Chapter 103 – Headache Disorders

Episode Overview:

1. Classify headache based on the IHSC (international headache society classification).
2. What are the IHSC for migraine without aura (common migraine) and migraine with aura (classic migraine)
3. Describe the pathophysiology of migraine
4. Outline a stepwise approach to treating acute migraine attacks? List 8 therapy options for Migraine.
5. List five migraine variants.
6. What can be done to prevent migraine headache recurrence? List 8 common precipitants.
7. What are the clinical features of a cluster headache? How are they treated? Tension headache?
8. List 6 causes and 6 risk factors for SAH.
9. Describe a clinical grading scale for cerebral aneurysms and SAH and how it relates to prognosis.
10. Describe a diagnostic strategy for SAH.
11. List 6 components of management for SAH.
12. What are features of a HA from CNS malignancy? What primary cancers spread to CNS? What are two components of management of a new CNS malignancy?
13. Describe clinical features and treatment of carotid and vertebral artery dissection
14. List 8 common clinical features for patients with giant cell arteritis; and its Dx and Rx.
15. What are the criteria to diagnose idiopathic intracranial hypertension? How is it managed?

Wisecracks

1. How do migraines differ between children and adults?
2. Describe four unique presentations of migraine headache.
3. What is the pathophysiology of idiopathic intracranial hypertension? Describe the typical patient.
4. List 3 LP factors that reduce the incidence of post-LP HA; and describe the management of post-LP HA.
5. Describe post-traumatic headache & hypertensive headache.
6. What is reversible cranial vasoconstriction syndrome?
7. List 10 HA Red Flags.

Rosen's in Perspective

Hey! Check out https://canadiem.org/crackcast-e020-headaches/
For a basic approach to headaches, red flags, ddx and the fundamentals!

1) Classify headache based on the IHSC (international headache society classification)

Primary	**Secondary** "Organic, identifiable, distinct pathologic process"	**Others:**
1. Migraine 2. Cluster 3. Tension	1. H/A attributed to trauma or injury to the head or neck 2. Cranial or cervical vascular disorder 3. Nonvascular intracranial disorder 4. A substance or its withdrawal 5. Infection 6. Disorder of homeostasis 7. Headache or facial pain attributed to disorder of cranium, neck, 8. Eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures	PAINFUL CRANIAL NEUROPATHIES, OTHER FACIAL PAINS, AND OTHER HEADACHES
Other benign primary headaches	9. Headache attributed to psychiatric disorder	

2) What are the IHSC for migraine without aura (common migraine) and migraine with aura (classic migraine)

- 80% of migraines are without an aura.
- Migraine with aura is primarily characterized by the transient focal neurological symptoms that usually precede or sometimes accompany the headache. Some patients also experience a premonitory phase, occurring hours or days before the headache, and a headache resolution phase. Premonitory and resolution symptoms include hyperactivity, hypoactivity, depression, cravings for particular foods, repetitive yawning, fatigue and neck stiffness and/or pain. - From ichd-3.org

Here are the classifications: Copied from https://www.ichd-3.org/1-migraine/1-2-migraine-with-aura/

1.1 Migraine without aura

Previously used terms:

Common migraine; hemicrania simplex.

Description:

Recurrent headache disorder manifesting in attacks lasting 4-72 hours. Typical characteristics of the headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity and association with nausea and/or photophobia and phonophobia.

Diagnostic criteria:

A. At least five attacks[1] fulfilling criteria B-D

B. Headache attacks lasting 4-72 hr (untreated or unsuccessfully treated)[2;3]

C. Headache has at least two of the following four characteristics:

- 1. unilateral location
- 2. pulsating quality
- 3. moderate or severe pain intensity

4. aggravation by or causing avoidance of routine physical activity (eg, walking or climbing stairs)
D. During headache at least one of the following:

- 1. Nausea and/or vomiting
- 2. Photophobia and phonophobia

E. Not better accounted for by another ICHD-3 diagnosis.

1.2 Migraine with aura

Previously used terms:

Classic or classical migraine; ophthalmic, hemiparaesthetic, hemiplegic or aphasic migraine; migraine accompagnée; complicated migraine.

