

Evaluation of Papilledema in the Pediatric Population

Todd A. Zelczak, OD, FAAO

Midwest Optometric Society

“

...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...”

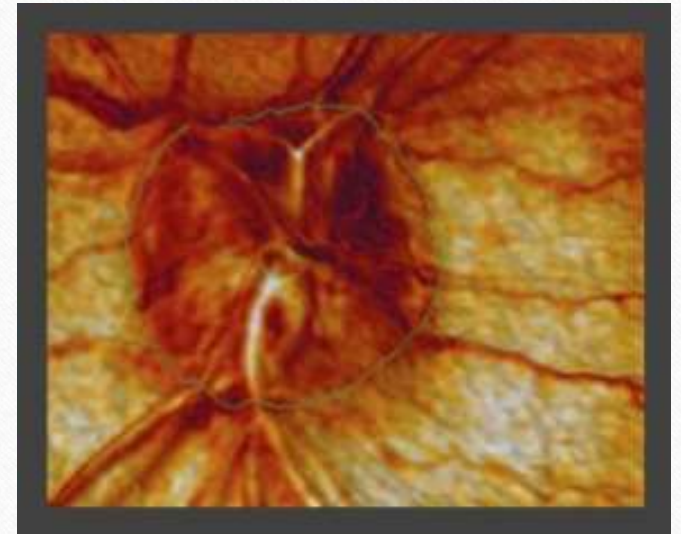
Pseudopapilledema

Accurate diagnosis important to avoid anxiety-provoking and expensive testing

Pseudopapilledema

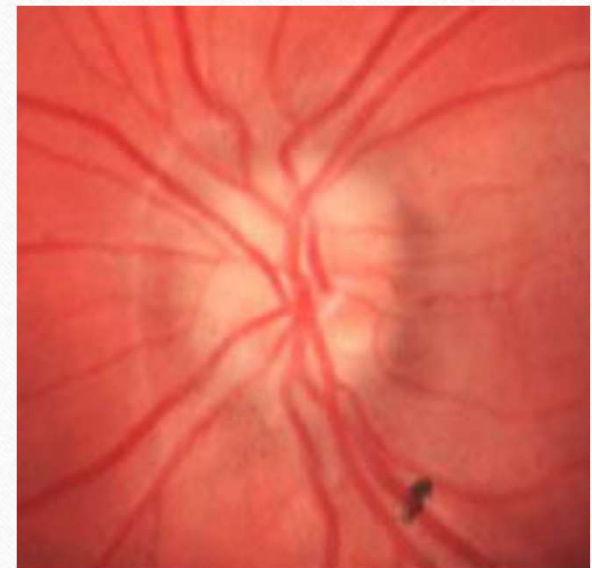
Optic Disc Drusen (ODD)

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



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



“

...optic disc swelling due to elevated
intracranial pressure (ICP)

”

Papilledema

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

Intracranial Hypertension

meningitis serosa, pseudotumor cerebri, benign intracranial hypertension, idiopathic intracranial hypertension, pseudotumor cerebri syndrome

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.

Medications
associated with
elevated ICP

Medications That Can Cause Pseudotumor⁵

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

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



Diagnosis of Pseudopapilledema vs Papilledema

History/Symptoms

Pseudopapilledema

- More common in the absence of symptoms
- Incidence and type varies
 - Headaches?
 - Transient visual obscurations

Papilledema

- Asymptomatic?
- Symptoms of elevated intracranial pressure
 - Headaches
 - Nausea/vomiting
 - Pulsatile tinnitus
 - Binocular horis diplopia (6th n palsies)
 - Transient visual obscurations
- Weight gain history and obesity
- Medication

Diagnosis of Pseudopapilledema

Clinical presentation

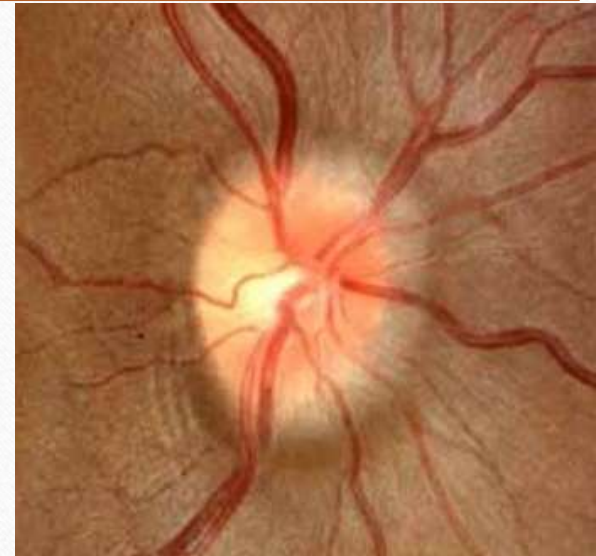
- Visual acuity/Color vision/Pupils
- Cupless, elevated optic disc
- Visible vessels in peripapillary NFL
- Peripapillary hemorrhages
- Spontaneous venous pulsation
- Surface refractile ODD



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



Diagnosis of Pseudopapilledema

Ancillary testing

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



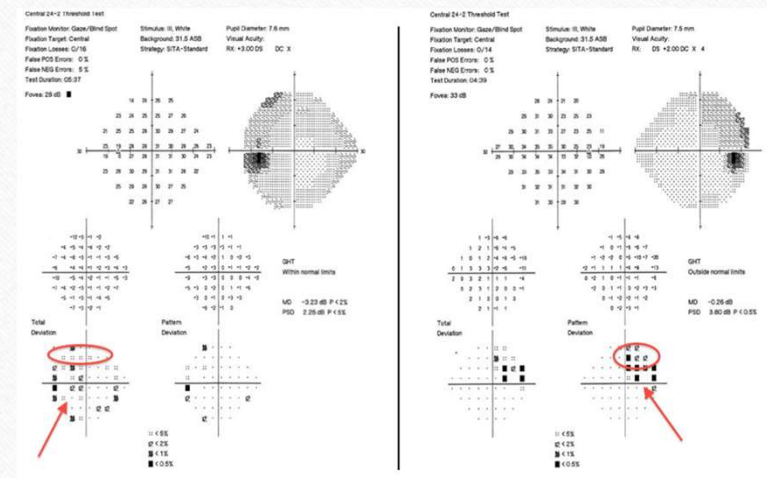
Diagnosis of Papilledema

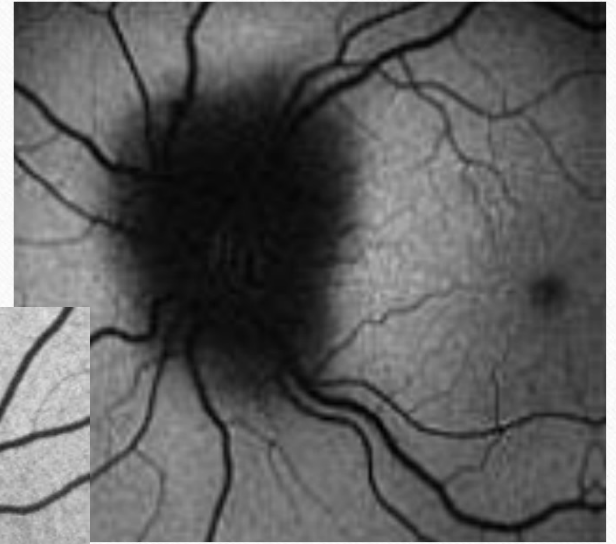
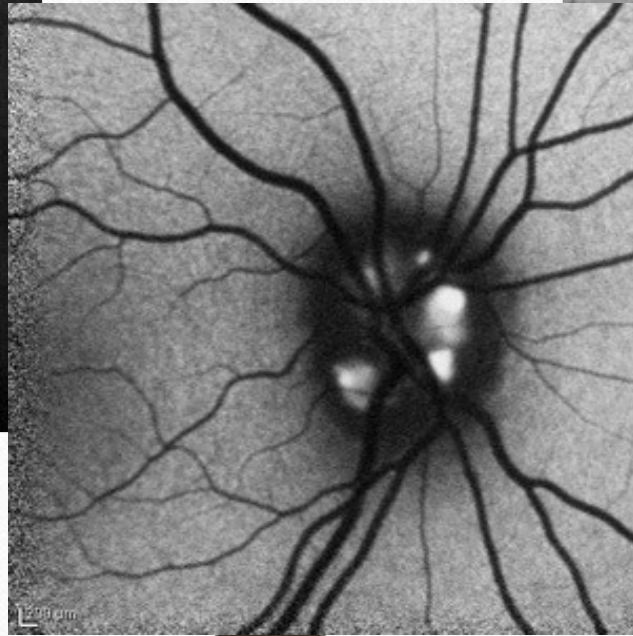
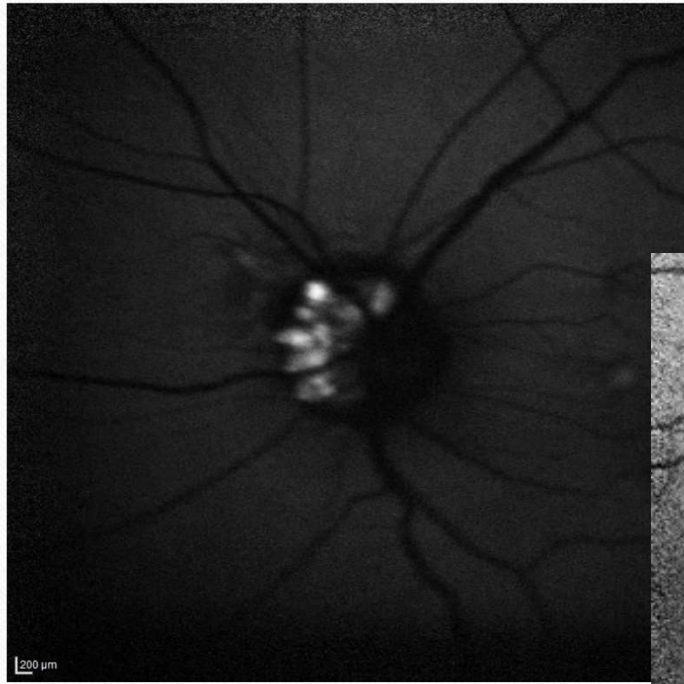
Ancillary testing

