinner frontal cortical thickness compared to those with normal pulmonary function ($\beta = -0.06$, $p = 0.02$).

Conclusion: The presence of obstructive pattern in the pulmonary function test was inversely associated with cortical thickness in brain MRI.

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Abstract – WCN 2013

No: 1837

Topic: 36 – Other Topic

SPG4 mutations in Brazilian patients with hereditary spastic paraplegia

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Background: Hereditary spastic paraplegia (HSP) is a heterogeneous group of neurodegenerative disorders characterized by progressive lower limb weakness and spasticity. There are 31 causative genes identified to date, but mutations in the *SPG4* gene are the major cause of autosomal dominant (AD) HSP in Europe/the USA accounting for 40–50% of all patients. Little is known about the frequency of *SPG4*-related HSP in Brazil.

Objective: To determine the frequency of *SPG4*-HSP among Brazilian patients with AD-HSP and to describe the mutational spectrum in this population.

Patients and methods: We recruited 34 unrelated families with AD-HSP from three university hospitals; patients underwent a complete clinical evaluation as well as detailed family history. Genomic DNA was extracted from lymphocytes and used in PCR reactions with primers designed to cover the 17 exons of the *SPG4* gene. Mutation screening was performed by automatic sequencing and multiplex ligand probe amplification (MLPA).

Results: We found eight previously described mutations in *SPG4*: c.839delAG (frameshift), c.1267G>T (missense), c.1378C>T (missense), c.1413+5G>A (splicing-site mutation), c.1495C>T (missense), c.1651G>C

(missense), c.1741C>T (nonsense) and c.1849T>G (nonstop). Three novel mutations were identified, each one in a single family: c.162C>T (splice-site mutation), c.1255G>T (nonsense) and c.1667delCA (frameshift). MLPA revealed no large deletion/duplication in these patients.

Conclusion: SPG4 mutations were found in 32% of the Brazilian families with AD-HSP. In this cohort, we found no large deletion/duplication in SPG4.

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Abstract – WCN 2013

No: 1821

Topic: 36 – Other Topic

Hypokalemic paralysis caused by de novo mutation of the Scn4a gene

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The aim of the case presentation is to point out the importance of de novo mutations as a cause of sporadic periodic paralysis.

32 year old, previously healthy male was admitted with sudden proximal tetraparesis. He had also weak trunk muscles, but his breathing was normal. Deep tendon reflexes and sensation appeared normal. Cervical MRT, ENMG and lumbar puncture were performed a couple of hours after admission with normal results. Blood test showed K⁺ 2.3 mmol/l. All other tests, including thyroid markers were normal. The patient received K⁺ replacement. After 9 h he was totally improved. Despite negative family history and late onset hypokalemic periodic paralysis was considered and his tests were sent for genetic analysis to GENDIA lab in Antwerp, Belgium.

Result of genetic test: a likely pathogenic heterozygous SCN4a: c.3404G>A variant was identified in exon 18 of the SCN4A gene by sequencing.

To prove the de novo origin of the variant analysis of the parents is planned.

Conclusion: The phenotype of sporadic periodic paralysis with de novo mutations could be very different. The reason for later onset and fewer attacks remains unclear. To provide a better understanding in this group of patients a larger study is needed.

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Abstract – WCN 2013

No: 1778

Topic: 36 – Other Topic

Cognitive impairment and depression in geriatric patients

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Background: The elderly often suffer from cognitive impairment (50%) and depressive symptoms (120.3%). These two pathologies have a bilateral connection and may influence each other.

Objective: To study the correlation between cognitive status and depression level of patients with encephalopathy of different genesis.

Material and methods: 29 patients with encephalopathy of different genesis were proposed to undergo a battery of tests which consisted of 2 blocks: cognitive – the Montreal Cognitive Assessment (MoCA), the Short Cognitive Performance Test (SKT) and emotional – Hamilton Rating Scale for Depression (HDRS). Inclusion criteria became MoCA score of ≤ 25 .

Results: According to the level of depression all patients were divided into 3 groups: without depression – 12 persons (410.4%),

mild – 10 (34.5%), moderate and high – 7 (24.1%). Patients without depression had average score of SKT 6.8, with mild depression – 4.3, with moderate and high depression – 7.3. In the group without depression was found a moderate negative statistically significant relationship between the scores of depressive symptoms and cognitive impairment ($\rho = -0.66 \pm 0.2$; $p < 0.05$). In the group with moderate and high depression was found a moderate positive statistically insignificant relationship (in this group it is necessary to increase the sample).

Conclusion:

1. Patients with encephalopathy of different genesis often suffer from depression (58.6%), which requires obligatory medical, psychological and social correction.

2. With increase of cognitive impairment depressive symptoms decrease in patients without depression. With increase of cognitive impairment depressive symptoms increase in patients with moderate and high depression.

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Abstract – WCN 2013

No: 1243

Topic: 36 – Other Topic

Connectivity changes after peripheral end-to-side coaptation following brachial plexus avulsion: a dynamic causal modeling (DCM) study

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Introduction: Following end-to-side coaptation of the phrenic to the muculocutaneous nerve using autologous nerve grafts the phrenic nerve has to perform a double function: independent control of breathing and elbow flexion. This results in activation of the diaphragm area with breathing but also with diseased arm flexion (Beisteiner et al. 2011). However, it is unclear whether this activation is driven by the SMA or other changes in connectivity.

Methods: One patient participated in this study, one year post surgery. fMRI was acquired while elbow flexion of the diseased arm or forced abdominal inspiration was performed. To assess effective connectivity changes with DCM, several classes of models with forward and/or backward or without connection between M1 and diaphragm were generated.

Results: Model comparison indicated the necessity of a forward connection between M1s of the arm and the diaphragm. Introducing a backward connection slightly increased the performance of this model. Within this model the instruction to perform arm flexion showed a strong enhancing effect on the forward connection of SMA to M1 arm (increasing M1 arm and M1 diaphragm activity) and a weak, but still significant positive effect on the SMA to M1 diaphragm connection (increasing M1 diaphragm activity).

Conclusion: Our results demonstrate the importance of a forward connection between the M1s of the arm and the diaphragm. We conclude that this specific cortical neuroplasticity as outlined by our final DCM model provides the neurophysiological basis for rehabilitation after peripheral end-to-side repair.

This study was supported by the Austrian Science Fund (P23611).

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Abstract – WCN 2013**No: 998****Topic: 36 – Other Topic****Beyond Kluver–Bucy syndrome: Colorful clinical picture of bilateral anterior cerebral artery infarction**

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Introduction: Kluver–Bucy syndrome, which is mostly caused by bilateral anterior temporal lobe lesions, is characterized by typical triad of hypersexuality, hyperorality and hyperphagia. Bilateral anterior cerebral artery (ACA) territory infarction is very rare and it has been reported even more rarely to cause Kluver–Bucy syndrome. **Case:** 51 year-old male patient was hospitalized with the complaint of difficulty in speech and left sided hemiparesis. Hemiparesis improved quickly. The neurological examination revealed apathy, loss of spontaneity and impoverishment of speech, loss of normal intonation, he was giving one-word answers to all questions. Diffusion weighted MRI images showed bilateral acute ischemic lesions in ACA territories, more prominent on the right side. Cerebral and cervical BT angiography showed aplasia of A1 segment of left ACA. In the follow-up, we have observed that the patient had hypersexuality, playing with his penis and talking inappropriately about women around; hyperoralism, taking all objects to his mouth; hyperphagia, trying to eat his wife's and other patients' meals after his own; disinhibition and utilization behavior. Detailed neuropsychological tests revealed prominent impairment in verbal and categoric fluency, motor sequencing (Luria's three step test), inhibitory control (go/no-go test), resistance to interference (Stroop test) and mild impairment in recent memory (enhanced cued recall). Visual attention and task switching (trail making A and B), abstract thinking and conceptualization were intact. The patient was then followed up under low dose olanzapine treatment and was getting better quickly.

Conclusion: Bilateral ACA infarction may lead to a colorful clinical picture including Kluver–Bucy syndrome triad.

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Abstract – WCN 2013**No: 1954****Topic: 36 – Other Topic****Immunopathological significance of ovarian teratoma in patients with anti-N-methyl-d-aspartate receptor encephalitis**

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Background: Although the clinical importance of ovarian teratoma in anti-N-methyl-d-aspartate receptor (NMDAR) encephalitis has been considered, investigations of ovarian teratoma in anti-NMDAR encephalitis remain limited.

Objective: To clarify differences in NMDAR distribution and lymphocyte infiltration in ovarian teratoma between patients with and without anti-NMDAR encephalitis.

Methods: Participants initially comprised 26 patients with ovarian teratoma. NMDAR distribution and lymphocyte infiltration in ovarian teratoma were examined using immunopathological techniques. Clinical, laboratory, and radiological data were compared between patients showing features of encephalitis. Anti-NMDAR antibodies in serum and cerebrospinal fluid were also measured in encephalitis patients.

Results: Neuronal tissues were obtained from ovarian teratomas in 22 patients (after excluding 4 patients who did not satisfy the inclusion criteria), and presence of NMDA receptor subunits was

revealed in all patients. The encephalitis group (n = 3) showed frequent lymphocyte infiltration, unlike the non-encephalitis group. In particular, dense B-lymphocyte infiltration near neural tissues was observed in the severe encephalitis group.

Conclusions: Differences in lymphocyte infiltration of ovarian teratomas between anti-NMDAR encephalitis and non-encephalitis patients suggest the immunological importance of ovarian teratoma as the site of antigen-presentation in anti-NMDAR encephalitis.

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Abstract – WCN 2013**No: 1970****Topic: 36 – Other Topic****Computer-based diagnosis of neurological and psychiatric illness in historical persons**

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Retrospective diagnosis of illness in historical figures is a popular pastime, but the paucity of detailed information about clinical features and disease progression often make its results less than fully reliable. Modern computer-based diagnostic programmes have been used to supplement historical documents and accounts, offering more objective approaches to the retrospective investigations of the medical conditions of historical persons. In the case of King George III, modern technology has been used to strengthen the findings of previous reports rejecting the popular diagnosis of variegated porphyria in the King, his grandson Augustus d'Esté, and his antecedents King James VI and King James I. Alternative diagnoses based on these programmes are suggested. The Operational Criteria in Studies of Psychotic Illness (OPCRIT) programme and the Young mania scale have been applied to the features described for George III and suggest a diagnosis of bipolar disorder. The programme SimulConsult was applied to Augustus d'Esté and suggests a diagnosis of neuromyelitis optica rather than acute porphyria as the leading diagnosis, with multiple sclerosis, as proposed by others, as a secondary possibility. James VI's and James I's complex medical history, clinical features and behavioural traits were also subjected to SimulConsult analysis; acute porphyria was rejected and the unexpected diagnosis of attenuated (mild) Lesch–Nyhan disease offered. We will review these approaches with regard to the methodology and validity. Textual analysis of the written and verbal outputs of historical figures indicates possible future developments in the diagnosis of medical disorders in historical figures.

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Abstract – WCN 2013**No: 1975****Topic: 36 – Other Topic****Prevalence of asymptomatic and symptomatic spondylotic cervical spinal cord compression**

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Background: Cervical spondylotic myelopathy (CSM) is the most frequent cause of the lower paraparesis in subjects above the age of 50. Magnetic resonance imaging (MRI) is able to detect asymptomatic

spondylotic cervical cord compression (ASCCC). The prevalence of both CSM and ASCCC increases with age, but the reason is not known.

Objective: To estimate the prevalence of both ASCCC and CSM in a general population above the age of forty.

Patients and methods: Fifty-nine randomly chosen healthy volunteers, recruited irrespective of the presence of signs of CSM, 37 women and 23 men, aged 65 (median), 42–82 (range) years participated in the study. All underwent MRI examination on a 1.5 T device using standard images and diffusion tensor imaging (DTI) at the level of maximum compression and at C2/3 as a reference. Subjects with MRI signs of cervical cord compression were examined clinically.

Results: MRI signs of cervical cord compression were found in 31 individuals (52.5%). Focal impingement was present in 6 cases (10.2%), and wide compression in 25 subjects (42.4%). The cross-sectional area at the level of compression <50 mm² was present in 2 cases and T2 hyperintensity in 2 subjects. DTI parameters showed no significant difference between subgroups with and without signs of compression. Clinical signs of symptomatic CSM were found in 2 cases (3.4%).

Conclusion: Prevalence of ASCCC in our group is higher than previously reported. The predictive significance of different types of compression remains to be established in future prospective evaluations of larger groups of subjects.

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Abstract – WCN 2013

No: 1757

Topic: 36 – Other Topic

Effects of hyperventilation on axonal excitability parameters in diabetic polyneuropathy

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Axonal excitability is a physiological concept connected with the characteristics of myelin and membrane of the peripheral nerve. Evaluation of axonal excitability provides important information about the peripheral nerve. It has not been one of the routine electrophysiological evaluations but suitable for use in experimental studies and helps in understanding pathological conditions and gives important information about pathophysiology. The mechanisms of diabetic polyneuropathy have not been understood clearly.

We used the short TROND protocol to measure the axonal excitability parameters in 11 healthy subjects and 7 patients with diabetes mellitus (DM) with polyneuropathy. The short TROND protocol was performed before and at the 20th minute of deep hyperventilation of healthy subjects and diabetic patients. Venous blood pH, pO₂, and pCO₂ values were recorded before hyperventilation and at the 20th minute of hyperventilation. The “hyperventilation score” was evaluated before and after the hyperventilation by asking the patient the degree of the numbness in the extremities and face or by seeing the carpopedal spasm. Although, significant changes in pH values occurred, the “hyperventilation scale score” of the DM group was lower than the score of the control group.

When the values of DM and control group after hyperventilation were compared, Ted (10–20 ms), Ted (40–60 ms), Ted (90–100 ms), Ted (peak) and superexcitability at 5 ms were statistically significant. There was a significant change in the hyperpolarization section of threshold electrotonus during hyperventilation in patients with DM.

DM patients have different responses to hyperventilation, and pH changes have different effects on diabetic polyneuropathy when compared with healthy subjects.

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Abstract – WCN 2013

No: 1703

Topic: 36 – Other Topic

A case of Hashimoto encephalopathy: Clinical presentation, neuroimaging and treatment

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Background: Hashimoto encephalopathy (HE) is a rare encephalopathy with positive antithyroid antibodies and normal thyroid function.

Objective: The objective of this study was to describe the clinical presentation, neuroimaging and treatment of Hashimoto encephalopathy.

Patients and methods: We report a 62-year-old female patient whose clinical presentation, neuroimaging, and response to steroid therapy fulfilled the diagnostic criteria of HE.

Results: The neuroimaging changes in this patient were multiple lacunar infarction in bilateral cerebral hemispheres. She was found with high serum anti-thyroid antibody concentrations. After high doses of corticosteroid therapy, her clinical symptoms were significantly improved. At the same time, the titers of serum antithyroid antibodies obviously decreased after steroid treatment.

Conclusion: This case illustrates the importance of considering rare but treatable causes of encephalopathy in a patient presenting with rapidly progressive cognitive dysfunction with/without the history of abnormality of thyroid function.

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Abstract – WCN 2013

No: 1946

Topic: 36 – Other Topic

Cross-border mobility of junior neurologists to and within the European Union: A major challenge for the health care system

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Background: There is a dynamic flow of medical doctors within and to Europe, particularly since the entry of more countries to the European Union (EU) in 2004 and 2007.

Objective: To assess interest and motives for cross-border mobility among residents and junior neurologists from member states of the EU and neighboring countries.

Methods: Questionnaire-based paper survey among 118 participants of a neurology course for young neurologists in 2011.

Results: Ninety-seven (82%) returned the survey. Most of them had at one point considered relocating within or to the EU for postgraduate education (87%) or employment (71%). Common motives were superior prospects for clinical training (85%), better resources at work and academic environment (both 80%), and higher income (70%). 52% of the surveyed would intend to return to their home country some time, yet 43% would not mind moving to another EU country. The attractiveness of the EU for migration is ranked over the United States, Australia/New Zealand or Switzerland (only 5% would instead prefer these options). The most common reasons which discourage cross-border relocation are the loss of family connection (53%) and uncertainty (40%), whereas language issues were less relevant (20%).

Conclusion: The interest of the upcoming generation of neurologists to relocate within and to the EU is strong. The driving forces are additional training and career opportunities as well as inadequate remuneration in certain regions. Therefore it is critical to take action to harmonize training and working conditions in Europe in order to ensure neurology service and patient care.

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Abstract – WCN 2013

No: 1920

Topic: 36 – Other Topic

Cotard's syndrome as an adverse effect of acyclovir treatment in renal failure

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Background: Aciclovir (ACV) is a well-known drug for treating herpes virus infections. ACV is generally considered safe and atoxic. However ACV-induced neuropsychiatric side effects (AINS) occur in about 1% of treated patients, mainly in patients with renal failure. We have earlier shown that AINS correlates to elevated levels of the ACV main metabolite, 9-carboxymethoxymethylguanidine (CMMG). AINS can present as confusion, agitation, visual/auditory hallucinosis and psychosis. In some patients, we have noted an acutely appearing anxiety with delusions of being dead, or variations thereof, which we interpret as equivalent to the rare Cotard's syndrome.

Objective: To pool reports of Cotard's syndrome in acyclovir-treated patients and to investigate the relation of symptoms to CMMG concentrations.

Patients and methods: We searched different sources – the PubMed, the Swedish adverse drug reaction database SWEDIS, and the Therapeutic Drug Monitoring (TDM) database – for symptoms of Cotard's syndrome in ACV-treated patients. Signs and symptoms were collated along with clinical and laboratory findings.

Results: We describe seven cases of ACV-treated patients with renal failure with what we interpret as Cotard's syndrome. In two cases the delusion was accompanied by an 'alien hand' variant of hemispatial neglect and in one case with Capgras delusion. In cases with known concentrations of CMMG, it relates to AINS and Cotard's syndrome.

Conclusion: In patients with impaired renal function, ACV treatment can result in AINS and Cotard's syndrome. High CMMG levels may be used as a marker for AINS.

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Abstract – WCN 2013

No: 1912

Topic: 36 – Other Topic

Atypical case of influenza vaccinated acute encephalopathy diagnosed by pathological approach. A case report and review of the literature

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Background: Influenza is one of the viral infections that cause epidemic or pandemic disease every year, and it is generally known as the most common and often causes serious complications. Preventive medicine for influenza achieved remarkable results by the spread of worldwide influenza vaccine. On the other hand, it may cause unusual neurological disorders such as acute encephalopathy by influenza vaccination. Influenza vaccinated acute encephalopathy (IVAE) is rare and its pathology is unclear.

Objective: We describe a 24-year-old male who presented with headache and difficulty of speech but no focal neurological deficits the day after influenza vaccination. 10 days later, generalized convulsive seizure presented and urgent hospitalization in neurology. Neurological examination presented amnesic aphasia. Furthermore, blood examination, viral cultures and CSF were negative. Brain MRI, DWI and FLAIR showed localized subcortical white matter hyperintensity area in the left inferior temporal gyrus. After hospitalization, convulsion attack became frequent and aggravated. Brain biopsy showed central chromatolysis around the Nissl bodies without inflammatory or demyelination changes. We started steroid pulse therapy after brain biopsy, and his neurological disorders and convulsive seizure improved immediately. Radiological examination improved and subcortical white matter lesion completely disappeared three months later with favorable outcome.

Conclusion: We concluded in this case that neuronopathy occurred due to toxic influence. Permeability obstacle of cell membrane and excitatory amino acid was excessively released triggered by the influenza vaccine. This case showed a different pathology from ADEM. IVAE is very important as influenza vaccination may have neurological sequels.

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Abstract – WCN 2013

No: 1910

Topic: 36 – Other Topic

Cytomegalovirus infection presenting with multiple low cranial nerve palsies

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Background: Cytomegalovirus (CMV) is a viral genus of the viral family known as herpes viruses. Although they may be found throughout the body, CMV infection is typically unnoticed in healthy people, but can be life-threatening for the immunocompromised patients. Diagnosis is usually done histologically by finding the inclusion bodies in infected tissue. We experienced one unusual case of CMV infection involving multiple cranial nerves.

Case: A 55-year old man had severe left eyeball pain, three months before admission. He then presented with throat pain, dysarthria and severe dysphagia with dehydration. On admission, neurological examination showed left seventh, ninth, tenth and twelfth nerve

palsies with severe pain in occipital area. He had uncontrolled diabetes mellitus and chronic otitis media. The CSF test revealed increase in white cell counts compatible with viral meningitis. HIV antigen test and brain MRI were negative. The initial diagnosis was herpes occipitocollaris. After treatment of steroid and acyclovir, the low cranial nerve palsy improved partially, but dysphagia and peripheral type facial palsy were persistent. One month later, he complained of abdominal pain, and colonoscopy with pathology found CMV colitis. He took gancyclovir and then the cranial nerve palsy disappeared, in addition to abdominal pain.

Conclusions: While spinal nerve root involvement by CMV may occur in around 1% of HIV patients, involvement of multiple cranial nerves is distinctly rare in healthy persons. The CMV infection must be considered in patients with low cranial nerve palsy.

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Abstract – WCN 2013

No: 2024

Topic: 36 – Other Topic

Differential diagnosis of vertigo using computer vertigometry data

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Background: Vestibular dysfunction is divided into: vertigo, dizziness, visual, psychogenic, and proprioceptive.

Objective: New method developed for detection of objective signs of vestibular dysfunction (vertigometry) in persons with vertigo paroxysms in history. Method is based on videostimulation of patient with panoramic image in horizontal plane, which is modeling rotatory vertigo.

Methods: We examined a group of 35 patients, 29 of them women and 6 men with complaints of dizziness in the anamnesis. An average age in the group was 51.2 ± 2.1 yr. Research group included diagnoses such as: 8 patients with position dependent peripheral vestibular syndrome, 12 patients with vestibular paroxysms in vertebral artery syndrome, and 5 patients with vegetative instability. At the moment of study patients did not complain of dizziness and no spontaneous nystagmus was registered. Vertigo attack was provoked under electronystagmographic control, rotatory vertigo direction and intensity were established using indicators of image scrolling while videostimulation and indicators of provoked nystagmus.

Results: It has been found that in patients with position dependent peripheral vestibular syndrome sensation of vertigo corresponded to stimulation presentation with velocity of $132.9 \pm 16.6^\circ/s$ and provocation nystagmus had velocity of slow phase $112.9 \pm 16.6^\circ/s$. In patients with vestibular paroxysms in vertebral artery syndrome sensation of vertigo corresponded to stimulation presentation with velocity of $66.4 \pm 16.6^\circ/s$ and provocation nystagmus had velocity of slow phase $48.5 \pm 16.6^\circ/s$.

Conclusion: The new method (vertigometry) allows finding objective signs of vestibular dysfunction in people with vertigo paroxysms in the anamnesis.

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Abstract – WCN 2013

No: 2040

Topic: 36 – Other Topic

Development of the Oxford Participation & Activities Questionnaire: Semi structured interviews with potential users

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Background: There is growing interest in the management of long term conditions and maximising the cost effectiveness of treatment, in part by keeping people active and participating in the community. The Oxford Participation and Activities Questionnaire (Ox-PAQ) initiative aims to develop and validate a patient reported outcome measure (PROM) for the assessment of participation and activity in patients experiencing a range of health conditions.

Objective: To gain the views of a variety of professionals on current PROMs and discuss the challenges and possible future developments that this area might face. Such views would be used to inform the on-going development of the Ox-PAQ.

Material and methods: Fifteen participants from across Europe with a range of expertise including regulation, commissioning, health economics, reimbursement, clinical practice, academic research, government and clinical trials, took part in semi-structured interviews.

Results: A range of views were expressed in relation to the appropriate length of PROMs, presentation of PROM data, use of PROM data and utility measurement. There was general agreement that existing measures have a number of limitations, including difficulty in interpretation, poor sensitivity to change and limited coverage across domains of health.

Conclusion: There is significant diversity in the professionals' views of PROMs. This is largely, but not exclusively, explained by the background of the individual whose views are being expressed. Developing an instrument such as the Ox-PAQ may benefit from such input, whilst at the same time recognising that a 'one size fits all' approach is unlikely to be successful.

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Abstract – WCN 2013

No: 1949

Topic: 36 – Other topic

Characteristics of horizontal positional nystagmus

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Background: Direction changing horizontal positional nystagmus is commonly observed in the context of horizontal canalithiasis, cupulolithiasis and central vestibular disorders.

Objectives: Our aim was to describe the spatio-temporal characteristics of 62 consecutive patients with symptomatic horizontal positional nystagmus.

Materials and methods: All patients underwent positional testing on the Epley Omniax Rotator with left and right ears down. Monocular video data was collected at 30 Hz and analysed offline. Nystagmus Slow Phase Velocity (SPV) was plotted as a function of time.

Results: Thirty-one subjects diagnosed with horizontal canalithiasis showed bursts of horizontal geotropic nystagmus with the affected ear down (onset: 0–1.33 s, duration: 11.7–47.9 s, peak SPV $79.17 \pm 67.0^\circ/s$). The SPV peaked at 5–10 s and dropped to 1.8% of the peak by 40 s. Nine subjects diagnosed with horizontal cupulolithiasis showed persistent apogeotropic horizontal nystagmus (onset: 0–4.36 s, peak SPV $54.21 \pm 31.82^\circ/s$ and $26.62 \pm 12.22^\circ/s$ with unaffected and affected ears down). At 40 s, nystagmus SPV had decayed to 81% and 65% of the peak for unaffected and affected ear down positions. Horizontal direction changing nystagmus was observed in 22 further subjects. 19 had Vestibular Migraine; in these patients persistent geotropic ($n = 5$) or apogeotropic ($n = 14$) nystagmus was recorded. Unlike the subjects with BPV, migraineurs had symmetrical peak SPVs (Left: $13.0 \pm 8.1^\circ/s$ and Right: $14.6 \pm 10.9^\circ/s$). At 40 s from onset, average SPV was 61% of the peak value. Three subjects with Meniere's Disease had persistent apogeotropic nystagmus.

Conclusion: The SPV profile of horizontal positional nystagmus can be useful in diagnosing canalolithiasis, cupulolithiasis and diverse central and peripheral vestibulopathies.

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Abstract – WCN 2013

No: 2033

Topic: 36 – Other topic

Ganglioglioma of the conus medullaris: A case report

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Background: Gangliogliomas are rare primary tumors of the CNS and are exceptionally found in the conus medullaris.

Objective: To present a case of ganglioglioma in the conus medullaris.

Methods: A 33-year old male presented with a 3-year history of morning low back pain, gait disturbances and increasing weakness of both legs. There were no bladder and bowel dysfunctions. Neurological examination revealed weak patellar reflexes, decreased ankle and plantar reflexes, numbness in L4-S1 dermatomes and normal strength of the group of hip muscles. The remaining muscle groups were graded 3 in the right lower limb and 4 in the left lower limb on a scale of 5.

Results: T1-weighted MRI disclosed expansive intradural mass at the Th12-L1 vertebral levels, with postcontrast heterogenous signal enhancement. Patient underwent Th12-L2 laminectomy, followed by maximal tumor resection. Microscopical examination revealed two components typical for gangliogliomas. The one was composed of irregularly oriented Synaptophysin, Neurofilament and Chromogranin A positive ganglion cells with large vesicular nucleolated nuclei and abundant eosinophilic cytoplasm. The other one, glial component, consisted of elongated bipolar glial fibrillary acidic protein positive pilocytic astrocytes. Rosenthal fibers, occasional granular bodies, conspicuous calcifications, and hyalinized blood vessels with typical perivascular lymphocytic cuffing were also observed. Proliferative Ki-67 index was low. Six months later, there was no tumor recurrence or change in neurological status.

Conclusion: Clinically and on MRI, conus medullaris gangliogliomas could be mistaken for more frequent ependymomas or astrocytomas. Accurate diagnosis depends on appropriate tumor sampling followed by detailed pathohistological and immunohistochemical analysis.

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Abstract – WCN 2013

No: 1738

Topic: 36 – Other topic

Chronic subarachnoid neurocysticercosis: A therapeutic challenge

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Introduction: Neurocysticercosis has a marked clinical pleomorphism, related to parasite number and location and host immune response. Parenchymal and subarachnoid forms are described. The latter is frequently resistant to cysticidal drugs and treatment protocols for these situations are lacking.

Case report: Fifty-six year-old female, with past history of partial epilepsy, admitted for persistent headache, in a context of intracranial hypertension. The investigation was compatible with neurocysticercosis.

In the following two years she was admitted six times for symptom recurrence, each with similar investigation results: CSF with hyperproteinorrhachia, hypoglycorrhachia, lymphocytic pleocytosis, both positive serologies and PCR for *Taenia solium*; brain and spinal-cord MRI showing loculation and cystic areas in the basal cisterns that extended to the cervical subarachnoid space and adhesive arachnoiditis in the lumbo-sacral region. In all admissions the same treatment was performed: albendazole 15 mg/kg/day for 15 days plus corticosteroids. Considering the symptomatic persistence despite several therapeutic cycles, a higher dose and longer duration of treatment with albendazole was attempted: 30 mg/kg/day for 90 days. There was a good tolerance to the treatment, aside from a slight increase in liver enzymes. Six months later there's a sustained clinical improvement. The CSF parameters have improved and the PCR for *T. solium* was negative for the first time. The imaging aspects remain identical, possibly corresponding to sequelae.

Conclusion: In this patient, there was a favorable response to the treatment with albendazole in a higher dose and superior duration, with good tolerance, possibly a safe and more effective therapeutic choice in these patients.

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Abstract – WCN 2013

No: 2016

Topic: 36 – Other topic

Role of ionic derangement and inflammatory factors in the development of hepatic encephalopathy following bile duct ligation

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Background: Encephalopathy and brain edema are serious central nervous system complications of liver failure characterized by various neurological symptoms. A growing body of evidence demonstrates that inflammation causes worsening of hepatic encephalopathy (HE) along with proinflammatory mechanisms that act synergistically with ammonia toxicity causing alterations in neurotransmission, leading to neuropsychiatric problems.

Objective: The study was designed to investigate the role of edema, ion shifts, neurotransmitters and inflammatory cytokines in the progression of HE.

Material and methods: HE was induced by bile duct ligation (BDL) surgery in adult male Wistar rats. Development of animal model of HE was assessed by routine liver function tests along with ^{99m}Tc labeled mebrotfenin hepatic biliary clearance test. BDL animals were compared with sham controls (SC) in terms of edema, Na, K, Ca levels, inflammatory factors (ELISA), neurotransmitters (HPLC) and neurobehavioral tests.

Results: A large fraction of ^{99m}Tc-mebrofenin was cleared from the liver of SC rats within 5 min, whereas ^{99m}Tc-mebrofenin was retained in BDL rats. Significant edema was observed in the cerebellum region which accounted for an increase in water content in whole brain. The tissue sodium and calcium concentration changed in a manner similar to that of water content. Preliminary data showed altered inflammatory cytokines, neurotransmitter levels following BDL along with considerable decline in the cognitive and motor capabilities of BDL rats.

Conclusion: Overall, prevention of ionic derangement, inflammation and restoration of neurotransmitter levels can provide new strategies for the prevention of HE.

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Abstract – WCN 2013**No: 2037****Topic: 36 – Other topic****Ethanol leads to poor impulse control in a T-maze based impulsivity task in rats**

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Background: Impulsivity often is accompanied by violent acts. Aggressive individuals are likely to experience general difficulties with impulse control and emotional regulation, and they show impaired social cognition. Several studies suggest that different neurochemical mechanisms can influence impulsivity, and that impulsive behaviour has no unique neurobiological basis. Alcohol intoxication is often associated with poor decision making, disruptive and irrational behaviour.

Objective: The present study investigated the effects of acute administration of ethanol on tolerance to delay reward in rats. Thus the possibility that alcohol may shift the choice towards the immediate reward was examined.

Material and methods: After acclimatization, different groups of rats were given the choice in a T-maze between two magnitudes of reward: Small (one food pellet) delivered immediately versus large (eight pellets) delivered after programmed delays (15, 30 and 60 s). After training trials different doses of ethanol which was diluted with water were administered IP in a volume of 5 ml/kg before impulsivity tests. In statistical analysis, the percent of animals that chose the arm baited with large reward were signified as response and evaluated compared to the development and sort of trials.

Results: Ethanol at a dose of 1.0 g/kg produced a significant decrease in preference for large and delayed reinforcer throughout the session, although there were marked individual differences in the size of the effect.

Conclusion: These results indicate that ethanol increases preference for the immediate reinforcer, which can be constructed as evidence of an enhancement in impulsive behaviour and reduction in self control.

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Abstract – WCN 2013**No: 2041****Topic: 36 – Other topic****Adrenomedullin, calcitonin gene-related peptide and urotensin may be involved in obstructive sleep apnea syndrome (Osas) in a complex manner**

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Background: Endothelial dysfunction may be an important component in pathophysiological mechanisms in obstructive sleep apnea (OSA) that is often accompanied by cardiovascular disease, hypertension and diabetes mellitus.

Objective: The aim of the present study was to determine if OSA cases differ from controls regarding the plasma levels of three vasoactive peptides; adrenomedullin (AM), calcitonin gene-related peptide (CGRP) and urotensin (U-II).

Patients and method: We assessed the plasma AM, CGRP and U-II levels in 39 OSA and 41 control cases. We evaluated the results

according to the presence and severity of OSA and to the associated medical conditions. Also, we evaluated if the plasma levels of these three peptides have an interrelation according to each other.

Results: Thirty out of 39 OSA cases were diagnosed as severe OSA. Mean plasma levels of CGRP and urotensin were significantly lower, whereas AM level was significantly higher in the OSA cases according to the plasma levels of the control group ($p = 0,005$, $p = 0,002$, $p = 0,017$, respectively). Only the mean plasma level of urotensin was significantly lower in severe OSA cases according to moderate OSA cases' ($p = 0.011$). The changes in plasma levels of AM, CGRP, and urotensin were significantly related to each other in OSA group.

Conclusions:

- i) CGRP, AM and urotensin may have an involvement in OSA;
- ii) significant interrelation of plasma AM, CGRP, and urotensin levels reveals the complex relationship of vasoactive peptides in this condition;
- iii) if these changes are causes or consequences is a question that should be confirmed in future studies.

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Abstract – WCN 2013**No: 1997****Topic: 36 – Other topic****Clinical data packages of drug approval for neurological diseases in Japan**

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Background: Many new drugs for neurological diseases have been approved in Japan since the foundation of Pharmaceuticals and Medical Devices Agency (PMDA) in 2004. However, it is difficult to plan and conduct clinical trials of neurological diseases because many rare and intractable diseases are included in this area.

Objective: To describe and analyze the drug review reports of neurological disease after the foundation of PMDA in 2004.

Methods: Drug review reports of neurological diseases from 2004 to 2012 were included. Three neurologists evaluated review reports independently. Target condition, clinical trial design for efficacy, primary outcome, and whether the drug was designated for orphan drug were abstracted.

Results: Forty-four review reports except for three diagnostic use drugs were evaluated. With regard to target condition, 8 drugs were approved for epilepsy, 6 for Parkinson disease, and 5 for spasticity or dystonia. In analysis of pivotal clinical trial design including Japanese for efficacy, randomized controlled trial (RCT) data was conducted in 30 drugs, open-label single-arm trial in 8 drugs. Eight reports out of 15 reports with orphan drug designation had RCT data of Japanese. Five drugs were approved based on the public knowledge-based application.

Conclusion: Many drugs were approved in various indications and clinical data package depended on the target condition. This analysis is useful and helpful in the drug development in neurological diseases.

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Abstract – WCN 2013**No: 1536****Topic: 36 – Other topic****Alternative splicing of canonical transient receptor potential type3 (TRPC3) ion channel expression in the human brain**

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Background: TRPC3 channels are nonselective cation channels in the TRPC subfamily. TRPC3 channels are activated by low cytosolic Ca²⁺ and G protein-coupled receptors (via phospholipase C–diacylglycerol signaling). TRPC3 channels are expressed throughout the central and peripheral nervous systems, and in particular associated with neuronal survival, development, and neurotransmission in the cerebellum. Recently, we have discovered a novel TRPC3 splice variant (designated TRPC3c) in rodents. TRPC3c isoform lacks exon 9, and has a much higher activity and cerebellar expression than the unspliced TRPC3b isoform.

Objective: To compare TRPC3c versus TRPC3b mRNA transcript expression in the human brain.

Methods: Total RNA was extracted from several post-mortem brain regions of 10 control subjects (donated to NSW Brain Bank Network). The cDNA obtained from reverse-transcription was amplified using SYBR-green qPCR reagent with primers specific to TRPC3b or TRPC3c isoforms. Amplicon templates of the isoforms enabled quantitative analysis of transcript levels.

Results: Among the brain regions, TRPC3c isoform represented from ~4% to 30% of the total TRPC3 mRNA level (TRPC3b + TRPC3c). The cerebellum had the highest relative proportion ($0.29 \pm 0.051, n = 8$), significantly greater ($p < 0.01$, ANOVA, Holm–Sidak multiple pairwise comparison) than the hippocampus ($0.067 \pm 0.011, n = 7$); midbrain ($0.036 \pm 0.0096, n = 5$); and pons ($0.075 \pm 0.047, n = 6$). The relative TRPC3 levels from the motor cortex ($0.11 \pm 0.03, n = 6$; $p = 0.116$) and medulla ($0.13 \pm 0.035, n = 2$; $p = 0.664$) were not significantly different from the cerebellum.

Conclusion: These findings show that TRPC3c expression is broadly significant in the human brain. The abundance and high activity of the TRPC3c subunit is compatible with significant roles in neuronal Ca²⁺ homeostasis, such as Ca²⁺-induced excitotoxicity during hypoxia.

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Abstract – WCN 2013**No: 1207****Topic: 36 – Other topic****Efficacy and biomarker findings from AVaglio, a phase III trial of bevacizumab plus temozolomide and radiotherapy in newly diagnosed glioblastoma**

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Background: Glioblastoma has a high disease burden and poor prognosis. AVaglio was the first double-blind, placebo-controlled phase III study evaluating bevacizumab in newly diagnosed glioblastoma.

Objectives: To evaluate efficacy, safety and potential biomarkers (plasma VEGF-A/VEGFR-2 prioritised).

Patients and methods: Patients received single-agent bevacizumab or placebo plus standard-of-care treatment (radiotherapy plus temozolomide) until progression/unacceptable toxicity. Co-primary endpoints were investigator-assessed PFS and OS. Secondary endpoints included health-related quality of life (HRQoL). Exploratory endpoints included correlative biomarker analysis, KPS, corticosteroid use.

Results: Baseline characteristics were balanced for intent-to-treat ($n = 921$) and biomarker-evaluable ($n = 571$) populations. Bevacizumab significantly prolonged PFS (HR = 0.64, 95% CI 0.55–0.74, $p < 0.0001$; median 10.6 vs 6.2 months) and delayed HRQoL time to definitive deterioration ($p < 0.0001$). Interim OS did not cross the threshold for significance (HR = 0.89, 95% CI 0.75–1.07, $p = 0.2135$). Functional independence (KPS $\geq 70\%$) was maintained during PFS in both arms (median bevacizumab vs placebo: 9 vs 6 months). Patients treated with bevacizumab had a diminished corticosteroid requirement. Patients with low and high baseline plasma VEGF-A concentrations derived similar PFS benefit with bevacizumab ($p = 0.610$): low VEGF-A (HR = 0.64, 95% CI 0.48–0.84) versus high (HR = 0.59, 95% CI 0.45–0.78). Similarly, PFS benefit in patients with low and high baseline VEGFR-2 levels was comparable ($p = 0.736$): low VEGFR-2 (HR = 0.54, 95% CI 0.41–0.71) versus high (HR = 0.66, 95% CI 0.50–0.87).

Conclusion: Addition of bevacizumab to standard-of-care therapy provided a significant clinically meaningful PFS improvement with stable/improved HRQoL/KPS and reduced corticosteroid requirement. No VEGF-A/VEGFR-2 predictive/prognostic effect was observed. Interim OS analysis did not cross the threshold for significance.

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Abstract – WCN 2013**No: 2103****Topic: 36 – Other topic****Flow cytometric analysis of cerebrospinal fluid is low diagnostic yield without atypical morphology or prior history of hematologic malignancy**

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Background: Flow cytometric analysis (FCA) of cerebrospinal fluid (CSF) has utility in detecting central nervous system (CNS) involvement by hematologic malignancies. Yet, the majority of samples are negative by FCA.

Objective: Identify pre-test characteristics of CSF specimens that will allow the rational use of FCA in the diagnosis of hematologic malignancy in CSF.

Design: Retrospective data was collected from the medical record and flow cytometric reports for all CSF samples submitted for FCA between 2007 and 2009.

Patients: 423 patients for whom 501 CSF samples were submitted for FCA.

Results: A positive diagnosis of a hematologic malignancy was made in 41 specimens (8.2%). The FCA-positive specimens showed atypical morphology, either blasts or atypical lymphocytes, in 98% of

specimens (40/41) on Wright-stained slides. There was a history of a hematologic malignancy in 89% of positive specimens (34/38). All FCA-positive specimens had atypical morphology or a history of hematologic malignancy. An enhancing brain lesion and radiologic leptomeningeal involvement were more common in FCA-positive cases while peripheral neurologic deficits were more common in FCA-negative cases. PCR for Epstein–Barr virus DNA was positive in two specimens. 406 specimens (81%) were negative by FCA. 7% of FCA-negative specimens (30/406) had atypical morphology and only 3% of FCA-negative specimens (12/404) had future positive CNS involvement within 30 days.

Conclusions: These data support a policy in which FCA of CSF is actively discouraged unless atypical lymphocytes or blasts are seen on Wright-stained slides or a history of hematologic malignancy is present.

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Abstract – WCN 2013

No: 2063

Topic: 36 – Other topic

Prevalence of restless legs syndrome in pregnancy and associated conditions in a Turkish population

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Background: Restless Legs Syndrome (RLS) is a common disorder that may severely impair sleep. According to the results of Turkish Adult Profile and Epidemiology of Sleep Study (TAPES), the prevalence for RLS was reported as 5.2% in general population, with a higher risk for women (7.3%).

Objective: The aim of the present study was to determine the risk of RLS and to find out its range according to the trimesters and associated demographic and medical conditions in pregnant women in a Turkish population. Local Ethics Committee approved the study and informed consents were obtained from all cases before inclusion.

Patients and methods: 241 pregnant women were included and given a face-to-face interview in the present study. The demographic features; past medical history; detailed history for the past gravidities, and the current pregnancy; sleep characteristics during the current pregnancy and the presence of RLS symptoms according to the International RLS Study Group criteria were recorded on a previously structured form.

Results: RLS was diagnosed in 94 out of 241 pregnant (39%). The RLS diagnosis was aforementioned in only two out of 94 RLS cases. Long pregnancy period and having an associated chronic disease showed statistical significance in RLS cases ($p < 0.001$, $p < 0.001$, respectively).

Discussion: In accordance with the previous reports from other populations, the present study revealed that RLS risk is remarkably high in the pregnant women, with an increasing frequency towards delivery. It also disclosed that RLS is widely underdiagnosed. Increasing awareness of health professionals may change this situation.

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Abstract – WCN 2013

No: 2112

Topic: 36 – Other topic

3D visualization of human spinal cord microanatomy demonstrated with contrast-enhanced x-ray microtomography

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Current methods for visualizing CNS microstructure in three dimensions depend mainly on reconstructions from serial sections, which are labor intensive and time consuming, or on methods such as confocal microscopy, which requires cleared samples of very limited size. Using a new combination of contrast staining and x-ray microtomography imaging (microCT), we have demonstrated high-resolution 3D visualization of neural microanatomy in whole slices of human spinal cord several mm thick. Staining the spinal tissues with potassium dichromate alone resulted in strong x-ray contrast between gray matter and white matter compartments, and allowed clear differentiation of Rexed laminae and the dorsal and ventral gray commissures. Similar contrasting and visualization were obtained with a simple inorganic iodine (Lugol) stain, which required only two days for 4–5 mm thick samples. Both methods are simpler and faster than Weigert-type stains. Using standard software, details and paths of axonal bundles entering the gray matter from the white matter could be clearly visualized in 3D. Tomographic imaging yields size-calibrated images, so that accurate in situ measurements of microanatomical features are possible. Because microCT imaging is non-destructive, the potassium dichromate samples can be post-processed for Weigert-type myelin stains allowing validation of the microCT data with light microscopy. These methods establish the potential of ex vivo microCT imaging for quantitative assessment of microstructure on spinal cord for a wide range of normal and pathological studies.

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Abstract – WCN 2013

No: 358

Topic: 36 – Other topic

A long-term evaluation of latency of pediatric brain tumors diagnosis in province of Delta-Egypt

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Background: Tumors of CNS represent the principal solid neoplasm in children and the second one after leukemia in frequency. Previous studies concluded that delay might exist between symptom onset and diagnosis (latency).

Objectives: This study was designed to assess the hospital based features of latency of pediatric brain tumor diagnosis in six main hospitals in the province of Delta-Egypt, as well factors that are important in predicting early clinical diagnosis.

Methods: From 2005 to 2011, 456 children with brain tumors were enrolled in this study. Clinical presentation, site, pathology and outcome of tumors were assessed. Pre-diagnostic symptomatic period (PSP) was analyzed as regard clinical presentations to make clear the common reasons of delay in diagnosis.

Results: 250 patients were male and 206 were female. Median age at diagnosis is 8.2 years with range from <1 year to 16 years. 60% of tumors were infratentorial. The most common tumors were low grade glioma (27.5%), medulloblastomas (25.7%), high grade glioma (15.3%) and craniopharyngioma (8.7%). The median PSP interval was

60 days. 35% of our patients were properly diagnosed within the first month of symptom onset. Medulloblastomas exhibited significant shorter diagnostic period in comparison to the other pathological subtypes.

Conclusion: Most frequent pathology in our experience was low-grade astrocytoma, then medulloblastoma. The appropriate diagnosis of pediatric brain tumors still generally requires months and frequent visits to various physicians. High level of awareness, a detailed medical history, and repeated and serious neurological sign interpretation should lead to earlier diagnosis and higher prospect of total tumor resection.

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Abstract – WCN 2013

No: 2050

Topic: 36 – Other topic

CPAP therapy reversed paradoxical weakness in myasthenia gravis with obstructive sleep apnea

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Introduction: Myasthenia gravis (MG) is a disease in which auto-antibodies against the acetylcholine receptor at the neuromuscular junction result in muscular weakness. Easy fatigability of the muscles and restoration of strength after rests are characteristic. Therefore, most patients with MG usually feel refreshed after nocturnal sleep. However, rare patients with MG complain morning weakness so called paradoxical weakness (PW). We report patient with OSA showing PW. CPAP therapy successfully terminated PW.

Case: Sixty-year-old obese woman with MG was admitted to the hospital because of general weakness, which was more severe in the early morning and improved slowly over a few hours of resting in awaking. She complained of weakness, especially in swallowing and eye-opening. She was diagnosed as MG and had a thymectomy five years ago. She took a fixed dose of pyridostigmine and azathioprine and 20–40 mg of prednisolone. In history, she had been loud snoring and witnessed apnea. In addition, she complained of recent body weight gain. In-lab polysomnography revealed severe OSA. Following successful CPAP therapy reversed PW within a week.

Conclusion: Sleep-disordered breathing is common in MG. We speculate that PW might be due to chronic exposure of repetitive hypoxia despite ongoing respiratory efforts during nocturnal sleep which resulted in accumulation of muscular fatigability in the morning. Weight gain secondary to chronic steroid therapy might be another one. It can aggravate the severity of underlying OSA or even lead to development of OSA. Early screening of OSA and adequate management such as CPAP therapy might be crucial to treat MG patients.

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Abstract – WCN 2013

No: 2092

Topic: 36 – Other topic

A case of primary Sjögren's syndrome presented with dysautonomia as initial manifestation

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Neurological involvement occurs in 10–42% of the primary Sjögren's syndrome (pSS). Distal sensory neuropathy is the most common, pandsautonomia without sensory neuropathy is extremely rare in the pSS as initial manifestation. We describe the case of pSS

presented with bladder dysfunction, paralytic ileus and postural intolerance. A 27-year-old woman developed paresthesia in the both arms 1 week before admission and progressed paresthesia in the legs and trunk below the chest with myalgia. At admission she couldn't urinate and defecate. On examinations she was revealed with abdominal distension, decreased deep tendon reflexes and sensation for the temperature in the extremities. Autonomic symptoms and signs included no bowel sound during auscultation, increased perspiration on the whole body, bladder distension and postural intolerance with normal pupil sizes and light reflex. Abdomen CT revealed urinary bladder distension and diffuse dilated colon without obstruction suspected paralytic ileus. She had positive anti-SS-A/Ro Ab, anti SS-B/La Ab, positive anti-nuclear antibody (titer 1:40, homogeneous pattern) and anti dsDNA antibody of 38.05 IU, but with negative rheumatoid factor, anti-Sm Ab and Anti-RNP Ab. Salivary scan revealed decreased activity of both the parotid glands and sublingual glands. Motor and sensory nerve conduction studies of median, ulnar, radial, tibial, peroneal and sural nerves are mild delayed terminal latency with normal amplitude and velocity. During months her symptoms recovered without steroid or IVIG. This case suggests that pSS should be listed in the differential diagnosis of acute autonomic failure as well as chronic form.

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Abstract – WCN 2013

No: 2156

Topic: 36 – Other topic

Paleocerebellar electrical stimulation prevents streptozotocin retinopathy

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Background: Oxidative stress and inflammation are implicated in the pathogenesis of retinopathy in diabetes. Fastigeal electrical stimulation (ES) alleviates experimental ischemic brain trauma.

Objective: To investigate effectiveness of electrical stimulation of paleocerebellum upon streptozotocin (STZ)-induced retinopathy in rats.

Methods: A group rats which received ES of culmen/pyramis lobules ES (0.5 s, 100 Hz, 0.25 ms) daily during two months from the moment of STZ administration, and another group received analogous stimulations of the parietal brain cortex. The retina was used to quantify oxidative stress and pro-inflammatory markers.

Results: Antioxidant capacity and the levels of intracellular antioxidant, GSH (reduced form of glutathione) levels were decreased by about 25–30%, and oxidatively modified DNA (8-OHdG) and nitrotyrosine were increased by 55–65% in the retina of diabetic rats. The levels of interleukin-1-beta (IL-1-beta) and vascular endothelial growth factor (VEGF) were elevated by 27% and 85% respectively, and the nuclear transcription factor (NF-kB) was activated by 1.7 fold. ES prevented diabetes-induced decrease in the antioxidant capacity, and increase in 8-OHdG and nitrotyrosine; however, it had only partial beneficial effect on retinal GSH. ES also inhibited diabetes-induced elevation in the levels of IL-1-beta, VEGF and NF-kB. Haematoxylin and eosin stained sections revealed outer reticular layer as well as inner nuclear layer thinning were prevented as well.

Conclusion: Paleocerebellar ES causes the beneficial effects of the metabolic and morphological abnormalities induced in the retina of rats treated with STZ.

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Abstract – WCN 2013**No: 576****Topic: 36 – Other topic****Neurological disorders in hospitalized HIV infected patients: Experience from a European Central Hospital**

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Background: Neurological manifestations of HIV infection are frequent and diverse. Combination antiretroviral therapy (ART) has changed HIV-associated manifestations including the majority of neurological disorders. Few studies in Europe have accessed pattern of NDs associated with HIV.

