Evaluation of Papilledema in the Pediatric Population

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> "…classified as the ostensible elevation of the optic disc secondary to local underlying structural conditions, such as optic disc drusen without edema of the nerve fiber layer…"

Pseudopapilledema

Accurate diagnosis important to avoid anxiety-provoking and expensive testing

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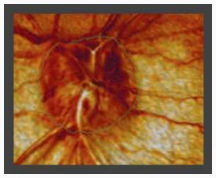
Pseudopapilledema

Optic Disc Drusen (ODD)

- Etiology?
- Genetic predisposition?
- Incidence
  - -Approx 0.4% in children
  - -Up to 2.4% in adults
  - -higher in white pop



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Buried Drusen

- Typically begin as buried deposits
- Migrate toward surface in adulthood
- Appearance mimics papilledema
- Mistaken for true papilledema in up to 76% of cases

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"…optic disc swelling due to elevated intracranial pressure (ICP)"

Papilledema

…can represent a harbinger of vision or life-threatening etiologies…

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Intracranial Hypertension

Idiopathic (IIH)

- Elevated intracranial pressure without clinical, radiologic, or laboratory evidence of a secondary cause
- Little is known about the pathophysiology of idiopathic intracranial hypertension

Secondary (SIH)

- Used for those cases in which an underlying cause is identified
- What is currently termed "idiopathic" may be termed "secondary" as the disease process becomes better understood.

Pseudotumor Cerebri

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Medications associated with elevated ICP

Medications That Can Cause Pseudotumor[5]
Accutane (isotretinoin, Roche)
Oral steroids
Vitamin A
Tetracycline, doxycycline, minocycline
Synthroid (levothyroxine sodium, Abbott)
Growth hormone
Isoniazid
Hormone replacement therapy
Birth control pills
Lithium
Nitroglycerin

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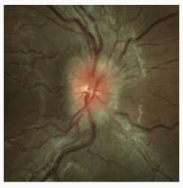
Pediatric Idiopathic Intracranial Hypertension and Weight Gain

- Pre-Pubertal pediatric patients
  - Less association w obesity and gender
  - More likely to be secondary than idiopathic in nature
  - Pathology not fully understood
- Post-Pubertal pediatric patients
  - Demographics and characteristics similar to adults



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"Most of the time, but not always"

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Diagnosis of Pseudopapilledema vs Papilledema

History/Symptoms

Pseudopapilledema	Papilledema
• More common in the absence of symptoms	• Asymptomatic?
• Incidence and type varies	• Symptoms of elevated intracranial pressure
Headaches?	Headaches
Transient visual obscurations	Nausea/vomiting
	Pulsatile tinnitus
	Binocular horiz diplopia (6th n palsies)
	Transient visual obscurations
	• Weight gain history and obesity
	• Medication

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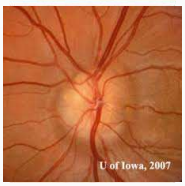
Diagnosis of Pseudopapilledema

Clinical presentation

- Visual acuity/Color vision/Pupils
- Cupless, elevated optic disc
- Visible vessels in peripapillary NFL
- Peripapillary hemorrhages
- Spontaneous venous pulsation
- Surface refractile ODD



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Diagnosis of Papilledema

Clinical presentation

- Visual acuity/Color vision/Pupils
- Disc appearance and presence of cup
- Vascular changes
  - Venous engorgement
  - Capillary leakage and hemorrhage
  - Peripapillary lipid and cotton-wool spots
- Blurring of disc margin/retinal NFL

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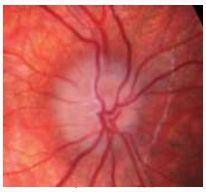
Diagnosis of Pseudopapilledema

Ancillary testing

- Visual Field Defects
  - Increased frequency w age
  - Types
- Fundus Autofluorescence (FAF)
- B-Scan Ultrasonography
- Optical Coherence Tomography (OCT)
  - RNFL, NFL defects
- Orbital Imaging



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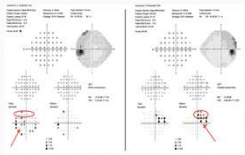
Diagnosis of Papilledema

Ancillary testing

- Visual Fields
- Fundus Autofluorescence (FAF)
- B-Scan Ultrasonography
- Optical Coherence Tomography (OCT)
- CT scan
- Magnetic Resonance Imaging MRI/MRV
- Lumbar Puncture (LP)
  - With opening pressure/CSF analysis



