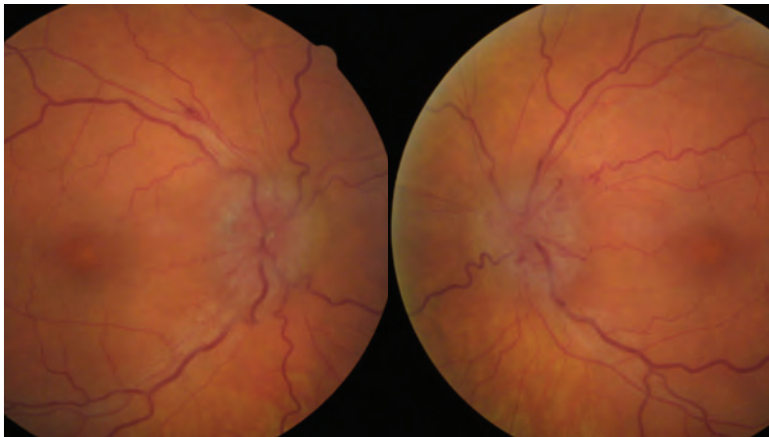


Differential Diagnosis of ONH Edema

Beth A. Steele, OD, FAAO
bsteele@uab.edu

Disclosures 2022-23– Dr. Beth Steele

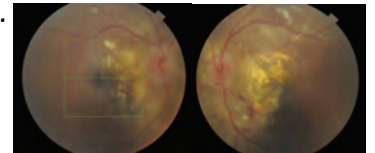
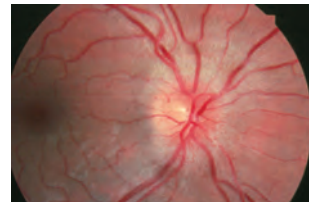
- Nothing to disclose



Causes of disc edema ...

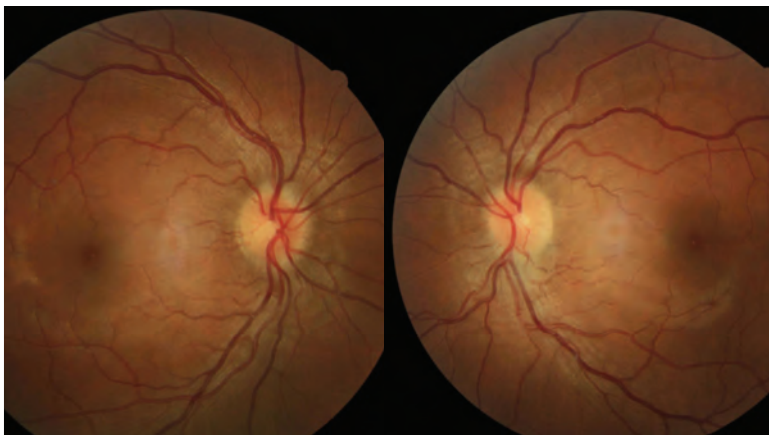
Unilateral

- Vascular
- Infectious
- Diabetic Papillopathy



Bilateral

- Hypertensive
- Toxic
- Inflammatory
- Infectious
- Compressive
- Hydrocephalus
- IICHTn



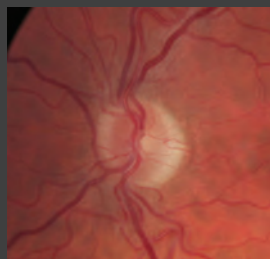
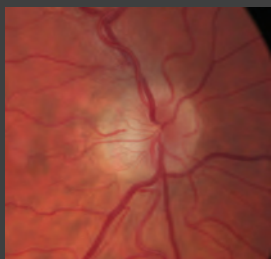
Is it...?
And if so then why is it?

Pseudopapilledema (H47.33_)

Disc Edema

Optic Disc
Drusen

Anatomical



Myopic / tilted discs?

Be careful not to hide behind a comfortable label...

Some things make you look twice....

Worrisome findings....

- Elevation
- Pallor
- Discoloration
- NFL defects
- Vascular changes

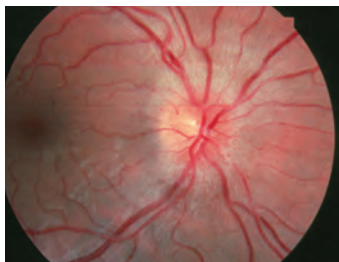
Tools you have...

- Stereoscopic DFE!
- Swinging flashlight test
- Pupil cycle time
- Red-free filter
- SVP
- VF
- OCT
- FAF
- B-scan



Terminology – 30% error rate

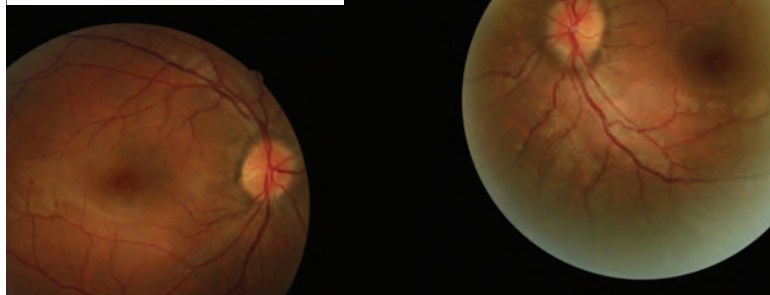
- Disc Edema
- Papilledema
- ~~Bilateral papilledema~~
- Pseudopapilledema
- ~~Pseudotumor~~
- ~~Benign~~ Idiopathic Intracranial Hypertension



FULL TEXT ARTICLE
Inappropriate Use of the Term "Papilledema" in the Medical Literature: A Systematic Review of Case Reports Across Specialties
Brendan Tan, MD, PhD, Edward S. Grunstein, MD, PhD, and Jonathan S. Siegel, MD, PhD
Ophthalmology
Copyright © 2015
DOI: 10.1016/j.ophtha.2015.05.010
Read Full Article
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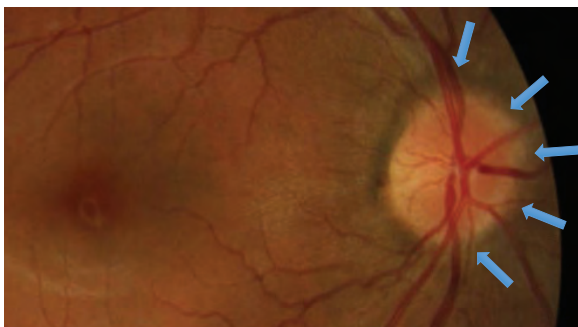
31 AAF

