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Beyond Tension, Migraine, and Cluster Headaches: The “Other” Primary Headaches

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Objectives

[illegible]

Musical notation example showing various accidentals and time signatures.

7 $\frac{3}{4}$ | F $\frac{3}{4}$ | $\frac{3}{4}$ | D n $\frac{3}{4}$ | B $\frac{3}{4}$ | I = n $\frac{3}{4}$ | N — N o d n — F e c l u i n m i k
 $\frac{3}{4}$ | $\frac{3}{4}$ | f n r j t x f — $\frac{3}{4}$ | f i $\frac{3}{4}$ | p $\frac{3}{4}$ | E O — L e N k $\frac{3}{4}$ | n $\frac{3}{4}$ | k

Primary vs. Secondary Headaches

Primary Headache

- Has no underlying cause
- The headache itself is the main medical issue

Secondary Headache

- Caused by other conditions, often serious and life-threatening
 - space-occupying lesions
 - CNS infections (meningitis or encephalitis)
 - subarachnoid hemorrhage
 - giant-cell arteritis
 - cerebral venous thrombosis
 - Increased intracranial pressure

Headache **Red** Flags

- Systemic symptoms including fever
- Onset of headache is sudden or abrupt (“thunderclap”)
- History of cancer
- Neuro deficit or decreased LOC
- Onset after age 50
- Pattern change of headache
- Positional headache
- Papilledema
- Brought on by sneezing, cough or exercise
- Posttraumatic headache
- Pregnancy
- HIV or immunosuppression

Headache **Green** Flags

- Current type of headache has been present since childhood
- Patient has headache-free days
- Headache is related to the menstrual cycle
- Close family members have similar type headaches
- Headache occurred or stopped over a week ago

Types of Primary Headaches

- *International Classification of Headache Disorders, Third Edition (ICHD-3)* includes 4 Categories of Primary Headaches
 1. Migraine
 2. Tension-Type Headache
 3. Trigeminal Autonomic Cephalgia (cluster headaches are most common type)
 4. Other Primary Headaches

The “Other” Primary Headaches

- Primary Cough Headache
- Primary Exercise Headache
- Primary Headache Associated with Sexual Activity
- Primary Thunderclap Headache
- Cold Stimulus Headache
- External Pressure Headache
- Primary Stabbing Headache
- Nummular headache
- Hypnic Headache
- New Daily Persistent Headache

Key Points about the “Other” Primary Headaches

- Pathophysiology is often not well understood
- Epidemiology is difficult to determine
- Treatment recommendations are often based on case reports and expert opinion with a lack of randomized clinical trials
- Many of these headaches will need to be managed by or co-managed with a neurology/headache specialist
- Secondary causes must always be considered
- Some of these headache types overlap

Cough Headache



Cough Headache

Relatively rare

Pathophysiology

- coughing causes a sudden increase in intra-abdominal and intrathoracic pressures that are transmitted via the venous system into the venous sinuses, thus activating intradural or perivascular nociceptive neurons

Cough Headache

Clinical Presentation

- Sudden onset
- Precipitated by cough or Valsalva
- Bilateral pain
- Can last up to 2 hours
- Pain described as “sharp” and moderate/severe
- Not usually associated with nausea/vomiting, photo/phonophobia

Cough Headache

******Remember that headaches precipitated by cough are a red flag and a serious secondary cause must be ruled out first before it can be diagnosed as Primary Cough Headache

Treatment

- Indomethacin 50-100mg/day (consider giving with a PPI for GI protection)
- Propranolol 40mg-120mg/day
- Vagus Nerve Stimulation
- Lumbar Punctures

Exercise Headache



Exercise Headache

Relatively rare

Pathophysiology

- One theory is that incompetent venous vasculature causes retrograde flow leading to an increase in intracranial pressure that leads to headache pain

Exercise Headache

Clinical Presentation

- Precipitated by strenuous exercise
- Bilateral
- Lasts 5 minutes to 48 hours
- Pulsatile or throbbing quality

