Journal of the Neurological Sciences (2015) xxx-xxx

ELSEVIER

Contents lists available at SciVerse ScienceDirect

Journal of the Neurological Sciences



journal homepage: www.elsevier.com/locate/jns

Miscellaneous Topics 2

598 WFN15-1483 Miscellaneous Topics 2 Neurological complications in Pfeiffer syndrome

<u>C.J. Amlang</u>^a, A. Nouh^b, D.E. Anderson^c, J. Biller^d. ^anone, Charité Universitätsmedizin Berlin, Berlin, Germany; ^bNeurology, Hartford Hospital, Hartford CT, USA; ^cNeurological Surgery, Stritch School of Medicine Loyola University Chicago, Maywood IL, USA; ^dNeurology, Stritch School of Medicine Loyola University Chicago, Maywood IL, USA

Background: Pfeiffer syndrome is a rare genetic disorder characterized by craniosynostosis, broad thumbs, big toes and partial syndactyly of the hands and feet. It may also be associated with respiratory, ocular, otologic or neurologic problems that can complicate treatment. Raised intracranial pressure in Pfeiffer syndrome has been attributed to different mechanisms.

Methods: Single case report of a young man with a Pfeiffer-like syndrome presenting with severe headaches and progressive visual loss found to have high-normal to elevated intracranial pressure.

Results: Imaging studies showed a right internal jugular vein occlusion raising concerns as to whether venous sinus hypertension may have been involved in the increased intracranial pressure. However, the main concern in our patient was the appearance of bilateral disc edema and his sudden loss of vision (see figure A). He presented a more severe optic atrophy and a complete loss of vision on the left eye, associated with a narrowing of the left optic canal in a subsequent CT scan (see figure B). **Conclusion:** Neurologists should be aware of Pfeiffer syndrome, a rare genetic disorder which may possibly lead to several neurological complications.



doi:10.1016/j.jns.2015.08.669

599 WFN15-0039 Miscellaneous Topics 2 Prevalence of brain diseases in adolescents and adults served in the physical therapy services

<u>G. Baquero Sastre</u>. Physical Therapy, Manuela Beltran University, Bogotá, Colombia

Background: The brain diseases are important events in the neurology and in the public health for the negative impacts in the life process of the persons with this problems that have repercussions in the functionality and independence of the persons reducing the quality of life.

Objective: To know the prevalence and characteristics of presentation of the brain diseases in the adolescents and adults.

Material and methods: The investigation is a cross sectional study, the population of study was the patients over 15 years old that was served in the physical therapy services with practices of the Physical Therapy Program of Manuela Beltran University in the year of 2012 for neurology diseases, in the results analysis was calculate general and specific prevalences with the respective standar error, and the Chi Square Test with one grade of freedom, p < 0.05.

Results: 1062 persons was served for neurology diseases in the year of 2012, the prevalence of brain diseases was 68.36% (n=726, standar error=0.009), 56.19% of the persons have stroke (n=408, standar error=0.01), 53.92% of this persons was of female gender (n=220, standar error=0.02) (X2=0.002 p >0.05, O.R.= 1.00, C.I 95% 0.82 - 1.20), 28.78% of the persons with brain diseases have Alzheimer Disease (n=209, standar error=0.02), 77.51% (n=162, standar error=0.01) of this persons was of female gender (X2=49.76, p < 0.05, O.R.= 3.11, C.I 95% 2.27 - 4.22). **Conclusion:** The brain diseases have a important prevalence between the neurology diseases and the two diseases more important are the stroke and the Alzheimer

doi:10.1016/j.jns.2015.08.670

600 WFN15-0512 Miscellaneous Topics 2 Epidemiological and clinical aspects of amyotrophic lateral sclerosis in Republic of Moldova

<u>A. Belenciuc</u>^a, D. Gaina^a, V. Lisnic^b, M. Leone^c, E. Beghi^d. ^aDepartment of Neurology, Institute of Neurology and Neurosurgery, Chisinau, Moldova; ^bDepartment of Neurology, State University of Medicine and Pharmacy, "Nicolae Testemiţanu", Chisinau, Moldova; ^cComplex Structure of Neurology, Istituto di Ricovero e Cura a Carattere Scientifico (IRCCS) Casa Sollievo della Sofferenza, San Giovanni Rotondo, Italy; ^dLaboratory of Neurological Diseases, "Mario Negri" Institute for Pharmacological Research, Milan, Italy

ARTICLE IN PRESS

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

Background: Amyotrophic lateral sclerosis (ALS) is the most common motor neuron disease. Loss of pyramidal and anterior horn motor neurons leads to progressive limb weakness, disability, dysarthria, dysphagia and respiratory insufficiency with a progressive fatal course. **Objective:** To evaluate the epidemiological and clinical characteristics of amyotrophic lateral sclerosis (ALS) in the Republic of Moldova (3,559,497 inhabitants) during a 5-year period (2009–2013).

Materials and methods: To ensure complete case ascertainment, multiple sources of information were used, including neurologists, other medical specialties, neurophysiology laboratories. ALS diagnosis was based on El Escorial criteria. Although all patients with motor neuron disease were enrolled, only probable and definite cases were included in the study.

Results: A total of 94 cases was collected. The average annual incidence of ALS was 0.42 per 100,000 inhabitants. On December 31, 2013 the prevalence was 2.64 per 100,000. Mean age at onset was 55.7 years, with no sex difference. The peak incidence was in the age group 51-60 years. The male: female ratio was 2,03:1. The mean duration of the disease at diagnosis was 13.6 months. The clinical spectrum comprised spinal onset, beginning in the upper or lower limbs, or bulbar onset.

Conclusion: The clinical features of ALS in Moldova are similar to those of other epidemiological studies in Europe. Incidence and prevalence are lower than in other European countries. This could be explained by under ascertainment of cases due to misdiagnosis and late neurological consultation.

doi:10.1016/j.jns.2015.08.671

601 WFN15-1021 Miscellaneous Topics 2 Comparison of intravenous immunoglobulin (ivig) with plasmapheresis for treatment of pediatric Guillain-Barre syndrome

<u>E. Bidabadi</u>. Child neurology, Guilan University of Medical Sciences, Rasht, Iran

Objective: To make a comparison between 2 methods of treatment of Guillain-Barre syndrome in pediatric patients.

Material and methods: Fifty one children with a diagnosis of Guillain-Barre syndrome were treated randomly with IVIG (400 mg/kg/day for 5 days) or plasmapheresis (finally 250 cc/kg plasma removal, replaced with human albumin 5% & normal saline), in Rasht 17th Shahrivar University Hospital, Iran. Demographic data, patient's complaints, physical exam, paraclinical data, response to treatment, complications, length of hospitalization, and recurrence were collected and analyzed.

Results: The mean age of patients were 67.97 ± 39.78 months for IVIG and 79.28 ± 43.99 months for plasmapheresis groups. Thirty three (64.7%) patients were boys and 18(35.3%) were girls. Complications of treatment were more frequent in plasmapheresis group [the most frequent complications in IVIG group were aseptic meningitis, fever, and vomiting (6.06% for each) and in plasmapheresis group were hypokalemia and anemia (33.33% for each)]. Finally, 15 patients needed ventilator; mean ventilation period in IVIG and plasmapheresis groups were 11.67 ± 2.89 and 24.75 ± 20.77 days, respectively (non-significant); mean hospitalization period for these two groups were 9.52 ± 7.00 and 47.00 ± 45.07 days (p<0.001), and mean ambulation period were 58.45 ± 45.07 and 140.33 ± 50.91 days, respectively (p<0.001). Only one case of recurrence was seen in IVIG group after 70 days of discharge.

Conclusion: It seems that IVIG is better than plasmapheresis for treatment of pediatric Guillain-Barre syndrome.

doi:10.1016/j.jns.2015.08.672

602 WFN15-1155 Miscellaneous Topics 2

Thyroid hormones alterations in children with phenobarbital usage

<u>E. Bidabadi</u>^a, A. Mir Emarati^b. ^aChild neurology, Guilan University of Medical Sciences, Rasht, Iran; ^bpediatrics, Guilan University of Medical Sciences, Rasht, Iran

Objective: To make a comparison of thyroid hormones before and after phenobarbital treatment in pediatric patients.

Material and methods: Fifty two children with a diagnosis of epilepsy who treated with phenobarbital were included in this study, in Rasht 17th Shahrivar University Hospital, Iran. Abnormal neurological exam, abnormal brain CT and/or MRI, hepatic or renal dysfunction, thyroid disease or endocrinopathy, chromosomal abnormality, uncontrolled seizures, and multi-drug therapy were as exclusion criteria. Thyroid hormones were measured before and 6 months after initiation of drug. Demographic data, as well as type, duration, and family history of seizure were recorded and analyzed.

Results: The mean age of patients were 65.55 ± 54.60 months; Thirty patients (57.7%) were boys and 22(42.3%) were girls. Mean T3 level was 3.09 ± 0.70 before and 1.70 ± 0.30 pg/ml after phenobarbital (p<0.001); as well as mean T4 level 12.03 ± 2.40 before and 7.10 ± 0.90 ug/dl after (p<0.001), mean T3RU level 1.40 ± 0.70 before and 0.40 ± 0.40 percent after (p<0.001), and mean TSH level 1.41 ± 0.82 before and 4.63 ± 1.86 mU/l after phenobarbital usage (p<0.001). Gender and age of patients could not influence in this alteration.

Conclusion: It seems that phenobarbital can significantly change thyroid hormones in pediatric patients.

doi:10.1016/j.jns.2015.08.673

603 WFN15-1516 Miscellaneous Topics 2 Profile of patients who have died with multiple sclerosis in Brazil

<u>E. Cardoso de Sousa</u>, I.L. Leite Pereira, D.H. Vieira Vilaça, I.F. Barbosa, Alencar e Silva, K. Dantas de Sousa Torquato, W. da Silva Queiroz, R. Farias Tavares, E. Freires Marques, P.K. de Figueiredo Medeiros, L. Costa Reis, O. Duarte Leite, J. Tavares Santos, A.M. de Carvalho Pereira. *Bacharelado De Medicina, Faculdade Santa Maria, Cajazeiras, Brazil*

Multiple sclerosis (MS) is the most common cause of chronic neurological disability in young adults. We evaluate, through retrospective study, epidemiological characteristics, the patient profile of dying with multiple sclerosis in 170 patients. This pathology has its distribution more common breed in white, the gender distribution is more common in women and the relationship with age is rare in adolescence and higher than 60 years and is very common at 30 years old. Data were collected from the DATASUS Health Information (Tabnet). Multiple sclerosis morbidity was selected. An analysis was made between the years 2009 and 2013, the variable sex, age and race. For analysis techniques were used descriptive statistics getting the absolute frequency, using the statistical program SPSS18. Total number of deaths in the past five years were 170 cases, of these, 81 (47.648%) male and 89 (52.352) female. The number of deaths by age group is given as follows: 10-14 years 1 case (0.588%), 15-19 years 1 (0.588%), 20 to 29 years, 8 (4,705 %), 30-39 years old, 17 (10%), 40-49 years old, 32 (18.823%), 50-59 years old, 36 (21.117%), 60-69 years 28 (16.470%), 70-79 years old, 26 (15.294%) and over 80 years, 21 cases (12.415%). Deaths from whites race, 80 cases (46.058%), black, 4 (2.301%), Brown, 35 (20.288%), yellow, 3 (1.664%), unreported, 51 (30%). Note is a

concordance of the results with the literature, although the age group that has the largest number of deaths and between 50 to 59 years.

doi:10.1016/j.jns.2015.08.674

604

WFN15-0719 Miscellaneous Topics 2 Autoantibody-associated autoimmune-encephalitis in Sri Lankan patients

<u>T. Chang</u>^a, T. Moloney^b, L. Jacobson^b, N. Malavige^c, J. Lohitharajah^d, S. Peach^b, M. Woodhall^b, P. Waters^b, A. Vincent^b. ^aClinical Medicine, Faculty of Medicine University of Colombo, Colombo, Sri Lanka; ^bNuffield Department of Clinical Neurosciences, University of Oxford, Oxford, United Kingdom; ^cMicrobiology, Faculty of Medical Sciences University of Sri Jayawardenapura, Nugegoda, Sri Lanka; ^dFaculty of Medicine, University of Colombo, Colombo, Sri Lanka

Background: Autoantibody-associated autoimmune-encephalitis has emerged as the main potentially treatable cause of encephalitis, but remains under-diagnosed. The prevalence of autoimmune-encephalitis in Sri Lanka is yet unknown.

Objective: To determine the frequency and characteristics of autoimmune-encephalitis among patients presenting with encephalitis. **Patients and methods:** Sera and CSF from 105 consecutive patients with a clinical diagnosis of encephalitis/meningoencephalitis admitted to two tertiary-care hospitals in Colombo, Sri Lanka were tested on primary embryonic mouse hippocampal neuronal cultures for surface-protein-binding antibodies and on cell-based assays for NMDAR, LGI1, Caspr2 and Contactin2 antibodies. Selected sera with neuronal and glial surface-binding antibodies were tested on cell-based assays for AMPAR, GABA_AR, GABA_BR, aquaporin-4 and MOG antibodies.

Results: Patient age ranged from 1 month to 73 years (median=18 vears; mean=25.5;SD=21.4) with a male:female ratio of 1.76:1. Sera of 4 patients showed high-positive neuronal staining while 5 showed low-positive neuronal staining. Of the patients showing neuronal staining, one (3.5-year-old female) was positive for NMDAR and one (59-year-old male) was positive for GABA_BR antibodies. Serum of one patient (19-year-old male) showed a glial pattern of staining and was positive for GABA_BR antibodies. Of patients who had no neuronal or glial staining, one (58-year-old male) was positive for Caspr2 and another (39-year-old male) was positive for NMDAR antibodies. In total, 1 patient had Caspr2antibodies (0.95%), 2 had NMDAR-antibodies (1.90%) and 2 had GABA_BR-antibodies. Screening for bacterial and viral aetiologies of meningoencephalitis was negative. Clinical features, brain imaging, EEG and CSF analysis were not suggestive of any specific aetiology. Conclusion: Autoimmune-encephalitis accounted for 4.76% of patients presenting with encephalitis.

doi:10.1016/j.jns.2015.08.675

605

WFN15-1551

Miscellaneous Topics 2

Using quality improvement methods to change clinical practice: a case example of multidisciplinary approach to tuberous sclerosis management

<u>M. Ciobanu</u>, A. Railean, M. Wong, M. Silvia. *Neurology, Wake Forest University School of Medicine, Winston Salem, USA*

Objective: Tuberous sclerosis complex (TS) is a highly variable inherited condition presenting a relentlessly progressive course over the lifetime of an affected individual. A TS program was designed at Wake Forest Medical Center in 2013 to improve patient care. We identified quality improvement (QI) practice changes and implemented a multidisciplinary approach that focuses on surveillance, anticipatory and preventive measures to address the primary and secondary aspects of the disorder.

Methods: As part of a QI initiative to develop a multidisciplinary TS clinic for achieving the surveillance and management recommendations of the 2012 International TS Consensus Conference, we retrospectively reviewed the charts of 22 patients. Longitudinal clinical data from electronic medical records were included in the study. Surveillance adherence was compared in the cohort.

Results: There were 22 patients (8 females and 14 males) enrolled in our clinic since June of 2013. The age ranged from 2 months to 15 years, with a median of 6 years and a mean of 3 years. Six patients had TSC 2 and two patients had TSC 1 mutations. Three patients had negative testing for both TSC 1 and TSC 2 mutations. Only 12/22 (55%) children were partially meeting surveillance guidelines. Only 6/22 (27%) patients received longitudinal dermatological and dental care.

Conclusions: Morbidity and treatment burden of TS manifestations is significant, suggesting substantial economic and humanistic burden for the families and society. Medical care is thus best provided by a team of medical professionals working closely together in an interdisciplinary fashion.

doi:10.1016/j.jns.2015.08.676

606

WFN15-0609 Miscellaneous Topics 2 The Brunei epidemiological study in stroke and multiple scelrosis (best) - a door-to-door survey

<u>U. Meyding-Lamadé</u>^a, E. Craemer^a, A. Lupat^b, M. Luissin^b, B. Bassa^c, J. Hengelbrock^d, H. Becher^d. ^aDepartment of Neurology, Krankenhaus Nordwest, Frankfurt am Main, Germany; ^bPARPS Institute of Health Science, Universiti Brunei Darussalam, Jalan Tungku, Brunei; ^cDepartment of Neurology, Krankenhaus Nordwest, Frankfurt, Germany; ^dInstitute for Med. Biometry and Epidemiology, Universitätsklinikum Hamburg-Eppendorf, Hamburg, Germany

Background: Stroke is a major disease at the present time. The expected increase in the number of stroke patients will require substantial changes in health care services unless the incidence of stroke decreases in the future.

