WCN 2013 TC 36 Emergency Neurology

Management of Acute Headache

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Classification of Headaches ICHD3

Primary headaches

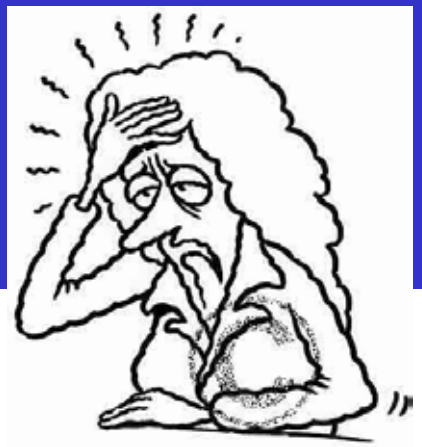
1. Migraine
2. Tension-type headache
3. Trigeminal autonomic cephalalgias
4. Other primary headache disorders

Secondary headaches attributed to

5. Trauma or injury to the head and/or neck
6. Cranial or cervical vascular disorder
7. Non-vascular intracranial disorder
8. Substance or its withdrawal
9. Infection
10. Disorder of homoeostasis
11. Disorder of cranium/neck/eyes/ears/nose/sinuses/teeth/mouth/cervical
12. Psychiatric disorder

Painful cranial neuropathies classical / secondary

Headache in the Emergency Department



Main complaint in 1-16% of all visits

- **Mostly young adults, female preponderance**
- **Mostly primary headaches, serious conditions in 5-15%**

Top priority = precise etiologic diagnosis

Crucial part = interview

Diagnosis determines management of the patient

Symptomatic treatment may be needed but a good response should not be a reason for postponing etiologic investigations

Have you ever had this type of headache before?
When did your actual headache start?

Patient able to say that he has **already suffered** from several **similar** headaches for **months or years** and that he is recognizing a usual headache attack

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A **primary headache disorder** is most likely

Diagnosis = **detailed interview**

Treatment relies on **specific acute headache treatments** as in/out patient

Patient reports having headaches for the **first time in his life** for hours/days/weeks/months

Patient reports history of definite primary headaches but states that his acute headache is **different** from his usual headaches attacks

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A **secondary headache disorder** has always to be excluded

Diagnosis = **emergent investigations**

Treatment of the **underlying cause**

Have you ever had this same type of headache before?
When did your actual headache start?

Patient has already suffered from several similar headaches for months or years and is recognizing his usual headache attacks

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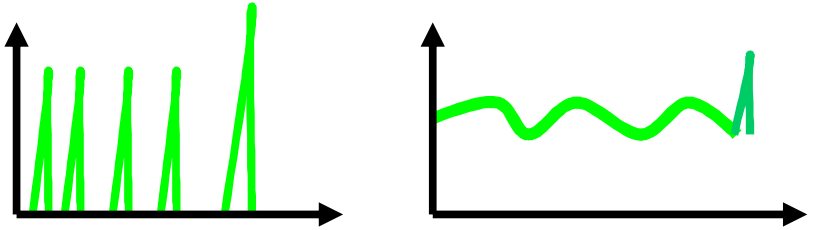
A primary headache disorder is most likely

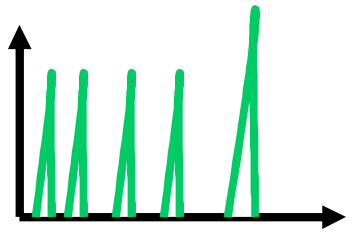
Diagnosis = detailed interview

→

Do you have headache attacks or do you have headaches all the time?



Attack of a Primary Headache Disorder



Since months or years

Migraine > tension type > cluster headache

Patient says he has already suffered from several similar headaches for months or years

Make a precise diagnosis (duration of attacks)

- Interview about characteristics of headache attacks
- Age of onset, duration, localisation, intensity, type, associated signs and symptoms, triggers
- Normal neurologic or physical examination
- Investigations not necessary, normal if done

Specific acute headache treatments

Primary Headache Attacks : Diagnosis

Migraine without aura

- 4-72 hours, irregular frequency of attacks all over the year
- Moderate or severe, ? by physical activity, unilateral, pulsating (2/4)
- Nausea/vomiting and/or photophonophobia
- Migraine with aura : aura is characteristic

Cluster headache

- 15-180 minutes; 1-8 attacks/day; periodicity; night, regular
- Severe to very severe, unilateral, always same side, agitation
- Autonomic signs: eyelid edema, miosis/ptosis, tearing, nasal congestion
- Mostly episodic, rarely chronic

Tension type headache

- 30 minutes to 7 days, limits not clear
- Mostly bilateral, pressing or tightening, not ?CH by physical activity
- Setting of stress, anxiety or depression or “can no longer cope”

Classical trigeminal neuralgia

- Severe, electric shock like or stab, seconds to 2 minutes, trigger zone

Primary Headache Attacks: Treatment

Migraine: treatment varies according to local protocols

- Specific drugs: subcutaneous sumatriptan or infusion of DHE
- Nonspecific drugs: IV paracetamol or NSAIDs
- Add antiemetics and/or tranquilizers (clorazepate 20-50 mg)
- IV fluids (vomiting), ice packs, calm/quite room, deep relaxation
- Intractable migraine: IV amitriptyline, IV sodium valproate
- *Status migrainosus*: hospitalisation may be requested

Cluster headache

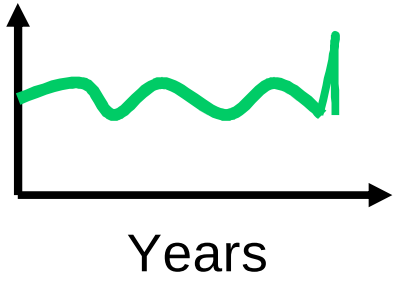
- Attack : subcutaneous sumatriptan and/or high flow oxygen 15L/min
- Initiate prophylaxis: verapamil 120 mg
- Consider transitional treatment with steroids: oral or GON injections