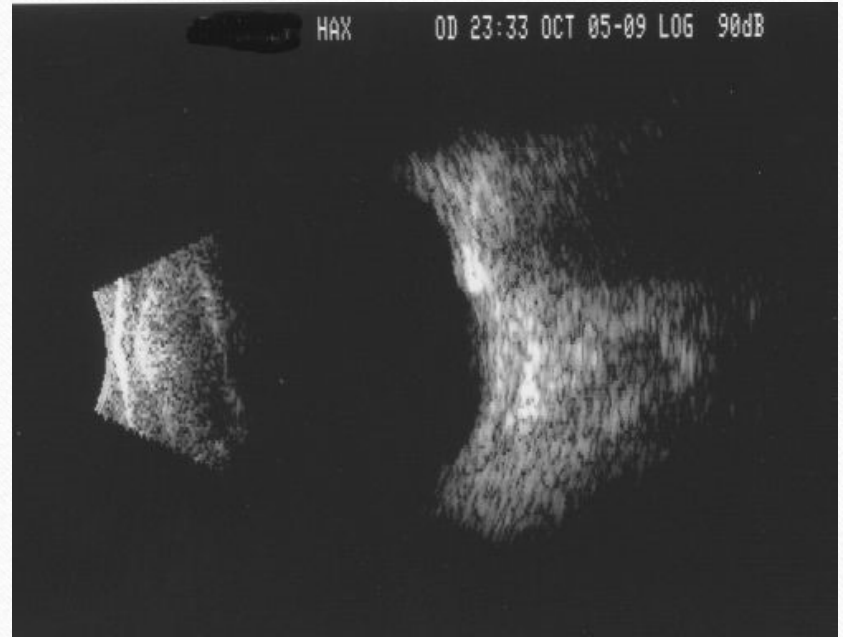
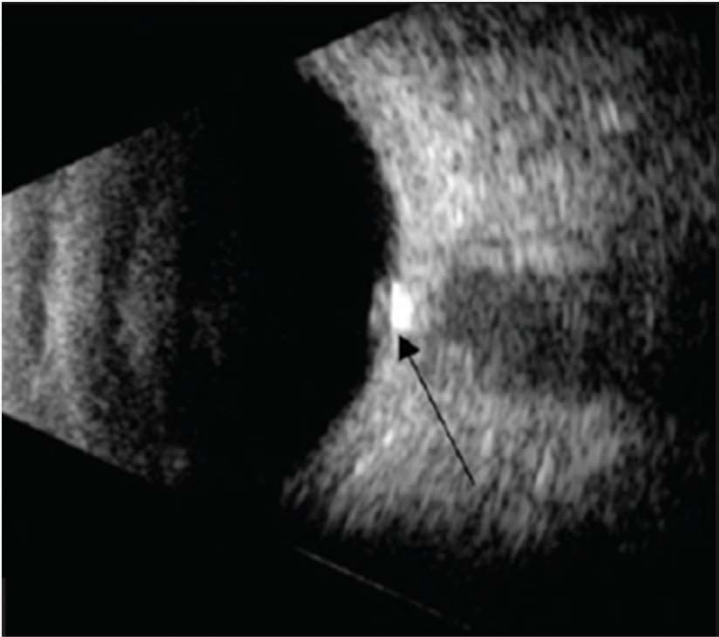
- Visual Fields
- B-Scan Ultrasonography
- Optical Coherence Tomography (OCT)
- CT scan
- Magnetic Resonance Imaging MRI/MRV
- Lumbar Puncture (LP)

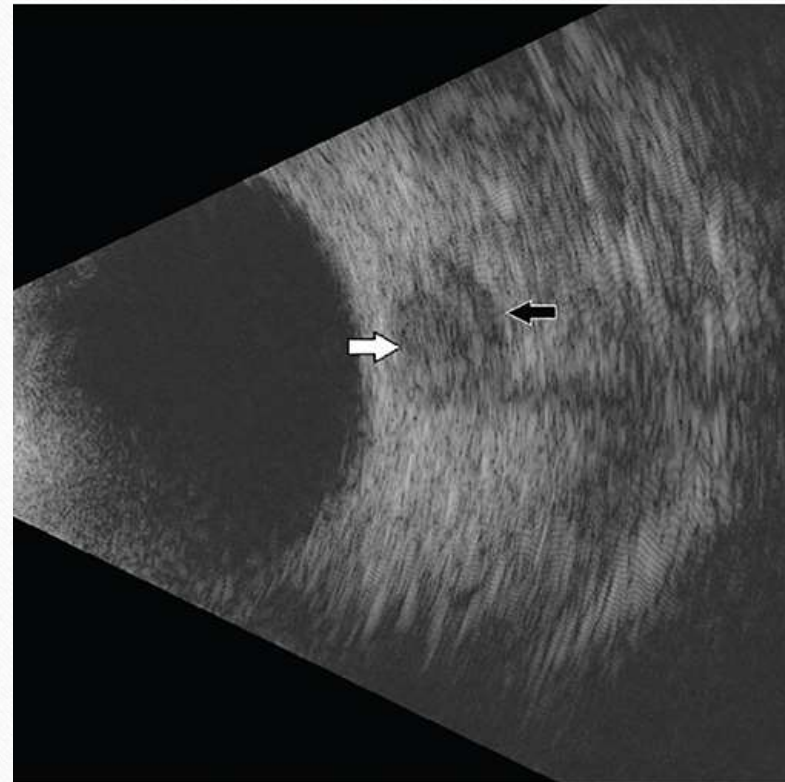
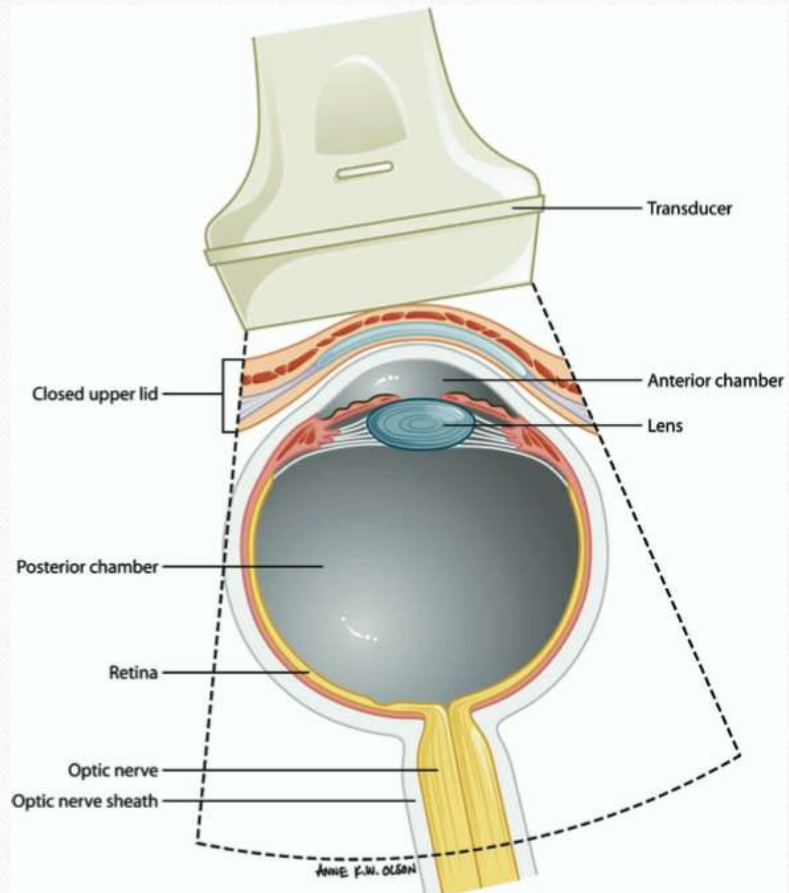
With opening pressure
CSF analysis

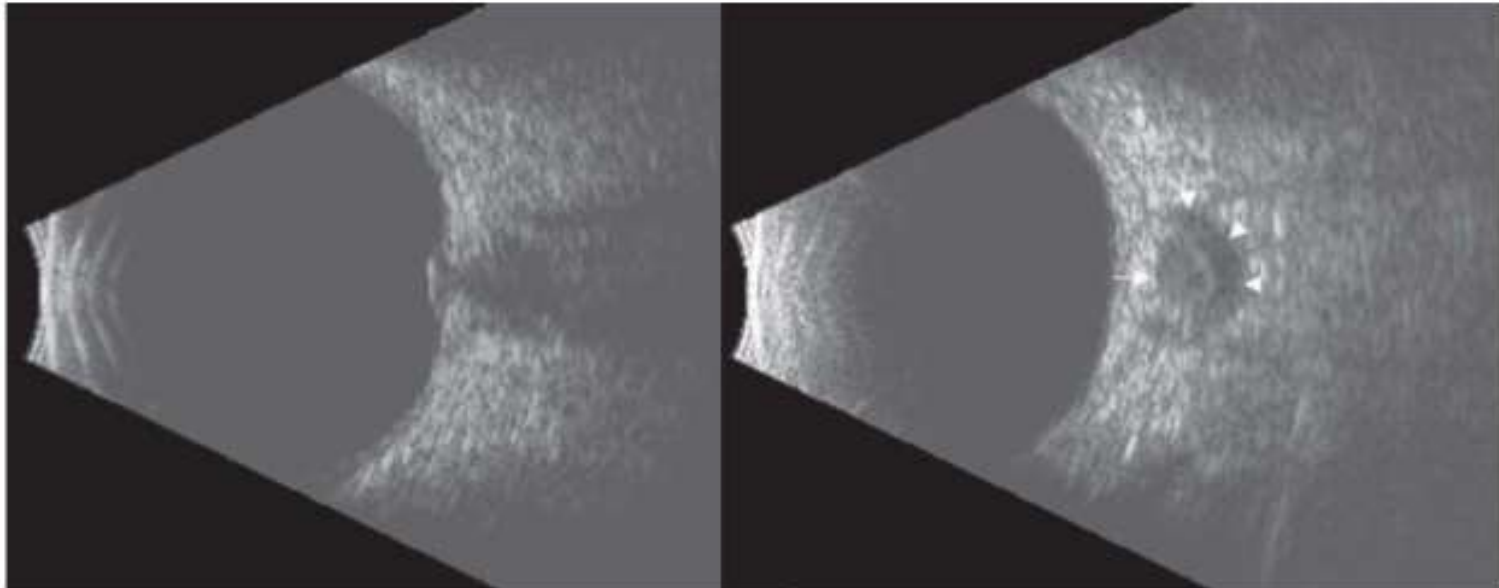
*Other testing to rule out secondary causes should be guided by the history and physical findings.







