

Idiopathic Intracranial Hypertension

Not a simple headache

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Disclosures

“Neither I nor my spouse/partner has a relevant financial relationship with a commercial interest to disclose.”

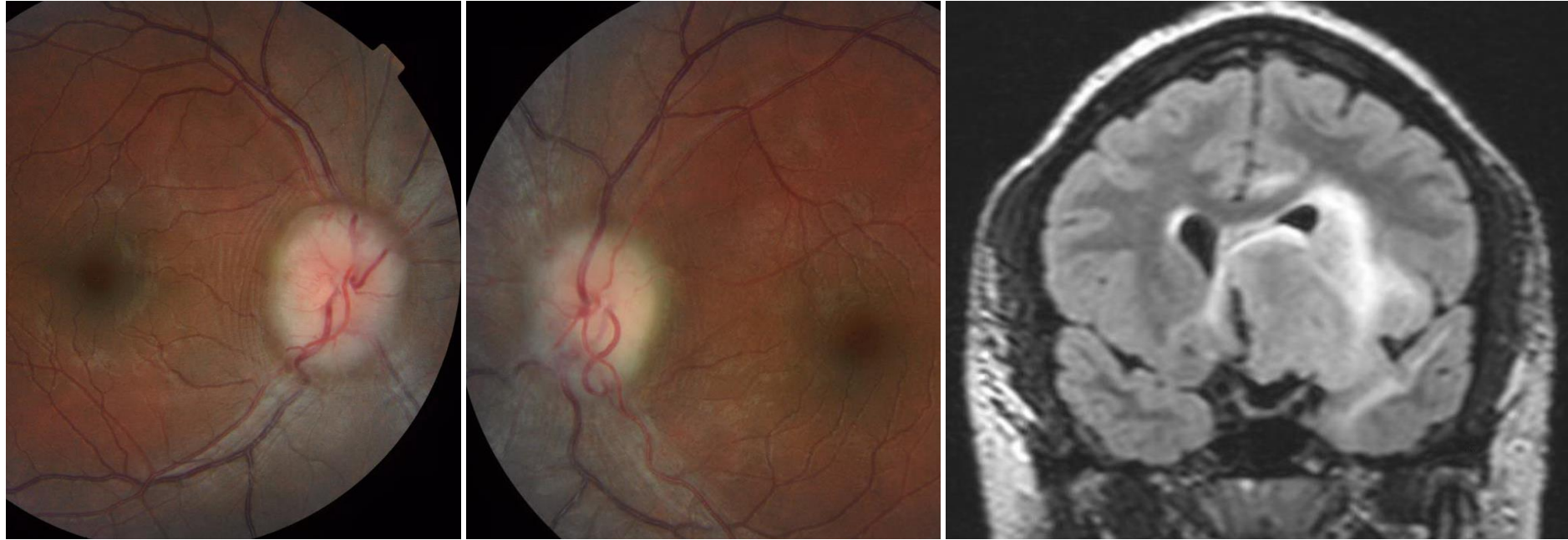


Snoop out Secondary Headache

- **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



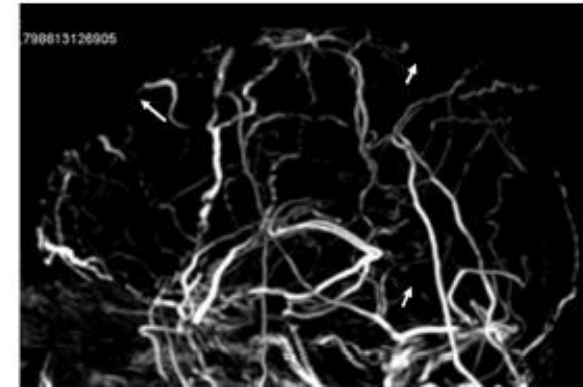
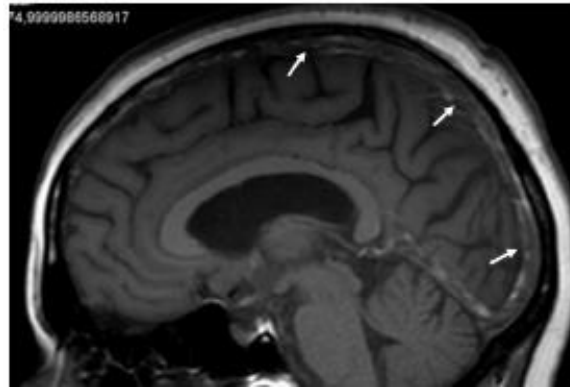
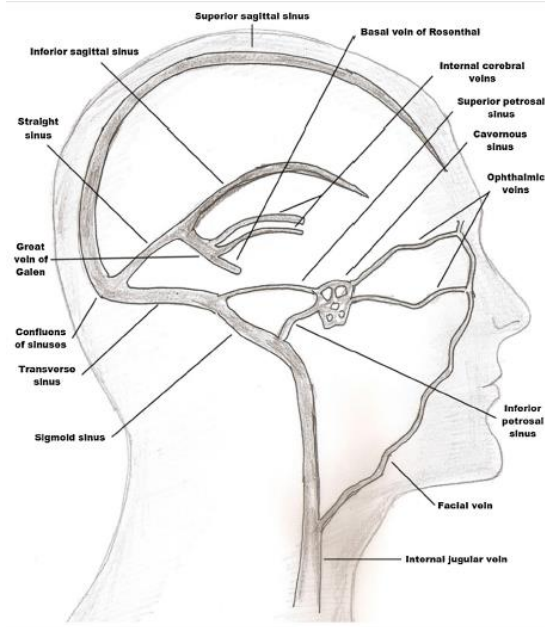
Papilledema changes everything!



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



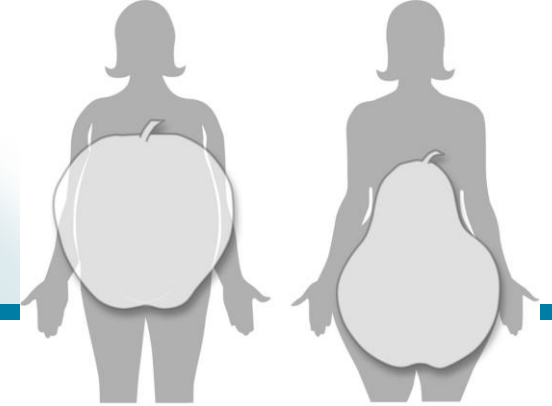
Ferro, 2016

- 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

Idiopathic Intracranial Hypertension (IIH, aka Pseudotumor Cerebri)

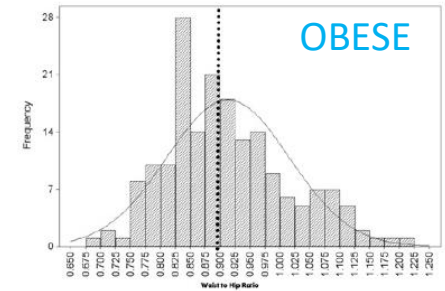