Tension type headache

- If needed IV paracetamol or NSAIDs often with tranquilizers

Classical trigeminal neuralgia: carbamazepine

Chronic Daily Headache
« I have headache all the time since years »



Mostly primary headaches

- **Chronic migraine**
- **Chronic tension-type headache**
- **Chronic post-traumatic headache**
- **Medication overuse**
- **Can present to an emergency department in the setting of stress, anxiety, depression or “can no longer cope”**

Investigations mandatory if never done before to exclude a secondary headache disorder

Management by headache specialist

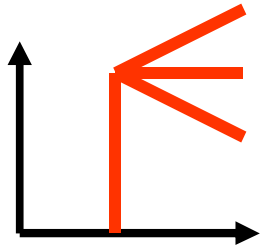
Have you ever had this type of headache before? When did your actual headache start?

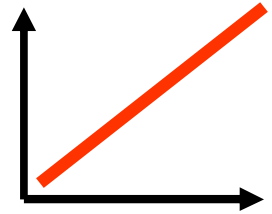
Patient reports having headaches for the first time in his life for hours/days/weeks/months

Patient reports history of definite primary headaches but states that his acute headache is different from his usual headaches attacks

A secondary headache disorder has always to be excluded

How did the headache begin?
How long the headache took to peak?
How has the pain changed since its onset?





Sudden Vascular Disorders >>> Nonvascular disorders

Progressive Nonvascular Disorders >> Vascular disorders

Unusual Acute Headache: Interview Headache Characteristics ?

Mode of onset: time to peak

Maximum severity

- 11-point scale (0-no pain to 10-worst ever pain)
- Headache can be mild in a serious condition

Type and location of pain: not specific

Spontaneous or triggered: Valsalva, exertion, sexual activity…

Avoids standing up: intracranial hypotension, cerebellar stroke

Avoids lying flat: SAH, intracranial hypertension, cerebral venous thrombosis, sinusitis

Unusual Acute Headache: Interview Circumstances? Medical history?

Mild trauma: subdural hematoma, cervical arterial dissection, intracranial hypotension

Intake of vasoactive substances: serotoninergic and sympathomimetic medications, illicit drugs: reversible cerebral vasoconstriction syndrome (RCVS)

Dural puncture: intracranial hypotension

Fever: infectious disorders

Postpartum: RCVS, cerebral venous thrombosis, eclampsia, post dural puncture intracranial hypotension

Ear, nose, and throat symptoms: complicated sinusitis

Cardiovascular disease and hypertension: stroke

Cancer: cranial metastases

Unusual Acute Headache: Interview Associated Symptoms?

Any central neurological symptoms: consciousness impairment, seizures, focal deficits): intracranial disorder

Visual symptoms: eclipses, visual loss, diplopia : intracranial hypertension

Any headache in a >50 years old: giant cell arteritis

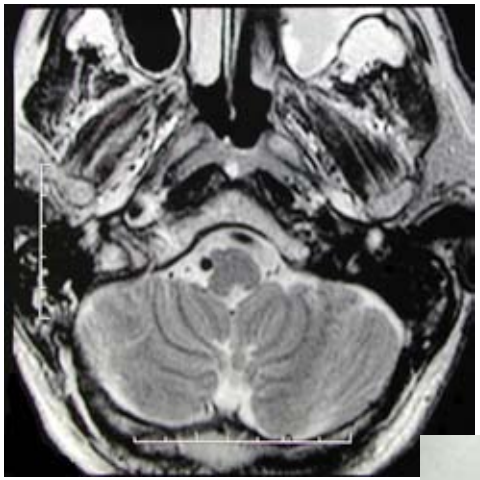
Fever: infectious disorders

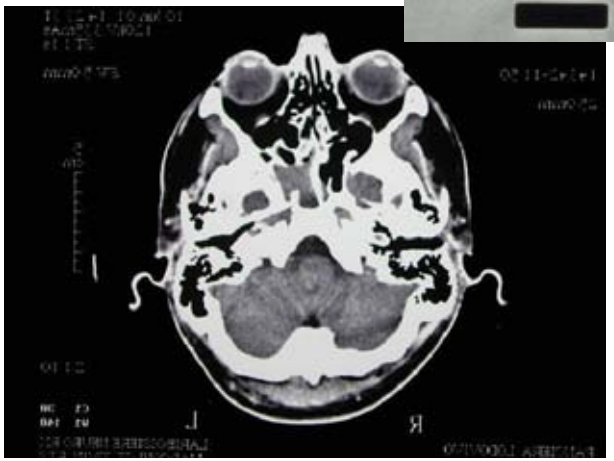
Nausea and vomiting: non specific

Unusual Acute Headache
Clinical Examination

- **Any abnormality in the clinical examination increases the need for rapid evaluation**
- **General:** blood pressure, temperature, skin
- **Consciousness and neck**
- **Always check eyelids, pupils, visual field, and cerebellar function for subtle signs**
  - Cerebellar ataxia (stroke)
  - Hemianopia (stroke)
  - Unilateral mydriasis (aneurysm)
  - Complete third nerve palsy complet (aneurysm)
  - Myosis and ptosis (carotid artery dissection)
  - Sixth nerve palsy (intracranial hypertension or hypotension)
- **Fundoscopy**

All these 3 males have an acute, recent, unusual right sided headache





The absence of any associated symptoms and a strictly normal examination do not exclude a serious cause

Urgent diagnostic work-up is still needed

Investigations are guided by the list of all possible underlying causes

Thunderclap Headache

High intensity headache of abrupt onset mimicking that of a ruptured cerebral aneurysm