"nasal margins mildly indistinct;
RTC x 1 week for further
evaluation"

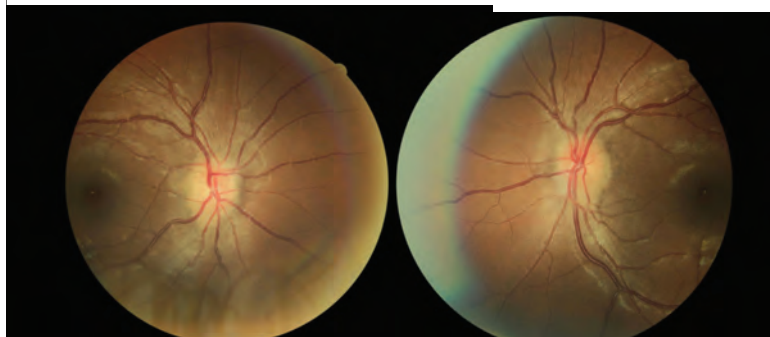


Early on – can appear as a nasal "C-shaped" edema

- Temporal area spared
- VA often normal



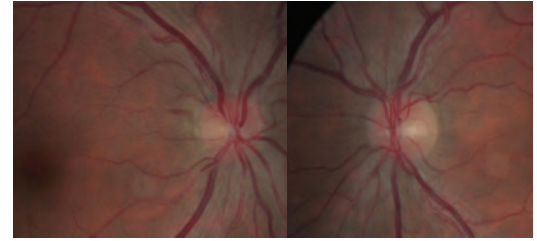
6 years later....



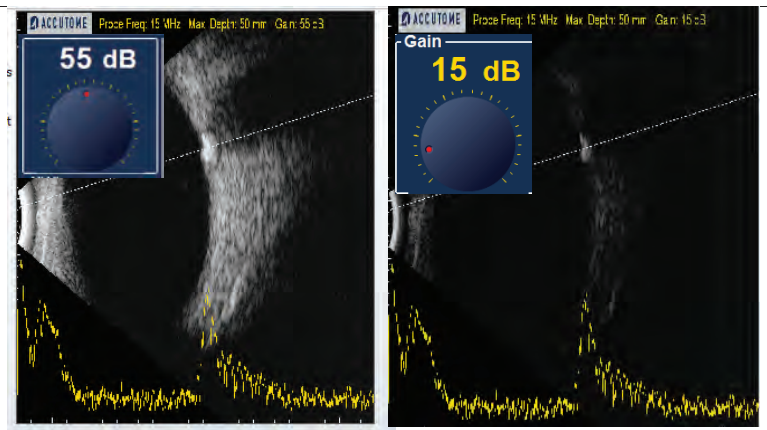
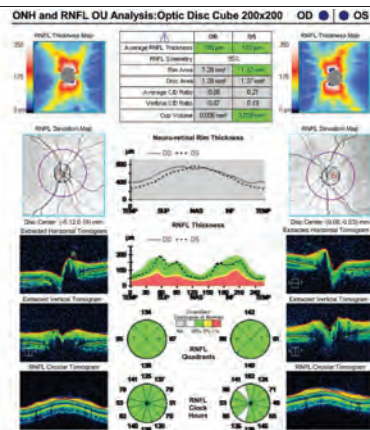
Bilateral Buried Disc Drusen – A common diagnostic dilemma

43 AA Female

- 20/15
- 140/90
- DFE: drance heme
OD and disc elevated
nasally

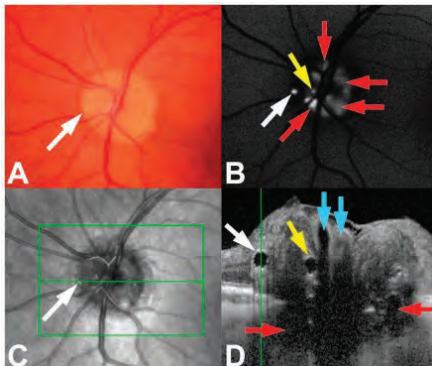


Normal RNFL

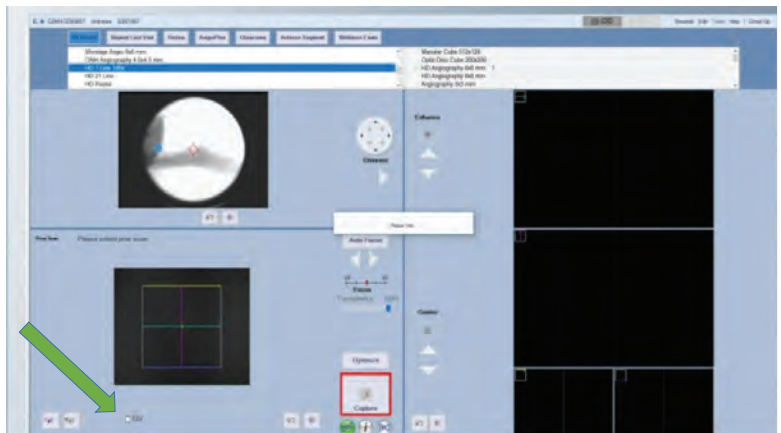


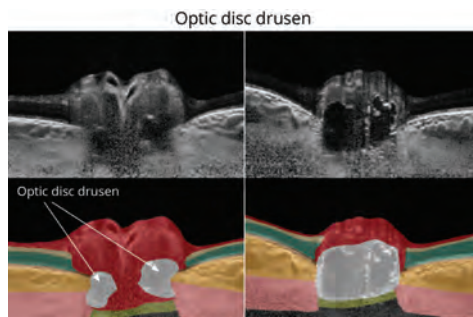
Recommendations from the ODD Consortium

- Radial ONH scans with EDI
 - RNFL scans (no EDI)
 - FAF
-
- Located above lamina
 - Signal poor core, with superficial hyperreflectivity
 - Often conglomerates
 - Can mimic vessels



Malmqvist et al. Optic Disc Consortium
Recommendations for Diagnosis of ODD using
OCT. Neuro-Ophthalmology 2017.





Girard M et al. J of Neurology 2022.

