

Idiopathic Intracranial Hypertension Update

Chris Borgman, OD, FAAO
Southern College of Optometry
Memphis, TN



COPE Disclosures:


--I do not have any relevant financial relationships to disclose.

--The content and format of this course is presented without commercial bias and does not claim superiority of any commercial product or service.

Disclosures...

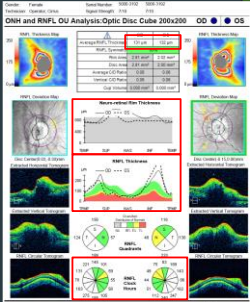
- ▶ I have no disclosures to report.
- ▶ I'm not perfect...
- ▶ I can email you my reference list...
 - ▶ 117 articles for this lecture!
- ▶ Questions??
- ▶ Email me: cborgman@sco.edu

Case 1 --- 35 YO AAF (20/20 sc OD, OS)



Baseline OCT

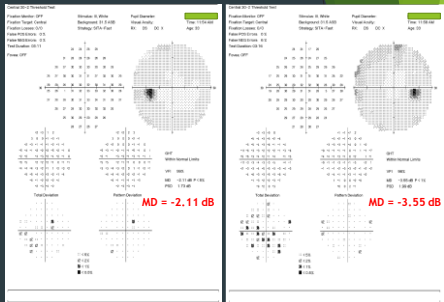
Avg RNFL:
131 μm OD
132 μm OS

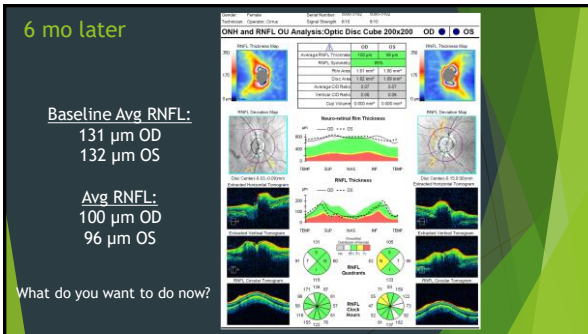
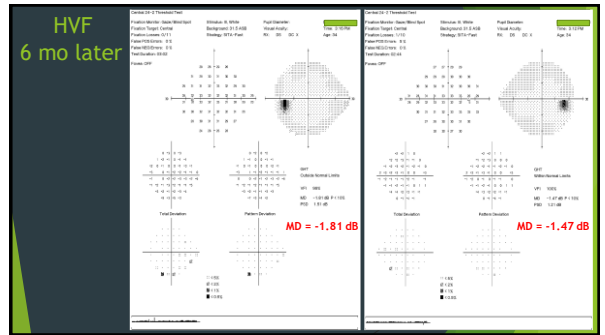
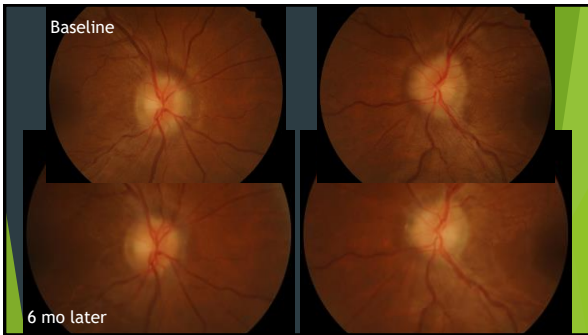


Baseline HVF

--workup → IIH!

--Started on Diamox 500mg BID PO by neurology

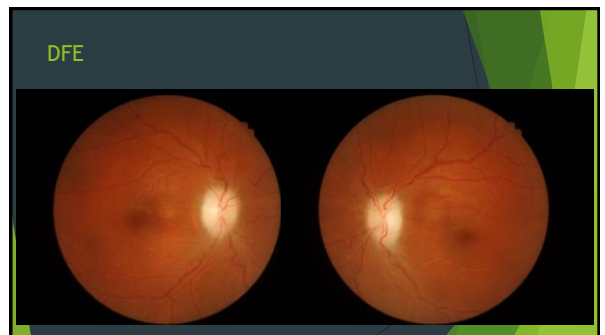


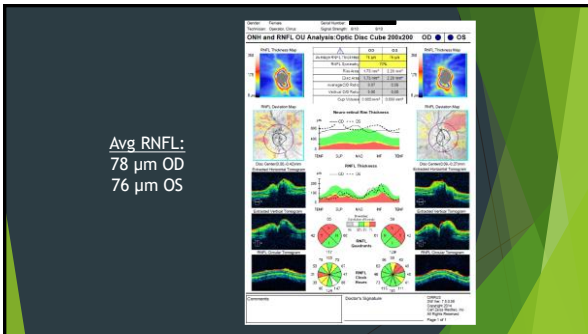
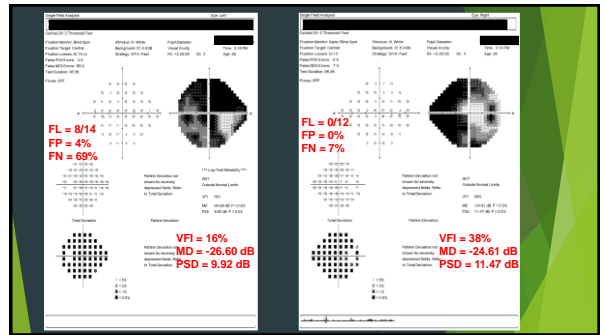
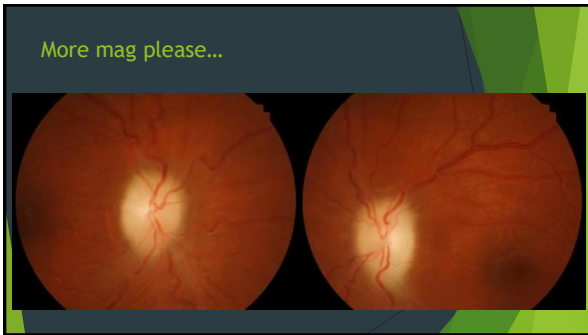