Multiple Sclerosis (MS) is a chronic, inflammatory, demyelinating disease of the CNS. MS signs can range from relatively benign to disabling and devastating symptoms, as communication between the brain and other parts of the body is disrupted.

The information on incidence, prevalence and mortality of stroke and multiple sclerosis is of utmost importance.

Aim: BEST will set a milestone in stroke and MS studies in South-East-Asia; the study aims to determine the prevalence of stroke risk factors and other neurological diseases. Those data are crucial for adapting treatment options and rehabilitation facilities in Brunei Darussalam.

Methods: This door-to-door survey will be a descriptive epidemiological investigation. Such studies require data on the population and its subgroups, and statistical methods like the calculation of confidence intervals for prevalence, incidence and mortality of stroke and Multiple Sclerosis.

Results: Here we report preliminary findings from the first 1600 participant whereby the data is used and analysed to examine the

ARTICLE IN PRESS

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

risk factors and incidence of stroke and MS.Initial demographic finding shows that 44% are male and 56% are female with the age range from 20 to 50 years old

Conclusion: BEST will provide a data basis for planning and establishing adequate resources to help patients and eventually be helpful for the development of treatment of stroke and multiple sclerosis.

doi:10.1016/j.jns.2015.08.677

607 WFN15-1439 Miscellaneous Topics 2 Stroke in México: mortality trends 1980-2012

<u>C. Cruz</u>^a, C. Hernandez^b, S. Parra^b, J. Campuzano^c, J. Calleja^d, A. Hernandez^c. ^aD Sc Student, Mexico Public Health School Mexico City, Mexico; ^bChronic Disease Division, National Public Health Institute, Mexico City, Mexico; ^cReproductive Health and Chronic Disease Division, National Public Health Institute, Mexico City, Mexico; ^dNeurology Investigation, Neurosurgery and Neurology National Institute, Mexico City, Mexico

Worldwide, stroke is a significant cause of mortality, disability and burden of disease. In the last decades, the stroke mortality rates have declined in high-income countries. However, stroke mortality has increased in low-and-middle-income countries.

Accordingly, the objective of this study was to provide information of changes in the stroke mortality trend from 1980 to 2012, in Mexico a low-and-middle country.

Methods: Age-adjusted mortality rates were computed for period 1980-2012 for all strokes.

The data were obtained from Health Information National System (HINS) and Population Census in Mexico. The cause of death from stroke was determined using the International classification of diseases (ICD), 9th Revision (ICD-9) codes 430-438 and 10th Revision (ICD-10) codes 160.0-169.8.

The results were analyzing by sex, type stroke and age groups.

The computer program STATA 13.0 was used to calculate ageadjusted mortality. Also Jointpoint analysis was used to identify changes in trends. The change of mortality trend is showed like annual percentage change (APC), α <0.05.

Results: The stroke age-adjusted mortality rates in Mexico, in both sexes decreased moderately between 1980 and 2012.

The average APC to ischemic stroke in the same period was – 3.8 points, <0.05. Additionally hemorrhagic stroke was -0.5 APC, <0.05.

The average APC -0.7 points was in Intracerebral hemorrhage while subarachnoid hemorrhage was APC 1.6 points, <0.05

In Mexico the stroke mortality trend by pathological type: ischemic stroke, hemorrhagic stroke and intracerebral hemorrhage decreased between 1980 to 2012. However differences between sex and age group exist. Also subarachnoid hemorrhage age-adjusted mortality rates increased.

doi:10.1016/j.jns.2015.08.678

608

WFN15-0197 Miscellaneous Topics 2 Enilency prevalence in children and adoless

Epilepsy prevalence in children and adolescents aged 0 -19 years – a door-to-door study

J.S. Dalbem^a, A.S. Dalbem^b, H.H. Siqueira^a, R.M.P. Alvarenga^c, M.E.C. Andraus^c. ^aMedicine School, Federal University of Mato Grosso, Cuiabá, Brazil; ^bMedicine School, Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo, Brazil; ^cMedicine School, Federal University in the State of Rio de Janeiro, Rio de Janeiro, Brazil

Introduction: Epilepsy is a cronic disease that affects 50 million people in the world. The clinical manifestations of epilepsy are similar in developed and developing countries, but the dimension with which patients are diagnosed, investigated and treated is differente.

Objective: Determine the prevalence of epilepsy in children and adolescents aged 0-19 years in the city of Barra do Bugres, Mato Grosso, from August 2012 to July 2013.

Metodology: Cross-sectional study, door-to-door, performed in two phases.

Results: The prevalence of epilepsy was 8.02/1000 population (95% CI: 6.50/1000, 9.79/1000). There was a predominance in male. The first seizure occurred in adolescence in 45,83% of the cases. Generalizes seizures were the most frequente. The most used antiepileptic drugs were fenobarbital and carbamazepine. Family history of epilepsy in first degree relatives was present in 42.71 % of the cases. The presence of associated diseases was found in 60.42 % of patients, and the most frequent were cerebral palsy and mental retardation.

Conclusions: The prevalence of epilepsy in children aged 0-19 years in Barra do Bugres city was 8.02/1000 inhabitants. The improvement in primary care with a better pre and perinatal assitance and childcare follow-up can reduce the number of cases of structural/metabolic origin epilepsy, since 60.4 % of the patients had associated diseases such as cerebral palsy and mental retardation, usually due to complications in the pre/perinatal period.

doi:10.1016/j.jns.2015.08.679

609 WFN15-1314 Miscellaneous Topics 2 Paraneoplastic CNS lesions secondary to multiple myeloma: a report of 2 cases

P.S. Dhawan, B.P. Goodman. Neurology, Mayo Clinic AZ, Scottsdale, USA

Background: CNS involvement of multiple myeloma (MM) is rare and involves presence of cranial plasmacytoma or myelomatous meningitis. Literature review reveals three cases of suspected MMassociated paraneoplastic leukoencephalopathy.

Objective: We present two patients with relapsing-remitting CNS lesions, ultimately diagnosed with MM. Absence of direct disease and improvement with treatment suggests paraneoplastic etiology.

Case 1: A 64 year-old man presented with right hemibody pain, with recent transient neck stiffness and later diplopia. Examination showed right-sided reduction in pain perception. Brain MRI showed acute (enhancing) and chronic bihemispheric white matter abnormalities. No leptomeningeal enhancement. Cord, CSF and paraneoplastic evaluation was unremarkable. Workup revealed IgG-kappa monoclonal gammopathy, elevated creatinine, vertebral compression fractures and 61% kappa light chain-restricted plasma cells on marrow biopsy. The patient was treated with CyBorD and stem cell transplantation. CNS lesions progressed until transplantation, then reduced. The pain syndrome stabilized.

Case 2: A 67 year-old man presented with left hemiparesis. He recently had three episodes of right hemiparesis associated with brainstem lesions, from which he recovered. Examination revealed left hemiparesis, spastic-ataxic dysarthria, right hemiataxia, spastic-ity and hyperreflexia. Brain MRI showed brainstem, internal capsule and callosal T2-hyperintensities without contrast enhancement. Cord, CSF and paraneoplastic evaluation was unremarkable. Workup revealed lytic lesions, IgA-lambda monoclonal protein and 16% lambda light chain-restricted plasma cells on marrow biopsy. Modest improvement in MRI findings and hemiparesis occurred with CyBorD treatment.

Conclusion: Evaluation of atypical CNS lesions should include investigation for lytic bone lesions or monoclonal protein. Paraneoplastic leukoencephalopathy can be associated with MM.

doi:10.1016/j.jns.2015.08.680

610 WFN15-0155 Miscellaneous Topics 2 Changes in cerebral metabolism in a preclinical model of blast-induced traumatic brain injury

<u>A. Divani^a</u>, Y. Wang^b, A. Talan^a, A. Murphy^a, X.H. Zhu^c, W. Chen^c, M. Pomper^b. ^aNeurology, University of MN, Minneapolis, USA; ^bRadiology, Johns Hopkins University, Baltimore, USA; ^cRadiology, University of MN, Minneapolis, USA

Background: There are very few published studies looking at metabolic changes in blast-induced traumatic brain injury (bTBI).

Objectives: The objective of this study was to assess changes in brain metabolism over time as a consequence of bTBI using an animal model developed in-house.¹ We conducted PET studies with [¹⁸F]fluorodeoxyglucose (FDG).

Material and methods: We divided 12 rats into two bTBI severity groups: 1) bTBI with 5 shockwave (SW) pulses and 2) bTBI with 10 SW pulses. To induce bTBI, anesthetized rats were placed on a lithotripsy machine to deliver 5 or 10 SW pulses to the right side of the frontal cortex. Next, animals were placed in a prone position in a microPET scanner for 10 min static scan. PET and CT images were obtained at baseline followed by 3 hours and 3 days post-injury to compare metabolic changes in cerebral glucose uptake in different regions of the brain using FDG tracer.

Results: A trend of lower standardized uptake value ratios (SUVRs) were observed from all post-blast FDG-PET studies of the rat brains, particularly in the motor cortex and retrosplenial regions. While those rats receiving 5 SW pulses showed stable or recovering SUVR from acute (3 hours) to the sub-acute (3 days) phase, those receiving 10 pulses demonstrated even lower SUVRs at the sub-acute phase.

Conclusions: Our study suggests that bTBI might cause hypometabolism on the impact side of the rat brains. Different bTBI impact location and severity may result in differing affected brain regions and degree/duration of hypo-metabolism in these regions.

doi:10.1016/j.jns.2015.08.681

611 WFN15-0564 Miscellaneous Topics 2 Neurological disease surveillance in Cameroon. A rural and urban-based in/outpatient population study

<u>J. Doumbe^a</u>, Y. Mapoure^a, T. Nyinyikua^a, K. Kompoliti^b, H. Shah^c, E. Cubo Delgado^d. ^aclinical sciences, Faculty of medicine and pharmaceutical sciences, Duala, Cameroon; ^bMedical center, Rush university, Chicago, USA; ^cThe Neurological institute, Columbia university medical center, NY, USA; ^dNeurology, Hospital Universitario Burgos, Burgos, Spain

Background: There is paucity of literature on the burden of neurological diseases in sub-Saharan Africa.

Objective: To create a registry and surveillance of neurological diseases from urban and rural health centers in Cameroon, from 2012-to 2014. **Methods:** The records of out-and inpatients were reviewed. In the urban areas, the diagnosis was made by a neurologist but this was not the case in the rural areas. The following variables were analyzed: demographics, medical center characteristics, presenting neurological

complaint, medical history, neurological diagnoses, death and disability. Neurological diseases were classified according to ICD-10.

Results: Out of 20131 medical charts reviewed (13% from the rural area), 4187 cases with neurological complaints were identified (20.79%), mean age 48.67 \pm 18.62 years, females 54.7%. Overall, besides diagnosis, other rural neurological data was scarce. The most frequent neurological complaints were: paresis/weakness (G.82, 31.4%) and headache (R.51, 21.1%). Most common concurrent medical history was hypertension (I10, 25.3%), and HIV (B20, 10.5%). Most common neurological diagnoses were: cerebrovascular disease (G46, 30.6%), epilepsy (G40, 12.8%), and headache (G43, 12.1%). Death due to neurological cause was recorded in 428 patients (19.1%), and disability in 1072 (57.2%). Overall, patients from the rural area were more likely to be diagnosed with intracranial infection (G06), malaria (B50), and nutritional deficiency (E90).

Conclusion: The present study supports the fact that neurological disorders are frequent in Cameroon and cerebrovascular disease constitutes a medical and economic burden. Although data are limited, neurological disease pattern differs in rural areas. A health policy geared towards prevention is warranted.

doi:10.1016/j.jns.2015.08.682

612

WFN15-0767 Miscellaneous Topics 2

Pediatric neurological disease surveillance in Cameroon. An urban-based outpatient population study

H. Shah^a, J. Doumbe^b, Y.N. Mapoure^b, T. Nyinyikua^b, K. Kompoliti^c, M.E. Cubo Delgado^d. ^aDepartment of Neurology, Columbia University, NY City, USA; ^bDepartment of Neurology, University of Douala, Douala, Cameroon; ^cDepartment of Neurology, Rush University Medical Center, Chicago, USA; ^dDepartment of Neurology, Hospital Universitario Burgos, Burgos, Spain

Background: There is limited information available regarding the frequency of pediatric neurological disorders in sub-Saharan Africa. We report here the rates of neurological disease among the pediatric population cared for at urban health care districts.

Objective: To record the morbidity and mortality resulting from pediatric neurological diseases using a registry and surveillance of neurological diseases from urban health care districts in Cameroon from 2012 to 2014.

Methods: The records of patients from 2 urban outpatient sites were reviewed. The following variables were analyzed: demographics, presenting complaint, neurological diagnosis, and medical history. Diagnoses were made by history, examination and investigations, as indicated. Neurological diseases were classified according to the ICD-10. Results: A total of 125 pediatric cases out of a total of 2237 charts were identified (6% of all cases) in an urban setting. 12 (9.6%) were between ages 0-2, 26 (20.8%) were between ages 3-5, 45 (36%) were between ages 6-10, and 42 (33.6%) were between ages 11-14. The most frequent neurological complaints were seizure (33.6%), headache (12.8%), any hyperkinetic movement (12%), and mental status change (8%). The most frequent neurological disorders were epilepsy (66.4%), headache (5.6%), and cerebral palsy (4.8%). Among infants (ages 0-2) and toddlers (ages 3-5), the most common neurological complaint was seizure (33.3% and 69.2% respectively) and most common neurological diagnosis was epilepsy (33.3% and 84.6% respectively). Conclusions: Pediatric neurological disorders are a major cause of

Conclusions: Pediatric neurological disorders are a major cause of morbidity in childhood in Cameroon with the most common neurological disorder being epilepsy.

doi:10.1016/j.jns.2015.08.683

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

613 WFN15-1601 Miscellaneous Topics 2 Neurological manifestations of Behçet's disease: 161 cases in one moroccan center

<u>S. AIDI</u>, M. Benabdeljlil, M. Rahmani, D. Sefiani, <u>M. El Alaoui Faris</u>. Departement of Neurology and Neuropsychology, Hôpital des Spécialités, Rabat, Morocco

Introduction: Behçet's disease (BD) is a chronic multisystemic inflammatory disease, predominant within the Mediterranean countries and characterized by oral, genital mucous ulcers and uveitis. Neurological involvement is one of the most serious complications of BD and represents a major cause of morbidity.

Patients and methods: 161 moroccan cases of neurobehçet disease (NBD) have been categorized into two main groups: "Parenchymal" CNS involvement, which includes hemispheric, brain-stem, spinal, and multifocal presentations; and "non parenchymal" CNS involvement, which includes dural sinus thrombosis and arterial involvement.

Results: Parenchymal CNS pattern was observed in 74% of our cases with a predominance of symptoms related to brainstem involvement (30%) such ophthalmoparesis, bulbar and pseudobulbar palsy. Other symptoms have been present with varying frequencies: cerebellar signs(52%), headache(52%), psychiatric and cognitive symptoms (34%), sphincter disturbances (22%), epilepsy (9%). Some features are less common such as optic neuropathy (2%) or peripheral neuropathy(1%).

Non-parenchymal form was noted in 26% of our series essentially represented by cerebral venous thrombosis (CVT). Only 4 of cases have cerebral arterial involvement, three with intracranial aneurysm.

All patients have cerebral imaging. Magnetic resonance imaging showed a caracteristic lesion in one third of our patients, located at the mesodiencephalic junction and the pontobulbar region.

Treatment was based on corticosteroids in the two categories. Long-term maintenance with immunosuppressive agents was administrated in patients with parenchymal CNS involvement and anticoagulants in case of CVT.

Conclusion: Neurological involvement represents a major complications of BD which requires early diagnosis and long terme management.

doi:10.1016/j.jns.2015.08.684

614

WFN15-1318 Miscellaneous Topics 2 Age of independent sitting posture acquisition for children with myelomeningocele

M.R.C. Sá^a, C.T. Ribeiro^a, F.G. Fracho^a, A.S. Ferreira^b, M. Xavier^c, V. Guedes^c, M.F. Cunha^c, R. Custódio^c, <u>A.C.A.F. Ferreira^c</u>, M. Orsini^d. ^aPediatrics, Instituto Fernandes Figueira, Rio de Janeiro, Brazil; ^bRehabilitation Science, Centro Universitário Augusto Motta – UNISUAM, Rio de Janeiro, Brazil; ^cNeurology, Universidade Federal Fluminense, Rio de Janeiro, Brazil; ^dRehabilitation Science, Universidade Federal Fluminense/Centro Universitário Augusto Motta – UNISUAM, Rio de Janeiro, Brazil

Objective: To study the acquisition of independent sitting posture in patients with thoracic MMC and high lumbar.