Ultr
con

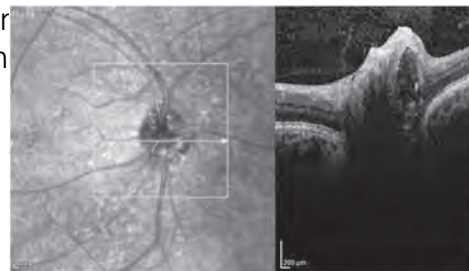


Figure 3. Optical coherence tomography: Large optic disc drusen, visible as a hypo-reflective structures with a total or partial hyperreflective margin, as described by the Optic Disc Drusen studies (ODDS) Consortium. The hyperreflective margin is often more evident superiorly and can be difficult to detect.

Rosa N et al. J of Clinical Medicine 2022.

Peripapillary Hyperreflective Ovoid Mass Structures (PHOMS)

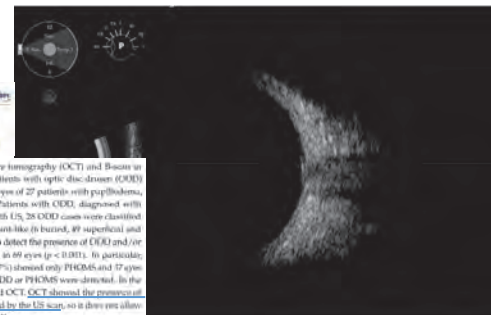
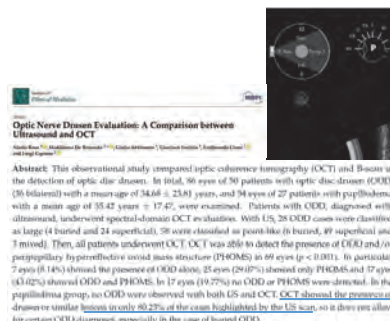
- Described initially as possible ODD precursors or variants
- Also present in a variety of diseases, including papilledema, NAION, RVO, acute demyelinating optic neuritis



Figure 4. Optical coherence tomography: A hyperreflective peripapillary structure similar to ovoid mass peripapillary hyperreflective ovoid mass structure (PHOMS), as described in patients with optic disc drusen.

Rosa N et al. J of Clinical Medicine 2022.
Fraser, J.A. et al. J Neuroophthalmol. 2021.

But...OCT may miss up to 20% of ODD



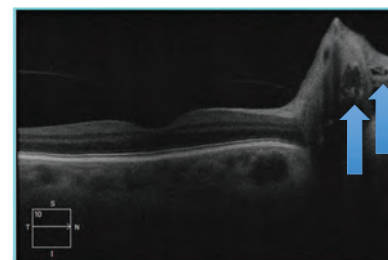
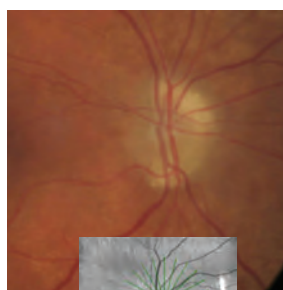
OCT preferred for ODD in kids

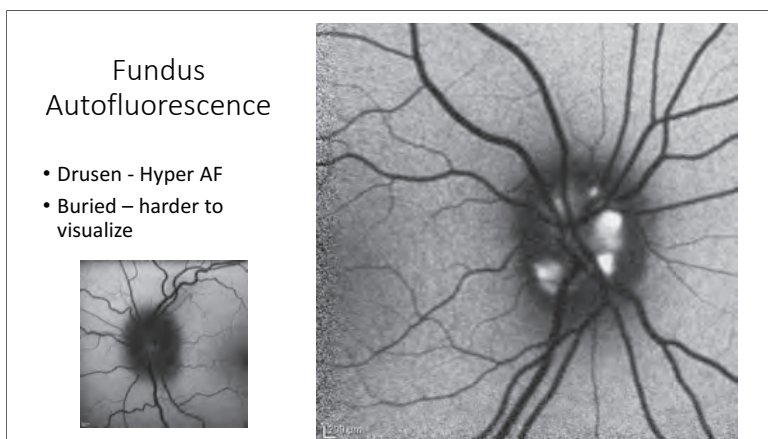
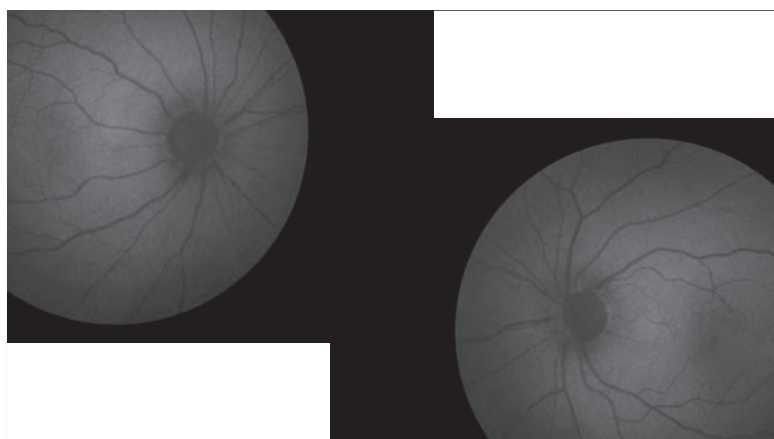
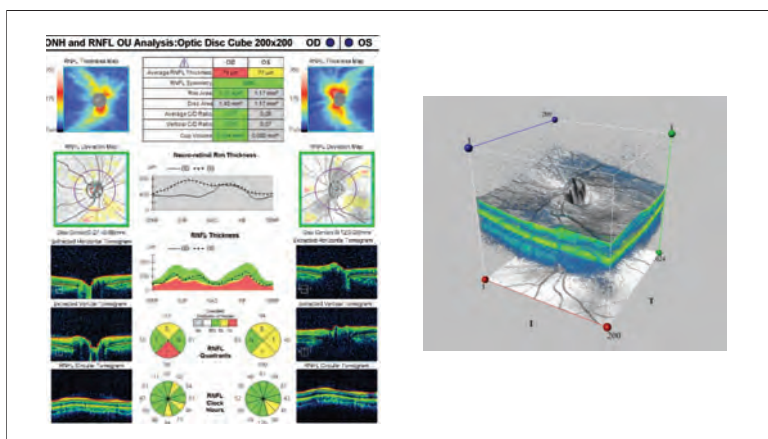
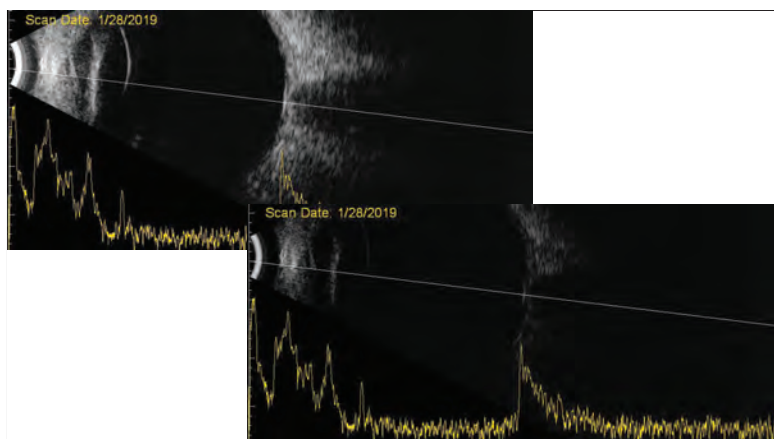
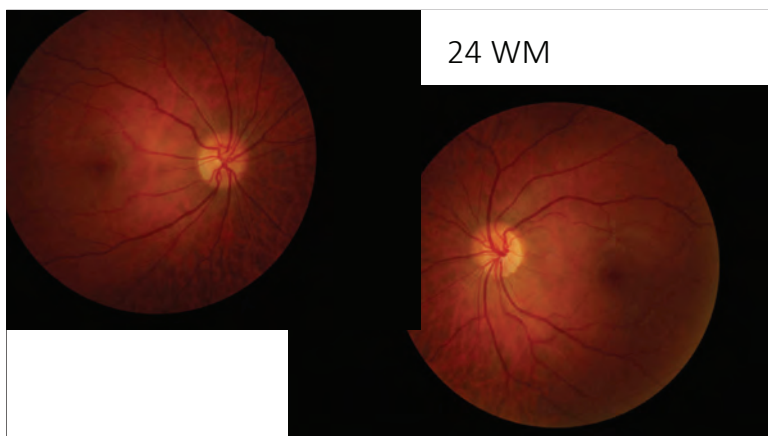
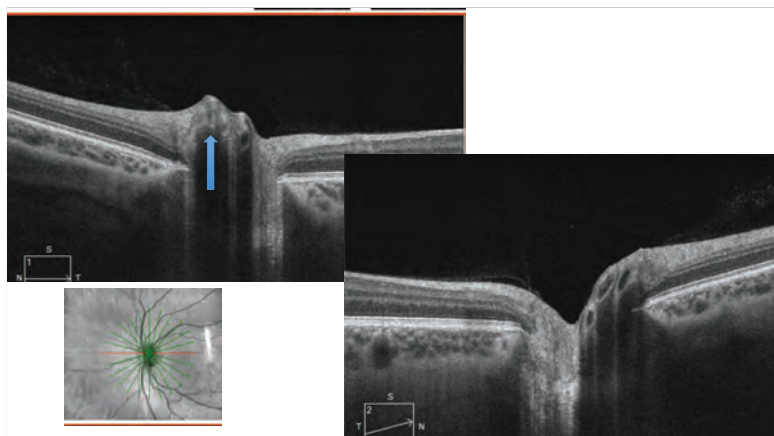
- Mainly with US, Leon et al. [12] examined 46 children with calcific ODD. Furthermore, they claimed that since drusen are often not calcified in children, they may not be visible under B-scan, and so the preferred device should be EDI-OCT