- Symptoms and signs from increased ICP (intracranial pressure)
 - E.g., headache, vision loss
 - Must have papilledema or 6th nerve palsy
- + elevated ICP
- + normal CSF
- And no other cause of intracranial hypertension evident on imaging
- Definitions:
 - Pseudotumor cerebri = idiopathic intracranial 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

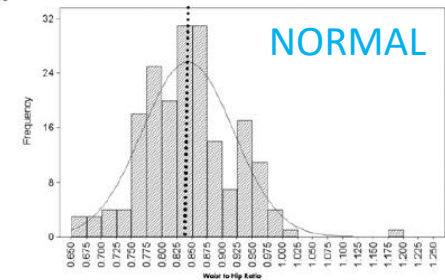


- Incidence: 1-2/100,000
 - Obese women age 15-44: 4-21/100,000
 - Rare in males, young children, elderly
- Risk factors
 - **Obesity**
 - Esp. lower body-predominant “gynecoid” or “pear” pattern
 - **Recent weight gain**
 - Average: 20 lb over preceding year, or 1.8 kg (~ 4lbs) over 2 months
 - **Medications:** 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

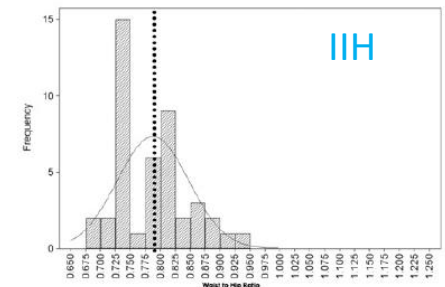
Obesity Clinic WHR distribution (Median =0.89)