Method: Cross-sectional study in children aged between 6 months and 4 years diagnosed with thoracic and high lumbar MMC. Data was collected from medical records.

Results: There were 16 children with MMC, 9 (56%) female. Regarding the type of MMC, there were 13 (81.2%) children with high lumbar level, and 3 (18.8%) with thoracic level. The most common comorbidities were: Arnold Chiari II Malformation, hydrocephalus and neurogenic bladder. Thirteen (81.2%) achieved the independent sitting posture. The average to reach this posture was 16 months (9-31 months). The average starting time of the physiotherapeutic monitoring in the physical therapy sector was of 3 months (1-8 months). From the 13 children who sat, 2 (15.4%) were thoracic level and 11 (84.6%) were high lumbar level. Most of the sample (15 = 93.8%) performed regular physical therapy. The average age to start the physical therapy was of 3 months (1-8 months).

Discussion: It was observed that most part of the sample acquired independent sitting posture, including thoracic levels that reached this milestone with age that was less than the one found in the literature. **Conclusion:** The results can be considered positive, adding the possible contribution for the early physiotherapy intervention in the acquisition of motor milestones in the promotion of functionality and in the quality of life improvement for children with MMC

doi:10.1016/j.jns.2015.08.685

615

WFN15-0186 Miscellaneous Topics 2 Influence of visceral adiposity on cardiovascular autonomic function in type 2 diabetes

<u>R. Goit</u>. Department of Physiology, Nepalgunj Medical College Teaching Hospital, Koholpur, Nepal

Patients with type 2 diabetes have two to four fold increase risk of developing cardiovascular diseases, which represent their leading cause of mortality. Heart rate variability (HRV), which is used as an assessment of the cardiac autonomic function, is frequently reduced in type 2 diabetes. The autonomic nervous system is an important determinant of the regulation of both the cardiovascular system and energy expenditure, and it is assumed to play a role in the pathophysiology of obesity. The aim of this study was to determine the influences of visceral adiposity on cardiovascular autonomic functions in type 2 diabetes. One hundred twenty patients with type 2 diabetes participated in this study. Anthropometric, metabolic parameters, visceral fat area, and HRV were measured in all the patients. Correlation between the visceral fat area and indices of HRV were evaluated. The patients were 48.23 \pm 4.7 years old with body mass index (BMI) of $25.1 \pm 1.2 \text{ kg/m}^2$, waist circumference (WC) of 101.67 \pm 4.2 cm and duration of diabetes 14.3 \pm 7.3 years. BMI and WC were inversely correlated with low frequencies. The visceral fat area correlated negatively with standard deviation of all RR intervals (SDNN), the square root of the mean of the sum of the squares of differences between adjacent RR intervals (RMSSD), percentage of consecutive RR intervals that differ by more than 50 ms (pNN50) and with low and high frequencies. These data suggest that increase in visceral adiposity contributes to an autonomic imbalance, as demonstrated by lower HRV in patients with type 2 diabetes.

doi:10.1016/j.jns.2015.08.686

616 WFN15-0729 Miscellaneous Topics 2 Epidemiology and disability due to Parkinson`s disease in Ukraine (12 years experience)

<u>V. Golyk</u>^a, T. Mischenko^b, A. Ipatov^a, N. Gondulenko^c, A. Pivnyk^d, V. Pogorelova^a, S. Sliva^a. ^aNeurology and Border States, Ukrainian State Institute of Medical and Social Problems of Disability (MoPH), Dnipropetrovs k, Ukraine; ^bCerebrovascular pathology, Institute of Neurology Psychiatry and Narcology (Academy of Medical Sciences),

6

Kharkov, Ukraine; ^cInternal Diseases, Ukrainian State Institute of Medical and Social Problems of Disability (MoPH), Dnipropetrovs `k, Ukraine; ^dRehabilitation, Ukrainian State Institute of Medical and Social Problems of Disability (MoPH), Dnipropetrovs `k, Ukraine

Parkinson's disease (PD) is major extrapyramidal disorder worldwide and the main part of parkinsonism syndrome with prevalence rates ranged 108-257 per 100000 inhabitants.

We investigated dynamics of PD (IDC X G20, G21) prevalence and disability incidence rates in Ukraine for 12 years (2003-2014, per 100000, per region). Disability status in Ukraine is set in case of at least grade 2 ICF domain impairment. The systemic mistake in Ukrainian neurology is widespread practice of false positive "vascular parkinsonism" ("chronic cerebrovascular insufficiency") diagnostics, followed epidemiological data incorrectness and inadequate neurological healthcare planning (including pharmacological treatment and social support).

PD prevalence increased 44,4–48,1–50,5–51,2–54,1–56,0–58,1– 59,6–61,4–63,7–65,0–63,0 consequently showing local neurologists PD awareness increasing. Surprisingly, PD disability incidence rates were gradually decreased: 1,9–2,0–1,9–1,5–1,7–1,5–1,4–1,3–1,3– 1,4–1,3–1,2 consequently. Highest prevalence rates in 2014 were detected in Kyiv city (111,6), Vinnitsa (126,1), L'viv (109,5), Kyiv (102,4) and Cherkassy (90,0) regions where movement disorders centres located. Highest primary disability rates (2014) observed in Chernihiv (2,4), Ternopol'(1,9), Zaporizhya (1,8) regions and Kyiv (2,1) city. Lowest PD prevalence rates in 2014 detected in Chernivtsi (39,1), Odessa (39,9) and Kirovograd (41,8) regions. Lowest PD disability incidence rates were observed in terms of primary disability rate: Kherson (0,2), Khmelnitsky (0,4), Cherkassy (0,6) regions.

Revealed data are the subject of further primary educational and awareness corrections both in clinical and medical-social expertise settings.

doi:10.1016/j.jns.2015.08.687

617 WFN15-0062 Miscellaneous Topics 2 Head-up tilt table test in differentiating neuropathic versus

L. Crnosija, M. Krbot Skoric, I. Adamec, M. Lovric, A. Junakovic, A. Mismas, V. Miletic, R. Sprljan Alfirev, A. Pavelic, <u>M. Habek</u>. *Neurology, University Hospital Centre Zagreb, Zagreb, Croatia*

hyperadrenergic postural orthostatic tachycardia syndrome

Background: Measuring standing plasma norepinephrine levels is standard way of differentiating between postural orthostatic tachy-cardia syndrome (POTS) subtypes.

Objectives: To investigate differences between neuropathic and hyperadrenergic POTS in hemodynamic response to head-up tilt, and to test if those differences could be used to effectively differentiate between POTS subtypes.

Patients an methods: Forty-three consecutive POTS patients underwent the head-up tilt table test (HUTT) consisting of a 10-minute supine phase and 30-minute 70° tilted phase. Patients were categorized into hyperadrenergic (group H) and neuropathic group (group N) according to standing norepinephrine values. Heart rate, systolic and diastolic blood pressure, and heart rate variability parameters of the two groups were compared.

Results: 10 patients had hyperadrenergic, and 33 had neuropathic type POTS. Using the following criteria neuropathic POTS can be differentiated with 76% sensitivity and 80% specificity: tilted phase endurance (TPE) of less than 14 minutes (TPE was 21.2 ± 3.5 min in group H, compared to 8.9 ± 6.8 min in group N (p=0.001)), or drop

of average dBP in the 1st minute of the tilted phase compared to average dBP during supine phase (mean dBP difference was significantly lower in group N compared to group H (-1.7 \pm 6.9 bpm vs. 5.6 \pm 7.46 bpm, p=0.007). When looking into heart rate analysis, group H showed a greater degree of sympathetic cardiac activity during the supine phase and during first 5 minutes of tilted phase.

Conclusion: Hyperadrenergic and neuropathic type of POTS can be effectively differentiated based on their differences in response to HUTT.

doi:10.1016/j.jns.2015.08.688

618 WFN15-0142 Miscellaneous Topics 2 Autonomic dysfunction in clinically isolated syndrome suggestive of multiple sclerosis

L. Crnosija, I. Adamec, M. Lovric, A. Junakovic, M. Krbot Skoric, <u>M. Habek</u>. *Neurology, University Hospital Centre Zagreb, Zagreb, Croatia*

Background: Autonomic nervous system has never been systematically investigated in patients with an initial demyelinating event suggestive of MS (clinically isolated syndrome - CIS).

Objectives: The aim of this study was to determine the extent of autonomic dysfunction in patients with clinically isolated syndrome (CIS) by using standardized battery of autonomic tests in the form of the composite autonomic scoring scale (CASS).

Methods: This was a prospective, cross sectional study which included 24 consecutive patients who were diagnosed with CIS and 17 healthy controls. In all participants, heart rate and blood pressure responses to the Valsalva maneuver, heart rate response to deep breathing and blood pressure response to passive tilt were performed. In 16 patients, Quantitative Sudomotor Axon Reflex Test (QSART) and catecholamine measurement was performed.

Results: The proportion of CIS patients with pathological adrenergic index was statistically significant higher compared to healthy controls (12 vs 2, p=0.018), while there was no difference in cardiovagal index between groups. Five patients had a sudomotor index of 1 (in 4 there was hypohydrosis < 50% and in 1 persistent foot hyperhydrosis). When combining adrenergic, cardiovagal and sudomotor index into CASS, 8 patients (50%) had evidence of autonomic dysfunction, 7 mild and one moderate. There was no difference in catecholamine levels between patients with or without sympathetic nervous system dysfunction.

Conclusion: CASS is able to detect autonomic nervous system dysfunction in CIS patients, in whom the sympathetic nervous system is frequently affected.

doi:10.1016/j.jns.2015.08.689

619

WFN15-1552 Miscellaneous Topics 2

Fatal paraneoplasic encephalo-myelo-neuropathy associated with anti-Hu antibodies: a case report

<u>Y. Habtany</u>, M.A. Rafaii, F.Z. Mouni, H. El Otmani, B. El Moutawakkil, I. Slassi. *Neurology, 20 August Hospital IBN ROCHD university Hospital, Casablanca, Morocco*

Background: Paraneoplasic neurological syndromes are rare. The association between a central and peripheral location is particularly infrequent.

ARTICLE IN PRESS

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

Objective: Report the case of central and peripheral location in a fatal paraneoplasic syndrome with anti-Hu antibodies, revealing a bronchial tumor.

Case report: We report the case of a 61 years old man patient, admited for a limbic encephalitis syndrome, complicated with an amyotrophic tetraparetic condition. MRI showed hippocampal lesions, and eletromyography showed an anterior horn syndrome. Further investigations discovered positive anti-Hu antibodies and a bronchial neoplasm.

Conclusion: The association of multiple neurological paraneoplasic syndromes in the same patient is possible, but rare. The authors illustrate a documented case of an «encephalo-myelo-neuropathy» with anti-Hu antibodies that revealed a bronchial neoplasm.

doi:10.1016/j.jns.2015.08.690

620

WFN15-1542 Miscellaneous Topics 2 Multiple severe and isolated cranial nerves involvement in childhood: idiopathic or CIDP localized form? Case report

<u>A. Hazim</u>, M.A. Rafai, N.A. Camara, B. El Moutawakil, H. El Otmani, B. Slaoui, I. Slassi Sennou. *Neurology, CHU IBN ROCHD, Casablanca, Morocco*

Introduction: Childhood chronic inflammatory demyelinating polyradiculoneuropathie is rare and difficult to diagnose, however, some clinical, electrophysiological and evolutive aspects are helpful for a better distinguishing of this uncommon disease.

Objectives: Reporting CDIP case in pediatric patient revealed by multiple cranial nerves impairment

Case report: We report a 4 year old child with no family history of neuropathy, referred for a rapidly progressive bilateral facial diplegia with chewing trouble and decreased tongue mobility, associated to a converging strabismus. This symptomatology evolved for over than 60 days.

Clinical examination revealed cranial nerve impairment of V, VII, III (Partial) and XII (bilateral) with an oval aspect of the mouth, without other associated anomalies.

Brain and spinal cord MRI was normal. The study of CSF highlighted a mild hyperproteinorachie (protein level at 0.5g/l). Inflammatory, immunological and infectious explorations were negative.

Facial nerve conduction velocity and blink reflex studies showed abnormalities of distal latency and R1R2 responses with severe neurogenic graph in EMG of facial and trigeminal and hypgloss muscle. The patient presented a slight improvement after immunoglobulin injections.

Discussion and conclusion: Given the clinical presentation, progressive profile, the negativity of the biological explorations and the electrophysiological features, cranial CDIP was discussed by analogy to acute polyneuropathy. To the best of our knowledge, this is the first description of a cranial childhood CDIP.

doi:10.1016/j.jns.2015.08.691

621

WFN15-0328 Miscellaneous Topics 2 Long-term outcome of chronic progressive neurological manifestations in behcet's disease

<u>S. Hirohata</u>^a, H. Kikuchi^b, T. Sawada^c, H. Nagafuchi^d, M. Kuwana^e, M. Takeno^f, Y. Ishigatsubo^f. *aRheumatology and Infectious Diseases, Kitasato University School of Medicine, Sagamihara, Japan; ^bInternal*

Medicine, Teikyo University School of Medicine, Tokyo, Japan; ^cInternal Medicine, Tokyo Medical University, Tokyo, Japan; ^dInternal Medicine, St. Marianna University School of Medicine, Kawasaki, Japan; ^eInternal Medicine, Keio University School of Medicine, Tokyo, Japan; ^fInternal Medicine, Yokohama City University School of Medicine, Yokohama, Japan

Purpose: Chronic progressive nuuro-Behcet's disease (CPNBD) is characterized by progressive deterioration leading to disability with or without previous attacks. The present study was designed to explore the effects of various treatment regimension the prognosis of patients with CPNBD.

Methods: Thirty-seven patients, who met the international classification criteria for BD, and developed CPNBD after 1988, were followed up until October 2013. The effects of various treatment regimens on prevention of death or severe disability of bedridden state were examined by Kaplan-Meier analysis and Cox's proportional hazard model.

Results: In 37 patients with CPNBD, 28 patients (75.7%) received MTX. Among the 28 patients with MTX, no patients died and only 5 patients progressed to disability with bedridden state. By contrast, among the 9 patients without MTX, 5 patients died and 3 patients progressed to the bedridden state. Thus, MTX significantly improved the survival of patients with CPNBD (HR 0.0507, 95% CI: 0.0077-0.334, p=0.020), but any of steroid pulse, azathioprine or cyclophosphamide did not. MTX also significantly reduced the proportion of the patients who were progressed into the bedridden state or death (HR 0.0694, 95% CI: 0.0047-0.7327, p=0.0258). Of note, CSF IL-6 levels, examined just before the final follow-up assessment, were significantly higher in more severely disabled patients.

Conclusion: These results indicate that MTX, but not high doses of steroids, azathioprine or cyclophosphamide, is effective to prevent the progression of CPNBD. Thus, it is recommended that MTX should be started as soon as possible the diagnosis of CPNBD is made.

doi:10.1016/j.jns.2015.08.692

622

WFN15-1296 Miscellaneous Topics 2

Ambulatory blood pressure profile and autonomic symptoms poststroke patients

<u>J. Idiáquez</u>^a, P. araya^b, J. Gigoux^b, G. Matamala^b. ^aNeurologia, Universidad de Valparaiso, Valparaiso, Chile; ^bNeurologia, Hospital Naval, Vina del Mar, Chile

Background: Autonomic symptoms are frequent in patients during poststroke period. Increased sympathetic and decreased parasympathetic cardiac function was reported.

Objective:

Patients and methods: In 20 hypertensive patients with symptomatic ischemic stroke we assess autonomic symptoms with a quantitative score SCOPA-AUT (scale for outcome in Parkinson disease) and 24-hr ambulatory blood pressure (BP) monitoring (AMBP). In 55 hypertensive patients without symptomatic stroke we assess autonomic symptoms.

Results: In stroke patients the AMBP showed that day time mean systolic BP was 137 \pm 22.8 mmHg and night time BP was 126.8 \pm 26.8 mmHg, The difference between day and night BP was not different (P = 0.4). Cardiovascular autonomic symptoms score for stroke patients was 2.4 \pm 2 and for the subjects without stroke was 1.1 \pm 1.5 (P $^{\circ}$ 0.05).