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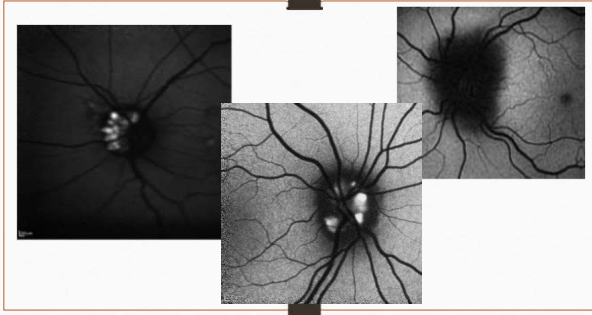

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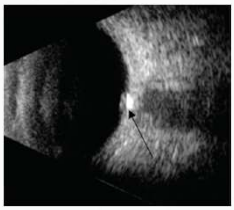

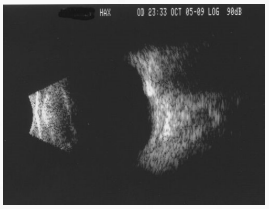

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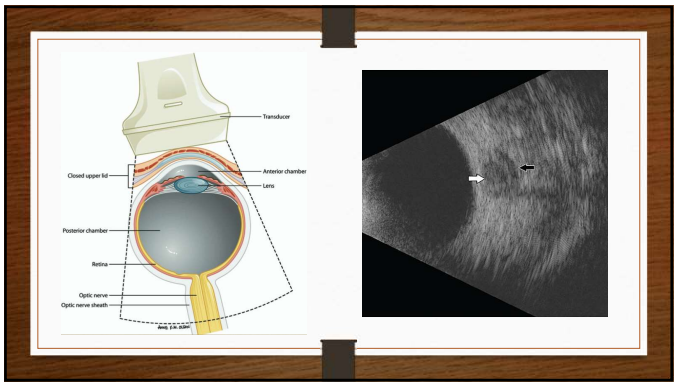

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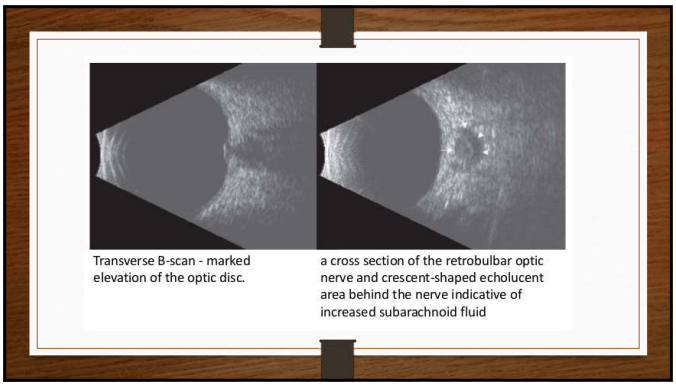

Transverse B-scan - marked elevation of the optic disc.

a cross section of the retrobulbar optic nerve and crescent-shaped echolucent area behind the nerve indicative of increased subarachnoid fluid

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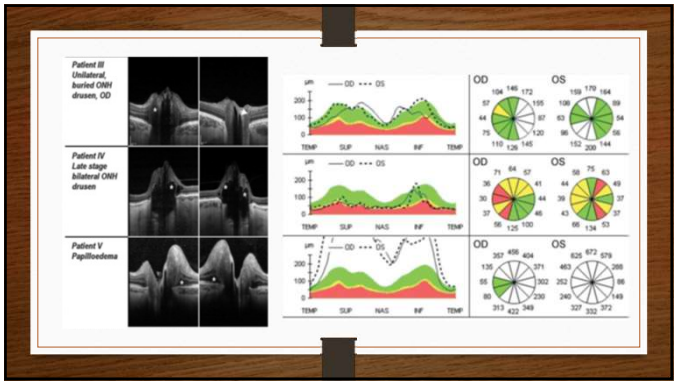

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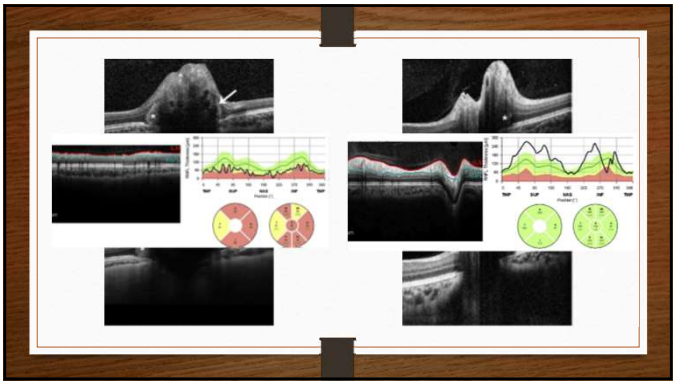