Case 2:

“Doc these headaches are killing me!... will new glasses help?”

- Case History...**
- ▶ 26 YO AAF----- “fits the profile”
 - ▶ CC: “progressive loss of side vision and now central vision”
 - ▶ OS->OD, (+)HA's behind eyes, onset 2-3 mo, referred by PCP after no improvement with oral antibiotics for “sinus infection”
 - ▶ BCVA = 20/25- OD, 20/1000 OS with Snellen
 - ▶ Color Vision = 1/7 OD, UTT OS with HRR Plates
 - ▶ CT = ortho at D & N
 - ▶ EOMs = Full OU
 - ▶ IOP = 16 mmHg OD, 18 mmHg OS with Goldmann
 - ▶ BP= 114/73 in-office
 - ▶ PMH = (+)HTN—controlled, (-)DM, (-)pregnant
 - ▶ Meds = Atenolol
 - ▶ Allergies = NKDA

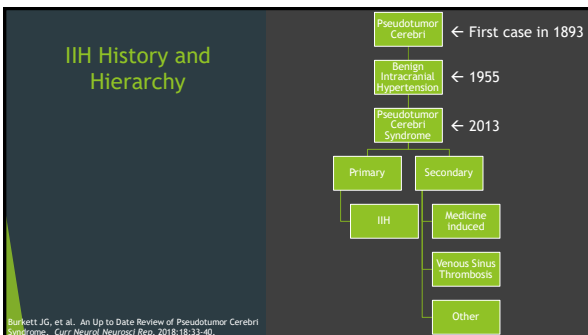




MRI results for patient

- ▶ MRI = WNL
- ▶ MRA = WNL
- ▶ MRV = bilateral stenosis of transverse sinuses
- ▶ LP opening pressure = 295 mm/H2O
- ▶ LP Cytology = normal

▶ Management? What would you do now?



Idiopathic Intracranial Hypertension (IIH)

- ▶ **AKA** → Pseudotumor Cerebri (PTC) or Benign Intracranial Hypertension (BIH)
- ▶ **Defn:** increased ICP without a mass effect and with normal CSF composition
- ▶ **MOA:** intracranial venous drainage obstruction ; decreased CSF drainage
- ▶ **F>M** (90% vs. 10%) ; females of child-bearing age
- ▶ Risk factors = obesity (70% of IIH), delayed CSF absorption, venous outflow abnormalities/increased cerebral venous sinus pressure
 - ▶ Recent estimates of IIH & obesity → 90-95% patients are obese!
- ▶ ***Headaches = 90-94% of cases**
 - ▶ Most common Sx
- ▶ Blurred vision, **loss of VF (up to 96%)**, visual obscurations, permanent visual loss (25%), legal blindness possible (10%)
- ▶ ***Papilledema = most common Sn ; 89-95% of cases**

Available theories of IIH pathophysiology...

1. CSF hypersecretion
 - ▶ But why no hydrocephalus/ventricular hypertrophy noted?
2. **Reduced CSF resorption**
3. Obesity
 - ▶ Increased intra-abdominal pressure causes ↓ decreased venous return from brain
4. Hormonal
 - ▶ Women of child-bearing age are most common group afflicted with IIH/PTC
 - ▶ No hormonal profile has been found though...
5. Endothelial dysfunction & inflammation
 - ▶ Obesity increases both of these
6. Aquaporin (CNS water channel proteins) deficiencies
 1. Acetazolamide lowers expression of aquaporin molecules...

Unproven!

Bottom Line: We don't know yet!

IIH prevalence expected to increase!

- ▶ US population is becoming more obese
- ▶ IIH cases expected to increase
- ▶ 57% IIH patients go on disability
- ▶ 31% IIH patients change jobs 2' IIH
- ▶ \$444 million/year spent on IIH management in US!

Chan JW. Current concepts and strategies in the diagnosis and management of idiopathic intracranial hypertension. J Neurol. 2017;264:1622-33.

Modified Dandy's Criteria (Revised 2013)

1. Absence of mass lesion or hydrocephalus with CT or MRI
2. Elevated CSF opening pressure upon lumbar puncture with normal CSF profile
 - ▶ ~~Non-obese patient >200 mmH2O = Abnormal~~
 - ▶ ~~Obese patient >250 mmH2O = Abnormal~~
3. Intact neurological exam with the exceptions of visual disturbances, and/or 6th nerve palsy, and/or papilledema

Adult patient >250 mmH2O = Abnormal
Pediatric patient >280 mmH2O = Abnormal **New!**

Friedman DI, Liu G, Digre KB. Diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. Neurology 2013; 81:1159-1165.