- **Peaking in < 1 minute, no limits for duration**

Often reveal serious causes

Investigations should be expedient and exhaustive

The absence of any associated symptoms and a strictly normal examination do not exclude a serious cause

Diagnosis is based on plain brain CT and, if tomogram is normal, on lumbar puncture but these first line investigations can be normal in several serious conditions

Subarachnoid hemorrhage

11-25% of patients with thunderclap headache

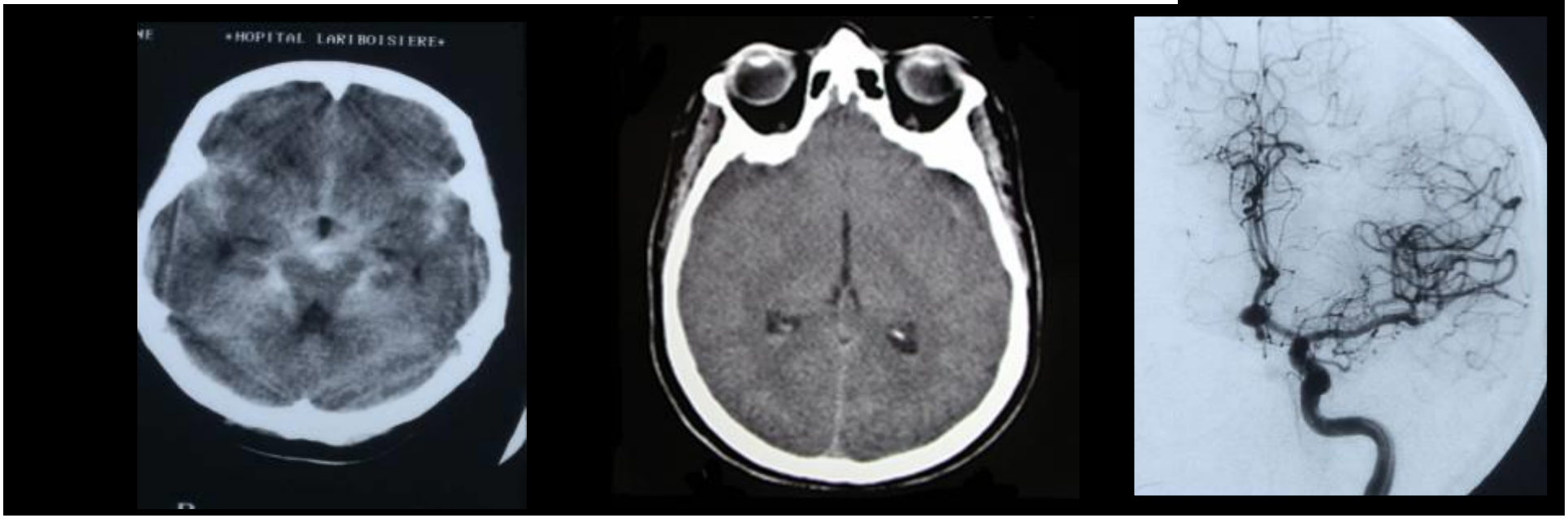
70% present with headaches, 50% thunderclap

CT : sensitivity decreases with time

MRI (FLAIR, T2*) : superior to CT after day 1

CSF analysis after normal imaging

Ruptured aneurysm 85% => angiography



Other intracranial hemorrhages

Intracerebral hemorrhage ICH

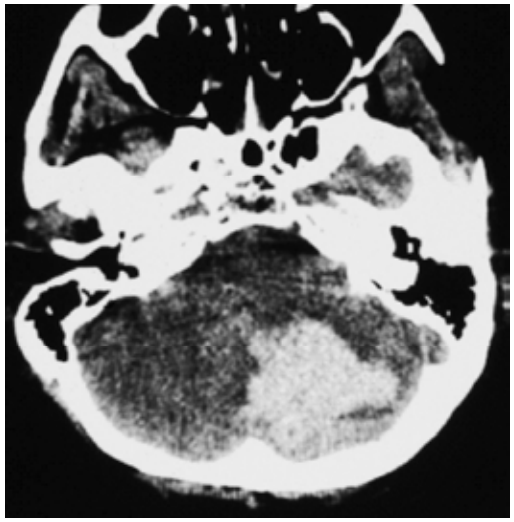
Headache >50% but often overshadowed by focal deficit and/or coma

Isolated (thunderclap) headache: cerebellar ICH, frontal, temporal or occipital ICH (non dominant hemisphere)