Objective: To characterize the occurrence of neurological disorders (NDs) in a European central hospital.

Methods: Retrospective case series of HIV hospitalized patients with NDs from an infectious disease service between 2004 and 2010.

Results: We identified 140 patients with NDs; the majority were males – 105 (75%); the mean age 42 years (range 19 to 89 years old). The most common manifestations were focal signs (55/39.2%), followed by confusional state (29/20.7%), seizures (23/16.4%), and cognitive deterioration (22/15.7%). Mean and median CD4⁺ count were 130 cells/mm³ and 71 cells/mm³ respectively. Mean HIV viral load was 232,434 copies/mm³. Frequent NDs were: cerebral toxoplasmosis (36/25.7%), progressive multifocal leukoencephalopathy (PML – 35/25%), cryptococcosis (12/8.6%), and HIV dementia (12/8.6%). Stroke, tuberculosis, meningoencephalitis, neurosyphilis and lymphoma were also among the NDs found. Few had more than one ND. Most (120/85.7%) had other associated severe nonneurological opportunistic disorders. Occurrence of NDs led to the diagnosis of HIV infection in 37 (26.4%). In-hospital mortality was 12.8% (18/140). The deceased patients had PML (6), toxoplasmosis (6), HIV dementia (3), stroke (2), and tuberculosis (1).

Conclusion: This case series illustrates the burden and diversity of neurological disorders in HIV patients. The relative high in hospital mortality is in alignment with the coexistence of severe neurological disorders with opportunistic diseases typical of advanced HIV infection.

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Abstract – WCN 2013**No: 2130****Topic: 36 – Other topic****Recording cervical vestibular evoked potentials on different muscle groups**

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Objective: Cervical vestibular evoked myogenic potential (cVEMP) is a short-latency myogenic response recorded over the sternocleidomastoid muscle (SCM) in response to saccular stimulation and is accepted to provide a non-invasive method of assessing vestibular-otolith function. The aim of this study was to try to make recordings by placing the active electrode on different muscle groups to investigate the origin of the potentials.

Methods: 20 healthy volunteers; 12 women and 8 men, with a mean age of 35 years were included in the study. The reference electrode was placed on the upper third of sternum and to record the surface EMG activity, an active electrode was first placed on the upper half of the ipsilateral SCM and then contralateral. Ipsilateral tibialis anterior muscle was selected as the third recording site. The volunteers were

asked to activate the muscles maximally during the recordings. Two stimulation consequences consisting of 250 sound stimuli were given. The acoustic stimuli were clicks at an intensity of 110 dBnHL of 0.1 millisecond duration, delivered at a frequency of 5 Hz through a headphone unilaterally to each ear. The latencies of peaks p13 and n23 and peak-to-peak amplitude were measured.

Results: In all individuals studied responses could be gathered from all the sites studied.

Discussion: Responses recorded from different active sites showed nearly the same features which are difficult to explain with the known electrophysiological criteria. Further studies involving different sites must be performed to be more confident about the origin of these potentials.

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Abstract – WCN 2013**No: 2162****Topic: 36 – Other topic****Subacute cerebellar degeneration associated with metabotropic glutamate receptor-1 antibodies: New paraneoplastic accompaniments**

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Background: Subacute cerebellar degeneration associated with metabotropic glutamate receptor type 1 (mGluR1) autoantibodies is an uncommon syndrome known to be part of the spectrum of paraneoplastic cerebellar degenerations associated with neuronal autoantibodies.

Objective: To report new paraneoplastic accompaniments of subacute cerebellar degeneration associated with antibodies to mGluR1.

Methods: Case report.

Results: We describe a patient with a history of mycosis fungoides who developed a subacute cerebellar ataxia. The ataxia worsened after the removal of a prostate adenocarcinoma. Autoantibodies specific for mGluR1 were detected both in the patient's serum and cerebrospinal fluid. The patient was treated with intravenous immunoglobulins (2 g/kg every month for six months) and steroids with improvement of the cerebellar ataxia.

Conclusions: To our knowledge, this is the first case of subacute cerebellar degeneration with mGluR1 antibodies associated with mycosis fungoides and prostate adenocarcinoma.

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Abstract – WCN 2013**No: 1876****Topic: 36 – Other topic****A case report of a congenital encephalon malformation and giant macrocephaly associated with hydrocephalus**

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Background: Congenital hydrocephalus is a common neurological diagnosis in newborns, with prevalence of 4.65 per 10 000 births. In 3.3% of cases there is a previous case in family history.

Objectives: Report a case of congenital giant macrocephaly associated with hydrocephalus and encephalon malformation.

Methods: J.G.P.G., male, 13-month-old, weight 15 kg, was admitted with progressive increase of cephalic perimeter (CP) since intrauterine

life, reaching 89.0 cm. The patient was born with CP: 35 cm, Apgar: 7, 9. Mother informs that the child presents tonic crisis 3–4 times/day, lasting 10 s, with diuresis. A respiratory discomfort is noticed depending on the position of the head. There is a similar family history, who died with 9 months.

Results: MRI shows the presence only of hindbrain and midbrain structures with extensive malformations. The cerebellum has a significant shift to the right and severe compression by the presence of fluid beneath the tent of the cerebellum. Only a small portion of the telencephalon, near the tent of the cerebellum and the occipital bone, is visible. The huge volume of the skull is due to the fluid accumulated. There was no closure of the cranial vault, with persistence of the fontanelles. The midbrain aqueduct has its diameter size noticeably reduced, which implies difficulty in CSF flow.

Conclusion: This is an unusual case of a congenital giant macrocephaly associated with hydrocephalus and encephalon malformation, which reflects its difficult treatment choice.

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Abstract – WCN 2013

No: 1729

Topic: 36 – Other topic

Blink reflex excitability abnormalities in multiple sclerosis

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Background: Brainstem dysfunctions in multiple sclerosis (MS) may be caused by either local brainstem lesions or changes in excitability modulation from supranuclear inputs. We hypothesized that while brainstem lesions lead to abnormalities in latency of BR (AbLat), supranuclear lesions lead to abnormalities in excitability measures (AbExc).

Objective: To assess abnormalities in BR excitability in MS patients and their correlation with MRI-supranuclear lesions.

Patients and methods: In 20 patients and 10 control subjects, we measured latencies and area of R1, R2 and R2c to single unilateral supraorbital stimulation of either side. We calculated the R2c/R2 ratio as a measure of asymmetric excitability enhancement. Patients were classified as having AbLat or AbExc when the values were above the mean + 2SD of our control group reference data. Brainstem and hemispheres (hLL) MRI-lesion load were measured with computer-assisted software (FSL). T-test and Spearman's test were used for statistics.

Results: AbLat were found in 8 patients (7 of them had pontomedullary lesions).

AbExc were found in 7 out of the remaining 12 patients. In these patients, a significantly larger R2c/R2 ratio ($p = 0.03$ with respect to controls) coincided with a significant enhancement of R1 amplitude ($p = .0001$ with respect to the contralateral side). They also showed a positive correlation between R2c/R2 ratio and ipsilateral hLL ($r = 0.357$), but only one had a lesion that compromised the trigeminal-facial circuit.

Conclusion: Asymmetric BR excitability enhancement correlates with unilateral brain lesions in MS. These tests add information for the functional evaluation of the effects of brain LL on brainstem circuits.

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Abstract – WCN 2013

No: 2141

Topic: 36 – Other topic

Homozygosity for P.Cys183ser mutation in Notch3 gene may influence the severity of clinical presentation? Report of an Italian family

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Background: Most of causative mutations of the cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) are missense mutations either creating or deleting one cysteine residue, inherited in a heterozygous state, in the NOTCH3 gene. Only two homozygous patients have been reported to date with minimal phenotypic peculiarities.

Objective: To report a homozygous patient comparing his clinical profile with five subjects throughout three generation of his pedigree, carrying the same mutation in heterozygosity.

Patient and methods: The index patient (IP) was a 44 year-old man, born from consanguineous parents. Other five relatives were examined. Personal and family histories, laboratory, neuropsychological and MRI assessments, skin biopsy (IP) and genetic testing have been collected.

Result: Symptoms started at 23 years in our IP, progressed with recurrent ischemic stroke. Diffuse leukoencephalopathy at MRI and a severe cognitive decline were also present. GOMs were detected in skin specimens and a homozygous p.Cys183Ser mutation of the NOTCH 3 gene was found. Among the heterozygous relatives, both parents developed stroke in advanced age and a sister was clinically asymptomatic at the same age.

Conclusion: Although homozygosity is not generally associated with worsened phenotype in autosomal dominant diseases, some differences may be noticed in clinical presentation or disease outcome. The case reported here would indicate that homozygosity is associated with an earlier onset of symptoms and a most severe clinical course compared to the heterozygous subjects of the same pedigree. However, we cannot exclude that other genes may influence clinical variability.

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Abstract – WCN 2013

No: 2143

Topic: 36 – Other topic

Neurologist brain-drain from Central-Eastern Europe: The effect on neurologist workforce in Hungary

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Background: Since joining the European Union (EU) the workforce of Central-Eastern Europe has become free to be employed in most West-European countries. Working conditions – including salary – are more favorable in West-Europe, where new EU regulations on working hours in healthcare have increased the demand for well-trained physicians. Therefore, a migration of neurologists was expected from formal communist countries of the EU to the West. We set forth to evaluate the effect of this process on recent changes in the number of active neurologists in Hungary.

Methods: The number of active neurologists was identified from the database of the National Health Insurance Fund – the only and universal state health insurance organization in Hungary – for 2009 and 2012. We selected only those board certified neurologists who reported the examination of more than 1 case per week (>52 cases per year) to exclude those who do not perform regular activity in

patient care. We analyzed the data in 5-year age categories of the neurologists.

Results: In 2009 the examination of >52 cases was reported by 827 neurologists. In 2012 this number decreased to 674. The drop of 153 neurologists included 21%, 24%, 27%, 22% and 27% in the 41–45; 46–50; 51–55; 56–60 and 61–65 years age groups, respectively.

Conclusions: The Hungarian healthcare system lost 22% of its active neurologists between 2009 and 2012. Retirement cannot be held responsible for this decrease, and other factors – including migration out of the country – are suggested to be in the background.

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Abstract – WCN 2013

No: 2178

Topic: 36 – Other topic

A case of isolated hypoglossal nerve palsy secondary to an internal carotid artery dissection

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An isolated hypoglossal nerve palsy is uncommon, dysfunction of the nerve usually occurs as part of a complex syndrome and therefore this case is unusual.

We present a 49 year old gentleman who noted altered sensation of the left side of his tongue when brushing his teeth; he denied all other symptoms apart from a mild headache. Examination only revealed an isolated left hypoglossal lesion with deviation of the tongue to the left on protrusion. Investigation with MRI and CT scans in the stroke unit was normal. The diagnosis was finally revealed with MRA imaging. The scan demonstrated images consistent with compression of the hypoglossal nerve and features of a dissecting left sub-cranial internal carotid artery aneurysm. Further imaging with a CT angiogram confirmed a 12 mm dissecting aneurysm at the high cervical segment of the left ICA. The consensus from the neurovascular MDT was to take a conservative approach, given the thromboembolic risk posed by the aneurysm, aspirin 75 mg daily was initiated.

This case, a young man with a combination of isolated hypoglossal nerve palsy and headache, highlights the need to consider all differential diagnoses affecting the hypoglossal nerve along its course from the brain stem nucleus to the carotid artery. The case demonstrated the importance of thorough clinical examination and history taking as well as collaboration with neuroradiology to reveal the diagnosis.

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Abstract – WCN 2013

No: 396

Topic: 36 – Other topic

Changes in dementia diagnostic category and diagnostic confidence after DaTSCAN™ imaging in subjects with possible Dementia with Lewy bodies

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Background: A clinical diagnosis of Dementia with Lewy bodies (DLB) has good specificity but low sensitivity and is particularly challenging in patients with an uncertain diagnosis (possible DLB). DaTSCAN™ (Ioflupane ¹²³I) is a radiopharmaceutical able to visualize the dopamine transporter receptors in the brain.

Objective: To evaluate the impact of DaTSCAN™ SPECT imaging on dementia diagnostic category and the diagnostic confidence of clinicians in patients with a diagnosis of possible DLB.

Methods: 187 patients with possible DLB were recruited from 21 centers in 6 European countries. Patients were randomized to have a DaTSCAN™ (n = 127) or to have no-imaging (n = 60). The proportion of patients with changes in clinical diagnosis (from possible DLB to probable DLB or non-DLB) and changes in the confidence of diagnosis was compared at 8 and 24 weeks of follow-up.

Results: More patients in the DaTSCAN™ group had a change in diagnostic category after 8 weeks (61% vs 4%; $P < .0001$) and 24 weeks (71% vs 16%; $P < .0001$) compared to the control group. Significantly more patients in the imaging group were given more confident diagnoses during follow-up. Clinicians were more likely to change the diagnostic category if the DaTSCAN™ image was abnormal (82%) than if the image was normal (48%).

Conclusions: DaTSCAN™ SPECT imaging significantly contributed to change of diagnostic category and improved diagnostic confidence, proving to be a useful adjunct in the diagnosis of dementia in patients with possible DLB. Changes in diagnostic category were less frequent in the control group despite a six-month prospective follow-up.

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Abstract – WCN 2013

No: 2177

Topic: 36 – Other topic

Voxel-based morphometry in Friedreich's ataxia: A prospective study

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Background: Friedreich's ataxia (FRDA) is the most common autosomal recessive ataxia, characterized by spinal cord and cerebellar damage. However, little is known about progression of such damage over the time.

Objective: To evaluate progressive gray and white matter (GM and WM) atrophy in FRDA.

Material and methods: Thirteen FRDA patients and 13 age-and-sex matched controls underwent a VBM protocol in a 3T scanner (Philips Achieva) two years apart. To examine differences, we used flexible factorial design assessing time × groups, using uncorrected height threshold of $P < 0.001$.

Results: Mean age of patients was 27.4 years. In FRDA, there was GM atrophy in right inferior temporal gyrus (peak MNI coordinates $x = 54, y = -6, z = -34.5$), left superior temporal gyrus ($x = -40.5, y = 13.5, z = -37.5$), left inferior temporal gyrus ($x = -51, y = -24, z = -33$), left middle temporal gyrus ($x = -64.5, y = -54, z = -1.5$), left parietal lobe ($x = -58.5, y = -51, z = 43.5$), and right frontal middle gyrus ($x = 36, y = 42, z = 34.5$). We found WM atrophy in FRDA patients in left cerebellum ($x = -6, y = -43.5, z = -58.5$), left caudate ($x = -9, y = 15, z = -3$), right postcentral gyrus ($x = 61.5, y = -9, z = 16.5$), left cingulate gyrus ($x = -6, y = -3, z = 33$), and left superior frontal gyrus ($x = -6, y = 31.5, z = 49.5$).

Conclusion: VBM analyses showed that FRDA patients have progressive GM and WM loss in two years.

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Abstract – WCN 2013

No: 2115

Topic: 36 – Other topic

Two new heterozygous mutations of *Htra1* gene in a Caucasian patient affected by CARASIL

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Background: Cerebral Autosomal Recessive Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CARASIL) is a hereditary cause of recurrent lacunar stroke and cognitive decline associated with alopecia and spondylosis deformans, due to mutations in the *HTRA1* gene on chromosome 10q. Firstly described in Asian pedigrees, CARASIL has been reported so far in only one Caucasian family.

Objective: To report a Caucasian case of genetically-confirmed CARASIL patient with new compound heterozygous mutation of the *HTRA1* gene.

Patient and methods: The proband was a 29 y-o Romanian patient, born from a non-consanguineous marriage, who presented two previous ischemic stroke episodes. Clinical examination and brain MRI, as well as direct sequencing of exons 1–9 of *HTRA1* gene, were evaluated.

Result: Neurologically she showed ataxic gait, pyramidal signs with severe leukoencephalopathy at MRI. Common inherited and genetic causes of early-onset leukoencephalopathy were excluded. Direct sequencing of exons 1–9 of *HTRA1* in the proband identified two new heterozygous mutations: a G to A transition at position 961 (c.961G > A) in exon 4 (inherited from the father) and a G deletion at nucleotide position 126 (c.126delG) in exon 1 (inherited from the mother). Both parents showed diffuse white matter changes in absence of vascular risk factors.

Conclusion: The clinical features and the absence of parents consanguinity suggest that this diagnosis should be considered in otherwise unrecognized forms of sporadic early-onset leukoencephalopathy with stroke-like episodes. Moreover CARASIL could be underestimated in Caucasian population.

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Abstract – WCN 2013

No: 2201

Topic: 36 – Other topic

Frontal and temporal cortices of both hemispheres participate in relationship between autonomic and cognitive functions in patients with vascular encephalopathy

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Since the middle of the 19th century it has been recognized that several higher cognitive functions, including language, are lateralized in cerebral cortex. Recently a different influence of left and right hemispheres on autonomic functions was demonstrated (Craig, 2005).

Aim: To examine the role of cortex of left and right hemispheres in relationship between cognitive and autonomic functions in patients with vascular encephalopathy (VE).

Methods: DC potentials and rCBF were investigated in 40 patients with VE before and after word fluency test (WFT). DC potentials were registered from head using 12 Ag/AgCl electrodes. Standard characteristics of computed tomography perfusion were measured in different areas of cortex. Differences between symmetrical characteristics of DC potentials and rCBF in right and left hemispheres were calculated. Blood pressure and pulse were estimated.

Results: Interhemispheric differences of CBV, CBF in temporal areas correlated positively with characteristics of blood pressure and pulse. Positive correlation between interhemispheric difference of DC potentials in frontal areas and pulse was revealed. Interhemispheric differences of TTP in the frontal areas, correlated positively with characteristics of blood pressure and pulse and negatively with verbal fluency.

Conclusion: Thus activation of left temporal and frontal area in comparison with right one is associated with the increase of verbal fluency and the decrease of blood pressure and pulse. Probably, the cortical interaction between left and right hemispheres is involved in a joint regulation of cognitive and autonomic functions.

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Abstract – WCN 2013

No: 2204

Topic: 36 – Other topic

Clinical manifestation of non-stiff anti-amphiphysin syndrome (NSAS)

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Background: The neurologic syndrome most associated with anti-amphiphysin antibody is the paraneoplastic stiff-person syndrome by small-cell lung cancer, breast cancer, or other tumors. However, other neurologic syndromes have been reported in several patients.

Objective: We analyzed the clinical manifestation of non-stiff anti-amphiphysin syndrome (NSAS).

Patients and methods: This study was conducted in a single center for six months. To diagnose any paraneoplastic syndromes, 340 samples were tested for onconeural antibodies by immunohistochemistry and western blotting, and those with anti-amphiphysin antibody were included in this study. The possible clinical syndromes included limbic encephalitis, brainstem encephalitis, cerebellar ataxia, encephalomyelitis, sensory neuropathy, and dysautonomia.

Results: Total 19 patients with NSAS were identified. Ten (53%) were male and the mean age was 57 (from 28 to 75 years). In six patients (32%), cancers preceded the neurologic syndrome, and in remaining 13 patients (68%), the neurologic syndrome preceded the cancer. Of the 19 patients, eight were with limbic encephalitis (including two with brainstem encephalitis), five with cerebellar ataxia, three with sensory neuropathy, two with encephalomyelitis, one with brainstem encephalitis, and one with dysautonomia. Clinical responses to immune modulation therapies were variable irrespective to the syndromes.

Conclusion: Here we show that NSAS is not rare, and the other neurologic syndromes, such as limbic encephalitis and cerebellar ataxia, can be associated with anti-amphiphysin antibody.

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Abstract – WCN 2013**No: 2173****Topic: 36 – Other topic****A blood brain barrier biorepository for disease-specific human brain microvascular endothelial cells: A new tool for neurologic and psychiatric research**T.R. Roesel, *Cervoe. Org, McLean, VA, USA*

Background: Blood brain barrier (BBB) dysfunction has been implicated in many illnesses such as stroke, traumatic brain injury, Parkinson's disease, Alzheimer's disease, schizophrenia, multiple sclerosis, as well as more uncommon conditions such as chronic Lyme disease, chronic fatigue syndrome, Gulf War-related illness, and HIV dementia. Investigations on BBB neurophysiology are limited by nonavailability of disease-specific human brain microvessel endothelial cells (HBMECs) for in vitro culture experiments, despite routine availability of normal, non-disease related HBMECs.

Objective: The objective is two-fold: 1) To confirm the need for a biorepository for disease-specific HBMECs, and 2) to present a schematic for establishing the first BBB bank to make disease-specific endothelial cells from patient donated brains available for clinically relevant translational medical research.

Material and methods: Non-oncological brain tissue banks were identified worldwide. These repositories were queried by website evaluation and/or email questionnaire. The scientific literature was reviewed for HBMEC isolation and cryopreservation techniques for protocol development.

Results: Seventy-seven non-oncological brain banks or their umbrella networks in 16 countries were identified, along with 8 commercial vendors. None offer disease-specific HBMECs, although 11 non-commercial banks provide disease-specific HBMECs with advanced notification, should a disease appropriate donor become available. The administrative requirements, equipment, material, and protocols essential for a functioning disease-specific HBMEC biorepository are reviewed.

Conclusion: Establishment of a biorepository for cryopreserved disease-specific HBMEC is feasible. Such a bank would provide needed cells for BBB-related translational medicine studies in neurology and psychiatry. With greater pathophysiologic knowledge, interventions to increase BBB resiliency in specific diseases can be explored.

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Abstract – WCN 2013**No: 2191****Topic: 36 – Other Topic****Necroptosis modulation by chitooligosaccharide presents a neuroprotective issue in Ngf-differentiated PC12 cells**P. Sadeghi, F. Khodagholi, *Neuroscience Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran*

Background: Necroptosis is an emerging form of cell death that pro-necrotic kinases in the receptor interacting protein (RIP) family are of crucial mediators. Chitooligosaccharide (COS) was applied for its application in medical therapies. Lately, evidence indicated that an elaborate biochemical network issue from receptors in the TNF superfamily can induce necrotic cell death.

Objective: Determination of necroptosis needed to be clarified by satisfying number of researches. Evaluating factors taking part in this pathway could reveal important indications of the cell death occurrence. Also, neuroprotection effect of COS was needed to be clarified as well.

Material and methods: PC12 cells were incubated with different concentrations of Necrostatin-1 30 min prior to being exposed to 150 μ M of H₂O₂. Also, different concentrations of COS were utilized 24 h prior to stress condition. The levels of Rip-1, Rip-2, Rip-3 and Caspase-8 were evaluated using western blot technique. Also, cell viability was evaluated by MTT reduction assay. AO/EB staining and propidium iodide (PI) staining were applied for detection of apoptosis and necroptosis, respectively.

Results: Necroptosis factors increased in H₂O₂ insulted cell groups. Also, applying Necrostatin-1 inhibited Rip kinases family members in all three different concentrations, having the most effective concentration at 100 μ M. Pretreatment of PC12 cells with COS modulated necroptotic factors. The most effective concentration observed at 150 μ g/ml was so similar to control levels. In addition, PI staining was utilized to confirm necroptosis.

Conclusion: Our data imply that chitooligosaccharide strikingly guards neurons against oxidative stress by modulating necroptosis signaling ways.

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Abstract – WCN 2013**No: 2167****Topic: 36 – Other Topic****Sleep and breathing in 100 healthy Caucasians—A polysomnographic study in self-perceived good sleepers**B. Högl^a, D. Gabelia^a, T. Mitterling^a, M. Biermayr^a, E. Brandauer^a, S.V. Schönwald^b, W. Poewe^a, B. Frauscher^a. ^aDepartment of Neurology, Innsbruck Medical University, Innsbruck, Austria; ^bDepartment of Neurology, Hospital de Clinicas de Porto Alegre, Porto Alegre, Brazil

Background: Normative studies for sleep using polysomnography have been performed, but are mainly historic and published prior to the introduction of the currently valid scoring criteria.

Objective: To investigate objective sleep parameters in healthy adults and establish normative polysomnographic values according to AASM.

Methods and participants: One-hundred subjects (f/m 40/60) aged 20–80 years who perceived themselves as healthy sleepers were recruited from the representative Austrian population. In a two-step procedure (telephone and face-to-face interviews) patients with any evidence for sleep disorders were excluded. Full-night polysomnography was performed and scoring of sleep, respiration and arousals was performed according to AASM.

Results: Total PSG sleep time decreased with age, from 6.9 (5.3–7.6) for those in their 20s to 6.3 h (3.6–7.3) (median; range) in the over 60s age group. Sleep efficiency decreased with age from 87% (71.9–94.1) in those in their 20s to 79.7% (44.5–90.9) (median; range) in those >60 yrs. The apnea hypnoea index (AHI) increased with age, with those in their 60s having a significantly greater AHI than subjects below 50 years of age.

Conclusion: This study provides normative PSG data according to AASM criteria. Our data show that there is unrecognized sleep-disordered breathing in the population. Even in subjectively healthy sleepers, and after exclusion of suspected sleep-disordered breathing, BMI > 30 and excessive daytime sleepiness, 3 subjects had moderate to severe sleep apnea. Based on these data screening for sleep apnea in asymptomatic elderly is recommended.

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Abstract – WCN 2013**No: 1302****Topic: 36 – Other Topic****Evaluating variability of fMRI in the sensorimotor cortex—A multicenter study***

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Background: More and more patients with planned neurosurgical interventions undergo fMRI-scans with localization of the primary sensorimotor cortex being one of the most frequently requested clinical tasks. Reliability of fMRI-localization is an important issue since changing the resection margins in the range of millimeters might dramatically change the functional outcome of surgery. Up to now dependence of fMRI-localization on the investigating site was only assessed in healthy volunteers, but is not known for patients with pathological brains.

Objective: To evaluate between-site-variability of fMRI in the sensorimotor cortex using a somatosensory and a motor task.

Methods: 15 patients with planned neurosurgical intervention due to unilateral brain pathology underwent fMRI-scans at 3 experienced clinical sites. fMRI-scans included a motor (fist clenching of hand contralateral to pathology) and a somatosensory task (vibrotactile stimulation of digitus 2/3 contralateral to pathology). fMRI-variability was assessed with

- (1) center-of-activation (COA) variability,
- (2) activation-size-ratio (SR),
- (3) activation-overlap-ratio (OR) and
- (4) intraclass-correlation-coefficients (ICC).

Results: Median COA variability was 5.7 mm (IQR 3.2–7.9 mm, maximum 16.5 mm) for the motor task and 5.8 mm (IQR 3.4–8.0 mm, maximum 12.0 mm) for the sensorimotor task. Median SR was 48.7% (IQR 40.8–66.1%; motor) and 30.8% (IQR 14.0–39.3%; somatosensory) respectively. The figures for OR were 33.0% (IQR 26.6–43.2%; motor) and 4.0% (IQR 1.1–10.2%; somatosensory). ICC was 0.23 (motor) and 0.20 (somatosensory).

Conclusion: Correspondence of fMRI-localizations is quite good between centers. In single patients localization differences can amount up to 16.5 mm. This figure needs to be considered in clinical practice and for clinical studies with multicentric designs.

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Abstract – WCN 2013**No: 2174****Topic: 36 – Other Topic****Effects of aging on supraspinal motor control of ankle movements**

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Background: Age-effects on functional Magnetic Resonance Imaging (fMRI) activation obtained by hand/finger movements have been reported previously in healthy subjects. However, despite the importance of changing gait capacities with age, analog studies for lower limb movements are scarce.

Objective: The purpose of this study was to explore the effects of age on motor control of ankle movements, aggregating data from several studies.

Patients and methods: We analyzed the fMRI data of 102 right-foot dominant healthy subjects aged 20–83 years, obtained during ankle movements. We both used FSL higher level analyses as standard approach and a meta-analysis employing coordinate-based activation likelihood estimation (ALE).

Results: Unilateral ankle movements elicited activation in the primary motor cortices and supplementary motor areas bilaterally (with contralateral peaks), the contralateral somatosensory cortices and the ipsilateral cerebellum. With increasing age, increasing cerebellar and precuneus activity was observed.

Conclusion: Similar to previous findings for fine motor skills of the upper limb, the BOLD-response associated with ankle movement scales with age. However, these changes predominantly concerned the precuneus, an area that has been implicated in neurodegenerative disorders like Alzheimer's disease.

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Abstract – WCN 2013**No: 2140****Topic: 36 – Other Topic****The phenomenology and epistemology of konzo: Readings from the Kahemba Outbreak, Democratic Republic of Congo. Part I: Socioeconomic aspects**

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Objectives: To determine the demographic characteristics of households affected by konzo; quantify the level of economic distress associated with konzo and elucidate the relationship between economic distress and cyanide poisoning.

Subjects and methods: 210 children aged 8.83 ± 2.57 years, 123 konzo and 87 non-konzo according to WHO criteria, were recruited in sectional study on demographic and economic profiles of patients affected by konzo. The work had included a literature review, a survey by structured interviews, observations and sampling of cassava flour and urine samples for biochemical analyzes in search of markers of cyanide poisoning. The socio-economic profile was evaluated by the Home. All children were tested negative for HIV and HTLV-1.

Results: The frequency of konzo was 79.47% for children aged <15 years and 56.9% for females. Exposure to HCN cassava flour is equivalent konzo children and non-konzo (p = 0.503), the severity of konzo was proportional to the poverty of the family socio-economic environment overall (p = 0.008). The nutritional status of

children with konzo was more degraded than non-konzo ($p < 0.001$). The presence of at least one parent with konzo in restricted or extended family was 79.8% for cases vs. 67.4% konzo non-konzo ($p = 0.041$, OR = 1.92 (95% CI: 1.019 to 3.587)).

Conclusion: Konzo is a toxico-nutritional, social and poverty associated with neurodevelopmental disease, affecting especially women and children during the school year. Studies on the psychoneuro-development environment of konzo is needed to understand the psychosocial vulnerability to this disease and better prevention.

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Abstract – WCN 2013

No: 2086

Topic: 36 – Other Topic

Predictors of Poor self-rated health in a Senegalese male elderly population utilizing the social and health center of Ipres, Senegal

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Background: Self-rated health is associated with morbidity and mortality. It is an indicator of quality of life. The objective of this study was to identify predictors of poor self-rated health in a Senegalese male elderly population of patients utilizing the Social and Health Center of IPRES, Dakar-Senegal for health care.

Methodology: During a cross-sectional study conducted in a sample of 546 elderly male 55 years and over utilizing the health center, data on sociodemographic characteristics, lifestyle, social network, past medical history, and self-rated health were collected from 2004 to 2005. Self-rated health was assessed with a single question as proposed by Mossey and Shapiro: "How do you perceive your health?" with responses in five categories: excellent, very good, good, fair and poor. Uni, bi and multivariate logistic regression analyses were performed.

Results: The patients had a mean age of 68 years (± 7.4), were married, and educated with high social network. Alcohol consumption and smoking were rare. Hypertension, arthritis, gastrointestinal and genitor-urinary diseases were more frequent. Self-rated health was poor (72%). Family history of dementia, stroke, epilepsy, head injury, illiteracy and weak social network were predictive of poor self-rated health while being married and age 60–74 years were protective.

Conclusion: It is necessary to take into account these results for primary prevention of poor self-rated health in this population.

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Abstract – WCN 2013

No: 2216

Topic: 36 – Other Topic

AQP4-neuromyelitis optica following NMDAR-encephalitis:

A case report

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Background: NMDAR-encephalitis is an autoimmune disorder often associated with teratoma. Neuromyelitis optica (NMO) is an inflammatory demyelinating disorder of the CNS associated with Aquaporin-4 antibodies.

Objective: To report a case of limbic encephalitis followed by NMO.

Case report: A 50-year-old woman presented with subacute memory loss, confusion and behavioral changes. A brain-MRI showed a T2-weighted hyperintense medial temporal cortex. Two months later an ovarian teratoma was removed. One year later she started to exhibit inflammation in the spine and subsequently in the optic nerve resembling neuromyelitis optica. She was treated with plasma exchange and steroids with benefit. A subsequent anti-neuronal antibody screening proved positive for anti-AQP4 antibodies in the serum and for anti-NMDAR antibodies in the serum and CSF. Human leukocyte typing disclosed class I allele B8 and class II DR3-DQ2 (DRB1*03-DQB1*02).

Conclusion: NMDAR-encephalitis can present with attacks that can mimic a demyelinating disease. However, to our knowledge this is the first case of NMDAR-encephalitis followed by AQP4-Ab positive NMO. The hypothesis that our patient had two distinct syndromes is supported by the typical association of NMDAR-encephalitis with teratoma whereas NMO is generally not a paraneoplastic disease. The possible coexistence of multiple autoantibody-associated neurological syndromes is increasingly recognized. The HLA typization suggests that a genetic predisposition played a significant role in the development of these two uncommon autoimmune disorders.

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Abstract – WCN 2013

No: 2237

Topic: 36 – Other Topic

Neural plasticity: Role of PPAR α activation

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Peroxisome proliferator-activated receptor (PPAR α) is expressed in neurons and astrocytes and its activation provides neuroprotection against noxious biological reactions induced by cerebral ischemia. The present study has focused on the role of PPAR α activation in neural plasticity. For this purpose, SV129 wild type mice were treated with either fenofibrates (FEN) or oleuropein, PPAR α activators, for 2 and 4 days, respectively. Similar treatment protocol was followed in Ppar α -null mice. The data of this study revealed that fenofibrates significantly up-regulated the brain-derived neurotrophic factor (BDNF), the neurotrophic factors NT-4/5 and NT-3, as well as the corticotrophin releasing factor receptor-1 (CRF-R1) and PPAR α in the prefrontal cortex (PFC). NT-4/5 and CRF-R1 were increased by FEN and oleuropein in the hippocampus. Oleuropein also markedly up-regulated CRF-R1 mRNA transcripts in the PFC. The FEN- and oleuropein-induced effects are mediated by PPAR α , because no change in the abovementioned neural plasticity indices was observed in Ppar α -null mice. In conclusion, drugs acting as PPAR α activators could presumably improve synaptic function/plasticity and dendritic outgrowth in the PFC and hippocampus, thus affecting cognitive functions, among others. PPAR α activation may also have an impact on the CRF-R1-regulated neuroendocrine and behavioral responses to stress.

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Abstract – WCN 2013

No: 2239

Topic: 36 – Other Topic

Low-grade glioma growth kinetics before and after CCNU alone

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Background: Previous studies have reported that low-grade gliomas (LGG) grow continuously and that temozolomide and PCV have an impact in their growth kinetics.

Objective: No data exists on the impact of CCNU alone. The aim of this study was to evaluate the effect of first-line CCNU alone chemotherapy.

Material and methods: The mean tumor diameter (MTD) of LGG was evaluated on serial magnetic resonance images before ($n = 28$), during, and after ($n = 36$) treatment with neoadjuvant CCNU.

Results: Before CCNU onset, MTD increased linearly over time (range 2.1 to 4.9 mm/year). During treatment with CCNU alone, 91% of patients experienced a decrease of MTD. After CCNU discontinuation, 28 of 33 patients who responded, an ongoing decrease of MTD was observed. Unresponsive tumors resumed their progressive growth within a year.

Conclusion: Our results confirmed that CCNU alone is an effective treatment for progressive LGG and its pattern of MTD decrease is comparable with previous study using temozolomide and PCV chemotherapy.

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Abstract – WCN 2013

No: 1259

Topic: 36 – Other Topic

Cardio- and cerebrovascular risk is associated with decreased slow wave sleep in patients with chronic kidney disease

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Background: Previous studies reported that sleep fragmentation and decreased slow wave sleep related to certain cardiovascular risk factors e.g. cholesterol levels, obesity in general population.

Objective: Our study aims to analyze association between cardio- and cerebrovascular risk and slow wave sleep in patients with chronic kidney disease.

Patients and methods: 100 kidney transplanted and 50 wait-listed hemodialyzed patients underwent polysomnography. The ten-year coronary heart disease risk was estimated for all patients using the Framingham Score. Moreover, the ten-year estimated risk of stroke was calculated according to the modified version of the Framingham Stroke Risk Profile. In multivariate linear regression models logarithmic transformed Framingham scores were used for estimating association between slow wave sleep and cardiocerebrovascular risk.

Results: Mean age was 51 ± 13 yrs, ratio of males was 56% in the population. Slow wave sleep was reverse related to cardiovascular and stroke risk (Spearman's rho: 0.259, $p = 0.003$; -0.209 , $p = 0.011$). In multivariate linear regression models the lower ratio of slow wave sleep was independently associated with the logarithmic transformed Framingham cardiovascular (beta = -0.211 ; 95% CI: -0.45 to -0.07) and cerebrovascular scores (beta = -0.155 ; 95% CI: -0.033 – 0.0) after adjusting for important co-variables (apnea-hypopnea index, serum albumin and hemoglobin levels, type of renal replacement, Charlson comorbidity index, sleep efficiency).

Conclusion: Decreased slow wave sleep was found to be an independent predictor of higher cardio- and cerebrovascular risk score in patients with chronic kidney disease.

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Abstract – WCN 2013

No: 673

Topic: 36 – Other Topic

Is transient global amnesia (TGA) hereditary?

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Background: TGA is a well established clinical syndrome, with an overall annual incidence of 3 to 8/100,000. 75% of the cases occur between the ages of 50 and 70 but rarely before 40. Stress, physical exercise and migraine are reported risk factors. MRI diffusion abnormalities in SCA2 are frequently demonstrated. Once affected, a patient has an annual relapse rate of 6–10%.

Objectives: To draw attention to a possible genetic factor in TGA for this individual predisposition.

Patients and methods: Hodge's diagnostic criteria were applied. Only first degree affected relatives were enrolled in the database. We review the published familial TGA and include 7 families reported recently by one of us. 6 of them were of the same hospital and were compared to a database of 127 consecutive TGA.

Results: The 6 families represent 4.7% of 127 TGA cases (95 CI 1.05–8.45%). We found 9 published familial TGA. Together with our 7 families, this topic gathers 41 cases in 16 families (mean age: 61.8–22 women–12 migrainous). Migraine and stress were frequent risk factors. These familial cases were not distinguishable from sporadic cases.

Conclusion: We suspect that familial TGA is globally underestimated and under-reported. If confirmed, a genetic link, even if rare, could provide helpful information in the understanding of the physiopathology of TGA as did the rare cases of familial Parkinson's or Alzheimer's disease. We propose a systematic inquiry of familial cases and their reporting, and DNA sampling of these families to set up a genetic research.

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Abstract – WCN 2013

No: 2303

Topic: 36 – Other Topic

Telemedicine—Is tele-EEG, tele-electrophysiology and telecytology possible—A feasibility study

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Background: A high end class stroke and teaching network by the help of telemedicine is established between the Department of Neurology, Krankenhaus Nordwest (KHNW), Germany and Bruneian Neuroscience Stroke and Rehabilitation Centre (BNSRC), since 7/2010.

Objective: This pilot study was conducted to validate the feasibility of tele-electrophysiology, tele-cytology and tele-electroencephalography carried out between KHNW and BNSRC.

Methods: 20 randomly selected patients for tele-electrophysiology, somatosensory evoked potential on the median nerve of the arm and tibial nerve of the leg and the nerve conduction were examined. 10 EEGs and 10 samples of CSF were examined and analyzed in both hospitals via a secure internet connection.

Results: Tele-electrophysiological part exhibits that findings made by neurologists from both sides are identical, tele-cytological part shows that both sides made the same results; tele-electroencephalographical part shows feasibility although there was a considerable inter-rater variability.

Conclusion: All studies revealed, evaluation of tele-cytology, tele-EEG and tele-electrophysiology is feasible. Evaluation of tele-electrophysiology and tele-cytological studies showed a high concordance between the evaluations. Concordance of the results of the tele-EEG examination was lower. This is most likely due to a high inter-rater variability, since EEG interpretation is highly influenced by examiner characteristics.

Quality of diagnosis and treatment of acute stroke depends on ability to recognize know and treat differential diagnosis and stroke mimics. As there is a lack of neurological knowledge in many areas of the world, this might be a useful future tool to deliver expert advice from one part of the world to another part of the world.

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Abstract – WCN 2013

No: 2312

Topic: 36 – Other Topic

Evolution in modern and future neurology and neuroscience

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Background: Evolutionary theories are ubiquitous in modern neuroscience. There is ongoing research on scales from the molecular and genetic, to organs, organisms and populations. After an introduction to the diversity of current evolutionary neuroscience, this presentation will focus on a possible future. Utilizing John Hughlings Jackson's evolutionary neurophysiology and his doctrine of concomitance, a new direction for evolution in cognitive neuroscience will be suggested.

Objective: To examine the diversity of evolutionary notions in neuroscience and a possible future.

Material and methods: Research.

Results: A commonality of the diverse applications of evolutionary ideas to neuroscience is identified, and a potential future for evolutionary neurophysiology is suggested.

Conclusion: Evolutionary theories have been essential in the development of neuroscientific ideas. Though the diversity of systems and scales is extensive, the shared concept of evolution provides a unifying feature. By carefully examining the past and present of evolution in neurology and neuroscience, one possible future application of evolution is identified.

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Abstract – WCN 2013

No: 2286

Topic: 36 – Other Topic

The highest incidence of traumatic spinal cord injury in the world

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Background: According to the retrospective study based on medical records, the incidence of traumatic spinal cord injury (TSCI) in Estonia is one of the highest in Europe (39.7 per million population, 1997–2007).

Objective: The aim of the study was to study the incidence of TSCI by including the individuals who die at the scene of the accident in Estonia from 2005 to 2007.

Patients and methods: Medical records of patients with TSCI from all hospitals of Estonia from 2005 to 2007 were studied. With collaboration of the Estonian Forensic Science Institute we included the data of the victims of TSCI who died before hospitalisation.

Results: One hundred and eighty-two patients were found retrospectively from medical records and 212 cases were detected from autopsy reports.

The annual incidence rate was 97.0 per million population (95% CI: 87.4–106.6). The mean age at injury was 44.4 ± 18.7 years. Motor vehicle accidents were the leading cause of TSCI among the individuals who died before hospitalisation (75%). Falls accounted for the highest number of TSCIs (43%) among the patients who reached hospital. More than half of the patients (55%) had consumed alcohol prior to the injury.

Conclusion: Our study shows that when we include the cases who die at the scene of the accident, the real state in the field of TSCI is more shocking that was earlier found. In order to reduce the number of TSCI, we must take into account the deaths in the pre-hospital phase.

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Abstract – WCN 2013

No: 2280

Topic: 36 – Other Topic

Etiologic agents of central nervous system infections in Georgia

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Background: Central nervous system (CNS) infections are severe diseases, caused by a wide variety of pathogens. The rapid diagnosis and management of these infections can reduce mortality and neurological sequelae but diagnostic laboratory tests are not readily available in all settings, especially in resource-limited countries.

Objective: In 2010, hospital-based surveillance was initiated in Tbilisi, Georgia to determine pathogens causing acute meningitis and encephalitis, and to enhance laboratory capacity for the diagnosis of CNS infections.

Patients and methods: Hospital staff collected cerebral spinal fluid (CSF) and acute and convalescent sera from patients meeting a clinical definition for meningitis/encephalitis. Specimens were tested by bacterial culture and RT-PCR for HSV types 1 and 2, mumps virus, enterovirus, varicella zoster virus (VZV), *Streptococcus pneumoniae*, *Haemophilus influenzae* type B (Hib), and *Neisseria meningitidis*.

Results: From October, 2010 through February, 2013, 199 patients were enrolled. One hundred eleven (56%) were children <18 years old. Pre-testing clinical diagnosis was bacterial meningitis in 82 (41%), viral meningitis in 95 (48%), viral encephalitis in 13 (7%), and TB meningitis in 9 (5%). By PCR, 25 CSF samples were positive for *S. pneumoniae*, six for *N. meningitidis* and one for Hib. Viral multiplex PCR of CSF identified enterovirus in 37 (19%) cases; other viral pathogens identified included VZV (6), HSV1 (2), and HSV2 (1). *S. pneumoniae* was isolated by culture in five cases.

Conclusion: This study expands the understanding of the range of pathogens causing CNS infections in Georgia. These findings affirm enteroviruses and bacteria as important CNS pathogens.

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Abstract – WCN 2013

No: 2302

Topic: 36 – Other Topic

Deficits in episodic memory relate to reduced hippocampal volumes in very-low-birth-weight (VLBW) young adults

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Background and aims: The hippocampus is regarded as a core structure for learning and memory functions. Academic problems are commonly reported in VLBW (bw ≤ 1500 g) children and adolescents, and the aims of this hospital-based, long-term follow-up study were to perform a comprehensive memory assessment in VLBW young adults and investigate the relationship between hippocampal volume and episodic memory compared to age-matched term born controls.

Methods: Forty-two non-disabled VLBW and 61 age-matched controls were examined at age 19 with the WAIS-III, Wechsler Memory Scale-III and cerebral MRI. An automated MRI technique (FreeSurfer ver. 5.1) at 1.5 Tesla for morphometric analysis of hippocampal volumes was applied.

Results: The VLBW group was at a disadvantage compared to controls on tests assessing visual immediate/delayed memory and working memory. The VLBW group had similar **relative** left (mean 0.244 (SE 0.003) mm³, p = 0.242) and smaller right (mean: 0.243 (SE 0.004) mm³, p = 0.027) hippocampal volumes vs. controls (left 0.249 (SE 0.003) mm³; right 0.253 (SE 0.003) mm³). Significant correlations were found between left (r = 0.344, p = 0.030) and right (r = 0.038, p = 0.016) relative hippocampal volumes and visual immediate memory. Working memory function was associated with right hippocampal volume (r = 0.392, p = 0.015).

Conclusion: We found that being born preterm with VLBW has long-term negative consequences regarding memory functions, persisting into young adulthood. The relative hippocampal volumes were reduced in VLBW young adults compared to term born controls. Positive correlations were found between hippocampal volume and immediate visual and working memory function in the VLBW group.

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Abstract – WCN 2013

No: 2310

Topic: 36 – Other Topic

Oculomotor changes in multiple system atrophy: Clinical, MRI and laboratory features in eight patients

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Background: Multiple system atrophy (MSA) is a progressive adult-onset α -synucleinopathy characterized by dysautonomia associated with cerebellar symptoms (MSA-C) or parkinsonism (MSA-P). Ocular motor changes, including saccadic dysmetria, saccadic intrusions and nystagmus, have been reported.

Objective: To determine the ocular motor changes in MSA that may differentiate these patients from other neurodegenerative diseases with overlapping clinical features.

Methods: Eight patients, six MSA-C and two MSA-P, were recorded. Mean age was 60 ± 3; mean disease duration was 4.87 ± 1.45 years. Horizontal (± 10°; ± 18°) and vertical (up/down 8°) visually-guided saccades and antisaccades were tested and compared with EVALab's normative data. Three minutes of steady fixation were performed. Saccadic parameters, antisaccade errors with relative corrections and fixation changes were evaluated.

Results: All patients showed significant reduction of amplitude (6.73° ± 2.11° vs 8.3° ± 0.09°) (P value = 0.026), and accuracy (2.01 ± 1.44 vs 1.17 ± 0.49) (P value = 0.004) of downward saccades. Amplitude at 18° of horizontal eccentricity (15.44° ± 2.56° vs 18.3° ± 0.9°) (P value < 0.0001) and accuracy at 10° (2.37 ± 0.86 vs 0.88 ± 0.52) (P value < 0.001) and 18° (4.86 ± 2.21 vs 1.22 ± 0.09) (P value < 0.001) of horizontal saccades were also decreased. Direction error in antisaccade task was about 45% with a 96% of spontaneous fast correction. Steady fixation was interrupted by saccadic intrusions (square wave jerks subtype, average number of 12/min and mean amplitude of 1.52° ± 0.31°).

Conclusions: Our findings substantiate peculiar changes of saccade dynamics in MSA. Downward saccades are more impaired than horizontal. Voluntary saccades and fixation are also impaired and lesser than in other similar neurodegenerative diseases. Oculomotor profile helps to differentiate MSA from other neurodegenerative diseases with overlapping clinical features.

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Abstract – WCN 2013

No: 2272

Topic: 36 – Other Topic

Serial magnetic resonance imaging of acute necrotizing myelitis

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Background: Acute necrotizing myelitis (ANM) is a rare spinal cord disorder occurring in the setting of infections, systemic autoimmune conditions, paraneoplastic syndromes or neuromyelitis optica; occasionally it may be idiopathic.

Objectives: To describe serial MRI findings in a case of idiopathic ANM involving the entire length of the spinal cord, including the anterior horns.

Patients and methods: A 25 year-old male was admitted to our department for severe back pain, numbness and leg weakness progressing over a few hours. On examination he presented paraplegia, lower limb areflexia, a T4 sensory level and loss of sphincter control. On admission, spinal MRI was unremarkable; however one week later it revealed lower cervical cord swelling and diffuse T2 hyperintensity of the entire thoracic cord extending to the

conus medullaris and involving white matter as well as ventral horns. Brain MRI was unremarkable. CSF examination showed neutrophilic pleocytosis with increased protein levels, and negative oligoclonal bands. Serum and CSF PCR testing for infectious pathogens, immunological assays and antibodies to aquaporin-4 were negative. The patient received methylprednisolone pulses followed by plasmapheresis, with no clinical improvement. Neurophysiological testing revealed axonal degeneration in lower extremities with diffuse fibrillation potentials. An MRI obtained 3 months later showed resolution of swelling with residual areas of T2 hyperintensities, cord atrophy and hydromyelia. The patient remained wheelchair-bound but relapse-free during a 22-month follow-up.

Results and conclusion: ANM is a rare disorder with a dramatic clinical presentation. Anterior horn involvement on MRI may be associated with poor prognosis despite aggressive immunosuppressive treatment.

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Abstract – WCN 2013

No: 2235

Topic: 36 – Other Topic

Epidemiology and etiology of acute flaccid paralysis of infectious origin in Georgia

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Background: With the ongoing elimination of poliovirus worldwide, understanding of other causes of acute flaccid paralysis (AFP) is increasingly important. Although studies of AFP have been conducted in developed countries, the epidemiology of non-poliovirus AFP in much of the developing world, specifically Central Asia and the Caucasus, has not been systematically investigated. This hospital-based surveillance study is the first attempt to identify and characterize cases of AFP in Georgia.

Objective: This study aims to characterize clinical, laboratory, and electrodiagnostic features of AFP admitted to four tertiary clinics in Tbilisi, Georgia.

Patients and methods: We enrolled patients >6 weeks of age meeting a clinical case definition for AFP. For each patient, we collected CSF, acute and convalescent serum, nasopharyngeal and stool samples to test for: *Campylobacter jejuni*, cytomegalovirus, enteroviruses, influenza virus, and tick-borne encephalitis and West Nile viruses. Nerve conduction studies and electromyography were performed in each patient.

Results: Between January, 2012 and December, 2012, we enrolled, nine patients; three were children <15 years of age. Based upon clinical and electrodiagnostic features, we classified six cases of Guillain-Barré syndrome, one case each of Fisher syndrome, transverse myelitis, and post-vaccine poliomyelitis. Laboratory testing for the pathogens assessed was all negative.

Conclusion: These preliminary findings suggest that the etiologies of AFP in Georgia vary, and GBS contributes substantially. Future investigations

will focus on continuing to define the etiologies of AFP in the Caucasus, determine subtypes of GBS, and identify concurrent or antecedent infectious etiologies of AFP.