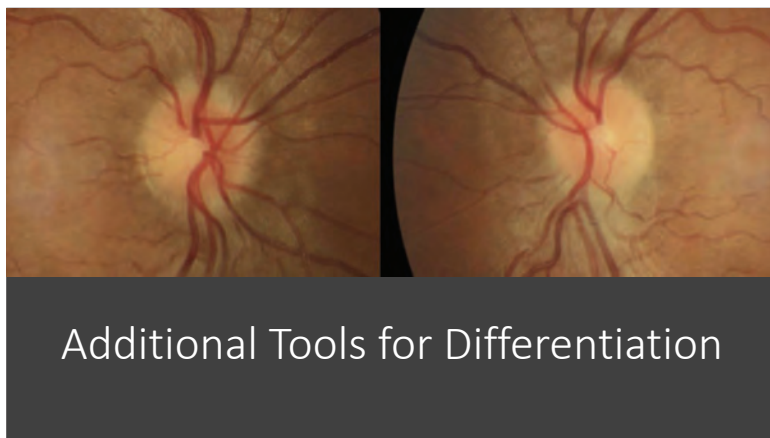
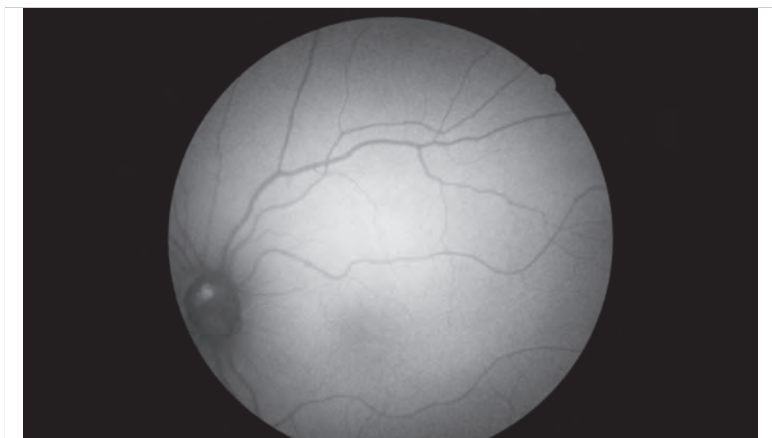


Article Optic Nerve Drusen Evaluation: A Comparison between Ultrasound and OCT

Nicola Rosa ¹, Maddalena De Bernardo ^{1,*}, Giulia Abbinante ¹, Gianluca Vecchio ¹, Ferdinando Cione ¹ and Luigi Capasso ²



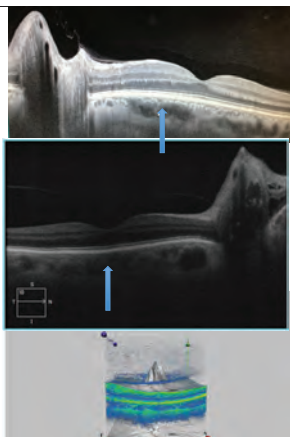




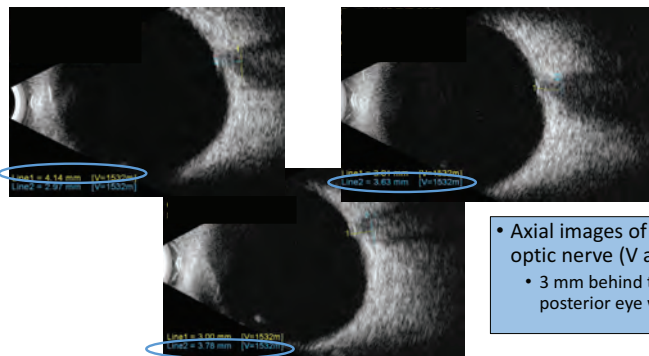
Additional Tools for Differentiation

OCT: Globe Convexity

- Increased ICP will push the globe anteriorly
- Easiest to appreciate with a 9mm scan
- With EDI, can see an anteriorly displaced Bruch's membrane
- What about with high myope? Look at most posterior aspect of sclera...