Intraventricular hemorrhage and acute subdural hematoma

Isolated headache

Plain CT, IRM T2*

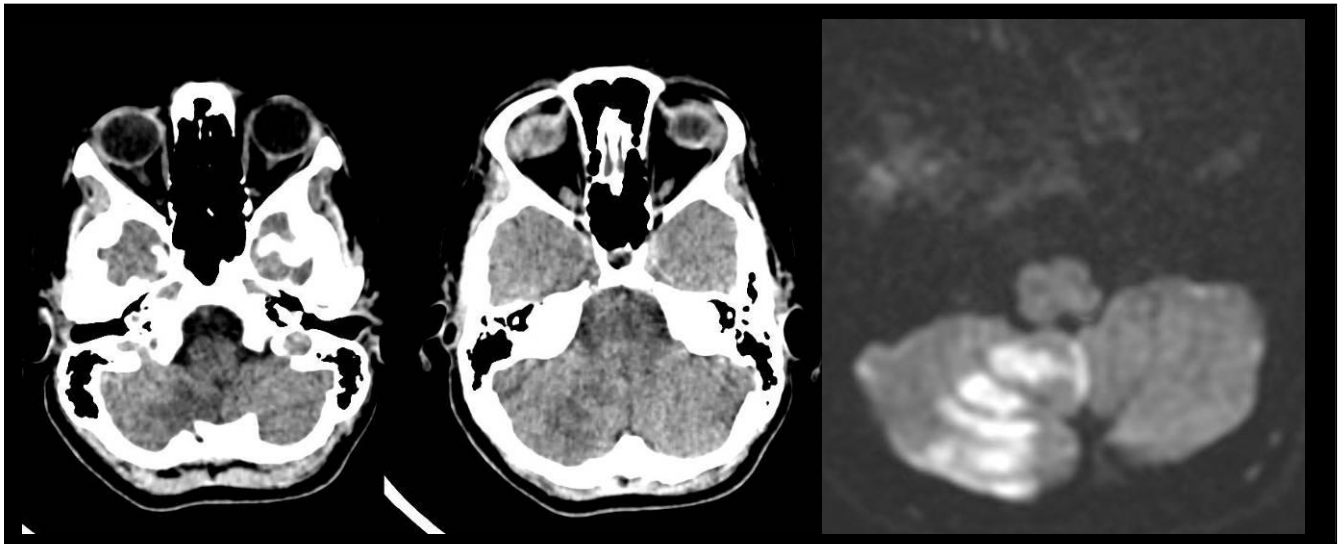


Ischemic stroke

Headache in 17-34% of all ischemic stroke

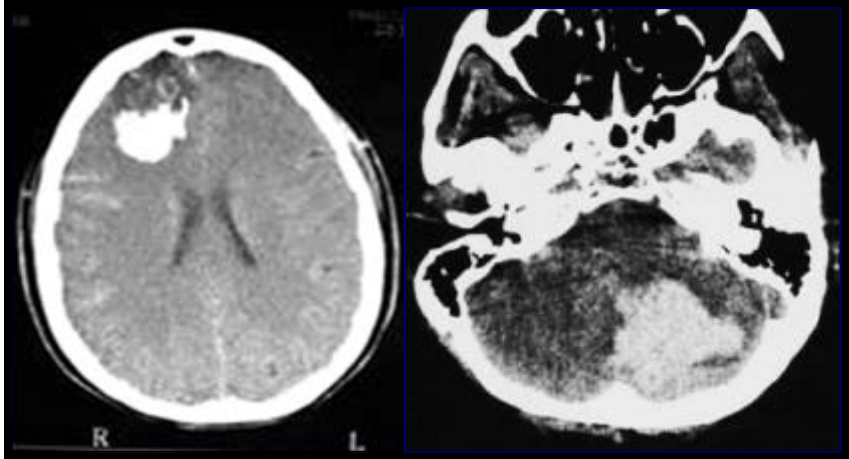
- Often overshadowed by focal deficit and/or coma
- Isolated (thunderclap) headache possible: cerebellar, temporal or occipital regions

Headache in ischemic stroke points to certain etiologies cervical artery dissection, RCVS, SAH followed by vasospasm, angeitis

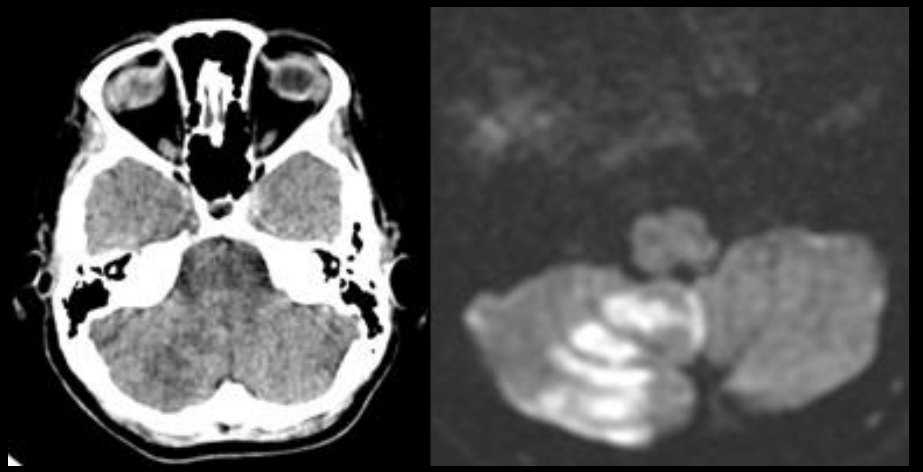


MRI diffusion weighted images +++

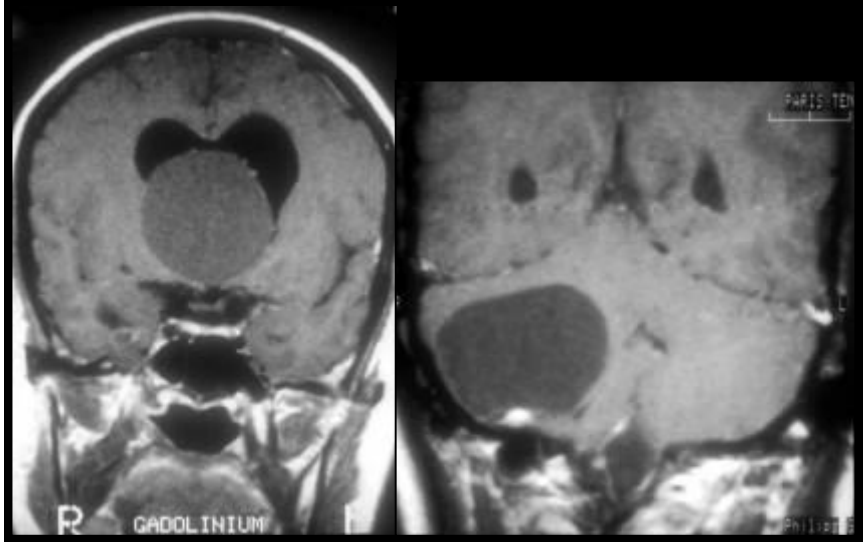
Other causes detected by CT and LP 10-12%