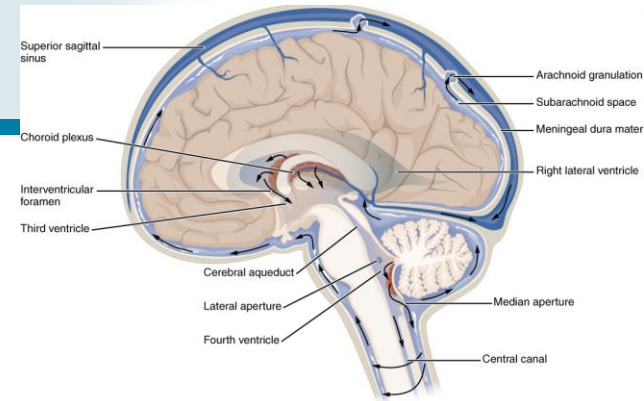
National Survey WHR distribution (Median =0.842)



WHR distribution in IIH patients (Median WHR= 0.784)



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

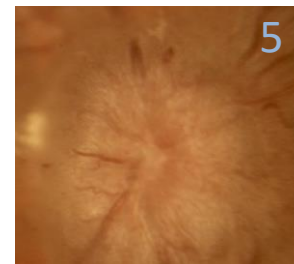
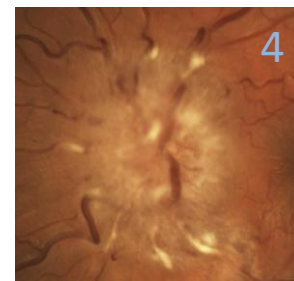
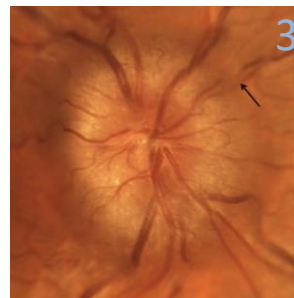
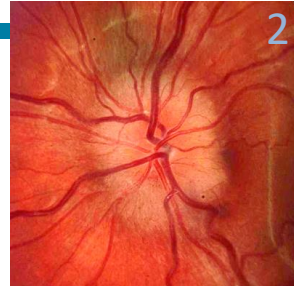
IIH: Presentation