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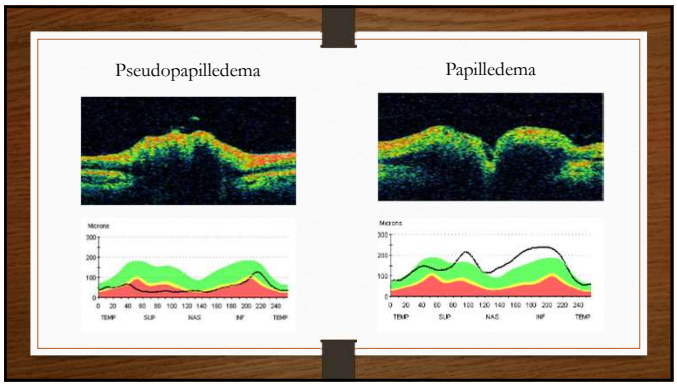

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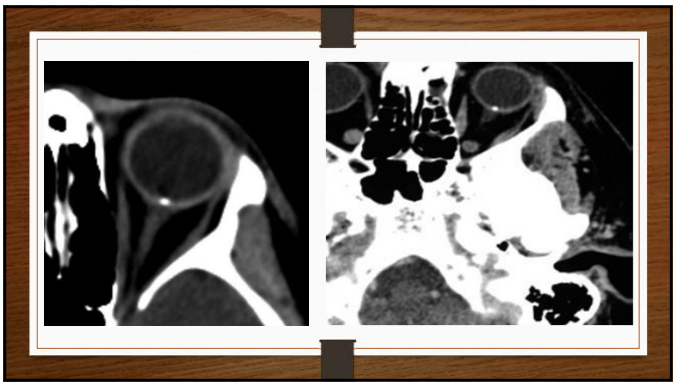

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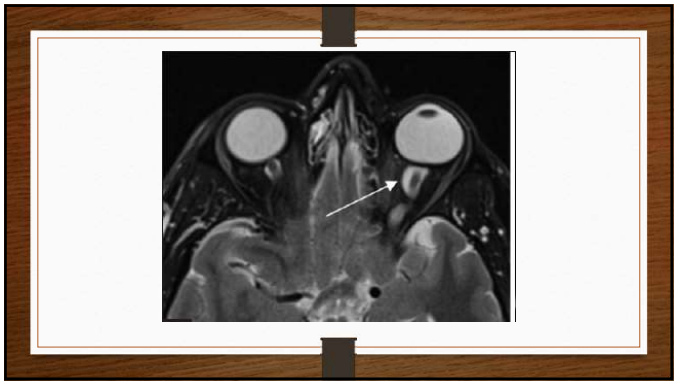

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Optic Disc Drusen

Complications and Management

Complications

- Rare
- Vision loss/VF defects
- Vascular

Management

- Annual exam/VFs
- Ocular hypotensive drops
- Neuroprotective agents?
- Serial OCT to confirm if suspicious for Papilledema

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Treatment of Pediatric Idiopathic Intracranial Hypertension

Based on severity of presentation/Goals

Treatment Goals: alleviate symptoms and prevention of permanent vision loss

- Steroids (oral or IV)
- **Acetylzolamide** (oral or IV)
- Weight loss (post pubertal)
- Mgmt of secondary/predisposing factors
- Surgical: Optic nerve sheath fenestration
  CSF shunt
- Serial testing: Visual acuity, Perimetry, ONH appearance

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CASES

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Case #1: 15yo IF

Routine exam
Cc: mild distance blur ou
BCVA: 20/20 OU
IOP: 19/19
Pupils: PERRLA –APD
C/D: 0.1/0.1 OU
Fundus: Elevated disc margins OS

Ancillary testing?