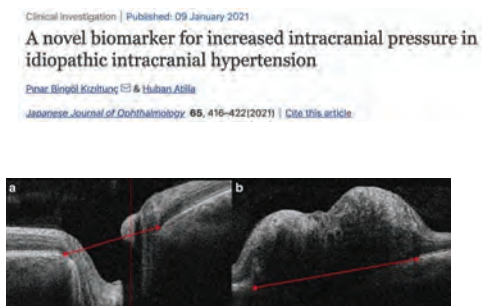
Normal ONH Sheath Diameter can ease your mind as well...



- Axial images of the optic nerve (V and H)
- 3 mm behind the posterior eye wall

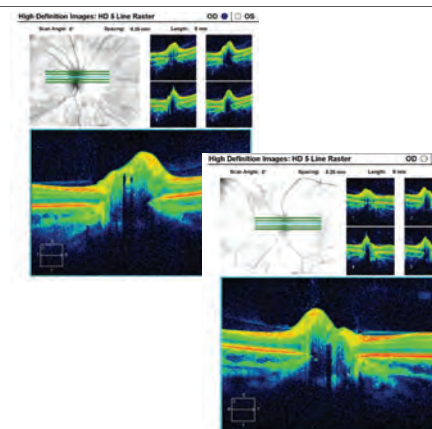
Bruch's Membrane Opening?

- Predictive: 1785 microns
- 78.8% sensitivity
- 81% specificity

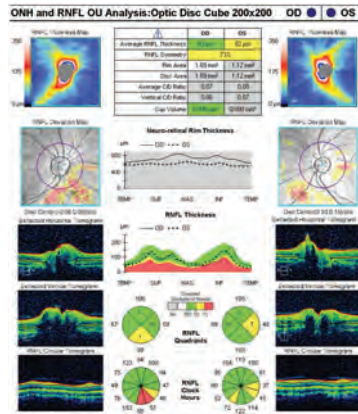


34 WF

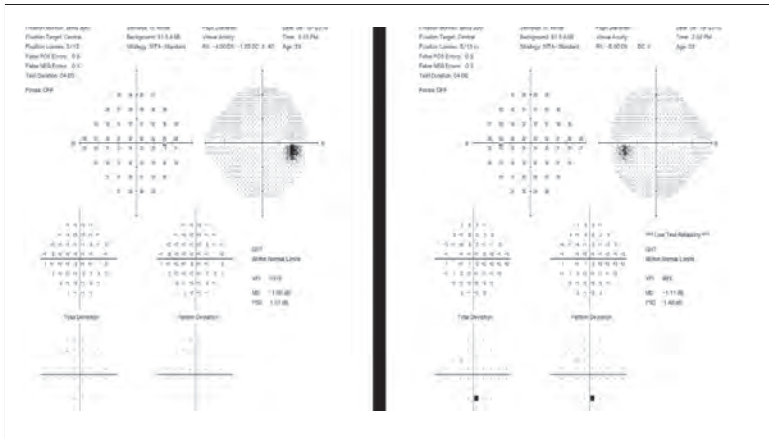
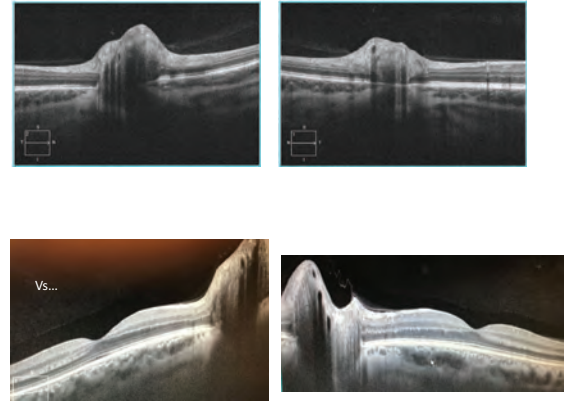
- 20/20, pupils normal, CF normal
- -7.00 OU
- BMI 31
- Denies H/A
- Elevated ONH
 - .1/.1 CD
 - +SVP
- Normal RNFL thickness



- Normal RNFL thickness

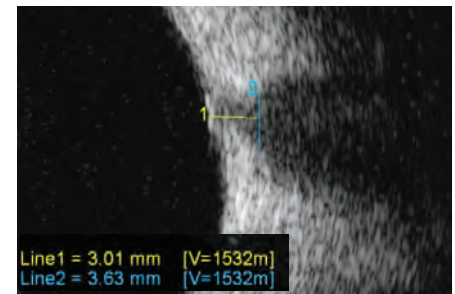


Globe convexity is normal

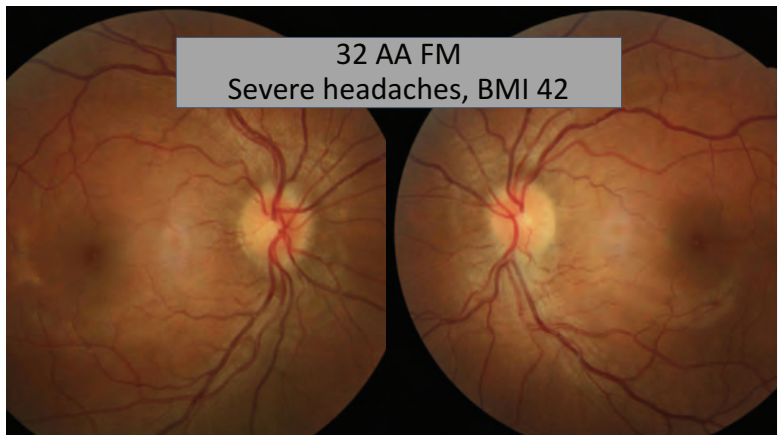
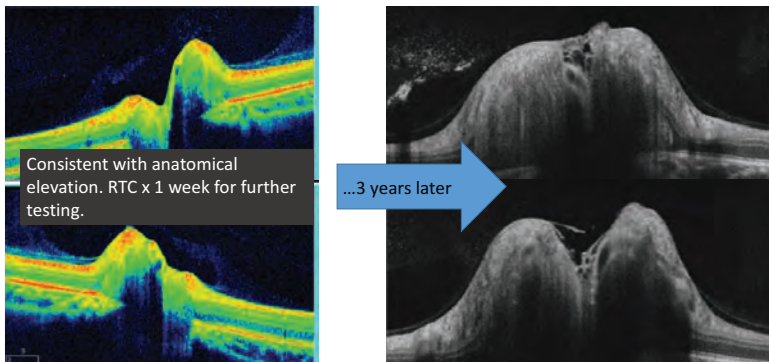