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

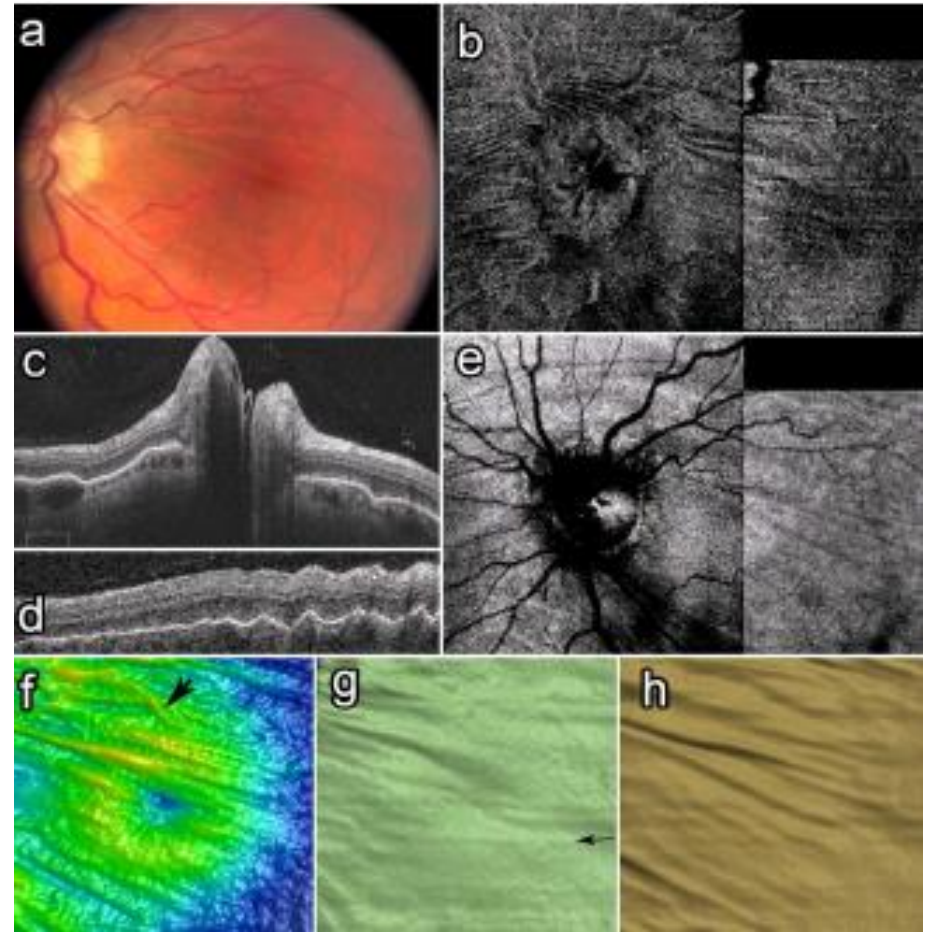
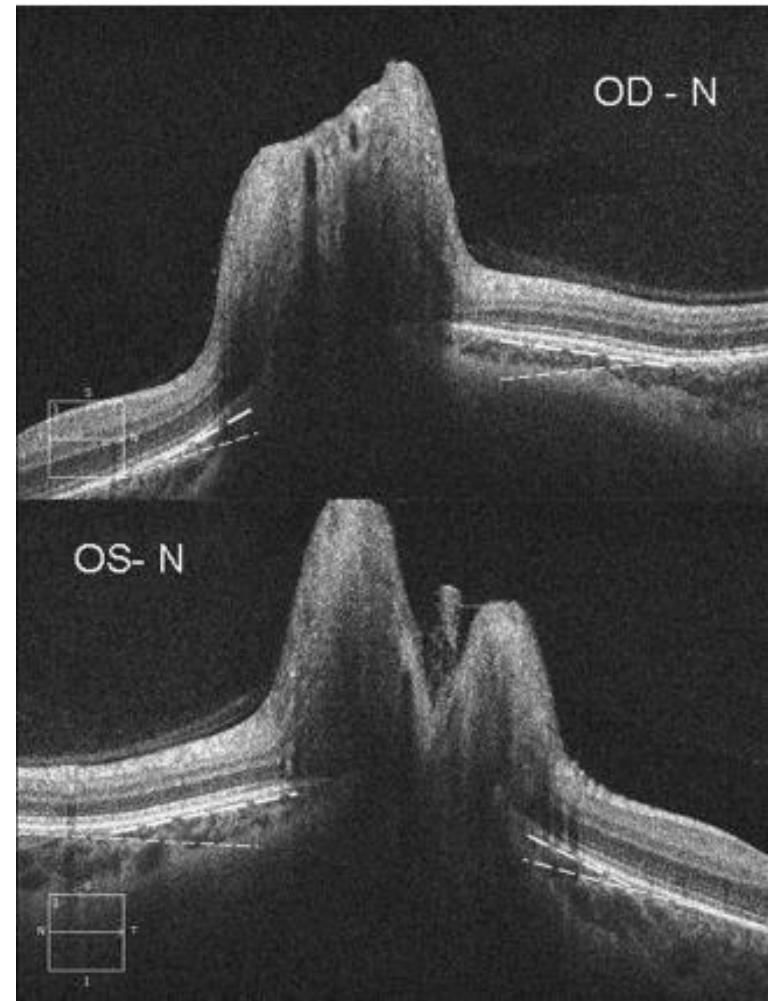
- Symptoms
 - Headache (84-92%)
 - Daily/intermittent, variable features
 - Transient visual obscurations (68-72%) *lasting seconds (<10 sec), rarely longer*
 - Pulsatile tinnitus (52-60%): *hearing heartbeat, water or wind inside head*
 - Photopsia (48-54%): *brief flashes or sparkles*
 - Neck/back 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