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Topic: 36 – Other Topic

Rare manifestations of Fahr's syndrome: A report of 3 cases

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Introduction: Fahr's syndrome (FS) is a rare entity characterized by bilateral calcifications involving mainly the basal ganglia. A variety of manifestations are described in this syndrome especially movement disorders, dementia and psychiatric disturbances. Seizures and ischemic stroke are rarely reported.

Materials and methods: We studied 3 patients aged between 25 and 58 years. All patients had bilateral calcifications of the basal ganglia on CT scan. Electroencephalogram (EEG) and neuropsychological tests were practiced in one patient. Blood levels of calcium, phosphate, and parathyroid hormone were ruled out.

Results: The 3 patients are a woman and two men, aged between 25 and 58 years old. One patient has a familial history of mother with psychosis, one patient had a total thyroidectomy. The epilepsy was generalized in two patients, with comital EEG, and responded to phenobarbital. One patient has several transient ischemic strokes without diabetes, arterial hypertension and hypercholesterolemia. Physical examination has found pyramidal signs in one case and a mild dementia with impairment of episodic memory and executive functions in the second patient, and choreoathetotic movements in the third patient. Hypoparathyroidism was diagnosed in two cases.

Discussion: In FS, extracellular calcium deposits occur mainly in the basal ganglia, and also in cortical areas, that may early contribute to the epilepsy, and sometimes in the walls of capillaries and small vessels which may explain the stroke episodes in these patients.

Conclusion: The heterogeneity of the clinical manifestations is noted in Fahr's syndrome. Seizures or stroke is rarely revealing of the disease.

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Abstract – WCN 2013

No: 2186

Topic: 36 – Other Topic

Neurophysiological endophenotype in non-demented carriers of apolipoprotein E and apolipoprotein J/clusterin Alzheimer's risk variants

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Background: GWAS studies have provided evidence that in addition to apolipoprotein E (ApoE), CLU CC (rs11136000) genotype is associated with risk of Alzheimer's disease (AD) (Harold et al., 2009; Lambert et al., 2009; Golenkina et al., 2010). The use of multilocus genotype patterns that combine SNPs at different susceptibility genes provide a helpful approach to predict genetic risk for AD. Brain endophenotype derived from EEG may be valuable marker for genetic predisposition to AD.

Objective: This study was aimed at determining the possible effect of ApoE and CLU polymorphisms on spontaneous EEG and EEG reactivity in response to letter fluency task (LFT) and to hyperventilation in non-demented adults.

Material and methods: We examined quantitative EEG during resting, LFT and under hyperventilation in 142 non-demented adults, stratified by ApoE and CLU polymorphism.

Results: A pattern involving ApoE ϵ 4 and CLU CC genotypes was associated with the decreased upper alpha power in spontaneous EEG, more pronounced event-related desynchronization (ERD) during LFT and the presence of high-voltage delta-activity, theta-activity and sharp-waves under hyperventilation. The increased ERD in the carriers of ApoE ϵ 4 and CLU CC genotypes may reflect greater "effort" to perform the task and/or may result from neuronal hyperexcitability.

Conclusion: The data suggest that neurophysiological endophenotype of non-demented individuals at genetic risk for AD, characterized by dysfunction of alpha rhythm-generating structures and hyperexcitability, may be revealed decades before the clinical symptoms of AD.

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Abstract – WCN 2013

No: 2166

Topic: 36 – Other Topic

Fulminant intravascular malignant lymphomatosis manifesting as cerebral vasculitis: A case report

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Background: Intravascular malignant lymphomatosis (IVL) is a rare and aggressive non-Hodgkin type lymphoproliferative disorder with a predilection for the CNS and skin.

Objectives: To describe a fulminant case of IVL with CNS involvement.

Patients and methods: A 46-year-old male was admitted to our department because of gait instability and right leg weakness. Previous hospitalization for a longstanding history of fever of unknown origin had failed to demonstrate any evidence of infectious, inflammatory or systemic disease and the patient had been discharged, remaining afebrile for the following two weeks. Neurological examination in our department revealed right cerebellar and pyramidal signs. Brain MRI demonstrated multiple bilateral T2 hyperintense lesions, suggesting CNS vasculitis. DSA was normal. CSF examination disclosed a slight lymphocytic pleocytosis with elevated protein and positive oligoclonal bands. The patient was put on intravenous methylprednisolone but his condition deteriorated rapidly and a repeated MRI demonstrated a new lesions. Due to respiratory failure he was transferred to intensive care unit. A new bone marrow biopsy showed clusters of abnormal large B-lymphoid cells in bone marrow sinusoids, consistent with IVL. Despite receiving the adequate treatment, the patient died a month later.

Results and conclusions: IVL must be considered in the differential diagnosis of cerebral vasculitis. The diversity of clinical manifestations and the absence of specific hematological and neuroradiologic findings contribute to a potentially lethal diagnostic delay. Although in cases with CNS involvement, brain biopsy remains the gold standard and repeated bone marrow biopsies can be elusive.

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Topic: 36 - Other Topic

Anti-NMDAR encephalitis: Clinical and immunological characterization of a case series

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Background: Anti-NMDAR encephalitis is an anencephalic multi-stage disorder that can be associated with ovarian teratoma. It is generally responsive to tumour removal and immunotherapy.

Objective: To report clinical and immunological data of a series of patients with autoimmune encephalitis and NMDAR-Ab.

Patients and methods: Between 2005 and 2012, 295 patients suspected for autoimmune encephalitis were screened for NMDAR-Ab on a cell-based assay. NMDAR-Ab positive patients' clinical data were collected.

Results: Sixteen patients were included in the study (9 females; age range: 9–77 years; mean follow-up: 23 months. CSF inflammation was found in 9 patients; brain MRI abnormalities in 7 cases. After encephalitis onset a tumour was found and removed in three patients (two teratomas and a germ cell testicular tumour). One patient had a teratoma removed 8 years before and in another a breast tumour was diagnosed. Fifteen patients received an immunomodulatory treatment and in 13 patients there was a confirmed improvement. Eight patients had prominent psychiatric sequelae.

Conclusion: This study confirms some known features of anti-NMDAR encephalitis: involvement of both sexes and all ages; low rate of paraneoplastic cases; and general response to immunotherapy. Psychiatric involvement was a striking clinical feature, at onset, but also with major consequences in terms of outcome. Atypical findings included a diagnosis of teratoma several years before encephalitis and breast tumour. The full spectrum of the syndrome and the role of tumour and other triggering factors need to be explored. Prompt recognition of the syndrome and antibody screening is fundamental to improve the outcome.

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Topic: 36 – Other Topic

L2 hydroxyglutaric aciduria (L2HGA): Phenotypic variability in C.241A>G mutation patients

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Introduction: L2HGA is a rare AR metabolic encephalopathy. We report clinical studies in 8 Tunisian families with 17 affected patients. Inter and intra familial phenotype variability is discussed.

Methods: We performed a retrospective study of L2HGA patients carrying the same homozygous mutation. Clinical, biological and radiological data were analyzed.

Results: The mean age of our patients was 15.82 years. Mean age at onset was 2.8 years. The disease onset was progressive except in two patients. Symptoms at onset were variable (psychomotor delay, gait difficulties...). Clinical examination showed mental retardation, cerebellar syndrome (16), pyramidal syndrome (15), extra pyramidal stiffness and chorea (2). Epilepsy was noted in 7 patients and febrile seizures in 9. MRI revealed demyelinating subcortical leukodystrophy with the basal ganglia and caudate nucleus involvement in all patients.

Discussion and conclusion: L2HGA is characterized by progressive mental and neurological deterioration. Marked intra and inter familial variability has been reported. Our study confirms this variability even in patients carrying the same gene mutation. There is a phenotypic variability from one family to another and even in the same family since some siblings had clinical symptoms (epilepsy, ophthalmoplegia, extra pyramidal stiffness...) not found in others. There is also a variability regarding age and mode of onset. It appears that the type of mutation does not play a complete role in different clinical phenotypes. Environmental factors, oxidative stress and disturbances of other unknown secondary metabolic pathways related to L2HGA may contribute to this variability in phenotype.

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Topic: 36 – Other Topic

Anti-NMDA-receptor antibody encephalitis in a teenage boy

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Background: Anti-NMDA-receptor antibody encephalitis is a neuro-autoimmune disease associated with the presence of antibodies targeting synaptic proteins. It most frequently occurs in young women, most common in the context of an ovarian teratoma. Clinical presentation includes variable combinations of psychiatric symptoms, autonomic dysfunction, alterations of consciousness and movement disorders. Treatment consists of tumor removal and immunosuppressive measures. Prognosis is usually favorable, but recurrence of encephalitis has been reported.

Objective: We describe an unusual presentation of anti-NMDA-receptor antibody encephalitis.

Material and methods: One 16-year old boy was followed during fourteen months.

Results: This teenage boy with Klinefelter, presented with impaired alertness. He was first hospitalized elsewhere after developing paresthesias and a nonfluent aphasia. There, a non-organic problem was suspected. During hospitalization with us, he developed orofacial dyskinesias, choreiform movements and autonomic instability. Radiography of the thorax showed a mature teratoma and gave us, together with the clinical findings, the clue to test for anti-NMDA-receptor antibodies. These were positive in CSF and blood. Treatment was performed by tumor resection, high-dose intravenous corticoids, plasma exchange and cyclophosphamide pulse therapy. After four months the patient was discharged in good condition. Recently, we restarted immunosuppressive therapy because of behavioral problems and hyperphagia in the context of persistent presence of antibodies.

Conclusion: A teenage boy with Klinefelter, who presented with impaired alertness, orofacial dyskinesias, choreiform movements and autonomic instability, was diagnosed to have anti-NMDA-receptor antibody encephalitis on a mature mediastinal teratoma. Teratomas occur more often in patients with Klinefelter. Follow-up until today is favorable.

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Topic: 36 – Other Topic

Systemic juvenile lupus erythematosus long lasting remission of severe neurological symptoms after treatment with rituximab

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Background: Systemic lupus erythematosus (SLE) has a much higher incidence in Asian and African populations than in Western populations. Up to 50% of patients suffer from neuropsychiatric symptoms of different pathogenesis.

Case presentation: We report a 13-year-old girl with a diagnosed SLE, predominantly rheumatic symptoms. She rapidly developed confusion, focal epileptic seizures with secondary generalization and left sided hemiparesis combined with high fever and tachycardia. Initial showed right hemispheric cortical DWI-positive lesions in the MCA territory. After 2/12 progressive MRI lesions to subcortical regions in both hemispheres with additional microbleeds appeared and neither arterial occlusion, typical vasculitic changes, nor meningeal enhancement was observed. CSF was without significant pathology.

Aggressive treatment with high dose corticosteroids and iv immunoglobuline G was started, followed by one cycle of iv cyclophosphamide and plasma exchange. The patient deteriorated with a septic syndrome, respiratory failure, increase of liver enzymes, severe thrombopenia and anemia, colitis and secondary infectious complications.

After stabilization of vital functions, again thrombopenia developed together with EEG abnormalities, since 2011 rituximab was given with no side effect. The patient recovered tremendously without any persistent motor dysfunction, was able to resume schooling and has no neuropsychological deficits except occasional headache and fatigue. CD 20 lymphocytes are mentioned. The patient has been free of new somatic and neuropsychiatric SLE manifestation since two years.

Conclusion: Rituximab was well tolerated and long term effective in this case of juvenile SLE with life threatening cerebral lupus vasculopathy. Further studies have to establish the therapeutic significance of rituximab in severe neuropsychiatric lupus.

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No: 2087

Topic: 36 – Other Topic

C9orf72 expansion in atypical parkinsonism

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Background: Recently, an intronic expansion in C9orf72 was reported as a major cause of familial and sporadic FTD and ALS. These cases exhibit TDP-43 pathology. Screening in other neurodegenerative diseases has been performed and expansions detected in patients with FTD-ALS and parkinsonism, clinical-PSP and clinical olivopontocerebellar-atrophy.

Objective: To screen for the C9orf72 hexanucleotide repeat expansion in pathologically-confirmed atypical parkinsonism cases including MSA, PSP and CBD; and in clinical cases with a phenotype consistent with CBS.

Methods: To provide a qualitative assessment of the presence of an expanded (GGGGCC)_n in C9orf72, we performed a repeat-primed-PCR reaction. PCR products were analyzed on an ABI3730 and visualized using GeneMapper-software.

Results: Screening for the expansion was performed in 96 MSA, 177 PSP and 18 CBD samples and no expansion was detected. Screening in 102 CBS cases detected a massive expansion in 3 cases. The initial

presentation was an akinetic–rigid syndrome followed by cognitive decline. One case developed bulbar palsy in the late stage. Two cases had a family history of a similar disorder. Interestingly we also detected an expansion of 27 repeats in another case presenting with atypical parkinsonism. The significance of this short expansion remains unknown.

Conclusions: We conclude that the *C9orf72* expansion can be detected in clinical cases presenting with parkinsonian syndromes that may have overlapping features with the FTD–ALS spectra. We failed to detect an expansion in pathologically proven cases with MSA, PSP or CBD, and this confirms that *C9orf72* is not related to alpha-synuclein or tau pathology.

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Topic: 36 – Other Topic

A rare cause of bilateral and symmetric high intensity T2 signal of corticospinal tracts at brain MRI: Neurolyme

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Neurolyme is defined as the neurological involvement secondary to systemic infection by the spirochete *Borrelia*. Acute meningoradiculitis is the most common presentation of neurolyme whereas meningoencephalitis is less frequent. We report a case of borreliosis meningoencephalitis with corticospinal tract involvement.

A 60-year-old man developed a progressive gait disturbance for 6 months and weight loss of 15 kg for 1 year. The neurological examination disclosed an ataxospasmodic walk with a positive Romberg sign, a reflex tetrapyramidal syndrome with a paraparesis. Cerebral MRI showed bilateral corticospinal tract FLAIR hyperintensities (internal capsules, mesencephalon, pons and cerebellar peduncles) without contrast enhancement. Medullar MRI was normal. Lyme serology was positive. The lumbar puncture revealed a lymphocytic pleocytosis (11 elements), protein 1.48 g/l, normal glycorachia and intrathecal anti borrelia antibody synthesis. The electroneuromyogram showed a sensorimotor axonal polyneuropathy. Paraneoplastic, immunologic, metabolic and others infectious causes were excluded. Later, he mentioned a tick bite 1 year ago, without erythema, in an endemic zone. He was treated with IV ceftriaxone 2 g/day for 4 weeks with clinical improvement. Cerebral MRI at 3 months was clearly improved but remains abnormal. Bilateral and symmetric hyperintensity signal on T2-weighted images of corticospinal tracts has been reported mainly in amyotrophic lateral sclerosis and more rarely in metabolic diseases such as Krabbe disease, X-linked adrenoleukodystrophy or infectious diseases such as HTLV1. To our knowledge this MRI pattern has not been previously reported in neurolyme.

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No: 2354

Topic: 36 – Other Topic

The impact of recurrence: Brain tumor patients' quality of life

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Background: Few studies have investigated the differences of psychological outcomes between patients with brain tumor recurrence and those without recurrence.

Objective: The objective of this study was to determine whether the quality of life of patients with brain tumor recurrence differs from quality of life of those without recurrence.

Patients and methods: The study enrolled 52 patients with brain tumor recurrence and 63 without it. Mini mental state examination score ≥ 25 was patients' inclusion criteria for the study. We used the following tools to assess psychological effects of patients' specific condition: functional assessment of cancer therapy-brain (FACT-Br), psychological distress inventory (PDI) and hospital anxiety and depression scale (HADS).

Results: Our study found that patients with brain tumor recurrence showed a more compromised social/familiar [$t(88.15) = 2.53$, $p < .05$] and emotional well-being [$t(106) = 2.15$, $p < .05$] as compared with patients without recurrence. In addition, patients with brain tumor recurrence experienced more stress than those without it. Also levels of anxiety and depression were higher in patients affected by brain tumor recurrence. Moreover patients with brain tumor recurrence reported a worse functional status.

Conclusion: Our results highlight that while the patients without recurrence of brain tumor showed a significant psycho-physical well-being, the patients with recurrent brain tumor lived an impairment of their overall quality of life, both in physical and psychological well-being. The results underline the necessity to support patients with recurrent brain tumor, in order to help them to improve their level of self-acceptance and their adjustment to tumor-related problems.

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Topic: 36 – Other Topic

Diagnosis of functional motor disorder: 10 years later

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Background: Functional motor disorders are medically unexplained symptoms including paralysis, hyperkinesia and dysphonia diagnosed as psychogenic in origin. The diagnosis of a functional movement disorder is a clinical challenge although rate of misdiagnosis has declined in last decades.

Objective: The purpose of the study was to describe the course of the disease. Process of making the initial diagnosis and reaching to organic diagnosis later on in 9 cases is described in detail. Other results of the study have been reported elsewhere.

Patients and methods: Patients (84) diagnosed with functional motor disorders in Neurology Clinic of Tartu University Hospital from 1998 to 2002 were included in the study. Questionnaires were sent to patients for investigating the course of the disease and new neurological diagnoses and an invitation to specialist consultation was added to evaluate the medical condition of the patient. Indirect data on patients was obtained from medical databases.

Results: During follow-up period of 8 to 12 years 9 patients (11% out of 84) were diagnosed with organic disease explaining the previous symptoms. 2 patients were diagnosed with multiple sclerosis, 2 Parkinson's disease, 1 cerebellar ataxia, 1 hereditary sensorimotor neuropathy type II, 1 idiopathic non-familial dystonia, 1 manganese-related encephalopathy, and 1 rheumatoid arthritis. The organic diagnosis was ascertained average 4.5 ± 2.7 years after hospitalization.

Conclusion: During follow-up period of 8 to 12 years previous functional diagnosis changed in 9 cases. Most of these patients had

developed organic movement disorder. We suggest examining patients repeatedly to reach correct diagnosis.

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No: 2375

Topic: 36 – Other Topic

Mild cognitive disorders with background psycho-vegetative syndrome. Some of the metabolic aspects

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Background: Often the mild cognitive disorders develop in background of psycho-vegetative syndrome (PVS) not infrequently in frames of anxiety or anxiety depressive disorders. Cognitive disorders coexist with anxiety and depressive conditions, and later they can aggravate resulting in professional and social disadaptation. These studies of cerebral metabolism in mild cognitive disorders with background PVS will allow to make patient's complaints objective, and to improve the treatment and prevention of cognitive disorders.

Objective: To investigate cerebral metabolism in patients with PVS to improve the treatment and prevention of cognitive disorders.

Material and methods: To study cerebral metabolism, 29 patients with age under 55 with mild cognitive disorders and 20 healthy volunteers without signs of cognitive disorders as a control were examined. Cerebral metabolism was estimated with the help of neuroenergy mapping (NEM) – the detection of level of constant potentials (LCP), slowly varying potential of millivolt range reflecting membrane potentials of neurons, glia and hematoencephalic barrier.

Results: Normal results of LCP were obtained in 17.2%. Increased metabolism was observed in 58.6%. Decreased metabolism was seen in 24%. Background metabolism in patients with PVS significantly differed from the reference values. In control group of healthy volunteers the background metabolism didn't differ from the reference values.

Conclusions: Assuming all the data on increased metabolic processes in 58.6% of the examined subjects that indicate to the activation of backup pathways, it is reasonable to recommend sedation in combination with antioxidants and non-stimulating neurometabolic drugs to the patients with mild cognitive disorders with background PVS.

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Abstract – WCN 2013

No: 2398

Topic: 36 – Other topic

Risk factors for cerebral palsy in children with Hiv/aids in Botswana

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Background: Cerebral palsy is common among children with HIV/AIDS in the developing world, but there are no published studies that address potentially preventable risk factors.

Objectives: To identify risk factors for cerebral palsy in children with HIV/AIDS compared to uninfected controls.

Methods: We conducted a prospective cohort study with a nested case-control component to identify risk factors and outcomes in children with cerebral palsy seen in pediatrics clinic at a major referral center in Gaborone, Botswana.

Results: Risk factors for cerebral palsy in HIV uninfected children include prematurity, birth asphyxia, and CNS infections. Risk factors for cerebral palsy in children with HIV include CD4 count <200, advanced clinical stage, history of HIV encephalopathy, and history of CNS infections.

Conclusions: Cerebral palsy in children with HIV has distinct risk factors compared to HIV uninfected controls, and is largely acquired postnatally. Risk factors emerge in more advanced HIV and may be preventable by earlier initiation of antiretroviral treatment.

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Abstract – WCN 2013

No: 2392

Topic: 36 – Other topic

Bickerstaff brainstem encephalitis – A case report

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Background: Bickerstaff brainstem encephalitis (BBE) is a rare, postinfectious, immuno-mediated disorder clinically presenting with acute ophthalmoplegia, gait ataxia and occasionally decreased level of consciousness. Clinical signs result from the inflammatory lesion in the region adjacent to the IVth ventricle and the mesencephalic reticular formation.

Objective: We present a characteristic case of BBE.

Patients, methods and results: A week after a nonspecific enteritis the 58 year-old male patient noticed paraesthesia on his face and arms. A day later, vertigo developed and his gait got unstable. On the next day he became drowsy, and we found mild abducent palsy, gaze-evoked horizontal nystagmus, severe dysarthria, tetraataxia, and Babinski sign on his right side, serious gait ataxia and forced crying. In his T2 weighted MR images the region around the IVth ventricle and the left middle cerebral peduncle showed increased signal with blurred edge, spectroscopy was characteristic for inflammation. CSF analysis proved elevated protein level with normal cell count. Anti-ganglioside (GQ1b) antibody tests were negative. Five-day high dose methylprednisolone therapy was followed by slow improvement. A week later the patient was able to walk without help. His ophthalmoplegia and dysarthria ceased but forced crying persisted. Repeated CSF proved to be normal; his MRI abnormalities did not change. Two months later the patient was symptom free.

Conclusion: Majority of post infectious central nervous system disorders are acute disseminated encephalomyelitides. If brainstem signs are dominated, BBE has to be taken into account. The differential diagnosis is based on cranial MRI.

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Abstract – WCN 2013

No: 1776

Topic: 36 – Other topic

Isolation and culture of hair follicle stem cells (HFSCs) and their use for regeneration of the sciatic nerve in rat

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Background: Hair follicle stem cells (HFSCs) can differentiate into neurons, astrocytes, Schwanns and oligodendrocytes. The aim of this study was to evaluate the effects of transplantation of HFSCs on functional recovery after injury to the sciatic nerve in the rat.

Objective: The evaluation of the effects of transplantation of hair follicle stem cells on regeneration of the sciatic nerve injury in rat.

Materials and methods: The middle of the sciatic nerve of Wistar rat was cut (N = 40). Three groups were studied afterward. First, control group in which the nerve cut remained intact. Second, the epineurium at the cuts was sutured together. Third, the end cut was sutured and 500,000 hair follicular stem cells (HFSCs) injected in the site of nerve cut. For this purpose the bulge region of rat was isolated and the cells were cultured. After eight weeks, electrophysiological and histological assessments were performed.

Results: The results of amplitude (12/26 mV), latency (1/6 ms) of electrophysiology study showed better results in the third group (P < 0.05).

The results of histology showed that the number of axons was significantly higher in the HFSC group (806/67 n) than the other experimental groups (P < 0.05). Also, the myelin sheat was thicker in third group than others (1/46 μm).

Conclusion: We showed that the transplantation of HFSCs had potential capability of regeneration of sciatic nerve injury in Wistar rat.

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Abstract – WCN 2013

No: 2396

Topic: 36 – Other topic

Posterior reversible encephalopathy syndrome associated with tacrolimus and renal failure after liver transplantation

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Posterior reversible encephalopathy syndrome (PRES) is a small vessel microangiopathy of the cerebral vasculature that occurs in 0.5–5% of solid organ transplant recipients, most commonly associated with tacrolimus. Clinical manifestations include hypertension and neurologic symptoms like headache, visual disturbances, focal deficits, mental status changes, and seizures. We report an adult liver transplant recipient who developed PRES associated with tacrolimus and acute renal failure. A 53-year-old woman with familial Mediterranean fever underwent liver transplantation due to cryptogenic cirrhosis. Two months after liver transplantation, she presented with nausea, vomiting, dizziness and poor oral intake and was hospitalized due to renal dysfunction, hyponatremia and pancytopenia. At the fifth day of admission, she had an acute mental deterioration with a tonic-clonic seizure, but neurologic examination and brain CT scan was unremarkable, and tacrolimus level was normal. Her hospitalization was further complicated with vancomycin resistant enterococci septicemia and renal failure requiring hemodialysis. In the meanwhile tacrolimus levels remained normal, except for three days. At one month of hospitalization, five days after the first hemodialysis, her mental status suddenly worsened and a

second generalized seizure occurred. Bilateral parietooccipital hypodensities and brain edema seen on control brain CT were suggestive of PRES. Brain MRI showed T2 and FLAIR hyperintensities in the parietooccipital, frontal regions, basal ganglia and brain stem which were also compatible with PRES. She did not show any clinical improvement, despite cessation of Tacrolimus and intensive renal replacement therapy. In this case, although PRES was primarily associated with tacrolimus neurotoxicity, renal failure could not be ruled out in the etiology. PRES is a rare but potentially serious complication of solid organ transplants.

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Abstract – WCN 2013

No: 1484

Topic: 36 – Other topic

Combination of neonatal cerebral ischemia and systemic endotoxin modulates expression of oligodendroglial genes

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Background: Periventricular leukomalacia (PVL), defined as gliotic and dysmyelinating lesions in the periventricular white matter, is the leading cause of cerebral palsy and other neurologic disabilities in infants born extremely premature. Perinatal inflammation and hypoxia-ischemia are presumed to be the leading causes of PVL.

Objective: The study's objective was to evaluate the myelin-specific transcription factor response, and subsequent myelination in an animal model of combined neonatal cerebral ischemia and systemic lipopolysaccharide (LPS)-induced inflammation.

Methods: CD-1 mouse pups underwent unilateral common carotid artery ligation on postnatal day P5. At 24 h and 6 h prior to ligation pups were injected intraperitoneally with 0.6 mg/kg bodyweight of LPS, or with saline. Reverse transcription real-time PCR and immunohistochemistry (IHC) were performed at P14 (n = 7–12/group). Expression of CNPase, MBP, SIP1, SMAD7, OLIG1, MRF, SOX10, CNPase, and MBP genes was measured. For IHC analysis, sections were stained for MBP and evaluated by densitometry in selected areas.

Results: A significant decrease in all promyelinogenic transcription factors but OLIG1 was seen between the naive control and the LPS-injected/ligated group. The transcripts of myelin components MBP and CNPase were also significantly decreased. While anteriorly there were no significant changes in MBP optical density, this did hold true for white matter in mid and posterior sections.

Conclusion: Human autopsy studies have demonstrated arrested differentiation of the oligodendrocytic lineage in PVL lesions. Here, we demonstrate that the combination of systemic inflammation with cerebral ischemia leads to decreased expression of the oligodendroglia specific transcription factors, which play a key role in myelinogenesis.

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Abstract – WCN 2013

No: 2386

Topic: 36 – Other topic

The “dormant potential” of the non-dominant hemisphere: Interemispheric asymmetry of cortical plasticity

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Background: Transcranial magnetic stimulation (TMS) studies have documented handedness related functional asymmetries in corticomotor excitability.

Objective: To use TMS in comparing changes in cortical excitability of dominant (DH) and non dominant hemisphere (NDH) in both right-handed and left-handed subjects in function of a bimanual non-fatiguing motor task.

Material and methods: Thirty enrolled subjects were classified as right or left handed according to their description of the hand used for writing and completed the Edinburgh Handedness Inventory. For each subject, cortical excitability was separately assessed in each hemisphere. By means of TMS, MEP amplitude, motor threshold, and silent period were assessed. Then subjects performed regular repetitive bilateral opening-closing movements of the index finger onto the thumb. MEPs of the first dorsal interossei were recorded before exercise, immediately after exercise periods of 30, 60, 90 s and after 15 min of rest. We evaluated if post-exercise facilitation and delayed facilitation were present. Hemispheric differences of MEP amplitudes registered after rest were related to handedness score.

Results: Mean threshold was lower for DH compared to NDH. MEP amplitude was increased in exercise condition, independently of the hemisphere stimulated. In rest condition, only NDH presented increased MEP amplitude, showing delayed-facilitation. Handedness scores and MEP asymmetry in rest condition were positively correlated.

Conclusions: DH and NDH answer differently to motor requests and recover differently after exercise. Bimanual motor task induces different changes in cortical excitability in the two hemispheres, in favour of the non-dominant. NDH has a “dormant potential” that can be activated if necessary.

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Abstract – WCN 2013

No: 2371

Topic: 36 – Other topic

A comparative-study on selective attention and executive function between patients with/without bipolar disorder based on Stroop test

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Background: Investigations indicate that patients with bipolar disorder are often accompanied by changes in cognitive processes and abilities. These include reduced attentions and executive capabilities.

Objective: The objective of present study is a comparison on selective attention and executive function between patients with/without bipolar disorder based on Stroop test.

Materials and methods: In this study, comparative and analytical methods are used. The statistical sample was 34 males with bipolar disorder selected by available sampling method in psychiatry hospital and 34 men without disorder selected by random sampling method in Tehran city. The measuring tool is Stroop test. The results were analyzed using *T*-test method.

Results: The obtained results showed a significant difference between two groups in selective attention, executive function and their dependent variables including the experiment time, reaction time, and numbers of errors, unanswered questions and correct answers in response to consistent and inconsistent stimuli ($P < 0.05$).

Conclusion: It was found that patients with bipolar disorder in selective attention, executive function and their dependent variables in response to consistent and inconsistent stimuli have inappropriate conditions compared to the normal individuals.

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Abstract – WCN 2013

No: 2374

Topic: 36 – Other topic

Listeria rhombencephalitis in an immunocompetent young adult complicated by cerebral venous thrombosis

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Background: *Listeria monocytogenes* rhombencephalitis (LMR) is a rare complication with high level of mortality and sequelae.

Objective: We present a case of seronegative LMR in a healthy person. Diagnosis was based on clinical, imaging findings and improvement after specific antibiotherapy.

Case report: A 19-year-old woman was admitted with diplopia, inability of mastication and swallowing, preceded by fever, headache and nausea. The neurological examination showed isolated asymmetrical palsies of almost all cranial nerves. Brain magnetic resonance image (MRI) found exclusive brainstem lesions. Cerebrospinal fluid (CSF) examination revealed lymphocytic pleocytosis, hyperproteinorrhachia without hypoglycorrhachia. Blood cultures and Gram staining of the CSF were negative. Treatment with acyclovir was introduced with worsening and hypoventilation requiring mechanical ventilation. LMR was suspected and empirical therapy with ampicillin and gentamycin was started. On follow-up MRI, an extensive CVT was noted and treated by heparin. Her neurological condition improved gradually. Gastrostomy was carried out.

Discussion: In contrast to other listerial CNS infections, the majority of LMR cases occur in previously healthy adults. Isolated brainstem localization is rare but very suggestive of the diagnosis. Pons and medulla's cranial nerves are the most commonly affected, our patient has an involvement of almost all cranial nerves. Sepsis and meningeal signs are often absent. Gram stains of CSF resulted positive in only 14%, and CSF cultures are positive in 45% of cases. Two mechanisms are discussed: hematogenous spread or migration along axons of LMR.

Conclusion: To our knowledge, this is the first case of CVT due to LMR in immunocompetent adult.

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Abstract – WCN 2013

No: 2372

Topic: 36 – Other topic

Acute myelopathies following epidural and spinal anesthesia

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Background: Spinal and epidural anesthetics (E-SA) are widely used and are generally regarded as safe with a very low frequency of permanent neurological sequelae.

Objective: To assess cases of Myelopathy following E-SA.

Methods: In order to describe the different clinical and imaging features of E-SA induced myelopathy, we have retrospectively reviewed cases of patients hospitalized and diagnosed in our institute during 2004–2012.

Results: Ten patients developed progressive spinal motor, sensory and autonomic dysfunction within 3–24 h following E-SA. The different entailed mechanisms had different clinical phenomena.

Three 25–30 years-old women manifested with acute myeloradiculitis related to peripartum epidural spinal anesthesia causing toxic damage to the spinal cord.

Six 32–71 year-old patients had a direct traumatic damage to the spinal cord. Their manifestations ranged from a Conus Medullaris injury that resulted in Brown-Sequard like spinal cord injury and detrusor instability (5 patients) to direct cervical spinal cord injury

causing Tetraparesis (1 patient). Empirical steroid treatment was beneficial in the disappearance of the local edematous effect and hence in rapid clinical improvement.

MRI in these nine patients revealed corresponding T2-hyperintense spinal cord lesions.

In one 74 year-old patient, the spinal anesthesia contributed to local hypoperfusion and spinal ischemia.

Discussion: Although rare, myelopathy caused by E-SA occurs usually in a perisurgical clinical setup or following a procedure of regional pain control. Main clinical characteristics include an acute spinal deficit and corresponding specific imaging findings. Spinal MRI in this clinical study appeared highly specific and diagnostic of inadvertent spinal cord injury and toxic or ischemic effect.

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Abstract – WCN 2013

No: 2327

Topic: 36 – Other topic

Is cortical excitability affected by sleep fragmentation?

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Background: Sleep deprivation (SD) influences cortical excitability. Alterations in movement-related cortical plasticity are present in Restless legs Syndrome (RLS), characterized by sleep fragmentation (SF).

Objective: We evaluated the effect of SF on cortical excitability using transcranial magnetic stimulation.

Material and methods: After a night of spontaneous sleep (basal condition–BC) and after a night of fragmented sleep (fragmented condition–FC), MEP amplitude, motor threshold (MT), silent period (SP) and intracortical inhibition were assessed in healthy subjects. In both conditions, MEPs of the first dorsal interosseus were recorded before exercise, immediately after the exercise periods of 30, 60, 90 s and after 15 min of rest. We evaluated if post-exercise facilitation and delayed facilitation were present. Before each session, Stanford Sleepiness Scale (SSS) was completed.

Results: MT and SSS were increased in SF. No significant differences for MEP amplitude or SP or intracortical inhibition were found. In both conditions MEP amplitude was larger than baseline after 30 and 60 s of exercise (post-exercise facilitation) and also after rest (delayed facilitation). Comparing the two conditions at each time point we found no significant differences in MEP amplitude.

Conclusions: We suggest that SD and SF represent different phenomena that can depend on various networks acting on motor cortex. SF seems to impair the restorative cognitive benefits of sleep via alterations in hippocampal synaptic plasticity, involving mechanisms different from altered SD. We speculate that the contradiction between our SF data and our previous results in RLS may not be associated to SF, but related specifically to RLS pathophysiology.

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Abstract – WCN 2013

No: 1800

Topic: 36 – Other topic

Diffusion tensor imaging reveals cerebellar, thalamic and global white matter abnormalities in a rat model of kernicterus

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Background: A mutant Gunn rat was evaluated as a model of kernicterus by diffusion tensor imaging and correlated with motoric changes observed.

Objective: MRI data was searched for imaging correlates of profound motoric changes observed, namely ataxia ± dystonia.

Methods: Animals were assigned to one of 4 groups: wild type controls (WT, n = 6), jaundiced Gunn rats without (jjC, n = 6) and with sulfonamide injections either manifesting with severe (jjSsev, n = 6) or mild dystonia (jjSmild, n = 6).

Fractional Anisotropy (FA), radial, axial and mean diffusivity were evaluated in white matter structures. T2 volumetric studies were used for the evaluation of the cerebellum.

Results: Cerebellar volume analysis showed highly significant decreases in all jaundiced animals compared to WT controls. Evaluation of white matter tracts revealed significant FA changes in the middle and inferior cerebellar peduncle in jjSsev- and jjSmild-animals. Interestingly, mean diffusivity and particularly axial diffusivity demonstrated changes in the corpus callosum below the motor cortex, the external capsule and the internal capsule in the jjSsev-group only.

Discussion: We report global white matter abnormalities and changes in the thalamus in dystonic animals. Pathological kernicterus staining was found in the Globus Pallidus (GP) of the jjSsev-group, however it did not correlate with any imaging findings. Whereas the cerebellum and GP certainly play a role in the development of dystonia, our novel finding is that there have to be thalamic and global white matter changes for the development of acute severe dystonia. We postulate that cerebellothalamocortical pathways may be indeed a signature of dystonia in general.

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Abstract – WCN 2013

No: 2307

Topic: 36 – Other topic

Psychological burden of brain tumor patients and their caregivers

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Background: Brain tumors not only impact patients' quality of life, but also reduce seriously the caregivers' quality of life.

Objective: We aim to describe brain tumor patients and their caregivers' quality of life during the illness and assess the existing relation between clinical and psychological features of patients and their caregivers.

Patients and methods: The study involved 72 patient/caregiver couples. Mini Mental State Examination score ≥ 25 was patients' inclusion criteria for the study. To evaluate various psychological features, we used the following tools: Hospital Anxiety and Depression Scale (HADS), Functional Assessment of Cancer Therapy-Brain (FACT-Br) for patients and HADS, Caregiver Reaction Assessment Scale (CRA), 36-Item Short-Form Health Survey (SF-36) for caregivers.

Results: Quality of life was more compromised in caregivers than in patients. The impairment of caregivers' quality of life appeared mainly in a significant reduction in their mental health. Most caregivers experienced more anxiety [$t(142) = 6.11, p < .001$] and depressive symptoms [$t(142) = 2.87, p < .01$], as compared with patients. Clinical and psychological features of patients did not correlate with psychological patterns of their own caregivers.

Conclusion: Although an anxiety and depression condition is somewhat expected both in brain tumor patients and in their caregivers, high levels of anxiety and depression in caregivers must become an important end-point in clinical trials in that such levels

compromise their quality of life and worsen their ability to provide care. It is important for physicians paying attention not only to the patient's pathology, but also to potential caregiver's physical and psychological disorders.

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Abstract – WCN 2013

No: 2368

Topic: 36 – Other topic

Neuro-Behcet's disease: Report of 51 patients

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Background: Behcet's disease (BD) is a chronic, relapsing, systemic disorder of unknown etiology. Neurologic involvement in BD is one of the most devastating manifestations of the disease.

Objective: This study was conducted to describe the clinical and prognosis aspects of neurologic involvement in BD among patients attending a department of neurology in a military hospital in Rabat (Morocco).

Material and methods: Fifty one patients had evidence of and fulfilled the diagnostic criteria of neurobehcet's disease (NBD). These patients underwent laboratory and imaging investigations, including lumbar puncture and computed tomography scanning or MRI.

Results: The study included 33 men and 18 women aged from 17 to 54 years. The analyses of the result found the typical parenchymal NBD in 69% characterized by lesion of brainstem and atypical parenchymal NBD in 31% dominated by venous occlusion. All patients received high dose steroids, and 59% were treated with a combination of steroids and cytotoxic agents. The follow-up was good in the atypical parenchymal NBD.

Conclusions: Our study shows that the atypical parenchymal NBD had good prognosis than parenchymal NBD, and aggressive intervention with corticosteroids and cytotoxic agents may be early in order to diminish and stabilize the negative effect of neurologic involvement.

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Abstract – WCN 2013

No: 2152

Topic: 36 – Other topic

Dual effect of H89, a potent protein kinase A inhibitor, on H₂O₂-induced apoptosis and Nrf2 stabilization

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Background: We investigated the effect of H89 as a selective and potent inhibitor of protein kinase A (PKA) on PC12 neuronal cells. It has been reported that high levels of neural PKA produce more synapses; however its possible involvement in apoptotic cell death has been shown. Nuclear factor erythroid-2 related factor 2 (Nrf2) as a cytoprotective factor, enhances the antioxidant capacity in the cells and protects them against cell death.

Objective: Effect of H89, a PKA inhibitor, on H₂O₂-induced apoptosis and Nrf2 stabilization.

Material and methods: Morphological analysis of apoptosis was determined by AO/EB and Hoechst staining. PC12 cells were differentiated and treated with different concentrations of H89 (5, 7, 10, 15 and 20 μM) with or without exposure to 150 μM of H₂O₂. Caspase3 and Nrf2 levels were determined by western blotting.

Results: Following H₂O₂ treatment, cleaved caspase3 increased and Nrf2 level was downregulated. In PC12 cells, treated with H89 (5, 7 and 10 μM) and H₂O₂, Nrf2 level increased and caspase3 level was significantly modulated, in contrast to H₂O₂-exposed group. However, high concentrations of H89 (15 and 20 μM) upregulated caspase3, by about 2 and 4 folds, and inhibited Nrf2 stabilization. Consistently, H89 treatment in 15 and 20 μM concentrations, solely, upregulated pro-apoptotic factor, caspase3, and Nrf2 level decreased, in contrast to control cells. Hoechst and AO/EB staining confirmed the data obtained from western blot analysis.

Conclusion: H89, a PKA inhibitor, has dual effects in apoptosis and survival depending on its concentration.

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Abstract – WCN 2013

No: 2421

Topic: 36 – Other Topic

Correlation between errors committed by patients with bipolar disorder in Bender-Gestalt test and their pessimism and guilty conscience

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Background: Bender-Gestalt test is one of the most widely used neuropsychological tests, and also an important tool for evaluation of motor and visual coordination. Bender test expanded this concept on how the performance of a person may be affected by the perceptual and motor maturation as well as organic or functional impairments. Investigations using this test have also been used in patients with bipolar disorder.

Objective: The objective of this research is to study of the correlation between errors committed by patients with bipolar disorder in Bender-Gestalt test and their pessimism and guilty conscience in PANSS test.

Materials and methods: In this study the correlation method was used. The statistical sample included 35 males with bipolar disorder selected by available sampling method in a psychiatry hospital in Tehran city. The measuring tools were positive and negative symptom scale (PANSS) and Bender-Gestalt test. The results were analyzed using Spearman's correlation test.

Results: According to the obtained results a meaningful correlation among pessimistic and guilty conscience scores, and the twelve committed errors in Bender-Gestalt test in patients with bipolar disorder was not observed.

Conclusion: The results of this study showed that the pattern of errors committed by bipolar patients in Bender-Gestalt test did not show any significant relationship. Based on this finding, diagnostic evaluation, therapeutic interventions and rehabilitation for these patients should be improved.

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Abstract – WCN 2013

No: 1812

Topic: 36 – Other Topic

Vogt-Koyanagi-Harada syndrome: Reminding for a rare cause of uveomenigeal syndrome

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Background: Vogt–Koyanagi–Harada syndrome is a rare disease defined as bilateral chronic granulomatous panuveitis that may be associated with CNS, auditory and integumentary manifestations.

Objective: To describe 3 cases of Vogt–Koyanagi–Harada syndrome.

Patients and methods:

Case 1: A 27-year-old woman presented with recurrent bilateral anterior uveitis in the previous three months. In the last episode she received oral steroids with partial improvement of the visual acuity. One month later she complained of blurred vision and headache with photophobia. The examination revealed bilateral papilloedema. CSF had lymphocytic pleocytosis. She didn't have skin changes.

Case 2: A 2-year-old woman presented with two-months of complaints of redness, blurred vision and discomfort in both eyes. She also had left tinnitus, severe headache, photophobia, and vomiting. On examination she had bilateral posterior uveitis with retinal detachment. One month later hair poliosis and achromatic patches in the face/chest were noticed.

Case 3: A 25-year-old man complained of painful red eye, bilateral low visual acuity and headache with photophobia in the previous month. The examination disclosed posterior uveitis with retinal detachment. CSF showed lymphocytic pleocytosis. Two months later a hypopigmented lesion developed in the periorcular area.

Results: In the three cases other causes of uveomeningeal syndrome were ruled out and treated with high-dose systemic steroids. Methotrexate/azathioprine was additionally required because of recurrent ocular involvement. In all, inflammation decreased and vision was preserved enabling tapering of the steroid therapy.

Conclusion: Early diagnosis of this syndrome is crucial because prompt administration of high-dose steroids may prevent permanent visual loss. Usually additional immunosuppression is needed with a steroid-sparing effect.

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Abstract – WCN 2013

No: 2420

Topic: 36 – Other Topic

Bereavement in brain tumor: Psychological reactions in caregivers

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Background: When brain tumor patients' conditions decline and death occurs, caregivers deal with the adaptation to a new life condition.

Objective: We aim to describe changes in quality of life, anxiety and depression experienced by caregivers after bereavement of their loved one.

Material and methods: The study involved 44 caregivers of brain tumor patients in two different periods. In order to evaluate the psychological features the following tools were provided: Hospital Anxiety and Depression Scale (HADS) and 36-Item Short-Form Health Survey (SF-36). First data collection (T1) occurred when caregivers, together with the patients, were attending the Neuro-oncology Unit of IRCCS Istituto Neurologico C. Besta, Milan, Italy. The same variables were detected in a period of time ranging from 18 months to 3 years after bereavement (T2).

Results: Of the 44 caregivers involved, 4 could not be reached and 3 did not give consent to participate in the research. The 37 caregivers tested in T2 reported levels of anxiety and depression lower than those experienced in T1. In T2 higher scores were detected in psychological well-being scales (Social Functioning, Role Emotional, Mental Health) while scores related to physical health remained almost unchanged from T1 to T2.

Conclusion: Data shows that brain tumor impacts more on caregivers' mental health. From 18 months after bereavement onward, caregivers reported a psychological well-being higher than the one experienced during the illness of their loved one. Even if grief elaboration is a long and painful experience caregivers seem to have elaborated the loss of their loved one and have restored interest in life.

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Abstract – WCN 2013

No: 2170

Topic: 36 – Other Topic

Alginate oligosaccharide upregulates Ampk and induces mitochondrial biogenesis in H₂O₂-exposed Pc12 cells

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Background: Many studies show mitochondrial dysfunction in aging and some diseases such as Alzheimer's disease (AD). Mitochondrial biogenesis is a special defensive mechanism that many factors involved in it include:

- 1) AMPK and CREB as stimulator of PGC-1 α ;
- 2) NRF1 that is stimulated by PGC-1 α ; and
- 3) Tfam expression and initiation of mitochondrial DNA replication.

Objective: Effect of alginate oligosaccharide on mitochondrial biogenesis.

Materials and methods: PC12 cells were pretreated by AOS for 12 h, then exposed to compound C (10 μ M), AMPK inhibitor, for 12 h. Then, cells treated by H₂O₂ (150 μ M) for different times. In addition, we compared the effects of enzymatically depolymerized unsaturated AOS with heat-degraded saturated AOS. The level of mentioned factors was determined by Western blotting and the mitochondrial number was measured by Mito Tracker Red fluorescent staining.

Result: After H₂O₂ treatment, AMPK, PGC-1 α , NRF1, Tfam and Cytochrome c increased and reached the highest level at 8 h after exposure, and then continually started to decrease. Also, phospho-CREB/CREB ratio increased but the highest level was at 12 h. Pretreatment with AOS caused a significant increase in the level of AMPK, PGC-1 α , NRF1, Tfam, Cytochrome c and phosphor-CREB/CREB ratio in H₂O₂-induced cells and stayed higher than H₂O₂-treated cell till 24 h. In the group with compound C, we found a weaker effect of AOS on the induction of mitochondrial biogenesis. The results showed that heat-degraded saturated AOS has more effect on this pathway compared to enzymatically depolymerized unsaturated AOS.

Conclusions: Mitochondrial biogenesis is involved in AOS-mediated protection.

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Abstract – WCN 2013

No: 2406

Topic: 36 – Other Topic

Duane syndrome: A case report

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Background: Duane's syndrome is characterized with a defect in the nucleus of the sixth cranial nerve and/or a developmental defect of the sixth cranial nerve axon. It was first described in 1879 by Heuck in a patient with a serious eye movement restriction and globe retraction. Alexander Duane reported a series of 54 cases in 1905 and the syndrome was named after him. Heterochromia can be seen in Duane's syndrome.

Objective: In this report we would like to present a rare case of Duane syndrome with heterochromia.

Patients and methods: A 38-year-old male patient presented to our outpatient clinic with complaints of double vision which he had since childhood. He stated that his complaints increased especially when looking to the left. The color of his eyes had been different since birth. His right eye was green, and his left eye brown. On the neurological exam left sided sixth cranial nerve palsy was present. Other neurological findings were normal.

Results: On his brain MRI and brainstem thin slice MRI, the intracysternal segment of the sixth cranial nerve was visualized on the right but not on the left side.

Conclusion: Because of its rare nature and the absent left sixth cranial nerve on the brain MRI we found this case worth reporting.

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Abstract — WCN 2013

No: 2413

Topic: 36 — Other Topic

Avicenna's description of Willis circle

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Background and objective: Willis circle is an interconnection between anterior and posterior, and right and left cerebral circulations. It supplies brain and neighboring structures. Although this circle was noticed briefly and incompletely by ancient doctors, it is described completely by Thomas Willis in the book of Cerebri Anatome in 1664. In this study, the description of Avicenna, who was one of the most important scientists of the medieval period, about Willis circle is investigated. It can be important to shed light on a part of history of neurology.

Method: Arabic and Persian versions of Canon on Medicine, Avicenna's medical encyclopedia were studied and his point of view about Willis circle was compared with current and also ancient terms.

Results: Avicenna (980–1037) was one of the great scholars in medieval time. He wrote about this circle in the third volume of the Canon, in the section about head diseases. According to Avicenna's description, Willis circle was well known. Persian scholars can dissect in the ancient and medieval era. With regard to this, they had good information about human bodies and progress the previous Greco–Roman knowledge.

Conclusion: There is a gap between the knowledge of the medieval Persians and current era. It was acquired in the Renaissance, when western scholars refused the medieval knowledge, created the new paradigm of medicine and became the leadership of science. It seems that many of the eastern medieval findings were forgotten in this transmission. Therefore many well known subjects in the medieval era were rediscovered by western scientists after 16th century AD.

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Abstract — WCN 2013

No: 1717

Topic: 36 — Other Topic

Rhinocerebral mucormycosis: Clinical features and prognosis, two case reports and review of the literature

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Background: Rhinocerebral mucormycosis is a rare opportunistic fungal infection of the upper airways. Cerebral involvement is exceptional and life threatening.

Objective: We report two cases of rhinocerebral mucormycosis and we discuss their clinical features and prognosis.

Case report: Patient A, a 60 year old diabetic woman presented with fever, acute multiple right cranial nerve palsy (Garsin's syndrome). Few days later, a stroke with left side paralysis occurred. Cerebral-MRI showed right middle cerebral artery stroke with occlusion of the right carotid and a thickening of cavum mucosa. CSF analysis showed lymphocyte-type meningitis. Viral, bacterial and fungal serologies and cultures were negative. Cavum biopsy confirmed the diagnosis of mucormycosis.

Patient B, a 69 year old man with history of mucosa-associated lymphoid tissue pulmonary lymphoma, has presented a left fronto-temporal and retro-orbital headache with exophthalmia for one year. Cerebral-MRI showed liquid filling ethmoidal cells with a thickening of the left maxillary sinus mucosa. Facial CT-scan showed a left retro-orbital tissular process with bone lysis. Biopsy confirmed the diagnosis of orbital mucormycosis.

Discussion: Rhinocerebral mucormycosis is a life threatening infection and outcome is highly dependent on early diagnosis and treatment. Diagnosis is based on the biopsy. Both patients were treated by Amphotericin B, Fluconazole IV for 4 weeks. The outcome was good with clinical, biological and radiological improvement for patient B, while patient A had died of a massive pulmonary embolism.

Conclusion: Our observations highlight the various clinical features of rhinocerebral mucormycosis. Some features like cranial nerve palsies and strokes should be considered of severe prognosis.