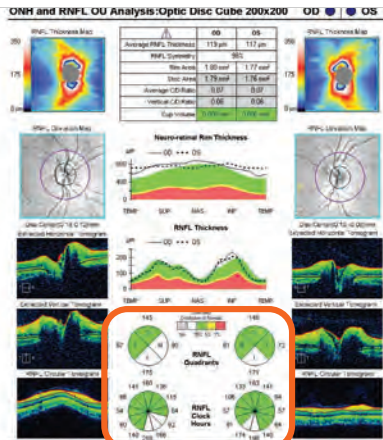
This patient...

- Exam
 - Elevated disc
 - +SVP
 - B-scan
 - (-) hyperreflectivity consistent with drusen
 - (-) crescent sign
 - Normal ONH sheath diameter
 - OCT
 - Normal RNFL thickness
 - Normal globe convexity
 - Stability....
- Presumed crowded discs but careful follow up



But sometimes...despite your tools....





Papilledema Suspected ? Now what...

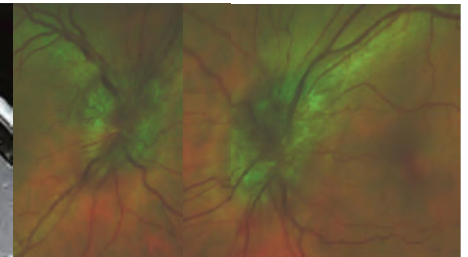
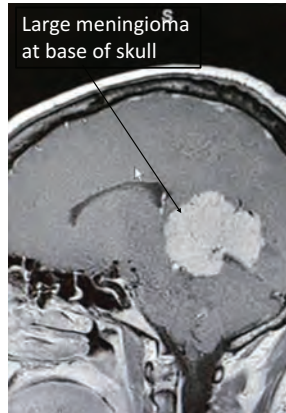
- Brain Imaging
 - MRI – rule out space occupying mass
 - MRV – rule out cerebral venous thrombosis
- Lumbar Puncture
 - With opening pressure
 - Higher than 20/25/30 cm H2O is abnormal
- Co-management
- Determine underlying cause/association if any
 - Weight
 - Associated medications
 - Modified Dandy criteria, 1985; Friedman criteria 2022

MRI features

- Order with/without contrast, T1/T2-weighted, with fat suppression
- Empty sella
- Enlarged ON sheath
- Increased tortuosity of ON
- Flattened sclera
- Anterior protrusion of ONH
- Attenuation of cerebrovenous sinuses

**Lumbar Puncture
opening pressure =
28cm H₂O**

Large meningioma
at base of skull

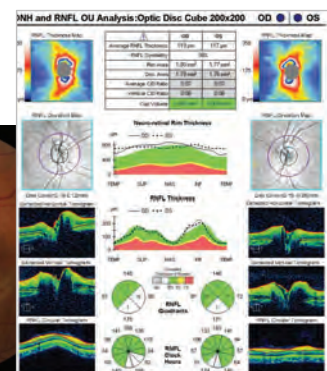
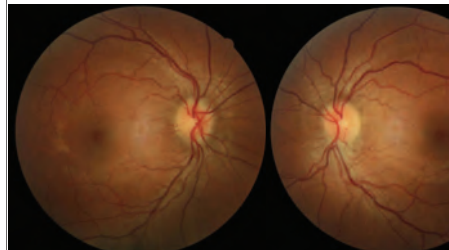


- 41 year old male
- Severe headaches
- BMI 38

26 WF, "blurry vision"
Hx hydracephalous, multiple previous surgeries
VA 20/40 OD, OS
H/A developing with more intensity
BMI 42



Our Patient:
MRI Clear
Opening Pressures of 28



Idiopathic Intracranial HTN

- 90-98% complain of headache
- Nausea/vomiting/dizziness – 40%
- Pulsatile tinnitus – 16-60%
- Visual disturbances – 30%
- No other neurologic findings (some with VIth palsy)
- Eventually:
 - Vision loss
 - Cognitive dysfunction

Thaller M et al. J of Neurology 2022
Mollan SP, et al. Pract Neurol 2018

IIHT – Who Gets It?

- BMI >30
 - >40 – worse visual outcome
 - Rapid weight gain – more severe
- Mostly females
 - Males 10% of the time
 - Not as likely to have H/A – may not come in
- Race – more aggressive in AA
 - 3 x more likely to have vision loss
 - 5 x increase in blindness
- Co-morbidities
 - Increased intra-abdominal pressure
 - HTN
 - Sleep apnea
 - Anemia
 - SLE
 - Uremia
 - hypothyroidism

J. Neuroophthalmol. 2002 Oct 16; 21(4):257-263. doi: 10.1097/00006123-200210000-00007.
Comparison of Idiopathic Intracranial Hypertension Patients With and Without Anemia
Lisa Capovilla¹, Gregory M. Hirsch², Jonathan A. Hirsch³
Affiliations: ¹ report
PMID: 12071466 DOI: 10.1097/00006123-200210000-00007

Abstract
Background: Idiopathic intracranial hypertension (IIH) is a condition of elevated intracranial pressure without an identifiable cause. It usually affects young obese women but has been reported in individuals with diagnosed with anemia or with chronic anemia. The relationship between anemia and the onset and duration of IIH is unclear. This study aims to determine the course of IIH in a series of patients.
Methods: Consecutive patients with IIH were recruited from neuro-ophthalmology clinics. Subsequent ophthalmologic and systemic laboratory data on complete blood counts (CBCs), iron studies, iron levels, and iron status were obtained. Patients with IIH were followed up for 12 months. **Results:** One hundred twenty-three patients with IIH were recruited for this study and 118 (96%) had anemia. More severe individuals had more severe visual field defects. Regression of visual field defects was more likely in patients with anemia (P = 0.001) and iron studies (P = 0.001). The median follow-up time was 4.6 months (range 0.1 to 10.8). Phases of IIH patients with anemia required more treatment and showed a trend toward increased response to treatment. There were no differences in the visual acuity, but patients with anemia had more improvement in visual field defects (P = 0.001). **Conclusions:** IIH patients with anemia had more visual field defects at presentation and more visual field improvement. This suggests that IIH should be considered for patients with anemia because it may influence IIH visual outcome.