New Updated Guidelines for IIH Dx

Table 1. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children [2]

1. Required for diagnosis of pseudotumor cerebri syndrome*

(A) Papilloedema

(B) Normal neurologic examination except for cranial nerve abnormalities

(C) Neuroimaging: normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, (or typical patient (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others, if MRI is unavailable or contraindicated, contrast-enhanced CT may be used)

(d) Normal CSF composition

(E) Elevated lumbar puncture opening pressure (250 mm CSF in adults and 280 mm CSF in children; 250 mm CSF if the dM is not isolated and not obese in a properly performed lumbar puncture)

2. Diagnosis of pseudotumor cerebri syndrome without papilloedema

In the absence of papilloedema, a diagnosis of pseudotumor cerebri syndrome can be made if B-E from above are satisfied, and in addition, the patient has a unilateral or bilateral abducens nerve palsy

In the absence of papilloedema or sixth nerve palsy, a diagnosis of pseudotumor cerebri syndrome can be suggested but not made if B-E from above are satisfied, as in addition at least three of the following neuroimaging criteria are satisfied:

- (i) Empty sella;
- (ii) flattening of the posterior aspect of the globe;
- (iii) distention of the peritrigonal subarachnoid space with or without a tortuous optic nerve;
- (iv) transverse venous sinus stenosis.

* A diagnosis of pseudotumor cerebri syndrome is definite if the patient fulfills criteria A-E. The diagnosis is considered probable if criteria A, D are met, but the measured CSF pressure is lower than specified for a definite diagnosis.

Friedman DI, Liu G, Digre KB. Diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. Neurology 2013; 81:1159-1165.
 Burkett Jr, et al. An Up To Date Review of Pseudotumor Cerebri Syndrome. Curr Neurol Neurosci Rep. 2018;18:33-40.
 Chan JW. Current concepts and strategies in the diagnosis and management of idiopathic intracranial hypertension. J Neurol. 2017;264:1622-33.

PTC/IIH Symptoms

1. ***Headache** (worse upon awakening) (84-94%)
2. Transient Vision Loss (62-68%)
 - Unilateral and/or bilateral possible
3. Pulsatile Tinnitus (48-52%)
4. Blurred Vision
 - Severe vision loss (25%)
 - Blindness (10%)
5. Vomiting
6. Diplopia (18%)

PTC/IIH signs...

- ▶ **Papilledema!** In up 95% of cases! (Puffer et al. 2014)
- ▶ "With rare exception, all PTC/IIH patients have papilledema, a hallmark of subacute intracranial hypertension." ---Galgano et al. (2013)
 - ▶ But...
- ▶ Although papilledema is present in the vast majority of PTC/IIH patients, its absence is not an exclusionary criteria." ---Galgano et al. (2013)

Ocular work-up in IIH...

- ▶ Visual Acuity
- ▶ **Visual Fields**
- ▶ **EOM's**
- ▶ **Fundus Exam**
- ▶ Retinal Imaging (FP, OCT, etc.)
- ▶ Color Vision
- ▶ Contrast Sensitivity

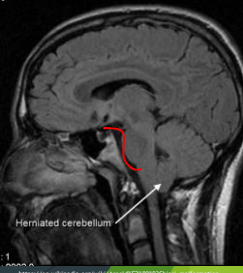
} Most important to assess in IIH

Visual Acuity and Color Vision in IIH

- ▶ **Visual Acuity:**
- ▶ Acuity tests foveal function
- ▶ Not typically affected unless edema extends into central 10° of fixation
- ▶ **Color Vision:**
- ▶ Only been found to be abnormal in ~20% of cases
- ▶ Ishihara defects only noted in the existence of moderate to marked visual loss and optic atrophy
- ▶ Not the most reliable way to follow patients

EOM's/VF's in IIH...

- ▶ **EOM's:**
- ▶ If present, uni/bilateral 6th nerve palsies are present 2" stretching nerve between apex of clivus bone/Dorello's canal and exit zone of 6th nerve on brainstem
- ▶ Dilution required in all 6th nerve palsies to rule out/in papilledema per Will's Eye Manual
- ▶ **Visual Fields:**
- ▶ **"Most important test to follow for changes"**
- ▶ Enlarged blindspot first to show, followed by generalized constriction, and nasal defects.
- ▶ Any kind of defect is possible though...



https://en.wikipedia.org/wiki/Arnold%E2%80%93Chiari_malformation

VF's in neuro-optometry.... Is testing the central 30° enough?

- ▶ "Humphrey SAP has replaced Goldmann perimetry in clinical practice despite fears that peripheral visual field defects may be missed. This fear seems unwarranted as **only 1-2% of patients with nonglaucomatous VF defects have abnormalities in the peripheral field beyond 30° degrees in the absence of central field defect.**"
- ▶ Alternatively said.... **98-99% of neurological VF defects will show up in the central 30° when tested....** pretty good odds!