Description:

Recurrent attacks, lasting minutes, of unilateral fully-reversible visual, sensory or other central nervous system symptoms that usually develop gradually and are usually followed by headache and associated migraine symptoms.

Diagnostic criteria:

A.

At least two attacks fulfilling criteria B and C

B. One or more of the following fully reversible aura symptoms:

- 1. Visual

"scintillating scotomas (bright rim around an area of visual loss), teichopsia (subjective visual image perceived with eyes open or closed), fortification spectra (zigzagged lines that slowly drift across the visual field), photopsias (poorly formed brief flashes or sparks of light), and blurred vision."

- 2. Sensory
- 3. Speech and/or language
- 4. Motor
- 5. Brainstem
- 6. Retinal

C. At least two of the following four characteristics:

- 1. at least one aura symptom spreads gradually over ≥5 min, and/or two or more symptoms occur in succession
- 2. each individual aura symptom lasts 5-60 min
- 3. at least one aura symptom is

	unilateral 4. the aura is accompanied, or followed within 60 min, by headache D. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack has been excluded. Notes: When for example three symptoms occur during an aura, the acceptable maximal duration is 3×60 minutes. Motor symptoms may last up to 72 hours. Aphasia is always regarded as a unilateral symptom; dysarthria may or may not be.

3) Describe the pathophysiology of migraine

- No longer thought to be vascular in origin.
    - This hypothesis has been disproven
- Now vascular changes are thought to be an “epiphenomenon” of what is a primary neurologic event triggered by abnormal trigeminal nerve activation.
    - cortical spreading depression or, less likely,
    - a sterile neuropeptide-induced inflammatory process, leads to pain and sensitization of higher order neurons in the brainstem and thalamus. Descending modulation is likely to be compromised as well. It is not yet known what initiates the pathophysiologic process that leads to a migraine attack.

4) Outline a stepwise approach to treating acute migraine attacks? List 8 therapy options for Migraine.

There are many ways to approach migraine treament. Most of the ER therapies are abortive with the goal of limiting the intensity and duration of the attack. We want to relieve pain, minimize recurrence, decrease side effects, and “street em”.

We need to consider:

- Patient's previous response to medications
- Pt. comorbidities
- Presence of N/V
- And know that gastric stasis is a common problem in an acute migraine attack (PO meds probably won’t help much)

Here’s a starting approach:

1. Acetaminophen or NSAIDs
2. First line agents for moderate-severe attacks:

2.1. DHE
2.2. Triptans
    2.2.1. Sumatriptan
2.3. Prochlorperazine
2.4. Metoclopramide
2.5. Droperidol
2.6. Ketorolac

3. Second line agents:
    3.1. Morphine
        3.1.1. Hydromorphone less effective
    3.2. Magnesium
    3.3. Valproic Acid

More details:

MILD TO MODERATE
Acetaminophen 500–1000 mg PO
Ibuprofen 600–800 mg PO Gastrointestinal upset
Naproxen sodium 275–550 mg PO Gastrointestinal upset

MODERATE TO SEVERE

First-Line Agents:

Dihydroergotamine (DHE) 1 mg IV or IM; may be repeated in 1 hour Nausea (pretreat with antiemetic), not safe in pregnancy.
Caution in inhibitors of enzyme CYP450 3A4

Triptans
Sumatriptan 6 mg SC Chest pain, throat tightness, flushing
Contraindicated with hypertension, coronary artery disease, peripheral vascular disease, and pregnancy.
Cannot be used within 24 hours of ergot use

Prochlorperazine 10 mg IV Sedation and dystonic reaction
Metoclopramide 10 mg IV Dystonic reaction
Droperidol 2.5 mg IV QT prolongation; dystonic reaction
Ketorolac 30 mg IV or 30 to 60 mg IM Gastrointestinal upset; avoid this medication in elderly and in patients with renal insufficiency

Second Line Agents:

Morphine 4 to 8 mg IM or IV Opioids less efficacious than other treatment modalities
Magnesium 2 g IV More efficacious in migraine with aura
Valproic acid 1 g IV Contraindicated in pregnancy

5) List four migraine variants.