Brousse V et al. Am J Oph 2007, 2012.

Etiologies

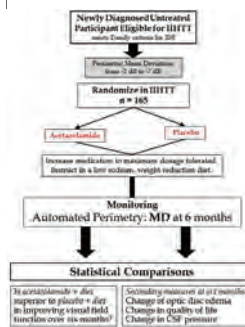
- Neuro-metabolic disorder
Thaller M et al. J of Neurology 2022
- Idiopathic
But what causes the papilledema ?

Axoplasmic flow stasis → swollen nerve fibers → compression of venules in the area and so venous stasis/leakage → accumulation of extracellular fluid

Toxic	Oral contraceptives Tetracyclines Steroids Vitamin A Retinoids Tamoxifen Nalidixic Acid
Other	Obesity

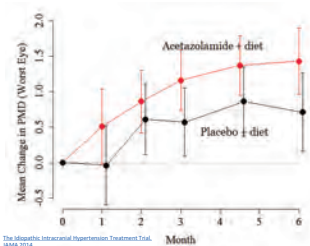
Neurological Advances in Drug Safety
Risk of intracranial hypertension with intrauterine levonorgestrel
Mollan SP, et al. Pract Neurol 2018

IIH Treatment Trial – JAMA, April 2014



- acetazolamide + a low-sodium weight-reduction diet vs. diet alone
→ modest improvement in visual field function
- OCT Substudy of the IIHT, Ophthalmology, Sept 2015
 - RNFL and Total Retinal Thickness (TRT) useful in following and monitoring response to treatment
 - Better RNFL thickness, TRT, and ONH volume swelling measurements

Acetazolamide + Low Sodium/Weight Loss



- Contraindications:
 - Sulfa allergy?
 - chemical structure different than antibiotics – little evidence of cross-sensitivity
 - consider avoiding if hx of severe reaction
 - Long-term: liver, kidney disease, severe COPD
 - Caution with sickle cell
 - Caution with low potassium

Common Side Effects	
paresthesia	metabolic acidosis
metallic taste	electrolyte imbalance (including hypokalemia)
fatigue	renal calculi
malaise	blood dyscrasias
GI disturbances	
decreased libido	

Treatment Goals ...

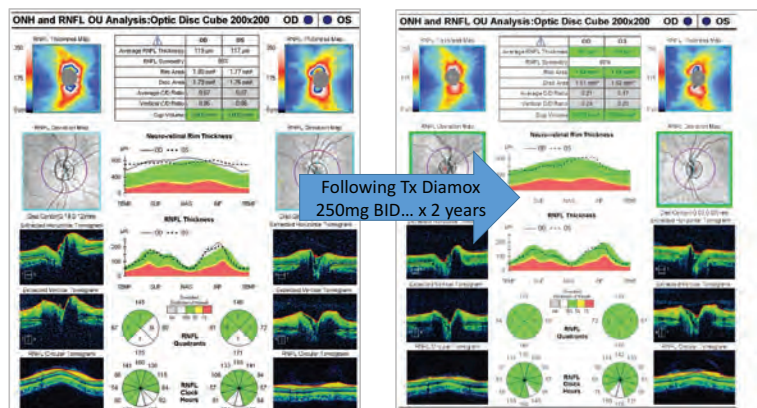


- Also impacted: cognitive function
- Papilledema is a good measure of response to treatment
 - Visual Fields, OCT – chronically
- Co-management
 - Labs, imaging
 - Refractory cases

Thaller M et al. J of Neurology 2022
Mollan SP, et al. Pract Neurol 2018
Cello KE J Neuroophth 2016

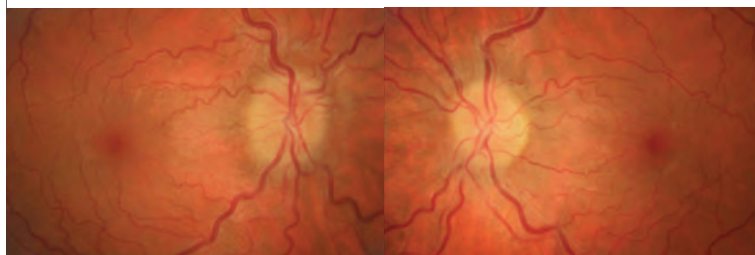
Other treatment options....

- Topiramate – for weight loss
- Surgical – for refractory, non-responsive cases
 - CSF flow diversion/shunting (Venous sinus stenting, VSS)
 - Ventricular-peritoneal shunt
 - Serial lumbar puncture
 - Optic nerve sheath fenestration
- Bariatric surgery
 - Effective, promising but no long term data
 - when BMI is >35
 - Neurology 2021, JAMA 2022*

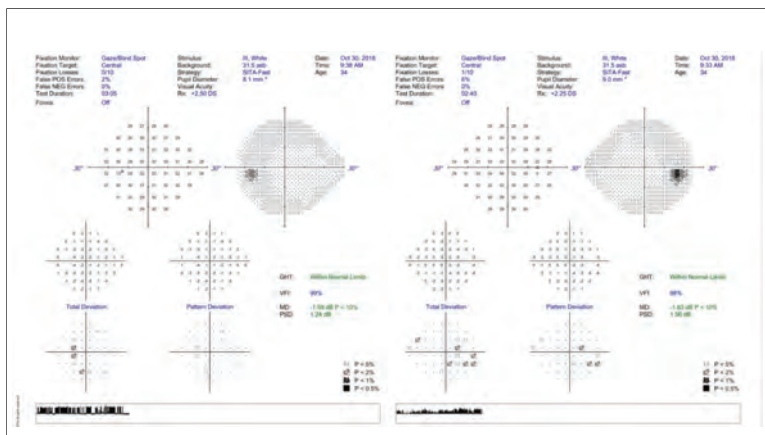
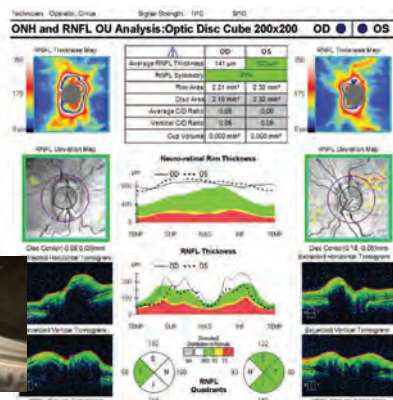
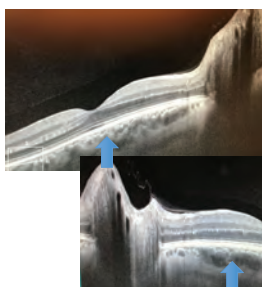