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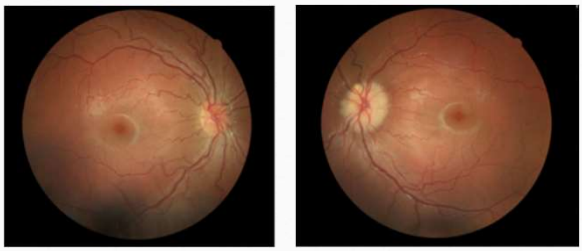

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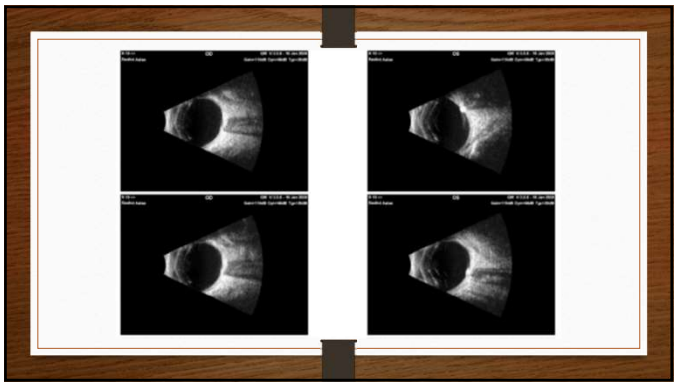

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Case #1: 15yo IF

Diagnosis: Pseudopapilledema OS
Secondary to Optic Disc Drusen

Mgmt: 1. Education
2. Baseline visual acuity
perimetry
fundus photography/FAF

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Case #2: 14yo WF (Pearle Vision)

Cc: Reports headache behind eyes 1-2 weeks, worsening over last week with increasing blurry vision
Reports feels eyes crossing. Horizontal double vision worse on left gaze, onset last 3 days, increasing frequency. Neg hx cold, sickness, or trauma

Medical Hx: OTC Allergy med, Minocycline 100mg/day (acne)for past 2 mos

Entering VA: 20/30+ OD 20/20- OS
Pupils/Color: WNL OU
EOMs: Left ESO in primary position
Left abduction deficit
IOP: 18/21
Fundus: Disc edema OU

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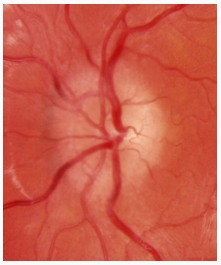

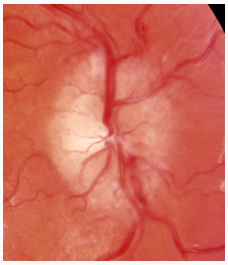

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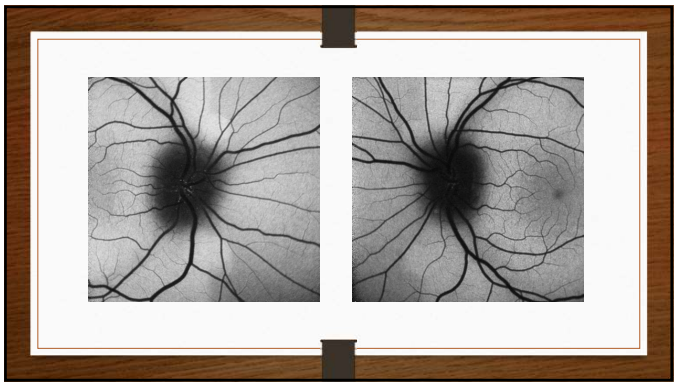

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Case #2: 15yo WF

Diagnosis: Presumed Bilateral Papilledema
Symptoms consistent w elevated ICP
Left CN VI Palsy

Mgmt: Urgent referral CCH for MRI to r/o space occupying lesion

Cinti Children's Hosp: MRI neg for intracranial mass
Diff diagnosis for Minocycline induced increased ICP
Pending results Lumbar Puncture, initiate Diamox
Stop Minocycline!

Prognosis: Good. Vision and color intact, minimal scattered peripapillary VF defects

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Case #3: 17yo WF

Cc: Patient presents for REE. Reports on/off HA's past 6-12 mos increasing w chores. Recently reports blurred vision ou and occasional nausea. Neg for trauma, DV, or ear ringing. Went to local OD earlier this year and got new glasses.
Hx: patient is 5'3" 178lbs, reports weight gain ~40lbs over 1 ½ yrs

BCVA: 20/20 ou
Color plates: 14/14
EOM's: full in all positions
Pupils: PERRLA –apd ou
IOP: 14/13
C/D: 0.1/0.1 Fundus: Elevated, blurred disc margins ou

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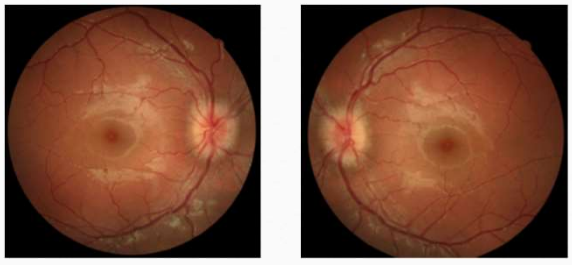