Kedar S, Ghate D, Corbett JJ. Visual fields in neuro-ophthalmology. Indian J Ophthalmol. 2011;59:103-109

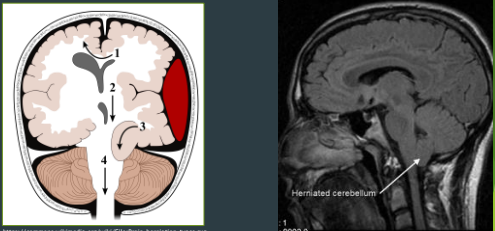
Neuroimaging/workup in IIH...Order is important...why???

1. Order MRI/MRV first
2. Followed by lumbar puncture if MRI/MRV is normal
 - ▶ >250 mmH2O in adult patients = abnormal
 - ▶ >280 mmH2O in pediatric patients = abnormal

- ▶ Herniation through foramen magnum can compress upper medulla which is where the respiratory and cardiovascular centers are located → Death

Van Crevel H, Hijdra A, de Gans J. Lumbar puncture and the risk of herniation: when should we first perform CT? J Neurol. 2002;249:129-137

What does herniation look like?



https://commons.wikimedia.org/wiki/File:Brain_herniation_types.png

https://en.wikipedia.org/wiki/Arnold%E2%80%93Chiari_malformation

What are we looking for in work up?

- ▶ **MRI**
 - ▶ Rules out space occupying mass, hemorrhage, etc.
 - ▶ Empty sella, pituitary deformities, distention of ON, posterior globe flattening
- ▶ **MRV**
 - ▶ Rules out transverse sinus stenosis and/or venous sinus thrombosis
- ▶ **Lumbar Puncture (LP)**
 - ▶ Document elevated opening/intracranial pressure
- ▶ **LP cytology and culture**
 - ▶ Rule out infectious meningitis, blood, and other possible issues/causes



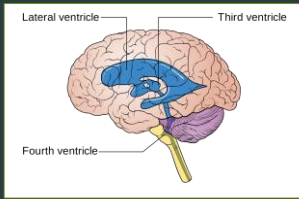
Question I've received from previous CE attendees:

- ▶ **Is LP required in a patient with normal MRI/MRV and papilloedema?**
 - ▶ "Lumbar puncture to measure opening pressure and spinal fluid examination are critical for the diagnosis." - Peralta, et al. *Curr Opin Ophthalmol.* 2018
 - ▶ "Following brain imaging, lumbar puncture is mandatory to record the CSF opening pressure and exclude a secondary cause." - Chatziralli, et al. *Graefes Arch Ophthalmol.* 2018
 - ▶ "Despite the value of neuroimaging in the diagnostic workup of IIH, it does not replace the need for measurement of lumbar opening pressure as imaging abnormalities show a large interindividual variation and none of the findings are pathognomonic of IIH.....LP is mandatory in the diagnostic algorithm of IIH." - Hoffmann, et al. *J Headache Pain.* 2018
 - ▶ "Following normal imaging, all patients with papilloedema should have a lumbar puncture to check opening pressure and ensure contents are normal." - Mollan, et al. *J Neurol Neurosurg Psych.* 2018

• Peralta GM, Cestari DM. An update of idiopathic intracranial hypertension. *Curr Opin Ophthalmol.* 2018;29:495-502.
 • Chatziralli I, et al. Perspectives on diagnosis and management of adult IIH. *Graefes Arch Clin Exp Ophthalmol.* 2018;56:1217-24.
 • Hoffmann J, et al. European Headache Federation guideline on idiopathic intracranial hypertension. *J Headache Pain.* 2018;19:93.
 • Mollan SF, et al. Idiopathic intracranial hypertension: consensus guidelines on management. *J Neurol Neurosurg Psychiatry.* 2018;89:1089-1100.

CSF Hydrodynamics

Where is CSF made again???

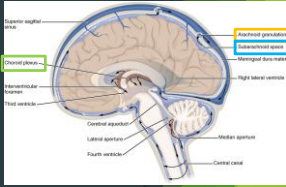


https://commons.wikimedia.org/wiki/File:Diagram_showing_where_the_ventricles_are_in_the_brain_CSF_107.png

Answer: Choroid Plexus in Lateral, 3rd, & 4th Ventricles

Where does CSF drain to?

- ▶ CSF is absorbed from the **subarachnoid space** across the **arachnoid villi** into the **venous circulation**.
- ▶ The arachnoid villi act as one-way valves between the subarachnoid space and the dural venous sinuses. The rate of absorption correlates with the CSF pressure.
 - ▶ "Pressure gradient valves"
- ▶ Dural Venous Sinus Thrombosis important to rule out with MRV!



https://en.wikipedia.org/wiki/Ventricular_system

Pathophysiology of IIH...

- ▶ Any blockage along this CSF drainage pathway or in the venous sinus drainage can result in increased ICP!
- ▶ Bottom Line MOA = obstruction of intracranial venous drainage

PTC /IIH Treatment Options...

1. Weight loss (5-10% is sometimes curative!)
2. Carbonic anhydrase inhibitors (6-50% less CSF production)
 - ▶ Acetazolamide and/or Topiramate
 - ▶ No oral steroids → weight gain
3. Ventriculoperitoneal Shunt / Lumboperitoneal Shunt
 - ▶ Headaches only ; vision stable
4. Optic Nerve Fenestration
 - ▶ Vision/Visual Field worsening ; no headaches
5. Venous Sinus Stenting
 - ▶ In venous sinus stenosis

Topamax vs. Diamox?