34 year old Caucasian FM

- (+) H/A
- BMI 34
- Meds – Mirena IUD

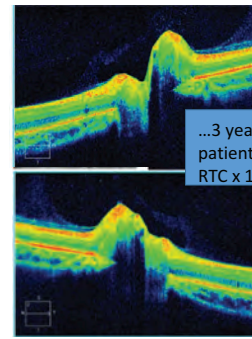
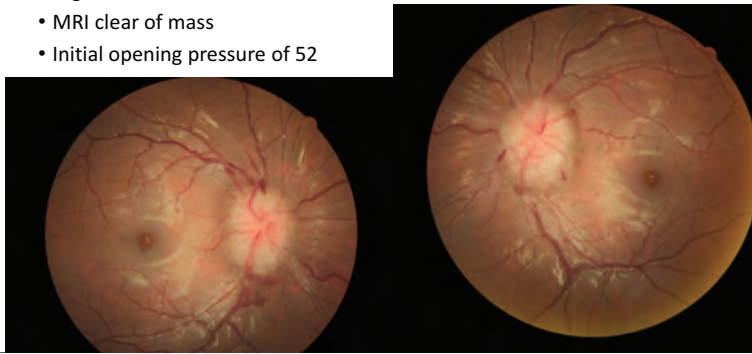


- MRI – empty sella, flattened sclera, distended ONH sheaths

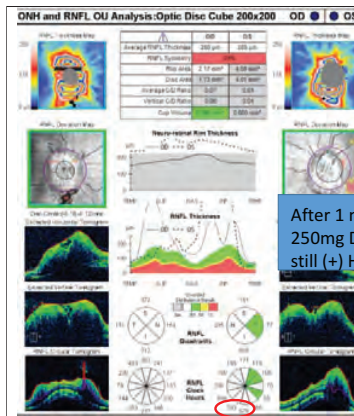
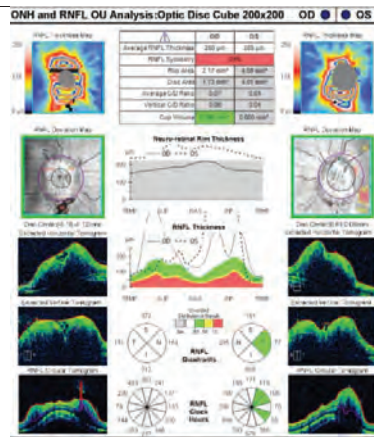
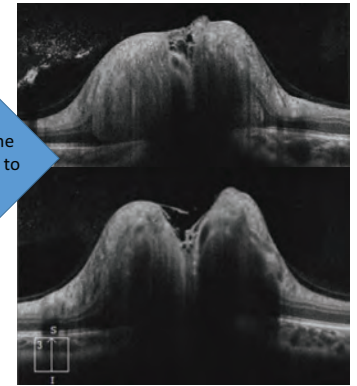


21 AAF

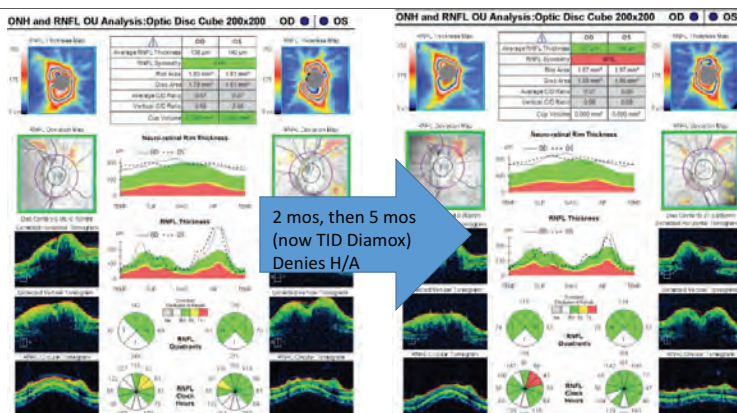
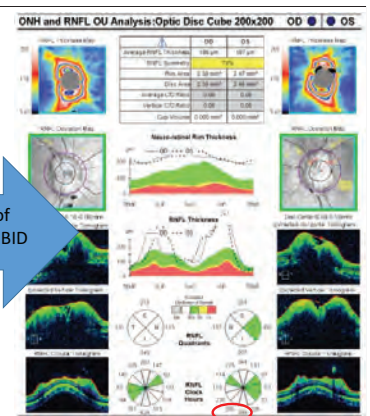
- High BMI
- MRI clear of mass
- Initial opening pressure of 52



...3 years earlier the patient was asked to RTC x 1 week



After 1 month of 250mg Diamox BID still (+) H/A



2 mos, then 5 mos (now TID Diamox) Denies H/A

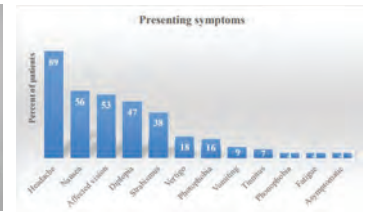
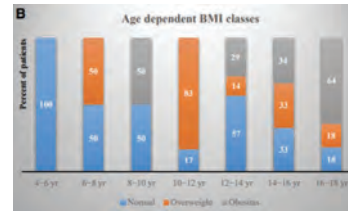
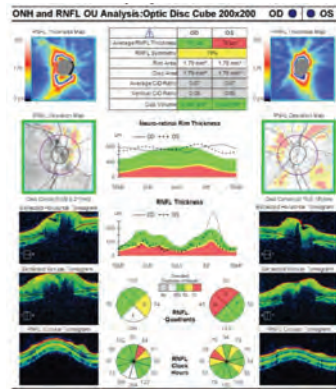
Watching VF carefully...

- H/A's improved
- RNFL thickness reduced
- Total volume reduced



And now...

- Extracellular edema leads to venous stasis...



IHTN in Pediatric Population

- Ehrstedt C, et al. Pediatr Neurol. 2023