Neuroimaging should be considered for older or immunocompromised patients with new-onset headaches, headaches associated with unexplained neurologic abnormalities, and headaches with an abrupt onset.

Don't forget to think about worst first in every patient:

- SAH
- TIA / stroke
- Brain mass
- Idiopathic intracranial hypertension!
- **Retinal migraine**
    - rare syndrome consisting of recurrent attacks of monocular visual dysfunction, including positive features (such as, scintillations) or negative features (such as, blindness). As with aura, these symptoms are completely reversible.
- **Hemiplegic migraine**
    - motor aura consisting of hemiparesis or hemiplegia. The progression of the motor deficit is gradual and in most cases is accompanied by a visual,
    - sensory, or speech disturbance. The neurologic symptoms last
    - up to 60 minutes, followed by headache. Rarely, the motor
    - deficit is persistent, resulting from a true migrainous stroke. A
    - familial version of hemiplegic migraine is associated with genetic
    - channelopathies.
- **Migraine with brainstem aura**
    - an aura referable to
    - the brainstem. Common neurologic findings include dysarthria,
    - tinnitus, vertigo, diplopia, and altered level of consciousness.
- **Status migrainosus**
    - Severe, unremitting migraine headache persisting unabated for more than 72 hrs

6) What can be done to prevent migraine headache recurrence? List 8 common precipitants.

To Prevent Headache Recurrence After Emergency Department Discharge:

Steroids Various regimens. Commonly used: **dexamethasone 10 mg IV**
Risks: Gastrointestinal bleeding, infection, cataracts, aseptic necrosis, memory disturbances

Other options:

- Naproxen 500 mg
- Sumatriptan 100 mg

Prophylaxis:

- Beta blockers
- TCAs

- Antiepileptic drugs

Precipitants:

- Sleep deprivation
- Stress
- Hunger
- Hormonal changes (menstruation)
- Drugs:
    - OCPs
    - Nitroglycerin
- Chocolate
- Caffeine
- Tyramine rich foods
- MSG
- Nitrates
- Strong glare - odour - loud noises - weather changes

7) What are the clinical features of a cluster headache? How are they treated? Tension headache?

Tension h/a = most common ha disorder in the USA.

Cluster headache	**Tension headache**
Young, middle aged men who smoke Multiple clusters of headaches in a day in a week/month period Precipitated by stress, etoh, climate change Thought to be related to trigeminal nerve activation	Common in women in the 4th decade Last 30 mins to 7 days Unknown pathophysiology, may be similar to migraine etio. Tight, band, pulsating, dull ache. Usually non-debilitating, and no associated symptoms.
Sudden onset, lasting 15 mins to 3 hrs Unilateral, sharp, stabbing pain in the eye, awakens from sleep Agitated, anxious, rocking, rubbing the head, pacing. Often ipsilateral ptosis, miosis, sweating, lacrimation, rhinorrhea. Injected eye. Nasal congestion.	Normal neuro exam. Normal vital signs. The most common disorders mimicking tension headache are migraine, IIH, oromandibular dysfunction, cervical spondylosis, sinus or eye disease, and intracranial masses. Subtle indolent infections (such as, cryptococcal meningitis) should be considered in the immunocompromised.
Treatment: • High flow O2 via NRB at 12 L/min ○ Most resolve in 15 mins • Sumatriptan 6 mg • Octreotide 100 mcg SC Prventative therapy: • Prednisone 100 mg po daily x 5, then 12 day taper • Verapamil 120 mg po tid	Treatment: • NSAIDs • Acetaminophen • Metoclopramide • Acupuncture

Don’t forget to rule out carotid artery dissection with any first, or atypical presentation!

8) List 6 causes and 6 risk factors for SAH.