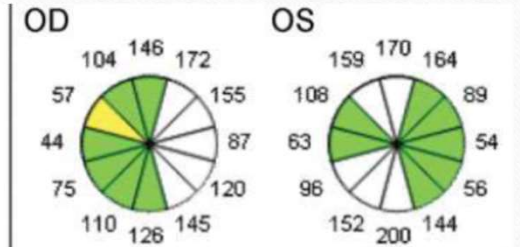
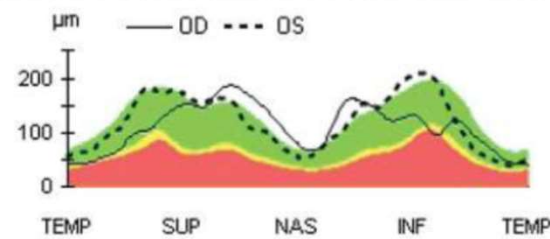
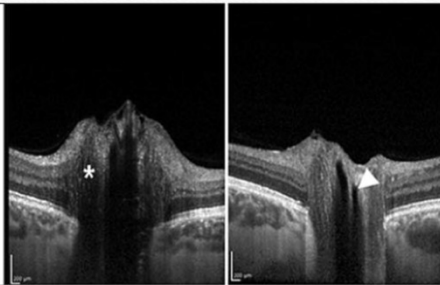




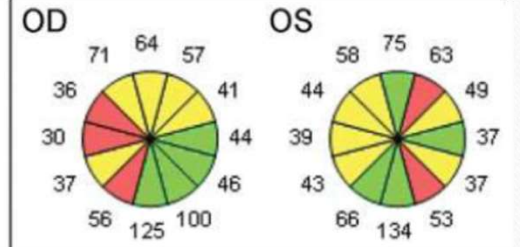
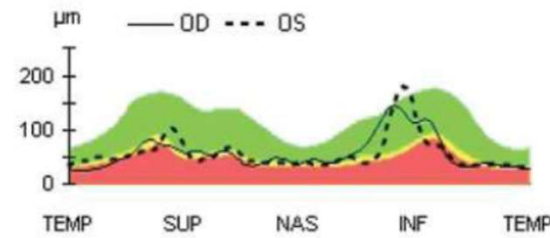
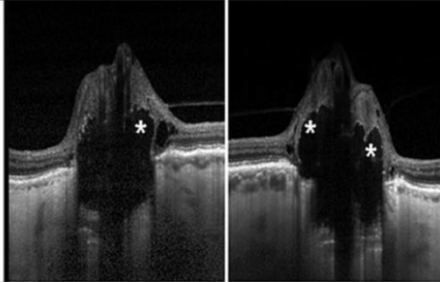
Transverse B-scan - marked elevation of the optic disc.

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

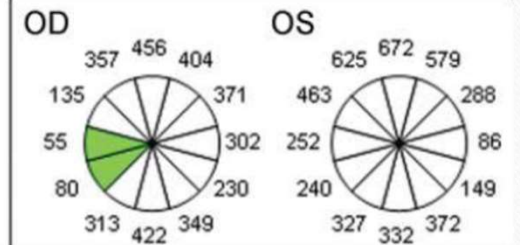
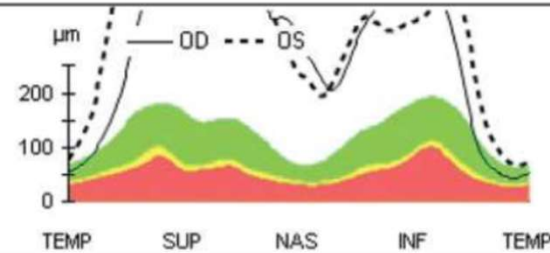
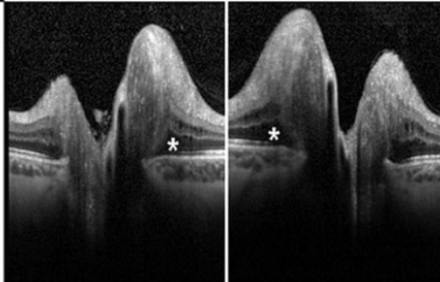
Patient III
Unilateral,
buried ONH
drusen, OD

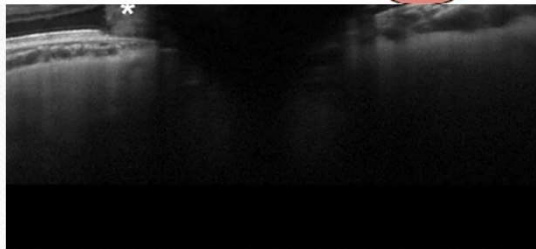
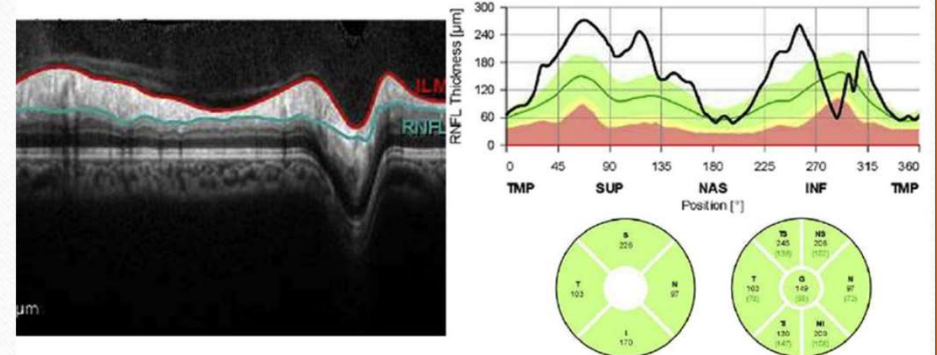
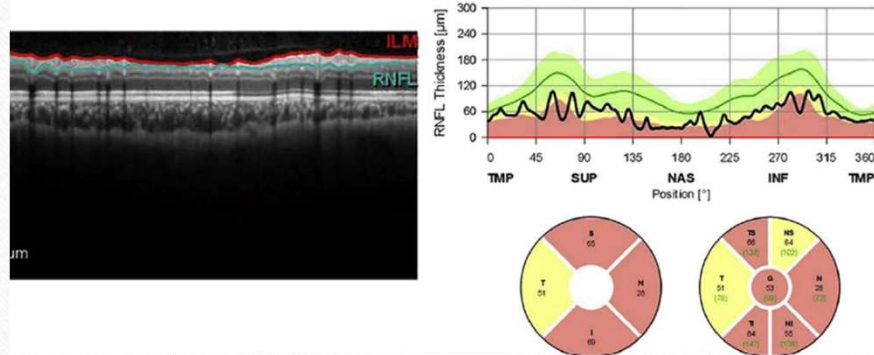
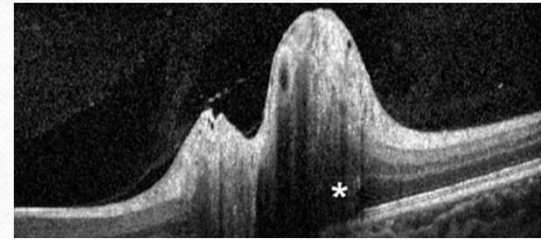
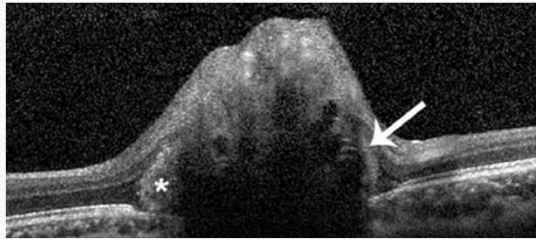


Patient IV
Late stage
bilateral ONH
drusen

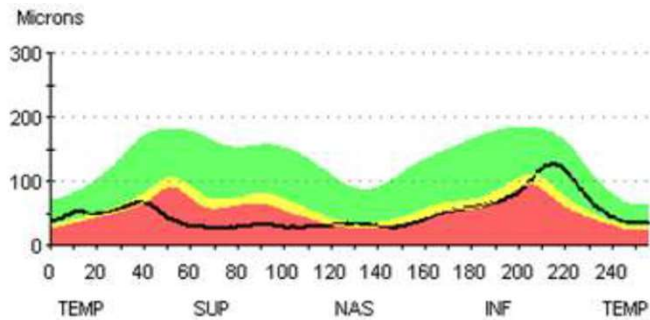
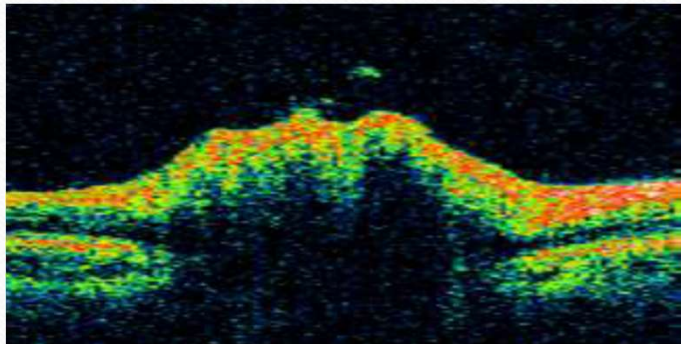


Patient V
Papilloedema

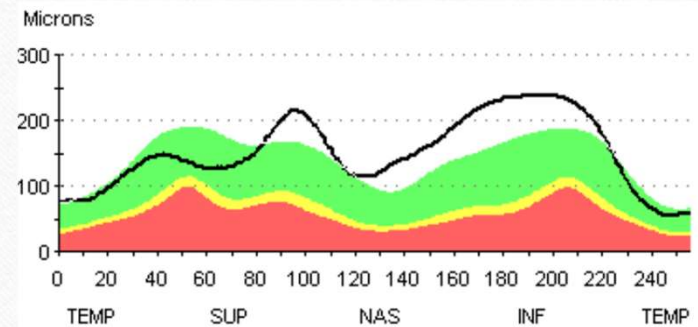
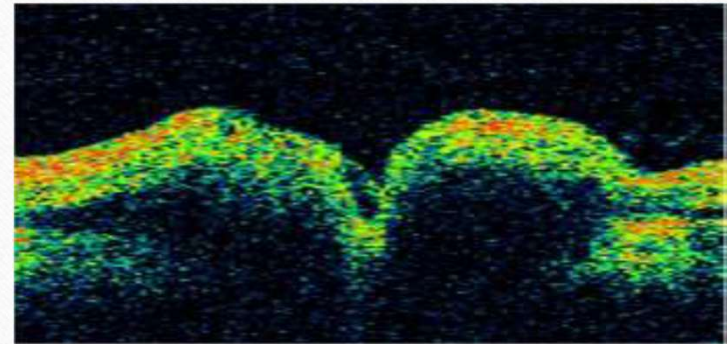


