Idiopathic Intracranial Hypertension
Not a simple headache

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Snoop out Secondary Headache

- **Mnemonic:**
- “**SNOOP 4 secondary causes**”
  - **S**ystemic conditions or symptoms:
    - HIV/AIDS, cancer, pregnancy, bleeding or clotting problems, fevers/chills, weight loss, malaise, cough etc.
  - **N**eurological signs or symptoms
    - Horner’s syndrome, dilated pupil, motor asymmetry
  - **O**lder patient
    - Primary headache disorders rarely start after age 40
  - **O**nset
    - Sudden vs gradual
    - Traumatic?
  - **P**ostural
    - Worse with standing: low pressure
    - Worse with lying down: high pressure
  - **P**rogressive
  - **P**recipitation by Valsalva
  - **P**apilledema
Papilledema changes everything!

Secondary intracranial hypertension:
Intracranial mass (tumor, abscess), venous sinus thrombosis, hydrocephalus, meningitis, increased CSF production (choroid plexus papilloma), malignant systemic hypertension, etc...
Cerebral Venous Thrombosis

- Easily missed diagnosis, often insidious
- Risk factors: Genetic, hematologic diseases, malignancy, head trauma, autoimmune disorders, intracranial & ear infections, dehydration, OCPs, post partum
- Clinical presentation
  - Intracranial hypertension syndrome
  - Sudden hemorrhage “worst headache of life”
  - Focal neurologic syndrome
  - Encephalopathy
  - Cavernous sinus syndrome, lower cranial nerve palsies
- Treatment: anticoagulation +/- endovascular treatment

Ferro, 2016
Idiopathic Intracranial Hypertension (IIH, aka Pseudotumor Cerebri)

- Symptoms and signs from increased ICP (intracranial pressure)
  - E.g., headache, vision loss
  - Must have papilledema or 6th nerve palsy
  - + elevated ICP
  - + normal CSF
  - And no other cause of intracranial hypertension evident on imaging
- Definitions:
  - Pseudotumor cerebri = idiopathic intracranial hypertension
  - Pseudotumor cerebri syndrome: intracranial hypertension from identifiable cause (e.g., drug toxicity)
  - Old term “benign intracranial hypertension” should not be used
    - Risk of permanent vision loss
    - Significant morbidity
  - “IIH without papilledema”
    - Unclear, controversial, probably rarely exists (anatomic variation)
    - Most patients have poorly taken or artificially inflated LP opening pressure measurement
    - Little or no risk of vision loss
IIH: Demographics

- Incidence: 1-2/100,000
  - Obese women age 15-44: 4-21/100,000
  - Rare in males, young children, elderly

- Risk factors
  - Obesity
    - Esp. lower body-predominant “gynecoid” or “pear” pattern
  - Recent weight gain
    - Average: 20 lb over preceding year, or 1.8 kg (~ 4lbs) over 2 months
  - Medications: growth hormone, tetracycline antibiotics (doxycycline, minocycline), vitamin A derivatives, corticosteroid withdrawal
    - Not oral contraceptives
  - Systemic illness: Addison’s, obstructive sleep apnea, severe anemia, systemic lupus erythematosus, Behçet’s, PCOS, renal failure
IIH Pathophysiology: theories

- Altered sodium & water retention mechanisms
- Decreased CSF absorption
  - ? Hormonally mediated on epithelial membranes
  - *Adipose is an endocrine organ!*
    - Adipose aromatase converts androgens to estrogen
    - More aromatase in buttock region fat
    - Also vitamin A metabolism, mineralocorticoids
  - Obesity-related increased abdominal and intracranial venous pressure
- Cerebral venous outflow abnormalities
  - Sometimes likely primary
  - In other patients, stenosis of transverse venous sinuses may develop secondarily, but then compound the problem: “feed-forward cycle”
- Diffuse cerebral edema
IIH: Presentation

Typical: *obese woman of childbearing age complains of headaches, found to have papilledema*

- **Symptoms**
  - **Headache** (84-92%)
    - Daily/intermittent, variable features
  - **Transient visual obscurations** (68-72%) *lasting seconds (<10 sec), rarely longer*
  - **Pulsatile tinnitus** (52-60%): *hearing heartbeat, water or wind inside head*
  - **Photopsia** (48-54%): *brief flashes or sparkles*
  - **Neck/back pain** (53%)
  - **Pain behind eyes** (44%), pain with eye movements or globe compression
  - **Double vision** (18-38%): *binocular, horizontal*
  - **Sustained visual loss** (26-32%)
  - Rare: other cranial nerve palsies, CSF leak (rhinorrhea, otorrhea)
IIH: Ophthalmic Evaluation

- Visual acuity
- Visual fields
- **Optic nerve assessment**
  - Papilledema
  - Pallor
  - Other features: hemorrhages, cotton wool spot, glistening pseudodrusen, optociliary collateral vessels, gliosis, peripapillary wrinkles & retino-choroidal folds
- OCT
IIH: Optical Coherence Tomography (OCT)