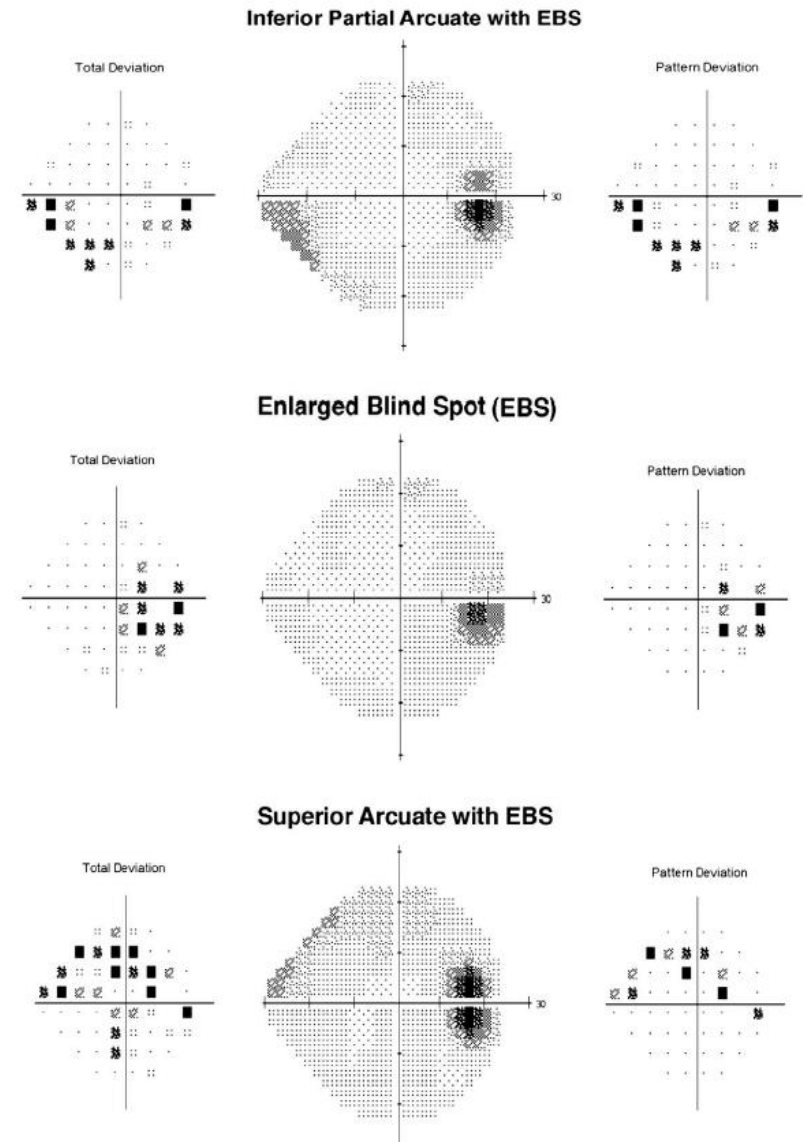


IIH: Optical Coherence Tomography (OCT)