SAH refers to extravasated blood in the subarachnoid space. Presence of the blood activates meningeal nociceptors, leading to diffuse occipital pain along with signs of meningismus. SAH accounts for up to 10% of all strokes and is the most common cause of sudden death from a stroke.

Approximately 80% of patients with nontraumatic SAH have ruptured saccular aneurysms. Other causes include arteriovenous malformations, cavernous angiomas, mycotic aneurysms, neoplasms, and blood dyscrasias. SAH may be caused secondarily by an intraparenchymal hematoma that dissects its way into the sub-arachnoid space.

Risk fx:

- Age > 40
- HTN
- Smoking
- Excessive ETOH
- Sympathomimetic drug use
- Familial cerebral aneurysm disorders
    - Autosomal dominant polycystic kidney disease
    - Coarctation of the aorta
    - Marfan syndrome
    - Ehler-Danlos syndrome

Don't forget about other things:

Cervical artery dissection (CAD), cerebral venous thrombosis (CVT), reversible cerebral vasoconstriction syndrome, hemorrhagic or ischemic stroke, and primary headache disorders, including migraine and cluster headaches, CNS infections.

9) Describe a clinical grading scale for cerebral aneurysms and SAH and how it relates to prognosis.

Table 93.2 – Hunt & Hess Clinical Grading Scale for Cerebral Aneurysms and SAH

Grade 0 = Unruputured aneurysm

Grade 1 = Asymptomatic or minimal headache and slight nuchal rigidity

Grade 2 = Moderate or severe headache, nuchal rigidity, no neurologic deficit other than cranial nerve palsy

Grade 3 = Drowsiness, confusion, or mild focal deficit

Grade 4 = Stupor, moderate to severe hemiparesis

Grade 5 = Deep coma, decerebrate posturing, moribund appearance

Prognosis

- **1-2 = good px**
- **3 = risk for rapid deterioration**
- **4-5 = poor px**

10) Describe a diagnostic strategy for SAH

Coles notes version:

1. Think about the dx
2. Get a non-contrast CT head
    a. Do an LP in CT negative patients if SAH is still suspected.
    b. If > 100 RBCs in tube 4 get a CT angiogram to hunt for any aneurysms.

Now for more detail….

Clinical features:

- Thunderclap h/a, kaboom, WHOML - 80% of people
- Exertional onset h/a - 20%
    - Valsalva
    - Sex
    - Non-strenuous physical activity
- *peaking intensity in seconds to minutes* - pearl
- Associated symptoms:
    - N/V
    - Syncope
    - Neck stiffness
    - Photophobia
    - Seizures
- Physical exam:
    - Meningismus - 50%
    - Focal neuro abnormalities - 20%
    - May have fundoscopic findings
    - 3rd, 6th nerve palsies
    - Mydriasis (expanding PCOM aneurysm)
    - *restlessness, altered LOC*
    - Preceding sentinel headache days to weeks before.

Imaging findings

- CT head - non contrast is the golden ticket here
    - 90% sensitivity for hemorrhage < 24 hrs old
    - 50% sensitive for hemorrhage up to a week old
    - If you get the CT within 6 hours of headache onset - CT has been shown to be 100% sensitive and 100% specific for SAH
    - But according to Rosen's:

        - "However, current guidelines still recommend follow up lumbar puncture (LP) when CT is nondiagnostic. Until additional studies are performed, we recommend a LP be performed in CT negative patients suspected of having a SAH."

- Other debates:

    - When to order the CTA? At the same time?
        - "In lieu of a non-contrast CT followed by LP, the emergency clinician can order a non–contrast-enhanced head CT scan followed by computed tomography angiography (CTA); this is reasonably sensitive and may be appropriate for patients considered to be at lower risk of disease, patients refusing a LP, and patients in whom a LP cannot be performed. Concerns of additional radiation exposure and contrast agent toxicity need to be weighed against the risks of a LP.

A normal non–contrast-enhanced head CT scan followed by a normal spinal fluid analysis definitively rules out SAH and does not need to be followed with angiography, even in patients at high risk of disease.

