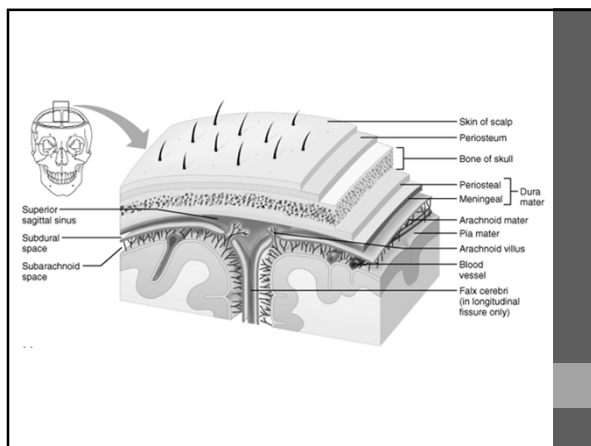


Headaches in PHACE Syndrome

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Head pain

- The brain does not have sensory receptors
- Blood vessels and tissues surrounding the brain *feel* pain



Headache types

- International Classification of Headache Disorders (ICHD)
 - Primary headaches
 - Secondary headaches

Tension-type headache

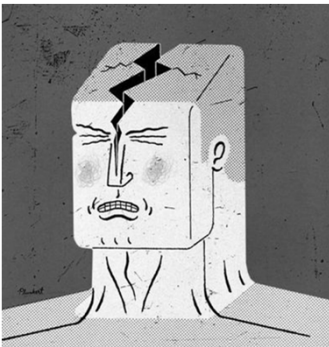


Tension-type headache

- Commonest headache type
 - Ranging from 30-78%
- Can be related to physical and psychological stress
- Tension headaches are 'episodic' or 'chronic'

- (A) At least 10 episodes
- (B) Headache lasts 30 minutes to 7 days
- (C) At least 2 of the following:
 - Bilateral location (both sides of head)
 - Pressing/tightening pain quality
 - Mild or moderate pain intensity
 - Pain not made worse by activity
- (D) No nausea or vomiting and
No more than one light/noise sensitivity
- (E) Not explained by another disorder

Migraine headache

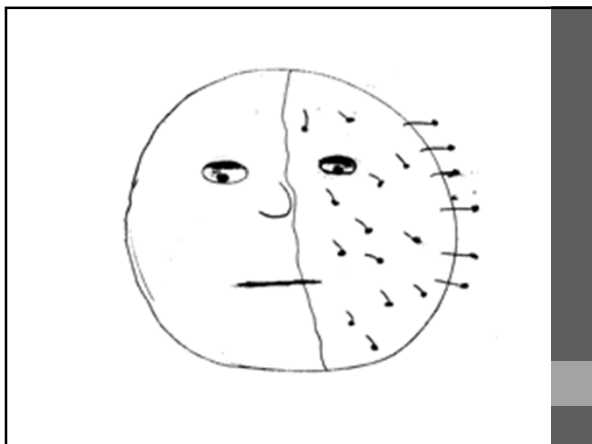


Migraine headache

- Up to 10.6% of children 5-15 years of age
- Affects about 17% of adult women
- And 6-8% of adult men

- (A) At least 5 attacks
- (B) Headache lasting 4-72 hours
- (C) At least 2 of the following:
 - Unilateral location (one side of head)
 - Pulsating pain quality
 - Moderate to severe pain
 - Worsening of pain with activity
- (D) Nausea and/or vomiting or light/noise sensitivity
- (E) Not explained by another disorder





Migraines in young children

- ICHD migraine diagnostic criteria do not account for young children
- Migraines often diagnosed retrospectively as the child grows older
- Several 'periodic syndromes' suggest increased migraine risk

Migraines in young children

- Early migraine features:
 - Vomiting
 - Irritability
 - Pale appearance
 - Inactive
 - Covers eyes and ears
 - Sleeps

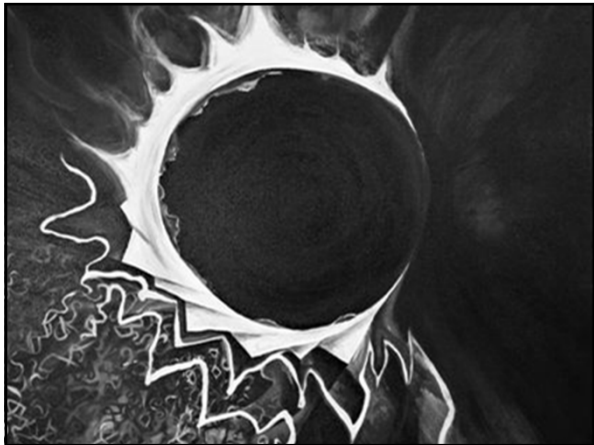
Periodic syndromes in children

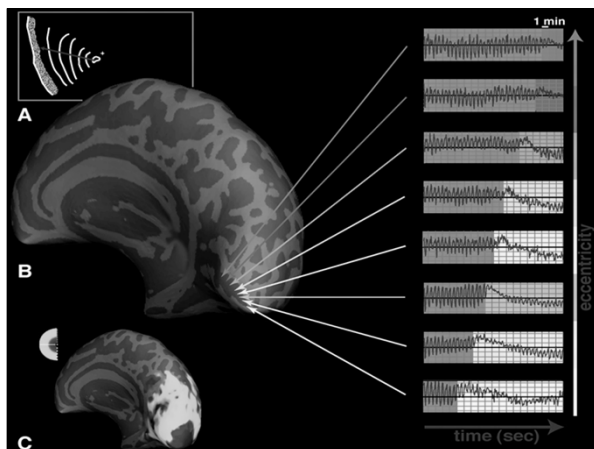
- Possibly 'precursors' to migraine
- Periodic syndromes include:
 - Cyclical vomiting
 - Abdominal migraine
 - Benign paroxysmal vertigo