Kupersmith, 2011

Sibony, 2015
IIH: vision loss

- Visual prognosis mostly excellent
- Vision loss is most serious complication
  - Early or late
  - High blood pressure may be risk factor
- Visual acuity: < 20/20 in 10-29% at presentation
  - Poor measure in most patients
  - Frank vision loss at onset may predict poor visual outcome
- Visual field
  - Peripheral field loss can be insidious and asymptomatic for a long time
    - Similar to glaucoma
  - *Sequential visual field testing is essential!*

Keltner, 2014
Fulminant IIH

- **Acute onset** of symptoms and signs of increased ICP
- **< 4 weeks** from initial symptoms to severe visual loss
- **Rapid worsening of visual loss** over few days
- Typically young (14-39 yo.), obese women
- Risk factors: high blood pressure, anemia
- Mean CSF opening pressure 54.1 cm H2O (range 29-60)
- Surgical treatment required in most cases
MRI

- MRI brain w/ & w/o contrast
  - Masses: tumor, cyst, abscess, etc.
  - Stigmata of increased intracranial pressure
    - Empty sella
    - Distended/tortuous optic nerve sheaths
    - Flattened posterior sclera
    - Protrusion of optic nerve heads (may enhance with IV contrast)
    - Others: low-lying cerebellar tonsils, dilated Meckel’s Cave & cavernous sinus, meningocele, widening of foramen ovale
- MRV
  - Rule out cortical venous sinus thrombosis
  - Distal transverse venous sinus stenosis

https://radiopaedia.org/articles/idiopathic-intracranial-hypertension-1
Lumbar Puncture

- Avoid with intracranial mass, significant Chiari/cerebellar tonsillar herniation
- Patient relaxed, on side, legs extended
  - Other positions (prone, sitting) can give falsely high readings!
  - Avoid Valsalva (crying, breath holding)
  - Anxiety, pain can raise pressure, also anesthesia/sedating medicines (raise CO2)
- Upper limit:
  - Traditional 20 cm H2O
  - IIH Criteria: 25 cm H2O
  - Children: 28 cm H2O
- Complications: rarely serious
  - Back pain
  - Headache: *IIH patients can have post-LP headache!*
IIH: Treatment

• Goals
  • 1) Preserve vision
  • 2) Alleviate symptoms (usually headache)

• Some patients with normal vision and minimal symptoms need no treatment!

• Stop offending drugs (e.g., tetracyclines, retinoids)
• Consider assessing for sleep apnea
• Weight loss: as little as 6% can be helpful!
  • Diet: low calorie, low carb, high protein
  • Consider bariatric surgery
• Medicines
• Surgery

Kupersmith, 2011  www.mghcme.org
Effect of Acetazolamide on Visual Function in Patients With Idiopathic Intracranial Hypertension and Mild Visual Loss

The NORDIC Idiopathic Intracranial Hypertension Study Group Writing Committee

- In patients with IIH and mild visual loss, the use of acetazolamide with a low-sodium weight reduction diet, compared with diet alone, resulted in modest improvement in visual field function.
- Greater weight loss, reduction in waist circumference, lower CSF pressure.
- Participants also had significant improvement in quality-of-life measures.
IIH: Medicines

• Acetazolamide
  • *Only drug supported by a large randomized trial!*  
  • Inhibits carbonic anhydrase  
  • Sulfa drug, but rare cross reactions with sulfa antibiotic allergy  
    • Careful with severe reactions  
  • Side effects: finger/mouth paresthesiae, *appetite + weight loss*, taste disturbances, fatigue, nausea, electrolyte changes, kidney stones  
  • Pregnancy category C (birth defects in animals but probably not humans)

• Topiramate
  • Seizure drug, weak carbonic anhydrase inhibitor  
  • Side effects: *weight loss*, cognitive symptoms, kidney stones, acute-angle glaucoma  
  • Teratogen: cleft lip/palate

• Loop diuretics: furosemide
  • Sulfa drug, but rare cross reactions with sulfa antibiotic allergy  
  • Others: indomethacin, short course of IV steroids
IIH Surgery

• Indications
  • Worsening vision
  • Intractable headache
  • Anticipated hypotension (blood pressure treatment, renal dialysis) - *may precipitate ischemia in swollen disc*
  • Patient factors: e.g., unable to follow up, impaired cognition, unreliable exam
• Optic nerve sheath fenestration
  • Primarily to stabilize visual function
  • Headache relief often only temporary
  • Complications: temporary diplopia, pupil dysfunction, vision loss (up to 11%, usually transient)
• VP Shunting
  • Rapid headache relief
  • Complications: shunt failure requiring revision (may have rapid vision loss!), hemorrhage, infection
• Venous sinus stenting
  • 6% complication rate (incl. SDH requiring surgical decompression)
THANK YOU!

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