- ▶ **Acetazolamide** = CAI inhibitor ; works on ciliary body and choroid plexuses
 - ▶ It has been shown to reduce CSF production in humans by 6% to 50%. This inhibition appears to require a higher dosage than is routinely used (routine dose: 1 g/day po)
- ▶ **Topiramate** = novel anticonvulsant with many MOA; epilepsy/migraines
- ▶ Sulfonamide drug...be careful of sulfa allergies
- ▶ Also has carbonic anhydrase inhibition component; and decreases appetite
 - ▶ Weight loss of 5-10% alone may be curative in some cases of IIH
 - ▶ Average weight loss of 7.3% was obtained in one year on medication
- ▶ Monitor for angle closure glaucoma and myopic shift!!!
 - ▶ 85% of this happens within first 2 weeks of therapy
 - ▶ Ciliochoroidal infundus occurs
 - ▶ MOA = lenticular/uveal effusion and ciliary edema causing forward displacement of the lens-iris diaphragm with resultant narrowing of the anterior chamber.
 - ▶ D/c med; cycloplege and IOP lowering meds may be needed. LPI not helpful.

Alore PL., Jay WM, Macken MP. Topiramate, pseudotumor cerebri, weight-loss and glaucoma: an ophthalmologic perspective. *Semin Ophthalmol*. 2008;21:15-17.

How long should CAI's be maintained?

- ▶ Can/Should Tx ever be discontinued once Sn/Sx are under control?
- ▶ A long-term follow up study was done in PTC patients using a CAI (acetazolamide) over 6.2 years.
 - ▶ 54 total patients followed for **over 6 years**
 - ▶ 60% of patients experienced multiple recurrent episodes over this time span
 - ▶ None of the recurrences occurred while maintained on acetazolamide!
- ▶ Good evidence to maintain longterm Tx???

Wester A, Hildayer A, Goldhammer Y, Almog Y, Korczyn AD. Idiopathic intracranial hypertension: risk of recurrence. *Neurology*. 2004;63:1772-9.

Idiopathic Intracranial Hypertension Treatment Trial (IIHTT)



HHS Public Access

Author manuscript
JAMA. Author manuscript; available in PMC 2015 March 17.

Published in final edited form as:
JAMA. 2014 April 23;311(16):1641-1651. doi:10.1001/jama.2014.3312.

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

What was being investigated?

- ▶ To determine whether acetazolamide is beneficial in improving vision when added to low-sodium weight diet in patients with IIH and mild vision loss over a 6 month period of Tx
- ▶ N=165 (86=acetazolamide & diet, 79=placebo & diet)
 - ▶ Completed: n=126 (69 vs. 57)
- ▶ -2 to -7 dB perimetric mean deviation at baseline
- ▶ Acetazolamide initial dosage = **500 mg BID PO**
- ▶ Increase in 250 mg tablet every week up to **4000 mg/day!!!**

Outcomes being measured...

- ▶ Primary outcome = change in PMD from baseline → 6 mo in most severe eye
- ▶ Secondary outcomes
 - ▶ change in PMD from baseline → 6 mo for better eye
 - ▶ papilledema grade
 - ▶ CSF pressure
 - ▶ visual acuity
 - ▶ QOL
 - ▶ vital signs
 - ▶ lab results
 - ▶ presence of HA
 - ▶ treatment failure

Visual Field Findings...

- ▶ Acetazolamide/Diet = 1.43 dB improvement
 - ▶ Placebo/Diet = 0.71 dB improvement
- } Most severe eye
- ▶ Acetazolamide/Diet = 0.87 dB improvement
 - ▶ Placebo/Diet = 0.42 dB improvement
- } Better eye

Papilledema Grading...

- ▶ Papilledema grade 3-5 = 2.27 dB improvement
- ▶ Papilledema grade 1-2 = -0.67 dB improvement
- ▶ Significant improvement in acetazolamide groups in study and fellow eyes with FP's
 - ▶ QOL improved too
- ▶ Weight change
 - ▶ -7.50 kg in acetazolamide at 6 mo
 - ▶ -3.45 kg on placebo at 6 mo
- ▶ CSF Pressure
 - ▶ -112.3 mmH2O on acetazolamide at 6 mo
 - ▶ -52.4 mmH2O on placebo at 6 mo
- ▶ No change in headache severity or visual acuity
 - ▶ 69% still had headaches on acetazolamide at 6 mo
 - ▶ 68% still had headaches on placebo at 6 mo

Adverse Events

- ▶ 9 total patients in study dropped out
 - ▶ Decreased CO2 levels
 - ▶ Increased Cl- levels
 - ▶ Mild decrease in potassium levels (no supplementation needed)
 - ▶ No changes to sodium
 - ▶ No liver function changes
 - ▶ Conclusion:
 - ▶ "In patients with IH 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. The clinical importance of this improvement remains to be determined."
- NORDIC IIHTT 2014

1. Monitor K levels?

- ▶ No participant on acetazolamide alone required potassium supplementation; thus, **routine monitoring of potassium levels is not recommended.**

2. Monitor with CBC with differential?

- ▶ No patient experienced aplastic anemia and **periodic monitoring of blood cell counts is not necessary or cost-effective.**

Ten Have MW, et al. Safety and tolerability of acetazolamide in the IIHTT. *J Neuroophthalmol.* 2016;36:13-19.