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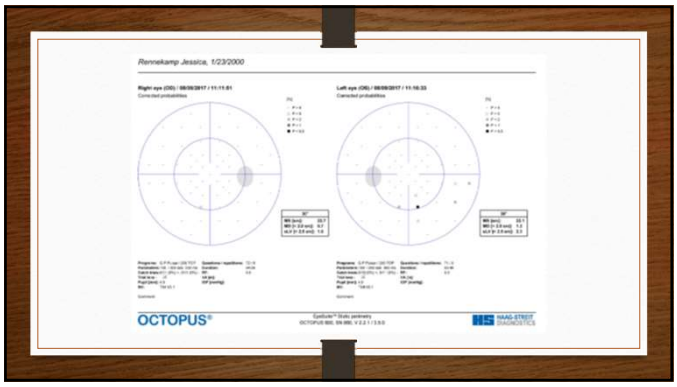

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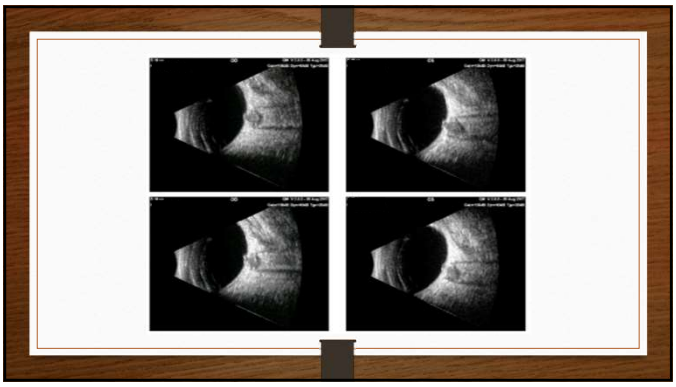

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Case #3: 17 WF

Diagnosis: Papilledema OU
Presentation consistent for IIH sec to weight gain, young female

Mgmt: MRI w and w/o contrast r/o intracranial mass, hydrocephalus

Results: MRI neg for intracranial pathology
Diagnosis IIH
Neuro referral for medical mgmt.
Initiated oral Diamox 500mg/day

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Case #4: 17yo WF* (Pearle Vision)

Cc: Reports "black spots" in peripheral vision OS>OD onset 4 weeks
Hx: Previous exam name noted as "Olivia". Now goes by "Lucas"
Further discussion reveals patient taking testosterone and minocycline
Going through gender reassignment

Evaluation: BCVA 20/30 OD 20/20-OS
Color 14/14 OU
Pupils: PERRLA –apd OU
IOP: 15/11
C/D: 0.25/0.25 OU
Fundus: Disc edema OU

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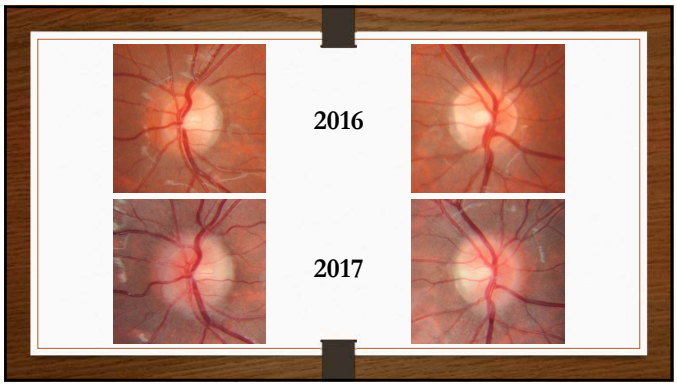

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Case #4: 17yoWF*

Diagnosis: Papilledema OU

Mgmt: Immediate neuro consult
MRI to r/o intracranial mass/pathology
Presentation and history consistent w Minocycline or Hormonal therapy induced elevated ICP

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Case #5: 11yo AWF

10/15/19
Cc: REE, gradual blur dist ou. No other oc/vis complaints

BCVA: 20/20 OU, hi myope, increase -050/-075 correction
EOMs: full in all positions
PERRLA –apd ou
IOP: 18/18
C/D: 0.15/0.25
Fundus: Blurred disc margins OU, nasal>temp, peripapillary heme OS?