However, this strategy does not rule out other causes of thunderclap headache that may be in the differential diagnosis, such as carotid artery dissection, cerebral venous sinus thrombosis, and reversible cerebral vasoconstriction syndrome." From Rosen's 9th.

Lab findings

- Often have abnormal ECG findings including arrhythmias, ST-T wave changes, U waves, QT prolongation.
- LP CSF studies to look for:
    - Xanthochromia (takes up to 12 hrs to occur after hemoglobin has been metabolized; poorly supported prospectively)
    - RBC count of < 100 in tube 4 makes SAH unlikely

11) List 6 components of management for SAH

- Treat acute medical and neurologic complications (supportive care)
    - Consider early intubation of patients with HH Class 3 SAH (drowsy, confused)
    - Analgesics
    - Antiemetics
    - Bed rest in a quiet and dark environment
    - Pneumatic compression stockings
    - Reverse any anticoagulation on board
    - Treat seizures with anticonvulsants
    - Comprehensive brain resuscitation
        - Normal ICP
        - Normal MAP
        - Avoid anemia

        - Avoid hyperthermia
        - Euglycemia
        - Normoxia
        - Normal electrolytes and euvolemia

- Prevent recurrent hemorrhage
    - Aminocaproic acid (antifibrinolytics) or TXA to reduce rebleeding.
    - Blood pressure control (based on neurosx opinion)
        - But ideally: SYSTOLIC BP < 160 mmHg or MAP < 130 mmHg unless in vasospasm.

- Stalling vasospasm
    - Start Nimodipine ASAP (one of the only evidence-based treatments)
        - 60 mp Po/NG q 4hrs

12) What are features of a HA from CNS malignancy? What primary cancers spread to CNS? What are two components of management of a new CNS malignancy?

- Headaches - most common sxt of brain tumour in the young. Usually due to traction on pain-sensitive structures: the meninges and/or blood vessels
    - Less common symptom in the older adult
    - Clinical features:
        - Depend on size, location, etc.
        - *essentially the pain can be anywhere and doesn't correlate with location of the tumour*
        - Classic symptoms:
            - Worsening headache for weeks to months
            - Was initially present only on awakening and is now continuous
            - Vomiting not preceded by nausea
            - Symptoms of increased ICP:
                - Bilateral h/a worse with coughing, sneezing, bending, defecation, sexual intercourse
                - Seizures
                - Personality changes or cognitive difficulties
        - Classic" triad: rarely seen
            - **Brain tumour headache:**
                - **Sleep disturbances**
                - **Severe pain**
                - **Nausea and vomiting**
    -
- Can be primary brain malignancy, or secondary
    - Most common metastatic sources: lung > breast > melanoma > GI cancer

- Dx with CT, MRI or CT with contrast - which helps improve the identification of underlying mass lesions from abscess, hematoma, vascular malformation.

Management:

- Urgent referral to neurosx

- **For those with signs of increased ICP** (h/a, N/V, confusion, weakness)
    - Treat with corticosteroids for ICP reduction and symptom relief +/- analgesia
    - Dexamethasone 8-16 mg po daily
    - In the ER: can give 10 mg Po/iv then 4 mg q6hrs

- **Anyone with a seizure:**
    - Should receive an AED
        - Phenytoin, carbamazepine, valproate
    - Don't give AED prophylactically if no seizure has occurred.

13) Describe clinical features and treatment of carotid and vertebral artery dissection

This is the most common cause of ischemic stroke in the < 50 yr old (10-25% of cases!)