Conclusion: During poststroke period patients showed no significant difference in circadian systolic BP and showed increased day time

cardiovascular autonomic symptoms. The combined use of 24-hr AMBP combined with a quantitative assessment of autonomic symptoms can be a tool for management.

doi:10.1016/j.jns.2015.08.693

623

WFN15-1244 Miscellaneous Topics 2 Adult-onset opsoclonus-myoclonus-ataxia syndrome: clinical features and diagnostic findings

<u>A. Jaureguiberry</u>, V. Ibarra, G. Moretta, C. Torres, R. Ceruzzi, S. Rodriguez-Perez, E. Reich. *Neurology, Sanatorio Julio Mendez, CABA, Argentina*

Background: Opsoclonus-myoclonus-ataxia (OMA) is a rare neurological disorder characterized by the presence of abnormal saccadic ocular movements, ataxia and myoclonus. OMA is an autoimmune disease probably caused by a dysfunction in Purkinje cells secondary to an altered humoral and cellular response. The most frequent etiologies in adults are paraneoplastic, idiopathic and parainfectious. **Objective:** to describe two cases of opsoclonus-mioclonus-ataxia.

Patients and methods: We reviewed the clinical history and diagnostic tests of two patients from December 2012 to April 2015. Case one: 32 year/old woman who, a week after a gastroenteritis, started suddenly with opsoclonus, ataxia and generalized myoclonic jerks. Case two: 60 year/old man who, after flu-like syndrome, started suddenly with generalized myoclonus, ataxia and opsoclonus. **Results:** Both patients were studied in the same way. In table one we show the results of diagnostic tests performed.

They were diagnosed as idiopathic OMA, received steroid therapy and had a good outcome.

Conclusion: As in the patients we describe, an important number of idiopathic cases are preceded by infectious symptoms without confirmation of an infectious cause. Even though both patients presented typical features of idiopathic OMA, the result of both PET scans showed cortical unilateral cortical hypometabolism without cerebellar involvement, which is not usual in this syndrome.

doi:10.1016/j.jns.2015.08.694

624

WFN15-0285

Miscellaneous Topics 2 Cognitive impairment in chronic progressive neuro-behçet's disease: comparative study of brainstem and hippocampus region using brain MRI

<u>H. Kikuchi^a</u>, S. Hirohata^b. ^aDepartment of Internal Medicine, Teikyo University School of Medicine, Tokyo, Japan, ^bDepartment of Rheumatology and Infectious Diseases, Kitasato University School of Medicine, Kanagawa, Japan

Background: We examined the volumes of hippocampus in order to determine the responsible lesions for neurobehavioral changes in chronic progressive neuro-Behçet's disease (CPNB).

Patients and methods: A total of 26 patients were studied, including 13 patients with CPNB (11 males and 2 females, age 51.2 \pm 12.1 [mean \pm SD]) and 13 patients with Behçet's disease without NB (non-NBD) (10 males and 3 females, age 54.4 \pm 11.4). The sagittal sections of T1-weighted images on brain MRI were obtained from each patient. Severity of gray matter loss in the hippocampal region and whole brain were investigated by the software for VSRAD (Eisai

Co., Ltd), calculating the indicators of the degrees of hippocampal region atrophy (HAI) and those of whole-brain atrophy (WBAI). The areas of midbrain tegmentum and pons (brainstem) were measured on mid-sagittal sections of T1-weightened images of brain MRI using image analysis software Image J (ver.1.45, NIH, U.S.).

Results: The VSRAD analysis showed that HAI was significantly increased in CPNB ($1.46 \pm 0.70 \pmod{5}$ compared with in non-NB (0.77 ± 0.40) (p=0.0016). Although less markedly, WBAI was significantly higher in CPNB (10.6 ± 5.0) than in non-NB (6.9 ± 1.7) (p=0.0240). Neither HAI nor WBAI was correlated with age. Whereas all the patients with CPNB showed brainstem atrophy, there was no significant correlation between HAI and the rate of brainstem atrophy.

Conclusions: These results indicate that hippocampus, in addition to brainstem, is a commonly affected lesion in CPNB, accounting for progressive neurobehavioral changes.

	Case 1	Case 2
Brain MRI	Normal	Normal
EBV, VZV, HSV PCR	Negative	Negative
HIV serology	Negative	Negative
Anti-Hu, anti-Yo, anti-Ri antibodies	Negative	Negative
Routine laboratory test and CSF analysis	Normal	Normal
Whole-Body FDG-PET scan	Right cortical	Left cortical
	hypometabolism	hypometabolism



doi:10.1016/j.jns.2015.08.695

625 WFN15-0113 Miscellaneous Topics 2 Children with the recurent headaches and migraine syndrome health care

<u>M. Knezevic-Pogancev</u>. Developmental neurology and epileptology, Child and youth halth care Institute of Vojvodina, Novi Sad, Serbia

The aim of the studu was to analyse children with recurent headaches, and migraine syndrome (as the most frequent), medical care.

ARTICLE IN PRESS

Investigations was carried out in the period from 1988 to 2014, on 24232 children aged 3 to 16 years, in nine towns of south part of Serbia - Vojvodina.

Recurrent headache was found with 27,46%, and migraine syndrome with 8,63% children. To a general practitioner in primary health care turn 92,6% of children with the migraine (97,7% of them with aura, 91,05% without aura and 89,3% with other migraine syndromes) and only 35,3% with recurrent nonmigraine headaches. Children with migraine headaches in 68,2% (76,8% with aura, 62,5% without aura and even 90,0% with other migraine syndromes) have other specialist check-ups. Children with migraine headaches in 16,7% cases are monitored and treated only by a general practitioner in primary health care, in 27,9% by a specialist (pediatrician, children`s neurologist or neuro-psychiatrist), and in 55,3% by a subspecialist (neuropediatrician, epileptologist). Children with headaches of different etiology are in 50,0% cases monitored and treated only by a general practitioner in primary health care.

Children with migraine headaches most often used analgetics (53,3%) and triptans (27.2%) to abort the attack; children with nonmigraine headaches used nonsteroidal anti-inflammatory drugs (63,5%) and analgetics (31,5%).

Children with recurent headache and migraine syndrome do have a good complete diagnostic medical care in Vojvodina, but not adecvate medication. The consuming of analgetics and unconventional therapy is unjustified, while the consuming of triptans is not high enough.

doi:10.1016/j.jns.2015.08.696

626

WFN15-0547 Miscellaneous Topics 2 Mortality and morbidity among hospitalized patients with neurological diseases in Cameroon

<u>K. Kompoliti</u>^a, J. Doumbe^b, Y. Mapoure^b, T. Nyinyikua^b, H. Shah^c, E. Cubo^d. ^aNeurology Movement Disorders, Rush University Medical Center, Chicago, USA; ^bNeurology, University of Douala, Douala, Cameroon; ^cNeurology Movement Disorders, Columbia University Medical Center, NY, USA; ^dNeurology, University Hospital of Burgos, Burgos, Spain

Background: There is paucity of systemically collected information on the morbidity and mortality (M&M) from neurological diseases in sub-Saharan Africa.

Objective: To record the M&M from neurological diseases in Cameroon from 2012 to 2014 using a registry and surveillance from urban and rural health care centers.

Methods: Patient records from 2 urban hospitals (out- and inpatients) and 2 rural clinics were reviewed. A neurologist made the diagnosis in the urban but not in the rural areas. Variables analyzed were: demographics, presenting complain, neurological diagnosis, medical history, medical center characteristics, M&M. Neurological diseases were classified according to ICD-10.

Results: Out of 20131 medical charts reviewed (13% rural setting), 4187 cases with neurological complaints were identified (53.5% inpatients, 46.5% outpatient). Death and disability data were collected from hospitalized patients only, N = 2237. Death from neurological disease was recorded in 427 patients (19.1%), and disability in 1072 (57.2%). The diseases resulting in the greatest M&M were G46 (cerebrovascular disease) 53.1%, and G04 (encephalitis) 17.6%. Among disabled patients, 21% had mild to moderate disability (able to perform some ADLs but needing help) and 25% were fully dependent. Death and disability were more common in older patients (p < 0.0001). Patients with low education were more likely to be fully disabled (p < 0.0001).

Conclusions: As expected, older patients with neurological diseases had more M&M. Morbidity was inversely associated with education, which given that cerebrovascular disease is by far the most common cause of morbidity, indicates the power of risk factor control in preventing neurological disability.

doi:10.1016/j.jns.2015.08.697

627

WFN15-1455 Miscellaneous Topics 2 Arterial ischemic stroke and diarrhea-associated hemolytic uremic syndrome in childhood

<u>M. López^a</u>, I. Huete^b, M. Hernández^a. ^a*Pediatría, Facultad de Medicina, Pontificia Universidad Católica de Chile, Santiago, Chile;* ^bRadiología, *Facultad de Medicina, Pontificia Universidad Católica de Chile, Santiago, Chile*

Background: Arterial ischemic stroke (AIS) due to diarrhea-associated hemolytic uremic syndrome (D + HUS) in childhood is an underreported complication in the course of the disease.

Aim: To describe neurological features at onset, neuroimaging, and 1-year neurologic disabilities of pediatric patients.

Patients and methods: We present three pediatric cases of D + HUS associated with AIS.

Results: Two boys and one girl, all previously healthy, median age was 18 months (range, 15 – 21). The median hematocrit, platelet count and glomerular filtration rates at onset were 27% $(range, 25 - 29), 51/mm^3$ (range, 16 - 72) and 22.6 ml/min/1.73 m² (range, 12.9 - 37.3), respectively. All patients have a negative detection of verotoxin-producing Escherichia Coli in stool samples by PCR. The median duration from diagnosis of D + HUS to the onset of neurological symptoms was 7 days (range, 3 - 13). Neurological manifestations are generalized seizures and impairment of consciousness, no focal signs were detected. CT an MR images showing extensive brain involvement in different locations, in one case, bilateral MCA vasculitis was demonstrated by conventional angiography (CONA). Neurologic disabilities were of varying severity, but present in all patients at year of follow-up. Permanent motor deficits, drug-resistant epilepsy and global delays in development were present in 3, 2 and 2 patients, respectively.

Conclusion: Despite good prognosis of neurologic complications in D + HUS, AIS is associated with long-term neurologic disabilities. CONA is a useful tool in diagnosis of AIS and vasculitis due to D + HUS.

doi:10.1016/j.jns.2015.08.698

628

WFN15-1457 Miscellaneous Topics 2 Neurological involvement and infective endocarditis in childhood: a review of 4 cases in a University Hospital

<u>M. López</u>^a, I. Schmidt^a, I. Huete^b, M. Hernández^a. ^aPediatría, Facultad de Medicina, Pontificia Universidad Católica de Chile, Santiago, Chile; ^bRadiología, Facultad de Medicina, Pontificia Universidad Católica de Chile, Santiago, Chile

Background: Neurologic manifestations are seen in up 40% of cases of infective endocarditis (IE), includes cerebrovascular entities and other complications. Few publications reports in pediatric population.

Aim: To describe neurological features, neuroimaging and hospital discharge outcomes of pediatric patients with IE.

Patients and methods: We present four pediatric cases of neurological involvement associated with IE seen in a tertiary hospital.

Results: Of 4 children, 2 boys, 2 with previous heart disease (rheumatic fever and septal defect), median age was 8 years (range, 3 - 14). 2 patients have a positive detection of pathogen in blood cultures (Staphylococcus Aureus, Streptococcus Viridans). Common clinical presentation were fever, abdominal pain and neurologic manifestations in 3, 2 and 2 patients, respectively. The median duration from diagnosis of IE to the onset of neurological symptoms was 42 days (range, 1 - 120). Neurological manifestations are impairment of consciousness, seizures and focal signs (3, 2 and 1 patients, respectively). All the patients had cerebrovascular complications, 3 cases due to cardioembolism (2 cases with mycotic aneurysm, additionally, 1 case with a left renal artery aneurism demonstrated by conventional angiography) and one due to vasculitis. 2 patients required surgical management of heart disease due to cardioembolic complications. Outcomes at hospital discharge were mild hemiparesis, normal neurological exam and death in 2, 1 and 1 patients, respectively.

Conclusion: Cerebrovascular complications of IE should be considered in every child with impaired consciousness and fever, with or without a history of heart disease.

doi:10.1016/j.jns.2015.08.699

629

WFN15-0515

Miscellaneous Topics 2

Phenotypic and biochemical features of pyruvate dehydrogenase complex deficiency: a retrospective cohort study at the hospital for sick children

M. Inbar-Feigenberg^a, J.M. Cameron^b, J.T. Clark^a, A. Feigenbaum^a, S. Hewson^a, K. Siriwardena^c, B.H. Robinson^b, <u>S. Mahmutoglu^a</u>. ^aPediatrics, University of Toronto, Toronto, Canada; ^bLaboratory Medicine, University of Toronto, Toronto, Canada; ^cPediatrics, University of Alberta, Toronto, Canada

Background: Pyruvate dehydrogenase complex (PDHC) deficiency is an inherited disorder of energy metabolism caused by one of the three catalytic enzyme deficiency including pyruvate dehydrogenase (E1), dihydrolipoamide transacetylase (E2), dihydrolipoamide dehydrogenase (E3).

Objective: To characterize the phenotypic and biochemical features of patients with PDHC deficiency, we performed a retrospective cohort study.

Methods: We obtained Institutional Review Board approval. We included all patients with a confirmed diagnosis of PDHC deficiency. Electronic patient charts were reviewed for all investigations.

Results: There were 9 patients (5 girls; 4 boys): 5 with E1, one with E2 and 3 with E3 deficiencies. All patients presented with developmental delay and 8 patients had cognitive dysfunction. Additionally, hypotonia (central and/or peripheral) was present in 6, seizures in 5, microcephaly in 2 and movement disorder (ataxia, dystonia, tremor) in 5 patients. Lactic acidemia was present in 8 patients: in 2 patients severe (>10 mmol/l); in 6 patients mild to moderate. Brain neuroimaging showed increased signal intensities in globus pallidus in 3 patients, thin corpus callosum in 3 patients, increased signal intensities in white matter in 4 patients and periventricular cysts in one patient. Brain magnetic resonance spectroscopy showed lactate peak in 3 patients. All patients had deficient PDHC enzyme activity in skin fibroblasts and mutations in the responsible genes.

Conclusions: We present 9 new patients with PDHC deficiency. Targeted next generation sequencing panels and whole exome sequencing will likely identify more patients with non-typical phenotypic and biochemical features. The ketogenic diet treatment will likely improve outcomes.

doi:10.1016/j.jns.2015.08.700

630 WFN15-0811 Miscellaneous Topics 2

Assessment of neuro-muscular function tests in mouse models of obesity and diabetes

<u>S. Martinez Huenchullan</u>^a, J. Power^a, C. Yee^a, M. Morsch^b, S. McLennan^a, S. Twigg^a, C. Tam^c. ^{*a*}Sydney Medical School Charles Perkins Centre, University of Sydney, Sydney, Australia; ^{*b*}Motor Neuron Disease Research Group, Macquarie University, Sydney, Australia; ^{*c*}School of Biological Sciences Charles Perkins Centre, University of Sydney, Sydney, Australia

Non-invasive muscle function tests in mouse models are widely used to investigate effects of neuromuscular disorders on muscle strength, coordination and endurance. In obesity and diabetes muscle function is impaired but the utility of these tests of muscle function in these conditions in mice is not known and was investigated.

Male C57BL/6 mice (5wks/old) were rendered diabetic (DM group;n = 19) using streptozotocin (3x55mg/kg), or fed a high-fat diet (HFD group;45%fat;n = 13). Mice fed laboratory chow acted as control group (CHOW;n = 21). After 10 weeks, hang wire (HW;1 attempt), four limb hanging (FLH;3 attempts) and grip strength (GS;3 attempts) tests were each performed. FLH and HW results were multiplied by body weight, and the mean and best of three attempts was calculated and associated with every attempt.

Testing showed GS data was highly reliable within all groups (Intraclass Correlation Coefficient = 0.7-0.9; p < 0.001).

GS is a reliable test in mouse models of obesity and diabetes. Furthermore, in these metabolic disorders the number of attempts executed per test may affect data precision and interpretation of neuro-muscular performance.

doi:10.1016/j.jns.2015.08.701

631

WFN15-0586 Miscellaneous Topics 2

Epidemiology of malignant neoplasms of the brain in Brazil from 2008 to 2013

M.A. Bannach^a, M.L. Caetano^a, D.G. Costa^a, R.V. Teles Filho^a, A.L. Souza^a, <u>H.H.S. Matozinho^a</u>, J.E.S. Cavalcante^b, L.C. Morais^a, W.N. Naves^b, W.S.S. Souza^b. ^aInternal Medicine Department, Federal University of Goias, Goiânia, Brazil; ^bNeurosurgery Department, Federal University of Goias, Goiânia, Brazil

Introduction: Malignant Neoplasms of the Brain (MNB) are originated from neurons or glial cells and represent 2% of the world's malignant neoplasms. Gliomas are the commonest (80%), but there are also ependymomas, meduloblastomas, among others. They are known for their high aggressiveness and death rate.