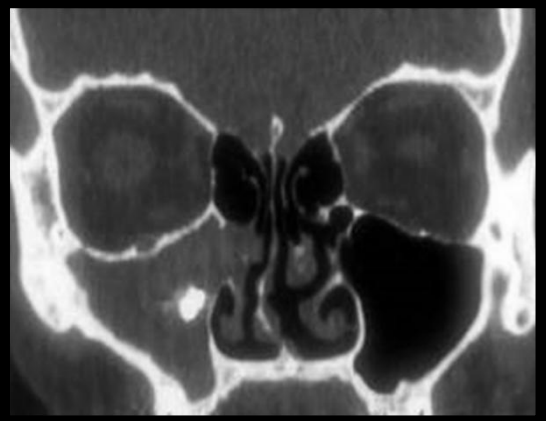
Other intracranial hemorrhages 5-10%



Ischemic stroke (rare)



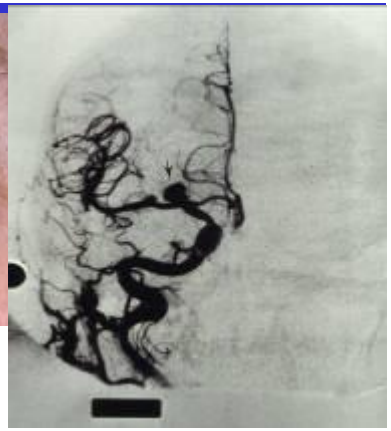
Hydrocephalus and tumours



Sinusitis

Meningitis 2%

Warning leaks?



Misdiagnosis of SAH 30%

50% aneurysmal SAH report previous thunderclap H

Painful third nerve palsy revealing aneurysm

=> Fear of warning leaks

Prospective series of sudden onset headache (CT and LP)

11-25% SAH, 10-12% stroke/tumour/meningitis

70% no cause and good outcome

=> Concept of « benign thunderclap headache »

=> Suggestion that cerebral angiography is not necessary in cases with thunderclap headache with normal CT and normal LP

