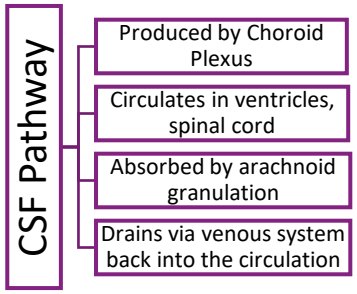
Idiopathic Intracranial Hypertension (IIH)

(aka: Pseudotumor Cerebri)

What is intracranial hypertension?

Intracranial hypertension refers to an increased pressure in the CSF that surrounds the brain and spinal cord. This pressure on the brain and optic nerves can lead to symptoms such as headache, tinnitus, and blurry vision.



Classic Headache Description:

- Constant, frontal headache
- Worsens with positions that increase ICP – Valsalva (stooling, coughing), lying down, bending over
- Transient visual obscuration ("graying out" of vision associated with postural changes)
- Pulsatile tinnitus ("a whooshing in my ears")

Differential Diagnosis:

- Before we call this IIH, we need to ask: is this truly ***idiopathic***?
- **Secondary causes** of IH include:
  - Medications:
    - Oral contraceptives
    - Tetracyclines
    - Isotretinoin
    - Sodium Valproate
    - Lithium
    - Withdrawal form prolonged corticosteroid use
  - Hypervitaminosis A
    - Source may vary – vitamins, foods: liver
    - Several above medications are vitamin A derivatives, including retinols, tetracycline antibiotics.
    - Mechanism of causing IIH currently unknown, but postulated that vitamin A affects arachnoid granulation cells, leading to decreased CSF absorption.
  - Venous drainage issue
    - Central venous thrombosis, outflow tract stenosis
  - Other, less common etiologies: Addison's disease, Growth hormone, Chronic kidney disease, Down Syndrome
- Idiopathic IH (aka IIH) is associated with **obesity** (mechanism unknown).

Differential-Driven Physical Exam:

- Fundoscopic exam - **papilledema** is a classic physical exam finding!
  - Why papilledema? The optic nerves exit the brain parenchyma and are briefly exposed to circulating CSF before they enter the orbit – this means that extra pressure from the CSF will push them forward, causing a "cupped" or blurred appearance on fundoscopic exam.
- Visual fields – are they restricted?
- Cranial nerves – particularly cranial nerve 6
  - Abducens Nerve Palsy = can't look ***laterally***
  - Occurs only when IH is severe
  - Why CN 6? → increased ICP causes brainstem to be pushed downward, stretching CN 6 and causing dysfunction
- Full neurologic exam – any focal deficits might suggest other causes of headache, such as intracranial mass, stroke, or bleed.

Workup:

- Review medications (including topical creams and supplements/vitamins)!
- Review growth chart!
- Do a fundoscopic exam (and have Ophthalmology do one as well).
- Lumbar Puncture **with Opening Pressure**:
  - Expect normal cell count/protein/glucose but **high opening pressure.**
- Imaging:
  - MRI Brain (typically without contrast, unless inflammatory process or tumor is suspected, in which case order with & without)
    - Empty Sella
    - Flattening of the posterior aspect of the globes
    - Tortuous optic nerve
  - MRV Brain (venogram)
    - Rule out cavernous sinus thrombosis
    - Assess for transverse venous sinus stenosis
    - Assess venous outflow tracts

Treatment:

Treatment is super important due to the risk of **permanent vision loss.**

- First line:
  - **Weight loss!**
  - Remove causative agents (birth control, retinols, etc)
  - **Acetazolamide** (brand name: Diamox) is first line for IH.
    - How does acetazolamide work?
    - Carbonic anhydrase inhibitor = *decreases* CSF production at the choroid plexus
- Second line:
  - Topiramate?
  - Furosemide?
- Third line: VP shunt

Sources:

Kliegman, Robert. **Nelson Textbook of Pediatrics**. Edition 21. Philadelphia, PA: Elsevier, 2020.

Paley, G. L., Sheldon, C. A., Burrows, E. K., Chilutti, M. R., Liu, G. T., & McCormack, S. E. (2015). Overweight and obesity in pediatric secondary pseudotumor cerebri syndrome. *American journal of ophthalmology*, *159*(2), 344–52.

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