Objectives: Analyze and characterize the prevalence of brain malignant neoplasms in Brazil, from 2008 to 2013.

ARTICLE IN PRESS

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

Materials and methods: Analysis of the numbers of MNB hospitalizations and death rate through Unified Health System (UHS), from 2008 to 2013 in Brazil. The Hospital Information System (HIS) – Unified Health System Informatics Department (UHSID) provided data.

Results: MNB corresponded to 1.89% (17th cause) of the hospitalizations by neoplasms in Brazil, which had been rising since 2008, but decreased 8.3% in 2013. São Paulo was the state with the highest absolute values, responsible for 27.15% of national hospitalizations. However, stratified data evidenced Rio Grande do Sul with the highest rates for all years analyzed, with an average rate of 483.83, and a peak in 2012 (512.49), which was higher than national average (315.55), while São Paulo had an average of 351.00. Regarding death rates, although there was a slight decrease in 2013, MNB remained the 7th cause (13.65%).

Conclusion: MNB are the 17th in hospitalizations in Brazil, corresponds to approximately 2% of the malignant neoplasms, befitting the literature, as well as the distribution pattern, presenting more cases in regions with higher HDI. They were also the 7th cause of deaths in the country, evidencing their high lethality.

doi:10.1016/j.jns.2015.08.702

632 WFN15-1140 Miscellaneous Topics 2 Signs and symptoms of brain tumour in childhood

V.C.J. Queiroz^a, <u>H.H.S. Matozinho^a</u>, L.C. Morais^a, J.E.S. Cavalcante^b, F.H.R. Silva^a, M.G.P. Costa^a, F.Y. Pereira I^a, L.C.P. Tavares^a, G.M. Guimarães^a, W.S.S. Souza^b. ^aInternal Medicine Department, Federal University of Goias, Goiânia, Brazil; ^bNeurosurgery Department, Federal University of Goias, Goiânia, Brazil

Background: Brain Tumour (BT) is a recurrent solid tumour in childhood, with variable prognosis that is highly influenced by the time spent between symptoms onset (SO) and diagnosis. Clinical history depends of intracranial pressure, direct compression of gray/white matter, shifting of intracranial contents, secondary cerebral ischemia. **Objective**: Analysis of clinical symptoms of pre-diagnostic symptomatic interval (PSI) of children with BT that would correlate with delay in diagnosis.

Materials and methods: A search was performed in Bireme using "signs and symptoms" AND "brain tumors" AND "childhood". Only English and Portuguese texts published in the last 5 years were considered. From 123 results, only 10 approached the subject of interest.

Results: Wilne et al. (2012) followed 139 children with BT. The symptom and the number of children with it at SO and at diagnosis, respectively, were: headache in 55:81 children, nausea/vomiting in 39:88, motor system abnormalities in 31:93, cranial nerve palsies in 24:75, visual system abnormalities in 23:96, endocrine or growth abnormalities in 10:35, behavioural changes in 4:55. The average of PSI was 3.3 months. Molineus et al. (2013), found that the main clinical symptoms were headache (66.7%), vomiting (57.7%), vision (46.2%) and gait (41.6) disorders and fatigue (41.0%).

Conclusion: Diagnosis of pediatric BT is often delayed due to unspecifics manifestations, which often lead to infections, gastrointestinal disturbances hypothesis. Therefore, BT should not be excluded before deeper investigations if parents report a clinical history of combination of headache with other neurological presentations.

doi:10.1016/j.jns.2015.08.703

633 WFN15-1303 Miscellaneous Topics 2 Case report of a newborn with Arnold-Chiari malformation type li associated with hydrocephalus and myelomeningocele

<u>H.H.S. Matozinho^a</u>, J.M. Rosa^a, J.A. Ferreira^a, S. Borges^a, V.F.P. Costa^a, M.S. Garrote^b, W.S.S. Souza^c, L.C. Morais^a, F.H.R. Silva^a, J.E.S. Cavalcante^c. ^aInternal Medicine Department, Clinics Hospital Federal University of Goias, Goiânia, Brazil; ^bPediatrics Department, Clinics Hospital Federal University of Goias, Goiânia, Brazil; ^cNeurosurgery Department, Clinics Hospital Federal University of Goias, Goiânia, Brazil

Background: Arnold-Chiari malformation type II is characterized by displacement of cerebellar tonsils, parts of the cerebella fourth ventricle, pons and medulla oblongata through the foramen magnum into the spinal canal. This is usually associated with hydrocephalus and myelomeningocele.

Objective: Report a case of a newborn diagnosed with Chiari II malformation, hydrocephalus and myelomeningocele.

Patient and methods: Female neonate was born full-term by cesarean surgery. Mother denies: illicit drugs abuse, complications during pregnancy, folic acid intake throughout pregnancy. Ultrasound examination, before giving birth, at admission evidenced ventriculomegaly and spina bifida. Physical examination afther birth detected Apgar score of 7 and 9, cephalic perimeter of 46 cm and presence of myelomeningocele. **Results**: Ventriculoperitoneal shunt was performed soon after birth and, in the 6° day of life the neonate underwent a correction surgery to close the toraco-lumbar myelomeningocele with a skin flap. On 11° day of life, cranium CT showed severe hydrocephalus, cortical thinning, herniation of the amygdala, cerebellar vermis, fourth ventricle and medulla oblongata through the foramen magnum. The neonate infant evolved with seizures from 13° to 17° days of life.

Conclusion: The ultasonographic prenatal screening is emphasized as the primary method of assessment of this early fetal malformation. MRI and CT are the modalities of choice for detecting and characterising the full constellation of findings associated with Chiari II malformations and combined with the clinical presentation allow a clear diagnose of this pathology.

doi:10.1016/j.jns.2015.08.704

634 WFN15-1131 Miscellaneous Topics 2 Morphofunctional changes in the brain during adolescence and their impact on social interactions

Y.C. Machado, <u>B.G. Mazzoni</u>, B.C. Gontijo. Medicine, Universidade Federal de Minas Gerais, Belo Horizonte, Brazil

Background: Relationships are a considerably important matter during adolescence. Histological and imaging studies have shown that, during this period, a large amount of structural changes which take place in the brain interfere with the teenager's social interactions and may be involved in the physiopathology of several psychiatric disorders, such as social anxiety, eating disorders, substance abuse and schizophrenia.

Objective: To present neurobiological aspects of social cognition that may contribute to the development of mental health and educational approaches.

Material and methods: An extended review was made using scientific articles found in Pubmed and Scielo.

Results: Social cognition depends on cortical neural networks related to mental functions, such as emotional processing, self-consciousness, empathy, social hierarchy perception, group belonging, social behavior monitoring and individual data filing. Biological and environmental

factors interfere with adolescents's social behavior. Hormonal synthesis, synaptogenesis, synaptic remodeling and myelination are the underlying causes of changes in specific brain areas (prefrontal, temporoparietal, insula, cingulate gyrus and amygdala), which are related to executive functions and social cognition improvements.

Conclusion: Biological studies concerning brain changes during adolescence allow us to better understand these individuals' behavior, helping society look for more effective ways to deal with teenagers and create methods which contribute to cognition and sociability development.

doi:10.1016/j.jns.2015.08.705

635 WFN15-0947 Miscellaneous Topics 2 Epidemiological study of stroke in a University Hospital School 2008-2012

<u>M.J. Medeiros</u>^a, J.P.U. Ferreira^b, R.B.C. Leão^c, L.F.M. Pereira^a, G.A.G. Silva^d, E.F. Pereira Filho^a, M.P. Ribeiro^e, P.F. Sablewski^f, F.A.G. Teixeira^f, J.D. Almeida^e. ^aFaculty of Medicine - Camilo Castelo Branco University, Santa Casa de Misericórdia de São José dos Campos Hospital, São José dos Campos, Brazil; ^bIntensive Care, São Francisco Hospital, Ribeirão Preto, Brazil; ^cE.R., Santa Casa de Misericórdia de São José dos Campos Hospital, São José dos Campos Hospital, São José dos Campos, Brazil; ^cE.R., Santa Casa de Misericórdia de São José dos Campos Hospital, São José dos Campos, Brazil; ^dFaculty of Medicine, Universidade da Região de Joinville, Joinville, Brazil; ^eFaculty of Medicine, Camilo Castelo Branco University, Fernandópolis, Brazil; ^fFaculty of Medicine, Santa Casa de Misericórdia de Fernandópolis University Hospital School, Fernandópolis, Brazil

Background: Worldwide, stroke is the second leading cause of death. In recent decades, Brazil has been changing its morbidity and mortality profile, one of the most important chronic disease is stroke, which is a major cause of hospitalization and mortality.

Objectives: To conduct an epidemiological study of stroke onset in patients enrolled in a University Hospital School

Patients and methods: Cohort retrospective study of cases of stroke. Statistical analysis was performed with use of chi-square test (χ 2), being accepted $\alpha = 0.01$ for interpreting the results.

Results: Of 490 patients studied (53.2%) were men and 431 (46.8%) women, the median age was 72 years (61-85), over 75% were retirees, 523 (56.8%) had a history of hypertension and 438 (47.55%) had more than one risk fator; 326 patients (35.6%) remained at the Hospital for up to five days, while 271 (29.4%) remained for 10 days. The mortality rate was 35.05 per 100,000 inhabitants, while the coefficient of mortality of the disease was 8%. Types of stroke, 783 (85.1%) were ischemic, while 138 (14.9%) bleeding.

Discussion: Our study shows stroke prevalent in older people which worsens the morbimortality table and dramatically decreases the quality of life of these people. More prevalent in men, white race and the hospital length of stay is high; the death rate from specific cause is also high, its lethality coefficient despite being below the world which is 24%, is worrying. Population, multicenter, long-term can show the epidemiological trend of cerebrovascular accidents in our midst.

doi:10.1016/j.jns.2015.08.706

636

WFN15-0367 Miscellaneous Topics 2 Mitochondrial neurogastrointestinal encephalomyopathy syndrome with an unusual pattern of inheritance

N. Rafiei^a, <u>S. Mishra^b</u>, H. Mohammadkhanli^c, B. Trikamji^c. ^aNeurology, UCLA David Geffen School of Medicine, Los Angeles, USA; ^bNeurology,

USC Keck School of Medicine, Los Angeles, USA; $^{\rm c}{\rm Neurology},$ Olive View UCLA Medical Center, Los Angeles, USA

Objective: we describe an unusual pattern of inheritance in a family with MNGIE syndrome. Background: Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE) is a rare autosomal recessive disorder defined by gastrointestinal dysmotility, external ophthalmoplegia, mitochondrial myopathy, peripheral neuropathy and leukoencephalopathy. It is caused by mutation in the gene of thymidine phosphorylase (TYMP).

Design/method: We report a Hispanic family with genetically proven MNGIE syndrome with affected members in two generations including three brothers of four siblings and their mother, maternal uncle and maternal aunt.

Results: Six family members in two generations were affected. Our patient and two of his siblings and their mother were among the affected members. Although the inheritance pattern of MNGIE is autosomal recessive, we observed an unusual pattern of maternal inheritance in this family which delayed the diagnosis prior to referral of the family to our center. We postulated that severe TYMP dysfunction resulted in de novo mitochondrial mutations in germinal cells of the mother. The eldest offspring was spared due to younger age of mother at the time of pregnancy.

Conclusions: We reported the first family with MNGIE syndrome and maternal pattern of inheritance. In clinically appropriate setting, diagnosis of MNGIE should not be dismissed due to a maternal pattern of inheritance as current available treatments may be beneficial to these patients.

doi:10.1016/j.jns.2015.08.707

637

WFN15-1343 Miscellaneous Topics 2 Organic acidemia: regarding two clinic cases in children

J. Montiel Blanco, <u>P. Muñoz Huerta</u>, J. Flores Bravo. Neuropediatria, Instituto Nacional de Salud del Niño, Lima, Peru

Summary: Report the clinic, neuroimaging and the laboratory in two children diagnosed with organic acidemias. Case. male infant of 29 days old began with symptoms of acute encephalopathy after initiation of breastfeeding, and develops coma with metabolic acidosis, hypoglycemia and elevated serum ammonia (503 umol / 1) associated with cerebral edema, He was diagnosed as propionic acidemia, and started to be fed with nutritional support (special formula) and breast milk, developing dermatitis enteropathica by isoleucine's déficit. He started a new nutritional support (special formula, breastfeeding and isoleucine and valine's supplements, He was discharged to home at 6 weeks old and 5 days after died of metabolic decompensation by an UTI. Second case: Male infant 10 months old with a history of seven previous hospitalizations by vomiting, poor sucking and dehydration, sometimes followed by respiratory illnesses; Ans was admitted as an emergency with metabolic acidosis, hypoglycemia, and seizures, we founded serum ammonium in normal range. A tándem mass spectrometry diagnosed Academy 3-hydroxy-3-methyl glutaric, and the neuroimaging and clinical outcome is described.

Conclusion: The organic acidemias are metabolic diseases by accumulation of organic acids in body fluids, whose suspicion and early diagnosis allows a better prognosis and less neurological sequelae.

doi:10.1016/j.jns.2015.08.708

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

638 WFN15-1273 Miscellaneous Topics 2 On neuro-vascular factors of idiopathic hypertension including cerebral apoplexy

<u>E. Neu^a</u>, M. Traub^a, M. Michailov^a, H. Schumitz^a, H. Dahlheim^b, G. Werner^c, J. Foltinova^d, P. Birkenbihl^a, H. Danler-Oppl^a, T.N. Senn^a, U. Welscher^a, N. Moro^a. ^aDept. Pharmaco-Physiology, Inst. Umweltmedizin c/o ICSD/IAS e.V. POB 340316 80100 Muenchen Germany (Int. Council Sci. Develop./Int. Acad. Sci. Berlin-Bratislava-Innsbruck-Muenchen-NewDelhi-Paris-Sofia-Vienna), Munich, Germany; ^bInst. Physiology, Med. Fac. Univ. München, Munich, Germany; ^cClinic Internal Med., Univ. Göttingen, Göttingen, Germany; ^dMed. Fac., Univ. Bratislava, Bratislava, Slovakia

Introduction: Since discovery of blood circulation (HARVEY 1628), homoeostasis of blood-pressure (CANNON) & blood volume (GAUER) is originated enormous information, but till today *genuine hypertension* pathogenesis incl. *apoplexy* (HIPPOCRATES) *cerebri* is not clarified. It will be given consideration of some neuro-vascular factors for discussion. **Methods**: Vascular motor activity (helical prep.), rat-blood-pressure (BP).

Results (recent/earlier): Reports about spontaneous-phasic (SPC: 0.5-2/min) and periodic slow-tonic contractions (STC: 0.1-0.2/min) induced by hormones (angiotensin-II/5-HT/PGs/vasopressin) & K⁺ in rat-aorta (Michailov et-al. 1978), human renal and uterine arteries (Michailov-Dahlheim-Werner et-al., Werner et-al., Ueda et-al. Neu et-al. 1981-85) open new approaches to vascular normal&pathophysiology. STC-relation to low-frequency bloodpressure fluctuations (Mayer-waves) & pericytes (Persson, Pohl et al. 2003/2006) is not clarified. Vasopressin (VP:10-20mIU) induce STC in human renal artery (Werner 1985) and SPC in cerebral basilar-artery (rat: Rusch & Hermsmeyer 1985). BP-observations about transformation of acetylcholine-depressor response into biphasic depressor-pressor (dR/pR) by nicotine & mercptoethylguanidine (MEG: inhibitor of NO-synthase), also of dR to electrical-central-vagalstimulation (CVS:55Hz,2ms,5s,5V), potentiation of pR by VP (5-100mIU), non- (AHR-600/McN-A-343) & nicotine-like (DMPP: 0.1µg-100mg/kg) ganglion-stimulating agents, also inverting serotoninergic dR into pR by MEG suggested new hypothesis about idiopathic hypertension. Conclusion: Drugs & endogen substances could sensitize BP regulatory central, spinal peripheral structures (ref.) causing idiopathic hypertension (independent of renal), leading to angio-cardiac/cerebral spasms (infarct & apoplexy) by biphasic dR/pR. Support of further experimental & clinical neurological investigations could open new approaches to neurovascular pharmacotherapy in context of UNO-Agenda21 for better health, education, etc. on global level.