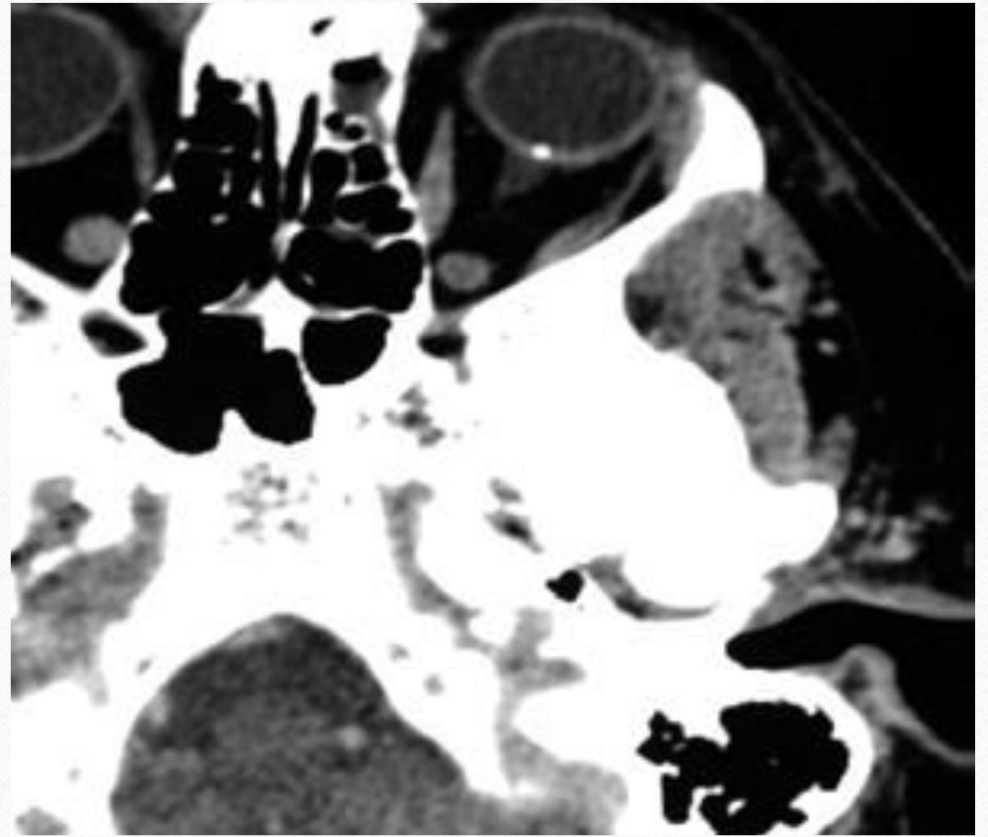
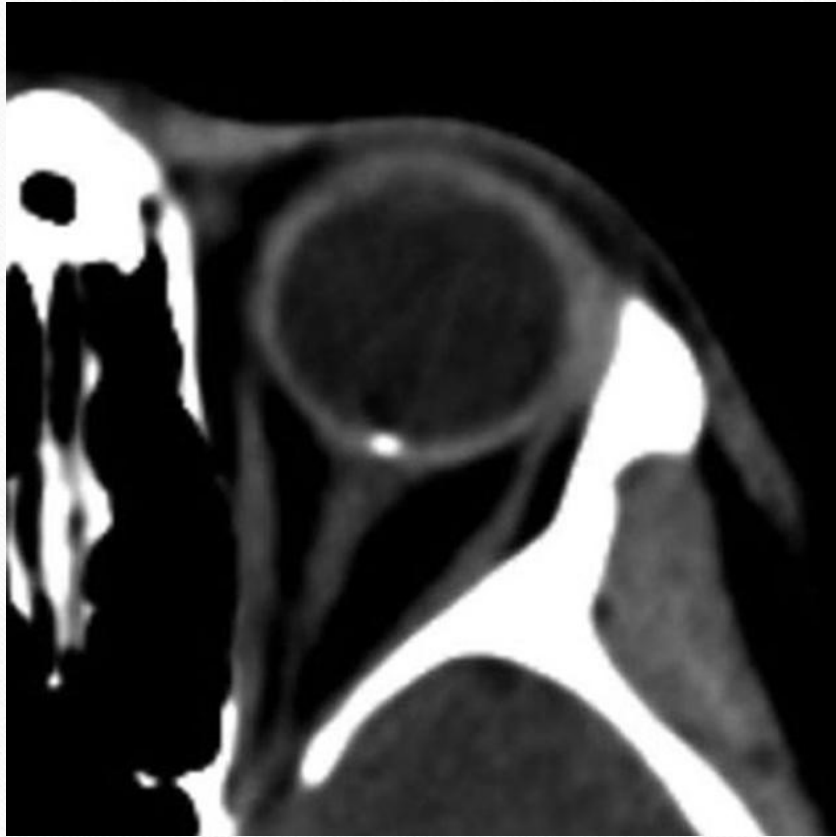