Further Hx: Patient is 5'4" 108lbs, neg for HA's, nausea, trauma, ear ringing, weight gain, prepubertal. Mother has hx migraines, but patient has non-specific, non-positional, occasional mild HA's

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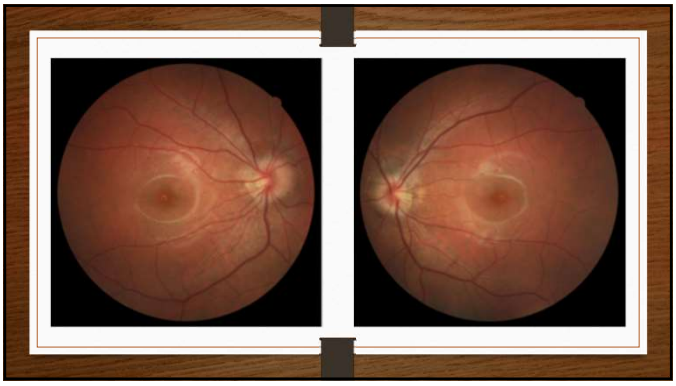

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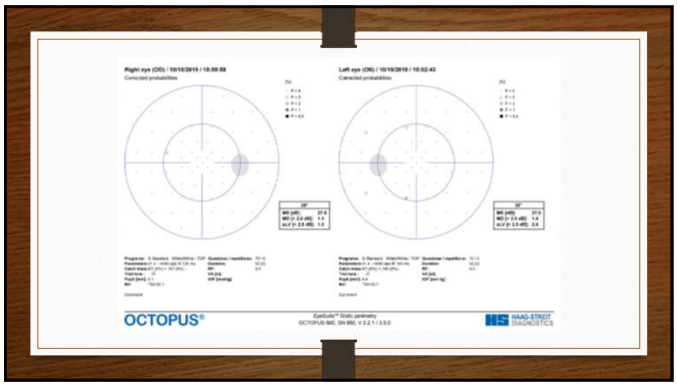

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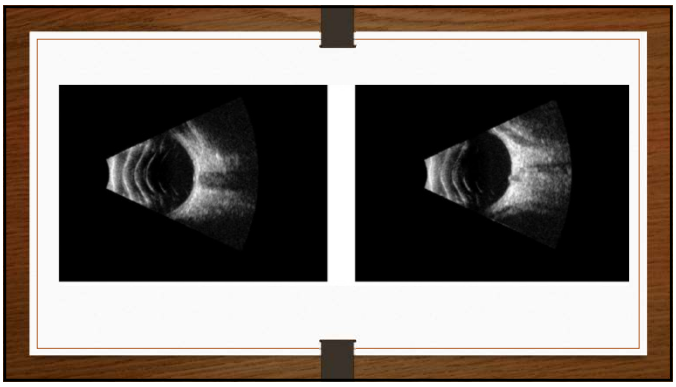

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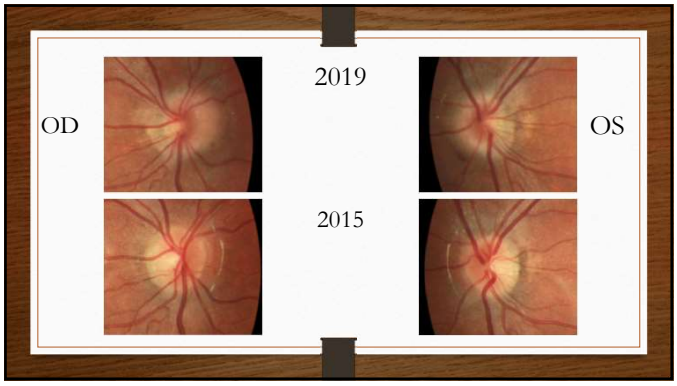

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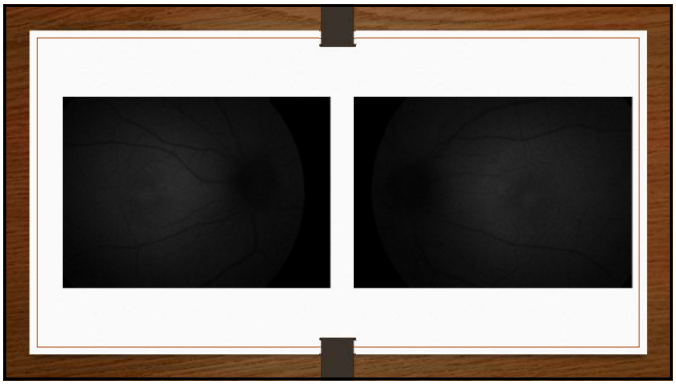