Ref., Methods & Dedication see Neu et al. Part I&II, WCN-2015.

doi:10.1016/j.jns.2015.08.709

639

WFN15-1560 Miscellaneous Topics 2 Neurocognitive assessment using coghealth battery in children with adhd before and after treatment

K. Kawabe^a, F. Horiuchi^b, <u>Y. Oka^c</u>, S. Kondo^a, M. Matsumoto^a, K. Seo^a, S. Ueno^a. ^aDepartment of Neuropsychiatry, Ehime University Graduate School of Medicine, Ehime, Japan; ^bCenter for Child Health Behavior and Development, Ehime University Hospital, Ehime, Japan; ^cCenter for Sleep Medicine, Ehime University Hospital, Ehime, Japan

Background: Attention-deficit/hyperactivity disorder (ADHD) is characterized by inattention, hyperactivity/impulsivity and is often

treated pharmacologically with methylphenidate. Subjective and objective assessment is necessary to determine the efficacy of treatment in children with ADHD.

Objective: The aim of the present study was to examine neurocognitive function, such as working memory and attentional performance, in children with ADHD before and after pharmacological intervention. The study also compared the results of the CogHealth battery in ADHD and age-matched healthy children, to assess its suitability as a brief and reproducible cognitive tool for children with ADHD in the clinical setting.

Methods: The CogHealth battery, an objective assessment tool, was used to assess the effects of treatment with methylphenidate on neurocognitive function in 11 male children with ADHD (mean age: 9.4 ± 2.2) and to compare such function with 33 age-matched male children.

Results: Significant abnormalities were noted in the accuracy and reaction time for correct answers in the Detection task in children with ADHD compared with the control. The scores improved significantly following treatment with methylphenidate. The accuracies of One Back Task and One Card Learning task improved significantly after treatment in children with ADHD.

Conclusion: The CogHealth battery is suitable for assessment of changes in neurocognitive function after treatment of children with ADHD.

doi:10.1016/j.jns.2015.08.710

640 WFN15-0116 Miscellaneous Topics 2 Cerebrotendineous xanthomatosis – a case report of two sisters

N. Oliveira, M. Lima, M. Prado, M. Acchar. NEUROLOGY, Federal University of Rio de Janeiro (UFRJ), Rio de Janeiro, Brazil

Introduction: Cerebrotendineous xanthomatosis (CTX) is a rare autosomal recessive disease characterized by deficiency of 27-hydroxylase, leading to lipid accumulation in various tissues.

Objective: Describe the clinical, radiological and laboratory findings of CTX in two sisters.

Case Reports: Patient 1: 34 year-old woman with a previous diagnosis of schizophrenia presented with gait impairment due to spasticity and cramps. Medical history revealed perinatal anoxia as well as congenital cataracts and chronic diarrhea since childhood. One sister (patient 2) has a similar clinical picture. Clinical examination disclosed xanthomas in the ankles, an ataxic gait with positive Romberg sign, spasticity and brisk reflexes in the four limbs and palatal tremor. Patient 2: 35 year-old woman, with history of seizures and gait impairment. Neurological examination showed universal hypotonia and hyperreflexia, bilateral intention tremor. The electrophisiologic finding of patient 1 shows a sensory-motor polyneuropathy with with axonal features restricted to the lower limbs. Brain Magnetic resonance imaging (MRI): hyperintense signal in T2 and Flair in the white matter, symmetric leukomalacia and hyperintense signal in the pons, cerebelar dentate nuclei and peduncles. Cerebellum atrophy. Hyperintense signal in the medula oblongata olive's. Serum Cholestanol was 20,58mcg/ml.

Discussion: CTX is a autosomal recessive disease with Pyramidal signs, peripheral neuropathy, ataxia, seizures and cognitive decline. The diagnosis can be confirmed by the dosage of colestanol. MRI usually demonstrates hyperintense lesions in T2 and Flair located in periventricular area, basal ganglia and dentate nuclei of the cerebellum with cerebellar atrophy. Chenodeoxycholic acid replacement therapy is used. Early recognition can prevent neurological injury.

doi:10.1016/j.jns.2015.08.711

14

641 WFN15-0385 Miscellaneous Topics 2 Functional outcome after first ever ischemic stroke: one-year cohort study

<u>T. Pekmezovic</u>^a, S. Medic^b, D. Kisic-Tepavcevic^a, A. Pavlovic^c, LJ. Beslac-Bumbasirevic^c. ^aInstitute of Epidemiology, University of Belgrade Faculty of Medicine, Belgrade, Serbia; ^bDepartment of Neurology, KBC Dr Dragisa Misovic, Belgrade, Serbia; ^cClinic of Neurology, University of Belgrade Faculty of Medicine, Belgrade, Serbia

Background: The purposes of this study were to determine the 1-year functional outcome after first-ever ischemic stroke and to identify the baseline predictors of post-stroke dependency.

Patients and methods: We prospectively and consecutively collected data on 300 patients with first-ever acute ischemic stroke admitted to two main neurological institutions for cerebrovascular diseases in Belgrade, Serbia. The predictive value of different variables in 1-year functional dependence was assessed by univariate Cox proportional hazard regression model.

Results: Data on functional outcome 1 year after stroke were available for 184 patients (109 died, 7 patients were lost to follow up). Among survivors 100 (54.3%) patients were functionally dependent at the end of the first year after stroke. The univariate predictive model revealed that National Institutes of Health Stroke Scale (NIHSS) score ≥ 11 (p = 0.019), modified Rankin scale (mRS) ≥ 3 (p = 0.001), Barthel index (BI) ≤ 40 (p = 0.001), diabetes mellitus (p = 0.034), glucose ≥ 7.3 mmol/L on admission (p = 0.035) and nursing-home residence (p = 0.019) were significant unfavorable predictors of dependence, while early physical therapy (p = 0.035) was significant favorable prognostic factor for 1-year functional outcome in our first-ever ischemic stroke patients. In the final multivariate analysis only diabetes mellitus (p = 0.020) and BI ≤ 40 (p = 0.002) remained significant independent predictors of 1-year functional dependency in our cohort.

Conclusions: The findings support the need for optimal control of hyperglycemia, especially in patients with diabetes, and emphasize the importance of early implementation of physical therapy, in order to improve initial dependency and long-term functional outcome after an ischemic stroke.

doi:10.1016/j.jns.2015.08.712

642

WFN15-1119 Miscellaneous Topics 2

The prevalence of familial forms of idiopathic inflammatory demyelinating disease in a referral center of Rio De Janeiro – Brazil

<u>F.F.C.C. Pereira</u>^a, R.M.P. Alvarenga^a, M. Bernardes^a, M.P. Alvarenga^a, C.C.F. Vasconcelos^a, C. Miranda^b, E. Batista^b, C. Paiva^c. ^aNeurology, Universidade do Rio de Janeiro, Rio de Janeiro, Brazil; ^bNeurology, Hospital da Lagoa, Rio de Janeiro, Brazil; ^cGenetic, Universidade do Rio de Janeiro, Brazil

Background: The influence of genetics in Multiple Sclerosis (MS) was suggested by Eichorst in 1896 when familial aggregation were identified. Differences in genetic susceptibility between MS and Neuromyelitis optica (NMO) have been described, introducing further evidence that they are different demyelinating conditions. **Objective**: To describe a study of familial forms of demyelinating diseases from a MS referral center in Río de Janeiro State, Brazil. **Methods**: a descriptive, cross-sectional study was done to identified familial idiopathic inflammatory demyelinating disease (IIDD) cases in a public hospital, where 75% of patients with IIDD who live in Rio de

Janeiro state (Southeast region of Brazil) are referred. The diagnoses of all consecutive patients followed in 2011 were reviewed to apply new diagnostic criteria (Wingerchuk et al 2008). The cases that had at least one other relative with IIDD were selected for the study.

Results: Familial forms were found only in the MS and NOM syndrome (NMOSD)categories. 23 MS families (MSf) were identified, 60,86% with first degree kinship. It has a Caucasian preponderance, 36 (90%) of whom were white. The frequency of early onset was 15% of patients and 20% of the MSf cases have progressive primary course. Two NMO patients and one OS-MS patient had relatives with monophasic demyelinating diseases.

Conclusion: The frequency of familial cases of IIDD was 6.12% among MS patients and 2.8% in NMO SD.

doi:10.1016/j.jns.2015.08.713

643 WFN15-113

WFN15-1133 Miscellaneous Topics 2 The prevalence of Neuromyelitis optica in a Brazilian City

F.F.C.C. Pereira, A.B.C. Pereira, R.M.P. Alvarenga, C.C.F. Vasconcelos. *Neurology, Universidade do Rio de Janeiro, Rio de Janeiro, Brazil*

Background: Neuromyelitis optica (NMO) is a demyelinating disease of central nervous system characterized by involvement of the optic nerves and spinal cord. Studies suggest that the incidence of NMO is very low and represents less than 1.5% of individuals with demyelinating disorders. The prevalence of NMO (per 100.000) rage to 0.52 (0.39–0.67) in Cuba to 4.4 (3.1–5.7) in Denmark.

Objective: To determine the crude prevalence of NOM in a city in the southeastern Brazilian region located at 22°31'23 " south latitude and 44°06'15 " west longitude and altitude of 390 meters.

Patients and methods: Patients were identified using three sources of information: 1) inform medical care from private clinics, public and hospitals; 2): MRI centers and 3) drug-dispensing center. Case records of these patients were reviewed and classified according to the most recent diagnostic criteria for NMO. Data were analyzed using SPSS to calculate the crude prevalence.

Results: One Caucasian woman patient was identified in a population of 257.803 residents - as NOM anti-AQP4 positive. The crude prevalence was 0,39/100.000 residents.

Conclusion: More studies were necessary to evaluate the prevalence and incidence of this disease.

doi:10.1016/j.jns.2015.08.714

644

WFN15-0128

Miscellaneous Topics 2

A national profile of neurodevelopmental disabilities in Canadian children: data from the National Longitudinal Study of Children and Youth

<u>A. Prasad</u>^a, J. Burneo^b, B. Corbett^c. ^aPediatrics and Clinical Neurosciences, Children's Hospital London Health Sciences Centre, London, Canada; ^bEpilepsy Program Clinical Neurosciences, University Hospital London Health Sciences Centre, London, Canada; ^cResearch Data Centre, Richard Ivey School of Business University of Western Ontario, London, Canada

Background: Chronic neurological conditions (epilepsy, cerebral palsy, intellectual disability, etc) contribute to the burden of neurological disorders in childhood. Population based data carry

ARTICLE IN PRESS

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

implications for childhood outcomes, health care costs, establishing priorities, and development of targeted health care interventions.

Objective: To establish prevalence rates and population based estimates of neurodevelopment disabilities based on survey data from Canada's National Longitudinal Study of Children and Youth (NLSCY) with ethics approval.

Methods: The NLSCY captured, socio-demographic information, age, sex, education, ethnicity, household income, health related conditions from birth to 15 years. The presence of neurodevelopmental disabilities was self-reported by the 'person most knowledgeable' (PMK) about the child. The list of responses included; Epilepsy, Cerebral palsy, Intellectual disability, Learning disability, Emotional and Nervous difficulties, ADHD treated with Ritalin. Prevalence rates were calculated from 3 cycles of the NLSCY using 1000 bootstrap weights to account for survey design factors.

Results: Cross sectional prevalence rates/1000 children(birth- 15 years) were determined as; Epilepsy (2.3-5.3), cerebral palsy (1.6-2.8), learning disability (37-57), intellectual disability (2.8-4.8) and emotional nervous difficulties (17-21.7). Population based estimates were calculated from Census data. There was a clear male preponderance observed across all the four neurological conditions surveyed (p<0.000).

Conclusion: These results provide prevalence rates and population estimates of neurodevelopmental disabilities in Canadian children. There is a clear gender effect seen with male children significantly more affected than females across the spectrum of neurodevelopmental disabilities. We discuss methodological aspects related to the ascertainment of epilepsy in the NLSCY, and to the validity and implications of our findings.

doi:10.1016/j.jns.2015.08.715

645

WFN15-1385

Miscellaneous Topics 2 Historical trends in epilepsy mortality in Uruguay. Preliminary results (1893-1996)

L. Rodríguez, P. Braga. Epilepsy Section, Institute of Neurology Facultad de Medicina Universidad de la República, Montevideo, Uruguay

Background: Scarce data on epilepsy mortality in LatinAmerica is available.

Objective: To explore the epidemiology of mortality attributed to epilepsy in Uruguay, and its temporal trends.

Methods: Available governmental mortality registries according to disease, sex and age ranges were reviewed. Population data was obtained from National Census or inter-Census estimates, according to the National Institute of Statistics. Epilepsy mortality rates (EMR) and proportion of mortality attributed to epilepsy (PMAE) were calculated for the total population and predefined age ranges.

Results: EMR varied from 0.79-3.08/100.000 inhabitants/year with a mean of 1.68 ± 0.44 , with random fluctuations in time (Fig. 1). EMR in females (1.30 ± 0.31) was significantly lower than in men $(1.76\pm0.59; p=0.049)$ (Fig. 2). Deaths attributed to epilepsy represented $16.25\pm4.47/10.000$ deaths. Up to 2-3 fold increase in PMAE was observed after 1950, mainly in infants and young adults.

Discussion and conclusions: Although limitations of epidemiological studies based on registries apply, these records allow for a unique historical view. Epilepsy MR was stable along one century. PMAE increase in the last decades may be explained by decrease in mortality due to other diseases, as infections.







WFN15-1486 Miscellaneous Topics 2 Social cognition in frontotemporal dementia and Alzheimer disease

<u>A. Ruiz-Tagle</u>^a, G. Musa^b, P. Lillo^c, A. Slachevsky^d. ^aUniversidad de Chile, Laboratorio de Neurociencias Cognitivas del Centro de Investigación Avanzada en Educación, Santiago, Chile; ^bUniversidad de Chile, Departamento de Ciencias Neurológicas Facultad de Medicina, Santiago, Chile; ^cUniversidad de Chile, Departamento de Neurología Sur Facultad de Medicina, Santiago, Chile; ^dHospital del Salvador, Unidad de Neurología Cognitiva y Demencias Servicio de Neurología, Santiago, Chile

The Mini SEA (MS) is the short version of the Social Cognition and Emotional Assessment battery, it evaluates emotion processing and theory of mind through a shortened version of the faux pas task and a facial emotion recognition test. The MS has shown neural correlate in VMPFC and has proven to be sensible and specific in distinguishing early Behavioural Variant Frontotemporal Dementia (bvFTD) from Alzheimer Disease (AD). We explored social cognition performance in bvFTD and AD patients at mild to moderate stage with the hypothesis to find a greater impairment in bvFTD compared to AD as seen in previous studies. 10 bvFTD and 9AD patients were recruited, we obtained patient and Institutional Review Board approval. Both groups were not significantly different with regard to age nor educational level. Results showed that AD patients scored lower than bvFTD patients in general cognition tests (ACE-III bvFTD mean 74,5 (SD:13) and AD 69,1 (12) p: 0,307; MMSE bvFTD 26,2 (3,6) and AD 23,3 (2,7) p: 0,022) but performed better than bvFTD at the MS; bvFTD 19 (4,9) and AD 20,4 (2,9) p: 0,870. bvFTD showing twice the variability in their performance at social cognition tasks. The preliminary data show only a mild difference at the MS performance comparing AD to FTD. Our results

suggest that MS as a tool for distinguishing AD from FTD may be more specific at earlier stages of AD.Funding: Fondecyt 1140423, 1130920 & Basal Funds For Centers of Excellence, Project FB0003- Associative Research Program – CONICYT-CHILE.

doi:10.1016/j.jns.2015.08.717

647

WFN15-0693 Miscellaneous Topics 2 Unusual progression of Miller Fisher syndrome with bilateral facial palsy

<u>M. Sakel^a</u>, H. Khan^a, M. Mazen^b, J. Sivagnanasundaram^b, K. Saunders^c, P. Pullicino^d. ^aNeuro-rehabilitation, East Kent University Hospitals NHS Foundation Trust, Kent, United Kingdom; ^bNeurology, East Kent University Hospitals NHS Foundation Trust, Kent, United Kingdom; ^cNeuro-rehabilitation, Gloucester NHS Hospital Trust, Cirencester, United Kingdom; ^dNeurology, East Kent University NHS Hospital, Kent, United Kingdom

Background: Miller Fisher syndrome (MFS) is part of the spectrum of Guillian Barre disorders comprising of multiple subtypes and course of disease.

Objective: We describe a case report of a patient with MFS who developed bilateral facial palsy after initial treatment with immunotherapy.

Patients and methods / **material and methods:** A 67 year old gentleman present complaining of feeling unsteady on his feet, tightness in his throat, double vision and slurred speech. He had bilateral ptosis, sluggish pupillary reflexes, diminished reflexes and down going planters, intention tremor and dysdiadochokinesia. The CSF protein was border-line raised with oligoclonal banding suggestive of systemic inflammation. CSF cell counts were normal. Anti-acetylcholine receptor antibodies were negative. MRI imaging was grossly normal. The patient was treated with a 5-day course intravenous immunoglobulin with a probable diagnosis of MFS and transferred to neuro-rehabilitation Unit.