Allergy to acetazolamide?

- ▶ "Allergic reactions to acetazolamide were uncommon. Furthermore, **there is no evidence to suggest that an allergy to sulfonamide antibiotics increases the risk of an allergic reaction.**"

Ten Have MW, et al. Safety and tolerability of acetazolamide in the IIHTT. *J Neuroophthalmol.* 2016;36:13-19.

Pregnancy Considerations...

- ▶ **Topamax** = FDA Category D; evidence shows up to 10-20% of dose can be found in infants who are nursing
- ▶ **Acetazolamide** = FDA Category C; case reports of placenta crossing ; has been avoided in pregnancy in the past....new evidence to suggest otherwise?
- ▶ If both are avoided in pregnancy, then sometimes repeat LP's may be necessary in short term to keep ICP down; inherent risks...

Lee AG, et al. The use of acetazolamide in idiopathic intracranial hypertension during pregnancy. *Am J Ophthalmol.* 2005;139:855-9.

National Collaborative Perinatal Project (NCPP) 1959-1974

- ▶ "The use of carbonic anhydrase inhibitors (CAIs) has a large pool of human data on which to base clinical decisions. The source is the National Collaborative Perinatal Project (NCPP) conducted by the NIH from 1959 through 1974. This study monitored more than 50,000 mother-child pairs and 1,024 instances of systemic usage of acetazolamide during pregnancy. In the resulting offspring, there were 18 instances of malformations. The predicted number due to chance was 18.06. This suggests that the incidence of malformations from acetazolamide exposure during pregnancy is no greater than the natural incidence. In the same study, there were 12 documented first trimester exposures to acetazolamide. No anomalies were observed in the resulting offspring."

---Steven Odrich, MD (Bronx, NY)

<http://www.aaa.org/publications/cyenet/200906/letters.cfm?renderoforPrint=18>

Lee et al. The use of Acetazolamide in IIH During Pregnancy. *Am J Ophthalmol.* 2005;139:855-9.

- ▶ 12 patients on Diamox 500 mg BID PO during pregnancy
- ▶ No adverse side effects nor congenital malformations noted
- ▶ Cited the results of the Collaborative Perinatal Project as well
- ▶ "In summary, there is no convincing evidence from the literature for the recommendation to limit the use of acetazolamide for IIH in pregnancy. Although the use of acetazolamide might be restricted in the first trimester, this recommendation may have a more medicolegal than medical rationale. It is our recommendation that acetazolamide be considered if the risk of nontreatment (e.g., progressive visual loss) is sufficiently high to warrant its use."

Falardeau J, et al. The use of acetazolamide during pregnancy in intracranial hypertension patients. *J Neuroophthalmol.* 2013;33:9-12.

- N= 158 pregnancies (101 women) with IIH diagnosis
 - N= 50 pregnancies who used acetazolamide used before 13 weeks gestation = study group
 - N=108 women with IIH but no acetazolamide use = control group
- Total spontaneous abortion rate → not statistically significant (p=0.36)
 - 28% acetazolamide users
 - 21.3% nonusers
- Also, no statistical difference when comparing groups in the following manner:
 - Untreated group → 13.9%
 - Acetazolamide <1000 mg/day → 22.2%
 - Acetazolamide >1000 mg/day → 17.4%

Falardeau J, et al. The use of acetazolamide during pregnancy in intracranial hypertension patients. *J Neuroophthalmol.* 2013;33:9-12.

- "There is clearly a lack of convincing evidence for a teratogenic effect associated with this use of acetazolamide during pregnancy. In all reported cases, there is no common morphological abnormality that one would expect with a teratogenic agent, and specifically, there are no cases of postaxial limb malformations in human offspring as found in animal models."
- "In summary, our study confirms the lack of convincing evidence for adverse effects of acetazolamide use in human pregnancy, even when prescribed prior to the 13th week of gestation. We agree with Lee et al that the avoidance of acetazolamide during the first trimester has very little medical justification and is mainly guided by medical-legal rationale."

Question to y'all:

Is it appropriate for O.D.'s to prescribe Diamox for these patients long term???

Surgical Considerations...

1. Headaches only, vision stable (can be used for both HA's and vision too)
 - LP shunt, VP shunt
2. Vision loss/VF worsening despite maximal medical Tx
 - Optic Nerve Fenestration
3. Venous sinus thrombosis
 - Anticoagulants
4. Venous sinus stenosis
 - Venous sinus stenting

- ▶ Majority can be managed via weight loss and oral meds (Diamox)

1A) Ventriculo-Peritoneal Shunts (VPS) and Ventriculo-Atrial Shunts (VAS) in IIH

The diagram shows a sagittal view of the brain with a ventricular catheter inserted into the lateral ventricle. The catheter is tunneled under the skin and leads to a reservoir. From the reservoir, a tube empties into the chest or abdominal cavity. Two axial CT scans show the placement of the catheter in the brain and the reservoir in the abdomen.

Labels: Ventricles, Ventricular catheter, Catheter lies tunneled under the skin, Tube empties into the chest or abdominal cavity.