34 yo female

Normal delivery 25/09/2007

Day 0, 26/09 first TCH

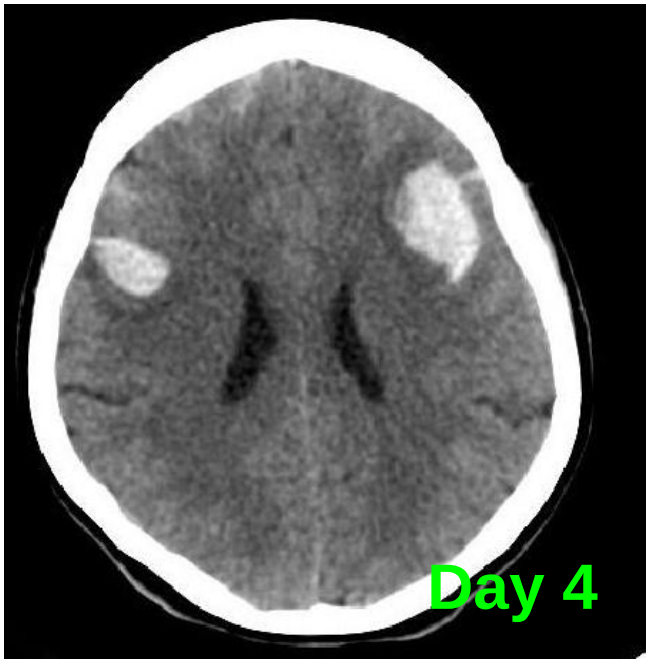
Day 1 normal CT scan

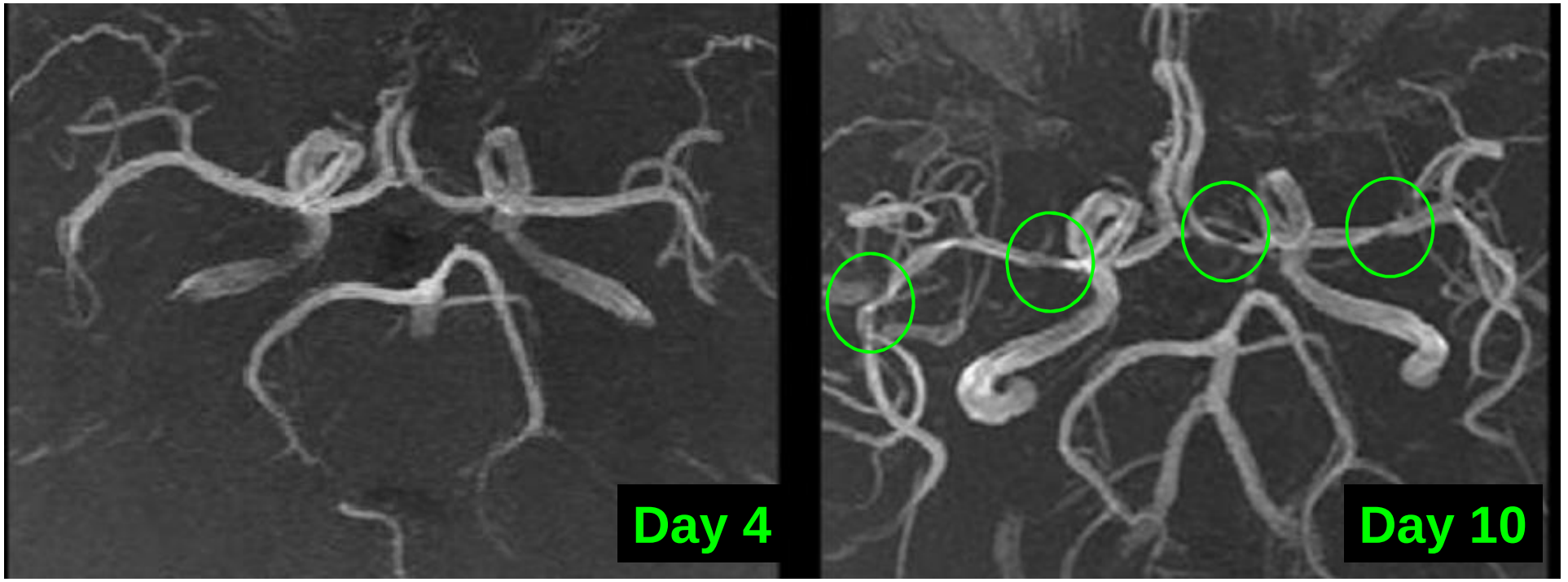
Day 3 2nd TCH + aphasia

Day 4 CT bilateral frontal ICH + cSAH, normal MRA and conventional angiogram

Day 6 partial seizure

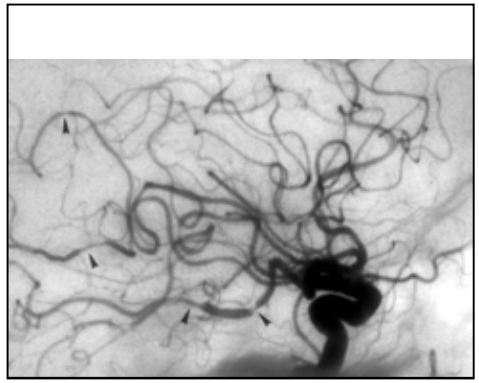
Day 10 third TCH





Reversible Cerebral Vasoconstriction Syndrome (RCVS) => Clinico-radiological syndrome

Calabrese LH, Dodick DW, Schwedt TJ, Singhal AB. Ann Intern Med. 2007; 146: 34-44



Acute severe headaches (often thunderclap) ± focal deficits or seizures

Segmental vasoconstriction of cerebral arteries on angio (CT, MR or IA)

Uniphasic course: no new symptoms >1 month

No evidence of aneurysmal SAH

Normalisation of arteries <12 weeks of onset

Isolated TCH in 75% of cases: RCVS accounts for most « benign thunderclap headache »

All ages, female preponderance

Stroke rare but often severe and sometimes lethal

Multiple causes (postpartum, vasoactive medications/drugs)

RCVS Clinical Presentations

Purely cephalalgic 55-75%

Recurrent thunderclap headaches over 1-4 weeks 94%

80% TCH triggered: sexual activity, leaning down, exertion, singing, emotion, defecation, cough, sneezing, shower, urination, laughing…..

=> RCVS, the most frequent cause of recurrent TCH

Headaches + other symptoms: 25-45%

Seizures 5-17%

Focal deficits 20-43%: transient (TIAs/aura like)

persistent (stroke)

Catastrophic, sometimes lethal < 2%

Multiple strokes and intractable vasoconstriction

Chen, Neurology 2006; Ducros, Brain 2007& Stroke 2010; Singhal A, NEJM 2009 & Arch Neurol 2011

Stroke in RCVS: a Dynamic Process

Intracranial hemorrhage 33%

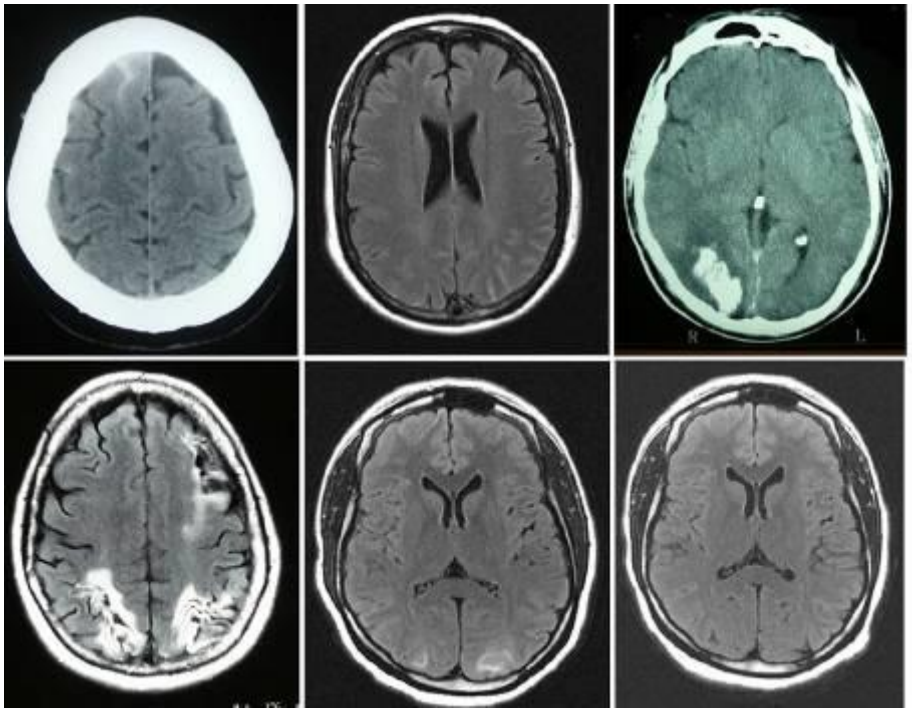
- Any variety of hemorrhage
- Convexity subarachnoid hemorrhage, intracerebral or subdural (associations)
- Early event 1st week
- 17% with normal initial CT