- Often people have minimal symptoms and the dx is missed!
- Usually caused by sudden neck movement or trauma or manipulation
    - Huge list! Coughing, chiro, lifting, sports, MVCs, etc.!
    - **Delay from symptom onset to diagnosis can be several days!**
- **The lesion is a hemorrhage into the media of the arterial wall**
    - This creates an intra-arterial HEMATOMA
    - A thrombus forms and clots can them embolise
- **the timing is variable and the ischemia can appear years later**
- Symptoms:
    - Sudden onset neck/head pain

Carotid artery dissection (CAD)	**Vertebral artery dissection**

• "Triad":
  ○ Unilateral h/a or neck pain
  ○ Ipsilateral horner's syndrome
  ○ Blindness OR contralateral motor deficits
• ****acute, severe retro-orbital pain in a previously healthy person with no hx of cluster h/a is suggestive of CAD**
	• Less common
	• "Young person with sudden, severe, unilateral, posterior headache."
	• Rapidly progressive neuro deficit and
	  ○ Vertigo, severe vomiting
	  ○ Ataxia
	  ○ Diplopia
	  ○ Hemiparesis
	  ○ Unilateral facial weakness
	  ○ Tinnitus

DDX for unilateral h/a or neck pain with horner's syndrome:

- Migraine
- Cluster h/a
- SAH
- CAD

Dx: CT angio or MRI

Management:

- If acute ischemic stroke symptoms ⇒ tPa candidates
- Current evidence and systematic reviews show:
    - Antiplatelets and anticoagulation show similar outcomes (3 month recurrent stroke incidence is rare)

Cerebral venous thrombosis

- Thrombosis of intracranial veins and sinuses are rare
- Think about this in the YOUNG patient without standard risk factors
    - Venous stasis
        - Neurosurgical procedures
    - Hypercoagulable state
        - Genetic
            - Antithrombin III, protein C and S deficiencies
            - Factor V leiden mutation
        - Acquired
            - pregnancy
            - Malignancy
            - Exogenous estrogen
            - Vasculitis
            - IBD
            - Connective tissue disease
    - Endothelial injury
        - Head trauma
        - Surgery
        - Parameningeal infections
- Presentation:
    - Increased ICP due to impaired venous drainage
        - Diffuse, increasing h/a over days-weeks.
        - Seizures
        - Papiledema
    - **Key symptoms:
        - **Ocular findings of pain and proptosis and paralysis of EOMs**
    - Focal brain injury due to ischemia, infarction, hemorrhage
        - Rarely present with a classic stroke syndrome

- Dx (from rosen's 9th ed)

"Routine blood work including a complete blood count (CBC), chemistry panel, ESR, and clotting studies, including a prothrombin time (PT) and partial thromboplastin time (PTT) should be obtained in all patients with suspected CVT. These studies are helpful in determining the presence of an underlying hypercoagulable state, an infectious process, or an inflammatory disorder contributing to the development of CVT. **A normal D-dimer is helpful for patients with a low probability of CVT and, along with the clinical findings,**

can be used to exclude the diagnosis in individual patients at low risk of disease. The definitive diagnosis of CVT is based on neuroimaging of the area of thrombosis. Noncontrast CT by itself is an insensitive test, but it may reveal nonspecific late lesions, such as an infarct, hemorrhage, or edema. Occasionally, hyperdensity of a cortical vein or sinus may be seen. The key to diagnosis is to image the venous system itself. This is best accomplished by a combination of MRI to visualize the thrombosed vessel and magnetic resonance venography (MRV) to detect nonvisualization of the same vessel. CTA and CTV may also be used to visualize the cerebral venous system, especially in patients who have a contraindication to MRI."

- Treatment
    - Full dose heparin or LMWH anticoag.
    - Consider thrombolysis or thrombectomy with IR

14) List 8 common clinical features for patients with giant cell arteritis; and its Dx and Rx.

GCA = inflammatory vasculopathy of med-lg arteries. Causes an inflammatory infiltrate into the wall → intimal hyperplasia, stenosis, and occlusion → ISCHEMIA

- Why we worry about vision loss - ischemic optic neuropathy

Involves:

- Aorta
- Temporal (MOST common) and occipital arteries
- Can also involve: ophthalmic, vertebral, distal subclavian, and thoracic aorta.