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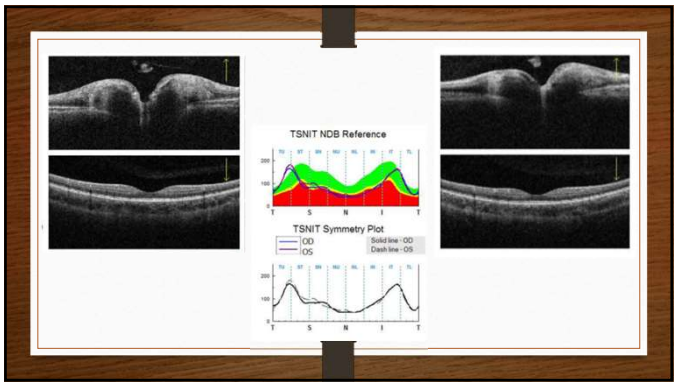

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Case #5: 11AWF

Diagnosis: Pseudopapilledema vs Papilledema OU
- Symptoms of HA's vague and non-diagnostic
- Lack of additional signs/symp related to elevated ICP
- Notable disc edema change from 2015 photo
- Inconclusive B-Scan to confirm Optic Disc Drusen

Mgmt: Consult Cincinnati Children's Neuro
- MRI to r/o intracranial pathology

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Initial Visit CCH Neuro 10/21/19

ASSESSMENT:

[redacted] is a 11 y.o. female found to have papilledema on a routine eye exam. Imaging found a pineal gland cyst. She present to Neurology for both findings. I am not certain the two findings are related. Given her lack of risk factors (no OCPs, no acne antibiotics, no head injury, no cerebral sinus venous thrombi, and she is not obese) for papilledema I think that an US to evaluate for the findings being drusen is warranted. I do not feel the cyst is large enough to cause such eye findings. The exception would be unless the cyst appeared very rapidly which we will only know after we get a short term repeat MRI. The MRI will be with gadolinium.

I discussed that for the papilledema the US will help determine if she has drusen or not. If it is determined that she clearly has drusen then she will get no further work up for papilledema and will only need a repeat MRI. If the US confirms papilledema or is inconclusive then she will need to get an LP with opening pressure to determine if treatment is needed.

It was also mentioned Valarie gets HAs. Currently they are not positional, they are infrequent and they are not severe. They meet criteria for probable migraines without aura, Her mother has migraines so I suspect she is just developing migraines as she gets closer to puberty.

PLAN: Ultrasound to r/o buried drusen, monitor HA's/symptoms, repeat MRI 3 mos

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2nd Visit CCH Ophthalmology 11/20/19

Dr. Connors' B-scan Interpretation and Report:
B-scan of both eyes complete today due to optic disc elevation.
Areas of superficial as well as buried optic disc drusen are present at both optic discs.

Dr. Connors' Octopus 32-TOP Visual Field Interpretation and Report:
Octopus 32-TOP Visual field exam completed today for both eyes due to optic disc elevation.
Right eye: FP errors: 14%, FN errors: 0%, MD: -0.4, within normal limits
Left eye: FP errors: 57%, FN errors: 0%, MD: -0.9, within normal limits

Dr. Connors' SD-OCT RNFL Interpretation and Report:
SD-OCT RNFL completed today for both eyes due to optic disc elevation.
Right eye: Average peripapillary RNFL thickness: 92 microns
Left eye: Average peripapillary RNFL thickness: 90 microns

PLAN:

[redacted] has 2+ elevation of both optic nerves. There is some lumpy-bumpy appearance as well as some smooth elevation. Confirmed superficial and buried optic disc drusen on B-scan today (see images above). Visual fields and SD-OCT RNFL do not support papilledema. MRI brain on 10/17/19 with pineal region cyst, otherwise within normal limits. No MRV was performed. Obtaining a MRV should be a consideration if all other testing is within normal limits. Recommend continuing with Neurology's recommendation for lumbar puncture with opening pressure on 11/22/19 due to some testing supporting optic disc drusen (B-scan, visual fields, and OCT RNFL), but other testing (Optos photos, clinical fundus exam) and symptoms (headaches getting worse) supporting possible papilledema.

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Initial visit CEI NeuroOPH 12/31/19

Cc: No other complaints other than HA's but thought to be "normal" HA's
Reports has stopped Diamox couple weeks due to intolerance/allergy

Evaluation: Nasal elevation of discs OU with otherwise normal vision and examination

Diagnosis: Psuedopapilledema OU

Plan: Has had changes documented in disc but by report ultrasound at CCHMC shows buried drusen. I am obtaining their records. Has had 3 VFs - all normal and no HAs c/w increased ICP despite LP showing OP of 28. Recent article shows normal OP in kids up to 28 however. Will review ultrasound from CCHMC, follow her VFs off meds as she had allergic reaction to the Diamox.