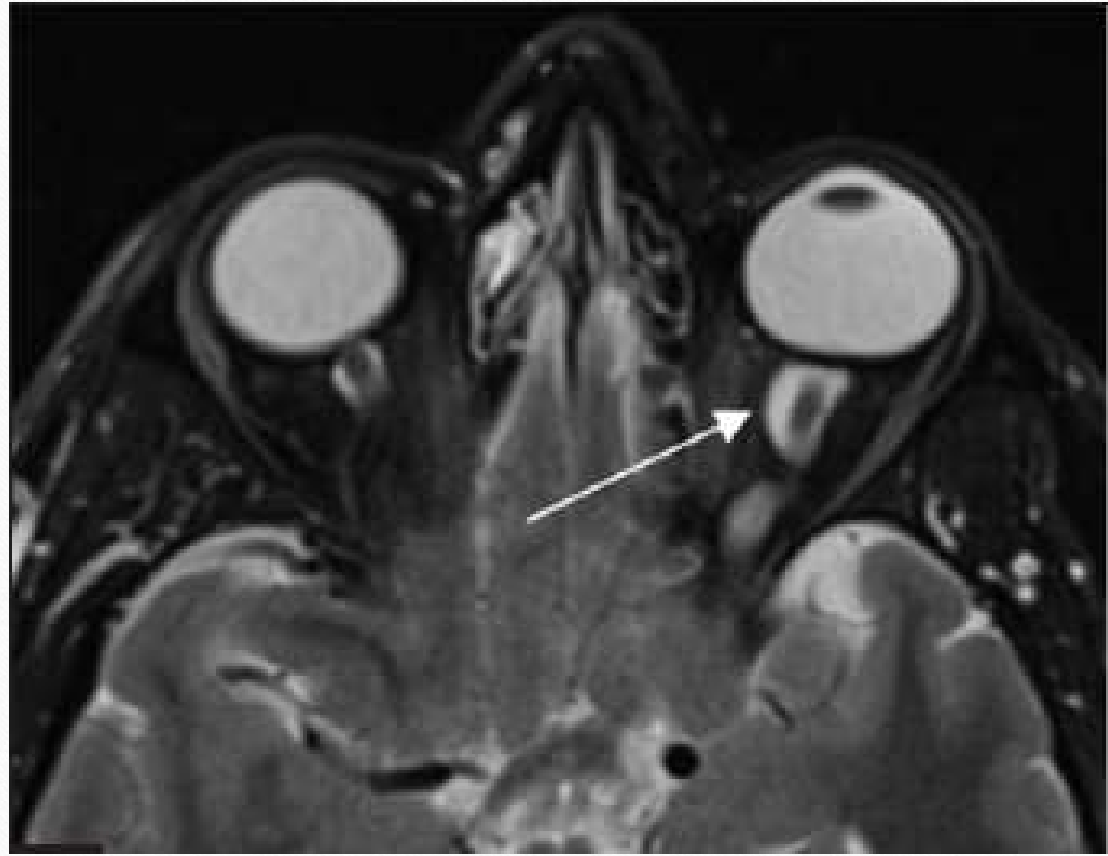
Pseudopapilledema



Papilledema







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

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



CASES

Case #1: 15yo IF

Routine exam

Cc: mild distance blur ou

BCVA: 20/20 OU

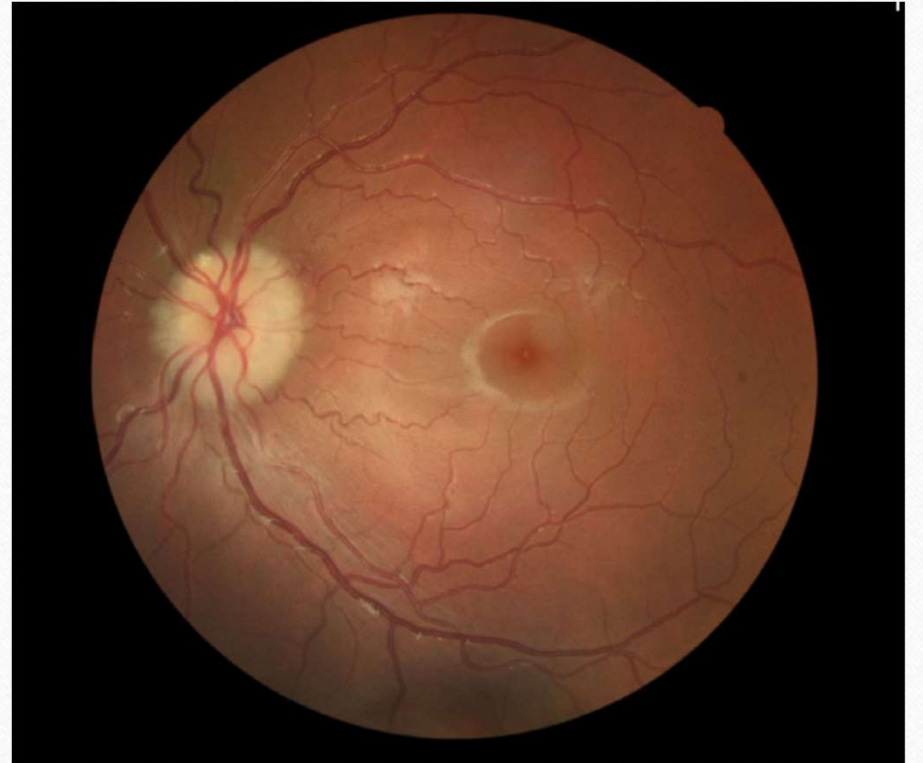
IOP: 19/19

Pupils: PERRLA –APD

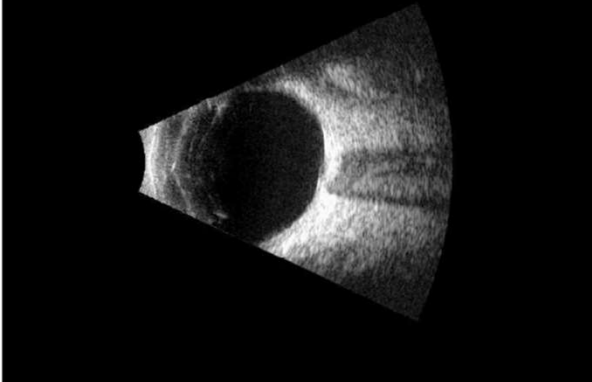
C/D: 0.1/0.1 OU

Fundus: Elevated disc margins OS

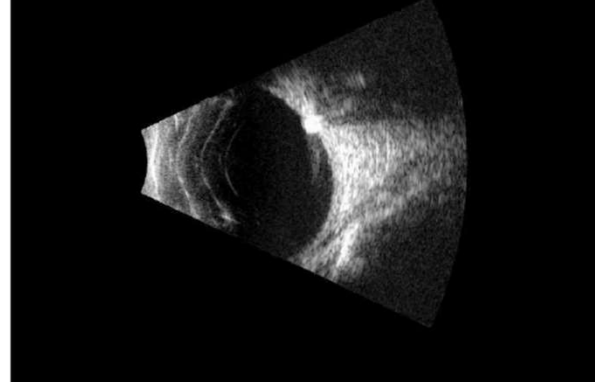
Ancillary testing?



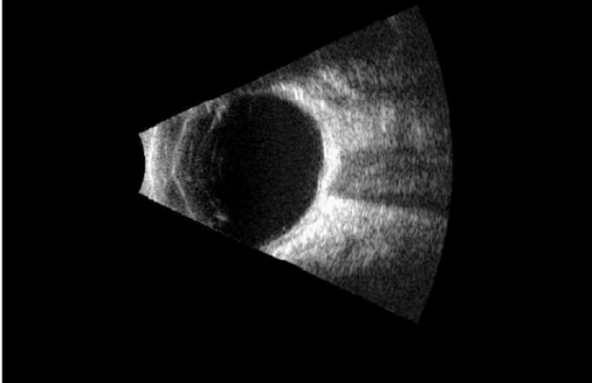
B 10 <> OD CM V:3.0.0 - 10 Jan 2020
Rashid Aalaa Gain=110dB Dyn=60dB Tgc=20dB



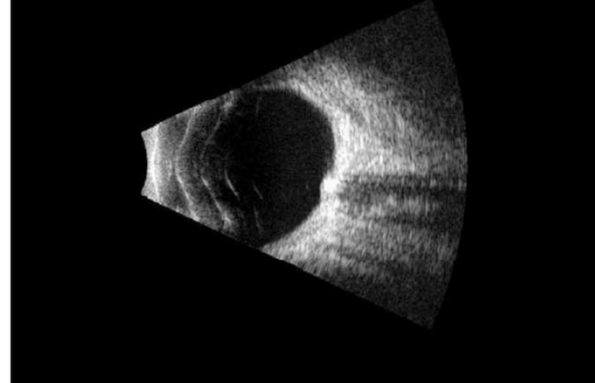
B 10 <> OS CM V:3.0.0 - 10 Jan 2020
Rashid Aalaa Gain=110dB Dyn=60dB Tgc=20dB



B 10 <> OD CM V:3.0.0 - 10 Jan 2020
Rashid Aalaa Gain=110dB Dyn=60dB Tgc=20dB



B 10 <> OS CM V:3.0.0 - 10 Jan 2020
Rashid Aalaa Gain=110dB Dyn=60dB Tgc=20dB



Case #1: 15yo IF

Diagnosis: Pseudopapilledema OS
Secondary to Optic Disc Drusen

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

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

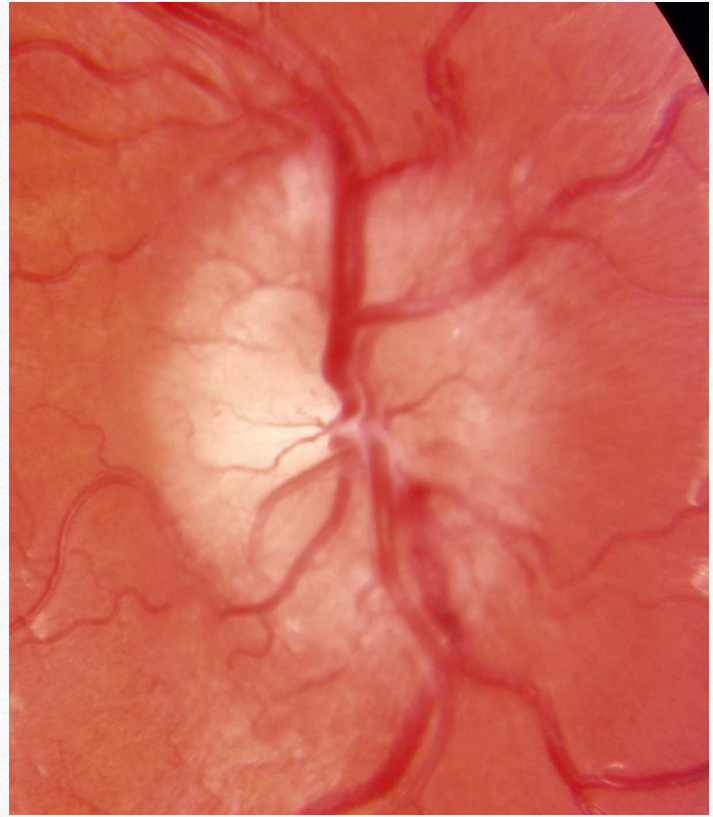
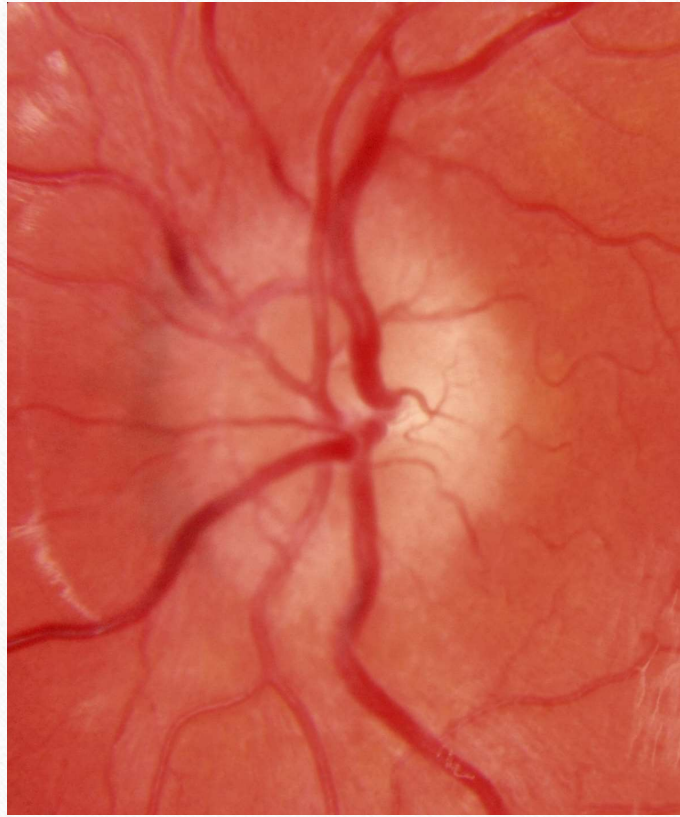
Pupils/Color: WNL OU