Results: Patient reported improvement in diplopia and in- coordination. However on day 10 of admission, neuro-rehabilitation professionals noted right facial nerve palsy, which progressed to bilateral facial nerve palsies by day 15. The patient was transferred back to Neurology ward to consider Plasma exchange. A further course of intravenous immunoglobulins resolved the neuro-deficits. A review of the literature noted that a delayed presentation of bilateral facial nerve palsies can occur in up to 3% of Miller Fisher syndrome cases.

Conclusion: Delayed presentation of bilateral facial nerve palsy can occur in MFS. Although previous cases have reported improvement in symptoms without repeat immunotherapy, our case required repeat immune-therapy without expensive Neuro-rehabilitation programme.

doi:10.1016/j.jns.2015.08.718

648 WFN15-0451

Miscellaneous Topics 2

Intrathecal 2-Hydroxypropyl-Beta-Cyclodextrin (HPBCD) therapy in adult-onset Niemann-Pick Disease Type C (NPC)

<u>Y. Sakiyama</u>^a, S. Shibata^a, H. Sanayama^a, S. Ono^a, M. Maekawa^b, M. Matsuo^c, T. Irie^d, Y. Eto^e. ^aNeurology, Jichi Medical University Saitama Medical Center, Saitama-city, Japan; ^bPharmaceutical Sciences, Tohoku University Hospital, Sendai, Japan; ^cPediatrics, Saga University Faculty of Medicine, Saga, Japan; ^dCenter of Clinical and Pharmaceutical Sciences,

Kumamoto University Faculty of Pharmacy, Kumamoto, Japan; ^eAdvanced Clinical Research Center, Institute for Neurological Disorders, Fukushima and Kanagawa, Japan

Background: NPC is a rare neurovisceral lysosomal storage disease. In many countries, Miglustat has been approved for the stabilization of neurological symptoms associated with NPC. Intrathecal HPBCD therapy is compassionately used for some child and adolescent cases in some countries.

Objective: We used HPBCD for siblings with adult-onset NPC.

Patients and methods: We obtained approval from patients' mother and the Institutional Review Board (IRB).

Patients are genetically proven (NPC1, G992R/ IVS6-3 C>G) NPC siblings (37-year-old male (Patient 1) and 28-year-old male (Patient 2)) who developed neurological symptoms despite the administration of Miglustat for two years.

HPBCD was administered by lumbar intrathecal injection once every four weeks, starting with a dose of 100 mg per individual that was increased gradually to 300 mg.

Results: No adverse effects were observed over the course of treatment. Patient 1 became neurologically stable, and the NPC-ADL score was improved in Patient 2.

Conclusion: Intrathecal injection was also safe and effective in adultonset NPC patients.

doi:10.1016/j.jns.2015.08.719

649 WFN15-1129 Miscellaneous Topics 2 Altered brain energetics causes mitochondrial fission arrest in Alzheimer's disease

<u>J. Salisbury</u>^a, L. Zhang^b, S. Trushin^b, T. Christensen^a, E. Trushina^b. ^aBiochemistry and Molecular Biology, Mayo Clinic, Rochester, USA; ^bNeurology, Mayo Clinic, Rochester, USA

Mitochondrial dysfunction and altered cellular energetics are implicated in the etiology of Alzheimer's Disease (AD). Excessive mitochondrial division has been observed in cellular and animal models of familial AD (FAD), and in AD patients. Thus, understanding regional cellular responses to changes associated with disease progression, particularly regarding the relationship between mitochondrial energetics and the balance of mitochondrial fission and fusion, has the potential to elucidate basic mechanisms of disease while also suggesting targets of therapeutic opportunity.

Using three-dimensional reconstruction electron microscopy (3D EM) of brain tissue from AD patients and animal models of familial FAD, we elucidated changes in mitochondrial morphology in the context of three-dimensional architecture of the brain and mitochondria per se. We demonstrated that AD neuronal mitochondria display a highly exaggerated fission arrest phenotype that resembles 'mitochondria-on-a-string" (MOAS) (Fig. 1b,c). MOAS formation was not associated with reduced levels of fission/fusion proteins or altered ability of Drp1 to translocate to mitochondria. The MOAS phenotype was mimicked in cultured neurons treated with the inhibitors of Drp1 GTPase activity and in mice under hypoxic conditions. Since fission is thought to provide a mechanism for quality control through the disposal of damaged mitochondria, we suggest that mitochondrial fission arrest may contribute residual mitochondrial functions extending a protective energetic margin that plays a role in neuronal cell survival.

doi:10.1016/j.jns.2015.08.720

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

650 WFN15-1076 Miscellaneous Topics 2 Autoimmune encephalitis as differential diagnosis of temporal encephalitis

<u>G. Saraiva^a</u>, G. Santana de Lima^a, R. Mendes Silva^a, A. Bueno Carvalho^a, Y. de Castro Machado^b, N. Matos Pereira^a, O. Ferreira Cunha^a, D. Magno^a, M. Alexandre Diniz Carneiro^a, D. Sisterolli Diniz^a. ^aNeurology, University Federal of Goias, Goiania, Brazil; ^bNeurology, University Federal of Minas Gerais, Belo Horizonte, Brazil

Background: The encephalitis matches to agude inflammatory diseases of the central nervous system associated to high morbidity and mortality, where the herpetic encephalitis is the main cause of temporal lobe impairment. Autoimmune encephalitis (AIE) enter the differential diagnosis with or without associated neoplasia. The most related antibodies with AIE are anti -NMDA and anti-glutamate.

Objectives: Report a case of autoimmune-mediated temporal encephalitis associated to anti-GAD antibodies.

Patients and methods: LFCN, 54, female, began in August/2014 with apathy, discouragement and evolved in one month to amnesia for recent events, confusion and somnolence. The Magnetic Ressonance (09/18/2014) showed hyperintense area in bilateral frontotemporal region. The electroencephalography (31/10/2014) showed nonspecific discreet intermittent disturbancein frontotemporal regions suggestive of cortico-subcortical dysfunction. Cerebrospinal fluid (CSF) with increased nonspecific proteins. All of the research for infectious viral diseases were negative, including serology and PCR search for cytomegalovirus, herpes virus and dengue. The patient was then treated with immunoglobulin IV and plasmapheresis with improvement of symptoms and PET-scan. Anticancer antibodies were asked in CSF and plasma, with positivation for anti-GAD in plasma.

Results: We reported a case of bilateral frontotemporal encephalitis and positive Anti-GAD antibodies as prime suspect of autoimmune encephalitis responsive to immunotherapy.

Conclusion: The anti-GAD usually is associated to diabetes mellitus and Stiffman syndrome. In this case, there is a temporal relation to the patient's condition and, therefore, we emphasize the importance to raise the possibility of anti-GAD relation with AIE.

doi:10.1016/j.jns.2015.08.721

651

WFN15-1034 Miscellaneous Topics 2 Orthostatic hypotension and delirium; an unusual presentation of the Anti-Hu antibody related paraneoplastic syndrome

<u>W.J. Scotton</u>^a, S. Chaal^a, M.S. Zandi^b, C. Allen^b. ^aNeurology, Queen Elizabeth Hospital Birmingham, Birmingham, United Kingdom; ^bNeurology, Addenbrookes Hospital, Cambridge, United Kingdom

We present an unusual case of the Anti-Hu antibody related paraneoplastic syndrome in a 74yr lady, presenting with orthostatic hypotension and delirium.

This lady had a two month history of recurrent falls and episodic confusion, initially presented to her local ED where she was diagnosed with orthostatic hypotension and confusion secondary to a UTI. She later presented to the neurology service at Addenbrookes Hospital with worsening falls associated with loss of consciousness.

On neurological examination the only abnormal findings were a mild impairment of verbal fluency, though she had a profound symptomatic orthostatic hypotension with a drop from 168/82 to 78/47 on sitting out in her chair. Her only abnormal investigations were

oligoclonal banding in her CSF and serum, along with a raised anti-Hu antibody. Although a PET scan demonstrated increased uptake in her left lower lobe airway, repeated bronchoscopic biopsies failed to show any malignant cells. Her pandysautonomia was confirmed with autonomic function testing.

She was managed with a 5 day course of iv Methylprednisolone, as well as compression stockings, Fludrocortisone, Midodrine, and Pyridostigmine. At 18 months follow-up she still had profound orthostatic hypotension with a developing gastroparesis, but no neoplasm was evident on a repeat imaging.

A literature review showed that an isolated orthostatic hypotension is a very unusual (<3% of patients) presentation for the anti-Hu paraneoplastic syndrome (Smitt et al.). We therefore present this case as to emphasise the importance of including the paraneoplastic syndromes within the differentials for an elderly patient presenting with falls and confusion.

doi:10.1016/j.jns.2015.08.722

652 WFN15-0793 Miscellaneous Topics 2 CNS-related mortality in HIV-1 infected persons in the Czech Republic

<u>D. Jilich</u>^a, A. Zjevíková^b, M. Malý^c, D. Sedláček^d. ^aDepartment of Infectious Diseases, Hospital Bulovka, Prague, Czech Republic; ^bDepartment of Infectious Diseases, University Hospital, Ostrava, Czech Republic; ^cDepartment of Biostatistics, National Institute of Health, Prague, Czech Republic; ^dDepartment of Infectious Diseases, University Hospital, Pilsen, Czech Republic

Background: CNS-disability in HIV-infected subjects is significantly involved in the morbidity and mortality. Pathologies include direct HIV-1 brain infection, opportunistic infections, and malignancies.

Objective: Analysis of all cases of CNS-related mortality in HIV-1infected persons from HIV centers was performed in the Czech Republic in the period from 1985 - to March 2015.

Patients and methods: Retrospective case-to-case data analysis using in total 336 patients' files was performed and the structure of 65 lethal cases of HIV/AIDS patients was analyzed. Their autopsies were conducted in regional hospitals in the Czech Republic and the patients' data were anonymized. An Institutional Review Board (IRB) has waived the requirement for their formal approval of the study.

Results: In the period from 1985 – to March 2015, the Czech Republic registered 2812 HIV-positive persons. 473 of them met the

CNS associated mortality in HIV-1 infected Czech Republic 1985-March 2015 (n=65)



18

criteria of AIDS. Out of the 336 documented deaths the main diagnosis of 65 persons (19.4%) was related to a CNS disability. The average age of the patients with AIDS at the time of death was 39.2 + -10.8 year. The period from the establishment of HIV-positivity to their death due to AIDS was 4.1 + -4.5 year. The most common CNS-disabilities are listed in Fig. 1.

Conclusion: Performed analysis shows that psychiatric illnesses, brain tumors and co-infections of HIV with parasitic, viral or fungal pathogens are significantly presented in total mortality rate.

doi:10.1016/j.jns.2015.08.723

653 WFN15-1251 Miscellaneous Topics 2 Autoimmune mania: when a diagnosis of exclusion is not enough

K. Steenerson, J. Drazkowski. Neurology, Mayo Clinic, Scottsdale, USA

Background: The understanding of autoimmune encephalitides is ever-expanding, but far from complete within the intersection of psychiatry and neurology. Although possible, most autoimmune and paraneoplastic entities associated with limbic encephalitis do not describe isolated neuropsychiatric symptoms, but instead usually include focal neurologic deficits as corroborative data in support of an encephalitis.

Case: The patient is a 51 year old, right-handed man who experienced a precipitous decline in behavior and memory in the last 11 months. The patient had no significant past medical history, medications-use or social history. His neurologic examination was significant only for tangential, pressured speech and mental status deficits in orientation, learning, abstraction and recall. His thorough, unremarkable workup included malignancy screen, MR imaging of CNS, EEG, lumbar puncture and serum studies including autoimmune encephalitis and paraneoplastic panels. However, CSF on pathology review showed a heretofore unclassified neural-restricted antibody staining pattern. His FDG PET was consistent with frontotemporal dementia. Due to suspicion of seronegative autoimmune-induced FTD, the patient was given 5 days of IVIG and subsequently placed on mycophenolate mofetil.

Conclusion: This case demonstrates a probable manifestation of isolated neuropsychiatric symptoms due to autoimmune encephalitis not otherwise specified, demonstrating the tenuous relationship between psychiatry and neurology in the realm of autoimmune diseases and the need to realize that the historical mantra of 'diagnosis of exclusion'' is not only not enough, but also may prove grossly neglectful in the comprehensive management of these patients. Further characterization of autoimmune staining patterns is needed to minimize the risk of incorrect diagnosis.

doi:10.1016/j.jns.2015.08.724

654

WFN15-0416

Miscellaneous Topics 2

Higher skeletal muscle mass may be associated with protective effect for ischemic stroke among community-dwelling adults: the present project

<u>S. Suk</u>^a, Y.K. Minn^b, S.H. Hwang^b, J.H. Park^c, I.S. Koh^d, J.H. Lee^e, S.Y. Kim^f. ^aNeurology, Wonkwang University Sanbon medical center and Ansan Municipal Geriatric Hospital, Gunpo-si, Korea; ^bNeurology, Hallym University Gangnam Sacred Hospitla, Seoul, Korea; ^cNeurology, Inje University Sangye

Baik Hospital, Seoul, Korea; ^aNeurology, National Medical Center, Seoul, Korea; ^eNeurology, National Health Insurance Ilsan Hospital, Koyang, Korea; ^fNeurology, Bundang Seoul University Hospital, Sungnam, Korea

Background: It is known that low skeletal muscle mass is associated with cardiovascular risk factor. However, it is unknown that sarcopenic is independent risk factor for stroke.

Objective: This study was design to establish association between low skeletal muscle mass and brain white matter changes and/or silent infarcts (WMC/SI). Methods: This is a community-based, crosssectional study supported by the regional government. Skeletal muscle mass (SMM) measurement and brain computed tomography were performed in 722 stroke- and dementia-free subjects (aged 50-75 years). Subjects were divided into quartile by SMM, which was checked by bioelectrical impedance (BIA) analysis method (InBody 770, InBody, Seoul, Korea).

Result: After adjustment for age, hypertension, diabetes mellitus, dyslipidemia, and current smoking status, the odds ratio (OR) of risk for WMC and/or SI was 0.46 in the highest quartile group (95 % confidence interval [CI] 0.22-0.98; P = 0.045). Among men, the OR of higher two quartile to lower two quartile was 0.35 (95 % CI 0.14-0.87; P = 0.024). In women OR was 0.77 (95 % CI 0.39-1.51; P = 0.452).

Conclusion: Increase of skeletal muscle mass may be associated with protective effect for brain WMC/and /or SI among community-dwelling adults without stroke and dementia. This finding is significant in men but not in women.

doi:10.1016/j.jns.2015.08.725

655

WFN15-0627 Miscellaneous Topics 2 Effects of meteorological factors on the onset of Bell's palsy in Athens, Greece

P. Zis, P. Leivadeas, <u>A. Tavernarakis</u>. Department of Neurology, Evangelismos Hospital, Athens, Greece

Introduction: The exact etiology of Bell's palsy(BP)remains unknown, although viral infection, either new or reactivation of a dormant infection is probably a major cause. The objective our study was to evaluate the effects of meteorological factors on the onset of BP in Athens,Greece.

Methods: We retrospectively looked into data of all patients presented in the Emergency Department(ED) with BP, over a period of 12 months.As per the Athens hospital rotation, the ED of our hospital is open every 4 days. Meteorological data were obtained from the official database of the National Observatory of Athens available at the website www.meteo.gr.The data included mean, minimum and maximum temperature, mean wind speed and rainfall for the day of onset, 3 days and 7 days prior to onset.

Results: Between 02/2014 and 01/2015, 126 patients (55.6% males, mean age 46.5 \pm 17.7 years) presented with BP over 91 shifts.Most patients presented in April and September(11.1%) and the lest in October(5.6%).

The difference between the maximum and minimum temperature 3 and 7 days prior to BP onset was negatively correlated to the number of BP presenting to the ED(respectively; spearman rho -0.223 and -0.208, p < 0.05).No other statistically significant correlations were observed.

Comparing the days with BP(78.0%) to the days without BP(22.0%)no significant differences were found.