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Abstract — WCN 2013

No: 2210

Topic: 36 — Other Topic

An unusual case of subacute sclerosing panencephalitis (SSPE) with cognitive decline and simultanagnosia

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Objective: To introduce a case of subacute sclerosing panencephalitis (SSPE) presenting with cognitive decline, hemiparesis, simultanagnosia, and diagnostic MRI findings.

Case report: A 21-year-old male patient was admitted to our hospital with hemiparesis, hesitant speech and progressive cognitive decline in the absence of myoclonus. His sister defined that the patient had an epileptic seizure one month ago and then developed disorientation. Neurological examination revealed prolongation of reaction time, disorientation of time and person, slight hemiparesis on the right side, hyperactive deep tendon reflexes and simultanagnosia. T2-weighted and FLAIR MR images showed extensive gray matter changes involving both temporal and parieto-occipital regions that are more prominent on the left side. EEG revealed recurrent periodic sharp and slow paroxysms. Extensive serum and BOS analysis for metabolic and infectious diseases revealed negative results. A diagnosis of SSPE was confirmed by elevated serum and BOS measles IgG titers.

Conclusion: SSPE is a slowly progressive degenerative disease of the central nervous system caused by a latent defective measles virus infection. Hallmarks of this entity include progressive cognitive decline, myoclonia, a generalized periodic pattern on EEG, and deep white matter abnormalities on MRI. In our case, although patient and relatives did not define any myoclonic jerks, we suggested a diagnosis of SSPE according to especially cognitive decline and MRI results. EEG and CSF examinations confirmed this disease.

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Abstract – WCN 2013

No: 2447

Topic: 36 – Other Topic

Carcinomatous meningitis from cervix uteri carcinoma

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A right handed female patient, 52 years old, was presented at emergency with a headache with nausea, vomiting and seizures; the symptoms started two weeks ago with nausea and vomiting. At another diagnostic center CT was performed on the head that was normal; after she was released from hospital she had an episode generalized seizure. The patient was diagnosed seven years ago with cancer of uterine cervix T1b. She had surgery, and then she was treated with chemotherapy. Six months before PET scan was performed where bone metastasis, mediastinal and peritoneal metastasis were seen, she has eight cycles of chemotherapy. After chemotherapy PET scan was performed, and full metabolic regression of the lesion was found. The neurological symptoms started two weeks after the last cycle of chemotherapy.

The patient is somnolent and has dysarthric and monotone speech, left monoparesis, bilateral Babinski, and meningeal signs.

MRI of the head and the spine with gadolinium revealed multiple vertebral metastases; lumbar puncture was performed on multiple areas of contrast enhancements of meningeal tissue in the brain and at the thorachal region. CSF histopathology examination revealed squamous cell poorly differentiated carcinoma. After oncologic consultation, it was recommended that the patient should have intrathecal methotrexate treatment but that was refused by her family members. Carcinomatous meningitis from uterine cervix-related malignancies is a rare complication; its incidence has increased probably survival time from the primary disease is extended.

This rare complication occurs in the advanced stage of cervical cancer. MRI of the head is a useful tool for the diagnosis but the lumbar punctures for neoplastic cells are the gold standard, and the prognosis remains poor.

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Abstract – WCN 2013

No: 799

Topic: 36 – Other Topic

Inborn errors of metabolism: Characterization of a group of patients with onset during adulthood

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Background: Inborn errors of metabolism (IEM) are a heterogeneous group of genetic conditions, with adult-onset occurring in 20% of patients.

Objective: Clinical characterization of patients with adult-onset IEM.

Methods: Identification of patients with adult-onset IEM who were screened for biochemical or molecular metabolic studies from January 2008 to December 2012. Review of clinical files.

Results: 142 patients were identified, and 56 were excluded after a clinical review identified symptoms' onset in childhood/adolescence. Of the remaining 86 patients, 22 had a neuromuscular disorder, 22 with stroke/TIA, 12 with movement disorder, 12 with leukodystrophy, 9 with neuropsychiatric symptoms, 6 with epilepsy, and 3 with optic neuropathy. In 17 patients it was possible to establish a diagnosis of IEM, 10♀, with an average age of onset of 38 years (20–63 years). When considering a clinical presentation, an IEM was confirmed in 22.7% of patients with neuromuscular disorder (5 with mitochondrial cytopathies, 2 of which have progressive external ophthalmoplegia), 22.2% have neuropsychiatric manifestations (2 with mitochondrial cytopathies), 18.2% with stroke/TIA (2 have Fabry's disease and 2 with mitochondrial cytopathies, one of which was MELAS), 18.2% have a movement disorder (3 with Wilson's disease, 1 with mitochondrial cytopathy), and 16.7% have leukodystrophies (1 with Krabbe's disease, 1 with mitochondrial cytopathy). In 35 patients other disorders were identified, in 18 a definite diagnosis was not established and in 16 investigations are still ongoing.

Conclusion: We wish to draw attention into adult-onset IEM. In our group of patients an IEM was confirmed in 19.7%, allowing the institution of appropriate treatment and genetic counseling. We believe that a systematic approach guided by clinical presentation will allow the identification of a significant percentage of these rare diseases.

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Abstract – WCN 2013

No: 2480

Topic: 36 – Other Topic

Concomitant radio-chemotherapy with CCNU after radical removal of newly diagnosed glioblastoma: Preliminary results of a single center retrospective study

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Background: When given concurrently with radiotherapy, Temozolomide has led to improved survival in GBM patients raising the possibility of its radiation potentiation.

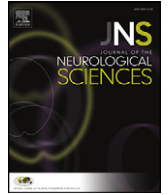
Objective: This study was conducted to determine the relative contribution of concomitant CCNU in patients with newly-diagnosed GBM.

Patients and methods: We identified all patients operated on for a supratentorial GBM and further treated with radiotherapy and CCNU-based chemotherapy either in a concomitant and adjuvant (group 1) or adjuvant only (group 2) setting. The primary endpoints of this study were progression-free survival (PFS), overall survival (OS) and the secondary endpoint was the toxicity.

Results: Forty-two patients (group 1, n = 20; group 2, n = 22) were included in this study. The two arms were well balanced according to their baseline characteristics (including age, KPS). The median follow-up was 15.7 months (95% CI, 12 to inf months). A borderline statistical difference (p = 0.07) was observed according to the PFS between two groups (12 vs 8 months). The median survival OS was 19 months (95% CI 14.1 and 24.7 months) with RT plus CCNU and 11.6 months (95% CI 11.2 and 17.2 months) with adjuvant group, p = 0.09 by the log-rank test. Hematological toxicity was absent during concomitant chemo-radiation and mild (9%) in adjuvant therapy.

Conclusions: Concurrent and adjuvant CCNU is associated with improved survival compared to adjuvant CCNU alone. These results highlight the contribution of CCNU in the radiation effect when given concomitantly.

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Topic: 36 - Other Topic

Abstract – WCN 2013

No: 2484

Topic: 36 – Other Topic

A case of spontaneous intracranial hypotension with cerebellar symptoms

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Spontaneous intracranial hypotension (SIH) is a rare clinical syndrome caused by spontaneous cerebrospinal fluid (CSF) leak, mainly characterized by postural headache and low CSF pressure. Besides postural headache, common accompanying clinical features include neck pain, nausea, vomiting, dizziness, vertigo, ataxia, blurred vision, and diplopia. More severe neurological manifestations such as parkinsonism and bulbar weakness occur infrequently.

Diagnosis of SIH should be considered in patients presenting with postural headache, which cannot be attributed to another cause. Neuroimaging, particularly MRI, is very useful in establishing the diagnosis and in identifying the site of leak.

Herein, we describe a case with postural headache, normal CSF pressure, and typical MRI findings of SIH. However, our case had quite atypical symptoms such as dysarthria, dysphagia, and ataxia for SIH.

A 64-year-old man, previously healthy, presented with postural headache, nausea, vomiting, dysarthria and ataxia. Onset of symptoms was 2 years ago. His medical and family histories were unremarkable. There were no trauma and surgery history especially on the spinal. On neurological examination, there were dysarthria, ataxia, bilaterally extensor plantar response, and bilateral dysmetria. Laboratory findings were normal. Opening pressure of CSF and the other biochemical tests were normal. Coronal and sagittal T1-weighted gadolinium-enhanced MRI revealed pachymeningeal enhancement, sagging of the brain, effacement of the perimesencephalic cistern.

In conclusion, the clinical spectrum of SIH can be wide as to include cerebellar and bulbar symptoms. The presence of clinical suspects for SIH and MRI findings is very important for diagnosis.

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Abstract – WCN 2013

No: 1889

Topic: 36 – Other Topic

Clinical features and outcome of ALS-like disorders: Two year follow-up of three cases and review of the literature

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Background: Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disease of unknown etiology. Various ALS-like disorders mimic the clinical feature of ALS, but prognosis is thought to be better. **Objectives:** We report 3 cases of ALS-like disorders and we discuss the clinical, electrophysiological features, etiologies and the outcome of this disorder.

Case reports: Three patients (2 men, 1 woman, mean age: 51 years) has presented with limb weakness and wasting associated with brisk muscle stretch reflexes and fasciculation, without any sensory involvement. EMG showed a motor neuron disease in 3 cases. Etiologic investigation had found respectively a sigmoid adenocarcinoma, a Gougerot–Sjögren syndrome (GSS) and a monoclonal gammopathy. All patients underwent etiologic treatment and had been followed for 2 years. Two patients have been stabilized without reversibility of the symptoms. The disease course has just slowed down in the third patient (with monoclonal gammopathy), which may imply that underlying etiology was not correctly treated.

Discussion: ALS-like must be searched in case of an anterior horn disease by an exhaustive assessment. Treatment of the causing pathology has been reported to improve the disease course and even reverse the symptoms, especially in infectious and toxic causes. In our patients, handicap didn't improve; instead we noted a stabilization or slow evolution of the symptoms, which extend the life span.

Conclusion: The importance of recognizing ALS-like syndromes lies in the potential reversibility of the disease, or at least, a delayed mortality.

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Abstract – WCN 2013

No: 2438

Topic: 36 – Other Topic

Primary dural MALT lymphoma mimicking a meningioma: A case report

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Background: Primary dural lymphoma is a rare subtype of primary central nervous system lymphoma. Mucosa-associated lymphoid tissue (MALT) lymphomas are the most common subtype of non-Hodgkin's lymphomas primarily involving the dura. They usually appear as single or multiple extra axial lesions with homogeneous contrast-enhancement mimicking a meningioma.

Case report: A 54-year-old woman presented with a two-month history of ataxia and left hearing loss. MRI showed an extra-axial tumor arising from the left cerebellar tentorium, measuring 5 cm × 3,7 cm × 2,3 cm, infiltrating the left internal auditory canal and the left foramen lacerum, T2 hyperintense, and T1 isointense, with homogeneous contrast enhancement. This pattern was highly suggestive of a meningioma. The biopsy showed a lymphoid infiltrate, without evidence of B-lymphocyte clonality. The patient improved after incomplete surgical resection and corticoid therapy. She was admitted one year later, for a one-month history of unusual headache, with nausea and vomiting. Brain MRI showed an enhancing tumor of the left tentorium with mass effect. A stereotactic biopsy established the diagnosis of a mucosa-associated lymphoid tissue, a subtype of low-grade marginal zone non-Hodgkin lymphoma (CD 20 +, CD3, CD79A).

Discussion: This case demonstrated the potential pitfall of imaging findings of dural MALT lymphoma, a rare condition which can lead to a misdiagnosis of meningioma.

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Abstract – WCN 2013

No: 1142

Topic: 36 – Other Topic

Depression in LRRK2-associated Parkinson disease

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Background: The prevalence of the leucine-rich repeat kinase 2 gene (LRRK2) at the PARK8 locus on chromosome 12p11.2–q13 in Parkinson's disease (PD) varies according to the geographical and ethnicity origins. In North Africa it represents about 35.7% of sporadic forms.

Depression is one of the most common non-motor symptoms of Parkinson's disease (PD). However the prevalence and the pattern of depression are lacking for LRRK2 (leucine-rich kinase 2)-associated PD patients.

Purpose: The purpose of this study was first to describe depression in LRRK2-associated PD patients, secondly to look for the impact of depression on activities of daily living and finally to compare prevalence of depression between LRRK2-associated PD patients and sporadic PD patients.

Methods: 40 patients diagnosed with PD-related LRRK2 G2019S mutation and 60 patients with sporadic PD were included in the study. All patients undergoing antidepressant, hypotension, corticoid or anti-tuberculosis treatment were excluded. Depression was screened by the Geriatric Depression Scale-long form. The impact on activities of daily living was assessed by Schwab and England Scale (indexed in UPDRS).

Results and discussion: Ninety six patients fulfilled inclusion criteria (thirty patients with PD-related LRRK2 G2019S mutation and sixty-six with PD sporadic form). 23 patients with mutation and 57 with sporadic PD had depression. Our results suggest that depression in PD is associated with increased disease severity and limitations in the activities of daily living but not with the l-dopa dose.

Conclusion: The association between depression and PD-related LRRK2 G2019S mutation needs further clarification.

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Abstract – WCN 2013

No: 2448

Topic: 36 – Other Topic

Psychological patterns of patients with recurrent brain tumor

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Background: Patients with brain tumor are rarely assessed for quality of life and psychological variables and even fewer studies have investigated these features in patients with a recurrence of brain tumors.

Objective: The aim of the present study was to investigate the reaction to the illness of patients with recurrent brain tumors.

Patients and methods: We enrolled 81 patients with recurrent CNS tumors. Multidimensional aspects of quality of life were assessed through "Functional Assessment of Cancer Therapy-Brain"; "Hospital Anxiety and Depression Scale" and "Psychological Distress Inventory". KPS was used to evaluate functional status of patients.

Results: The distress of our sample was significantly lower than the distress reported in patients affected by other cancer types. All mean Fact-Br sub-scale scores were significantly lower in patients as compared with normative data. There were significantly lower scores in our sample for functional well-being and for social/family well-being than a sample of patients with primary brain tumors. Unexpectedly, emotional well-being mean score was significantly higher in our recurrence sample than in patients with primary brain tumors. The anxiety seems not to be influenced by relapse diagnosis; depression instead was significantly higher than the normative data.

Conclusion: The recurrence of a brain tumor can have a major impact on patients' condition and their psychological response than KPS levels. The dissociation between patients' judgment on their quality of life (bad excepted for emotional) and their reported distress (low) is the most intriguing finding, suggesting highly preserved coping strategies in the emotional sphere despite intact judgment and disease awareness.

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Abstract – WCN 2013

No: 2456

Topic: 36 – Other Topic

Hirayama disease and IgA deficiency: An unusual coincidence or a complex syndrome

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Background: Hirayama disease (HD) is a rare lower motor neuron disease. Although various possibilities have been proposed for the etiology of HD, the existing theories are insufficient.

Objective: Here we present the first case of HD in Bulgaria and discuss the pathophysiological, immunological and imaging characteristics of the disease.

Patients and methods: The patient had a complete and thorough medical, neurological, neurophysiological (EMG, SSEP, MEP), radiological (3 T MRI), and immunological evaluation.

Results: A 21-year-old man presented with a 2-year history of gradual loss of muscle mass and weakness in his left hand extending to the forearm. Neurological examination revealed moderate atrophy and slight weakness of the intrinsic left hand and forearm flexors, as well as slight weakness of some intrinsic right hand muscles. EMG showed chronic neurogenic changes in both hands. Conventional cervical MRI and cervical MRI in the fully flexed position demonstrated typical features for HD. Additionally, immunological tests revealed selective IgA deficiency.

Conclusion: The most widely accepted hypothesis for the underlying pathogenic mechanism of HD is cervical myelopathy based on the spinal flexion mechanism. However, it does not explain the natural history of the disease. Several recent studies report different immunological changes in patients with HD. Together with typical MRI findings, our patient also has a selective IgA deficiency. This data suggests that additional immunological mechanism may play a role in the pathogenesis and progression of HD, but this finding warrants further study.

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Abstract – WCN 2013

No: 2548

Topic: 36 – Other Topic

Macular tuberculoma and optic neuritis: Rare association with tuberculosis meningo-encephalitis

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Introduction: Tuberculosis is an endemic disease responsible for death and morbidity in developing countries.

Case report: A 50-year-old man with no medical history was admitted to the emergency department for meningism associated with fever and confusion. The ophthalmic exam showed a decline in left visual acuity, a reduction of light perception, VIth nerve left oculomotor paralysis, and ocular fundus demonstrating a yellow tumor located on the posterior segment, measuring 1.5–2 mm, papillomatous and prominent in the vitreous cavity. Fluorescein angiography showed a peritumoral choroiditis area, miliary tubercles of the choroid, and sectorial papillomatous edema. Color retinography unmasked inflamed posterior vitreous areas. Echography demonstrated a 4- to 5-mm oval hyper-echogenic and calcified tumor along with hyper-echogenic vitreous areas. Lumbar puncture showed lymphocytic meningitis associated with hyponatremia. The CT scan and MRI demonstrated optic neuritis. The antibiotic therapy was initiated and the outcome was favorable.

Discussion: This case report shows the importance of systematic ocular fundus in the presence of systemic tuberculosis and outlines the assessment of color retinography to unmask vitreous lesions. It shows the importance of radiological imaging in the semiological study of orbital and cerebral lesions.

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Abstract – WCN 2013

No: 2541

Topic: 36 – Other Topic

Intracranial dural arteriovenous fistula: A rare cause of myelopathy

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Introduction: Intracranial dural arteriovenous fistula (DAVF) is a rare acquired abnormality whose clinical manifestations are dependent on the pattern of their venous drainage.

Case report: A 50 year old male presented with a progressive gait disturbance which started 4 months before. He also developed urinary incontinence and constipation. Cervical MRI revealed an extensive spontaneous hyperintensity involving medulla oblongata and cervical medullar transition until C3 suggesting a tumoral or inflammatory/infectious lesion. The serological and CSF syphilis reactions were positive. He was started with penicillin. In 3 weeks the lesion progressed rapidly to flaccid tetraplegia with a respiratory dysfunction. No other symptoms

were found including headache or cervical pain, tinnitus, fever or previous trauma. The cervical MRI showed the same lesion extending now until C5 with epidural venous plexus dilatation and “flow voids” suggesting an arteriovenous shunt. The angiography confirmed a DAVF at the level of jugular foramen fed by the jugular and hypoglossal branches of ascendant pharyngeal artery and a petrous branch of middle meningeal artery; venous drainage was to a suboccipital vein and this one drains to a perimedullar vein being that this phenomenon was responsible for the clinical presentation. The fistula was occluded and in the control angiography no abnormality was found. At a neurological exam he presents a flaccid tetraparesis grade 3 (MRC) and regained partial autonomous ventilation.

Conclusion: Rare reports of intracranial DAVFs with perimedullary venous drainage were found in the literature. These usually present with myelopathy, rarely hemorrhage. Endovascular therapy is the gold standard. An attempted recognition of this entity is crucial for a better neurological outcome.

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Abstract – WCN 2013

No: 2547

Topic: 36 – Other Topic

Isolated psychosis with an organic etiology: A diagnostic challenge

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Introduction: In a patient presenting with neuropsychiatric symptoms, especially in the absence of a previous psychiatric history, is obligatory to exclude an organic etiology.

Clinical cases:

1. A 68 yo female, with history of myocardial infarction, spontaneous abortion (1st trimester), persistently positive IgM anticardiolipin antibody titers presented with a 3 month neuropsychiatric syndrome (euphoria, disinhibition, hyper-religiosity, delusions of grandeur, executive dysfunction, visual hallucinations) and later seizures. She had no psychiatric history. Brain MRI showed old subcortical and basal ganglia ischemic lesions. She met the criteria for primary antiphospholipid syndrome. She improved with hypo-coagulation, anti-epileptics and neuroleptics.
2. A 47 yo female presented with Behçet's disease (BD) which was diagnosed at 32 yo (recurrent ulcers, uveitis, erythema nodosum). By then she had psychiatric symptoms, and was stabilized with neuroleptics. At 45 yo she had active BD and started with disinhibition, hyper-religiosity, delusions of grandeur and obsessive cleaning. Brain MRI excluded acute lesions, electroencephalogram and CSF were normal. Psychiatric symptoms improved under quetiapine and sodium valproate.
3. A 51 yo female, with unremarkable personal history, was admitted in hospital to donate a kidney to her husband. Two days after surgery she started behavioral (agitated, disinhibition, obsessive cleaning) and sleep changes, followed by persecutory delusion. CSF showed pleocytosis (45 leucocytes/uL–44 lymphocytes, 1 polymorphonuclear), proteins 0.26 g/dL and glucose 0.66 g/dL. PCR analysis was positive for enterovirus. She recovered with supportive treatment and was discharged asymptomatic.

Discussion: We presented rare causes of organic psychosis. Psychiatric symptoms, even if isolated, can be the form of presentation of neurological disorders. It highlights the importance of an early recognition in order to ensure proper treatment.

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Abstract – WCN 2013**No: 2559****Topic: 36 – Other Topic****Prophylaxis of pediatric headache with Topiramate**

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Background: Headache (HA) is a very common neurological problem in the pediatric population. Topiramate (TPM) is currently approved in children 2 years of age and older as an adjunctive treatment of partial and primary generalized seizures. Historically, anticonvulsants have proven effective for adult HA and, in fact, given this, and the availability of safety data on TPM in the pediatric population, it is an agent that holds promise for HA prophylaxis in children. There is at least one small case report indicating efficacy in pediatric migraine but there are no studies to date including patients with CDH and mixed type HA.

Objective: To determine if Topiramate is an effective agent in the treatment of headache, including migraine, chronic daily headache (CDH) and mixed type headache, in the pediatric population.

Methods: A retrospective chart review was conducted of patients seen in the pediatric HA clinic at Children's Hospital and treated with TPM.

Results: 30 patients were treated with TPM. Responses to TPM were as follows:

Ha free (11); significant (>75%) reduction in headache (9); moderate (50–75%) reduction in ha (5); four patients stopped TPM secondary to side effects; and three patients failed to respond.

Overall, 67% of the patients treated with TPM achieved HA freedom or significant improvement. In those patients with CDH, 73.6% achieved HA freedom or significant improvement and 36.8% became HA free.

Conclusions: TPM is a well tolerated and efficacious medication for the treatment of pediatric HA and is effective in much smaller doses than generally needed when treating seizures.

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Abstract – WCN 2013**No: 2555****Topic: 36 – Other Topic****Neurophysiological study in infantile neuroaxonal dystrophy: Retrospective review of seventeen North-African patients**

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Background: Infantile neuroaxonal dystrophy (INAD) is a rare autosomal recessive neurodegenerative disorder due to PLA2G6 mutations.

Objective: To characterize both the EEG patterns and EMG findings in INAD. To compare EEGs and EMGs according to the age in patients who had serial exams.

Patients and methods: Over 7 years (2005–2012), 17 patients were diagnosed INAD according to Nardocci criteria and carrying PLA2G6 mutations. EEGs were performed in 14 patients and reanalyzed according to background activity, fast rhythms, and epileptiform activity. EMGs were obtained in all but one patient.

Results: EEG was normal in seven patients and abnormal in seven with fast rhythms in six all aged more than 2 years old with additional few slow waves and spikes in three of them, while one child had generalized spike waves predominantly in the centro-temporal regions. Two patients with fast rhythms on EEG had earlier investigations that reported normal findings. EMG showed axonal motor neuropathy in eleven patients, sensory and motor neuropathy in two patients and horn cell involvement in one. The two remaining patients had normal

EMG which was performed before 2 years of age. Four patients with abnormal EMGs had earlier exams that were normal.

Conclusion: Although not specific, neurophysiological findings, namely the presence of fast rhythms on the EEG and signs of chronic denervation are highly suggestive of INAD in the appropriate clinical and imaging context. EEG pattern and EMG findings depend on the stage of the disease course and normal exams in the early stage do not exclude the diagnosis.

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Abstract – WCN 2013**No: 2564****Topic: 36 – Other Topic****Acoustic noise leads to an attention shift indicated by brain direct current (DC) potential changes**

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Background: Acoustic environmental noise is known to adversely affect information processing via attention mechanisms. Facilitation and inhibition of information processing are basic mechanisms of selective attention. Such mechanisms can be investigated by analyzing information processing under conditions of external directed attention (intake of environmental information) versus internal directed attention (rejection of environmental stimuli). This study investigated effects of noise on brain direct current (DC) potential shifts – which are discussed to represent different states of cortical activation – of intake and of rejection tasks.

Objective: It was hypothesized that without noise rejection tasks are associated with more positive DC potential changes compared to intake tasks and that under noise both kinds of tasks are associated with positive DC shifts as an expression of cortical inhibition caused by noise.

Material and methods: DC potential shifts were analyzed from scalp EEG at 16 standard locations in 46 persons. Noise effects were investigated by irrelevant speech and by white noise, and attention tasks consisted of figural and verbal tasks to take into account modality effects.

Results: Without noise, rejection tasks showed more positive DC potential changes compared to intake tasks, whereas under noise, this difference disappeared and both kinds of tasks led to more positive DC shifts.

Conclusion: More positive DC shifts under noise for intake tasks could represent an additional cognitive load as an expression of an inhibition of environmental information. Results indicate that noise leads to an attention shift towards an environmental information rejection mode.

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Abstract – WCN 2013**No: 2450****Topic: 36 – Other Topic****The relative interpeak latency I–V as an effective parameter for evaluation of neuropathophysiological disorders in neuters and children**

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Background: The relative interpeak latency I–V (rIPL I–V) is a parameter for description of the delay of conduction in auditory

tract, observable in brainstem auditory evoked potential (BAEP) examination. It is defined as the non-dimensional value, equal to the real number of the standard deviations (SD) of mean IPL I–V, calculated for a representative group of healthy subjects. Exemplarily, rIPL I–V of 2.0 means that the value of the IPL I–V for an examined child is delayed by 2.0 SD compared to a healthy child at this age.

Objectives: Implementation of the rIPL I–V parameter for neuroresearch on a larger scale.

Material and methods: We present the exemplary studies of BAEP in cytomegalia Brown syndrome and healthy subjects in children of ages from 1 week to 39 weeks.

Results: Statistical analysis revealed the predominance of the proposed relative interpeak latency I–V (rIPL I–V) parameter over frequently used absolute IPL I–V.

Conclusion: The relative IPL I–V value is an objective parameter for a description of the transmission in the brainstem part of the auditory tract and it is very useful for plenty of neurophysiological disorder studies.

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Abstract – WCN 2013

No: 2499

Topic: 36 – Other Topic

Opsoclonus-myoclonus syndrome – A case report

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Background: In adults opsoclonus-myoclonus syndrome (OMS) should primarily lead to the investigation of either a paraneoplastic or parainfectious etiology.

Materials and method: We present a case of a 34-year old female, who was referred with a 10-day history of dizziness, nausea, chaotic eye movements and involuntary jerks following a non-specific viral infection. In addition, the patient complained of a long-standing history of non-specific lower abdominal pain.

Results: Positive findings upon full neurological exam were: slight dysarthria, myoclonal jerks, opsoclonus, loss of gaze fixation, postural instability, positive Romberg, severe gait ataxia and dysmetric finger–nose–finger test.

Blood work was unremarkable. CSF exam revealed 26 lymphocytes, normal protein, no oligoclonal bands, and no paraneoplastic, autoimmune or viral antibodies. Brain MRI showed 5 unspecific non-contrast enhancing white matter hyperintensities. Whole-body PET-CT revealed a metabolically inactive ovarian cyst. Brain FDG-PET showed hypermetabolic activity in the cerebellar vermis.

The patient was treated with IVIG and high dose prednisolone. Histological diagnosis of the ovarian cyst was a benign teratoma. At follow-up 2 months after surgery, some but not complete clinical recovery was noted.

Conclusion: In this young woman with OMS no underlying malignancy was identified. Paraneoplastic and viral antibodies were negative. A benign ovarian mature teratoma was found and removed. Two previous cases have been reported with ovarian teratoma as the underlying pathology in OMS.

We suggest that the ovarian teratoma was the cause of OMS in the present case as a differential diagnosis to postinfection cerebellitis and that this benign tumor should be considered in younger women with OMS.

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Abstract – WCN 2013

No: 2530

Topic: 36 – Other Topic

Relationship between paraneoplastic cerebellar degeneration and overexpression of C-erbB-2 gene in breast cancer

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Background: Case report of a 38-year-old woman with cerebellar ataxia.

Objective: Case report.

Patients and methods: The cerebellar ataxia manifested itself through dysarthria, nystagmus, finger-to-nose test – nose indicating severe impairment mostly left – coarse tremor of the upper limbs, with bradykinesia, with dysdiadochokinesia and Babinski sign and Hoffman negative, also presenting a history of nodulectomy in the left breast. Presented with a new nodule of 3.1 × 2.1 cm, diagnosed as infiltrating ductal carcinoma grade 2, estrogen receptor positive, progesterone receptor negative, C-erbB-2 positive 3+, ki67 of 27% and CK5 negative, the patient was referred to neoadjuvant chemotherapy and posterior quadrantectomy of the left breast. Imaging and laboratory tests showed no indication of metastases in the central nervous system or in any other system. The final neurological diagnosis was paraneoplastic cerebellar degeneration (PCD), a rare syndrome that occurs in less than 1% of breast cancers.

Results: The case associated to a literature review demonstrated a possible connection between PCD and C-erbB-2 strongly positive.

Conclusion: Although most of the series with PCD and antibody positive show that this disturbance rarely respond to treatment, early recognition of this rare complication of breast cancer is imperative, since there may be neurological improvement with appropriate therapy, for example, removal tumor, plasmapheresis, IVIG, cyclophosphamide, rituximab or glucocorticoids.

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Abstract – WCN 2013

No: 2526

Topic: 36 – Other Topic

Frequency and clinical presentation of dizziness in neurological emergency department

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Background: Sensation of dizziness occurs frequently, and may be caused by various different disorders rendering the differential diagnosis challenging in the neurological emergency department (NED).

Objective: We investigated the frequency and clinical presentation of different dizziness syndromes at our NED to improve diagnosis assignment during patient evaluation.

Patients and methods: We retrospectively analyzed medical records of patients who attended the NED with a chief complaint of dizziness/vertigo over a 5 month period (n = 100). We collected demographic data, information about clinical presentation (disease course, triggers, associating symptoms), previous history and co-morbidities, findings of the neurological examination and diagnostic tests.

Results: Dizziness was more prevalent among elderly (mean: 59.21 ± 19.54) and far more frequent among females (7:3). The most common cause of dizziness was benign paroxysmal positional vertigo (BPPV), and the 5 most common occurring syndromes (BPPV [21%], vertebrobasilar stroke [20%], internal medicine disorders [17%], psychiatric disorders

[15%], vestibular migraine [10%]) from the 12 established diagnoses covered 83% of all cases. Migraneous vertigo and psychiatric disorders were underdiagnosed, and BPPV was overdiagnosed (false positivity). One-third of patients presented reoccurring dizziness episodes, and 32.3% of them had psychiatric disorders. Chronic dizziness (>3 months) occurred most commonly (47.4%) in patients with a psychiatric disease. Isolated vertigo occurred in 50% of vertebrobasilar stroke cases. Seventy percent of patients with vertebrobasilar stroke had two or more vascular risk factors.

Conclusion: Careful history taking and bedside evaluation provided in most of the cases the base for diagnosis, and laboratory and imaging techniques only had additional confirming and supporting roles.

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Abstract – WCN 2013

No: 2485

Topic: 36 – Other Topic

Radio-neuroprotective effect of a phosphatidylcholine derivative in a rat model of irradiation

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Ionizing radiation plays major role in the treatment of brain tumours, but side-effects may restrict the efficiency of therapy. In the present study, our goals were to detect the functional and morphological changes that result in the administration of l-alpha glycerylphosphorylcholine (GPC), a deacylated derivative of phosphatidylcholine, in a rodent model of hippocampus irradiation.

Anaesthetized SPRD rats were subjected to 40-Gy irradiation of one hemisphere of the brain, with or without GPC treatment (50 mg/kg per os). After the irradiation the rats received GPC or the vehicle at the same time every second day at 5 days per week. The effects of partial rat brain irradiation on the spatial orientation and learning ability of rats were assessed in the Morris water maze (MWM) test, which was performed once before and two times after the irradiation. At the same time morphological changes were detected by MRI to define the time-point for histological examinations.

Irradiation of 40-Gy resulted in moderate neurological deficit both at the level of cognitive function and morphology after 4 weeks of irradiation. The MWM test was found to be a highly sensitive tool for the detection of neurofunctional impairment. We found that the place navigational function of the rats was impaired by the irradiation and the treatment with GPC could prevent this damage. Marked protective effect of the GPC was detected as concerns of the histopathological findings.

Targeted brain irradiation induced notable changes in the central nervous system. GPC supplementation provides significant protection against irradiation-caused functional and morphological disturbances.

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Abstract – WCN 2013

No: 2459

Topic: 36 – Other Topic

Carotid intima-media thickness (IMT) and indices of arterial stiffness as the predictors of ischemic cerebral infarction (ICI) and myocardial infarction (MI)

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Background: New specific vascular inflammatory marker LP-PLA2 may predict future cerebrovascular and cardiovascular events in patients with iCI, MI, coronary artery disease (CAD) and arterial hypertension (AH). In the past years there were discussions regarding its relationship with intima-media thickness (IMT), arterial stiffness and plaque morphology.

Objective: The aim of this study was to assess the changes in IMT, stiffness, LP-PLA2 and their mutual relationships in a prospective, multidisciplinary, multicentric study.

Materials and methods: Materials consist of 436 subjects divided in four subgroups

1. iCI (n = 171), mean age 69 +/- 11 years, men 52%,
2. CAD (n = 87) mean age 70 +/- 9 years, men 32%,
3. AH (n = 124) mean age 60 +/- 10 years, men 46%, healthy controls (n = 56), mean age 47 +/- 13 years, men 55%. In all subjects, neurological and cardiological examination, ICI confirmed by CT/MRI, SPECT, battery of biochemical/haematological investigations,

1. IMT using ultrasonography by radio-frequency data analysis.
2. Alx and PWV on using applanation tonometry, LP-PLA2 using ELISA. Statistical software STATISTICA Base Cz Version 10, Pearson-Spearman-Kandell and Whitney U tests.

Results: The study showed statistically higher values of all followed parameters iCI, CHD, AH comparing them to controls (p < 0.01–0.0001). Also statistically significant changes between AH:CHD, AH:iCI and CHD:iCI (p < 0.05–0.02) were documented. Close correlations between LP-PLA2 and IMT stiffness were documented in all followed groups.

Conclusions: Our results documented significant changes in IMT, aortic stiffness and LP-PLA2 in all followed groups (iCI, CAD, AH) compared to controls. A close correlation was found between these parameters and LP-PLA2. The measurements of IMT, stiffness and LP-PLA2 are very useful parameters for assessing cerebrovascular and cardiovascular risk. They represent significant prognostic power to ascertain subjects with an increased risk for onset and outcome of cerebrovascular and cardiovascular events.

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Abstract – WCN 2013

No: 2411

Topic: 36 – Other Topic

Changes of esophageal peristalsis in patients with non-erosive reflux disease and functional heartburn following non-invasive brain stimulation

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Background: Gastroerosive reflux is a widespread disease and, although pH-metric and proton pump inhibitor (PPI) tests represent important tools, the differential diagnosis between non-erosive reflux disease (NERD) and functional heartburn (FH) can be challenging.

Objective: The aim of this study was to evaluate differences of cortical excitability in patients with NERD and FH.

Materials and methods: Nineteen subjects (18–65 years) with GERD symptoms, PPI trial and an endoscopy ruling out esophagitis were enrolled. Esophageal manometry was performed before and during transcranial direct current stimulation (tDCS, 1.5 mA) of the right precentral cortex. Half of the patients were randomly assigned to anodal and the other half to sham stimulation. Distal wave amplitude and pathological wave percentage (peristaltic waves <30 mm Hg, simultaneous or not propagated waves, or absent peristalsis) were measured after the patients swallowed ten subsequent water boli. Lastly, a 24 hour pH-bilimetry was performed to subdivide NERD by FH. The values obtained before and during anodal or sham tDCS were compared by repeated-measures ANOVA with Time (before–during) as within-subject factor, and Stimulation (anodal-sham) and Sub-group (NERD-FH) as between-subjects factor.

Results: We observed a significant increase of distal wave mean amplitude ($p = 0.02$) and a decrease of pathological wave percentage ($p < 0.01$) either in NERD and FH subjects exclusively with anodal tDCS. No significant differences were documented in distal wave mean amplitude following anodal tDCS in NERD compared with FH patients, while post-hoc analysis confirmed that pathological wave percentage significantly improved after stimulation only in NERDs ($p < 0.001$).

Conclusion: Anodal tDCS can differently improve esophageal manometric features in GERD sub-categories.

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Abstract – WCN 2013

No: 2508

Topic: 36 – Other Topic

Cerebral malaria with corpus callosum splenium lesion

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Cerebral malaria is the most severe neurological complication of infection with *Plasmodium falciparum* malaria. Mortality is high and some surviving patients with sustained brain injury manifest as long-term neuro-cognitive impairments. Cerebral malaria is seen in 2% of malaria cases. In endemic areas it affects mainly children. Occurrence in adults is far less. Here we report a 27-year-old man with cerebral malaria who has a rare but suggestive radiologic finding for malaria. He has been admitted to our emergency department with fever, diarrhoea, muscle pain, headache and progressive drowsiness. He visited Sierra Leone, Africa 5 days ago. His physical examination revealed icterus, hepatosplenomegaly and dark urine. At his neurological examination, he was lethargic and had a score of 6 on the Glasgow Coma Scale. He has meningeal irritation signs with neck stiffness. Cranial MRI performed urgently and showed focal restricted diffusion in corpus callosum splenium in diffusion weighted imaging. Epileptic attacks have been observed and his EEG revealed generalised severe background slowing. Blood smears showed *Plasmodium falciparum*. Urgent antimalarial treatments have been administered and the patient regained consciousness. Anti-epileptic medication was progressively reduced and the treatment was finally stopped after normal EEG was obtained. Awareness of clinical and radiological features of malaria and cerebral malaria is really important for early recognition and treatment. Taking the visiting history of an endemic area is helpful for diagnosis.

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Abstract – WCN 2013

No: 2510

Topic: 36 – Other Topic

Differentiate neglect and hemianopia with semantic priming paradigm. Evidences in three patients with acquired brain lesion

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Background: A prime word in neglected visual field should determine semantic activation effect even when the patient does not consciously perceive it, while no activation effect should be expected if the prime occurs in a blind hemifield.

Objective: We aim to provide evidences on different information processing between neglect and hemianopia and find a task which could differentiate between them.

Methods: We enrolled three patients: two with left neglect; one with a bilateral lesion which determined left neglect and right homonymous hemianopia.

Task: Patients were required to press the space bar when a target word was deemed as “living entity”; each target was firstly preceded by a prime word occurring in six possible positions on the horizontal line of the screen.

Three different prime-target conditions were present: related, unrelated and neutral.

Results: We analyzed results as a multiple single case with a Crawford analysis: patients with left neglect showed a significant activation effect in all portions of space (in average 60 ms quicker in “related condition”, $p < .05$), except when the prime occurred in the extreme left position. The patient with left neglect and right hemianopia showed semantic activation in the left neglected space (related condition, $p < .001$), but not in the right space (hemianopic field).

Conclusions: These results confirm the initial hypothesis: we found a semantic activation in the space affected by neglect but not in the hemianopic field; on these first data our task seems to allow a differentiation between the two deficits even though a test on a larger sample of patient is now in progress.

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Abstract – WCN 2013

No: 2494

Topic: 36 – Other Topic

Ictal characteristics of common vestibular disorders

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Background: Peripheral and central vertigo are associated with nystagmus, the spatiotemporal characteristics of which may point to its origin.

Objective: To capture the ictal attributes of episodic vertigo.

Methods: Thirty patients presenting to a neuro-otology clinic with episodic vertigo were taught to record spontaneous, gaze-evoked and positional nystagmus, using custom-made lightweight portable video-goggles. Monocular video data were collected at 30 Hz using infrared cameras. Portable audiometry was recorded when aural symptoms were present.

Results: Six subjects fulfilled criteria for clinically definite Meniere's Disease and had episodic vertigo associated with horizontal/torsional nystagmus; three demonstrated ictal fluctuation in hearing threshold (15–60 dB). Their horizontal slow phase velocities (SPV) ranged between 10 and 52°/s and the SPV profile was constant over a 60 s recording interval. Two undertook consecutive recordings over 10–15 min intervals

and demonstrated irritative, parietic and recovery nystagmus. Sixteen subjects fulfilled the criteria for clinically definite (6) or probable (10) vestibular migraine. Their ictal nystagmus was characterized by spontaneous horizontal (6) vertical upbeating (4) or downbeating (4) nystagmus with a constant SPV profile (SPV range: 5–25°/s) and 2–3 fold enhancement during positional testing. Four subjects had short-lived paroxysmal positional nystagmus consistent with posterior canal BPV. The slow phase velocities, plotted as a function of time, could be fitted into a 5th–8th order polynomial curve which peaked at 3–10 s (range: 15–95°/s) and had declined to 0 by 60 s.

Conclusion: Home-video nystagmography is feasible, enables a detailed study of the ictal profiles of central and peripheral vestibulopathies and facilitates differentiation between common causes of episodic vertigo.

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Abstract – WCN 2013

No: 2491

Topic: 36 – Other Topic

Neurosarcoidosis: A Tunisian case series

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Background: Neurosarcoidosis (NS) more commonly occurs in the setting of systemic disease. Neurologic manifestations are found in 5 to 20% of cases and symptoms may be mild or even severe. Clinical presentations and imaging findings in nervous system are highly variable.

Objective: The aim of this study was to determine the different clinical and radiological presentations of neurosarcoidosis and describe the evolution under corticosteroids.

Materials and methods: A retrospective study was conducted from 2005 to 2013, in the adult department of neurology at the National Institute of Neurology Mongi Ben Hamida in Tunis, including seven patients hospitalized for suspected neurosarcoidosis.

Results: The mean age of our patients was 36.85 ± 11.61 years with a female predominance (sex ratio 1.33). Multiple cranial nerve involvement was the most common clinical sign (6/7). Five patients had a headache with an increased intracranial pressure in three patients. Only one patient developed seizures. Endocrine disorders were noted in 3 patients. Lymphocytic meningitis was noted in 5 patients. MRI showed several radiological aspects: hydrocephalus, pachymeningitis, sellar and suprasellar infiltrates and hyperintense cranial nerves. The mainstay of medical treatment in neurosarcoidosis is corticosteroids, and all patients received this therapy alone. Six patients improved or stabilized, whereas only one deteriorated.

Conclusion: Sarcoidosis should be suspected in patients with unexplained neurological symptoms even in the absence of any systemic manifestations. Biopsy of neural tissue is often not practical and the diagnosis must be inferred through other tests, often coupled with biopsy of extra neural organs.

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Abstract – WCN 2013

No: 1770

Topic: 36 – Other Topic

Peripheral paraneoplastic neuropathy, an uncommon clinical onset of sigmoid cancer

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Introduction: Peripheral paraneoplastic neuropathy (PPN) is rare. Multiple myeloma and tumors of the lung, breast, kidney and ovary are most commonly implicated. Colorectal cancer rarely presents with neurological paraneoplastic disorder.

Objective: We report an unusual rectum adenocarcinoma presentation revealed by a sensory neuronopathy. Our message is that an occult malignancy should always be researched when we have subacute sensory neuronopathy.

Case report: A previously healthy 56-year-old woman was admitted because of progressive ascending weakness and ataxia gait, beginning 6 weeks ago. Neurological examination revealed a flaccid and areflexic quadriparesis associated with glove and stocking distribution of sensory loss. No sensory conduction velocity was detected in neurophysiological studies. Complete microbiological and virological investigations on blood, urine and CSF specimens, anti-nuclear antibody and tumor markers were negative. Lip biopsy also was normal. Only Anti-Hu antibodies were positive on blood and CSF. A whole body computed tomography scan was completely unremarkable. At the time of the diagnostic workup an episode of rectal bleeding occurred. Colonoscopy demonstrated an exophytic process of the rectum at 15 cm from the anal verge. No improvement was obtained after surgery and intravenous immunoglobulin. Our patient died from dysautonomic disorder related to her neuropathy.

Discussion and conclusion: Only 0.05% of cancer patients suffer from paraneoplastic neurological syndrome, of which less than 10% had rectal cancer.

We conclude that, an occult malignancy should always be researched when we have subacute sensory neuropathy.

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Abstract – WCN 2013

No: 2694

Topic: 36 – Other Topic

Time related patterns of blood pressure response in patients with orthostatic hypotension during prolonged tilt testing

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Introduction: Orthostatic hypotension (OH) is a common, frequently under diagnosed, disabling and treatable condition with prevalence up to 30%. OH is usually diagnosed with the tilt test (TT). The optimal duration of TT needed to detect OH, or whether standing longer than 3 min contributes to diagnosis, remained unclear. We aimed to assess various time-related patterns of blood pressure (BP) responses in patients with OH.

Methods: The protocols of prolonged (40 min), vertical position drug-free TT were studied in 745 consecutive individuals during 2010–2011 years.

Results: 296 subjects (40%) among the 745 referred for TT had OH. This group included 36 of the 83 patients (30%) younger 20 years (young group, YG), and 116 of the 260 subjects (46%) who were elder 65 years (elderly group, EG). The female: male ratio was 66:33 for the OH-YG group and 55:45 for the OH-EG group. Around 25% of the OH-EG patients developed a decrease only in systolic BP (SBP), around 25% only in diastolic BP (DBP), and 50% in both. In OH-EG, SBP dropped within 5–35 min, while DBP dropped within 10 min.

Conclusions: Individuals referred for TT had a 40% probability for OH. The prevalence of systolic and diastolic OH was similar, but their temporal patterns were different. Extending TT beyond 35 min is important to detect delayed systolic OH.

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Abstract – WCN 2013**No: 1846****Topic: 36 – Other Topic****Vestibular manifestations in a migrainous population**C.G. Videla, A. Bissonni, E. Doctorovich, E. Cristiano. *Neurology, Hospital Italiano de Neurología, Buenos Aires, Argentina*

Introduction and objectives: Vestibular symptoms are common manifestations observed in patients with migraine. This coincidence is greater than the statistic possibility of presenting two high prevalence diseases in general population.

The aim of this study is to describe the neuro-otological findings in patients assisted by a headache unit during a year.

Material and methods: 460 patients evaluated in the period from January 1, 2012 and January 1, 2013 met criteria for migraine according to the International Headache Society. All patients underwent an auto filling questionnaire, in order to evaluate the presence of vestibular symptoms. Those patients who reported vestibular symptoms were referred to neuro-otological section for a specialized examination.

Results: 400 patients had a diagnosis of migraine without aura and 60 migraine with aura. 108 patients (92:16; MSA:MCA) reported the presence of positive vestibular symptoms, the diagnoses were: migraine-vertigo (43), benign paroxysmal positional vertigo (30), Basilar-type migraine (12), acute vestibular hypofunction (10), central positional vertigo (6), Ménière syndrome (5) and acoustic neurinoma (2).

Conclusions: We confirmed a high prevalence of vestibular disorders in our migraine population (23.5%). Vestibular disorders occur more frequently in patients with migraine without aura. 55% of patients with vestibular pathology showed a clear association with migraine etiopathogenia; in 34% the relationship between the two disorders was likely/probably and 10% of the cases were attributable to merely statistical chance.

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Abstract – WCN 2013**No: 927****Topic: 36 – Other Topic****Paediatric narcolepsy – A seven-year experience**S. Aurangzeb, Z. Zaiwalla. *Clinical Neurophysiology, John Radcliffe Hospital, Oxford University Hospitals Trust, Oxford, UK*

Prevalence of narcolepsy varies from 0.05 to 0.02% in adults and although narcolepsy is considered to be a disease of adulthood, most cases have their onset in childhood. Diagnosis is frequently delayed or misdiagnosed because of unfamiliarity with paediatric narcolepsy and variable presentations.

Objective: To describe the clinical and polysomnographic features of children with narcolepsy.

Patients and methods: 38 children diagnosed with narcolepsy at John Radcliffe Hospital, Oxford, between January 2005 and October 2012, are included in this study.

Results: The mean age of onset was 9.61 ± 3.51 years. 12 children had a predisposing insult and two of them reported swine flu vaccination prior to symptom onset. Full tetrad of narcolepsy symptoms was seen in only 18.4% while 52.6% children presented with three out of four cardinal symptoms. Excessive daytime sleepiness was the most common symptom (100%), followed by cataplexy (86.84%), hypnagogic hallucinations (60.5%) and sleep paralysis (28.9%). Change in the facial expression was noticed by 15.62% parents while the characteristic “cataplectic facies” were seen in 36.8% children in the clinic. Disturbed night sleep was reported in 89.5% children, excessive dreaming in 71.1% and parasomnias in 68.4% children. The mean sleep onset latency on multiple sleep

latency test (MSLT) was 2.95 ± 2.98 min and mean sleep-onset rapid eye movement (SOREM) was 3.

Conclusion: Narcolepsy must be considered in the differential diagnosis of hypersomnia, even in the absence of classic signs and clinicians must be vigilant towards ancillary features. MSLT, following a strict protocol, is diagnostic in most children.

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Abstract – WCN 2013**No: 2688****Topic: 36 – Other Topic****IFCN guidelines for cervical vestibular evoked myogenic potentials**E.S. Papathanasiou^a, T. Murofushi^b, F. Akin^c, J.G. Colebatch^{d,e}. ^a*Clinical Sciences, Cyprus Institute of Neurology & Genetics, Nicosia, Cyprus;* ^b*Department of Otolaryngology, Teikyo University School of Medicine, Kawasaki, Japan;* ^c*Audiology, VA Medical Center, Mountain Home, TN, USA;* ^d*UNSW Clinic School, Sydney, NSW, Australia;* ^e*Prince of Wales Medical Research Institute, Sydney, NSW, Australia*

Background: Cervical vestibular evoked myogenic potentials (cVEMP) are electromyograms evoked by high-level acoustic stimuli recorded from the tonically contracted sternocleidomastoid (SCM) muscle, and have been accepted as a measure of saccular and inferior vestibular nerve function. As more laboratories are publishing cVEMP data, there is a wider range of recording methods and interpretation, which may be confusing and limit comparisons across laboratories.

Objective: To recommend *minimum requirements* or guidelines in the recording and interpretation of cVEMPs in the clinic and for diagnostic purposes.

Material and methods: We have avoided proposing a single methodology, as clinical use of cVEMPs is evolving and questions exist about its underlying physiology and its measurement. The development of guidelines by a panel of international experts may provide direction for accurate recording and interpretation.

Results: cVEMPs can be evoked using air-conducted (AC) sound or bone conducted (BC) vibration. The technical demands of galvanic stimulation have limited its application. For AC stimulation, the most effective frequencies are between 400 and 800 Hz below safe peak intensity levels (e.g., 140 dB pSPL). The high pass filter should be between 5 and 30 Hz, the low pass filter between 1000 and 3000 Hz, and the amplifier gain between 2500 and 5000. The number of sweeps averaged should be between 100 and 250 per run. Raw amplitude correction by the level of background SCM activity narrows the range of normal values. There are few publications in children with consistent results.

Conclusion: The present recommendations outline basic terminology and standard methods. Because research is ongoing, new methodologies may be included in future guidelines.