URLs: https://en.wikipedia.org/wiki/Cerebral_shunt, https://en.wikipedia.org/wiki/Brain_abscess, https://en.wikipedia.org/wiki/Brain_abscess

1B) Lumboperitoneal shunts in IIH

- L3/L4 or L4/L5 spaces most commonly used
- Drain into peritoneal space like VPS

The diagram shows a cross-section of the spine with a catheter inserted into the L3/L4 or L4/L5 space. The catheter is connected to a reservoir and drains into the peritoneal space.

Labels: Spinal cord, Epidural space, L3, L4, Subarachnoid space.

URL: https://en.wikipedia.org/wiki/Epidural_blood_patch

VPS vs. LPS...

- Ventriculo-Peritoneal Shunt (VPS):
 - Infection rate of 7-15%
 - 20% revision rate q2 yrs
- Lumbar Peritoneal Shunt (LPS):
 - Infection rate of 1%
 - 50% revision rate q2 years

• "In short, most shunted PTC patients require multiple revision surgeries during their lifetime."
---Galgano MA et al. (2013)

2) ON Sheath Fenestration...

- Described in 1872 by DeWecker; expanded upon by Heyreh in 1964
- Defn: make slits in ON sheath to reduce the local pressure around the optic nerves.
- ~50% of unilateral ON sheath fenestration procedures results in resolution of visual symptoms in both eyes.
 - Both optic nerves are connected via the subarachnoid tissue around the optic chiasm
- Typically only done for visual Sn/Sx without headaches...
 - If headaches → shunt procedure may be better option
- Safe and effective up to 10 years per several studies
- Revision rate is usually very low ; 1 procedure per lifetime generally

Optic Nerve Fenestration

- ONSF procedure is approximately 25-30 minutes in duration for a bilateral operation
 - (10-15 minutes per eye)

The diagram shows a cross-section of the optic nerve sheath with a fenestration (slit) made in the lamina propria. Labels include: External coating of sheath, Optic nerve sheath, Pia mater, Arachnoid, Subarachnoid space, Basilar part of optic nerve, External artery and vein of optic nerve.

URL: https://en.wikipedia.org/wiki/Optic_anc

Super hard question to answer: ---When should I send for ONSF ???

- "At present, evidence-based guidelines for when ONSF should be utilized in the management of vision loss in IIH do not exist."
- "Neither are there any prospective studies to guide surgical decision making, nor are there data comparing ONSF visual outcomes to CSF diversion or medical therapy."
- "When visual loss is noted at presentation, Corbett et al have recommended consideration of ONSF early in the clinical course. Banta and Farris recommend ONSF when progressive visual loss, as defined by loss of >2 lines of Snellen acuity or new onset or progression of VF defect, occurs despite initial medical management."

??? ↻

Mudumbai RC. Optic nerve sheath fenestration: indications, techniques, mechanisms, and results. *Int Ophthalmol Clin.* 2014;54:43-9.

Define "Progression of VF defect" please!?

- ▶ In IIHTT by NORDIC Group → MD of <-7.00dB used for surgical cutoff
- ▶ Treatment failure of Acetazolamide defined in NORDIC1
 - ▶ Baseline MD of: 0.00 to -3.5 dB → **worsened by >2 dB**
 - ▶ Baseline MD of: -3.5 to -7.0 dB → **worsened by >3 dB**

} Confirmed **TWICE** by repeat VF's Z4-Z

NORDIC Study Group. Effect of acetazolamide on visual function in patients with IIH and mild visual loss: The IIHTT. JAMA. 2014;311:1641-51.

Criteria for ONSF by OSU in Columbus, OH

- ▶ Indications for ONSF included persistent symptomatology (i.e., headache) and/or signs (i.e., papilledema) associated with increased intracranial pressure despite maximal medical therapy or neurosurgical shunting, or severe vision loss at presentation.
- ▶ ONSF procedure is approximately 25-30 minutes in duration for a bilateral operation (10-15 minutes per eye)

Vaidya NS, et al. Visual outcomes following optic nerve sheath fenestration via the medial transconjunctival approach. Orbit. 2016;35:271-7.

How often is ONSF actually needed for IIH?

- ▶ Only 14 patients (31 eyes) over a 7-year period actually needed ONSF in a tertiary referral center...
- ▶ Shows how uncommon the need for ONSF really is!

Obi EE, et al. Optic nerve sheath fenestration for idiopathic intracranial hypertension: a seven year review of visual outcomes in a tertiary centre. Clin Neurol Neurosurg. 2015; 137:94-101.