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2nd visit CEI NeuroOPHTH 02/26/20

Cc: Patient reports vision stable, no additional symptoms noted from previous visit

Evaluation: Nasal disc elevation OU, Stable vision and 24-2 visual fields w normal fixation losses
Stable RNFL with serial OCT

Diagnosis: Pseudopapilledema of the optic discs OU
Buried disc drusen OU

Mgmt: Reassurance
Monitor symptoms and VA
Serial visual fields, OCT and photos 6-12 mos

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Case #6 20yo WF

- CC: Blurry vision, occasional HA's?? Poor historian responses inconsistent
- Hx: +obesity, >50+lb weight gain <1yr
- BCVA: OD +175-100x180 20/25-
  OS +225-150x170 20/30+

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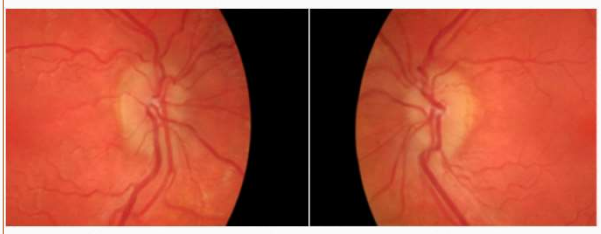

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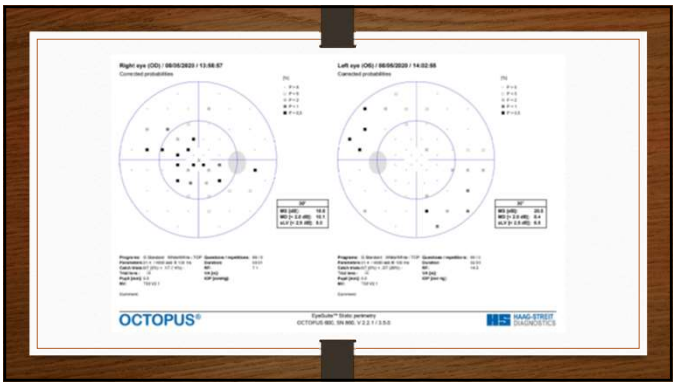
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HAAG-STREIT DIAGNOSTICS

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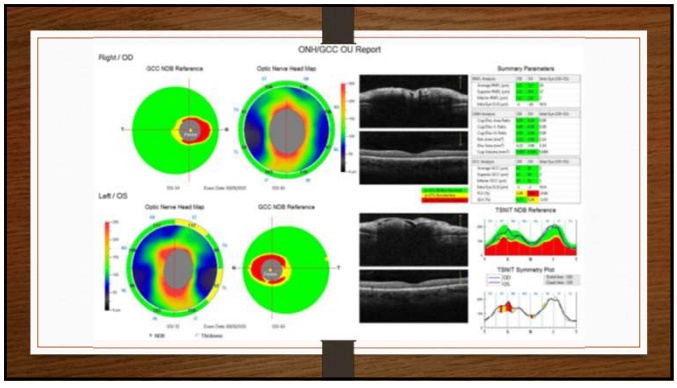
ONH/GCC OU Report
Right / OD
Left / OS
Summary Parameters
TSNIT NDB Reference
TSNIT Symmetry Plot

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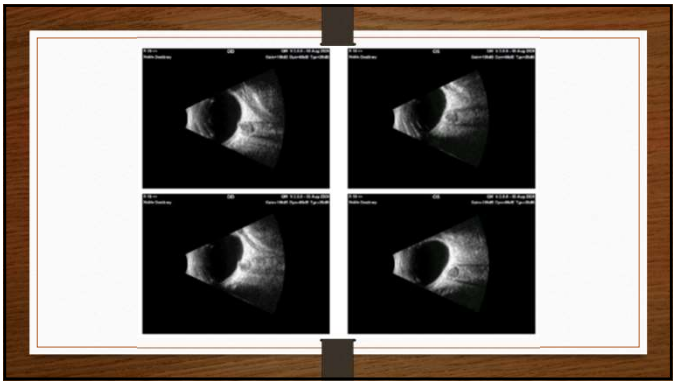

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Case #6 21yo WF

- Diagnosis: Probable Idiopathic Intracranial Hypertension
  Consistent profile and symptoms? Decreased vision?
  (+)Crescent sign B-Scan Ultrasound
- Plan: Immediate MRI imaging to r/o intracranial lesion
  Neuro consultation
  If MRI negative, initiate weight loss and Acetylzolamide

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Summary

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