Conclusion: Apart from the weak correlation of the difference between the maximum and minimum temperature 3 and 7 days prior to BP onset no other correlation was found, suggesting that

ARTICLE IN PRESS

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

the effect of meteorological factors on the onset of bell's palsy is minimal.

doi:10.1016/j.jns.2015.08.726

656

WFN15-0641 Miscellaneous Topics 2 Does cognitive decline in Parkinson's disease start before diagnosis? A population-based study

A. Foubert-Samier^a, M. Le-Goff^b, C. Helmer^b, H. Jacqmin-Gadda^b, F. Dartigues^b, H. Amieva^b, <u>F. Tison^a</u>. ^{*a*}*IMN, CHU Bordeaux, Bordeaux, France;* ^{*b*}*U897, ISPED, Bordeaux, France*

Objective: New concepts propose that Parkinson's disease (PD) starts decades before motor signs (pre-motorPD) with non-motor symptoms such as REM sleep behaviour disorder, hyposmia, constipation etc. It is accepted that 20-30% of newly diagnosed PD subjects have some cognitive decline but it is not known if it starts before diagnosis. Thanks to the very long-term follow-up study of the population based PAQUID study, we challenged the occurrence of cognitive symptoms over a 14-year period before the diagnosis of PD.

Methods: This is a case-control study nested in the PAQUID cohort. Of the 3,777 initial subjects of the cohort, 43 have developed a PD during the 14 years of follow-up. These cases were matched to 86 elderly control subjects. The evolution of scores on cognitive, functional, and depression scales was described throughout the 14-year follow-up using a semiparametric extension of the mixed-effects linear model.

Results: We have not found significant cognitive decline or emergence of depressive symptoms in future PD subjects before clinical diagnosis compared with controls. Only psychomotor speed was found to significantly decrease 4 years before PD diagnosis. Also, there was no difference in the impact on daily activities, except in using public transportation two years before diagnosis of PD.

Interpretation: This study shows that slowed psychomotor speed occurs 4 years before motor diagnosis while other cognitive functions seemed preserved until diagnosis. We could not confirm pre-motor depression in PD. Limits of our study are that of the test used and subject numbers.

doi:10.1016/j.jns.2015.08.727

657

WFN15-0283

Miscellaneous Topics 2

Combined screening for lysosomal and peroxisomal disorders by Flow Injection Liquid Chromatography Mass Spectrometry (FIA-MS/MS) in Dried Blood Spots (DBS)

<u>S. Tortorelli</u>, C. Turgeon, D. Gavrilov, D. Oglesbee, K. Raymond, P. Rinaldo, D. Matern. *Laboratory Medicine and Pathology, Mayo Clinic, Rochester, USA*

Background and objectives: Lysosomal and peroxisomal disorders are likely underdiagnosed in the adult population as shown by higher than expected prevalence of later onset disease variants found by newborn screening programs/pilot studies. We developed a rapid new method for the simultaneous analysis of six lysosomal enzymes and lysophosphatidylcholines (LPC) in dried blood spots (DBS) for screening at risk patients.

Method: DBS are extracted in buffer for enzyme activity determinations (after overnight incubation) and methanol for LPC analysis. Using FIA-MS/MS, concentrations of LPC (C26:0, C24:0, C22:0, C20:0) and reaction products of acid sphingomyelinase (ASM), β -glucocerebrosidase (ABG), α -glucosidase (GAA), α -galactosidase (GLA), galactocerebrosidase (GALC) and α -L-iduronidase (IDUA) are measured. Total FIA-MS/MS run time is 1 minute/sample.

Results: Specimens from subjects with MPS I (N = 5), Gaucher disease (N = 5), Nieman-Pick A/B (N = 2), Pompe disease (N = 5), Krabbe disease (N = 5), Fabry disease (N = 11), ALD (N = 8), heterozygous ALD (N = 5) and peroxisomal biogenesis disorders (N = 5) were correctly identified by the simultaneous analysis of the enzyme activities as well as C20 to C26 LPC concentrations in DBS by FIA-MS/MS. **Conclusions**: This method is a rapid, effective and high-throughput screening assay for six lysosomal diseases, and peroxisomal disorders using FIA-MS/MS.

doi:10.1016/j.jns.2015.08.728

658 WFN15-0695 Miscellaneous Topics 2 Epidemiologic investigation of amyotrophic lateral sclerosis in Trakya, Turkey, 2006-2010

N. Turgut^{a,*}, G. Varol Saraçoğlu^b, S. Kat^c, K. Balcı^d, O. Birgili^c, L. Kabayel^c, S. Yagıbasan^e, E. Ersoz^f, L. Kor^g, M. Küçük^h. ^aNeurology Dept., Namık Kemal University School of Medicine, Tekirdag, Turkey; ^bPublic Health Dept., Namık Kemal University School of Medicine, Tekirdag, Turkey; ^cNeurology Dept., Edirne State Hospital, Edirne, Turkey; ^dNeurology Dept., 19 Mayis University, Samsun, Turkey; ^eNeurology Dept., Corlu State Hospital, Tekirdag, Turkey; ^gNeurology Dept., Tekirdag, Turkey; ^gNeurology Dept., Optimed Hospital, Tekirdag, Turkey; ^hNeurology Dept., Medikent Hospital, Edirne, Turkey;

Background: There are no reports about the incidence and the prevalence of Amyotrophic Lateral Sclerosis (ALS) from Turkey.

Objective: The aim of the study was to estimate the incidence and prevalence of ALS in Trakya region of Turkey.

Material and methods: The ALS cases diagnosed between 2006 and 2010 were identified to study. The study used the El Escorial criteria for ALS diagnosis. Definite ALS patients included to study. We have obtained Institutional Review Board (IRB) approval.

Results: Between January 1, 2006 and December 31, 2010 we identified 87 patients (54 males, 33 females) with a new diagnosis of ALS. The mean age of diagnosis was 57.4+11.7. Ten cases were ALS with bulbar onset, 68 cases were ALS with spinal onset. The average annual incidence of ALS in the entire study was 3.95 per 100 000. On December 31, 2010, the ALS prevalence was 13.00 per 100,000. Five years fatality rate was 25.2%, and five years mortality rate was 4.67 per 100 000.

Conclusion: This is the first report on the prevalence and the incidence of ALS in a representative population of Turkey. Our incidence and prevalence rates were higher than the studies were conducted other countries. This result may be explained by an interaction with environmental factors.

Keywords: Amyotrophic Lateral Sclerosis, incidence, prevalance

doi:10.1016/j.jns.2015.08.729

659

WFN15-0596 Miscellaneous Topics 2

Photo-stimulating effect of low reactive level laser on lower urinary tract dysfunction in Parkinson disease model

<u>T. Uchiyama</u>^a, T. Yamamoto^b, Y. Watanabe^c, T. Kadowaki^c, K. Hashimoto^c, T. Shingo^d, K. Kaga^a, C. Shibata-Yamaguchi^a, T. Ymamanishi^a, R. Sakakibara^e, S. Kuwabara^b, K. Hirata^c. ^aDokkyo

Medical University, Neurourology and Continence Center, Tochigi, Japan; ^bChiba University, Neurology, Chiba, Japan; ^cDokkyo Medical University, Neurology, Tochigi, Japan; ^dDokkyo Medical University, Neurosurgery, Tochigi, Japan; ^eToho University Sakura Medical Center, Neurology, Chiba, Japan

Background: Photo-stimulation using low reactive level laser was reported to have neurobiological effects. As these effects, inhibition of Aδ- and C- fibre nerve conductions, activation of central descending inhibitory system, and suppression of local synaptic neurotransmission were reported. Micturition reflex is constructed by activation of peripheral Aδ- and C- fibre afferent nerves, and which is controlled by central descending inhibitory system. Then, the photo-stimulation will be applicable to modulate these neural controls.

Objective: Therefore, we investigate the photo-stimulating effect of low reactive level laser on lower urinary tract dysfunction (LUTD) in Parkinson disease(PD) model.

Methods: Experiments were performed on adult male Sprague-Dawley rats with bilateral injections to substantia nigra of 6OHDA (PD model). Cystometric investigation was performed, and interval time between voids, urine volume per void, and maximum bladder pressure during voiding were investigated. After 30-60 minutes' baseline recording, photo-stimulation using low reactive level laser or sham stimulation via prove was irradiated to bilateral L6/S1 intervertebral foramen via the probe contacted to body. Recording after the stimulation was continued for several hours until micturition cycle returned to baseline.

Results: Compared with the baseline record, in sham-stimulated groups, interval time between voids and urine volume per void were not unchanged. In photo-stimulated groups, interval time between voids and urine volume per void was significantly increased. These changes were stimulation-time dependent. Maximum bladder pressure was unchanged.

Conclusion: Photo-stimulation using low reactive level laser to bilateral L6/S1 root improved storage dysfunction without exacerbation of voiding dysfunction in LUTD of PD model.

doi:10.1016/j.jns.2015.08.7230

660 WFN15-1512 Miscellaneous Topics 2 A case series of Joubert syndrome

<u>N. Vasudevan</u>^a, S. Balakrishnan^a, L. Ranganathan^a, C. Leemapauline^b, D. Arjundas^c, K. Bhanu^a. ^{*a*}Neurology, Madras Medical College, Chennai, India; ^{*b*}Neurology, Institute Of Child Health And Madras Medical College, Chennai, India; ^{*c*}Neurology, Mercury Hospital, Chennai, India

Background: Joubert syndrome (JS) is a rare autosomal recessive disorder with the key finding of cerebellar vermis hypoplasia with a complex brainstem malformation that comprises the molar tooth sign (MTS) on MRI.

Objective: To study the clinical features and radiological pattern in short series of cases presented with cerebellar ataxia and developmental delay ,with MRI showing "MOLAR TOOTH SIGN".

Material and methods: Series of cases attending Paediatric neurology department with complaints of unsteadiness and developmental delay were analyzed clinically and evaluated with MRI brain. The joubert diagnostic criteria is used to diagnose the condition.

Case vignette: In our case series of four patients with JS, case one and two are siblings of age five[girl] and two[boy] years respectively. Both presented with developmental delay. On examination they had hypotonia and ataxia. MRI revealed MTS.

Case three is a six year old girl presented with developmental delay and difficulty in walking.Clinical examination revealed oculomotor apraxia, hypotonia, ataxia. MRI showed MTS. Case four is a two month old boy presented with abdominal distention, abnormal eye movements. Clinical examination revealed absent social smile, nystagmus, hypotonia, hepatomegaly. USG abdomen showed polycystic kidneys, hepatic fibrosis. MRI showed MTS.

Conclusion: Joubert syndrome is rare congenital malformation of CNS presenting as Cerebellar ataxia with delayed development. MRI Brain and clinical criteria is useful in diagnosing this condition.

1. I have obtained patient and/or Institutional Review Board (IRB) approval, as necessary.

2. An Institutional Review Board (IRB) and/or Animal Use Committee have waived the requirement for their formal approval of the study.

doi:10.1016/j.jns.2015.08.731

661

WFN15-0817 Miscellaneous Topics 2 Mild encephalitis with reversible splenial lesion and anti-NMDA receptor encephalitis

<u>A. Vitali da Silva</u>^a, M.A. Pires Lazaro Fay Neves^a, C. Parizotto^a, O. Batista de Rezende Filho^a, T. Koltermann^b. ^aNeurology, Irmandade Santa Casa de Londrina, Londrina, Brazil; ^bNeurology, Moinhos de Vento Hospital, Porto Alegre, Brazil

Mild encephalitis with reversible splenial lesion (MERS) was described in patients receiving antiepileptic drugs and associated with infectious diseases. There is no report about MERS and autoimmune encephalitis.

Case Report: A 24-year-old man was admitted with emotional and behavioral disturbances, including decreased cognitive skills, psychosis and hypersexuality. These symptoms progressed without prodromal phase during the course of 3 weeks. Neurological





ARTICLE IN PRESS

Abstracts / Journal of the Neurological Sciences (2015) xxx-xxx

examination on admission was normal, except for the mental status, as well as the laboratory studies. CSF was clear and revealed the presence of 13 leukocytes (99% lymphocytes), but normal protein and glucose contents. Brain MRI showed a well-defined lesion in the splenium of the corpus callosum hyperintense on T2WI, slightly hypointense on T1WI, associated with a restricted diffusion and no contrast enhancement. He was treated empirically for herpes virus and borreliosis until investigation was completed. Infection and vasculitis sources assays were negative. Anti-Hu was also negative, and anti-NMDA receptor tested positive in the CSF. Neoplasms were excluded by abdominal and thoracic CT, testicular US and CA19-9. The patient did not receive specific anti-NMDA receptor encephalitis therapeutics since he recovered throughout these weeks before results were ready. The MRI repeated after four weeks came out normal. He was discharged with near normal mental status, and recovered fully after six weeks.

This is the first case described about MERS originated by anti-NMDA receptor encephalitis, whose association enhances the immunological etiology of this clinical-radiological syndrome.

Figure: DWI and FLAIR MRI showing hyperintensity of the splenium of the corpus callosum

doi:10.1016/j.jns.2015.08.732

662 WFN15-0386 Miscellaneous Topics 2 Spectrum of neurological presentations in an outpatient clinic of Rural Zimbabwe

<u>M.V. Vyas</u>^a, A. Wong^b, J.M. Yang^c, P. Thistle^d, L. Lee^b. ^aDivision of Neurology Department of Medicine, University of Toronto, Toronto, Canada; ^bDivision of Neurology Department of Medicine, Sunnybrook Health Sciences Centre & University of Toronto, Toronto, Canada; ^cDepartment of Psychiatry, University of Toronto, Toronto, Canada; ^dDepartment of Obstetrics and Gynecology, University of Toronto University of Zimbabawe and Karnada Mission Hospital, Mt Darwin, Zimbabwe

Background: Previous studies to estimate burden of neurological disorders in Africa are limited to inpatients in urban hospitals. The spectrum of neurological conditions in rural Africa remains unclear. **Objective**: To determine the spectrum of neurological presentations in an outpatient setting in rural Zimbabwe.

Methods: Clinical data was collected from outpatient records at Karanda Mission Hospital, a rural community hospital in Northern Zimbabwe from February 2013 to February 2014. Each patient visit was entered in an outpatient record book by a registered nurse or a nurse trainee. Demographic details such as age, sex, weight and address of the patient, and clinical details such as diagnosis on discharge and medications prescribed were recorded in the record book following assessment by a physician or nurse practitioner. Each visit corresponded to a separate entry in the study.

Results: We recorded a total of 19 206 visits in the outpatient registry. The average age was 46.41 (standard deviation = 21.46), and there were more visits from women (57.81%). 11.63% (2 233) of all visits had a neurological diagnosis at discharge. The most

common neurological diagnoses were epilepsy/seizures (24.38%), followed by neuropathies (13.63%), headaches (11.4%) and strokes (4.6%).

Conclusions: One in ten cases in an outpatient setting in rural Zimbabwe were neurologically related. Further studies are required to determine the public health burden of neurological disorders in rural Africa. The development and funding of educational initiatives in resource-limited areas is needed to improve neurological diagnosis and care.

doi:10.1016/j.jns.2015.08.733

663 WFN15-0218 Miscellaneous Topics 2 Examination and effectiveness of traditional treatments used in neurological disorders in the inner part of Aegean region

<u>M. Yaman</u>^a, S. Oruc^a, H. Demirbas^a, F. Karakaya^b, G. Koyuncu^c, Z. Toklu^d, R. Demirel^e. ^aNeurology, Afyon Kocatepe University School of Medicine, Afyonkarahisar, Turkey; ^bNeurology, Isparta State Hospital, Isparta, Turkey; ^cNeurology, Nigde State Hospital, Nigde, Turkey; ^dNeurology, Afyonkarahisar State Hospital, Afyonkarahisar, Turkey; ^ePublic Health, Afyon Kocatepe University School of Medicine, Afyonkarahisar, Turkey

Introduction: Human beings looked for the remedy for the diseases in nature for thousands of years fight. In this study, we aim to determine which kind of traditional methods are used against neurological diseases and symptoms by individuals who live in Afyonkarahisar and other cities in the inner part of the Aegean region. We also would like to show the efficiency of these healing methods.

Materials and methos: This study was conducted between 2013 and 2014. We visited all provinces inner Aegean region. We have questioned the traditional medical methods that are used recently and were used in the past in order to cope with neurological disorders and symptoms. Additionally, the confidence was also questioned against these methods.

Findings: We have found that 131 plants, 33 plant mixture and 2 other techniques were used as a traditional remedial treatment for the all systematic disorders and symptoms. Out of these, we have detected that 67 plant, two plants mixture and two other techniques were used in neurological disease and symptoms. Societies accept these products as an adjunctive therapy.

Discussion: There should be further studies for the usage of these 8 plants (capers, beans, Akbaşlı, cinnamon, poppy, licorice, buckwheat seed, rose), two methods. People living in the inner Aegean choose predominantly the modern medical methods. Traditional methods are regarded as aftercare. However, people choose traditional methods instead of modern ones in order to heal the semptoms such as headache and drowsiness. Praying to God is also regarded as afteraid for disorders.

doi:10.1016/j.jns.2015.08.734