Migraine aura

• Aura defined as:

- Fully reversible
- Visible symptoms
- Sensory symptoms
- Change in speech
- Develops gradually over 5+ minutes
- Lasts between 5-60 minutes









Headache *Red Flags*

- Change in headache character
- Neurologic changes
- New-onset headaches
- Underlying brain (or blood vessel) problems
- Positional headaches

Headaches in PHACE Syndrome

• Headaches common in PHACE Syndrome

- 49 (67%) of 73 survey participants had headaches
- Average age of onset about 4 years
- 51% evaluated by physician
- 84% use non-prescription medicines
- 18% use prescription medicines
- 10% use daily prescription medicines

Physician diagnoses (n=25)	
Migraine headaches	13 (52%)
Non-migraine headaches	5 (20%)
Headaches caused by hydrocephalus	1 (4%)
Headaches caused by moyamoya vasculopathy	1 (4%)
Not known	5 (20%)

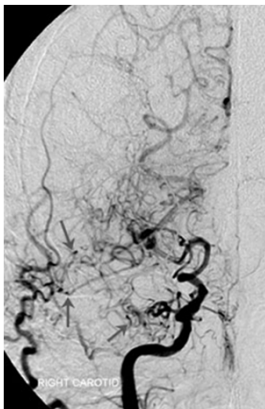
Quality	
Throbbing	15 (30.6%)
Aching/tight	10 (20.4%)
Stabbing	3 (6.1%)
Other/not known	20 (41.7%)
Intensity, scale 1-10	
Mean (median)	5.4 (5)
Associated symptoms	
Nausea	29 (59.2%)
Vomiting	18 (36.7%)
Photophobia (aversion to bright light)	34 (69.4)
Phonophobia (aversion to loud noise)	34 (69.4)
Osmophobia (aversion to strong smells)	4 (8.2%)
Vision disturbances	12 (24.5%)

Why headaches in PHACE Syndrome?

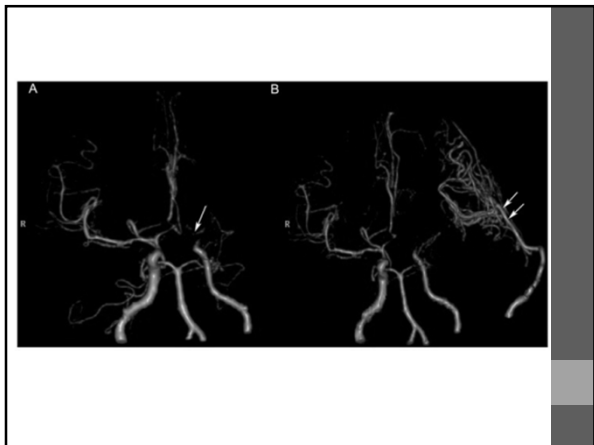
- Abnormal blood vessels (vasculopathy)
- Posterior fossa (back of the brain) cysts
- Hydrocephalus
- Stroke

Vasculopathy and headaches

- Changes in blood vessels can cause headaches in individuals without PHACE
 - Moyamoya syndrome
 - Aneurysms
 - Arteriovenous malformations (AVMs)
 - Venous clots







Hydrocephalus and headaches

- Dandy-Walker malformations are common in PHACE syndrome
- They can cause increased pressure in the head – causing headaches

Hydrocephalus and headaches



Even with PHACE syndrome most headaches are benign

- Headaches are common
- Some headaches in PHACE may represent blood vessel or other changes
- Many headaches will be primary (and coincidental)

Headache treatments

- Depends on severity and frequency
- Treatment can be given when headaches occur (abortive)
- Or treatments can be given to decrease headache frequency (prophylactic)

Abortive headache treatments

- Ibuprofen, acetaminophen, naproxen
- 'Triptans': Imitrex, Maxalt, Relpax, etc.
- Nausea medicines: prochlorperazine (Compazine), promethazine (Phenergan)
- Do not overuse pain medicines

Prophylactic headache treatments

- Amitriptyline (Elavil)
- Topiramate (Topamax)
- Propranolol (Inderal)
- Cyproheptadine (Periactin)

Lifestyle changes as headache treatments

- Stay hydrated
- Maximize sleep
- Avoid headache triggers
- Do not miss meals
- Daily exercise/activity
- Stress control

Always make neurologist or PCP aware
of headaches

- Often easily treated
- Can represent brain problems
- Some individuals with headaches need additional testing
- PHACE patients risk of blood-vessel problems is high

Questions?