******Remember that headaches precipitated by exercise are a red flag and a serious secondary cause must be ruled out first before it can be diagnosed as Primary Exercise Headache

• Treatment

- Indomethacin 50-150mg taken before strenuous exercise

Sexual Headache



Sexual Headache

Prevalence

- Lifetime prevalence estimated to be 1-1.6%
- More common in men
- First episode usually occurs in 30's or 40's

Pathophysiology

- sudden hemodynamic changes are responsible for the headache pain
- Patients with a history of sexual headaches found to have more of an increase in systemic increase in BP with exercise

Sexual Headache

Clinical Presentation

- Either dull, progressive, or abrupt exploding headache (or combination of the two)
- Usually bilateral and occipital
- May last several minutes to over 24 hours
- “Explosive” and/or “throbbing” quality
- May have nausea/vomiting, photo/phonophobia
- Patients may have co-morbid HTN, migraines, or tension HAs

Sexual Headache

******Remember that headaches that have a sudden or abrupt onset and are described as “explosive”, like a “thunderclap”, or are the “worst headache of their life” are a red flag and a serious secondary cause must be ruled out first before it can be diagnosed as Primary Sexual Headache

Treatment

- Usually self-limited but can become chronic for more than a year
- Prevention can include maintaining passive sexual role
- Indomethacin 25-50mg given 30-60 minutes before sexual activity
- Beta blockers, topiramate, triptans

Nummular Headache



Nummular Headache

Very rare

Pathophysiology

- epicranial in origin and involves the terminal branches of the sensory nerves. These nerves are responsible for connecting the structures of the scalp to the dura mater

Nummular Headache

Clinical Presentation

- Pain is located in rounded or elliptical areas of the scalp measuring 1-6cm in diameter
- Usually unilateral and more common on right than left side
- The area does not change in size or shape over time
- Pressure-like, sharp, or stabbing type pain
- Mild/Moderate pain but can be severe
- Can last minutes, hours or days
- Can have numbness, allodynia, hyperesthesia in area
- Pain can be chronic or can have remissions that last weeks or months

Nummular Headache

Treatment

- NSAIDs
- Gabapentin, pregabalin
- TCAs
- Peripheral nerve stimulation
- Transcutaneous nerve stimulation
- Botox

Cold-Stimulus Headache



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Cold-Stimulus Headache

Prevalence

- Lifetime prevalence estimated to be 15%
- More common in patients with migraines

• Pathophysiology

- one theory is that a cold stimulus may trigger a trigeminal response followed by a reflex vasoconstriction. This rapid constriction and dilation of vessels activated nociceptors in the vessel wall and causes referred pain in frontal or temporal areas of the head

Cold-Stimulus Headache

Clinical Presentation

- Precipitated by ingestion or inhalation of a cold stimulus
- Usually bilateral frontal or temporal
- Short-lived (30 seconds) but can last >5 minutes
- “Stabbing” type HA
- Patients with migraine may describe the pain as “throbbing” and on the same side as their migraines

Treatment

- Avoiding cold or icy foods
- Eating cold or icy foods slowly
- Curling tongue and placing underside against the roof of the mouth (not an old wives tale!)

External-Pressure Headache



External-Pressure Headache

Prevalence

- Increased during COVID-19 and use of masks and PPE

• Pathophysiology

- The pathophysiology of these headaches is thought to be caused by the compression of nerve endings of the branches of the trigeminal and occipital nerves

External-Pressure Headache

Clinical Presentation

- Brought on compression or traction to the scalp (i.e.. from wearing tight headband, goggles, mask, tight ponytail, etc.)
- Headache located at area of traction on scalp
- Usually resolves within an hour after the noxious stimuli removed
- Pain is non-pulsatile, constant, and moderate in intensity
- Not associated with nausea/vomiting or photo/phonophobia

Treatment

- Removal (or avoidance of noxious stimuli)
- Medications usually not needed

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