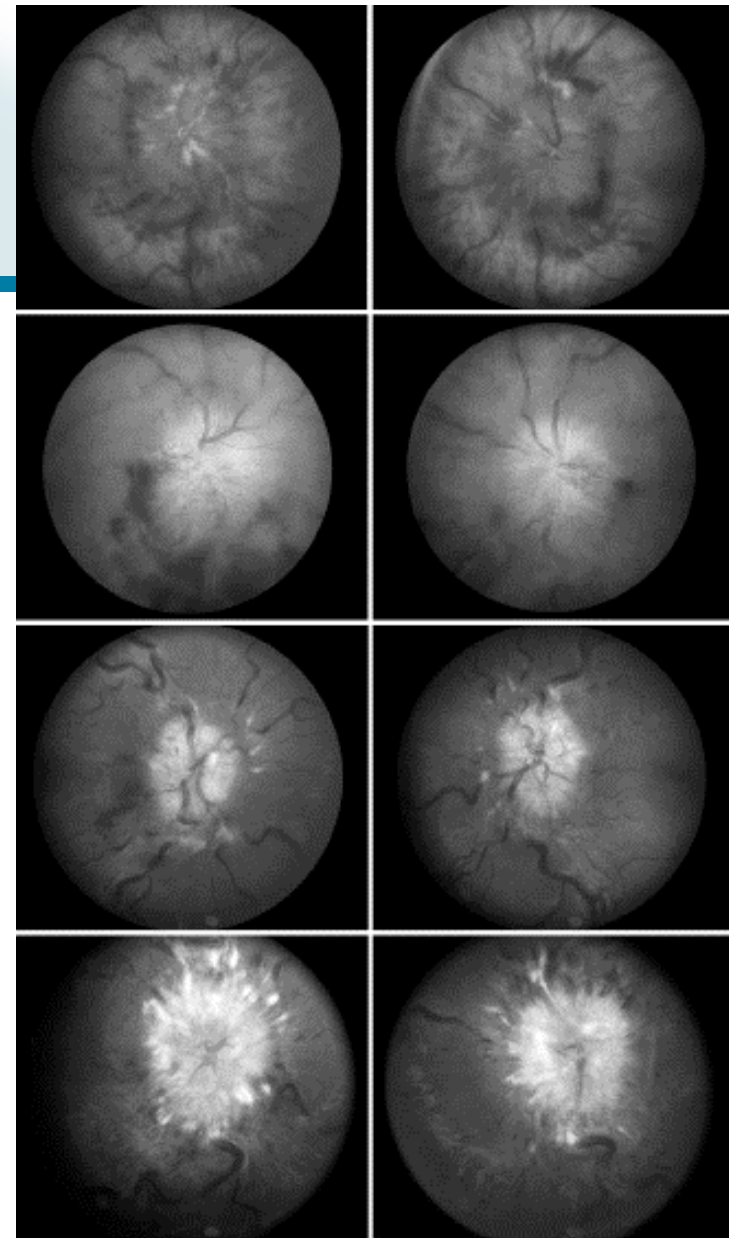
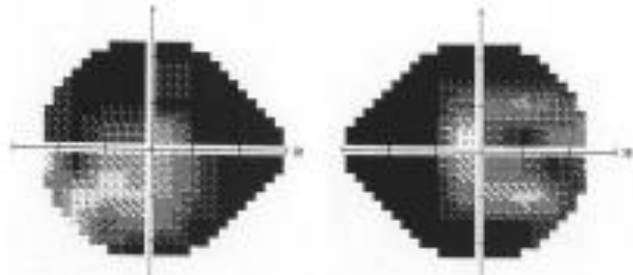
IIH: vision loss

- 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!*



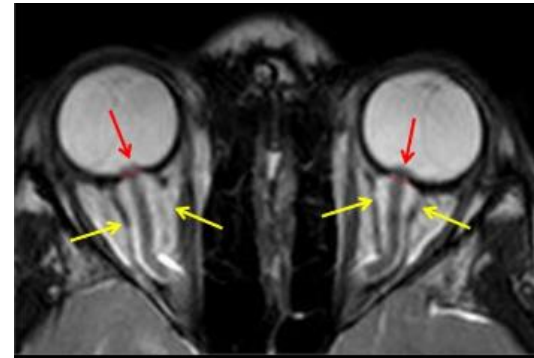
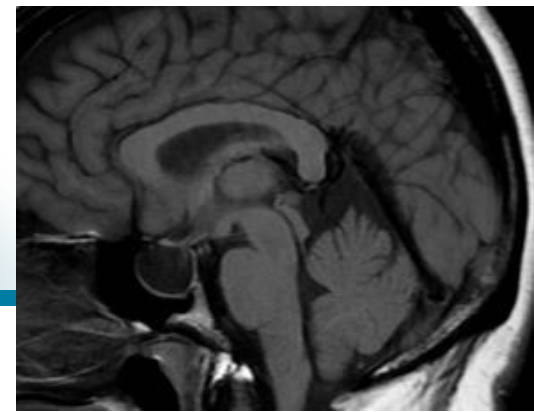
Fulminant IIH