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Abstract – WCN 2013**No: 2695****Topic: 36 – Other Topic****Successful treatment of carotid-cavernous fistula through a percutaneous orbital approach**A. Garcia de la Fuente^a, R.A. Rangel-Guerra^b. ^a*Hospital and University Federation of Adult and Geriatric Psychiatry Christus Muguerza, Mexico;* ^b*Neurology, Hospital Universitario Jose Eleuterio Gonzalez, Nuevo Leon, Mexico*

Material and methods: 72 y/o white female during the last 2 months developed gradually scleral congestion, ocular pain, proptosis, chemosis and decreased visual acuity of the left eye without pupillary involvement, and without extraocular nerve paresis.

The rest of the neurological and neuro-ophthalmologic examinations were within normal limits. Carotid cavernous fistula on the left side was suspected. Digital subtraction carotid angiography was performed which revealed a carotid cavernous fistula (type “C” of the Barrow classification). The fistula was fed by the meningohypophyseal trunk (Fig. 1).

Endovascular approach was tried though the meningohypophyseal trunk was unsuccessful.

Then, venous approach was intended to be done through the inferior petrosal sinus, but the cavernous sinus was thrombosed.

It was decided to perform a percutaneous transorbital approach with catheterization of the cavernous drainage vein and a glue injection occluded the fistula without injection occluded the fistula without complications.

Conclusion: Percutaneous orbital approach is a useful tool in the management of carotid cavernous fistula, when neither arterial nor venous access was possible.

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Abstract – WCN 2013

No: 2698

Topic: 36 – Other Topic

The “weekend effect” – Observations from a neurology inpatient service

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Background: Patients admitted to hospitals on weekends are at increased risk of adverse outcomes, including mortality. Whether this specifically holds true for neurological patients is unclear.

Objective: We undertook this study to investigate whether the so-called “weekend effect” extended to a neurology service providing care in a developing country.

Patients and methods: Mortalities occurring over a ten-year period (January 2000 through December 2009) on an inpatient neurology service at a tertiary-level hospital in Karachi, Pakistan, were reviewed. Deaths were categorized by primary diagnosis, day and month of admission, and day and month of death. Mortality rates were compared using the chi-square test.

Results: Of 9966 admissions during the study period, 601 (6 · 03%) patients died before discharge. Mortality rate for patients admitted on weekends (165 deaths from 2382 admissions, or 6 · 93%) was significantly higher than for patients admitted on weekdays (436 deaths from 7584 admissions, or 5 · 75%; $p = 0 · 035$ for weekend versus weekday comparison, reflecting an excess of 1 · 18 deaths per 100 admissions for patients admitted on weekends). When analyzed by the three leading primary diagnoses, this weekend effect was present in patients with stroke, although not in those with epilepsy or CNS infection.

Conclusions: Neurological inpatients admitted on weekends are at greater risk of dying in the hospital than those admitted during the rest of the week. These observations raise important questions about the appropriateness of work schedules, staffing patterns and, ultimately, the quality of care delivered in hospitals on weekends.

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Abstract – WCN 2013

No: 2677

Topic: 36 – Other Topic

Ethical quality of clinical trials in Lebanon: Result of a national survey

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Medicine shall exceed mere knowledge therefore every specialty shall remain a bridge to real medicine. Neurology cannot be applied without considering the context and its impact on the practice.

Objectives: This study is considered the first to analyze the situation in Lebanon concerning the ethical frame of therapeutic trials in order to compare the two largest groups of industrialized and developing countries. The national survey was held in 144 hospitals working in the Lebanese environment characterized by a plurality of communities.

Method: Three methodological tools were used: a questionnaire (IRB, pharmaceutical companies, patients), direct and semi-direct interviews. The analysis about the ethical quality of IRB works legitimacy was developed according to Berdeu chart.

Results and discussion: There is heterogeneity in the practices of essential definitions regarding biomedical research and its limitations. IRB regulations are rather permissive to researchers and industrial promoters. The future of research and the protection of concerned participants are at stake. The scientific expertise within IRBs, and the criteria applied in determining ethical opinions are major concerns. The absence of a pharmacovigilance system or of a competent authority in health safety increases the seriousness of the current situation.

Conclusion: The field is now open to industrial promoters, and the right goal of clinical trials is under questioning. The country has to decide to which extent it will accept and support clinical and scientific investigations implying citizens' participation. This mission is essential and necessary to avoid transforming research into oppression and manipulation of the human being.

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Abstract – WCN 2013

No: 1993

Topic: 36 – Other Topic

Valsalva maneuver shows prolonged sympathetic outflow in patients with a history of mild traumatic brain injury

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Background: Increased mortality rates persist years after mild traumatic brain injury (mTBI) and might result from central autonomic dysregulation (Hilz et al., J Neurotrauma 2011;28:1727–38).

Objective: To determine whether Valsalva maneuver (VM) refines autonomic pathophysiology in post-mTBI-patients.

Methods: In 25 patients (GCS 13–15; 34.0 ± 13.2 years), tested 4–98 months after mTBI and 29 controls (31.3 ± 12.2 years), we monitored respiratory frequency (RESP), RR-intervals (RRI) and systolic blood pressure (BP) at baseline and during three VMs (15 s, 40 mm Hg strain). At baseline, we assessed total autonomic modulation as RRI-coefficient of variation (CV), RRI-standard-deviation (SD), RRI-total-powers, sympathetic modulation as RRI-low-frequency-powers (LF),

parasympathetic modulation as RRI-RMSSDs and RRI-high-frequency-powers (HF), and baroreflex sensitivity (BRS).

We calculated Valsalva-ratios (VR) and – as indices of sympathetic withdrawal – intervals from the highest BP-value during phase-IV-overshoot to the time when BP had decreased by 60% (60%-BP-recovery-time) and 90% (90%-BP-recovery-time) of the differences between phase-IV-BP-maxima and baseline-BP. We compared patient- and control-parameters before and during VMs (ANOVA, post-hoc analysis; significance: $p < 0.05$).

Results: At baseline, RRI-CVs, RRI-SDs, RRI-total-powers, RRI-LF-powers, RRI-RMSSDs, RRI-HF-powers and BRS were lower in patients than controls. During VMs, RRI, BPs, and VRs were similar in both groups, while 60%- and 90%-BP-recovery-times were significantly longer in patients than controls.

Conclusions: Baseline autonomic modulation was reduced in mTBI-patients. During VM-challenge, only prolonged 60%- and 90%-BP-recovery-times unveiled altered autonomic adjustment in mTBI-patients with prolonged sympathetic hyperactivity. Sympathetic hyperactivity upon challenge may contribute to increased cardiovascular risk years after mTBI.

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Abstract – WCN 2013

No: 2673

Topic: 36 – Other Topic

Genetic modifications present in choroid plexus neoplasias and associated disturbances of psychoneural development

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Choroid plexus papillomas or carcinomas are rare forms of brain neoplasias. Some years ago, Steichen-Gersdorf et al. found a choroid plexus papilloma associated to a case of hypomelanosis of Ito and a 17p: Xq translocation in this female patient (Hum Genet 1993; 90:611–613). Recently, a 10-month-old girl with a Brachmann–Cornelia de Lange syndrome (BCLS) and a choroid plexus papilloma of the third ventricle of the brain was operated upon and studied at Hospital Infantil de México Federico Gómez (HIMFG) in Mexico City. Karyotypes (GTG bands) of the proband and her parents undertaken at HIMFG were normal. Array CGH analyses of blood DNA of these 3 persons carried out at BlueGnome's Laboratory in Cambridge, U.K., set in evidence amplification of genes SPNS2, GGT6, SMTNL2, PELP1, MYBBP1A and ALOX15 in chromosome 17p of the proband. Since MYBBP1A is a proto-oncogene and ALOX15 participates in development of cancer and metastases of tumors, further FISH analyses of these 2 genes were implemented at HIMFG. Amplification of both genes was also found in the tumor of the case under study but not in an unrelated papilloma of the choroid plexus. The possible genetic counseling, diagnostic and prognostic importance of the findings, especially regarding genetic modifications of chromosome 17p in choroid plexus tumors and associated clinical disturbances of psychoneural development of the type of hypomelanosis of Ito or BCLS, is discussed.

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Abstract – WCN 2013

No: 1160

Topic: 36 – Other Topic

BAFF controls neural cell survival through BAFF receptor

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Background: B cell activating factor (BAFF) is a fundamental survival factor for B lymphocytes and its effect on B cells has been extensively investigated. However, its effect on the central nervous system (CNS) has not been clarified.

Objective: To explore the novel mechanism of neuronal survival by BAFF receptor (BAFF-R) signals in CNS.

Material and methods: We examined BAFF and BAFF-R expression in neurons and cultured embryonic neurons of either wild-type or A/WySnj mice (*Baffr^{m/m}*), which carry a mutation in BAFF-R, to compare the survival of neurons of both groups in vitro. We also bred mice carrying the human Superoxide Dismutase-1 mutation G93A (*mSOD1*), animal models of familial amyotrophic lateral sclerosis (ALS) with *Baffr^{m/m}*, and body weight and survival were assessed.

Results: Immunohistochemical analysis and RT-PCR study revealed that both BAFF and BAFF-R were expressed in mouse neurons. Primary cultured neurons of *Baffr^{m/m}* showed decreased survival compared with those of wild-type mice (*Baffr^{+/+}*). Furthermore, *mSOD1/Baffr^{m/m}* displayed reduced survival (141.56 days \pm 2.25) vs control mice (*mSOD1/Baffr^{+/+}*) (152.3 days \pm 1.28); $p < 0.001$. *mSOD1/Baffr^{+/+}* at age 18 weeks displayed reduced body weight (90.80% \pm 0.83 [n = 20]), compared with *mSOD1/Baffr^{+/+}* (97.18% \pm 0.61 [n = 19]); $p < 0.001$.

Comparable numbers of microglia/macrophages and astrocytes were observed at early- and middle-stage disease in spinal cord section of both groups.

Conclusion: Our findings suggest a novel role of BAFF as a critical member of neuroprotective factors that directs neuronal survival independent of the action for glial cells.

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Abstract – WCN 2013

No: 2656

Topic: 36 – Other Topic

Severe cardiac dysautonomia in acute myelitis

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Background: Traumatic spinal cord injury is frequently complicated by cardiocirculatory impairment (bradycardia and hypotension). It is unknown whether the same phenomenon can manifest in acute myelitis.

Design and methods: Case report

Results: A 35-year-old woman presented with left hemisoma weakness and hypoesthesia. Neurological examination revealed left arm weakness and dysmetria, left lateropulsion at Romberg test and hypoesthesia in the left hemisoma with a C2 upper level. Laboratory tests and CSF analysis were unremarkable. Brain MRI was negative. Cervical spine MRI showed a T2-hyperintense C2 intramedullary lesion. An acute myelitis was diagnosed and the patient started on a course of methylprednisolone, with initial benefit. Five days after

symptom onset, she developed severe bradycardia episodes, that required atropine and dopamine treatment. Electrocardiograms revealed sinus bradycardia and prolonged QT interval. Cardiac MIBG-scintigraphy showed low early and delayed H/M ratio. Patient's status improved during the following months and the neuroimaging follow up was negative for new lesions.

Conclusion: Patient's lesion involved the dorsal funiculi, and the left lateral and ventral funiculi. The dorsal aspect of the lateral funiculi represents the localization of the descending vasomotor pathways. The most likely explanation for patient's bradycardia is the myelitis-related disruption of descendent pathways from the brainstem cardiovascular center to sympathetic neurons into T1–T4 segment intermediolateral nuclei, resulting in sympathetic hypoactivity. Myocardial MIBG-scintigraphy confirmed this hypothesis, revealing a decreased cardiac MIBG uptake. This case extends the spectrum of the clinical presentations of cervical myelitis and emphasizes the importance of careful cardiac rhythm monitoring in the acute phase.

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Abstract – WCN 2013

No: 2658

Topic: 36 – Other Topic

Activation of GABA-A receptors in the medial prefrontal cortex produces anxiolytic-like response

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Objectives: There has been increasing evidence that GABAergic system is involved in neurobiology of anxiety. The present study aims to investigate the role of GABAergic systems in the modulation of anxiety in medial prefrontal cortex (MPFC) of rats, using the elevated plus maze test.

Methods: Rats were anaesthetized with a mixture of ketamine and xylazine, and then special cannulas were inserted stereotaxically into the MPFC. After 5–7 days of recovery, the effects of intra-MPFC administration of GABAergic agents were studied.

Results: Bilateral injection of the GABA-A receptor agonist muscimol (0.25, 0.5 and 1 µg/rat) produces an anxiolytic-like effect, shown by significant increases in the percentage of open arm time (%OAT) and percentage of open arm entries (%OAE). Intra-MPFC administration of the GABA-A receptor antagonist bicuculline (0.25, 0.5 and 1 µg/rat) produces significant anxiogenic-like behavior. However, intra-MPFC injection of GABA-B receptor agonist baclofen (0.05, 0.1 and 0.2 µg/rat), and GABA-B receptor antagonist CGP35348 (5, 10 and 15 µg/rat) did not alter %OAT and %OAE significantly.

Conclusion: The results of the present study demonstrate that the GABAergic system of the MPFC modulates anxiety-related behaviours of rats via GABA-A receptors.

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Abstract – WCN 2013

No: 58

Topic: 36 – Other Topic

Cellular pathology and apoptosis in neoplastic myelopathy due to metastatic disease

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Solid neoplasms, whether primary or metastatic, may lead to compression of the spinal cord and development of a compressive myelopathy syndrome. Apoptotic processes of cell death are thought to contribute to cell death in chronic compressive myelopathy due to degenerative spondylosis but this has not previously been described in neoplastic compression. This study aimed to characterise the spatiotemporal distribution of apoptosis and pathophysiological changes in neoplastic compressive myelopathy using human post-mortem tissue from 7 cases with extramedullary or intramedullary tumour involvement. A loss of anterior horn cells and cystic necrosis was demonstrated, as well as the presence of APP immunopositive axonal swellings in all cases. Apoptosis was shown using a panel of immunohistochemical markers including TUNEL, DNA-PKcs, PARP and AIF seen at, above and below the compression. Apoptosis was maximal at the site of tumour compression. We conclude that axonal damage and neuronal loss, as well as oligodendroglial apoptosis may be important processes in the pathophysiology of neoplastic compressive myelopathy.

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Abstract – WCN 2013

No: 1008

Topic: 36 – Other Topic

Change in brain neural activation during stress due to a continued simple cognitive task

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Background and objective: Repetition of simple cognitive tasks causes stress. We report change in brain activity during such tasks.

Subjects and methods: Some 13 healthy subjects (mean age: 22.8 yrs) gave informed consent. Tokyo Metropolitan University Ethics Committee approved the study. Brain activity at the start and finish of cognitive tasks 3.0 T fMRI recorded activity in three 3-minute sessions of consecutive subtraction of 7, starting from 1000. Initial 36- and final 36-second stages of each session were compared. fMRI data was analyzed using SPM8 (uncorrected height threshold $p < 0.001$) and one-sample t-test second-order random effect model was performed to determine regions of significant change.

Results: Initially activity was prominent in the bilateral superior and inferior parietal lobules (SPL, IPL), left Brodmann area 44 (BA 44), left cerebellum, bilateral prefrontal cortex, and left supplementary motor cortex; finally in the left SPL and IPL, bilateral cerebellum, bilateral thalamus, cingulate gyrus, and insula (claustrum). Activity level in thalamus and cerebellum was higher in the final stage.

Conclusion: Initial activation in the parietal lobule and BA44, particularly significant in the region from the left parietal to frontal lobes suggests involvement of these sites in arithmetic concepts. Since temporary memory of numbers was necessary; sites related to working memory were also activated. Due to activation in the cingulate gyrus and insula (claustrum) over long-term, subjects felt stress resulting in prominent activation of sites unrelated to calculation. Repetition of simple tasks over time changes the sites of activation.

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Abstract – WCN 2013**No: 2097****Topic: 36 – Other Topic****Three new mutations in *CSF1R* gene in 3 Italian patients with hereditary diffuse leukoencephalopathy with axonal spheroids**

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Background: Hereditary diffuse leukoencephalopathy with axonal spheroids (HDLS), first described in 1984, is an autosomal dominant disorder characterized by white matter neurodegeneration, progressive cognitive decline and motor symptoms, and histologically, by axonal swellings (“spheroids”) and loss of axons and myelin. To date, 18 mutations have been identified in the colony stimulation factor 1 receptor (*CSF1R*) gene, located in exons 12–22 and affecting the tyrosine kinase domain of the protein. HDLS patients have been often misdiagnosed with CADASIL, CARASIL, fronto-temporal dementia, and multiple sclerosis.

Objective: We described the first 3 Italian patients affected by HDLS, showing three new mutations and expanding the phenotypic presentation of the disease.

Patients and methods: Out of 15 undiagnosed patients with cognitive decline and leukoencephalopathy, three (2 ♂, 1 ♀, age range of 37–58 years) showed *CSF1R* gene mutations. The clinical course is characterized by progressive cognitive and behavioural declines, preceded in all of them by a brain ischemic stroke-like.

Results: The genetic analysis of *CSF1R* gene identified 3 new heterozygous missense mutations located at the PTK domain (case 1: exon 19, c.2527A>T, p.Ile843Phe; case 2: exon 14, c.1957T>C, p.Cys653Arg; case 3: exon 21, c.2717C>T, p.Ile906Thr).

Conclusion: In all our cases, the onset of the diseases was characterized by acute neurological onset, mimicking a stroke, followed by severe neurocognitive decline. Our data suggest that the frequency of HDLS may be underdiagnosed and that *CSF1R* analysis must be performed in all unsolved cases of leukoencephalopathy and progressive dementia.

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Abstract – WCN 2013**No: 2546****Topic: 36 – Other Topic****Nerve conduction abnormalities in Gullain Barre syndrome in Nairobi**

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Background: Gullain Barre syndrome is an autoimmune polyradiculopathy. The major types are AIDP AMAN and AMSAN and MFS. No neurophysiological studies of GBS have been presented from this country.

Objective: To determine the neurophysiological types of GBS in Nairobi.

Materials and methods: We reviewed the neurophysiological reports of 36 cases of GBS who presented to our lab. The nerve conduction studies were performed on Nihon Kohden neuropack S1. The mean age was 40 + 7 years and range of 11 to 74 years. There were 20 males and 16 females. We used the distal latencies, conduction block, amplitude f and H response to determine the physiologic type. The types of neurophysiology abnormalities were AIDP 30 (83%), AMSAN 2 (5.5%) and AMAN 4 (11%).

Conclusion: AIDP is still the commonest form of GBS in our setting with a few cases of AMAN and AMSAN. This is in keeping with results found in the developing world.

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Abstract – WCN 2013**No: 2224****Topic: 36 – Other Topic****Vestibular–somatosensory interaction; A near-infrared spectroscopy study**

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Background: From animal experiments, it is known that a cortical area in which a vestibular afferent pathway is projected is diffusely present from the parietal lobe to the temporal lobe.

Objective: The aim of the present study with a near-infrared spectroscopy (NIRS) is to evaluate the correlation of the vestibulo-ocular reflex (VOR) and the somatosensory input.

Methods: Ten healthy, young adults with no known neuro-otological disorders participated in this study. A NIRS system holder was placed bilaterally over the parieto-temporal region of the head. Caloric stimulation was induced by irrigating the left external ear canal of a study participant, while the subjects lay supine with eyes closed. During the occurrence of nystagmus, active-somatosensory stimulation was achieved when the subjects pushed against a footboard with the soles of their feet. The oxygenated cerebral hemoglobin changes before and after the somatosensory stimulation was calculated using NIRS.

Result: During active-somatosensory stimulation, the amount of oxygenated hemoglobin increased in the parieto-temporal (parieto-insular vestibular cortex?) lobe bilaterally by the active-somatosensory stimulation.

Conclusions: Omnipause neurons (OPNs) are thought to control a number of oculomotor behaviors, especially in rapid-phase VOR. OPN inhibition originates from the superior colliculus (SC). The active-somatosensory input may enhance the activity of SC, OPN, and inhibit the RPEV via the central vestibular cortex. The present study suggests that NIRS is useful for confirming the existence of neural linkage between the peripheral vestibular organ and somatosensory input.

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Abstract – WCN 2013**No: 2577****Topic: 36 – Other Topic****Modified embryo organ culture model system for preclinical studies of growth factor-induced neural tissue differentiation in mammals**

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Aims: Fibroblast growth factor and nerve growth factor are found in various parts of human and vertebrate embryos during development. The development of the vertebrate nervous system begins with the inductive differentiation of the neural plate from the dorsal side of flat ectoderm at the end of gastrulation. The role of FGF and NGF in critical period for mammalian development, gastrulation, was studied.

Material and methods: In our laboratory a unique organ culture system of mammalian embryo was designed. Embryonic parts of the whole gastrulating rat embryos were cultivated on the stainless-steel grid supported lens paper, at the air–liquid interface during two weeks using liquid serum-free and protein-free medium. A signal-protein FGF or NGF (100 ng/ml and 200 ng/ml) were added, or both in a culture medium in a time frame of 2, 5 or 9 days.

Results: Embryos developed into teratoma built of intermixed tissues, including nerve tissue. In control serum-free medium neuroblasts were absent. NGF did never improve differentiation of neuroblasts. FGF significantly stimulated the differentiation of neural tissue during 5 days, and the best during 9 days. In FGF/NGF-treated

embryo differentiation of neuroblasts was the same as was in FGF-treated.

Conclusions: Our model system is simpler than the embryo *in vivo*, but closer to normal development than the cell cultures used in other studies. The neuroblasts found were direct answer to molecular signals added. This fact is in accordance with the idea that in a multi-level process of neural differentiation, FGF is the neural inducer in mammals.

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Abstract – WCN 2013

No: 2576

Topic: 36 – Other Topic

Spinal schistosomiasis: Relevance of epidemiologic and imaging features in an unusual case of myelopathy

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Background: Neuroschistosomiasis is a common cause of myelopathy in endemic regions but it is infrequent in developed countries.

Objective: Our goal is to present a case of spinal schistosomiasis where epidemiologic and imaging features played an important role.

Patients and methods: Clinical case.

Results: A 26-year-old woman from Guinea-Conakry was referred to a Portuguese Neurosurgery Department for progressive paraparesis and urinary retention. Spinal MRI showed an intramedullary lesion causing enlargement of the spinal cord from D9–10 to L1–L2. The lesion was hyperintense on T2-weighted images and showed a heterogeneous pattern of contrast enhancement. The patient improved to some extent without treatment and two weeks later she was transferred to our Neurology Department. Control MRI revealed slight decrease of the spinal enlargement and CSF analysis showed lymphocytic pleocytosis and elevated protein. Brain MRI, somatosensory evoked potentials of the lower limbs, and blood analysis including cyanocobalamin levels and autoimmunity were normal, and infectious serologies were negative except for schistosomiasis serology which came back positive. A diagnosis of spinal schistosomiasis was established based on epidemiologic and imaging features, supported by a positive schistosomiasis serology and exclusion of other causes. Treatment consisted of an anthelmintic and corticotherapy according to guidelines, as well as physical rehabilitation. On reevaluation 7 months later there was clinical improvement and imagiological resolution.

Conclusion: Spinal schistosomiasis is rarely encountered in developed countries, making diagnosis challenging. Epidemiologic and imaging features are essential for diagnosis, leading to prompt treatment and improved prognosis.

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Abstract – WCN 2013

No: 2589

Topic: 36 – Other Topic

Behavior and reproduction of mothers as well as behavior of the second generation pups are influenced by maternal separation

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Background: Daily separation of rat pups from their mothers while in the preweaning period has an impact on behavior and stress response of pups.

Objective: We hypothesized that maternal separation may create a depression like state in mother rats from which we separate the rat pups and decrease their reproductive function and fertility. Also, we aimed to evaluate the transmission degree of depression and anxiety across generations.

Material and methods: Females aged 3 months were divided in two groups of 10 rats each. After a first mating and a first parturition, we conducted maternal separation 3 h during 22 days for the experienced group while the control group mothers kept their pups. At 4 months of age, females underwent behavioral tests and a second mating. Second generation pups were also subjected to behavioral tests.

Results: Behavioral tests shown that mothers who experienced separation were more depressive and anxious than control ones, also they have a lower litter size. Rat pups of the second generation whose mothers' experienced maternal separation also revealed behavior changes akin to depression and anxiety but less noticeable than the first generation pups.

Conclusion: Thus, maternal separation causes depressive and anxious like-states on mother rats which experienced separation and has an impact on their litter size. Also, consequences of maternal separation seem to last throughout generations.

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Abstract – WCN 2013

No: 2591

Topic: 36 – Other Topic

Work stress in health care workers in Hospital El Idrissi

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Background: Health care workers are strongly exposed to work stress, their conditions of work and multiple other factors increase their susceptibility to this adjustment disorder. Our work is an epidemiological study designed to assess the state of total health, to identify risk factors of stress, its consequences among hospital staff in of the region of Gharb Cherarda Beni Hsen (Morocco).

Methods: This study is a survey based on a questionnaire covering the Total Health Test (TST) and the Global Stress Index (GSI), distributed to doctors and nurses of the CHR and collected for statistical analysis.

Results: Our results have shown the involvement of socio-demographic factors relating to professional careers in the initiation and maintenance of stress. Our study showed the dissatisfaction of health workers, particularly females, towards working conditions. Moreover, a majority of respondents, regardless of their profile, have high TST scores reflecting an alarming exposure to stressful factors.

Conclusion: Aware of the scarcity of works dealing with stress of health care staff in morocco and negative impact that this stress has in terms of morbidity and mortality, our work aims to improve the quality of hospital work in the Gharb region and this by understanding and reducing risk factor of this disease.

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Abstract – WCN 2013

No: 2193

Topic: 36 – Other Topic

Clinical features of aseptic meningitis

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Background: Aseptic meningitis is a self-limited common disease. But there are few reports about the clinical course, the laboratory

findings, and about the frequency of meningeal irritation sign. We studied the actual clinical features of aseptic meningitis.

Patients and methods: We retrospectively analyzed the clinical features of the patients with aseptic meningitis who admitted to our hospital from January 2003 to December 2012. Evaluated points were the mean length of hospital stay and that of febrile state, meningeal irritation sign (jolt accentuation of headache, neck stiffness and Kernig's sign), laboratory findings (white blood cells, C-reactive protein, etc.), and cerebrospinal fluid examination (cells, protein, sugar). We compared these points to those of control patients.

Results: 251 patients (134 males, mean age 34.7 years) and 221 control patients (115 males, mean age 35.8 years) were enrolled. The mean length of hospital stay was 13.8 days, and that of febrile state was 6.8 days. Jolt accentuation of headache was present 82% vs. 60% (the patients vs. the controls). Neck stiffness was 65% vs. 40%. Kernig's sign was 31% vs. 21%. Sensitivity of jolt accentuation of headache was 0.82, specificity 0.31. Neck stiffness was 0.65/0.60 and Kernig's sign was 0.31/0.79 retrospectively. Aseptic meningitis patients had lower white blood cell count (7209 ± 2568 vs. $9479 \pm 4118/\text{mm}^3$, $P < 0.01$), C-reactive protein (1.05 ± 2.12 vs. 3.91 ± 4.96 mg/dL, $p < 0.01$).

Conclusion: Jolt accentuation of headache is not useful for diagnosis of aseptic meningitis because of its low specificity. Almost normal white blood cell and C-reactive protein are common findings in aseptic meningitis.

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Abstract – WCN 2013

No: 2596

Topic: 36 – Other Topic

Association between pediatric non-REM parasomnias and obstructive sleep apnea

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Background: Parasomnias are neurological disorders characterized by undesirable physical events or experiences that occur during sleep. Obstructive sleep apnea (OSA) is increasingly recognized as an aggravating factor for parasomnias, however, the association between the two disorders has not been well elucidated among children.

Objective: To identify the association between non-REM parasomnia and OSA in cases of children especially focusing on the effect of OSA treatment.

Patients and methods: Two cases of non-REM parasomnia (9 and 11 years old) presented with sleepwalking, sleep terrors and snoring were involved. Polysomnography (PSG) was conducted to identify the characteristics of nocturnal events and the severity of OSA. Treatment of OSA was introduced with CPAP and the PSG parameters under CPAP were evaluated. Improvement of parasomnia events was found for at least three months.

Results: Nocturnal events were observed during slow wave sleep and severe OSA (apnea hypopnea index of more than 15) was also noted in both cases. Treatment with CPAP dramatically improved non-REM parasomnia in one case, however, nocturnal event did not decrease with CPAP in another case.

Conclusion: Presence of OSA could aggravate the severity of non-REM parasomnia in children. Importance of conducting the screening of OSA among children with non-REM parasomnia has been suggested.

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Abstract – WCN 2013

No: 2552

Topic: 36 – Other Topic

MR volumetry of amygdala and hippocampus in healthy controls

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Background: Amygdala and hippocampus play an important role in patients with Alzheimer disease, temporal lobe epilepsy or psychiatric disorders. Atrophy of these structures in comparison to volumes in healthy subjects has been described.

Objective: To measure amygdala and hippocampus volumes in healthy subjects by both manual and automatic segmentation and to determine variability of these structures in healthy population.

Patients and methods: 25 healthy right-handed volunteers (12 men, 13 women), median age 41 years, were enrolled. 3T-MRI T1-weighted acquisition with 1.3 mm slice thickness was performed. Left (L) and right (R) amygdala and hippocampus were outlined manually in coronal plane and automatically. Asymmetry index (AI) calculated as $2 * (L - R) * 100 / (L + R)$ and variation coefficient ($SD/MEAN * 100\%$) of normalized values were calculated.

Results:

Manual segmentation

Volume (cm^3)

Left hippocampus 2.70 ± 0.25 , Right hippocampus 2.78 ± 0.25 ,

Left amygdala 1.46 ± 0.25 , Right amygdala 1.52 ± 0.24 ,

Asymmetry index (%) hippocampus -2.9 , amygdala -4.1

Automatic segmentation

Volume (cm^3)

Left hippocampus 2.70 ± 0.25 , Right hippocampus 2.78 ± 0.25 ,

Left amygdala 1.46 ± 0.25 , Right amygdala 1.52 ± 0.24 ,

Asymmetry index (%) hippocampus -2.9 , amygdala 4.9

Hippocampal volumes showed similar slight right-left asymmetry both in manual and automatic segmentation, asymmetry of amygdala volumes was inconsistent and dependent on the method.

Use of automatic segmentation resulted in an increase of amygdala volumes.

Conclusions: There is relatively high variability in hippocampus and amygdala volumes of healthy subjects. Method of segmentation used, manual or automatic, could influence the results.

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Abstract – WCN 2013

No: 2568

Topic: 36 – Other Topic

Diffusion tensor imaging of the cerebellum-prefrontal area in ADHD

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Objectives: DTI studies have revealed developmental changes in cortical WM pathways in prefrontal regions and in pathways surrounding the basal ganglia and cerebellum in patients with ADHD, which presumably reflect decreasing myelination of axons. It is believed that these changes cause a decrease in the speed of neuronal communication. We hypothesized the presence of abnormal diffusivity in right prefrontal cortex and cerebellum brain regions assessed in vivo using diffusion tensor MRI.

Methods: Twenty three children (ages 7–12 yrs, 11 ADHD patients, 12 controls) were examined as follows: all subjects were scanned while in the supine position with thighs relaxed and parallel to the magnet magnetic field direction. Images were acquired on a 1.5 T imager Diffusion weighted gradients were applied along 15 non-collinear directions with a b-value = 800 s/mm². High-resolution images were acquired using 3DT1. Segmentation of the cerebellum CB was manually drawn on midline sagittal 3D-T1 images.

Results: Shows tract distribution in ADHD and healthy controls. We present preliminary results of white matter connectivity of tracts connected cerebellum-prefrontal area. There were no discernible ADHD-Control changes in ADC values along the connected white matter (Fig. 2) while generalized fractional anisotropy is increased ($p = 0.08$), as it was reported in recent literature [1]. Length of tracts vs ADC is shown in Fig. 3. In summary, as the study progresses, MD, I1, I2, and I3 will be used in order to further examine frontal-cerebellum tracts in patients with ADHD.

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Abstract – WCN 2013

No: 2571

Topic: 36 – Other Topic

Resting state in Latin American child with ADHD

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Objectives: ADHD is a neurological disorder in children. This illness is considered to be 80% originated by genetic factors. Here we compared infant Latin ADHD patients with healthy ones. We discuss the differences with previous studies that used different genetic pools.

Methods: 30 volunteers (8.4 ± 2 years, both sexes) were divided in two groups, healthy (H) and ADHA (AD). Volunteers laid in an MR scanner in silence while 150 brain volumes covering the whole of the brain were acquired. Resting state analysis was performed using DPARSF software. Low frequencies under 0.08 Hz were kept.

Results: Figure 1A and 1B present the results of a comparison between H and AD patients (H > AD in green and AD > H in red). H subjects presented strong left lateralization (80% vs. 20% structures). AD patients presented a stronger right lateralization (55% vs. 45%).

Conclusions: AD patients had a larger predominance of right hemisphere activations over left in contrast to healthy subjects. Previous work has reported strong involvement of the brain stem and the anterior cingulate gyrus for AD patients compared to H which we did not find. Never reported correlations with the frontal gyrus and the posterior cingulate cortex were found. Considering that similar analysis methods were followed as in previous studies, we believe that the differences shown arise by the different genetic origin of volunteers.

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Abstract – WCN 2013

No: 2455

Topic: 36 – Other Topic

Pelizaeus Merzbacher like disease in Tunisian children: A severe phenotype

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Background: Pelizaeus Merzbacher like disease (PMDL) is an autosomal recessive disorder related to GJC2 gene mutation. It resembles Pelizaeus Merzbacher disease (PMD) but with more preserved cognitive and motor functions.

Objective: To review clinical, radiological and neurophysiological characteristics in 4 Tunisian patients with PMDL.

Patients and methods: Four children belonging to 2 unrelated Tunisian families with PMDL were followed in our department. Medical history, physical examination, cerebral MRI and neurophysiological studies were analyzed.

Results: Mean age of patients was 7.2 years. Three patients presented with a psychomotor delay and one with psychomotor regression at 18 months. Examination showed nystagmus, severe mental retardation, axial hypotonia, cerebellar ataxia, and spastic tetraparesis in all patients. Clinical severity score was classified as form 0 in 3 out of 4, based on the best motor function. Diffuse cerebral hypomyelination was noticed on brain MRI in all patients. Nerve conduction velocities were normal. Brainstem auditory evoked potentials performed in 1 patient revealed increased I–V truncal conduction time. Direct sequencing of the GJC2 gene confirmed the diagnosis of PMDL.

Conclusion: PMDL is characterized by a less severe phenotype than PMD. Severity score ranged between 3 and 4 in most reported cases, unlike most of our patients who had severe clinical phenotype. Besides, radiological and neurophysiological findings in our patients were similar to those in previously reported cases. These observations highlight the expanding clinical spectrum and probably a severe phenotype of PMDL in the Tunisian population.

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Abstract – WCN 2013

No: 2562

Topic: 36 – Other Topic

Clinical features of myelopathies: A cohort study in Tunisia

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Introduction: Myelopathies represent a heterogeneous group of disorders with distinct etiologies, clinical and radiologic features, and prognoses, characterized by spinal cord dysfunction resulting in paresis, sensory level, and autonomic (bladder, bowel, and sexual) impairment.

Objective: The aim of this study was to determine the clinical characteristics and the management of patients with myelopathy.

Patients and methods: We conducted a retrospective study using population-based information from medical records of 100 patients with myelopathy, hospitalized in the neurology department at the National Institute of Neurology in Tunis over a period of 12 years.

Results: The mean age of patients was 38 years (9–76 years) with a sex ratio of 1.43. Most patients had a severe motor deficit. CSF studies revealed hyperproteinorachia in most patients and were normal in 31 cases. MRI showed a spinal hypersignal in almost all cases and was normal in 9 patients. Etiologies were dominated by infectious and dysimmune disorders. Intravenous methylprednisolone (1 gr/day for 3 to 5 days) was indicated for 55 patients, and an intravenous immunoglobulin cure was used in two cases. Rapid clinical improvement was observed in 33.3% of patients with full recovery in 4 patients.

Conclusion: In non-compressive myelopathies, an extensive work up is needed in order to distinguish between demyelinating, infectious, other inflammatory, vascular, neoplastic, and paraneoplastic etiologies. An algorithm for the diagnosis and management of myelopathies is necessary.

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Abstract – WCN 2013**No: 1880****Topic: 36 – Other Topic****Prediction of trochlear nerve absence by superior oblique muscle volumetry in congenital superior oblique palsy**

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Purpose: To determine the predictability of superior oblique muscle volumetry in diagnosing trochlear nerve absence in patients with congenital superior oblique palsy (SOP).

Methods: Retrospective study of congenital SOP patients with (present group, n = 38) and without a trochlear nerve (absent group, n = 87) and controls (n = 34) using high-resolution magnetic resonance imaging (MRI) to analyze superior oblique muscle volume (SOV) and trochlear nerve absence.

Results: The paretic side SOV and normal side SOV were not significantly different in controls (p = 0.536) and in the present group (p = 0.750). However, in congenital SOP patients of the absent group, the paretic side SOV was significantly smaller compared to the normal side (p < 0.001). The cut-off value of paretic/normal side SOV ratio for diagnosing trochlear nerve absence was ≤0.75 (sensitivity 100%, specificity 100%).

Conclusion: In congenital SOP patients, the paretic/normal side SOV ratio has an excellent predictability in diagnosing trochlear nerve absence.

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Abstract – WCN 2013**No: 2007****Topic: 36 – Other Topic****Relationship between the trochlear nerve diameter and superior oblique muscle volume**

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Purpose: To determine the relationship between the trochlear nerve diameter (TD) and superior oblique muscle volume (SOV) in patients with congenital superior oblique palsy (SOP).

Methods: Retrospective study of congenital SOP patients with (present group, n = 38) and without a trochlear nerve (absent group, n = 87) and controls (n = 34) using high-resolution magnetic resonance imaging (MRI).

Results: The non-paretic side TD (npTD) of the absent group was smaller (p = 0.04) and npTD of the present group was larger than the TD of controls (p = 0.047). SOV positively correlated with the TD of controls (p = 0.029), npTD of the present group (p = 0.02) and npTD of the absent group (p = 0.008).

Conclusion: The npTD was smaller in the absent group and larger in the present group compared with controls. The TD of controls and npTD of congenital SOP patients positively correlated with the SOV.

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Abstract – WCN 2013**No: 2624****Topic: 36 – Other Topic****The relationship between optical coherence tomographic and perimetric findings in patients with papilledema**

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Background: Optical coherence tomography (OCT) provides quantitative ocular imaging in conditions causing optic neuropathy.

Objective: We compared measures of retinal and optic nerve head (ONH) anatomy to standard perimetry in patients with papilledema associated with intracranial hypertension.

Patients: We studied 69 eyes in 26 patients with papilledema (85% idiopathic intracranial hypertension) and no alternative ocular pathology. 14 patients were tested twice, at least 3 months apart.

Methods: Using spectral-domain OCT, we quantified retinal nerve fiber layer (RNFL) thickness, total macular volume and ONH volume in each eye and compared values with standard automated static perimetry.

Results: For most eyes, there was no relationship between the degree of RNFL or ONH swelling and perimetric mean deviation. In this group, even eyes with severe swelling showed normal or only mildly reduced mean deviations and RNFL thickness demonstrated a positive linear correlation with ONH volume. In a smaller group of eyes with ONH volumes at the lower end of the supranormal range, however, RNFL thickness and macular volume were paradoxically subnormal and perimetry showed moderate to severe impairment. In 4 eyes of this latter group, serial testing showed stable or improved visual fields despite declining RNFL thickness within the subnormal range.

Conclusions: Quantitative measures of tissue swelling do not correlate with visual field loss in most patients with papilledema. In a minority of eyes, RNFL and macular thickness may be reduced despite optic disk swelling, implying a combination of swollen and damaged axons. Further RNFL thinning may not portend visual field deterioration.

doi:10.1016/j.jns.2013.07.2401

Abstract – WCN 2013**No: 2619****Topic: 36 – Other Topic****Biomarker development on alcohol addiction using EEG**

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Alcohol addiction is harmful to society, economy and personal health. Alcohol addiction treatments intend to help addicted individuals reduce and stop compulsive alcohol use. Using biomarker, the clinicians could determine if drugs are having a desirable effect much earlier and given in correct dose for alcohol addiction treatment. This paper focuses on the usage of electroencephalography (EEG) to develop biomarker for alcohol addiction treatment including EEG methodologies and their applications.

doi:10.1016/j.jns.2013.07.2402

Abstract – WCN 2013**No: 2629****Topic: 36 – Other Topic****Chronic Ritalin administration during adulthood increases serotonin pool in rat medial frontal cortex**

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Background: Ritalin is a type II medication with a high tendency to be abused. It has been the main indication to control ADHD and/or treatment of major depression. As the college students may seek for it to improve memory, decrease the need for sleep and improvement the sense of well being (especially during exams), which all at least partially, related to serotonergic system activity, it seems worthy to evaluate the effect of Ritalin intake on a mature, brain. Although there are many studies about the effects of Ritalin on developing brain, only few studies on adults are available. The aim of present study is to find out if Ritalin can affect the SERT (serotonin transporter) density in mature medial frontal cortex or not.

Methods: Thirty male Wistar rats were enrolled in the study. Rats were assigned into five groups; control, two Ritalin and two vehicle groups. Twelve rats received Ritalin 20 mg/kg/BD orally for eleven days. After one week of withdrawal and another two weeks rest to evaluate short-term effects of Ritalin, six rats were sacrificed. Another six were studied to detect the long-term effects of Ritalin, so, 12 weeks after the previous group, were sacrificed. The immunohistochemistry (IHC) was performed to compare the results.

Results: Immunohistochemistry studies showed a higher density of SERT in both 2 weeks and 12 weeks after withdrawal from Ritalin intake in medial frontal cortex of rat.

Conclusions: Our findings demonstrated both short and long-term effects of Ritalin on frontal serotonergic system after withdrawal period.

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Abstract – WCN 2013

No: 2595

Topic: 36 – Other Topic

Funicular myelosis with complete favorable evolution. Case report

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Background: Vitamin B12 deficiency may present neuropsychiatric disorders. The most common is the subacute combined degeneration of the spinal cord, affecting mainly the lateral and posterior columns of the spinal cord.

Objective: Report a case of funicular myelosis by vitamin B12 deficiency in a male patient.

Material and methods: Laboratory exams were collected, such as dosage of vitamin B12, folic acid, HIV and cytomegalovirus serology, among others. Additionally image exams were done, as MRI with contrast of cervical and thoracic spine during the diagnosis and after treatment.

Results: The patient's age was below the incidence peak of the disease, with progressive neurological deficits in six months. Surgical etiology, infectious or chronic diseases were discarded. He presented typical involvement of posterior and lateral cords of the spinal cord associated with Babinski, Romberg and Lhermitte signs.

Tests showed no anemia (probably as result of previous treatments), macrocytosis, vitamin B12 deficit and seropositive for cytomegalovirus. On MRI, there were hyperintense signals in posterior cervical and thoracic levels bilaterally on T2 sequence. Replacement of vitamin B12 was done, achieving complete recovery of neurologic deficits, keeping only residual cramps in the abdomen.

Conclusion: There was a complete recovery of neurologic deficits, despite the presence of Romberg and Babinsky signs, that is different from what the literature suggests. This finding may be due to patient age, under 50 years, which allows greater neuronal plasticity as described in other studies as well as for the rapid completion of treatment, even in the first six months of evolution.

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Abstract – WCN 2013

No: 1849

Topic: 36 – Other Topic

Full recovery of limbic encephalitis after ovarian teratoma resection

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Introduction: Limbic encephalitis includes a variety of diseases affecting the limbic system, many of them related to the immune system and disease paraneoplastic. Most cases of the patients are young and they have a good prognosis if properly diagnosed. We present the case of a woman of seventeen years old who has limbic encephalitis associated with ovarian teratoma that after surgical resection has a full recovery.

Patients and methods: A seventeen year old female patient with no morbidity history presents delirium, psychotic symptoms and behavioral changes. She was initially diagnosed as schizophrenia debut; so she received standard antipsychotic treatment. She has a poor outcome with greater impairment of consciousness, hemodynamic instability and fever. On physical examination meningeal signs and abdominal distention were noted.

Material and methods: In the laboratory tests, the cerebrospinal fluid has fifteen leukocytes (monocytes), normal glucose and protein. Normal brain magnetic resonance and slow diffusion in the electroencephalogram are found. Abdominal ultrasound was performed which showed a left anexial tumor and hemoperitoneum; so the patient underwent emergency surgery for oophorectomy and tumor extirpation.

Results: Delayed biopsy concludes a mature teratoma. The patient received four days of methylprednisolone one gram a day and after three weeks the patient has recovered completely. She only presents retrograde amnesia of the episode.

Conclusion: This is one of the first cases of limbic encephalitis with complete recovery after resection of teratoma ovatico described in Chile. Should have a high level of suspicion in young female patients with psychiatric symptoms.

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Abstract – WCN 2013

No: 2609

Topic: 36 – Other Topic

Experience with rTMS in refractory cases on a third-world reference private hospital

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Introduction: rTMS consists in a repetitive magnetic discharge circumscribed to a cortical brain area and subjacent subcortical structures aimed to modulate the local and distant brain activity. Constant neuronal and network activation will progressively change the basal activity. fMRI has shown changes in the connectivity strength of involved neuronal networks. rTMS effects have been described on different neuropsychiatric disorders: neuropathic pain, fibromyalgia, depression, OCD, stroke rehabilitation.

Methods: We report the experience of 1 year using rTMS on patients with refractory conditions in a third world reference private hospital.

Results: From December 2011 to December 2012 our department received 10 patients with refractory cases of neuropathic/neurogenic pain (5 patients), OCD (3 patients) and depression (2 patients). All were evaluated before and after treatment with Hamilton depression rating scale, SF-36, and Neo-FFI-R. Pain patients were also evaluated with an analog visual scale for pain, and OCD patients were evaluated with Maudsley OC Inventory. 7 of these 10 patients showed a positive result (at least a 50% improvement on the appropriate scales – pain, MOCI, HDRS – plus improvement on quality of life scales); 2 patients showed no significant improvement and 1 showed mixed results (improvement on MOCI without improvement on quality of life).

Conclusions: rTMS is a relatively underutilized tool that may be useful for the treatment of refractory cases of neuropathic pain, depression and OCD, even in an underdeveloped country.

doi:10.1016/j.jns.2013.07.2406

Abstract – WCN 2013**No: 2614****Topic: 36 – Other Topic****Hemispheric Arteriovenous Malformation (AVM): A case report**

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Background: Hemispheric AVM are rare and represent a serious therapeutic challenge, nowadays endovascular intervention has improved their survival and the secondary co-morbidity.

Objective: To present a 20 year old woman suffering Subarachnoid Hemorrhage (SAH) due a right Hemisphere AVM.

Materials and methods: CT, MRI scans and Cranial Arteriography were required.

Results: CT scan showed a SAH, while the MRI scan did show a parenchymal bleeding in a further study, Intracranial Arteriography demonstrated a massive AVM on the Cerebral Right Hemisphere.

Conclusions: The patient underwent three subsequent immobilization endovascular procedures and as a result there is no neurological deficit.

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Abstract – WCN 2013**No: 2616****Topic: 36 – Other Topic****Injuries to patients and their bedpartners are frequent in REM sleep behavior disorder**

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Objective: REM sleep behavior disorder (RBD) is widely recognized to involve potentially injurious dream enactment behaviors (DEB). However, specific clinical factors associated with RBD-related injuries remain largely unknown. We aimed to identify injury predictors in RBD.

Methods: We surveyed consecutive RBD patients seen at Mayo Clinic between 2008 and 2010 regarding RBD-related injuries, and reviewed medical records to determine idiopathic (iRBD) or symptomatic RBD diagnosis. Associations between injuries prior to treatment and demographic, clinical, and medication variables were then determined with odd's ratios and multiple logistic regression analyses. The primary outcome variables were injury and injury severity.

Results: Fifty-three (40%) responded. Mean age was 66.2 years and 35 (75%) were men. 23 (43%) had symptomatic RBD. Twenty-nine (55%) reported injury, including self (37.8%) and bedpartner (16.7%) injury. iRBD diagnosis (OR = 7.7, p = 0.002) and dream recall (OR = 10.3, p = 0.002) were associated with injury, and iRBD diagnosis was independently associated with injury and injury severity, adjusting for age, gender, and DEB frequency. Falls during DEB (p = 0.03) were also associated with more severe injury, while DEB frequency was not associated with injury, injury severity, or falls.

Conclusions: Injuries are a frequent complication of RBD. iRBD patients are more likely to suffer injury than symptomatic RBD patients. Recall of dreams was also associated with injury, and DEB-related falls were associated with more severe injuries. DEB frequency did not predict RBD-related injuries, highlighting the importance of timely initiation of treatment for RBD in patients having even rare DEB episodes.

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Abstract – WCN 2013**No: 2675****Topic: 36 – Other Topic****Plasmodium falciparum histidine-rich protein-2 (pHRP-2) is a cerebrospinal fluid (CSF) biomarker for cerebral malaria (CM)**

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Background: Cerebral malaria (CM) causes a rapidly developing comatose state and remains a major contributor to morbidity and mortality in malaria-endemic regions. CM may be diagnosed during life by the presence of *Plasmodium falciparum* parasitemia, clinical assessment and retinal changes. The plasma level of pHRP-2, a malaria histidine- and alanine-rich protein, identifies children with histologically confirmed or retinopathy-positive CM with high specificity and sensitivity. We sought to determine whether pHRP-2 levels in the CSF were detectable in CM cases.

Materials and methods: The immuno-PCR technique was adapted for malaria use by coupling a 60 base pair oligonucleotide to a monoclonal HRP2 antibody. The limit of detection was demonstrated to be 1 pg per sample with specificity over 90%. The immuno-PCR test was used on 84 CSF samples from Tanzanian CM patients and 33 non-malaria control samples.

Results: In our patient cohort, CSF pHRP-2 was present by a novel immuno-PCR in 98% (82/84) of CM samples. The geometric mean pHRP-2 concentration in CSF was 20 pg/μl with a mean and median of 10 pg/μl. A rapid diagnostic test (RDT) was positive in 81% (68/84) of the samples. In 11 matched plasma and CSF samples of CM patients, the ratio of plasma to CSF pHRP-2 was 168. The 33 non-malarial control samples did not have detectable levels of CSF pHRP-2.

Conclusions: pHRP2 is present in the CSF of patients with CM. Further studies are needed to determine the utility of CSF pHRP-2 testing in malaria-endemic regions.

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Abstract – WCN 2013**No: 2718****Topic: 36 – Other Topic****HbA1c is associated with sural nerve regeneration and degeneration in diabetic neuropathy**

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Background: Diabetic neuropathy (DN) is the most common, debilitating, and costly complication of diabetes. Patients with DN demonstrate variable degrees of nerve regeneration and degeneration; however, risk factors affecting these processes are not understood.

Objective: To identify risk factors closely associated with nerve regeneration in patients with DN.