Risk factors:

- Age > 50
- Women > men

Clinical features (Box 93.4)

- Attributed to ISCHEMIA and SYSTEMIC INFLAMMATION
    - Headache - 70% OF PTS. usually chronic for 2-3 months.
    - Can be anywhwere on the head
    - 40% of pts develop symptoms of PMR
- Risk for:
    - TIA's of the eye, peripheral neuropathies, strokes.
- On exam:
    - Temporal artery: Findings include tenderness, reduced or absent pulsations, erythema, and nodularity or swelling.
    - Visual acuity, visual field testing, and thorough funduscopic examination should be performed. The presence of a relative afferent pupillary defect (Marcus-Gunn pupil) should increase the suspicion for GCA, although these patients usually will also have a visual loss or a visual field defect.

Common Clinical Features in Patients With Giant Cell Arteritis

- Age older than 50 years old
- Female more than male
- Headache -
- Visual symptoms:
- Amaurosis fugax
- Visual loss
- Diplopia
- Jaw claudication
- Temporal artery tenderness
- Systemic symptoms:
    - Fever
    - Weight loss
    - Fatigue

Dx:

- Think about Ddx:
    - Takayasu's arteritis affecting younger patients with visual loss
    - Stroke causing h/a and visual loss
    - Polyarteritis nodosa
- Testing:
    - Guided by clinical suspicion
    - Labs:
        - ESR > 50-100
        - CRP elevated
            - These tests have a 85% sens, and only 35% spec.
            - 4% of people with GCA can have anormal CRP
        - Thrombocytosis
        - Anemia
    - Imaging is not yet validated - MRI - U/S
    - **Gold standard: Temporal / artery biopsy** (but because this is a skip lesion disease, some patches may be missed!

Treatment: involve neuro, optho, rheumatology

1. **IF amaurosis fugax or diplopia = treat with steroids emergently**
    a. Methylprednisolone 1000 mg daily for 3 days!
2. Without visual symptoms:
    a. Prednisone 40-60 mg PO daily

15) What are the criteria to diagnose idiopathic intracranial hypertension? How is it managed?

- **Young, obese, women of childbearing age**
    - CSF prod/absorp. Imbalance. Poorly understood
    - Often have signs of increased ICP, and may have temporary visual obscurations related to postural changes; pulsatile tinnitus, n/v, dizziness.

- **Criteria for Diagnosis of Idiopathic Intracranial Hypertension**
    - Headache that remits with normalization of CSF pressure
    - Papilledema
    - Nonfocal neurologic examination
    - May have CN VI palsy
    - Increased CSF opening pressure
        - >250 mm (>25 cmH20) in adults
        - >280 mm in children
    - Normal CSF diagnostic studies
    - Normal neuroimaging studies
    - No other cause of increased ICP identified
- Dx: MRI with MR venography ideally performed before LP (lateral decubitus position)

- **Management:**
    - With visual field loss:
        - Lower ICP with acetazolamide, lasix, steroids
        - If no response may be candidates for VP / LP shunts
    - Without visual field loss: symptomatic large volume CSF drainage - 20 mL

Wisecracks

1) How do migraines differ between children and adults?

"In children and adolescents (aged under 18 years), attacks may last 2-72 hours.

Migraine headache in children and adolescents (aged under 18 years) is more often bilateral than is the case in adults; unilateral pain usually emerges in late adolescence or early adult life. Migraine headache is usually frontotemporal. **Occipital headache in children is rare and calls for diagnostic caution**. A subset of otherwise typical patients have facial location of pain, which is called "facial migraine" in the literature; there is no evidence that these patients form a separate subgroup of migraine patients. In young children, photophobia and phonophobia may be inferred from their behaviour. Migraine attacks can be associated with cranial autonomic symptoms and symptoms of cutaneous allodynia." from ICHD-3.org

2) Describe four unique presentations of migraine headache

a. **Retinal migraine**
b. **Hemiplegic migraine**
c. **Brainstem migraine**
d. **Status migrainosus!**

See above for information!

3) What is the pathophysiology of idiopathic intracranial hypertension? Describe the typical patient.