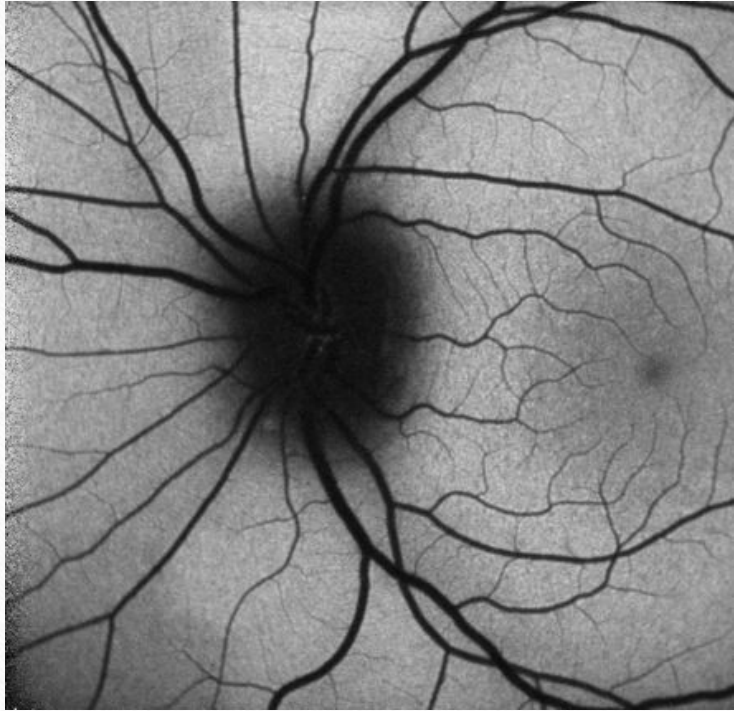
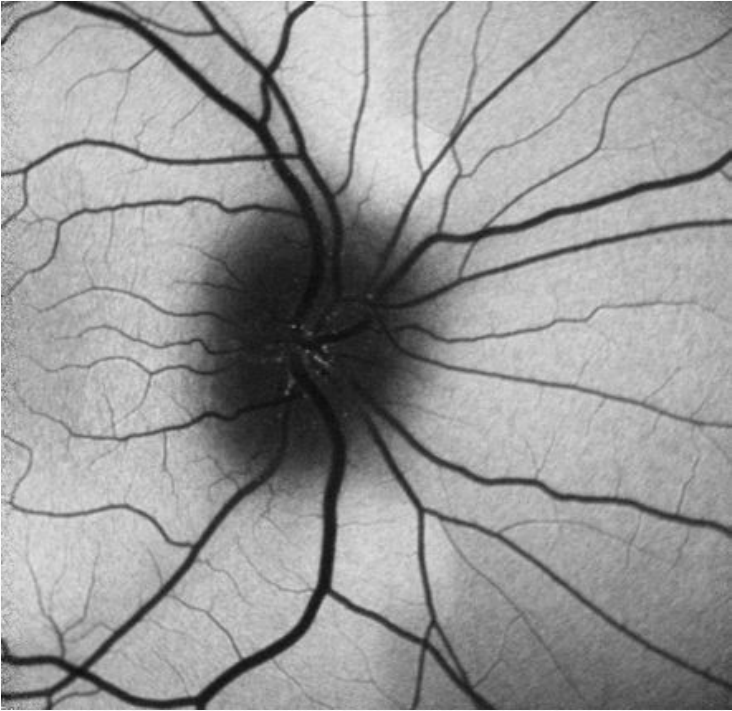
EOMs: Left ESO in primary position

Left abduction deficit

IOP: 18/21

Fundus: Disc edema OU





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

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

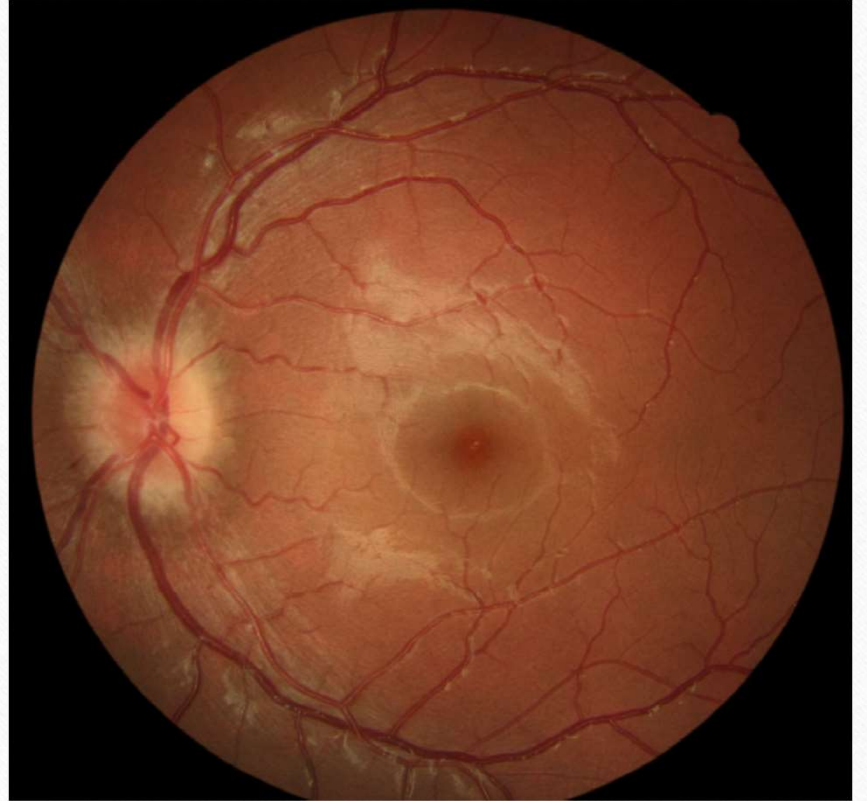
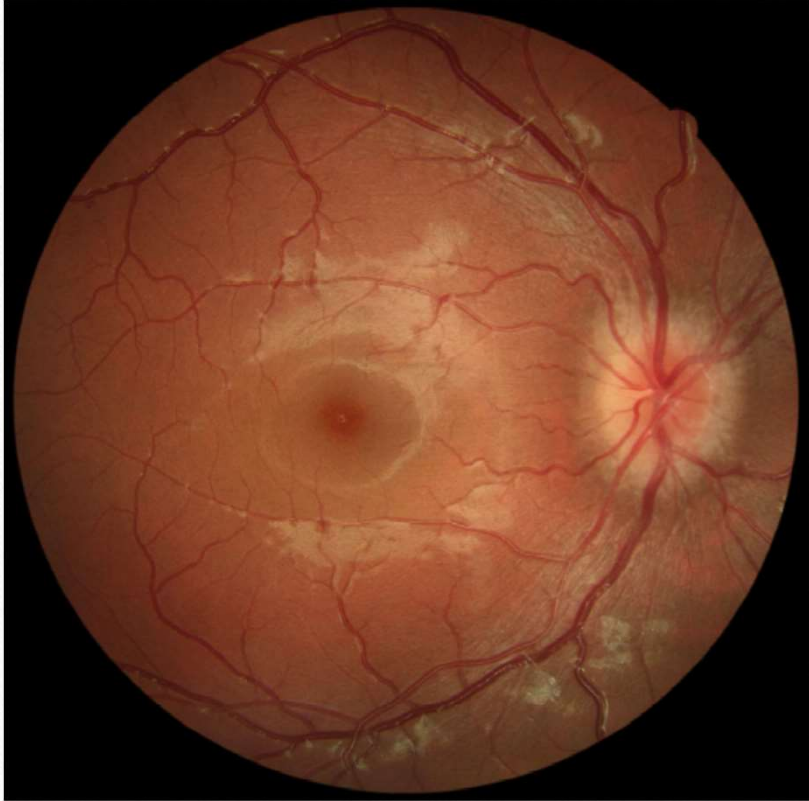
Color plates: 14/14

EOM's: full in all positions

Pupils: PERRLA –apd ou

IOP: 14/13

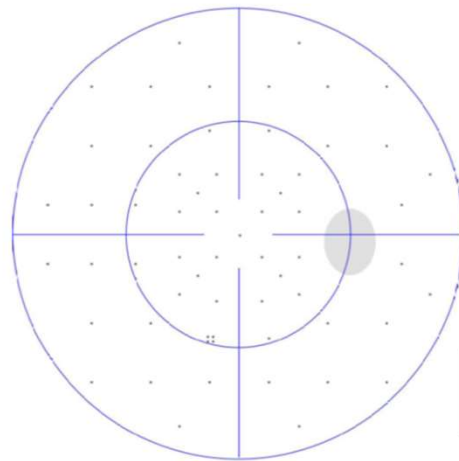
C/D: 0.1/0.1 Fundus: Elevated, blurred disc margins ou



Rennekamp Jessica, 1/23/2000

Right eye (OD) / 08/09/2017 / 11:11:51

Corrected probabilities



[%]
- P > 5
:: P < 5
■ P < 2
■ P < 1
■ P < 0,5

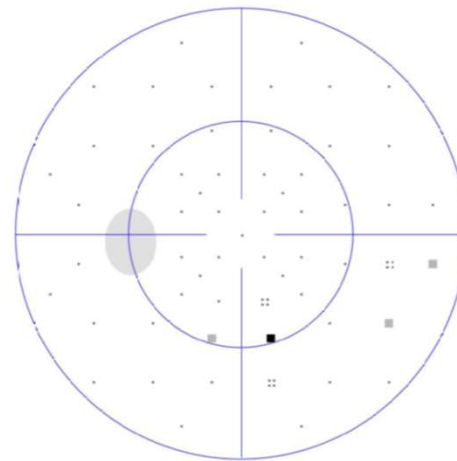
30°
MS [src]: 22.7
MD [< 2.0 src]: 0.7
sLV [< 2.5 src]: 1.6

Programs: G P Pulsar / 200 TOP Questions / repetitions: 72 / 8
Parameters: 100 / 200 asb 500 ms Duration: 04:09
Catch trials: 0/11 (0%) +, 0/11 (0%) - RF: 0.0
Trial lens: VA [m]:
Pupil [mm]: 4.9 IOP [mmHg]:
NV: T54 V2.1

Comment:

Left eye (OS) / 08/09/2017 / 11:16:33

Corrected probabilities



[%]
- P > 5
:: P < 5
■ P < 2
■ P < 1
■ P < 0,5

30°
MS [src]: 22.1
MD [< 2.0 src]: 1.2
sLV [< 2.5 src]: 2.3

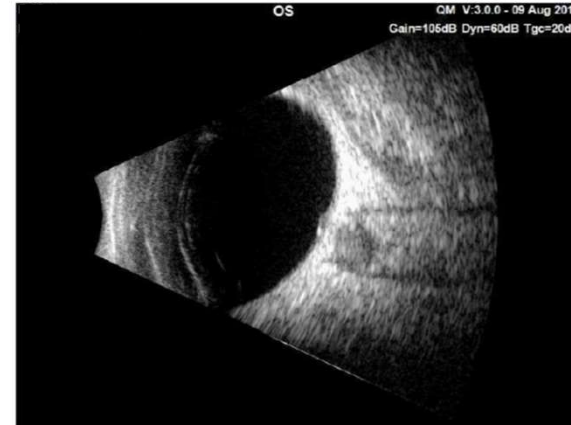
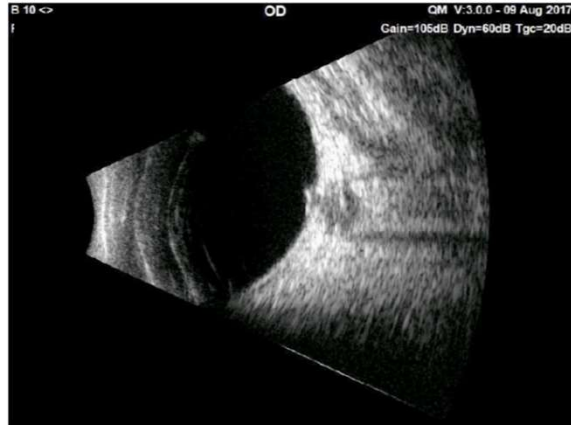
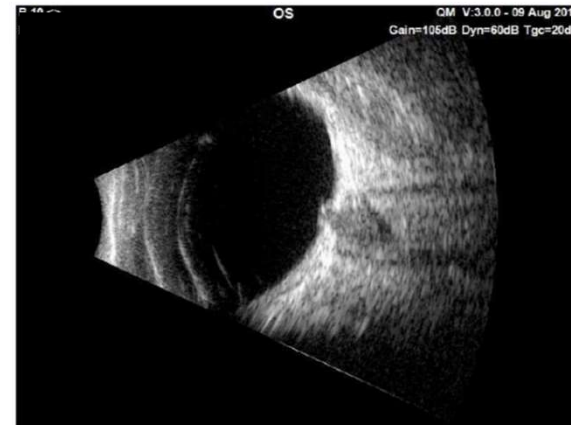
Programs: G P Pulsar / 200 TOP Questions / repetitions: 71 / 0
Parameters: 100 / 200 asb 500 ms Duration: 03:48
Catch trials: 0/10 (0%) +, 0/11 (0%) - RF: 0.0
Trial lens: VA [m]:
Pupil [mm]: 4.9 IOP [mmHg]:
NV: T54 V2.1