Materials and methods: We retrospectively examined demographic, anthropometric and biochemical data of diabetic subjects from a double-blind, placebo-controlled, 52-week trial of **acetyl-L-carnitine**. Based on the change of sural nerve myelinated fiber density (DMFD%), subjects were designated as Regenerator (top 16 percentiles, n = 67),

Degenerator (bottom 16 percentiles, $n = 67$), or Intermediate ($n = 290$), with dramatically increased, decreased, or steady DMFD%, respectively. Fisher's exact test, ANOVA, and multifactorial logistic regression analyses were performed to identify significant risk factors. A preliminary microarray experiment was also performed on a subset of samples to identify regeneration-related genes and pathways.

Results: DMFD% were 35.6 ± 17.4 (Regenerator), -4.8 ± 12.1 (Intermediate), and -39.8 ± 11.0 (Degenerator). HbA1c at baseline was the only risk factor significantly different between Regenerator ($8.3 \pm 1.6\%$) and Degenerator ($9.2 \pm 1.8\%$). Support Vector Machine classifier using HbA1c alone demonstrated 62.4% accuracy of classifying subjects into Regenerator or Degenerator. Gene expression studies revealed that up-regulated genes in Regenerator are enriched in cell cycle and myelin sheath functions, while down-regulated genes are enriched in immune/inflammatory responses.

Conclusions: Our data suggest that HbA1c level predicts myelinated nerve fiber regeneration and degeneration in patients with DN, and that maintaining optimal blood glucose control is essential to prevent continued nerve injury in patients with DN.

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Abstract – WCN 2013

No: 2720

Topic: 36 – Other Topic

Epigenetic miRNA dysregulation as a mechanism for sporadic amyotrophic lateral sclerosis

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Background: Amyotrophic lateral sclerosis (ALS) is a progressive and fatal neurodegenerative disease. The majority of cases are sporadic (sALS) with unknown causes, and the late onset of sALS suggests that long-term exposures to environmental factors may play a role in disease pathogenesis. Epigenetic mechanisms, such as miRNAs, may be driven by adverse environmental factors proposed to contribute to sALS etiology to promote an accumulation of altered gene expression, culminating in sALS.

Objective: Our goal is to address the role of miRNAs in sALS pathogenesis. We hypothesize that differential expression of miRNAs in human sALS spinal cord promotes dysregulation of key genes and biological pathways leading to sALS.

Materials and methods: We combined two high-throughput profiling assays to identify differentially expressed miRNAs (TaqMan OpenArray) and differentially expressed genes (DEGs; Affymetrix GeneChip Human Genome U133 Plus 2.0 Array) in postmortem human spinal cord tissue from sALS patients and controls. Since miRNAs negatively regulate gene expression, inverse correlations between miRNA and DEGs were identified and examined with multiple miRNA prediction target databases to determine potentially relevant miRNA/DEG pairs. Gene and miRNA expression was confirmed by qPCR.

Results: We identified 90 differentially expressed miRNAs and 1182 DEGs. Of these differentially expressed miRNAs and DEGs, prediction databases identified miR-577, let-7 miRNA family members, miR-133b, and miR-140b-5p as potential miRNA regulators of the gene targets FAS, CD4, EIF2C4, and CCL2.

Conclusion: Changes in miRNAs and their corresponding gene targets may represent, in part, a pathogenic mechanism leading to sALS.

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Abstract – WCN 2013

No: 2715

Topic: 36 – Other Topic

Spontaneous remission and relapse of primary central nervous system lymphoma: Case report and review

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There are numerous reports of lesions affecting the central nervous system which disappear without treatment. They are called vanishing tumors. The most common of these is the primary central nervous system lymphoma, which accounts for 4% of all intracranial neoplasms. There are other factors that induce remission such as corticosteroid therapy, chemotherapy and radiotherapy. However, many of these vanishing tumors relapse. We present the case of a 58 year old, male patient who presented with partial third cranial nerve palsy. The brain MRI showed a periventricular mass and the biopsy was informed as diffuse large B-cell non-Hodgkin lymphoma. A control MRI was requested displaying complete disappearance of the mass. Patient was followed during 3 months with MRI, full body CT and lumbar puncture showing no evidence of the disease. One year after discharge patient presented with subtle memory alterations and progressive temporal-spatial disorientation. MRI showed a heterogeneous, hypointense, expansive process with left temporal periventricular epicenter. Spectroscopy was suggestive of lymphoma. Thoracic, abdominal, and pelvic CT scans evidenced an L1 lumbar fracture; MRI was suggestive of lymphoma metastasis. Daily methylprednisolone pulses were initiated and treatment with Methotrexate, Cytarabine and Leucovorin was begun, with clinical improvement. Many reports present relapsing vanishing tumors, suggesting that full therapy should be initiated in spite of spontaneous remission. Clinical trials should be performed in order to prove the effectiveness of this proceeding.

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Abstract – WCN 2013

No: 2768

Topic: 36 – Other Topic

Syndroma Brown-Séguard as a rare manifestation of von Hippel-Lindau disease

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Case report: We report a 45-year-old female patient with Brown-Séguard Syndrome (BSS). The onset of disease was 14 years before admission to our Department, during the first pregnancy, when she felt thoracic pain. Five years later, during the next pregnancy, she felt numbness at umbilical region and drop of right foot, but all symptoms disappeared spontaneously 10 days after delivery. MRI of thoracic spine showed syrinx from Th5 to L1. Now, at the admission neurological examination revealed spasticity, increased muscle reflexes and positive Babinski's sign on the right leg, with ipsilateral loss of tactile discrimination, vibratory, and position sensation below the level of right rib arch. Contralaterally there was reduction in pain and temperature sensation below the level of Th8. MRI of thoracic spine showed multiple intradural nodular tumors at the levels of Th3–5 and Th11 with hydrosyringomyelia in whole spinal cord. MRI of the brain showed multiple hemangioblastomas in the craniocervical region with spreading at the level of C1 and in both cerebellar hemispheres. Multi-slice computed tomography (MSCT) of abdomen showed multiple pancreatic cysts and suspected liver hemangioma. Ophthalmological, otological and gynecological findings were normal. Genetic analysis

revealed a deletion in von Hippel–Lindau (VHL) gene 2 exon at codon 128. It was made a diagnosis of de novo VHL disease type I (without pheochromocytoma).

Conclusion: Hemangioblastomas of the spinal cord are a rare cause of BSS, especially as a manifestation of VHL disease. Detection of these tumors and their removal could contribute to the improvement of neurological finding and prevention of further complications.

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Abstract – WCN 2013

No: 2733

Topic: 36 – Other Topic

Common presentation – atypical diagnosis

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Background: Literature review reveals that the reported incidence of Dural Arteriovenous Fistulas (DAVFs) is approximately 10–15% of all intracranial vascular abnormalities. We report the first case where a DAVF caused an obstructive hydrocephalus in a patient.

Objective: The objective of our poster presentation is to make clinicians aware of the rare possibility of an intracranial vascular lesion causing obstructive hydrocephalus.

Patients and methods: A 76-year-old woman presented with poor mobility, frequent falls, confusion and urinary incontinence for a month. She was initially treated for a urinary tract infection. CT scan of her head showed obstructive hydrocephalus and an MRI brain scan confirmed that this was caused by a dilated vascular lesion in the posterior aspect of the third ventricle and also near the inferior aspect of the cerebral aqueduct. Digital subtraction angiography of her brain showed that the vascular lesion was a DAVF obstructing the flow of CSF.

Results: A conservative approach was taken for the DAVF and the patient had an endoscopic third ventriculostomy to treat her obstructive hydrocephalus. All her symptoms improved and the patient went home after a short period of rehabilitation.

Conclusion: There's no other reported case in the literature where a DAVF has caused obstructive hydrocephalus. Our patient improved with treatment and was discharged home. As clinicians, we need to be aware of common presentations with atypical diagnosis.

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Abstract – WCN 2013

No: 2786

Topic: 36 – Other Topic

Non-invasive detection of cortical population spikes: Functional discrimination of pre- vs. postsynaptic components in SEP at 1 kHz

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Introduction: Ultrafast EEG signals, featuring frequencies above 500 Hz, can be observed in scalp somatosensory evoked potentials (SEP). Usually, these recordings have a low Signal-to-Noise Ratio (SNR) because weak signals are overlaid by intrinsic noise of much higher amplitude as generated by biological sources and the amplifier. Scalp recordings of SEP show bursts above 400 Hz with submicro-volt amplitudes, which could be extracted from noise up to now only by the use of massive averaging ($n > 1000$) and digital signal processing.

Methods: Here, we investigate SEP up to 1 kHz elicited by median nerve stimulation with rates of either 0.99 Hz or 8.1 Hz. We exploit the increased SNR provided by a custom-made low noise amplifier technology together with an adapted implementation of Canonical Correlation Analysis algorithm, based on the assumption of high level stimulus phase locking across trials.

Results: It proved possible in 3/3 subjects to discriminate hf-SEP generators at subcortical and intracortical levels at the single trial level, relying on the different neuronal refractory properties of these components which are phase-locked to the stimulus onset.

Discussion: A custom-made low-noise EEG system enables the noninvasive detection of pre- and postsynaptic evoked activity at 1 kHz, i.e., in the frequency range housing spike-related multi-unit activity in intracortical recordings, providing the opportunity of non-invasive monitoring of multi-unit spike activity in man.

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Abstract – WCN 2013

No: 2772

Topic: 36 – Other Topic

Total ophthalmoplegia due to giant cell arteritis

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We report the case of a lady who presented with total ophthalmoplegia due to giant cell arteritis.

Case report: A 79 year old lady presented with two days of blurring of vision with abnormal gait. Observation on arrival was normal. She had bilateral ptosis, total ophthalmoplegia. Visual acuity in the left eye was able to count finger and on the right 9/6. She had wide base gait, positive Romberg's sign due to compensating of her visual impairment but her reflexes diminished bilaterally with down-going planters, with normal sensation. There was loss of proprioception of both feet. There was no incontinence of urine or faeces, with normal speech and swallow.

ECG, chest X-ray and MRI/DWI brains. Cerebral spinal fluid showed normal protein level. Superficial temporal artery biopsy was normal. Her ESR was 108. Vasculitis screen, HIV and syphilis serology, serum electrophoresis and urine for Bence Jones protein, anti Ro, anti La, anti GAD, TPO antibodies, and acetyl choline receptor antibodies were all negative. Nerve conductive study performed showed a sensory-motor polyneuropathy related to diabetes mellitus.

She was commenced on 60 mg Prednisolone with Bisphosphonate. Her ESR dropped from 108 to 54. She has had balance and gait improved since admission but patient did not show significant eye improvement.

Discussion: Giant cell arteritis (GCA) is a systemic inflammatory vasculitis. It can result in a wide variety of systemic, neurologic, and ophthalmologic complications including visual loss. Prompt treatment with steroids can prevent blindness and other vascular sequelae of GCA.

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Abstract – WCN 2013

No: 2816

Topic: 36 – Other Topic

Treatment for paraneoplastic neuropathies

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Paraneoplastic neuropathies are the most frequently reported paraneoplastic syndromes. They are, however, heterogeneous and require several therapeutic approaches. This review was undertaken to

systematically assess any data available from randomised controlled trials (RCTs) on the treatment of paraneoplastic syndromes of the peripheral nervous system.

Objectives: To assess the benefits and harms of treatments for paraneoplastic neuropathies.

Selection criteria: We planned to include all RCTs and quasi-RCTs (in which allocation is not random but is intended to be unbiased, for example alternate allocation) of any treatment for paraneoplastic neuropathies. Since we expected that there are to be few or no included studies, we also planned to assess and summarise observational studies, prospective and retrospective comparative cohort studies, case–control studies and case series that met minimum criteria in the discussion.

Main results: Despite many reports on paraneoplastic neuropathy, we identified no RCT or quasi-RCTs for inclusion in this review. We found only six studies, involving 54 participants, from among the non-randomised evidence that were judged by predefined criteria to be of suitable quality for inclusion in the discussion. These studies were not readily comparable. The treatments focused on tumour treatment and immunomodulation, mainly intravenous immunoglobulin.

Authors' conclusions: At present there are no RCTs or quasi-RCTs of treatment for paraneoplastic neuropathies on which to base practice. There is only evidence from case series, case reports or expert opinion (class IV evidence) for the effect of immunomodulation (intravenous immunoglobulin, plasma exchange, steroid treatment or chemotherapy) on paraneoplastic neuropathy.

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Abstract – WCN 2013

No: 2780

Topic: 36 – Other Topic

NMDA-receptor mediated excitotoxicity is involved in the pathogenesis of experimental cerebral malaria

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A major cause of morbidity and mortality of *Plasmodium falciparum* malaria is cerebral malaria (CM). Mortality is high and neurological sequelae are observed frequently. A deeper understanding of the pathophysiology is missing yet. In addition to ischemia and inflammation, excitotoxicity might be an important factor. The current study investigates the role of NMDA-receptor mediated excitotoxic cell death during CM and its potential for the development of adjunctive treatment strategies.

C57BL/6J mice were infected with *Plasmodium berghei* ANKA. Cerebral Microdialysis was performed and glutamate levels were measured. Animals with CM were randomized for treatment with artesunate, MK801 (a non-competitive NMDA-receptor antagonist), artesunate and MK801 or vehicle over 5 days. Glutamate levels were significantly elevated in mice with CM compared to control animals. Subsequently NMDA antagonisation was investigated. Twenty-eight animals were randomized. No animals survived in the MK801 or vehicle treated group. In contrast, 33.3% of the animals in the artesunate group and 74.1% in the MK801/artesunate combination treatment group survived. Kaplan Meier survival curves yielded a significantly longer survival of the animals in the combination treatment group compared to the vehicle or MK801 treatment group. Histological analyses yielded a lower number of micro-hemorrhages and Fluoro-Jade B positive cells in the artesunate/MK801 treated animals compared to artesunate treated mice. In conclusion, glutamate levels in the brain are increased early in the course of CM.

Treatment with MK801 rescues mice from CM. NMDA-receptor mediated excitotoxicity plays a role in the pathogenesis of CM and could represent a target for adjunctive treatment strategies.

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Abstract – WCN 2013

No: 2842

Topic: 36 – Other Topic

The effect of stress on long-term memory consolidation: An update

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Background: It has long been recognised that stress has a profound effect on long-term memory consolidation. The past decade has seen substantial growth in efforts to characterise this relationship. Numerous potential mechanisms have been highlighted, involving biochemical regulators of emotional arousal and specific neural pathways. More recently, research has led to increased understanding of interactions between regions of the brain previously not acknowledged to contribute to memory consolidation.

Objectives: We present a systematic review to expound the prevailing theories behind this relationship, in addition to the supporting experimental evidence and potential clinical implications.

Materials and methods: The review was performed using Medline/Pubmed and Embase, using search terms {(stress) AND (memory)}, under constraints of “English language” and “humans”.

Results: Behavioural and physiological evidence has begun to establish a convincing role for stress; consistently demonstrating enhanced memory under stressful conditions. Wide support is found for effects of epinephrine and glucocorticoids on consolidation, in addition to electrophysiological evidence for specific neural influences.

Functional imaging advocates an interaction between the basolateral amygdala, nucleus accumbens and locus coeruleus with the hippocampus, although uncertainty prevails regarding the pathways involved. Clinical implications of the stress–memory relationship are undoubtedly far-reaching, and are discussed herein.

Conclusions: Although precise mechanisms by which stress affects memory remain unclear, the advent of functional neuroimaging and growing interest in memory consolidation has produced rapid advances in understanding. Research clarifying the relationship between stress and memory consolidation undoubtedly merits further attention given the additional insights provided by these advances and their significant therapeutic potential.

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Abstract – WCN 2013

No: 2640

Topic: 36 – Other Topic

Comorbidities and mortality associated with intracranial bleeds and ischaemic stroke

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Background: Stroke is a leading cause of mortality and acquired disability, however to date, there exists no comprehensive comparison of risk factors between stroke sub-types.

Objective: The aim of the present study was to compare risk factors and mortality across different intracranial pathologies; subdural haematoma, subarachnoid haemorrhage, ischaemic stroke and haemorrhagic stroke.

Patients and methods: We compiled a database of all patients (n = 4804) admitted with these conditions to a large teaching hospital in Birmingham, UK during the period 2000–2007. The ICD 10th revision coding was used to identify these cases. Multinomial logistic regression was performed to analyse differences in the frequencies of comorbidities, associations were regarded as significant when $p < 0.001$ only.

Results: Subdural haematoma patients had lower prevalences of hypertension and pneumonia, whereas alcohol abuse was more common. In subarachnoid haemorrhage, epilepsy was less frequent (OR: 0.37; CI: 0.22–0.64). As would be expected, a range of cardiovascular risk factors were more common in ischaemic stroke including for example type II diabetes mellitus and ischaemic heart disease. Brain tumours were more common in haemorrhagic stroke. There was no difference in the duration of hospital stay between the four groups. Mortality was significantly higher in ischaemic and haemorrhagic strokes, and the duration to death was greater in ischaemic stroke.

Conclusion: Our findings provide novel associations to inform the preventative measures undertaken by healthcare professionals and this may lead to more rigorous control of risk factors. Further studies are required to elucidate the mechanisms by which subarachnoid haemorrhage protects against epilepsy.

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Abstract – WCN 2013

No: 2641

Topic: 36 – Other Topic

Predictors of mortality in ischaemic stroke patients – A hospital admissions study: 2000–2012

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Background: Ischaemic Stroke is a leading cause of mortality and morbidity in the UK, accounting for 9% of all deaths.

Objective: We aimed to identify predictors of mortality in Ischaemic Stroke patients, in a large hospital population in the UK.

Patient and methods: Anonymous information on patients with Ischaemic Stroke, attending a large multi-ethnic general hospital in Manchester, UK during the period 2000–2012 was obtained from the hospital activity analysis register using ICD-10 and OPCS coding systems. Statistical analysis was performed using SPSS version 20.

Results: Out of 458,410 patients admitted, there were 3541 (0.8%) patients with Ischaemic Stroke; mean age 69.7 yrs \pm 14.6 (SD). The main comorbidities were Hypertension (48.5%), Type 2 Diabetes Mellitus (21.5%), Atrial Fibrillation (18.7%), Hyperlipidaemia (15.3%), Myocardial Infarction (11.2%), and Heart Failure (9.2%). A multinomial logistic regression model accounting for variations in age, sex and ethnic group showed that Hypertension (OR: 0.8; CI: 0.7–0.9), Type 2 Diabetes Mellitus (OR: 1.2; CI: 1.0–1.5), Atrial Fibrillation (OR: 1.4; CI: 1.2–1.8), Hyperlipidaemia (OR: 0.4; CI: 0.3–0.6), Heart Failure (OR: 3.0; CI: 2.2–4.1) and Duration of stroke (OR: 1.0; CI: 1.0–1.0) as significant predictors of mortality ($p < 0.05$).

Conclusion: We have shown that Hypertension, Type 2 Diabetes Mellitus, Atrial Fibrillation, Hyperlipidaemia, Heart Failure and Duration of stroke are significant predictors of mortality in patients with Ischaemic Stroke from a large hospital based sample in the UK. Our improved understanding of these predictors will guide clinicians to focus on these high risk groups to improve clinical practice and outcomes in patients with Ischaemic Stroke.

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Abstract – WCN 2013

No: 2850

Topic: 36 – Other Topic

Waterhouse–Friderichsen syndrome induced by meningococcal meningitis in young adult with SLE

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Background: Although patients with systemic lupus erythematosus are more susceptible to infections, meningococcal meningitis is relatively rare among these patients.

Case report: We report a case of neisserial meningitis in an 18 years old male that was under treatment for SLE (monotherapy with hydroxychloroquine). The patient presented with one day fever, headache, arthralgias, purpuric rash on two sites (right gluteal region, left arm) and neck rigidity. CSF had a cell count of 5200 RBC/ μ L, 25 WBC/ μ L with 13 NEU/ μ L (corrected for 20 WBC/ μ L) and the stain and combo tests were negative for bacteria. Both CSF and blood cultures revealed *Neisseria meningitidis* group B. Total blood count displayed low platelet count. Biochemistry revealed elevated creatinine and CRP. Despite the prompt diagnosis and treatment, the disease progressed rapidly with major complications, such as sepsis, DIC, purpura fulminans, myocardopathy, respiratory distress and finally evolved to Waterhouse–Friderichsen syndrome.

Discussion and conclusion: This is a rare case of meningococcal serogroup B meningitis in a patient with SLE. In SLE meningitis, the most common pathogens are *Mycobacterium tuberculosis*, *Cryptococcus neoformans* and *Listeria monocytogenes*. It is interesting that the laboratory tests at presentation did not absolutely reflect the severity and rapid progression of the infection. In addition, although there are references of corticosteroid use as a predisposing factor to meningitis, there are no reports for hydroxychloroquine alone.

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Abstract – WCN 2013

No: 2890

Topic: 36 – Other Topic

Brain imaging in chorea in Tunisian children

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Background: Studies on brain imaging aspects of chorea are scarce, and there is no such study in Tunisia in children.

Objective: To determine imaging characteristics of chorea in Tunisian children.

Patients and methods: We conducted a descriptive study over an 8-year period (2004–2012) including all children diagnosed with chorea and followed at our department. All the patients had a brain MRI and 23 of them a brain CT-scan. The imaging aspects have been analysed.

Results: 54 children presented with chorea (sex ratio = 1.1, mean age = 7.6 years, mean age of onset = 3.1 years). Brain CT scan (23 patients) was abnormal in 18%, mainly in metabolic diseases. Brain MRI was abnormal in 74%, especially in metabolic causes (31%) and cerebral palsy (14%). Imaging abnormalities are dominated by abnormalities of the brain white matter (24%) and cortico-sub-cortical atrophy (14%). Basal ganglia (mainly putamen) and thalamus show abnormalities in only 5%, in association with other movement disorders.

Conclusion: Our study is the first to discuss imaging aspects of chorea in childhood in Tunisia. The high frequency of imaging abnormalities in metabolic diseases and cerebral palsy may be explained by the

vulnerability of basal ganglia to hypoxic-ischemia in cerebral palsy, and variations of metabolism and associated leukodystrophy in neuro-metabolic disorders. Atrophy could be a sequelae, or of degenerative origin. Therefore, brain imaging, especially brain MRI, should be done in every child with chorea.

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Abstract – WCN 2013

No: 2783

Topic: 36 – Other Topic

Cortical afferent inhibition is reduced in patients with idiopathic REM sleep behavior disorder and cognitive impairment: A TMS study

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Background: Impaired cognitive profile and electroencephalography (EEG) slowing have been reported in patients with REM sleep behavior disorder (RBD), but the neurobiological significance of these findings remains unknown. The cholinergic system is known to play a key role in all attentional processes and cognitive functions.

Objective: A transcranial magnetic stimulation (TMS) protocol may give direct information about the function of some cholinergic circuits in the human brain; this technique relies on short latency afferent inhibition (SAI) of the motor cortex. The objective of this study was to test the hypothesis that cognitive performance and cortical activation in RBD patients are associated with a dysfunction of the cholinergic system.

Materials and methods: We applied the SAI technique in a group of 10 patients with idiopathic RBD (iRBD) and compared the data with those from a group of 15 age-matched healthy subjects. All the iRBD patients and the control subjects also underwent an extensive neuropsychological evaluation.

Results: Mean SAI was significantly reduced in patients with iRBD when compared with controls. Neuropsychological examination showed mild cognitive impairment in six out of the 10 iRBD patients. SAI values correlated strongly with tests measuring episodic verbal memory and executive functions.

Conclusions: These results support the hypothesis of cholinergic dysfunction in some patients with iRBD who develop cognitive impairment. Our findings raise the possibility that the presence of SAI abnormalities may indicate increased risk of cognitive impairment in patients diagnosed with iRBD.

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Abstract – WCN 2013

No: 2895

Topic: 36 – Other Topic

Using clinical details to direct attention to the correct lesion in a patient with history of breast cancer

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Background: Sensitive imaging detects many asymptomatic brain lesions which can distract attention from significant problems in

patients with cancer. Continuity of care with imaging done for any CNS symptoms can provide a basis for longitudinal comparison.

Objective: To facilitate cooperation between clinicians and radiologists to avoid missing important but small lesions.

Patients and methods: Case report of a 62 year old nurse with 5 year history of breast cancer treated with chemotherapy, local resection and radiation presented with severe vertigo and vomiting. Dizziness had caused her to have CT scan three years earlier which showed a 1.5 cm round hyperdense frontal lesion. CT with contrast showed a tiny enhancing area above the fourth ventricle; MRI with 1 mm cuts detected a 3 mm lesion with slight edema, and no change or bleeding in the previously seen lesion.

Results: At a conference between oncologist, neuroradiologist and neurologist the significance of the new lesion in the area postrema was underscored. Arrangements were made to resume cancer treatment, with “gamma knife” followed by whole brain radiation to manage all potential micrometastases.

Conclusion: Radiologists need information from treating physicians to properly interpret imaging and direct attention to important lesions, in the presence of other (benign) lesions that are asymptomatic and stable. In patients referred to tertiary cancer centers a mechanism for sharing imaging with primary centers is mandatory.

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Abstract – WCN 2013

No: 2899

Topic: 36 – Other Topic

Identifying interactions between EphB3 and EphA4 dependence receptors using bimolecular fluorescence complementation

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Objectives: The goal of our study is to identify protein–protein interactions of membrane-bound receptor tyrosine kinases that regulate cellular functions after CNS trauma. We discovered that Eph receptors influence cell survival following CNS injury through pro-apoptotic ‘dependence receptors’, which induce cell apoptosis after cellular stress. To examine their interaction we used bimolecular fluorescence complementation (BiFC).

Methods: To visualize protein interactions, cDNA of EphB3 and EphA4 was amplified by PCR and cloned with sequences encoding Cyan Fluorescent Protein (CFP). Eph vectors were transfected into human embryonic kidney (HEK) cultured cells, with GFP and empty vectors as controls. The BiFC of Jun and Fos fragments fused to N- and C-termini of mutated yellow fluorescent protein (YFP) Venus was also performed as positive control.

Results: Green fluorescence was observed after 24 hours of transfection of GFP into HEK cells. Using the GFP filter, we also observed fluorescence with Venus-based fragments Jun and Fos. Introducing EphB3 fused with C terminus of CFP with EphB3 with N terminus did not show expected blue fluorescence, which might reflect inadequate linker lengths between the receptor and CFP fragments.

Conclusion: BiFC assay was used to study the interaction between EphB3 and EphA4 in live HEK cells. The N- or C-terminal halves of a CFP are linked to EphB3 and EphA4, respectively. Since blue fluorescence was not observed between two EphB3 receptors that are known to interact, the next step is to modify linkers. Future studies examine variable linker lengths required for membrane receptor interactions to occur in living cells.

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Abstract – WCN 2013**No: 2909****Topic: 36 – Other Topic****Paraneoplastic encephalitis due to Merkel cell neuroendocrine skin carcinoma**

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Background: We report a case of a 57-year old man with subacute cerebellar ataxia related to Merkel cell carcinoma.

Case description: A 57-year-old man presented with a subacute, progressive cerebellar ataxia with gait instability, marked trunk ataxia and explosive speech. Brain MRI, whole-body CT and complete blood work-up were negative. CNS showed elevated protein but a complete CNS serological (all known viruses), immunological and paraneoplastic antibody testing were negative. PET-scan showed an inflamed inguinal lymph node (attributed to skin infection). Genetic testing for inherited ataxias was negative. The patient received high-dose corticosteroids with remarkable improvement. One month later his symptoms reemerged. A second course of corticosteroids succeeded in controlling his symptoms but significant side effects led to their discontinuation. A third relapse occurred. Although the lymph node had diminished, a biopsy was decided which showed Merkel cell carcinoma. Complete skin examination revealed no abnormal skin nodule and a second PET scan was now negative. The patient showed remarkable improvement of his neurological symptoms after the lymph node removal and is receiving chemotherapy.

Conclusion: Merkel cell skin carcinoma (MCC) is a rare, aggressive cutaneous malignancy linked to UV radiation exposure, infection with the Merkel cell polyomavirus, and immunosuppression. To our knowledge no case of related paraneoplastic encephalitis has ever been reported and although no direct link is determined between the tumour and the cerebellar insult the fact that the removal of the lymph node (primary tumour), led to an almost complete neurological recovery is a strong indication that the two are connected.

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Abstract – WCN 2013**No: 2908****Topic: 36 – Other Topic****Effect of *Satureja khuzestanica* essential oil on acquisition, extinction and reinstatement of morphine induced conditioned place preference in male rat**

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Background: Drug addiction is an impulsive, unconscious drug seeking behavior associated with neurobiological changes. This neurobiological disorder accompanies high recurrence even after long period of treatment. The present study, investigated the effects of *Satureja khuzestanica* essential oil (SKEO) a well-known Iranian traditional medicine, on acquisition and reinstatement of morphine-induced conditioned place preference (CPP) in rats.

Methods: CPP was induced by intraperitoneal injection of morphine (10 mg/kg) as an 8-day conditioning schedule. The effects of SKEO on the rewarding properties of morphine were tested in rats receiving SKEO (10, 15, and 20 mg/kg i.p.) 30 min prior to each morphine injection (acquisition) or prior to the CPP test on day 6 (expression). Once established, CPP was extinguished by repeated testing, during

which conditioned rats were injected daily with different doses of SKEO. Finally, the efficacy of SKEO in blocking reinstatement of CPP provoked by priming injections was also evaluated.

Results: SKEO administration showed dose dependency for prevention of establishment of CPP. However it had no effect in facilitating extinction of morphine-induced CPP. SKEO suppressed priming induced reinstatement of CPP.

Conclusions: In conclusion, as SKEO was effective for reducing craving and vulnerability to relapse, it might be a very effective natural remedy for the treatment of opioid addiction.

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Abstract – WCN 2013**No: 2911****Topic: 36 – Other Topic****Apathy is associated with a single-nucleotide polymorphism in a dopamine-related gene**

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Background: Dopaminergic neurotransmission is an important factor in the pathogenesis of apathy. In addition, the contribution of genetic factors to the regulation of brain dopaminergic activity is widely acknowledged. Therefore, we hypothesized that genes associated with brain dopaminergic activity may have some effects on apathy.

Objective: In the current study, we evaluated the association between four functional single-nucleotide polymorphisms (SNPs) in specific genes related to dopaminergic neurotransmission and apathy in a general population.

Methods: Participants in the health examination at the Shimane Institute of Health Science were recruited for this study (n = 963). Apathy was assessed using the Japanese version of the apathy scale. SNPs were genotyped using the TaqMan method.

Results: In our population, 22.1% had apathy. We confirmed that apathy was associated with decreased cognitive function and depressive state. A significant association was found between an SNP in the catechol-O-methyltransferase (COMT) gene (rs4680) encoding the low-activity Met allele and apathy (GA: OR = 0.67, p = 0.01, CI = 0.48–0.92 and AA: OR = 0.47, p = 0.02, CI = 0.23–0.89). This relationship was still significant after adjustment for confounding factors (GA: OR = 0.59, p = 0.003, CI = 0.42–0.84 and AA: OR = 0.34, p = 0.002, CI = 0.15–0.67).

Conclusions: Our study indicates an association between rs4680, an SNP in the COMT gene, and lower risk of apathy. Considering the function of rs4680, the current study suggests the importance of dopaminergic neurotransmission in the pathogenesis of apathy in a general population.

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Abstract – WCN 2013**No: 2871****Topic: 36 – Other Topic****Sensory-motor integration after selective stimulation of intraepidermal Aδ nociceptors**

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Background and objective: Previous studies on sensory-motor integration (SMI) between somatosensory afferents and the motor

responses to transcranial magnetic stimulation (TMS) have been performed with electrical stimulation of the nerves, revealing early (20–30 ms) and late (after 100 ms) afferent inhibitions with an intervening facilitation. This study was planned to investigate the effects of selective electrical stimulation of intraepidermal A δ fibers (IES) on SMI.

Material and methods: Fourteen healthy volunteers aged between 22 and 40 were included. Paired stimuli at 2 times the sensory threshold were applied with custom made IES electrodes placed on the 1st dorsal web space of the dominant hand. A butterfly shaped coil was placed over the cortical hot spot for the first dorsal interosseous muscle adjacent to the IES and the TMS intensity was adjusted for responses >1 mV in amplitude at >5/10 stimulations. Inter-stimulus intervals (ISI) between the IES and TMS consisted of 18 steps between 20 and 450 ms. Eight stimulations for each ISI were performed randomly. Mean amplitudes and areas of the motor responses elicited at each ISI were divided by those evoked by TMS alone and graphs for SMI were created.

Results: SMI curves showed an afferent inhibition that began after ISI 80 ms and lasted until 300 ms.

Conclusion: Later beginning of afferent inhibition after IES can be considered as an additional evidence for selective excitation of the intraepidermal nerve fibers by the described method. The long duration of the inhibition may be a reflection of the different central processing for the nociceptive afferents.

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Abstract – WCN 2013

No: 2882

Topic: 36 – Other Topic

Functional outcomes after rehabilitation of tuberculous myelitis 14 case reports

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Introduction: Tuberculosis including its medullar location is a public health problem in Morocco.

Objective: We report the epidemiological characteristics, clinical and functional aspects in a group of patients suffering from tuberculous myelitis after rehabilitation.

Material: Over a period of 4 years, 14 cases of tuberculous myelitis (F/H = 6:8) were admitted for rehabilitation. We noted epidemiological, clinical and complication characteristics. The ASIA neurological status, the mobility and the sphincter control were assessed by the MIF.

Results: Most patients were male (F/M = 6:8), the average age was 29 years (19–39), the average length of stay in the rehabilitation was 79.1 days (21–103), paraplegia was complete in nine patients according to ASIA classification. Six patients had multifocal tuberculosis. All patients had sphincter dysfunctions, and had to learn self catheterization, three patients had complications. All patients returned to their homes after discharge, one patient died after 6 months, one patient, seen after 2 years, has recovered autonomous walking capacity. The results of the MIF averaged 54.1 out of 126 (42–59) at admission and 75 (69–89) at discharge.

Conclusion: Tuberculosis including its medullar location is a public health problem in Morocco. The main goal of rehabilitation is to increase patient independence and reduce disability. A large proportion of patients with non-traumatic spinal cord can reach a good level of mobility and sphincter independence. Disability was significantly reduced during rehabilitation.

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Abstract – WCN 2013

No: 2878

Topic: 36 – Other Topic

Narcolepsy secondary to a third ventricle glioblastoma

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Background: Secondary narcolepsy is a rare entity, as is a glioblastoma developing in the third ventricle. We report a case of narcolepsy secondary to a third ventricle glioblastoma.

Objective: To report the occurrence of narcolepsy secondary to glioblastoma.

Methods: Case report.

Result: A 24-year-old man presented with recurrent episodes of headache and vomiting. Imaging studies showed obstructive hydrocephalus and a third ventricle lesion. He underwent several surgeries, with partial removal of the tumor. After the last surgery, the histological diagnosis was glioblastoma, and further treatment with focal radiotherapy and temozolomide was started. During hospitalization, the patient was asleep during the day for short periods and sleep was interrupted by multiple awakenings at night. He was often found with eyes closed, even during the physical examination, while continuing to perform and respond adequately. Polysomnogram revealed a fragmented sleep pattern, with a predominance of REM sleep and superficial sleep, without sleep cycles. A multiple latency test was also performed, confirming the hypersomnia and showing average latency time of 8.6 minutes and 3 sleep-onset REM on 4 sleep periods. These findings were suggestive of narcolepsy. Modafinil was instituted, with clear and immediate benefit; cycles of sleep and wakefulness returned to his premorbid pattern.

Conclusions: In this case, we assume the existence of a causal relationship between tumor location and narcolepsy. The diagnosis and therapy directed to narcolepsy, in addition to cancer treatment, resulted in a significant clinical improvement.

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Abstract – WCN 2013

No: 2884

Topic: 36 – Other Topic

Pathologic reflexes of proximal segments in ALS patients

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Introduction: Amyotrophic Lateral Sclerosis is a neurodegenerative disease that affects both motor cortex and anterior horn motoneurons producing a progressive amyotrophy of all voluntary muscles including the ones innervated by the cerebral bulb. The atrophy progresses from the distal muscles of the limbs to the central ones, this is clearly detected by an electromyography study. The hyperreflexia of ankles, knees, wrists and elbow are pathognomonic signs of cortical liberation of the spinal cord reflex arches. Absences of this sign talk against the first neuron compromise inducing a more benign diagnosis. In the hospital setting several patients came with advanced stages of ALS, but their diagnosis became difficult because the myoatrophy of the distal members is hidden the above mentioned hyperreflexia. To find out a low cost way to overcome these problems there was a study of the presence of shoulder and hip muscular tendon reflexes.

Methods: 10 patients that met all El Escorial ALS Criteria were studied and followed for at least one year. The three joint reflexes of each limb were assessed every three months for, at least, a year.

Results: 10/10 patients showed the three reflexes present in the four limbs. After a year 8/10 had lost the two distal reflexes keeping the hyper reflexia of shoulder and limbs.

Discussion: There is a lack of previous reports on these findings but, even when the study of a larger number of patients should be studied to validate this clinical sign, it seems to help the first time diagnostic.

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Abstract — WCN 2013

No: 2691

Topic: 36 — Other Topic

Psychiatric manifestation of adult form of Niemann–Pick type C. A case report

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Background: Niemann–Pick type C (NPC) is a rare neurovisceral lipid storage disorder of autosomal inheritance, caused by abnormal intracellular cholesterol metabolism. In patients with adult form of NPC, psychiatric symptoms may precede neurological symptoms. If patients present with psychiatric manifestation of adult onset NPC, diagnosis is delayed.

Objective: The objective of this case report is to raise awareness of inborn errors of metabolism, in particular NPC, as an organic cause for psychosis.

Patients and methods: We describe the case of a young man, who initially presented with cognitive decline and psychotic symptoms at the age of 21 and was diagnosed with hebephrenic schizophrenia. He did not respond well to neuroleptics. At the age of 27 he was hospitalized at the neurology department due to progressive cognitive decline (MMSE 19/30).

Results: Neurological examination revealed a supranuclear vertical gaze palsy, saccadic eye movements, bradykinesia, dysarthria and ataxia. There was mild atrophy in the MRI of the brain and diffuse slow activity in the EEG. Transaminase levels were elevated, abdominal sonography revealed hepatosplenomegaly. The combination of neurological, visceral and psychiatric symptoms raised the suspicion of NPC. Genetic testing of NPC1 and NPC2 gene was performed and showed homozygosity for a mutation in exon 13 of the NPC1 gene.

Conclusion: NPC may initially present with psychosis in adults. In patients with psychiatric symptoms, in particular if they do not respond to neuroleptics, inborn errors of metabolism should be considered as organic cause of psychiatric manifestations, even in the absence of neurological and visceral symptoms.

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Abstract — WCN 2013

No: 2821

Topic: 36 — Other Topic

Cyclin D1 expression in Schwann cell nucleus associated with the stage of nerve regeneration

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Objective: To investigate the roles of cell cycle-associated proteins in peripheral nerve, we examined the expression of cyclin D1 and pRB using immunohistochemistry.

Background: D-type cyclins are required for the initial step in cell division and translocation into nuclei is crucial for promotion of cell proliferation. Cyclin D1 binds CDK4/6 and regulates cell cycle G1/S

phase transition by a phosphorylation of pRB. In CMT1A, a different mechanism from acquired neuropathy acts to the proliferating process of Schwann cell. Axonal involvement advances in proportion as demyelination and its severity is correlated with clinical symptom. To clarify the role of axon–Schwann cell interaction may be a clue for elucidating the pathogenesis.

Material and methods: Sural nerve biopsies from patients with CMT1A (n = 6), CMT2 (n = 5), CMTX (n = 2), CIDP (n = 6), acute axonal degeneration (n = 6), active nerve remyelination (n = 3) and control subjects (n = 3) were studied by immunohistochemistry. Each patient provided informed consent.

Results: In controls, cyclin D1 immunoreactivity was found weakly in a part of Schwann cell cytoplasm. Actively regenerating nerves showed distinct cyclin D1 expression in cytoplasm of denervated Schwann cell clusters and nuclei. Both cyclin D1 and pRB positive Schwann cell nuclei were found in a part of onion bulb in only early stages of CMT1A.

Conclusion: Cyclin D1 immunoreactivity was identified in Schwann cell nuclei under remyelinating process in early stages of CMT1A. These results seemed similar to axonal neuropathy, and suggested that a certain signal from remyelinating axons is involved in Schwann cell proliferation.

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Abstract — WCN 2013

No: 2504

Topic: 36 — Other Topic

Treatable cause of relapsing encephalopathy

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A 68 year old builder presented with a two month history of slowly progressive impairment of memory, confusion, shuffling gait and urinary incontinence. He gradually became unresponsive. On admission he was abulic and progressed to coma.

Examination: Neck and limbs were rigid. Reflexes were brisk with extensor plantars. Normal funduscopy. Full neurological exam was not possible.

Investigations: ESR, 77 mm CRP 250. CSF 86cells (Poly30% Lym 70%). Protein 1.2 g/l. normal glucose. Blood count and electrolytes were normal. MRI brain showed diffused white matter abnormality bilaterally suggesting leukodystrophy.

Progress: Treated empirically, with IV antibiotics and Acyclovir. He began to improve. Within two weeks his MMSE was 30/30. No focal neurology. Mobilised normally. Discharged home without a definite diagnosis.

Four months later he was readmitted with two weeks history of progressive confusion, shuffling gait and incontinence. He later became comatose. Family revealed that he suffered intermittent deafness and redness of pinna. CSF showed 46 cells, 58% lymphocytes. High proteins and normal glucose. MRI brain was similar to previous one and MRA showed areas of restricted diffusion with contrast enhancement. EEG was diffusely slow.

Brain biopsy showed vasculitis associated with giant cells without granuloma consistent with CNS vasculitis of relapsing polyorchondritis.

Conclusion: CNS vasculitis secondary to relapsing polyorchondritis. Patient was treated with steroids and remained well during the 3 year follow up without further relapse on a low dose of oral steroids.

Discussion: This paper describes a serious neurological complication of a rheumatological condition. Awareness of this association will help early diagnosis and correct treatment.

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Abstract – WCN 2013**No: 2928****Topic: 36 – Other Topic****A novel heteroplasmic mutation in the mitochondrial ATP6 gene associated with Parkinson's syndrome and epilepsy**

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Background: Mutations in the mitochondrial DNA are maternally inherited and may lead to multisystem disorders. A tissue-specific threshold-level of the respective mtDNA-mutation load must be exceeded to give rise to a phenotypic manifestation.

Objective and methods: We report on a 44-year-old male patient with a history of dysarthria and tremor since childhood and epilepsy starting at age 16. In addition, he is of short stature and has mild mental retardation. Previously, diagnosis of Parkinson's disease and multiple sclerosis had been established. At age 42, sensorineural hearing loss was diagnosed. When he was admitted to our hospital, we found that lactate was elevated in blood and CSF. Due to elevated lactate levels, short stature, extrapyramidal symptoms and epilepsy a muscle biopsy was performed. **Results:** A hitherto unreported point mutation in the mitochondrial ATP6 gene (mt.8969G>A) was detected (homoplasmic in the muscle, heteroplasmic in blood: ~80%, urine sediment: ~90%). The detected missense mutation is predictably pathogenic due to the replacement of a highly conserved serine residue by asparagine at position 148 within the fourth transmembrane domain of the ATP synthase F0 subunit 6. This mutation was also detected in the patient's asymptomatic 39-year-old brother, however, with lower heteroplasmy levels (~50% in blood and ~70% in urine sediment, respectively).

Conclusion: We present strong evidence, that a hitherto unreported mtDNA mutation (mt.8969G>A) affecting the ATP synthase F0 subunit 6 of the mitochondrial respiratory chain complex V, leads to the complex clinical presentation of a mitochondrial epilepsy syndrome accompanied by dysarthria, sensorineural hearing loss, and mental retardation.

doi:10.1016/j.jns.2013.07.2437

Abstract – WCN 2013**No: 3006****Topic: 36 – Other Topic****MicroRNA-214 expression in dorsal root ganglion neurons and response to peripheral nerve injury**

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Background: The complicated regenerative process of peripheral nervous system (PNS) can be impacted by many factors. MicroRNAs (miRNA(s), miR(s)) are non-coding RNA molecules that regulate gene expression in a wide variety of biological processes mainly at the post-transcriptional levels.

Objective: This study investigated the possible involvement of miRNAs in gene regulation relevant to peripheral neural regeneration.

Material and methods: A miRNA microarray analysis detected change in 23 miRNAs after sciatic nerve transection (ScNT) among which miR-214 was dramatically reduced. MiR-214 was predicted to target 3'-UTR of Slit-Robo GTPase-activating protein3 (srGAP3), a member of axon guidance pathway. The down-regulation of miR-214 was further

validated using quantitative reverse transcriptase PCR (qRT-PCR). srGAP3 was confirmed to be the target of miR-214 by in situ luciferase report experiment.

Results: In situ hybridization (ISH) experiments using locked nucleic acid (LNA) microRNA detection probe verified that miR-214 was located in primary neurons of DRG and down-regulated following ScNT. Combination of ISH and immunofluorescence revealed that miR-214 and srGAP3 were co-localized in primary DRG neurons. Analysis of western blot suggested that srGAP3 was negatively regulated by miR-214 in DRG neurons after ScNT.

Conclusion: These data demonstrate that miR-214 is located and differentially expressed in primary DRG neurons and may participate in the course of neural regeneration by regulating gene expression of srGAP3 after sciatic nerve transection.

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Abstract – WCN 2013**No: 2611****Topic: 36 – Other Topic****Carpal tunnel syndrome: The possibility of early diagnosis**

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Carpal tunnel syndrome (CTS) is a clinical disorder caused by compression of the median nerve at the wrist. Despite the long history of the disease (over 100 years), there remains the problem of early detection and diagnosis of carpal tunnel syndrome. Many people have classic symptoms without abnormal electrodiagnostic test results.

Purpose: The aim of this study was to develop an algorithm of early diagnosis of carpal tunnel syndrome in the prehospital setting by studying the features of neural conduction fibers of the median nerve in artificial compression.

Methods: 120 wrists of 60 patients were enrolled, along with the same number of age- and gender-matched controls. All patients had nerve conduction study (NCS) and imitation compression test. The patients were divided into three groups. The first group (control group) included healthy subjects (n = 17). The second and third groups (n = 43) are formed with the inclusion criteria, it was the presence of numbness in the finger joints for 3 months.

Results: In patients with carpal tunnel syndrome on the background of additional artificial compression of the nerve there was a significant inhibition of neuronal transport and reduced reserve capacity of the nerve fiber to recovery. At the stage of MCL indicated sufficient stability motor fibers to compression, the most susceptible to compression is sensory fibers.

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Abstract – WCN 2013**No: 3001****Topic: 36 – Other Topic****Use of Rituxan in neurological disease and outcomes in key disease groups**

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Background: This presentation outlines patients with myasthenia gravis, dermatomyositis, neuromyelitis optica, NMDA, encephalitis, vasculitis, and CIDPN who were treated with Rituxan. These diseases are considered treatable and even have the potential to go into remission. However, many patients only improve with multiple therapies – such as steroids, azathioprine, IVIg and plasmapheresis –

on a chronic basis. Rituxan is being increasingly used in order to improve the clinical status in patients who either do not respond or respond but remain dependent on these therapies long term.

Objective: This study is a description of the cases that received Rituxan and the response and lack of response as measured by clinical symptoms and dependence upon other therapies. The outcome measures include improvement to a minimally affected state and withdrawal of other therapies as an indicator of response to therapy and potential remission.

Patients and methods: Forty patients received Rituxan after being either intolerant or poorly responsive to traditional therapies.

Results: Patients with inflammatory myopathies responded significantly to Rituxan, allowing a decrease in other therapies. Patients with myasthenia gravis demonstrated dramatic improvement in clinical symptoms and were able to reach minimal clinical symptomatology and complete remission combined with withdrawal of other therapies, particularly IVIg and plasmapheresis. Little to no response was seen in patients with CIDPN; all patients with this disorder continued previous therapy.

Conclusion: Rituxan can be safely recommended for treatment of inflammatories and myasthenia gravis, however, it is not recommended for the treatment of CIDPN based on our local experience.

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Abstract – WCN 2013

No: 2781

Topic: 36 – Other Topic

Do young European neurologists know enough to comfortably treat patients with neurotrauma?

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Background: Traumatic brain injury (TBI) and spinal cord injury (SCI) are taking together total (in)direct medical costs among the diseases with the highest annual costs (US-40 billion dollars). Despite the clinical and societal impact of TBI, young neurologists lack the opportunities for formal training in this topic.

Objective: We wanted to evaluate motivation, knowledge and obstacles in treating patients with neurotrauma and how to improve the training.

Patients and methods: Sixty neurologists were included (33 women) from 10 central and eastern European countries. Survey consisted of fifteen questions testing both knowledge (nine multiple choice questions) and exploring opinions.

Results: The mean age of the participants was 30.8 ± 5.6 years. The majority (60%) were resident or consultant (up to 5 years of post-academic experience). They had at least one patient with traumatic brain, spine or peripheral nerve injury per week in their daily practice (45%). The majority (63%) didn't have any formal training and 82% was certain that the diagnosis and treatment of traumatic brain injured patients would significantly improve, if neurologists become more involved. In practical test no one achieved 0 or 9 points (mean 4.2 ± 1.6). Almost three-quarters (72%) of participants think they don't have enough knowledge to comfortably treat their patients and 98% of them would

like to get more practical training mostly in form of teaching sessions (37%) or focused workshops (30%).

Conclusions: Our results clearly indicate that young European neurologists need and wish to improve their knowledge in neurotraumatology. Since there is little formal training, teaching/special sessions/courses at European congresses may be first step to improve education in this field.

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Abstract – WCN 2013

No: 3003

Topic: 36 – Other Topic

Sleep disturbances in systemic lupus erythematosus

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Knowledge about sleep complaints in systemic lupus erythematosus (SLE) is limited.

The aim of this study is to characterize sleep complaints in patients with systemic lupus erythematosus.

Method: Prospective study on 42 consecutive patients with SLE (95, 23% women, the mean age 53, 43 ± 22, 69 years). There was a direct interview with a standardized questionnaire which contained demographic characteristics and validated scales.

Main outcome measures included: Pittsburgh Sleep Quality Index (PSQI), Epworth Sleepiness Scale (ESS), EQ-5D, Fatigue Severity Scale (FSS), Lupus QoL questionnaire, Hospital Anxiety and Depression Scale. The Pittsburgh Sleep Quality Index (PSQI) was used to quantify sleep quality. Poorer sleep quality was correlated with higher PSQI scores.

Results: Mean PSQI scores were significantly higher in patients with higher Lupus QoL scores, depression, and fatigue. PSQI had significant correlations with worse QoL ($p < 0.01$). Patients reported difficulties falling asleep (73.8%), reduced subjective sleep quality (88.09%), and disturbed sleep (90.47%).

Conclusions: Sleep disturbances are frequently encountered in SLE patients and are significantly linked to quality of life and depression. A better management of sleep alterations could improve these symptoms.

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Abstract – WCN 2013

No: 3014

Topic: 36 – Other Topic

Quality of sleep in medical students

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Objective: To determine the sleep quality and related factors of the 1st, 4th and 6th year medical students in Pamukkale University, Denizli, Turkey.