- **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



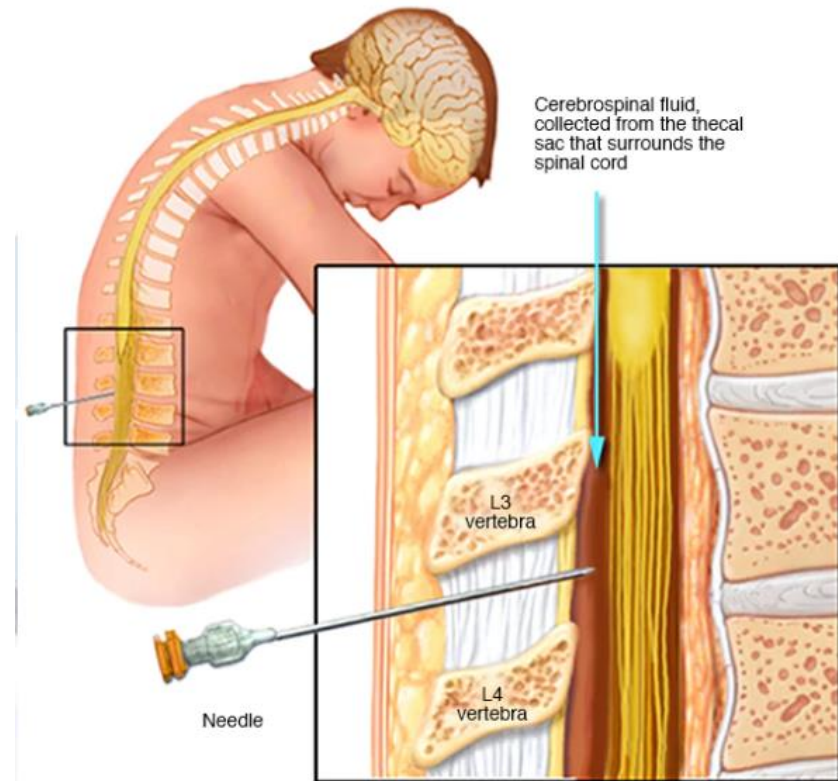
MRI

- 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



Lumbar Puncture

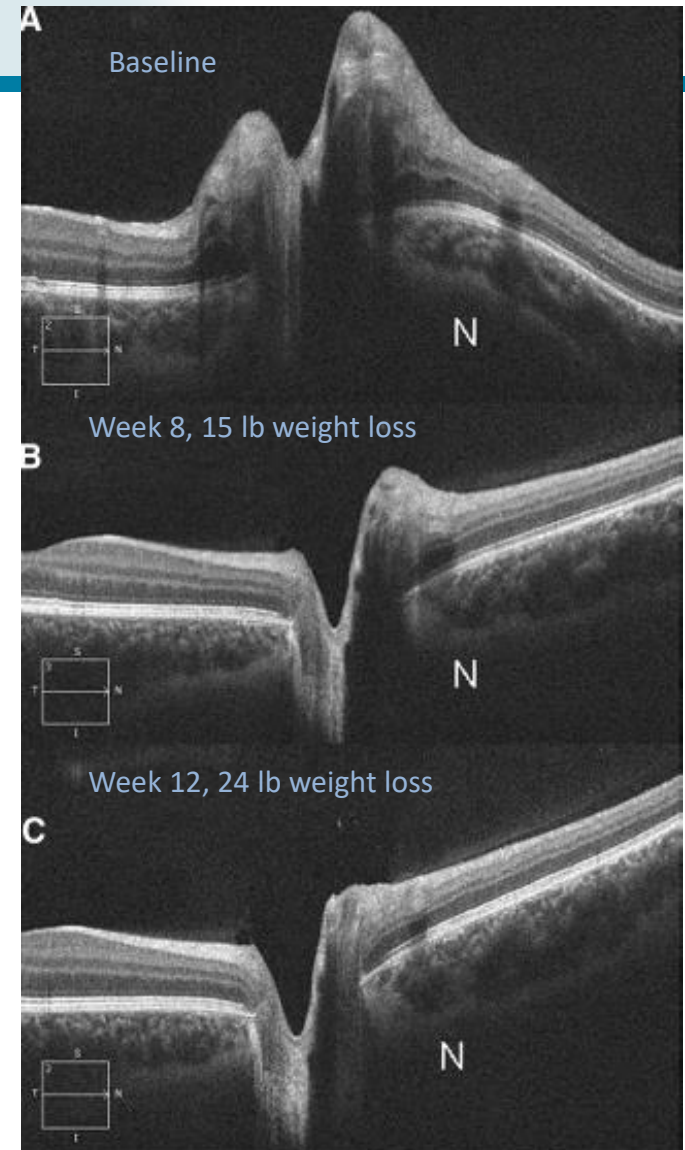
- Avoid with intracranial mass, significant Chiari/cerebellar tonsillar herniation
- Patient relaxed, on side, legs extended
 - Other positions (prone, sitting) can give falsely high readings!
 - Avoid Valsalva (crying, breath holding)
 - Anxiety, pain can raise pressure, also anesthesia/sedating medicines (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!**



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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