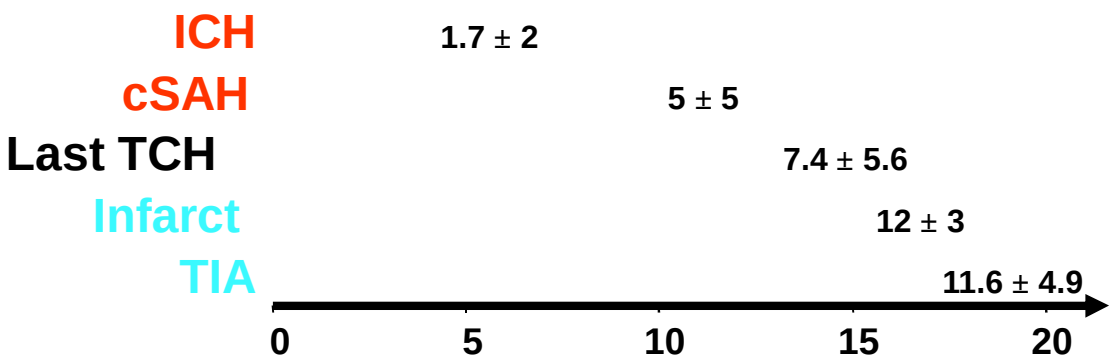
PRES 8-40%

- Early event 1st week

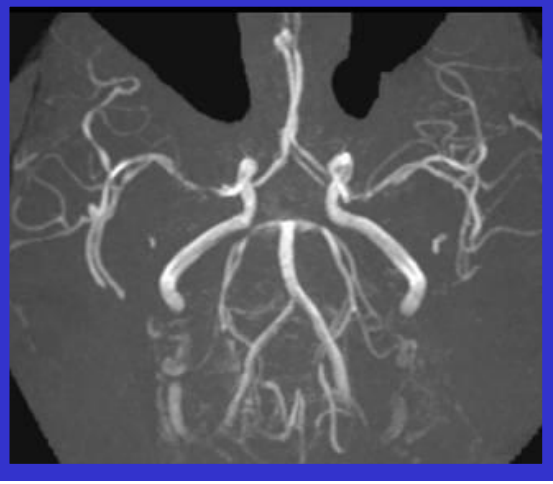
Infarcts 6-39%

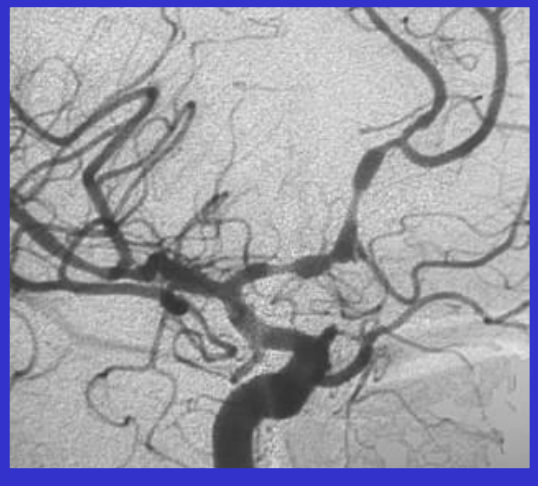
- Later on 2nd week





RCVS Diagnosis and Management





Sensitivity of investigations is incomplete but increases with time

Repeat brain imaging: Normal initial imaging in hemorrhagic RCVS 17%

Image cerebral vasoconstriction

- **Initial normal angiogram 20-40% (MRA/CTA, transfemoral rarely)**
- **MRA maximal at D16±10, close to headache resolution** *(Chen, Ann Neurol 2010)*
- **Intracranial velocities TCD maximal at D18-25** *(Chen, Ann Neurol 2008)*

Image cervical arteries: 12% cervical artery dissection *(Mawet, Meurology 2013)*

Prove reversibility of vasoconstriction < 3 months

Rest, removing vasoactive substances, nimodipine

Mostly good outcome, few permanent deficit, Case fatality < 1%

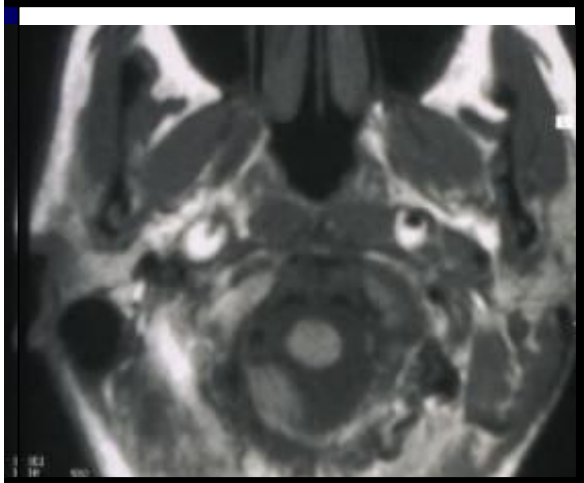
What are the Less Common Causes of Thunderclap and Sudden Headaches ?

Causes detected by plain CT or lumbar puncture

RCVS is possibly missed by plain CT and lumbar puncture

Several case series showed that some etiologies of thunderclap headache cannot be ruled out clinically and can present with normal plain CT and normal cerebrospinal fluid