New Study! Surgical IIH Treatment Trial (SIGHT)

- ▶ Currently underway and recruiting!
 - ▶ Starting date → October 2018
 - ▶ Estimated completion date → June 2021
- ▶ Randomized clinical trial n=180 patients
- ▶ Comparing the following surgical options in IIH patients:
 1. Medical therapy alone (ie. Diamox and diet/weight loss)
 2. MT + ONSF
 3. MT + VP shunting

Study Terminated Due To Low Enrollment

https://clinicaltrials.gov/ct2/show/NCT03501966

3) Dural Venous Sinus Thrombosis

The slide contains two images. On the left is an anatomical diagram of the dural venous sinuses, with labels for the Superior sagittal sinus, Inferior sagittal sinus, Great cerebral vein, Superior petrosal sinus, Middle petrosal sinus, Inferior petrosal sinus, Transverse sinus, Sigmoid sinus, Jugular bulb, Internal jugular vein, and Occipital sinus. On the right is a CT scan of the head at the level of the sigmoid sinus, with a yellow arrow pointing to a filling defect in the sigmoid sinus, indicating a thrombosis. The CT scan includes technical details: Im 11 (17/25), DICOM PARS COMBINA COMPRESS 512x512, MIP 1.75 mm 250 ms 120kV, LUT 100, Slices 51 @ 5mm, Window 512x512 (H, W, 40kV), and a URL: http://www.radiopaq.org/2007/01/14/transverse-sinus-thrombosis-2/.

DVS Thrombosis Considerations...

- ▶ Blood clots in young people are not normal...
- ▶ If DVST occurs, hematological workup and anticoagulant therapy is required.
 - Subramanian PS et al. (2014)
- ▶ Consider: CBC with diff, CMP, lipid panel, PT/PTT, Protein S, Protein C, Homocysteine levels, Lupus anticoagulant, anticardiolipin, Factor V Leiden, Prothrombin mutation, Antithrombin III mutation, Sickledex screen, hemoglobin electrophoresis

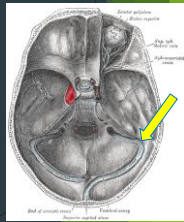
DVS Thrombosis Treatment...

- ▶ Rule out clotting disorder, infection, etc.
- ▶ Aggressive anti-coagulation (heparin, warfarin, clopidogrel)
- ▶ Not a candidate for DVS Stenting in vast majority of cases...
- ▶ If anticoagulation and oral CAI's do not work, then may need shunt surgery

Dural Venous Sinus Stenosis

Dural Venous Sinus Stenosis (DVSS)

- ▶ Defn: focal, narrowed section of dural venous sinuses causing back up/turbulent venous blood flow
- ▶ ***Most common at junction of Sigmoid and Transverse sinuses**
- ▶ Not a true blood clot like DVST is...
- ▶ Treatment = weight loss, oral CAI, and/or DVS Stenting procedure



DVSS in IIH vs. Normals...

- ▶ Focal stenosis has been demonstrated in **90+% of IIH patients** using advanced imaging techniques.
- ▶ Furthermore, focal stenosis in the same sinus territory was only demonstrated in **6.8% of asymptomatic control subjects**.
- ▶ Might be on to something here....

4) Dural Venous Sinus Stenting

- ▶ Right transverse sinus is dominant in **73% of cases**
- ▶ **MOA:** Increases drainage of venous blood from venous sinus system which helps with the pressure dependent valves, arachnoid villi granulations, allowing them to clear CSF in to the venous system more efficiently/quickly → decreasing ICP
- ▶ High frequency of resolved or improved HA's and papilledema with this method

DVS Stenting...

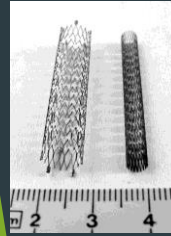
- ▶ Not every patient is a candidate for DVS Stenting
- ▶ **Criteria Needed:**
 1. **Presence of venous sinus stenosis (MRI/MRV) ; not thrombosis...**
 2. **Transvenous manometry across the stenosis >10 mmHg differential**
- ▶ Catheter with stent and manometer placed in femoral vein and "fished" upwards to location of sinus stenosis
- ▶ **Post-Op Medications:**
 - ▶ Plavix 75 mg x 6-12 weeks then d/c
 - ▶ ASA 325 mg for life

Transvenous Manometry in IIH

Significant =
 >10 mmHg pressure
 differential of proximal
 vs. distal locations in
 respect to stenosis

Differential Example:
 Pre Stent = 31 mmHg
 Post Stent = 1 mmHg

Dural Venous Sinus Stenting for IIH



Suboccipital/Subtemporal Cranial Decompression

- ▶ Very invasive ; but historically pretty successful...
- ▶ Remove part of skull to allow for more room inside...
- ▶ Not gold standard anymore
- ▶ Can be used in severely refractory cases unresponsive to traditional surgical procedures for ICP and IIH

Optometry IIH/PTC Summary...

- ▶ Make the diagnosis
- ▶ Get MRI/MRV
- ▶ Refer for LP
- ▶ Neurology should start Diamox/Topamax
- ▶ Monitor x 1 month post med Tx, then q3-4 months until resolution/stability (varies)
- ▶ Serial OCT, FP, and HVF's are necessary to gauge Tx/stability
- ▶ Relay findings to managing neurologist/PCP regularly
- ▶ Encourage weight loss

What happened to patient #2?

- ▶ "No-showed" to neurologist twice!
- ▶ Has not returned phone calls or letters...
 - ▶ Phone has been disconnected...
- ▶ Possible candidate for ONSF? DVSS? Diamox only? All 3?
- ▶ "You cannot care more for a patient than what they care for themselves."----Joseph Sowka, OD and Alan Kabat, OD

Thank you!!!

Questions??
 Accolades??

Email → cborgman@sco.edu



Complaints???



• Email →
wmcgriff@sco.edu



SOUTHERN
 COLLEGE OF
 OPTOMETRY