Comment:

OCTOPUS®

EyeSuite™ Static perimetry
OCTOPUS 600, SN 860, V 2.2.1 / 3.5.0

HAAG-STREIT
DIAGNOSTICS



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

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

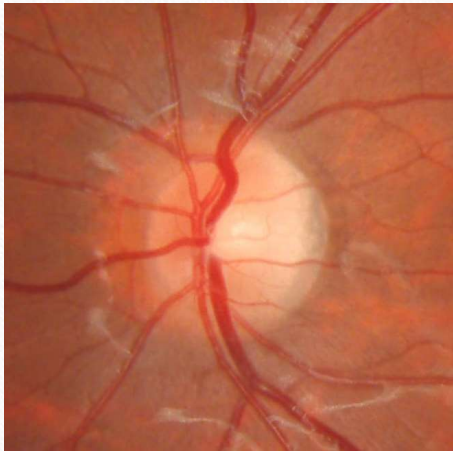
Color 14/14 OU

Pupils: PERRLA –apd OU

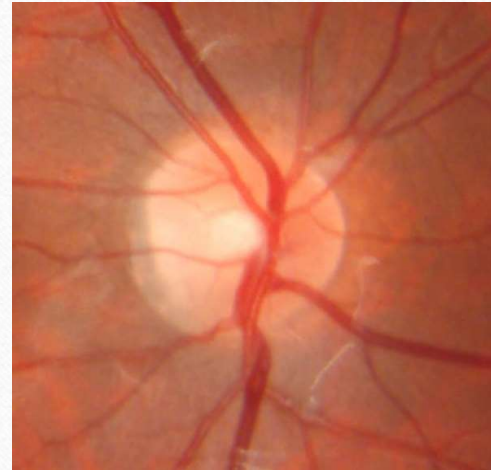
IOP: 15/11

C/D: 0.25/0.25 OU

Fundus: Disc edema OU



2016



2017



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

Case #5: 11yo AWF

10/15/19

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

BCVA: 20/20 OU, increase -050/-075 correction

EOMs: full in all positions

PERRLA –apd ou

IOP: 18/18

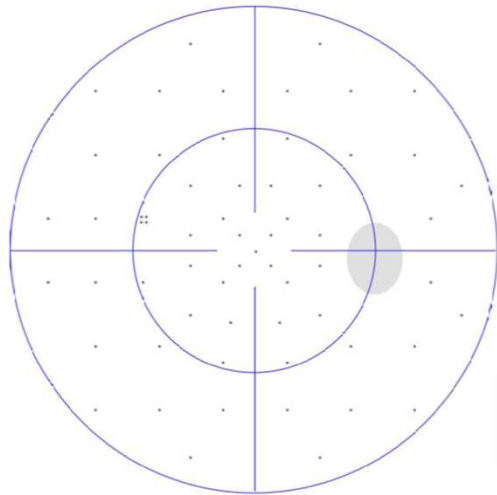
C/D: 0.15/0.25

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



Right eye (OD) / 10/15/2019 / 18:59:58
Corrected probabilities



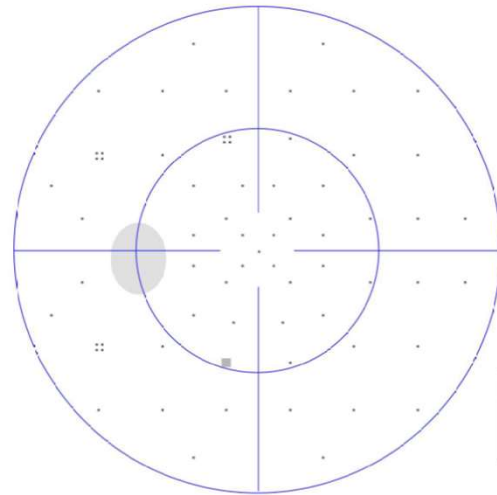
- [%]
 · P > 5
 ∴ P < 5
 ■ P < 2
 ■ P < 1
 ■ P < 0,5

30°	
MS [dB]:	27.8
MD [< 2.0 dB]:	1.1
sLV [< 2.5 dB]:	1.8

Programs: G Standard White/White / TOP Questions / repetitions: 70 / 0
 Parameters: 31.4 / 4000 asb III 100 ms Duration: 02:22
 Catch trials: 0/7 (0%) +, 0/7 (0%) - RF: 0.0
 Trial lens : 4 VA [m]:
 Pupil [mm]: 4.1 IOP [mmHg]:
 NV: T53 V2.1

Comment:

Left eye (OS) / 10/15/2019 / 19:02:43
Corrected probabilities

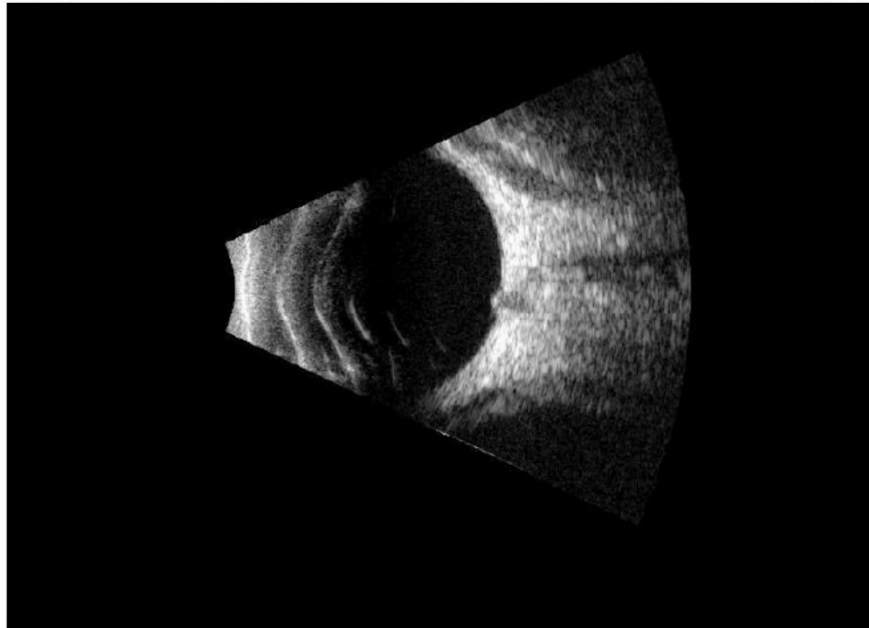
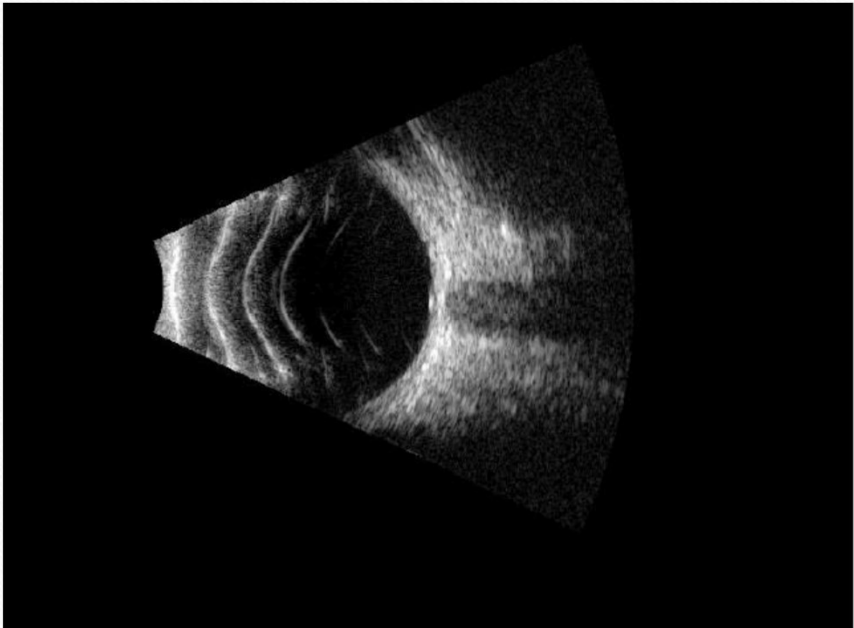


- [%]
 · F > 5
 ∴ F < 5
 ■ F < 2
 ■ F < 1
 ■ F < 0,5

30°	
MS [dB]:	27.0
MD [< 2.0 dB]:	1.9
sLV [< 2.5 dB]:	2.8

Programs: G Standard White/White / TOP Questions / repetitions: 72 / 0
 Parameters: 31.4 / 4000 asb III 100 ms Duration: 02:22
 Catch trials: 0/7 (0%) +, 0/8 (0%) - RF: 0.0
 Trial lens : 4 VA [m]:
 Pupil [mm]: 6.8 IOP [mmHg]:
 NV: T53 V2.1