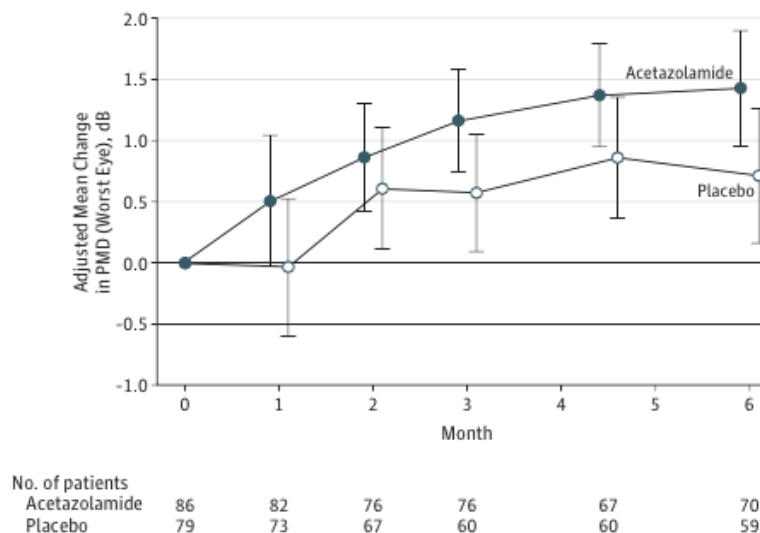


Effect of Acetazolamide on Visual Function in Patients With Idiopathic Intracranial Hypertension and Mild Visual Loss

The Idiopathic Intracranial Hypertension Treatment Trial

The NORDIC Idiopathic Intracranial Hypertension Study Group Writing Committee

Figure 2. Adjusted Mean Change in Perimetric Mean Deviation (PMD) Over Time by Treatment Group

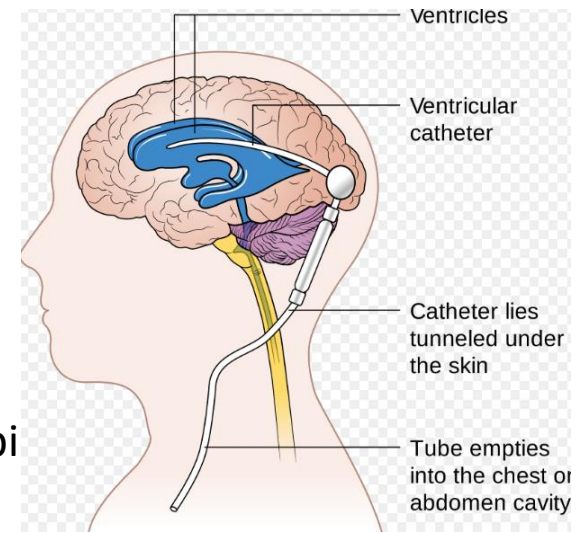
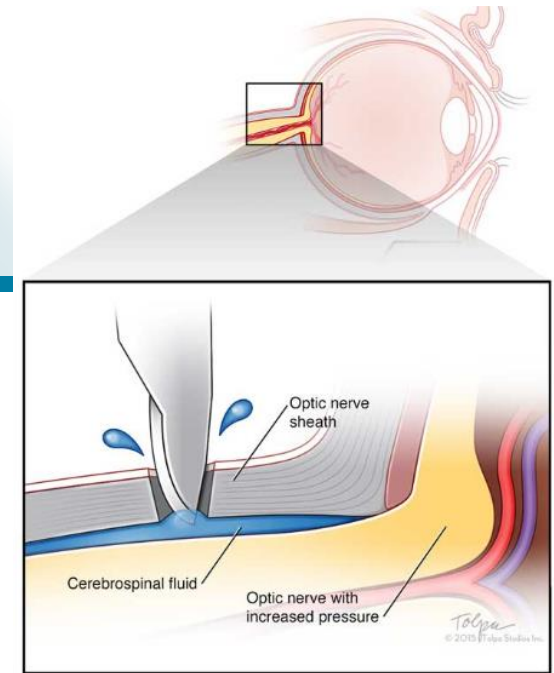


- In patients with IIH and mild visual loss, the use of acetazolamide with a low-sodium weight reduction diet, compared with diet alone, resulted in modest improvement in visual field function
- Greater weight loss, reduction in waist circumference, lower CSF pressure
- Participants also had significant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



Everyday E(B)M

- 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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