Methods: This study is a cross-sectional survey. It was carried out after obtaining an IRB permission. The study group included in the 1st, 4th and 6th year medical students. There were 113 (88.9%) participants from the 1st, 62 (73.8%) from the 4th, and 46 (93.8%) from the 6th year in the study. The questionnaire consists of two parts. The first part had questions on the demographic characteristics of the participants. The Family Welfare Scale was used to determine the socio-economic levels. The second part was consisted of the Pittsburgh Sleep Quality Index (PSQI).

¹ On behalf of the Scientist Panel on Neurotraumatology, European Federation of Neurological Societies (EFNS) and The European Association of Young Neurologists and Trainees (EAYNT).

Findings: The majority of participants are female (51.1%). The mean age is 21.6 ± 2.3 . The mean PSQI score of the students is 8.3 ± 2.9 . Only 14.9% of the student PSQI scores are 5 or under. The sixth year students' sleep quality is the worst (9.3 ± 3.2), however the 4th year students' sleep quality is the best (7.0 ± 2.2) ($p < 0.001$). Sleep quality was the worst in those their mothers' educational level was less than high school (9.0 ± 3.0) ($p = 0.017$). People with chronic disease (9.8 ± 3.5) and smokers (9.9 ± 3.2) were also with poor sleep quality.

Conclusions: Significant numbers of medical students have poor sleep quality. Interventions to improve sleep quality should be planned in the light of the findings of the studies on this issue.

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Abstract – WCN 2013

No: 3038

Topic: 36 – Other Topic

Is it post-malaria neurological syndrome or viral encephalitis?

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Introduction: Post-malaria neurological syndrome (PMNS) is a rare self-limited clinical syndrome in which patients develop neurological and/or psychiatric symptoms within 2 months of a severe falciparum malaria infection, after the parasites have cleared from the peripheral blood.

Case report: A 60-year-old Caucasian man presented with a confusional state associated with ataxia that had started 3 days earlier. On admission, he had a generalized seizure. He had recently returned from Angola, where he had been treated with quinine in an intensive care unit for a severe malaria infection, and later discharged, following full recovery and clearance of parasitemia. He had had no fever during the previous 15 days. CSF analysis revealed lymphocytic pleocytosis ($123 \text{ cel}/\mu\text{L}$) and hyperproteinorrhachia (188 mg/dL). Brain MRI showed no abnormalities. The EEG detected slightly slowed fronto-temporal activity. No evidence of central nervous system (CNS) infection was found, with negative CSF polymerase chain reaction (PCR) testing for common neurotropic viruses and negative CSF and blood cultures. The patient was empirically treated with ceftriaxone and acyclovir, with full symptomatic recovery following the first day of treatment.

Conclusion: We report the case of a patient who had fully recovered from a severe malaria infection and later developed a diffuse encephalopathy. PMNS, usually a self limited entity, should be considered in the differential diagnosis, taking into account the patient's recent history. Nevertheless, the lack of relevant changes on brain MRI and EEG, along with a clinical picture suggestive of viral encephalitis means we shouldn't exclude a false negative CSF PCR viral encephalitis.

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Abstract – WCN 2013

No: 2734

Topic: 36 – Other Topic

Different clinical presentations with similar reversible splenic lesions

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Background: Mild encephalopathy with reversible isolated splenic lesion (MERS) is an occasionally reported clinicoradiological entity. It has been associated with anti-epileptic drugs, viral encephalitis, metabolic disturbances, pre-eclampsia and posterior reversible encephalopathy syndrome. Magnetic Resonance Imaging (MRI) shows restricted diffusion in the splenium of the corpus callosum (SCC), along with hyperintensity on T2/T2-FLAIR weighted imaging, iso/hypointensity on T1 and non-enhancement after gadolinium injection. It is totally reversible, usually disappearing in days to weeks. Rarely, it involves the whole corpus callosum and bilateral white matter. Prognosis is mostly related to the etiology. Few clinical reports describe this imaging finding after cardiac arrest or metronidazole administration.

Objective: Comparison of two cases of reversible isolated restricted diffusion in the SCC with different etiologies and clinical presentations.

Patients and methods: Description of the clinical picture, initial and follow-up brain MRIs of two patients; one presenting with mild encephalopathy after metronidazole administration and another with coma after cardiac arrest.

Results: Both presented reversible restricted diffusion in the SCC. Complete clinical resolution and transient bilateral augmented ADC value on the dentate nucleus were found in one, compatible with metronidazole-induced encephalopathy. The follow-up MRI showed hypoxic-ischemic brain lesions in another with persistent vegetative state.

Conclusion: Restricted diffusion in the SCC seems to be unreliable for prognostic purposes. It can be observed not only in the setting of a mild encephalopathy, but also in the acute stage of a more aggressive brain insult. Only follow-up MRI can identify such an entity as being reversible.

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Abstract – WCN 2013

No: 1666

Topic: 36 – Other Topic

Mitochondrial DNA variations and evidence of haplogroups in Indian Friedreich's ataxia (FRDA) patients

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Objective: This study is aimed to evaluate FRDA patients for mtDNA sequence variations and identify related haplogroup in FRDA patients from Indian population.

Background: FRDA is an early onset autosomal recessive disorder, caused by loss of function of the frataxin protein. Frataxin is required for the biogenesis of the iron-sulphur-cluster (ISC) which is vital for the synthesis of the respiratory chain complexes I-III enzymes and aconitase in mitochondria.

Methods: Considering D loop, NADH dehydrogenase and ATP region as hotspots for the mitochondrial pathogenic mutations in neurodegenerative diseases, these regions were sequenced by direct sequencing method for 30 FRDA patients and 63 ethnically and age matched controls. The FRDA patients were genetically confirmed with southern blot and TP PCR protocols.

Results: 185 variations were observed in all 30 patients. 101 variations were in coding region (ND genes and ATP) while non-coding (D Loop) had 84 variations. Variations at positions 16519, 4883 and 5178 were found significantly associated with patients ($P < 0.05$). Haplogroup analysis using D loop results had revealed that 57% of patients belong to haplogroup M, while control constitutes only 18.6%. Variations at positions 489 and 195 were found in 100% of patients belonging to the M group, while control at these loci were under represented.

Conclusions: Variations at positions 16519, 4883 and 5178 suggest that these variations may act as modifier for the disease. Significant association of haplogroup M with patients suggests possible impact of

haplogroup M encompassing inherited mitochondrial DNA variations on predisposition to FRDA patients of Indian ethnicity.

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Abstract – WCN 2013

No: 3022

Topic: 36 – Other Topic

Neuroimaging study in 210 Iranian neurometabolic patients

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Inborn error of metabolism (IEM) involving CNS encompasses a wide spectrum of inherited disorders.

We present and discuss imaging features of the more common and important IEMs involving CNS that diagnosed molecularly in Iran.

Our pattern approach for neuroimaging classification of Common Metabolic Disorders was according to anatomic evaluation:

White matter (WM)

- Primarily affecting periventricular
- Primarily affecting subcortical

Gray matter (GM)

- Primarily affecting cortical
- Primarily affecting deep

Disorders involving WM and GM

During 3 past years, we studied systematically neuroimages of our patients and finally we classified our cases pre- and post-molecular diagnosis as below:

Initially affecting periventricular WM

Metachromatic leukodystrophy (12 cases)

Krabbe disease (4 cases)

X-linked adrenoleukodystrophy (6 cases)

Phenylketonuria (21 cases)

Maple syrup urine disease (18 cases)

Homocystinuria (7 cases)

Merosin-deficient congenital muscular dystrophy (10 cases)

Initially affecting subcortical WM

Canavan disease (8 cases)

Alexander disease (5 cases)

Lack of myelination

Pelizaeus–Merzbacher disease (5 cases)

Cortical gray matter

Neuronal ceroid lipofuscinoses (9 cases)

Mucopolysaccharidosis type I (5 case)

Deep gray matter

Leigh disease (13 cases)

Organic acidopathies (31 cases)

Cortical gray matter only

- Cortical dysplasia:
- Walker-Warburg syndrome (1 case)
- Muscle-eye-brain disease (1 case)
- No cortical dysplasia
- Menkes disease (4 cases)

Deep gray matter involvement

- Primarily affecting thalamus:
- Krabbe disease (8 cases)
- GM1 or GM2 gangliosidoses (18 cases)
- Wilson disease (6 cases)

Deep gray matter involvement

- Primarily affecting globus pallidus:
- Canavan disease (2 cases)
- Kearn–Sayre disease-subcortical U fiber (4 cases)
- Methylmalonic acidemia-deep WM (8 cases)
- Maple syrup urine disease-dorsal brainstem (18 cases)
- Primarily affecting striatum:
- Leigh disease (13 cases)
- MELAS (10 cases)
- Glutaric aciduria type I (6 cases)
- Wilson disease (6 cases)

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Abstract – WCN 2013

No: 1479

Topic: 36 – Other Topic

Levodopa-responsive parkinsonism and autonomic (small fiber) dysfunction in patients with Wilson's disease (WD)

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Background: Autonomic dysfunction has been rarely described in WD patients.

Objective: To describe a series of patients with WD, levodopa-responsive parkinsonism and autonomic dysfunction.

Methods: After IRB approval, we evaluated 4 patients with WD and parkinsonism for the presence of neuromuscular dysfunction and performed water-induced skin wrinkling test – SWT (Teoh, JNNP 2008;79:835).

Results:

Patient 1: A 37 year old female presented with cirrhosis at age 33 was diagnosed with WD and treated with ZnSO₄, penicillamine and pyridoxine/complex B vitamins. Since age 35 she developed pramipexole and levodopa-responsive parkinsonism, with burning and numbness in arms and genital area and urinary incontinence. NCS/EMG was normal. Water-induced SWT revealed small fiber dysfunction (mean 4-digit wrinkling of 1).

Patient 2: A 23 year old male was diagnosed with WD at age 14 and was treated with penicillamine + pyridoxine. Since age 21, he developed levodopa-responsive parkinsonism, with resting tremor, weight loss, dysphagia and dysphonia. NCS/EMG was normal but water-induced SWT was abnormal (mean wrinkling of 0.25).

Patient 3: A 34 year old male presented with psychosis, behavioral dysfunction, levodopa-responsive parkinsonism and episodes of loss of consciousness. He was treated with penicillamine and complex B vitamins. NCS/EMG and water-induced SWT were normal (mean wrinkling of 4).

Patient 4: A 38 year old female presented with liver dysfunction, behavior changes and parkinsonism at age 24. She was diagnosed with WD and treated with penicillamine and biperiden. Parkinsonism almost completely subsided with penicillamine. SWT was normal (mean wrinkling of 3.6).

Conclusions: A subset of WD patients with levodopa-responsive parkinsonism also exhibits abnormalities on SWT (autonomic dysfunction).

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Abstract – WCN 2013**No: 2583****Topic: 36 – Other Topic****Sleep disorders in HIV–AIDS patients in Cameroon, sub-Saharan Africa**

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Background: Sleep disorders have been reported to be more common among people living with HIV–AIDS (PLWHA) than matched controls in Western countries, but few data exist on the subject in Africa. In Cameroon, no study has been done on the prevalence and characteristics of sleep disorders in PLWHA, the object of this study.

Patients and methods: A case–control study including 39 PLWHA age- and sex-matched with 43 HIV negative controls. We used the Pittsburgh Sleep Quality Index (PSQI) to assess sleep quality, the Epworth Sleepiness Scale (ESS) for daytime sleepiness and the Berlin Questionnaire (BQ) for obstructive sleep apnea (OSA).

Results: HIV + participants had poorer sleep quality than the controls: 66.7% versus 11.6%, (AOR 20.60; 95% CI, 2.98–142.19) and took significantly longer to fall asleep (33.08 ± 27.35 min versus 20.81 ± 16.61 min, $p = 0.015$). Poor sleep quality was significantly more common in HIV₁ cases compared to HIV₂ (100.0 versus 0.0% respectively, $p = 0.001$). No significant differences were observed with respect to age at HIV diagnosis, disease duration, CD4 count, disease (WHO) stage, being on HAART or not, 1st or 2nd line treatment or being on Efavirenz. Daytime sleepiness and OSA were significantly more common in the cases compared to the controls: 23.1% versus 2.3% ($p < 0.001$) and 43.6% versus 14.0%, ($p = 0.003$) respectively.

Conclusion: Sleep disorders are common in Cameroonian HIV-positive patients. The use of sleep scales in the clinical assessment of patients could contribute to better management and improves quality of life.

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Abstract – WCN 2013**No: 2966****Topic: 36 – Other Topic****Elderly patients in vegetative state and minimally conscious state**

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Background: Current epidemiological and clinical information related to vegetative state (VS) and minimally conscious state (MCS) are mainly based on adult patients, however there is limited research on ageing with disorder of consciousness (DOC) and on elderly people with DOC.

Objective: The aim of this presentation is to report clinical and demographic data on patients with DOC above 65 years.

Patients and methods: The multicentric national projects “Functioning and disability in VS and MCS” and PRECIOUS “Taking care of people with acquired severe brain injury” collected information on a large sample of people with DOC in 107 post-acute rehabilitation and long term Italian institutions.

Results: Among 600 participants in VS and MCS, 201 (33.4%) were aged above 65 (mean = 74.0; DS = 6.44; range = 65–93). The majority of the elderly sample were male (55.7%), in VS (72.2%) due to a non-traumatic aetiology (84.1%), hosted in long term care facilities (78.6%) and with a mean disease duration equal to 2.5 (DS = 2.1; range = 0.1–11). The Disability Rating Scale indicated severe levels of disability

(mean = 22.7; SD = 1.2) and no differences of severity of the disease were found between age groups. Within 2 years, 111 (55.2%) patients, mainly non-traumatic ($n = 87$), died.

Conclusions: These results indicated a high prevalence of elderly non-traumatic patients in VS with complex disability outcomes and high needs in terms of rehabilitation. Similar outcomes and findings are found in the data collected in the Coma Research Centre of Neurological Institute Carlo Besta IRCCS Foundation that undertakes an in depth evaluation of patients with DOC.

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Abstract – WCN 2013**No: 2967****Topic: 36 – Other Topic****Pathways of care of Italian patients in vegetative state and minimally conscious state: Results from a national study**

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Evidence has described heterogeneous pathways of taking care of patients with disorders of consciousness (DOC) both at the national and local level, with inter- and intra-regional differences.

The aim is to collect data on the clinical condition and pathways of taking care of a sample of patients in Vegetative State (VS) and Minimally Conscious State (MCS) in Italy.

Based on the national projects “Functioning and disability in VS and MCS” (T0), the follow-up PRECIOUS “Taking care of people with acquired severe brain injury” (T1) collected information on a sample of patients with DOC in 107 post-acute rehabilitation and long term institutions, located in the North, Central and South of Italy.

Out of 600 patients collected in T0, 587 patients were followed up. After two years, 380 (64.7%) were alive and 109 of them (28.7%) changed place of care. About them, 22% changed region. Movements to rehabilitation centres range from 3% (South) to 7% (North). 51.3% of patients hosted in northern structures moved to a long term care and 41.0% to domicile. In the Centre, 69.6% went to domicile and 26.1% to long term care. In the South, instead, 82.8% moved to domicile and 13.8% to a long term care. As Italian Ministry of Health recommends, equal opportunities of care should be provided to all patients, wherever they live in the country. The high levels of people at home in centre and south Italy, where also home care is inadequately provided, represent a lack in the health and social systems.

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Abstract – WCN 2013**No: 2969****Topic: 36 – Other Topic****Italian study on functioning and disability of people with disorder of consciousness**

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Background: The vegetative state (VS) and minimally conscious state (MCS) are neurological conditions in which patients appear to be awake but show no or partial signs of awareness of themselves or of their environment. Patients require adequate health pathways but the lack of epidemiological information creates difficulty in the organisation of medical care and treatments.

Objective: The aim is to describe the results of a National Italian Research that collected epidemiological and clinical data of adults and children in VS and MCS and to highlight issues related to management, treatment and care.

Material and methods: This was an observational cross-sectional study, conducted in 69 Italian centres specialized in post-acute rehabilitation treatment or in long-term care of patient with disorder of consciousness (DOC). Data collection was performed between June 2009 and March 2010. An ad hoc protocol was used collecting sociodemographic, clinical and functional data.

Results: 600 patients were enrolled (69.7% in VS). 64.3% of cases had non-traumatic aetiology. Patients in VS and MCS were similar for age at acute event and at enrolment (>50 years old). Disease duration was similar for both conditions and 2.6% of VS and 4.8% of MCS patients survived for more than 10 years. Mean number of drugs per adult patient was four and decrease with increased disease duration.

Conclusion: This study reports data based on a large sample of patients with DOC. Results may be useful to analyse health and social issues on people in VS and MCS that need a large amount of environmental facilitators.

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Abstract – WCN 2013

No: 2971

Topic: 36 – Other Topic

Horizontal epidemiology approach: Functioning and disability in patients with neurological disorders

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Neurological disorders (ND) lack comprehensive understanding of psychosocial difficulties (PSDs) and determinants. PSDs are defined as impairments, activity limitations, participation restrictions and environmental factors.

This study aimed to identify PSDs that a person with ND experience in everyday life and their determinants according to the framework of horizontal epidemiology.

400 adult patients with stroke, epilepsy, Parkinson's disease, migraine and multiple sclerosis were interviewed using the PARADISE protocol to collect self-reported PSDs associated to brain disorders (www.paradiseproject.eu). PSDs reported by at least 45% of the patients in all the conditions are described below.

Mean age: 49.6 years (SD 11.5); 57.2% females; 68.1% married; 67.6% with at least high school education; and 53% working. Neurological patients mostly report PSDs related to restlessness (71.2–95.0%), sleeping problems (46.2–71.2%), being emotionally affected by health condition (66.2–88.7%), anxiety (61.2–87.5%), depressive symptoms (61.2–81.2%), coping functions (55.0–81.2%), pain (48.7–93.7%), and daily activities (45.0–68.7%). 52.1% of patients rated their health at least as good. Main determinants acting as moderate or strong environmental facilitators were family (92.1%), health professionals (90.6%), and medicines (94.8%), despite 54.0% of the sample reporting side-effects of medicines as environmental barriers. More than half of the sample (57.5–78.7%) reported that they felt to be a different person after the beginning of the disease/acute event.

Perceived PSDs and determinants that contribute to the course of PSDs were identified, suggesting that it is important to consider not only

impairments and symptoms but also environment's impact on health and disability to plan tailored intervention for person with ND.

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Abstract – WCN 2013

No: 2972

Topic: 36 – Other Topic

Disability, quality of life and well-being in patients undergoing brain surgery

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Background: The assessment of quality of life (QoL) has become an important endpoint in clinical trials in brain tumours and cerebrovascular diseases, helping clinicians in providing the best and most complete treatment and care.

Objective: This study aimed to evaluate disability, QoL and well-being in patients undergoing brain surgery at National Neurological Institute Besta.

Patients and methods: Adult patients with brain tumour and cerebrovascular diseases undergoing neurosurgery from May 2012 to February 2013 were enrolled and interviewed using a research protocol composed of evaluation of disability levels (WHODAS-12), self-perception of well-being (PGWB-S) and quality of life (EUROHIS-QoL).

Results: Eighty seven adult patients were included: mean age was 51 years, 55% were females, 49% has high school education or higher, 63% has a tumour and 37% cerebrovascular disease. On WHODAS-12, where epilepsy patients rank 13.6 and stroke patients 25.9, the levels of mean score of our sample were 22.9, suggesting high levels of disability compared to Italian population. Patients reported lower levels of well-being (63) when comparing with the general population (69.7). QoL was rated as slightly lower (3.43) than the general population (3.68).

Conclusions: This study shows that patients undergoing brain surgery present high disability levels, low levels of well-being and QoL. In this sense, data on functioning and disability and the impact of environment in which person lives should be considered for a prognostic evaluation of patients and are necessary to define tailored and personalised treatments.

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Abstract – WCN 2013

No: 2979

Topic: 36 – Other Topic

Neurological diseases of ageing people with Down syndrome: A review from 1960 to 2011

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Background: The scientific literature reports that people with Down syndrome (DS) start aging prematurely so that they can be considered old people at the age of 45.

Objective: To highlight the evidences of the scientific literature on neurological diseases of ageing people with DS.

Methods: Our review was conducted through two electronic databases: PUBMED and ScienceDirect covering the period from 1960 to 2011.

Results: In all studies the prevalence of Alzheimer's increases with age: from 30 to 39 years is 10%, from 40 to 49 years is 25%, from 50 to

55 years is 28% and after 60 years is 75%. The average age of onset is 57 years. The Dementia Alzheimer's type in people with DS is characterized by: changes in behaviour and personality, decline in cognitive functions, psychological changes, ataxia, seizures, urinary and fecal incontinence. Alzheimer's and Parkinson's disease with severe dementia occurring in about 15% of patients with DS aged over 40. 26% of adults with DS suffer from epilepsy and the average onset is 37 years. **Conclusion:** Due to early ageing, Alzheimer's and epilepsy in people with DS appear before and more frequently than in general population. So it is appropriate that we share more awareness about adult DS health needs while they age. It is also necessary to combine the medical evaluation of disorders presenting in people with DS due to ageing with the relevant information about social and personal life, according to a broader biopsychosocial perspective.

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Abstract – WCN 2013

No: 2959

Topic: 36 – Other Topic

Neuro expressionism: Contributions at the beginning of the 20th century

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The term expressionism is used to describe any art that raises subjective feelings above objective observations. Affected by the anxieties of accelerated social change, expressionist artists employed violent exaggerations and distortions of form and colour. Austrian expressionist artist Egon Schiele (1890–1918) made exhibitionism and persecution-mania in a dystonic way with stronger linear effects and harsher outlines, one of his major themes. Arthur Schüller (1874–1957), the father of neuroradiology, was born in Vienna. In 1905 his first book, *The skull base on the radiogram*, was published and expressionism developed almost simultaneously in different countries. It was a comprehensive description of normal and pathologic anatomy as well as of many special radioprojections, like Schiele's linear effects, of the skull base. Heinrich Quincke (1842–1922) who should be credited with the discovery of lumbar puncture, performed research in many fields of medicine, including internal medicine, surgery, paediatrics, neurology and dermatology. Christfried Jakob (1866–1956) a great neuropathologist, neurobiologist, philosopher, professor and artist was born in Germany and lived and worked for 57 years in Argentina. Otto Klemperer (1885–1973). First as revolutionary ground-breaker, later as champion of the classics from Bach to Mahler, amassed a recording legacy of masterworks, unequalled in their vision, depth, and power. Does neurology express art or art express neurology?

Did the neuroexpressionism contribute with the beginning of the age of globalization?

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Abstract – WCN 2013

No: 2964

Topic: 36 – Other Topic

A case of Pancoast tumor: Unusual presentation and successful therapy

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Background: The Pancoast syndrome (PS) presents with a variety of clinical manifestations. Neurological symptoms are pain, radicular sensory and motor syndromes, and Horner's syndrome. PS often bears a grim prognosis.

Patients and methods: A 45 year old lady came for neurologic evaluation, due to pain radiating into her right elbow for nine months and numbness in her 5th finger. Sensory loss in her fourth and fifth fingers resulted in clumsiness in her right hand. She presented right sided Horner's syndrome and a decreased triceps jerk; the right hand had a mild atrophy of the small hand muscles. NCV and EMG studies were not conclusive. CT-scan and MRI of the lung and brachial plexus revealed a large mass of the lung, infiltrating into the brachial plexus.

Results: After preoperative chemo- and radiotherapy the tumor could be surgically removed. Histologically, the nerve fascicles were embedded in tumor tissue, which also spread along the affected fascicles. Postoperatively she suffered from severe neuropathic pain, which could be successfully managed. The right hand has only crude sensory perception left and loss of function of lower arm and hand muscles.

Conclusion: This case report describes an atypical onset of lung tumor causing a PS. NCS were not conclusive. The diagnosis was confirmed by MRI, CT-scan and biopsy. Despite considerable loss of distally sensorimotor function of the right hand, the patient is able to use the extremity in her professional life. This observation demonstrates the successful management of lesions of the peripheral nervous system caused by cancer.

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Abstract – WCN 2013

No: 1828

Topic: 36 – Other Topic

Pathogenesis of the hemifacial hyperhidrosis

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Background: Facial hyperhidrosis may be often compensatory hyperhidrosis due to the contralateral hemifacial anhidrosis, but little is known on the pathogenesis.

Objective, patients and methods: We analyzed it for 14 patients with this symptom by defining the total body sweating patterns using Minor's starch-iodine test, total skin temperature distribution using infrared thermography, local skin blood flow using laser Doppler flow meter, and MRI to confirm the lesions.

Results: The sweating patterns were classified as follows: I. Systemic unilateral hyperhidrosis (4 patients from 41 to 68 years old): might be compensation due to the contralateral systemic unilateral anhidrosis. This might be caused by cervical disk herniation, protruding to a spinal median, which might induce minor circulatory disturbance of the central artery of the spinal cord on the compressed side; and II. Segmental unilateral hyperhidrosis (10 patients).

- Hyperhidrosis and flushing (harlequin syndrome) from hemifacial to the ipsilateral cervical innervation area during body heating (1 y 9 m, 2 y 1 m, 5 y 6 m, and 9 y male): might be compensation due to the contralateral anhidrosis caused by injury of cervical sympathetic trunk preganglionic fibers. Since cervical traction side at delivery corresponded with the disorder side, obstetric traumatism might contribute to the etiology;
- Segmental well demarcated hyperhidrosis along the cervical innervated areas (63, 74, and 37-year-old patient): might be compensation due to the anhidrosis of the adjacent lower area. They had cervical disk herniation protruding at 3 mm lateral from the median;
- Other symptomatic segmental hyperhidrosis.

Conclusion: Defining sweat distribution is prerequisite for elucidating the pathogenesis and treatment of hemifacial hyperhidrosis.

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Abstract – WCN 2013

No: 2973

Topic: 36 – Other Topic

7-Tesla MRI reveals no structural lesions of the vestibulocochlear nerve in patients with vestibular paroxysmia (VP)

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Introduction: Vestibular paroxysmia is a rare entity causing recurrent attacks of vertigo. A neurovascular cross-compression (NVCC) is assumed to be responsible for the symptoms. In conventional MRI the contact between the nerve and the artery can be best shown on CISS-sequences. After years of symptoms a structural lesion in the nerve may be assumed, which has to date not yet been confirmed. The aim of the current study is to investigate whether a pathologic enhancement in the vestibulocochlear nerve following NVCC can be shown by means of 7-Tesla-MRI.

Patients and methods: Six patients with chronic VP were included in this study. In all patients the diagnosis was established according to the clinical symptoms, the finding of a NVCC in MRI (1.5 or 3 T) and the cessation of symptoms after the administration of gabapentine or carbamazepine.

Results: All patients exhibited a NVCC on 7-Tesla-MRI, confirming the findings of conventional MRI. None of the patients showed an enhancement in the vestibulocochlear nerve at the site of the NVCC in T-Tesla-MRI.

Discussion: In all our patients the results of the CISS sequence in 1.5 T or 3 T MRI could be confirmed by FLAIR sequences in 7 T MRI. None of the probands showed pathological enhancement or a structural lesion in the vestibulocochlear nerve. The findings imply that the symptoms of VP cannot be understood as vestibular hypofunction due to structural pathologies but rather as the result of sensory stimulation of the 8th nerve.

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Abstract – WCN 2013

No: 1650

Topic: 36 – Other Topic

Posterior reversible vasoconstriction syndrome in congenital disorders of glycosylation due to a defect in Conserved Oligomeric Golgi complex (COG4)

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Background: Congenital disorders of glycosylation (CDGs) are a rare group of metabolic diseases that have been described in only a few individuals, where phenotypes are continually being updated. Some of these disorders present hemorrhagic or prothrombotic features, but there is not a clear correlation with each individual subtype.

Objective: The authors describe a patient with CDG that presented a posterior reversible vasoconstriction syndrome (PRES).

Patients and methods: An 11-year-old male, diagnosis of CDG at 2 years of age, due to a Conserved Oligomeric Golgi (COG)4 deficiency (identified p.R729W missense mutation) with psychomotor developmental delay, hyperactivity and poor language skills. Admitted with

fever, anorexia and increased inflammatory biomarkers; 3 days later presented a sleep/wakefulness pattern change, severe insomnia and visual hallucinations. A slight increase in the blood pressure profile was seen and some hours later a bilateral amaurosis was installed. Electroencephalogram revealed bi-occipital epileptiform activity. CT brain scan disclosed bilateral parieto-occipital hypodense lesions, with erasing cortical sulci, suggestive of PRES, with a restricted diffusion pattern on the MRI. Transcranial ultrasound was negative for vasospasm. He was started on corticotherapy and levetiracetam with recovery of visual acuity. All the remaining study was normal including coagulation factors and causes of secondary hypertension.

Discussion: CDGs have multisystemic manifestations and phenotypic presentation is in constant renewal. In the case described above, the difficulty of the diagnosis in a non-cooperative patient is added.

Conclusion: The authors describe the first PRES described in these groups of patients, raising the question of a possible relationship between CDGs and PRES.

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Abstract – WCN 2013

No: 2913

Topic: 36 – Other Topic

Sonography – A helpful method in diagnosis of carpal tunnel syndrome

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Carpal tunnel syndrome is the most common entrapment neuropathy of the upper extremity. Usually physical examination and nerve conduction velocity studies are used to confirm the diagnosis. In the last years ultrasound has gained wide acceptance for imaging of peripheral nerve diseases.

Our objective was to evaluate the wrist-to-forearm ratio as an imaging method for the diagnosis of carpal tunnel syndrome.

Patients with electrodiagnostically proven carpal tunnel syndrome underwent an ultrasonographic examination of the median nerve at the wrist and forearm. The cross-sectional area of the median nerve was measured at these points and used for calculating the wrist-to-forearm ratio. The results were compared with the values from healthy volunteers. The average cross-sectional area of the median nerve at the proximal carpal tunnel was $13.61 \pm 3.05 \text{ mm}^2$ in patients presenting with carpal tunnel syndrome and $7.77 \text{ mm}^2 \pm 1.64 \text{ mm}^2$ in healthy volunteers. At the forearm the average cross-sectional area of the median nerve was $6.98 \pm 0.92 \text{ mm}^2$ in patients and 7.34 ± 1.47 in the control group. The calculated wrist-to-forearm ratio was 1.95 ± 0.34 in patients and 1.06 ± 0.06 in healthy volunteers.

We were able to prove that using a wrist-to-forearm ratio of 1.4 gives a 100% sensitivity for detecting patients with carpal tunnel syndrome. Calculating the wrist-to-forearm ratio is a valid imaging method and is from our point of view superior to measuring the cross-sectional area of the median nerve at the wrist alone.

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Abstract – WCN 2013

No: 2668

Topic: 36 – Other Topic

Initiation of regular physical activity promotes quality of life in healthy elderly people

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Background: Healthy ageing is of great personal and economical interest for both the individual and the society. Known favourable prognostic factors are high physical, intellectual and spiritual activity during lifetime, high socioeconomic status and Mediterranean diet.

Objectives: We investigated the effect of the initiation of a regular physical activity in a cohort of non-demented senior citizens on quality of life, memory functions, and physical performance.

Patients and methods: 71 participants were randomised into a training and a control group. Structured physical training, comprising of 3 one-hour units of aerobic exercise per week including balance and strength training over a period of 3 months, was conducted in a collaborating fitness centre. The examinations before and after the 3-month period included: neurological examination, assessment of adverse events, stance and gait testing, neuropsychological testing (emphasis on working memory and executive functions), and answering of questionnaires (Wellbeing Five, SF-36, FOSQ, GDS).

Results: Age, sex, vascular risk factors, and baseline values of outcome parameters were equally distributed between the groups. The training group showed an improvement in quality of life measures (Wellbeing Five, $p = 0.005$), sleep quality ($p = 0.041$), gait velocity ($p = 0.046$) and mental speed ($p = 0.049$) compared to the control group.

Conclusions: Initiation of a regular physical activity promotes emotional, physical and mental wellbeing, as could be measured even after a short period of three months. Long-term effects and imaging measures of brain plasticity will be investigated in the on-going 12-month extension phase (until June 2013).

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Abstract – WCN 2013

No: 2930

Topic: 36 – Other Topic

Serum glucose adjusted cut-off values for cerebrospinal fluid/serum glucose ratio

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Background: Calculation of the cerebrospinal fluid/serum glucose (CSF/S_{Glu}) ratio is part of the routine CSF work-up, however, different cut-off values ranging from 0.3 to 0.5 have been suggested so far to distinguish physiological from pathological conditions.

Objective: To determine normal cut-off values of the CSF/S_{Glu} ratio.

Patients and methods: As an Austrian reference laboratory we screened our database for paired CSF and serum samples, which have been collected by lumbar puncture, were processed within 1 h after withdrawal, showed clear supernatant, cell count < 5/μl, erythrocyte count < 500/μl, total protein < 55 mg/dl, and albumin quotient < 8 and were considered as normal by the evaluating neurologist resulting in 318 sample pairs. Glucose concentrations in CSF and serum were measured by enzymatic spectrophotometry.

Results: Glucose concentrations in CSF were approximately 60% of those in serum. CSF/S_{Glu} ratios significantly correlated with glucose levels in CSF ($R = 0.225$, $p < 0.001$) and serum ($R = -0.572$, $p < 0.001$). For serum samples with a glucose concentration of < 100 mg/dl (median 85), 100–149 mg/dl (median 115) and ≥ 150 mg/dl (median 188) the 5 percentile of CSF/S_{Glu} ratio was 0.52, 0.43 and 0.28.

Conclusion: CSF/S_{Glu} ratio inversely correlates with serum glucose concentrations in a non-linear manner. These findings suggest that cut-off values for CSF/S_{Glu} ratio distinguishing normal from pathological conditions must be adjusted to serum glucose levels, and, furthermore, that an active mechanism is responsible for glucose transport from blood into CSF.

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Abstract – WCN 2013

No: 2921

Topic: 36 – Other Topic

Psychosocial factors relevant to brain disorders in Europe (PARADISE)

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Introduction: The EC funded project PARADISE was conducted in terms of the theoretical conceptualization of “horizontal epidemiology”, that is that the psychosocial difficulties (PSDs) people live with when they have a brain disorder – either psychiatric or neurological – and the determinants of those PSDs are common across disorders.

Objective: To develop and test as a proof of concept an innovative approach to collect clinical and epidemiological data on PSDs based on horizontal epidemiology. The specific aims are:

- 1) to determine the PSDs and determinants that are common across brain disorders;
- 2) to develop a metric to assess PSDs and their determinants across brain disorders.

Methods: For aim

- 1) literature reviews and focus groups in 9 brain disorders (dementia, depression, epilepsy, migraine, multiple sclerosis, Parkinson's Disease, schizophrenia, stroke and substance use disorders) as well as data analyses of surveys and expert consultations were carried out. For aim
- 2) a cross-sectional study with 720 patients across Europe was conducted. Data was analysed by means of Rasch analyses and Random Forest regression.

Results: 24 PSDs common across brain disorders and covering the complete severity continuum of PSDs made up the PSD metric. 11 determinants of the extent of difficulty were identified.

Conclusions: PARADISE is a proof of concept of the hypothesis of horizontal epidemiology. The developed metric to assess PSDs across brain disorders points to a novel approach to data collection that can profoundly change how we collect epidemiological data on the burden of brain disorders and assess the effectiveness of interventions.

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Abstract – WCN 2013

No: 2917

Topic: 36 – Other Topic

Ventricular biopsy for early diagnosis in tuberculous meningitis

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Background: Tuberculous meningitis (TBM), a diagnostic challenge because of inconsistent clinical presentation and also lack of rapid, sensitive and specific tests. The mortality is less than 20% in early stage and approximately 70% in late stage.

Objective: The definitive criteria include biopsy of brain with clinical features of TBM. Getting an accurate diagnosis in the form of tissue biopsy is another challenge. Here we are presenting a case of tuberculous meningitis confirmed by neuroendoscopic ventricular examination and ventricular biopsy.

Case presentation: A 17 year old lady presented 4 months history of fever, weight loss and meningism; she had inconclusive lumbar puncture and was started on anti-tuberculosis treatment empirically. She continued to deteriorate and presented to the neurosurgery team with low consciousness, hydrocephalous with trapped 4th ventricle. She underwent neuroendoscopy for VP shunt and during the procedure ventricular examination and biopsy of the ventricular wall. The histopathological examination confirmed the diagnosis; she had recovered well post procedure and the anti-tuberculosis treatment was modified as she developed abnormal liver function test with deranged INR.

Conclusion: This is an interesting case where neuroendoscopy has helped to confirm diagnosis of tuberculous meningitis. Literature searches reveal case series of neuroendoscopy procedure done to treat tuberculous meningitis with hydrocephalous in late stage but this is one of the few cases in the literature where ventricular biopsy had been used to yield early diagnosis of tuberculous meningitis. This procedure has so far ZERO mortality on procedure risk and had been performed as a day procedure.

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Abstract – WCN 2013

No: 2923

Topic: 36 – Other Topic

Reduced expression of hippocampal BDNF, NGF and their cognate receptors in postmortem brain of suicide victims

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Background: The role of neurotrophins in directing brain growth and neuronal functioning is being increasingly recognized. Neurotrophins not only play an important role in cellular proliferation, migration, and phenotypic differentiation and/or maintenance in the developing central nervous system but also their presence is required in the adult CNS for maintenance of neuronal functions, structural integrity of neurons, and neurogenesis, which suggests that neurotrophins are biologically significant over the entire lifespan.

Objectives: Investigation of expressions of BDNF, NGF with their respective receptors and few downstream signaling molecules in the hippocampus of the suicide victims.

Materials and methods: These studies were performed in the hippocampus obtained from 20 suicide and 20 non-psychiatric control subjects. The neurotrophin levels were quantitatively measured through Sandwich ELISA, the expression of neurotrophin receptors and the downstream signaling molecules were determined by Western Blot. Finally mRNA levels of BDNF, TrkB and NGF, TrkA were determined by RT PCR.

Results: A unique parallel decrease of the protein levels of BDNF, NGF and their cognate receptors in post-mortem suicide victims clearly correlated with the significant reduction of their mRNA levels at the hippocampal regions compared to the normal individuals. A marked decrease in the expression profiles of downstream signaling molecules like PLC γ , PKC δ , ERK 1, 2, Akt and CREB of suicide victims compared to non-psychiatric controls.

Conclusion: Significant reduction of BDNF, NGF and their cognate receptors along with the dysregulation in BDNF/TrkB and NGF/TrkA signaling cascades in post-mortem brains might be of relevance to its pathophysiology of depression.

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Abstract – WCN 2013

No: 2927

Topic: 36 – Other Topic

Symptoms and risk factors associated with carpal tunnel syndrome in east of Turkey: Evaluation of 767 cases

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Carpal tunnel syndrome (CTS) is the most common compression neuropathy with various etiologic factors and symptoms.

In this study, risk factors and symptoms associated with CTS diagnosed cases were determined in our electrophysiology laboratory.

The study involved 767 cases of suspected CTS who were examined by ENMG. Of these 97 (12.6 %) were male and 670 (87.4%) were female. The mean age of patients was 43. Twenty-eight (3.7%) of the cases were left handed and the remaining 739 (96.3%) were right handed. Of the cases 179 (23.4%) had complaints in the right hand, 110 (14.3%) in the left hand and 478 (62.3%) in both. The proportion of symptoms that developed during the night was 74.6%. The common symptoms were numbness (78%), weakness (75%) and pain in the shoulder and arm (77%). Examinations using the Tinel, Phalen and pressure tests revealed positive in 65%, 63.5%, and 72% of patients, respectively. The proportion of CTS patients having other complaints was 35% obesity, 12% DM, 6% hypothyroidism, 1% RA, and 0.5% acromegaly. Of the patients 14% were smokers. Female patients with CTS also had pregnancy (19%), nursing (16%), menopause (31%), and premenopause (23%). Of the women with CTS 89% were housewives and of these 49% were making handicraft and the remaining 21% were agriculture and animal rearing. CTS was diagnosed in 93% of these patients on ENMG examination. CTS is common in women, obese, and those making handicrafts. Animal rearing is another etiological factor.

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Abstract – WCN 2013

No: 2929

Topic: 36 – Other Topic

Hearing outcome in children after non-polio enteroviral meningitis

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Background: Long-term sequelae of meningitis including hearing impairments are concerns to parents and physicians alike. Hearing assessment of children, while useful, is logistically challenging requiring great skill and the frequent need for sedation. It is strongly recommended following bacterial meningitis in view of the significant risk for hearing loss. The situation is less clear with viral meningitis.

Aims: We reviewed outcomes for hearing in children following non-polio enteroviral meningitis to determine the utility of routine post-enteroviral meningitis hearing screening.

Methods: This is a retrospective records review of children <16 years with a cerebrospinal fluid polymerase chain reaction (PCR)-confirmed diagnosis of non-polio enteroviral meningitis who had undergone

hearing screen at 8–10 weeks utilizing otoacoustic emissions (OAE). Excluded were children who had concomitant bacterial infections. Children who failed the hearing screen or had caregiver concerns about hearing or language development underwent formal audiology evaluation. Data was collected on gestational maturity, Gentamicin administration and hyperbilirubinemia.

Results: We enrolled 103 children, 54 males and 49 females, aged 3 days to 16 years. Eighty-three (80.5%) were chronologically younger than 90 days. Eighty-four (81.5%) received Gentamicin at 5–7.5 mg/kg/day for a median duration of two days. Eleven (10.6%) were premature; none had significant hyperbilirubinemia. Eighteen children (17.5%), 16 younger than one year, failed the initial hearing screen. Formal audiology testing of these 18 children revealed no hearing loss.

Conclusions: Outcome for hearing in children after non-polio enteroviral meningitis is good. Routine objective hearing screening may not be indicated following enteroviral meningitis.

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Abstract – WCN 2013

No: 3123

Topic: 36 – Other Topic

Actigraphy in the assessment of sleep patterns in sickle cell disease patients in Cameroon (Sub-Saharan Africa)

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Background: Sleep disorders have been reported to be more common in sickle cell disease (SCD) patients than controls in Western countries, but almost no data exist on the subject in Africa. This study sought to characterize sleep patterns in SCD patients using actigraphy, a validated and user-friendly sleep study tool.

Patients and methods: 13 SCD children and 13 age and sex-matched controls wore wrist actigraphs for a minimum of 72 h and kept a sleep diary. Actigraphy data was analyzed with Action4® and MATLAB® software. Data on total sleep time, sleep onset latency (SOL), sleep cycle mesor, acrophase, amplitude, inter-daily stability (IDS), intra-daily variability (IDV) and F-ratio were collected, analyzed and reported.

Results: SCD patients took significantly shorter time to fall asleep than controls (mean SOL: 75 ± 15.35 versus 113.41 ± 11.79 min, $p = 0.02$), and slept significantly shorter (mean SOL: 422.66 ± 33.24 versus 528.75 ± 24.10 min, $p = 0.0084$). In addition, they also displayed a lower sleep ratio (0.593 ± 0.045 versus 0.424 ± 0.044, $p = 0.0071$), and less frequent night awakenings (14 ± 1.47 versus 20.42 ± 2.29, $p = 0.014$). But for the mesor (156.27 ± 4.34 and 142.55 ± 4.55, respectively for SCD and controls, $p = 0.02$), cosinor rhythmometry was not significantly different for both groups for the acrophase time, amplitude, the F-ratio and goodness of fit.

Conclusion: This is the first report in Cameroon showing that sleep and circadian rhythm disorders that are common in SCD patients, can be easily characterized using actigraphy. We propose the use of actigraphy in routine sleep evaluation in SCD patients after validation of the technique.

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Abstract – WCN 2013

No: 3068

Topic: 36 – Other Topic

Actigraphy in the assessment of sleep patterns in HIV-AIDS in Cameroon (sub-Saharan Africa)

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Background: Sleep disorders are more common among people living with HIV-AIDS (PLWHA) in Western countries, but few data exist on the subject in Africa. This study sought to characterize sleep patterns in PLWHA using actigraphy, a validated user-friendly sleep study tool.

Patients and methods: 16 PLWHA and 16 age and sex-matched HIV-negative controls wore wrist actigraphs for a minimum of 72 h and kept a sleep diary. Actigraphy data was analyzed with Action4® and MATLAB software. Data on total sleep time, sleep onset latency (SOL), sleep cycle mesor, acrophase, amplitude, inter-daily stability (IDS), intra-daily variability (IDV) and F-ratio were collected, analyzed and reported.

Results: Though PLWHA took significantly shorter time to fall asleep than controls (mean SOL: 83.50 ± 48.89 versus 174.94 ± 103.57 min, $p = 0.003$), they slept significantly longer than the controls (582 ± 12.00 versus 433.50 ± 170.57, $p = 0.008$) and had more frequent night awakenings (20.06 ± 11.072 versus 12.13 ± 6.41, $p = 0.019$). But for the acrophase time (13:14 ± 1:23 and 14:26 ± 1:09, respectively for PLWHA and controls, $p = 0.007$), cosinor rhythmometry was not significantly different for both groups for the mesor, amplitude, the F-ratio and goodness of fit. Intra-daily variability was higher in PLWHA than in controls (0.235 ± 0.023 versus 0.186 ± 0.009, $p = 0.03$). The reverse was true for inter-daily stability (0.97 ± 0.001 versus 0.96 ± 0.003, $p = 0.015$).

Conclusion: This is the first report in Africa showing that sleep and circadian rhythm disorders that are common in PLWHA, can be easily characterized using actigraphy. We propose the use of actigraphy in routine sleep evaluation in PLWHA after validation of the technique.

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Abstract – WCN 2013

No: 3103

Topic: 36 – Other Topic

Clinical predictors of sleep disorders in sickle-cell disease (SCD) patients in Cameroon

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Background: Good sleep quality is important for optimal brain function and quality of life while poor sleep may precipitate acute pain crises in sickle cell disease (SCD). Data on sleep disorders in SCD as well as their clinical predictors is limited in Africa and does not exist in Cameroon to the best of our knowledge.

Patients and methods: As part of a case-control study, binary logistic modeling was used to identify clinical predictors of obstructive sleep apnea (OSA) and restless legs syndrome (RLS) in 45 SCD patients (hemoglobin SS) aged 2 to 17 years screened for sleep disorders with the pediatric Sleep Quality (PSQ) scale.

Results: The prevalence of OSA and RLS was respectively 26.7% and 22.2%. An enlarged tonsil and vaso-occlusive crises were predisposing

factors for OSA (AOR 106.19 95% CI 2.52–4471.29 and AOR 3238.54 95% CI 4.76– 1.0×10^6 for ≥ 4 crises respectively). On the other hand having been hospitalized more than twice in the past year was a protective factor (AOR 0.002 95% CI 0.00–0.74). Sex, age of participant, age at diagnosis, number of transfusions in the past year, and past histories of stroke, enuresis, and hydroxyurea use were not significant predictors. None of the above-named factors were identified as predictors of RLS.

Conclusion: Vaso-occlusive crises and tonsillar hypertrophy predispose to OSA while hospital admissions are protective. Prevention and prompt management of pain crises and enlarged tonsils as well as hospital management of SCD crises appear to prevent sleep OSA in sickle-cell patients.

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Abstract – WCN 2013

No: 3108

Topic: 36 – Other Topic

Obstructive sleep apnea in sickle-cell disease patients in Cameroon

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Background: Obstructive sleep apnea is a major cause of poor sleep in sickle-cell disease (SCD). Poor sleep affects functional status, quality of life and may precipitate acute pain crises. Data on OSA in SCD is limited in Africa and does not exist in Cameroon to our knowledge.

Patients and methods: A case-control study of 45 SCD patients (hemoglobin SS) age- and sex-matched with 45 non-sickle cell disease controls aged 2 to 17 years employed the Pediatric Sleep Quality (PSQ) scale to screen for obstructive sleep apnea (OSA) and the Epworth Sleepiness Scale (ESS) for daytime sleepiness.

Results: Obstructive sleep apnea was significantly more common in sickle-cell patients compared to the controls (26.7% versus 2.2%, $p = 0.002$, OR 15.9 95% CI 1.98–125). Snoring as well as inattention hyperactivity PSQ scores were significantly higher in the cases compared to the controls (0.91 ± 1.18 versus 0.09 ± 0.29 , $p < 0.001$ and 2.11 ± 1.90 versus 0.96 ± 1.92 $p = 0.002$).

More sickle-cell patients with OSA had enlarged tonsils than not (47.4% versus 12.5% $p = 0.017$ OR 6.30 95% CI: 1.39–28.46). Excessive daytime sleepiness was more common in sickle-cell patients with OSA compared to those without (27.3% versus 0.0%, $p = 0.012$).

Conclusion: Obstructive-sleep apnea is common in sickle cell disease patients in Cameroon. The management and prevention of tonsillar hypertrophy should be part of OSA management in SCD as well as excessive daytime sleepiness, inattention hyperactivity and its consequences.

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Abstract – WCN 2013

No: 3115

Topic: 36 – Other Topic

Bedwetting and sleep disorders in sickle cell disease patients in Cameroon

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Background: Bedwetting and enuresis are known predictive factors of sleep disorders in sickle-cell disease (SCD). Poor sleep affects functional status, quality of life and may precipitate acute painful crises. Data on bedwetting in SCD is limited in Africa and does not exist in Cameroon.

Patients and methods: A case-control study of 45 SCD patients (hemoglobin SS) age- and sex-matched with 45 non-sickle cell anemia controls aged 2 to 17 years. We used the Pediatric Sleep Quality (PSQ) scale to screen for obstructive sleep apnea (OSA) and restless legs syndrome (RLS) and the Epworth Sleepiness Scale (ESS) for daytime sleepiness.

Results: Though cases gained bladder control earlier than controls (78.6% versus 66.7% before the age of 2 years, $p = 0.229$) bedwetting was two and a half times more common in the former than in the latter (56.8% versus 22.7%, $p = 0.001$, OR 4.54 95% CI 1.79–11.11). This was true for males as well as for females (54.5% versus 21.7%, $p = 0.023$ and 59.1% versus 23.8%, $p = 0.019$ respectively).

Enuresis was more common in the cases than in the controls though not significantly (63.2% versus 42.9% $p = 0.407$). Enuresis was significantly more common in sickle-cell patients with OSA compared to those without (100.0% versus 46.2%, $p = 0.044$). This was observed for RLS and daytime sleepiness, though not significantly (100.0% versus 56.2%, $p = 0.263$ and 100.0% versus 50.0%, $p = 0.106$).

Conclusion: Bedwetting and enuresis are common in sickle-cell disease in Cameroon and appropriate management strategies are needed.

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Abstract – WCN 2013

No: 3117

Topic: 36 – Other Topic

Vitamin B12 status does not influence central motor conduction time in asymptomatic elderly people: A transcranial magnetic stimulation study

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Background: Vitamin B12 deficiency causes neurologic and psychiatric disease, especially in older adults. Subacute combined degeneration is characterized by damage to the posterior and lateral spinal cord affecting the corticospinal tract.

Objective: To test corticospinal tract projections using motor evoked potentials (MEPs) by transcranial magnetic stimulation (TMS) in asymptomatic older adults with low vitamin B12 (B12) levels.

Patients and methods: Cross-sectional study of 53 healthy older adults (> 70 years). MEPs were recorded in the abductor pollicis brevis and tibialis anterior muscles, at rest and during slight tonic contraction. Central motor conduction time (CMCT) was derived from MEP latency and peripheral motor conduction time (PMCT). Neurophysiological variables were analyzed statistically according B12 status.