- **Young, obese, women of childbearing age**
    - CSF prod/absorp. Imbalance. Poorly understood

"PATHOGENESIS — Although many theories for IIH have been proposed, the precise pathogenesis of IIH remains unknown. Any theory must account for the high incidence of IIH in obese women of the childbearing years. Proposed etiologies include cerebral venous outflow abnormalities (eg, venous stenoses and venous hypertension); increased cerebrospinal fluid (CSF) outflow resistance at either the level of the arachnoid granulations or CSF lymphatic drainage sites; obesity-related increased abdominal and intracranial venous pressure; altered sodium and water retention mechanisms; and abnormalities of vitamin A metabolism" - From uptodate 2017

4) List 3 LP factors that reduce the incidence of post-LP HA; and describe the management of post-LP HA.

- **NOT** affected by the amount of time in a recumbent position post LP
- Things that due decrease risk of PDPH:
    - Small diameter needles 20-22 ga cutting needles
    - Inserted with the needle bevel up
    - Atraumatic needle (**whitaker or sprotte needle**)

Management:

- Most resolve spontaneously in 5-7 days post
    - Supportive care:
        - Bed rest,
        - Hydration
        - Mild analgesia
    - Unproven therapy:
        - Caffeine
    - Epidural blood patch is the definitive treatment for non-responders to conservative treatment (15-30 ml of autologous blood).

5) Describe post-traumatic headache & hypertensive headache.

PTH:

- Common post concussion or TBI
    - Often the person has both anatomic and functional symptoms consistent with post-concussive syndromes
    - Normal neuro exam, imaging normal

HH:

- Meet criteria when BP > 180/120 mmHg and h/a → then resolves when the blood pressure is normalized

- **Be sure to inquire about pregnancy or post-partum status (up to 8 weeks post!)**
- **No objective end-organ damage identified on labs or exam.**
- First treat with antidopaminergic and NSAIDs, reserve blood pressure medications for people with evidence of end organ damage.

6) What is reversible cranial vasoconstriction syndrome?

"A cerebral arteriopathy characterized by segmental areas of vasoconstriction within large- and medium-sized vessels. It is the same disease as postpartum angiopathy or migrainous vasospasm. RCVS causes recurrent thunderclap headache in susceptible patients and may cause ischemic or hemorrhagic stroke." - Rosen's 9th Ed.

- Diagnosed more frequently now given non-invasive neurovasc. Imaging
- Some data suggests this may be the common cause of thunderclap h/a
- Symptoms:
    - Thunderclap h/a, severe, throbbing, n/v
    - Photophobia
- Dx: by excluding the other emergent causes of H/A (Ct +/- LP)
    - "RCVS = Recurrent thunderclap h/a's in a discrete time frame"
    - Different diagnosis than "primary headache associated with sexual activity"
- Treatment:
    - No known optimal treatment
    - Some people use CCB's
    - Discharge with neuro f/u

7) List 10 HA Red Flags

1. Onset > age 50
2. Onset in immunocompromised patients
3. Hx of malignancy or "b" symptoms.
4. First ever headache / symptom
5. Febrile, with no explanation
6. Thunderclap headache
7. Post-traumatic with no imaging
8. Onset with exertion
9. Focal deficits
10. Signs of increased ICP

Remember:

First
Febrile
Forceful
Forty
Focal

Or:

“The mnemonic SNOOP is a reminder of the danger signs ("red flags") for the presence of serious underlying disorders that can cause acute or subacute headache [11,12]:

- Systemic symptoms, illness, or condition (eg, fever, weight loss, cancer, pregnancy, immunocompromised state including HIV)
- Neurologic symptoms or abnormal signs (eg, confusion, impaired alertness or consciousness, papilledema, focal neurologic symptoms or signs, meningismus, or seizures)
- Onset is new (particularly for age >40 years) or sudden (eg, "thunderclap")
- Other associated conditions or features (eg, head trauma, illicit drug use, or toxic exposure; headache awakens from sleep, is worse with Valsalva maneuvers, or is precipitated by cough, exertion, or sexual activity)
- Previous headache history with headache progression or change in attack frequency, severity, or clinical features” - From UptoDate 2017