Causes of TCH which can present with normal CT/LP



Cervical Artery Dissection

Isolated pain 8%

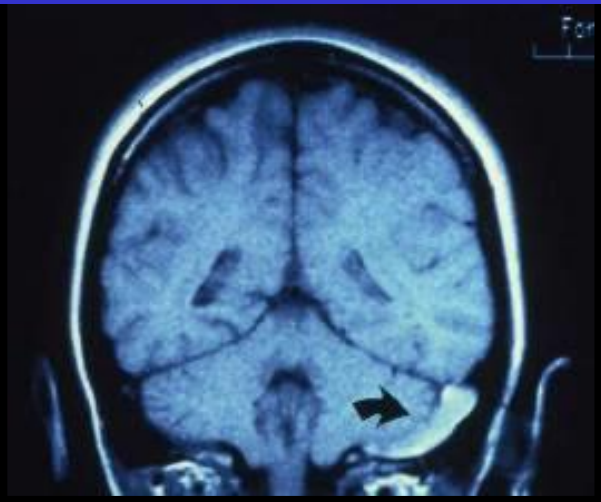
TCH 20%

Pain precedes stroke

Image cervical and cerebral arteries

- CT or MR angio
- Fat sat T1WI

Antithrombotics



Cerebral venous thrombosis

Inaugural TCH 2-16%

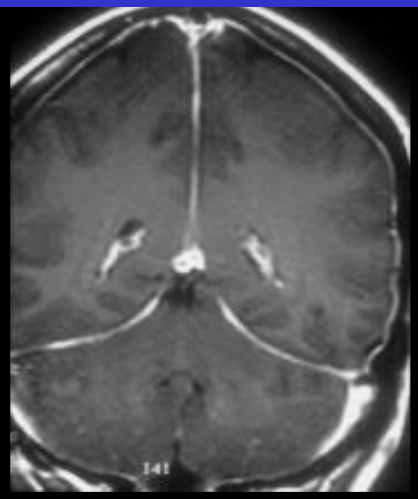
Elevated CSF pressure

Pain precedes stroke

Image cerebral veins to visualize thrombus

- IRM T1 T2, T2*
- Sinus manquant

Heparine



Intracranial hypotension

Inaugural TCH 15%

Orthostatic headache

Low CSF pressure

Complic= SDH and CVT

MRI signs PMGE, cranio-caudal descent, subduralcollections

Epidural blood patch

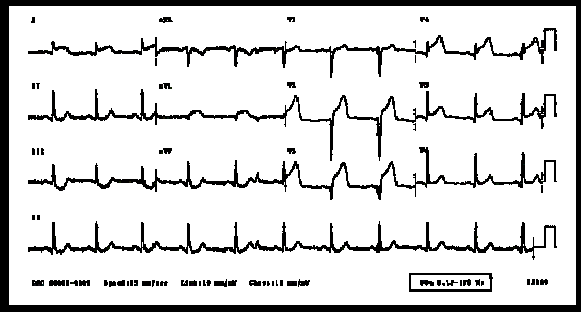
Other causes of sudden headache which can present with normal CT and normal LP

Posterior reversible encephalopathy syndrome

Pituitary apoplexy

Spontaneous retroclival hematoma

Giant cell arteritis



Myocardial ischemia

Aortic arch dissection

Etiologies of recent headache with progressive subacute onset

Causes diagnosed by physical examination followed by plain CT, and when CT is normal, by LP

Intracranial hypertension with abnormal CT

- **Space occupying lesion (tumour/abcess/subdural hematoma)**
- **Hydrocephalus**

Meningitis and meningoencephalitis

ENT and eye disorders: sinusitis, glaucoma…

Any causes of TCH can also present with progressive headache !!!

Other etiologies of acute headache with progressive onset which can present with normal CT

- **Any causes of TCH can present with progressive headache !!!**
- **Causes of intracranial hypertension with normal CT**
  - **Chronic meningitis**
  - **Cerebral venous thrombosis**
  - **Dural fistula**
  - **Hyperproteinorachia (horse tail tumour, PRN)**
  - **Idiopathic intracranial hypertension**
- **Intracranial hypotension (post dural puncture or with spontaneous CSF leak)**
- **Giant cell arterities**
- **CO poisoning**

Idiopathic intracranial hypertension

- Rare disorder
- Female predominance, obesity
- Normal imaging (CT, MRI + gado, MRV, MRA) + no hormonal or toxic causes
- Headache 75-99% progressive or rapidely increasing
- Visual symptoms 80%
  - Eclipses, bilateral decrease of VA
  - Diplopia with VIth nerve palsy
  - Bilateral papilloedema 95-100%
- Tinitus, nausea
- CSF pressure > 25 cm
- Tt : diet + LP + acetazolamide
- Surgery if intractable headaches/papilledema/visual loss

Diagnosis of Acute Headache

- **Rapid interview distinguishes a primary headache attack from a suspected secondary headache disorders**
- **Characterise primary headache attack and provide a specific treatment, discharge the patient with a treatment plan to avoid early readmission to the emergency department**
- **When a secondary headache disorder is suspected, the absence of any associated symptom and strictly normal examination do NOT exclude a serious cause, and investigations should always be performed**

18 years-old male without past medical history

- Sudden headache at 3:30 PM while working on his computer
- Headache peaked in 10 secondes reaching 9 on the 11-point verbal scale
- Bilateral and diffuse headache, pulsating, phono and photophobia
- Clinical examination at 5:20 PM
  - Painful but calm
  - Supple neck
  - Body temperature 36,8°C
  - Blood pressure 130/75 mmHg

A strictly normal clinical examination does not exclude a subarachnoid hemorrhage

SAH is initially misdiagnosed in ?Â of 3 patients

Investigations in Acute Headaches

- **Blood tests, ESR and CRP, electrocardiogram**
- **Plain head CT followed by LP if CT is normal**
- **Timing of LP**
  - **12 hours after headache onset (spectrophotometry)**
  - **Risks: meningitis, early rebleeding of ruptured aneurysm (15%)**
- **Further investigations after normal CT and LP**
  - **Cervical and cerebral angiography : alternate diagnosis**
  - **Normal CT/LP/angiogram: brain MRI may show cortical CVT, pituitary apoplexy, PRES, intracranial hypotension**
- **Four vessel angiography nowadays rarely necessary**

Conclusions

Acute headache, either of the thunderclap type, or sudden, or rapidely progressive is a warning symptom pointing to numerous underlying causes

Plain CT and LP whenever the CT is normal diagnose several causes

Several vascular disorders can present with isolated acute headaches, and, except intracranial haemorrhages, may be missed by plain CT and lumbar puncture

Large prospective series to establish the respective frequencies of underlying causes and to evaluate the diagnostic yield of angiography and brain MRI after normal CT and LP

Systematic cervical and cerebral CT or MR angiography