Results: Median age was 74.3 ± 3.6 years (58.5% women). Twenty-six out of the 53 subjects had low vitamin B12 levels ($B12 < 221$ pmol/L). MEPs were recorded for all subjects in the upper and lower extremities. There were no significant differences in either latency or amplitude of MEPs and CMCT between low and normal B12 groups. There was a significant PMCT delay in the lower extremities in the low B12 group ($p = 0.014$).

Conclusion: No subclinical abnormality of the corticospinal tract is detected in asymptomatic B12 deficient older adults. The peripheral

nervous system appears to be more vulnerable to damage attributable to this vitamin deficit.

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Abstract – WCN 2013

No: 3114

Topic: 36 – Other Topic

Paraneoplastic cerebellar degeneration (PCD) – Improvement after oophorectomy and PLEX. Case report

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Objectives: Paraneoplastic cerebellar degeneration (PCD) is a rare syndrome associated with systemic malignancies, most in ovarian cancer, breast cancer and lung cancer. Clinical symptoms develop within a few weeks or months. In the course of PCD most often found in incidence of anti-Yo, that accompany ovarian cancer.

Methods: The authors describe a case PCD without the presence of ovarian cancer, with a high titer of anti-Yo where clinical improvement occurred after ovariectomy and PLEX.

Results: A 52 year old postmenopausal female subacutely developing ataxia, tremor, nystagmus, dysarthria and dysphagia with progression of symptoms during several weeks, resulting in the inability of sitting and standing. The study demonstrated the presence of serum anti-Yo titer 1:10,000 in IIFT (confirmed test Euroline), and elevated levels of tumor marker CA-125–83.530 U/ml. The patient was previously treated with pulses of methylprednisolone and PLEX no clinical effect. A study of PET and CT of the chest, abdomen and pelvis. There was no cancer present. Observed accumulation of physical disability, the patient confined to bed. For this reason, an oophorectomy was performed, upon histopathology there was no presence of the tumor. A month after the operation was repeated treatments of PLEX provide clinical improvement – patient sitting alone.

Conclusion: This report shows efficacy oophorectomy and PLEX in PCD with the presence of anti-Yo and elevated Ca-125 without the presence of an ovarian tumor.

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Abstract – WCN 2013

No: 3106

Topic: 36 – Other Topic

A longitudinal study in a case of Köhlmeier–Degos-disease with CNS-involvement: Clinical and neuroradiological evolution

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Köhlmeier–Degos disease is a small vessel angiopathy with primarily cutaneous manifestation (papulosis atrophicans), targeting only the skin in the benign variant of the disease (majority of diagnosed cases); in 30% there may be also an additional involvement of other organ systems: gastrointestinal tract, urogenital tract, CNS, ophthalmic and cardiac manifestations.

As etiological factors, recently a C5b/IFN-alpha-mediated mechanism targeting the endothelium, altered fibrinolysis and platelet functions was identified as a cause for this systemic vaso-occlusive disorder. So far the literature database includes numerous singular case reports, rarely focusing on neurological cases, and then in a cross-sectional

setting. In our report we will show the clinical course in correlation to neuroradiological and laboratory findings, demonstrated in a case of a 35 yr old man. The strict sequential appearance includes the cutaneous manifestation 2 yrs prior to ocular affection with progressive occlusion of the central artery, prior to progressive development of disseminated vascular CNS-lesions in a territorial pattern including the capillary system and terminal vessels.

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Abstract – WCN 2013

No: 2314

Topic: 36 – Other Topic

Spastic paraparesis and sensorineural hearing loss in a case of neurobrucellosis

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Background: Brucellosis is a multisystem zoonosis with a broad spectrum of clinical presentations. Acute infection often goes undetected, and the disease runs an indolent course that can lead to irreversible sequelae. Nervous system involvement occurs in a minority of cases and early diagnosis is essential to avoid persistent neurologic deficits.

Objective: To report a rare case of spastic paraparesis and hearing loss due to neurobrucellosis, highlighting the importance of considering brucella infection in patients with unexplained neurological symptoms in endemic regions, even in the absence of fever.

Patients and methods: Case report.

Results: A 29-year-old Portuguese farmworker presented with a 1-year history of progressive difficulty walking. Three years earlier he had experienced a self-limited episode of headache and vomiting, and two years before admission he suffered sudden and sequential hearing loss affecting both ears within a week. No fever was reported and family history was irrelevant. Examination revealed profound sensorineural deafness, as well as spastic paraparesis, hyperreflexia and bilateral extensor plantars. Brain and spinal MRI showed leptomeningeal enhancement and serum Rose Bengal test was positive. CSF studies revealed lymphocytic pleocytosis, low glucose, elevated protein, oligoclonal IgG bands and positive *Brucella*-specific IgG and IgA antibodies. A diagnosis of neurobrucellosis was made and the patient was started on combined therapy with rifampicin, doxycycline and trimethoprim-sulfamethoxazole.

Conclusion: Neurobrucellosis should always be considered in the diagnosis of chronic inflammatory CSF changes in endemic areas, and should be kept in mind when evaluating a patient with acquired spastic paraparesis, particularly when associated with hearing loss.

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Abstract – WCN 2013

No: 3065

Topic: 36 – Other Topic

Neurological manifestations of visceral leishmaniasis

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Background: Visceral leishmaniasis is common in our country. The neurological manifestations were found in some patients as it carries a high risk of morbidity and mortality.

Objectives: The study aimed to investigate the presence of neurological manifestations in a group of Sudanese adult patients with visceral leishmaniasis.

Patient and methods: This is a descriptive prospective cross sectional hospital based study. It was conducted at Omdurman tropical teaching hospital in Khartoum state; it implies 100 beds and 3 units of general medicine and specialists in tropical medicine.

Hundred patients with visceral leishmaniasis were examined for neurological manifestations using standardized questionnaire including medical history, clinical examination and investigation including, EMG and brain MRI.

Results: Male to female ratio was found to be 3:1 and common age group affected was <30 years. Peripheral neuropathy was found to be the commonest neurological manifestations followed by epilepsy, cranial nerves involvement and encephalopathy.

Conclusion: Patients with visceral leishmaniasis can present with neurological manifestations.

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Abstract – WCN 2013

No: 3112

Topic: 36 – Other Topic

Neurological manifestations of sickle cell anaemia

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Introduction: Central nervous system involvement is one of the most devastating aspects of SCD.

Objectives: The aims of this review are to document the range of neurological complications of sickle cell disease.

Methods: This is a prospective hospital based, cross-sectional study. One hundred Sudanese patients with Sickle cell anaemia were included in the study during the period from March to July 2012.

Results: The most common age group affected was below 20 years. Male to female ratio was almost equal; irritability & headache were the most common symptoms, 41% and 40% respectively. Numbness was observed in 25%, hemiplegia in 24%, seizure in 19%, recurrence of hemiplegia occurred in 8%, gait was found to be spastic in 14%, limping gait (due to non neurological causes) in 7%, while inability to walk in 6% and cerebellar manifestation in 2%.

Conclusion: The study revealed high incidence of irritability and headache followed by numbness then hemiplegia. Hemiplegia is usually ischemic in children and haemorrhagic in adults. Silent brain infarcts occur in 17% of our patients. Convulsions occur as an isolated event but frequently associated with stroke.

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Abstract – WCN 2013

No: 1854

Topic: 36 – Other Topic

Ischemic stroke in cerebrotendinous xanthomatosis

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Background: Cerebrotendinous xanthomatosis (CTX) is an extremely rare treatable lipid storage disease, characterized by early-onset cataracts, tendinous xanthomas and progressive neurological dysfunction due to cholestanol accumulation. Despite an antiatherogenic lipid profile, premature atherosclerosis and heart disease are common, but cerebrovascular disease has seldom been reported.

Objective: Our aim is to describe an unusual manifestation of CTX, and discuss the relevance of cerebrovascular disease in this condition.

Materials and methods: Case report.

Clinical report: A 35-year-old man presented with acute right-sided weakness. He had a long-standing history of cognitive impairment, lower limb deformities, and a recent diagnosis of cataracts and hypertension. He had a sister with cataracts and cognitive impairment and no other known family history. Neurological examination revealed right-sided hemiparesis, dysarthria and ipsilateral facial and hypoglossal palsy, as well as pyramidal and extrapyramidal dysfunction. He had prominent xanthomas in the Achilles tendons, patellar tendons, and hand and elbow extensors. Brain MRI revealed diffuse cerebral and cerebellar atrophy, basilar dolichoectasia and an ischemic lesion involving the left thalamus and cerebral peduncle. Carotid ultrasound showed small bulbar plaques bilaterally and bloodwork revealed normal total and LDL cholesterol, low HDL and mild hypertriglyceridemia. Serum cholestanol was elevated, confirming the diagnosis of CTX, and the patient was started on antiplatelet, statin and chenodeoxycholic acid treatment.

Conclusion: Although cerebrovascular disease is not traditionally associated with CTX, it may be an underreported complication of accelerated atherosclerosis in these patients. Further data are required to define a causal association.

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Abstract – WCN 2013

No: 3045

Topic: 36 – Other Topic

Vernet syndrome as a presentation of varicella-zoster virus infection

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Background: Vernet syndrome is defined by a unilateral palsy of glossopharyngeal, vagus and accessory nerves and may occur due to different causes. We present a rare case associated with an infection by the varicella-zoster virus (VZV).

Case report: A 76-year-old man, with previous history of poliomyelitis, presented with one week-long symptoms of dysphonia and dysphagia, mild right temporo-parietal headache extending to the ipsilateral ear and worsening of previous right arm weakness. Neurological examination showed dysphonia, asymmetrical elevation of soft palate with left deviation, paresis of the right trapezius and several vesicular lesions in the right ear's concha. Laryngoscopic examination demonstrated right vocal cord palsy. Cerebrospinal fluid (CSF) examination revealed 6 cells/mm³ and normal levels of glucose and proteins. Head MRI disclosed thickening and gadolinium enhancement of right IX, X and XI cranial nerves in T1 sequences. Antibody titer for VZV was initially elevated in serum-IgM and IgG-progressing to isolated IgG elevation several weeks later. VZV-DNA was not detected by PCR in CSF. The patient had a remarkable clinical improvement after initiation of acyclovir and prednisolone therapy.

Conclusions: VZV infection with lower cranial nerves palsy is extremely rare. The absence of VZV-DNA in CSF does not exclude the diagnosis, since the test for this particular virus has a low sensibility (42–53%). In this case, the diagnosis of Vernet Syndrome, as a result of VZV infection, is demonstrated by clinical, laboratory and imaging data. Awareness to this rare presentation of VZV infection is important since it benefits from specific therapy.

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Abstract – WCN 2013**No: 3099****Topic: 36 – Other Topic****Diagnosis and management of neuro-Behçet's disease:****International consensus recommendations**

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Background: The neurological complications of Behçet's disease (BD) [neuro-Behçet's disease (NBD)] are amongst the most serious and disabling manifestations of BD. Evidence based recommendations on the main diagnostic and therapeutic aspects of NBD are scarce.

Objective: To develop internationally agreed consensus recommendations on the main diagnostic and management issues of NBD to assist clinicians involved in the care of such patients.

Methods: We conducted a literature review using Cochrane, Medline and Embase databases to generate draft recommendations. We followed this by a three-round Delphi consultation process to achieve consensus. The expert group consisted of 52 international members from 20 countries, including 23 neurologists and a voluntary patient representative. The draft recommendations were modified after each consultation round based on the expert feedback. This was followed by voting by a smaller consensus group consisting primarily of neurologists with interest in NBD. Draft recommendations with high level of agreement (score $\geq 7/9$) by $\geq 75\%$ of the Group members were accepted.

Results: Seventeen recommendations were agreed on by the group as consensus recommendations. These include new diagnostic criteria for NBD.

Conclusion: This international project has managed to agree and develop diagnostic and therapeutic consensus recommendations. We aim to present these at the World Congress of Neurology 2013.

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Abstract – WCN 2013**No: 3044****Topic: 36 – Other Topic****Results of video-EEG monitorization in the rats treated kainic acid in acute term**

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Background and objective: Kainic acid (KA) treatment causes neuronal injury primarily in the amygdala and hippocampus as well as in many regions of the rat brain and activates epileptic seizures. This model is used to research human temporal lobe epilepsy (TLE) in chronic term. We aimed to investigate clinic and electrophysiologic features of seizures in the KA-treated rats to form TLE models in acute term.

Material and methods: Sixteen young adult Wistar albino rats (220–270 g) were used in this study. Rats were divided into two groups as acute kainic acid and sham. Teflon coated steel wire electrodes were stereotactically inserted into the rat hippocampus bilaterally. A week after electrode implantation, intraperitoneal (i.p.) injections of KA

(5–15 mg/kg) were given to rats in order to follow with video-EEG monitorization.

Results: The first clinic seizure latency was measured as 85 m (29–132 m) while the first seizure duration was 95 s (14–428 s). 7 of 16 rats underwent convulsive status epilepticus (SE) clinically whereas one of them had nonconvulsive SE. One of them died during SE. 27 absence, 35 focal motor, 49 myoclonic seizures were observed in 24 h. Isolated or serial epileptiform discharges have been detected in the interictal period. No ictal seizure and interictal epileptiform discharges were observed in the sham group.

Conclusion: The first study to show the video-EEG synchronization of acute term KA-treated TLE models, a new semiological classification is made by classifying clinic and electrophysiologic seizures.

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Abstract – WCN 2013**No: 3050****Topic: 36 – Other Topic****Using photosensor for precise recording of the stimulus onset during N400 event-related potential registration**

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Background: Extracting event-related potentials (ERP) from multi-channel EEG recordings requires precise synchronization between stimulus presentation and EEG recording system.

Objective: The aim of this study was to develop a photosensor for a precise recording of stimulus onset during N400 ERP registration.

Material and methods: The sample consists of 15 healthy subjects who were visually presented with 104 semantically congruent or incongruent word pairs. ERPs were obtained by averaging EEG data registered with the 21 channel digital EEG system (impedance $< 5 \text{ k}\Omega$, bandwidth 0.1 Hz–30 Hz). Synchronization between stimulus presentation and EEG recording system was obtained using specially developed photosensor which was placed on the stimulus presentation screen and connected to the EEG bipolar channel. EEG signal was processed by ERPLAB software. Photosensor's sensitivity and response time were measured.

Results: Photosensor was made using silicon NPN phototransistor in hermetically sealed package with base terminal and glass lens. It was sensitive to changes in light intensity on the screen. Measured sensitivity was 0.2 $\mu\text{V}/\text{lux}$ with constant response time ($< 1 \text{ ms}$). ERPs were extracted by averaging EEG data based on precise timing information which was recorded as a spike at one of the EEG channels. A statistically significant difference in the N400 ERP component amplitude was registered between the semantically congruent and incongruent trials in the interval ranging from 300 to 400 ms after the stimulation ($p < 0.05$).

Conclusion: This research demonstrates a successful utilization of photosensor for a precise recording of the stimulus onset during N400 ERP registration.

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Abstract – WCN 2013**No: 2894****Topic: 36 – Other Topic****Astrocytes in infantile neuronal ceroid lipofuscinosis (INCL)**

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Background: INCL is the commonest pediatric neurodegenerative disorder caused by autosomal recessive mutation. Children affected

by INCL develop visual defects, seizures and cognitive deficits, leading to premature death. INCL is directly caused by mutation in CLN1 gene that encodes for lysosomal enzyme Ppt1. Despite our knowledge of the mutation and the role of the protein, we have no understanding of how faulty PPT1 enzyme leads to these phenotypes. Although INCL is classified as neurodegenerative disease, astrocytes are activated prior to neuronal death. Hence my aim was to observe the astrocytes of PPT1 $-/-$ and WT mice in vivo and in vitro.

Aims:

- To determine if PPT1 deficient astrocytes behave differently to wild type astrocytes.
- To identify any difference in lysosomes of WT and PPT1 mice brain.

Results: PPT1 $-/-$ astrocytes were abnormal in 4 domains: they were stimulated very early in development leading to increased proliferation, variable morphology, altered protein expression and secretion.

Initially these observations were localized to the thalamus and the cortex, however in older mice these abnormalities were generalized in the brain indicating progression of the disease.

Conclusion: PPT1 $-/-$ astrocytes behave rather different to the WT astrocytes in all fundamental cellular behaviors. They are stimulated early in the brain: with proliferation, altered protein expression and cytokine secretion. These alterations may result in secretion of substances which are harmful and halt secreting cytokines which are necessary leading to the development of a toxic environment for the neurons. Hence neuronal death is localized to these changes and observed soon after.

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Abstract – WCN 2013

No: 3063

Topic: 36 – Other Topic

A case of primary central nervous system lymphoma

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Background: Primary central nervous system lymphomas (PCNSL) are rare non-Hodgkin lymphomas, constitute 3–4% of primary brain tumors, and most cases are diffuse large B-cell lymphomas. Brain biopsy is essential to a precise histological diagnosis.

Objective: We present a case of a patient whose definitive diagnosis was delayed due to non-specific MRI and pathology findings.

Case: Previously healthy 53 year-old female patient, presented with a 2-month history of vertigo, ataxia and diplopia. Neurologic examination revealed left fourth nerve palsy and gait difficulty. MRI showed irregular nodular contrast enhancement, an increase in diffuse intensity on the left periventricular white matter. MRI findings were considered to be likely demyelinating disease or lymphoma. PET/CT evaluation revealed multifocal CNS lymphoma. A CT guided stereotactic brain biopsy was accomplished but histological examination revealed no tumoral cell. For differential diagnosis of vasculitis, demyelinating and metastatic diseases, extensive tests and imaging techniques were performed. CSF examination showed just a slight increase in protein content. Patient's symptoms gradually worsened, so intravenous steroid treatment was started (1 gr/day for 5 days) empirically. Her symptoms improved rapidly but exacerbated 4 weeks later when therapy was withdrawn. Cranial MRI was repeated and revealed lymphoma, consequently a second stereotactic brain biopsy was performed and pathology results were reported as PCNSLs (diffuse large B cell). Patient was treated with methotrexate and cytarabine therapy.

Conclusions: PCNSL forms solitary or multiple mass lesions with homogeneous contrast enhancement on brain MRI. For the differential

diagnosis steroid responsiveness is important and brain biopsy is essential.

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Abstract – WCN 2013

No: 3072

Topic: 36 – Other Topic

Brain metastases: A new Graded Prognostic Assessment (GPA) index

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Background: The Graded Prognostic Assessment (GPA) is a commonly used index based on prior randomized trials. The purpose of this study was to validate GPA index in a recent cohort of cancer patients with brain metastases (BM) at a large tertiary care center.

Methods: Cleveland Clinic Neuro-Oncology Center's database was used to identify BM treated in the recent era (2000–12). A proportional hazard model was used to assess overall survival (OS), measured from the date of diagnosis of brain metastases to death or last follow-up.

Results: 877 BM patients (46% males) median age 57 years (range 23–84) were included. 490 patients with lung cancer, 161 patients with breast cancer, 90 with melanoma and 136 with renal cell carcinoma were analyzed. The median number of brain metastases was 1 (range, 1–47). OS was 14.3 months (95% C.I. 12.9–15.5), and is highly dependent on the underlying primary tumor. The original GPA was prognostic for survival ($p < .0001$). However we found that these factors can be employed to propose anew GPA that improves on the conventional GPA based on the following characteristics: the primary is controlled, the number of extracranial metastases present, and the interval from initial diagnosis of the primary to the diagnosis of BM ($p < 0.0001$).

Conclusion: A new index developed based on a revised set of independent prognostic factors (primary controlled, diagnosis to be brain metastases, and number of extracranial metastases) is proposed.

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Abstract – WCN 2013

No: 3087

Topic: 36 – Other Topic

Limbic encephalitis, clinical features and autopsy findings

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Background: Paraneoplastic limbic encephalitis (PLE) is a rare disorder characterized by behavioral change, depression and memory loss, sometimes associated with dementia, psychosis and seizures. Antineuronal antibodies may be detected. Imaging results present variable results.

Case report: A 42 year old white winegrower developed an anti-Hu-antibodies positive of PLE. The patient presented initially with ataxic gait, peripheral facial palsy and loss in short term memory. Double vision and insecure gait developed within half a year. First MRI was unsuspecting. Follow-up MRI four months later demonstrated hyperintense temporomedial enhancement. Two lumbar punctures had unsuspecting results (cell count, cell morphology). Anti-Hu-antibodies and oligoclonal bands were positive. EEG findings revealed slowing, but no ictal pattern. The tumour screening (CT thorax and abdomen) was negative. PET scan described a tracer enhancement mesiotemporally in the brain. After intravenous steroids over 5 days the ataxia improved. Cognitive deficits

persisted unchanged. Episodes with visual hallucinations and psychomotor agitation occurred. After the positive MRI-scan, we administered two series of intravenous immunoglobulins (IVIG). The cognitive impairment improved slightly. His general condition and dysphagia deteriorated. The patient died of aspiration and cardiorespiratory arrest. The general autopsy did not reveal a tumour.

Brain autopsy showed no macroscopic changes; in histology patchy lymphoid infiltrates mainly perivascular were discovered in the brain parenchyma. Most of the lymphoid cell stained positive with T cell markers.

Conclusion: The case describes the clinical course of a Hu positive PLE, which could be followed up anatomically and confirmed the inflammatory changes in the brain parenchyma.

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Abstract – WCN 2013

No: 3056

Topic: 36 – Other Topic

Reversible encephalopathy in neuroborreliosis

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Background: Despite the fact, that borreliosis has a well documented involvement of the CSF and the nerve roots, reports on CNS manifestations of borreliosis are scarce.

Objective: Case report of a patient with neuroborreliosis and atypical MRI manifestations.

Case report: We report a 70 year old female with diplopia and unsteady gait for 3 weeks before admission. Medical history revealed no relevant diseases, except for unspecific symptoms like depression and weight loss of 15 k in the last 6 months. Neurological examination showed horizontal diplopia in all viewing directions and an unsteady gait with retropulsion. MRI exhibited T2-weighted, hyperintense lesions periventricularly and in the brainstem and rostrally up to the basal ganglia in both hemispheres resembling a perivascular spread of either infection or granulomatous process. CSF revealed increased cell count of 180/mm³ and a mixed cell type pattern consisting of lymphocytes, monocytes and plasma cells. The serology for borreliosis was positive, while diagnostic workup for autoimmune, neoplastic and infectious etiology remained negative. The patient improved well under intravenous antibiotics for 3 weeks and could be discharged in a normal condition without neurological symptoms.

Conclusion: Neuroborreliosis usually presents with a polyradiculitis of spinal and cranial nerves or varying symptoms arising from a meningoencephalitis, although rare manifestations like myelitis or borrelia induced cerebral vasculitis can be observed. The distribution of MRI lesions at first indicated a suppurative disease, and also demyelinating diseases were considered. This case with atypical MRI manifestation underlines the importance of considering neuroborreliosis in clinical diagnosis of inflammatory CNS diseases.

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Abstract – WCN 2013

No: 3071

Topic: 36 – Other Topic

Walter DeWitt Shelden: The father of neurology at the Mayo Clinic

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Background: Shelden started the neurology section at Mayo in 1913. **Objective:** To describe where Shelden trained, explain why he was chosen to start the neurology section at Mayo, and identify his local and national neurologic influence.

Material and methods: Review of material concerning Shelden in the Mayo and University of Vienna Archives.

Results: Shelden graduated from Rush Medical College in 1895, spent two years as an intern at Cook County Hospital, and worked as a general practitioner in Reedsburg, Wisconsin for four years. He then received internal medicine training at the University of Vienna for 1.5 years, working with Neusser, Kovacs, Frankl-Hochwart (neurology), Finger, and Monti. He subsequently had a private internal medicine practice and taught at the University of Minnesota for 10 years. He was known as an excellent bedside teacher and diagnostician, and a proficient neurologist. In 1913 he was asked by W.J. Mayo to start the Mayo neurology section. Shelden was head of the neurology section until 1930.

Conclusion: Shelden exemplified the benefit of post-graduate training in Vienna for the well-prepared American physician. His recruitment to Mayo Clinic gives insight into how W.J. Mayo chose diagnosticians and developed specialty sections. Shelden was an internist who focused primarily on neurology over time. The section of neurology at Mayo Clinic was more like internal medicine than neuropsychiatry because of Shelden's background. He did not publish much and therefore did not have a significant, direct, national influence, but he had an indirect influence via his trainees (H. Woltman, L. Eaton).

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Abstract – WCN 2013

No: 3070

Topic: 36 – Other Topic

Diffusion-weighted imaging in non-alcoholic Wernicke's encephalopathy

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Background: Wernicke's encephalopathy (WE) is a potentially dangerous neurological disorder, caused by thiamine deficiency. WE is most commonly seen in chronic alcoholics, yet it can manifest in any other patient with reduced thiamin intake.

Objective: To describe serial MRI findings, including diffusion-weighted imaging (DWI), during the course of WE in a patient with pyloric stenosis.

Results: A 52-year-old woman was admitted to our department with a 5-day history of diplopia. Cognitive functions were normal; there was no history of alcohol abuse. Neurological examination showed bilateral lateral rectus palsy with absent tendon reflexes. During hospitalization the patient's neurological condition deteriorated, with development of vomiting, complete ophthalmoplegia and anterograde amnesia. Gastroscopy showed severe pyloric stenosis. Brain MRI showed symmetrical lesions involving medial thalami, fornix and periaqueductal gray matter, with restricted diffusion on DWI, indicative of cytotoxic edema. During the ensuing week, the patient's condition worsened further, with ataxia, somnolence and finally coma. A new MRI scan showed spreading of lesions to the cerebellar vermis, as well as to the medullary tegmentum. Serum thiamine was well below normal limits. Following intravenous thiamine administration, consciousness was fully restored; moreover there was a complete resolution of ataxia and ophthalmoplegia. The patient was left with a residual Korsakoff syndrome; a follow-up MRI demonstrated complete remission of signal abnormalities.

Conclusion: DWI has the potential to become an invaluable tool for early diagnostic suspicion of WE; it may also monitor the clinical course of the disorder as well as its response to treatment.

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Abstract – WCN 2013**No: 3078****Topic: 36 – Other Topic****Vertebrobasilar Doppler sonography in patients with vertigo**

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Background: Vertigo as symptom of vertebrobasilar insufficiency (VBI) is a common neurovascular problem. It is caused by a reduction of vertebral artery flow, from atherosclerotic and degenerative changes of vertebral spinal structures. Color Doppler flow imaging has improved investigations of the vertebral arteries (VA). The aim of this study was to evaluate the blood flow characteristics of extra cranial segments of vertebral artery in patients with vertebrobasilar insufficiency.

Methods: Bilateral VA were examined sonographically in the prevertebral (V1 segment), intertransverse (V2 segment) and sub-occipital (V3 segment) in 80 patients with VBI. Angle-corrected peak systolic (Vps), end-diastolic (Ved), and time-averaged maximum blood flow velocity (TAV) were measured in pulsed Doppler mode, and the resistance index (RI) was calculated. The cross-sectional area (A) was measured on gray-scale images and flow volume was calculated.

Results: Left VA were dominant in 65% of patients. RI values increased and flow decreased with age. Blood flow velocity and volume were higher, and RI was lower in the left than in the right VA. In VBI patients peak systolic velocity and RI were significant higher in V1 segment, and lower in the V2 and V3 segments: VpsV1 = 62.7 ± 16.4 cm/s; VpsV2 = 46.5 ± 15.1 cm/s; VpsV3 = 45.6 ± 17.7 cm/s; RIV1 = 72.2 ± 10.1; RIV2 = 68.7 ± 12.9; RIV3 = 64.6 ± 14.4.

Conclusion: The Doppler sonographic assessment of extra cranial VA may be useful for the study of hemodynamic changes in patients with vertebrobasilar insufficiency.

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Abstract – WCN 2013**No: 3079****Topic: 36 – Other Topic****Pathology of the main arteries of the patients with controlled and uncontrolled arterial hypertension**

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Research objectives: To compare the structural changes of the carotid arteries of the patients with controlled and uncontrolled arterial hypertension.

Materials and techniques: 56 people have been examined (average age was 58.6). The first group (n = 24) included patients with uncontrolled arterial hypertension with episodic high blood pressure (>180 mm Hg) for the last 3 months while treating. The second group (n = 14) included patients with controlled hypertension, there was episodic high blood pressure in their medical history, but while treating for the last 3 months blood pressure was of ≤180 mm Hg. The third group (n = 17) included patients with no arterial hypertension. Patients of all 3 groups matched on age and sex.

Results: 33% of the patients of the 1st group (n = 8) and 7.1% of the patients of the 2nd group (n = 1) had episodic cerebrovascular accidents (NIHSS average score 4.5). Patients of the 1st group had more subjective symptoms than the others, especially concerning giddiness. 87.5% of the patients of the 1st group (n = 21), 85.7% of the patients 2nd group (n = 12) and 76.4% patients of the 3rd group (n = 13) had arterial sclerotic disease of the carotid arteries and vertebral arteries.

Conclusion: While researching we discovered that patients with arterial hypertension had arterial sclerotic disease of the extracranial artery and main artery deformities of the brain. At that, patients with uncontrolled arterial hypertension suffered from sclerotic disease of

the arteries more often. Detection frequency of the main arteries deformity of the patients with controlled and uncontrolled arterial hypertension was approximately the same.

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Abstract – WCN 2013**No: 2618****Topic: 36 – Other Topic****Response to treatment in lead exposed children with autonomic dysfunction**

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There is evidence that chronic lead exposure causes dysfunctions of the autonomic nervous system: impaired autonomic function assessed by heart rate variability (HRV) was shown in adults occupationally and children environmentally exposed to lead. Effects of lead exposure on treatment efficiency of autonomic dysfunction (AD) have not yet been assessed.

Repeated ECG five-minute recordings in supine position were conducted; standard time and frequency domain HRV parameters were obtained in 60 children with AD (13.25 ± 0.17 year old; 40 boys) before, in the middle, and after treatment. All children received same therapy during the 1 month period. Hair lead concentrations were determined using X-ray spectrophotometry; children were considered to be lead-exposed if lead level was above 5 µg/g.

No significant difference in the initial HRV parameters except for RRNN were observed in exposed group (n = 27) compared to that of non-exposed, with mean lead levels ten times higher in first (14,80 ± 4,03 µg/g, p < 0.001). The initial RRNN was significantly lower in lead-exposed group (775.81 ± 22.71 ms vs 836.15 ± 22.48 ms). Significant changes in HRV were found in both groups: non-exposed group showed gradual increase in RRNN from 1.2% to 6.1% by the end of treatment, while in lead-exposed – its initial rise was followed by 1.7% drop in the end (p < 0.05). RMSSD and HF values showed significant depression by the end of treatment in exposed children; Pb levels were directly correlated with RMSSD and HF (0.39 < r < 0.43, p < 0.05).

These data suggest that lead exposure affects cardiac autonomic functions through the parasympathetic nervous system.

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Abstract – WCN 2013**No: 2600****Topic: 36 – Other Topic****Sleep disorders in sickle cell disease patients in Cameroon**

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Background: Chronic symptoms such as sleep complaints in sickle-cell disease (SCD) patients are often ignored in favour of more acute phenomena such as pain and anaemia. Poor sleep affects functional status and may precipitate acute pain crises. Data on sleep in SCD is limited in Africa and does not exist in Cameroon.

Patients and methods: A case-control study of 45 SCD patients (hemoglobin SS) age- and sex-matched with 45 non-sickle cell anemia controls aged 2 to 17 years. We used the Pediatric Sleep

Quality (PSQ) scale to screen for obstructive sleep apnea (OSA) and restless legs syndrome (RLS), and the Epworth Sleepiness Scale (ESS) for daytime sleepiness.

Results: The prevalence of OSA, RLS and daytime sleepiness was significantly higher in the cases than in the controls (26.7% versus 2.2%, $p < 0.001$, 22.2% versus 2.2%, $p = 0.007$ and 6.8% versus 2.2%, $p = 0.051$ respectively). In adjusting for socio-economic status (SES) the odds of OSA in the cases compared to the controls was 12.53 (95% CI: 1.41–114.43) and on adjusting further for depression 5.90 (0.48–72.54).

With regard to RLS, in adjusting for SES and depression the OR became insignificant: AOR 8.19 (0.79–100.56). In doing same for ESS, the OR which was of borderline significance ($p = 0.051$) became significant: AOR 9.62 (1.47–62.5).

Conclusion: Sleep disorders are common in sickle-cell patients in Cameroon. Depression and socio-economic status may mediate the occurrence of OSA and RLS. Clinical sleep quality assessment (with the aid of sleep scales) in SCD may contribute to better management and improve life quality.

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Abstract – WCN 2013

No: 3165

Topic: 36 – Other Topic

Laparoscopic assisted ventriculoperitoneal shunting in patients with normal pressure hydrocephalus

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Background: Normal pressure hydrocephalus is a potential reversible neurological decline characterized by the clinical triad: gait disturbance, urinary incontinence, and dementia, with dilated cerebral ventricles and normal cerebrospinal fluid (CSF) pressure. Indications, techniques and outcome measures for shunting remain under debate.

Objective: We evaluated outcome of NPH patients 3 months and one year after ventriculoperitoneal shunt placement with either laparoscopic or conventional surgical technique.

Material and methods: A total of 67 patients (mean age, 71.2 ± 16.6 years) had VPS-surgery at BIDMC between 2004 and 2012. Clinical assessment included CT, neuropsychological evaluation and lumbar CSF tap.

Results: There was no significant difference in outcome or in surgical morbidity between the two groups of patients. At 3-month and 1-year follow-up, 95% of patients improved in gait, 90% in urinary incontinence, and 75% in memory. Only 3 of 67 patients did not benefit from shunting. Evans ratio in our cohort was 0.37 (range, 0.24–0.5) in the pre-operative CT and 0.28 (range 0.2–0.5) in the post-operative CT done after 3 months.

Conclusion: In patients with NPH, laparoscopic shunt surgery is equally safe and useful and shows a low morbidity and failure rate with overall improvement in >90% of patients at one year follow-up. Laparoscopic surgery carries a number of distinct advantages such as shorter OR times, shorter hospital stays, less pain medications and earlier mobilization as well as a lower incidence of ileus. This technique should especially be considered in obese and medically frail patients.

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Abstract – WCN 2013

No: 3156

Topic: 36 – Other Topic

Lateral antebrachial cutaneous neuropathy

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Compression neuropathy of the lateral antebrachial cutaneous nerve presents as purely sensory lesions, manifesting as elbow pain or dysesthetic pain over the lateral forearm. It is an infrequent clinical entrapment syndrome. In this report, we present a case of lateral antebrachial cutaneous nerve injury which occurs after splint use.

A 48 year old female patient had fallen 2 months ago and then a forearm splint was applied. Numbness and a painful dysesthesia over the right radius had started 15 days following splint use. On physical examination, there was tenderness and swelling at the lateral forearm. There was no motor deficit. Plain radiographs of the arm and the forearm showed no abnormality. Electromyography showed no differences between the SNAP amplitudes of right and left superficial radial nerves and median sensory nerve of the first digits. However SNAP amplitude of the right lateral antebrachial cutaneous nerve was significantly (70%) lower than the contralateral side. The patient was treated conservatively with physical therapy and pregabalin. The result of this intervention was that the patient's dysesthesia was reduced.

Compression neuropathy of the lateral antebrachial cutaneous nerve is uncommon, and is easily overlooked. It is important to rule out other conditions that may produce similar symptoms including lateral epicondylitis, biceps strain, cervical radiculopathy, musculocutaneous nerve injury, and brachial plexopathy.

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Abstract – WCN 2013

No: 3176

Topic: 36 – Other Topic

Application of MSCTA combined with VRT in the operation of intra-extra cervical spinal canal dumbbell-shaped tumors

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Background: The operative management of the dumbbell-shaped tumors of the intra-extra cervical spinal canal is challenging. The complicated spatial relationship between tumor and the surrounding tissues is hardly understood by MRI and CT scans. However, the reconstruction of the three-dimensional space around the dumbbell-shaped tumors through multi-slice spiral CT angiography (MSCTA) combined with volume rendering technique (VRT) can predict favorable preoperative imaging data for surgery.

Objective: To probe the significance of MSCTA combined with VRT in the surgery of dumbbell-shaped tumors of the intra-extra cervical spinal canal.

Patients and methods: We examined 11 patients from 2010 to 2013 who exhibited dumbbell-shaped configuration tumors of the intra-extra cervical spinal canal by using MRI. MSCTA was performed before surgery. The tumor three-dimensional images were reconstructed by MSCTA along with the use of VRT, and the relationships among spinal cord, vertebral artery and vertebral bodies were clearly enough to imitate the approach of operation. The reconstruction images were compared with the surgical findings as control.

Results: All of the reconstruction images clearly showed the shape of the tumors and their three-dimensional relationships with adjacent vessels and vertebra, which were in accordance with the intraoperative findings. By imitating the operation, all patients were designed the best approach for tumor resection, effectively avoided unnecessary nerve and vascular injury, and reduced the blood loss in surgery while maintaining maximum cervical spine stability.

Conclusion: By coupling MSCTA with VRT, we can imitate the surgical operations in multi-angle and choose proper surgical

approaches, thereby reducing surgical trauma and postoperative complications.

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Abstract – WCN 2013

No: 3170

Topic: 36 – Other Topic

Neuroprotective effect of melatonin against bisphenol-a-induced apoptosis and upregulation of CD95-fas, caspase-3 activation and p53 expression in brain

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Background: Epidemiological reports have indicated the correlation between the increase of bisphenol-A (BPA) level in the environment and the incidence of neurodegenerative diseases.

Objective: In the present study, the molecular mechanism by which BPA causes neurotoxic effects was studied. Furthermore, the mechanism of melatonin's ability to reduce BPA-induced apoptosis was examined.

Material and methods: Adult male rats were orally administered BPA at a dose of 50 mg/kg body weight 3 days a week for 6 weeks with or without melatonin (10 mg/kg) cotreatment.

Results: Consistent with its antioxidant properties, melatonin reduced BPA-induced lipid peroxidation and ameliorated the levels of superoxide dismutase and glutathione in brain of rats treated with BPA toward control group. Furthermore, melatonin treatment reduced the upregulation of p53 and CD95-fas and caspase-3 activation induced by BPA treatment. We propose that, in addition to its antioxidant properties, melatonin has the ability to protect brain cells against apoptosis mediated BPA neurotoxicity by modulating death receptor pathway of apoptosis in the brain of rats.

Conclusion: melatonin is a promising pharmacological agent for preventing the potential neurotoxicity of BPA following occupational or environmental exposure.

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Abstract – WCN 2013

No: 3134

Topic: 36 – Other Topic

Eric Kodjo Grunitzky, neurologist and the father of neurology in Togo

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Objectives: Make the history of neurology in TOGO and the life of a special neurologist in Africa.

Method: Review of the literature covering strokes from theses (1975–2013), memoirs and publications available at the bookshelf of the University of Lomé and interviews.

Results: Born in 1948, He had begun neurology in 1980 in Lomé in Tokoin Teaching school. The first service of neurology was created a few years later, and the second service in the Campus Teaching school was created in 1988. With this creation much research especially on epilepsy (NADOBA Study) and stroke was done.

Conclusion: The history of neurology in Togo is very intimate with the carrier Prof Eric Grunitzky.

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Abstract – WCN 2013

No: 2703

Topic: 36 – Other Topic

Evaluation of quality of life with SF36 test in patient with obstructive sleep apnea syndrome

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Objective: To evaluate the quality of life in patients with obstructive sleep apnea and the relationship between the severity of obstructive sleep apnea and impairment of quality of life.

Methods: 69 patients who had undergone polysomnography (PSG) examination at Harran University sleep laboratory were enrolled in the study. 49 of them were male and 20 were female. 32 patients whose Apnea–Hypoapnea index (AHI) was below 5, were taken as controls. The severity of obstructive sleep apnea syndrome (OSAS) according to AHI were as follows: 9 patients whose AHI was between 5 and 14.9 were considered to have mild sleep apnea, 7 patients whose AHI was between 15 and 29.9 were considered to have moderate sleep apnea, 21 patients whose AHI was above 30 were considered to have severe sleep apnea. Short Form 36 (SF-36) quality of life questionnaire was performed to all patients.

Results: In the control group 17 were male and 15 were female, and the average age of the 32 patients was $40.34 \pm (8.14)$. The study group consisted of 37 patients, including 5 women and 32 men. The mean age of the group was $44.27 \pm (8.87)$. The SF-36's scores for OSAS patients were lower than those of the control group in all parameters. All parameters (general health, bodily pain, physical functioning, social functionality, physical role, and emotional aspects) except mental health and vitality were statistically significant ($p < 0.05$).

Conclusion: This study of sleep apnea syndrome, compared to the healthy control group, suggests a major health problem affecting quality of life.

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Abstract – WCN 2013

No: 3181

Topic: 36 – Other Topic

Dynamic change of CSF flow at lumbosacral level

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The hydrodynamics of the cerebrospinal fluid (CSF) in the brain can be directly inferred from structural change of the ventricles, whereas that in the spinal canal cannot be. As such, CSF flow at the lumbosacral level has not been a subject of clinical interest and little has been revealed thus far. In this study, we investigated how the CSF flow changes according to the posture using phase contrast magnetic resonance image (3.0T Siemens, German). Velocity encoding parameter was optimized after trials and periodicity of CSF flow was normalized per cardiac cycle. Phase contrast image in the axial plane was obtained at the L2 and S1 levels and phase contrast image in the sagittal plane was also recorded. During the study, lumbar flexion was maintained with the hip flexed in supine position and was supported by cushions. Ten subjects (25–77 years old, 5 male) participated in this study. In every case, the flow rate at the sacral level was significantly slower than at the lumbar level. During one cycle, steady cephalard flow was dominant in the early cycle and phasic caudal flow was dominant in the late cycle, either in neutral or in flexion posture. With lumbar flexion, the cephalard flow

rate of the CSF at the L2 level increased marginally compared to with neutral (0.53 ± 0.41 m/s at neutral, 0.94 ± 0.79 m/s at flexion). The AP diameter of the spinal canal increased insignificantly with lumbar flexion. In conclusion, the hydrodynamics of the CSF at lumbosacral level changed according to postures.

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Abstract – WCN 2013

No: 3161

Topic: 36 – Other Topic

Global functional connectivity is related to age and memory performance in healthy adults: A resting-state fMRI study

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Background: Global functional connectivity (GFC) is considered an estimate of a region's overall synchronization with the brain. Recently GFC has been used to identify associations with cognitive performance and changes in neuropsychiatric conditions. We are not aware of any study that investigated the relationship between age, episodic memory and GFC obtained from resting-state functional MRI (rs-fMRI) in healthy adults.

Objective: To investigate whether GFC during resting-state is associated with age and performance in episodic memory in adults with no neurological or psychiatric disorders.

Methods: Episodic memory performance and rs-fMRI were obtained from 37 cognitively healthy adults (age range: 19–75 years). The rs-fMRI was preprocessed and then, for each grey matter voxel, the mean of the z-transformed correlation coefficient between this voxel and every other grey matter voxel was calculated, resulting in a GFC map for each subject. To test the relationship between GFC, age and episodic memory performance we used the partial least squares method for neuroimaging.

Results: There were significant negative correlations between age and performance on the Wechsler Logical Memory test, both immediate ($r = -0.36$; $p = 0.03$) and delayed recall ($r = -0.35$; $p = 0.03$). Increased GFC in the bilateral motor cortex, left dorsolateral prefrontal cortex, bilateral superior temporal gyri, right visual cortex and left thalamus was significantly associated with aging and with worse performance in memory tests (permuted $p = 0.022$; absolute bootstrap ratio > 3).

Conclusion: Age-related increases in GFC were associated with poorer episodic memory performance. Interestingly, the most reliable findings were in brain regions not related to the default mode network.

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Abstract – WCN 2013

No: 3163

Topic: 36 – Other Topic

Effect of mobile phones on hearing

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Background: In the last two decades the use of mobile phone has increased globally. This has focused interest on its possible biological effects. The surprising huge increase in the usage of mobile phone in Sudan has been a strong justification for conducting such a study, on the possible harmful effects of mobile phone on hearing sense.

Objectives: To explore possible effects of mobile phone use on hearing level in humans and to compare between level of exposure and threshold for hearing.

Methods: This is an observational cross-sectional population-based study, conducted in Khartoum state – Sudan. Two hundred volunteers filled questionnaires and had their hearing tested by diagnostic audiometer. Those who use the mobile phone for a minimum of 2 h per day are classified as heavy users, and those who use the mobile phone for less than 2 h are classified as moderate users, while those who do not use the phone are classified as non-users.

Results: Heavy users of mobile phones show statistically significant prevalence of mild sensorineural hearing impairment, with no other possible underlining causes or risk factors. This is in comparison with the moderate users and the non-users, who all show normal hearing. Heavy users also show elevated auditory threshold at the minimum speech tone, compared to the other two classes, which means that they might be, at more risk for developing sensorineural hearing impairment. This study does not show association between long-term use of the mobile phone in years and hearing impairment.

Conclusion: The use of mobile phones has adverse effects on auditory function in humans. These effects need to be more thoroughly investigated.

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Topic: 36 – Other Topic

Case report – Viral encephalitis complicated by cerebral venous sinus thrombosis

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Background: Encephalitis is the inflammation of the brain often of unknown cause. The most common cause is viral infection. Cerebral venous sinus thrombosis [CVST] as a complication of encephalitis is a very rare phenomenon.

Objectives: To report cerebral venous sinus thrombosis as a complication of viral encephalitis.

Case presentation: This is a case report of a 23 year old male Sudanese university student who presented on the 26th of December 2012 with fever, headache, confusion and convulsions. There was no history suggestive of hypercoagulable states, vasculopathies or head & neck infections. On examination, he was confused, disorientated to time, anxious, agitated with mild neck stiffness. Fundoscopy was normal. He refused lumbar puncture. Full blood count showed lymphocytosis. Malaria investigations were negative. CT brain showed signs of encephalitis [diffuse brain swelling evidenced by sulcal effacement and attenuated cisterns]. MRI T1 showed oedema and high signal intensity in the temporal lobe [haemorrhagic areas]. T2 showed areas of hyper and hypo-intense signals in the temporal lobe suggesting herpes simplex encephalitis. He was treated with acyclovir & carbamazepine which resulted in dramatic improvement. Seven days later he started to complain of severe occipito-temporal headache and vomiting. Re-examination revealed development of papilloedema. MRV confirmed

the presence of sigmoid sinus thrombosis. Acetazolamide, heparin & warfarin were added to the treatment protocol. The patient showed remarkable response and is being regularly followed up.

Conclusion: Cerebral venous sinus thrombosis is a very rare complication of viral encephalitis. To our knowledge, this is one of the very few documented case reports in Sudan & worldwide.

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Structural features of denticulate ligaments spinal cord.

Implications for transport cerebrospinal fluid

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Recent research demonstrated that at least one-fifth of the cerebrospinal fluid (CSF) is reabsorbed from the spinal part of subarachnoid space. However, until now the structure involved in this process has not been clarified. The aim of this research was to investigate the structural elements of denticulate ligaments using light optical and electron microscopic techniques. The material of this study is the denticulate ligaments of domestic ducks. Our findings determine that a denticulate ligament together with cellular elements consist of mainly collagen fibrils (with a diameter 23–199 nm) and elastic fibers (with a diameter 114–706 nm). The absence of latter in the subpial space proved that the source formation of denticulate ligaments is the dura mater of the spinal cord. The study of serial semi- and ultrathin sections shows that the barrier and reticular cell layers of arachnoid covered only the initial part of the denticulate ligaments. The other parts of denticulate ligaments in subarachnoid space are not fully covered by a reticular cell layer of arachnoid. It should be noted that between the structural elements of denticulate ligaments and dura mater there are no structures that perform a barrier function, because the fibrillary elements of denticulate ligament without any boundaries merge into the dura mater. Given the known role of collagen fibrils in the transport of interstitial fluid and high hydrostatic pressure of the CSF it may be noted that one of the main ways of outflow CSF from the subarachnoid space can serve as a denticulate ligament.

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Inhibitory effects of fulvestrant on the growth of estrogen-induced prolactinoma in Fischer 344 rats and its possible mechanism

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Background: Prolactinoma is an estrogen-related tumor, and some anti-estrogen agents have suppressive effects on prolactinoma. Previous studies have suggested that fulvestrant, a selective estrogen receptor antagonist, has inhibitory effects on the proliferation and prolactin secretion of MMQ and GH3 cells *in vitro*. However, there is still limited evidence of the inhibitory effects of fulvestrant on prolactinoma *in vivo*.

Objective: To investigate whether fulvestrant can inhibit the growth or the prolactin release of prolactinoma *in vivo*, and identify the signaling pathways that mediate the effects.

Material and methods: Pituitary prolactinomas were induced in male Fisher 344 rats by s.c. implantation of silastic tubes containing diethylstilbestrol. The effects of treatment with fulvestrant on pituitary weight, prolactin release, cell proliferation and apoptosis were evaluated. The expression of signaling molecules in the apoptosis pathways and the expression of Wnt signaling pathway-related proteins β -catenin, c-Myc and cyclin D1 were analyzed by Western blotting to investigate the possible mechanisms.

Results: Fulvestrant treatment caused significant reduction in pituitary weight and prolactin release. High β -catenin level in cytoplasm and low β -catenin level in nucleus were detected, and the expression levels of cyclin D1/c-Myc were significantly decreased; the expression levels of p53 and Bax were increased, and Bcl-2 was decreased; Caspase-8 and Caspase-9 cleave bands were detected; Caspase-3 and its substrate PARP were activated in the fulvestrant-treated pituitary tissues.

Conclusion: We demonstrate that fulvestrant could inhibit prolactinoma growth, decrease prolactin release, induce cell apoptosis by the mitochondrial and receptor apoptotic pathways, and inhibit cell proliferation through Wnt pathway.

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Revisiting the observation of anxiety-like behavior and assessing phenotypic progression in PGC-1 α -deficient mice

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Introduction: Mice deficient in full-length peroxisome proliferator-activated receptor gamma coactivator-1 α (FL-PGC-1 α) have been implicated as model animals for mitochondrial spongiform leukoencephalopathies. In previous behavioral studies that these mice were reported to exert hypomotility and weakness, and were proposed to show anxiety-like behavior. However, studies confirming this anxiety-like behavior in validated experimental models of anxiety have been lacking, and no studies have yet addressed the issue of progressivity of motor symptoms in larger scale studies.

Materials and methods: To revisit the issue of anxiety we applied elevated plus maze and light-dark box tests, whereas locomotor performance and muscle strength were assessed by automated open-field motimetry and wire-hang test. FL-PGC-1 α -deficient (n = 27) and wild-type (n = 33) mice were distributed and analyzed along two age categories (mature 26.2 \pm 2.21 w and aged 57.5 \pm 1.4 w).

Results: Locomotor exploratory behavior and muscle strength were significantly decreased in FL-PGC-1 α -deficient mice compared to wild-type controls. Unexpectedly, however, FL-PGC-1 α -deficient mice demonstrated a behavior consistent with the complete lack of anxiety behavior. Both routine models applied failed to generate normal anxiety in FL-PGC-1 α -deficient mice. The results were consistent in both age categories, and comparing to the age-related decline in motor performance observed also in the control group we could not detect additional phenotypic progression in the FL-PGC-1 α -deficient group in the observed alterations.

Conclusion: Deficiency in FL-PGC-1 α in mice evokes a behavior consistent with weakness and hypomotility, and associates with a decreased susceptibility to produce anxiety behavior in conventional models.

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