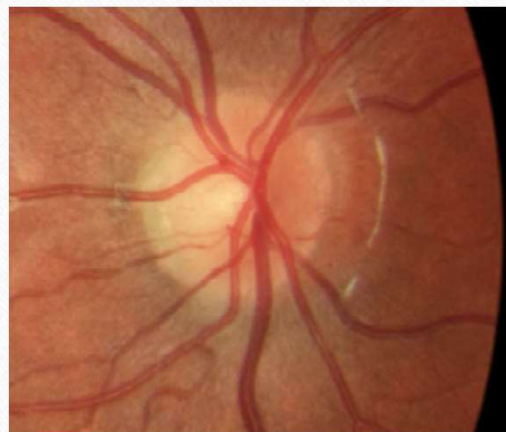
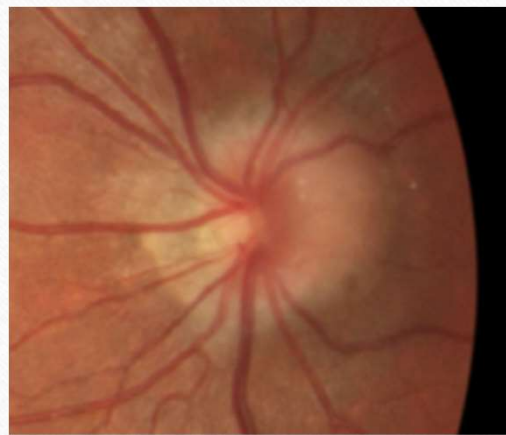
Comment:



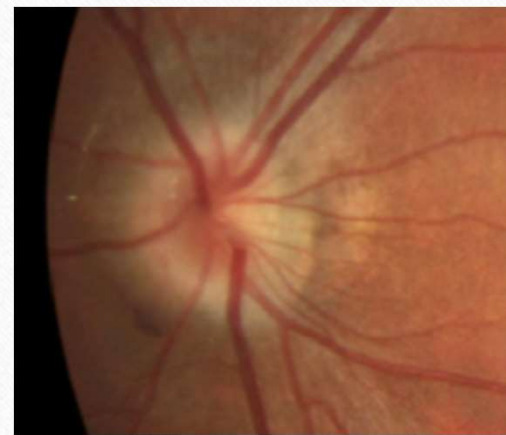
2019

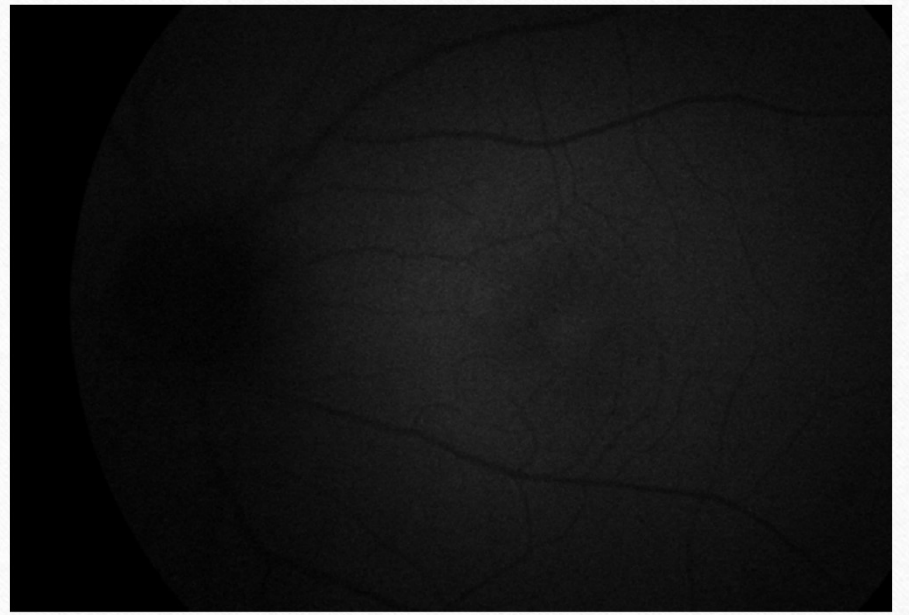
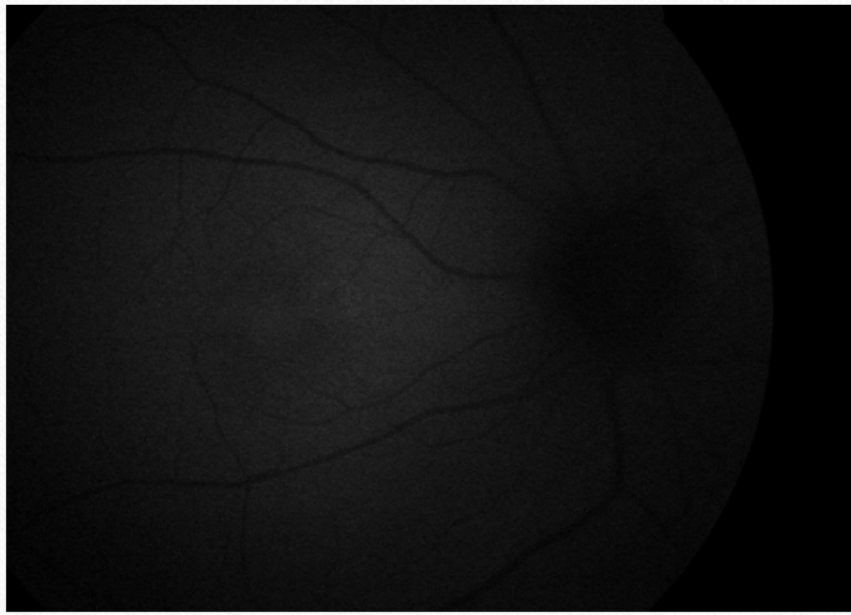
2015

OD



OS

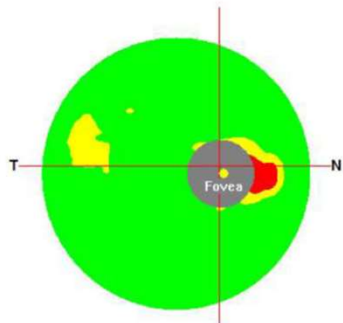




ONH/GCC OU Report

Right / OD

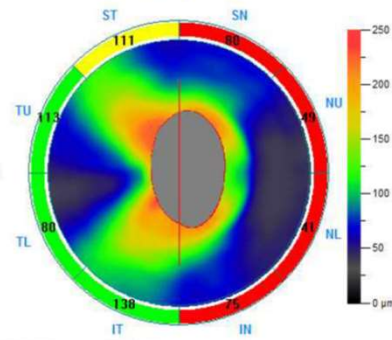
GCC NDB Reference



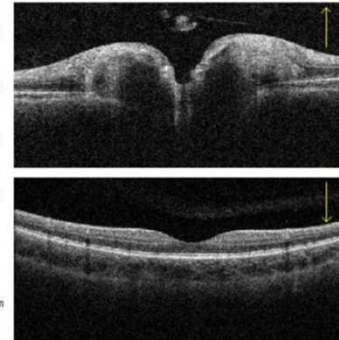
SSI: 63

Exam Date: 08/14/2020

Optic Nerve Head Map



SSI: 61



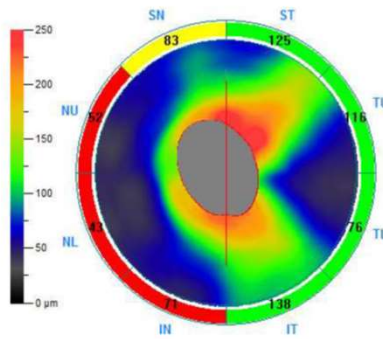
■ p>5% Within Normal
■ p<5% Borderline
■ p<1% Outside Normal

Summary Parameters

RNFL Analysis	OD	OS	Inter Eye (OD-OS)
Average RNFL (μm)	86	88	-2
Superior RNFL (μm)	88	84	-6
Inferior RNFL (μm)	83	82	1
Intra Eye (S-I) (μm)	5	12	N/A
ONH Analysis	OD	OS	Inter Eye (OD-OS)
Cup/Disc Area Ratio	0.00	0.00	0.00
Cup/Disc V. Ratio	0.00	0.00	0.00
Cup/Disc H. Ratio	0.00	0.00	0.00
Rim Area (mm ²)	2.33	2.08	0.25
Disc Area (mm ²)	2.33	2.08	0.25
Cup Volume (mm ³)	0.000	0.000	0.000
GCC Analysis	OD	OS	Inter Eye (OD-OS)
Average GCC (μm)	86	84	2
Superior GCC (μm)	85	82	2
Inferior GCC (μm)	87	85	2
Intra Eye (S-I) (μm)	-2	-2	N/A
FLV (%)	0.42	0.58	-0.16
GLV (%)	0.67	11.80	-1.68

Left / OS

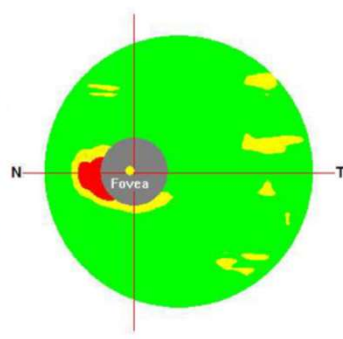
Optic Nerve Head Map



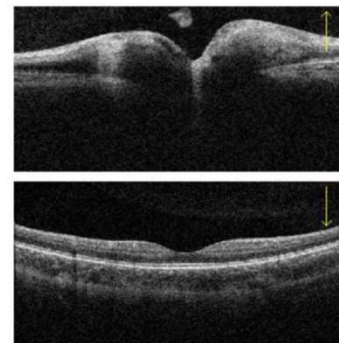
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Exam Date: 08/14/2020

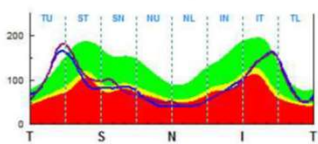
GCC NDB Reference



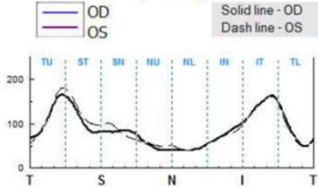
SSI: 57



TSNIT NDB Reference



TSNIT Symmetry Plot



● NDB
● Thickness

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

Initial Visit CCH Neuro 10/21/19

ASSESSMENT:

██████████ 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

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:

██████ 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.

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.

2nd visit CEI NeuroOPHTH 02/26/20

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

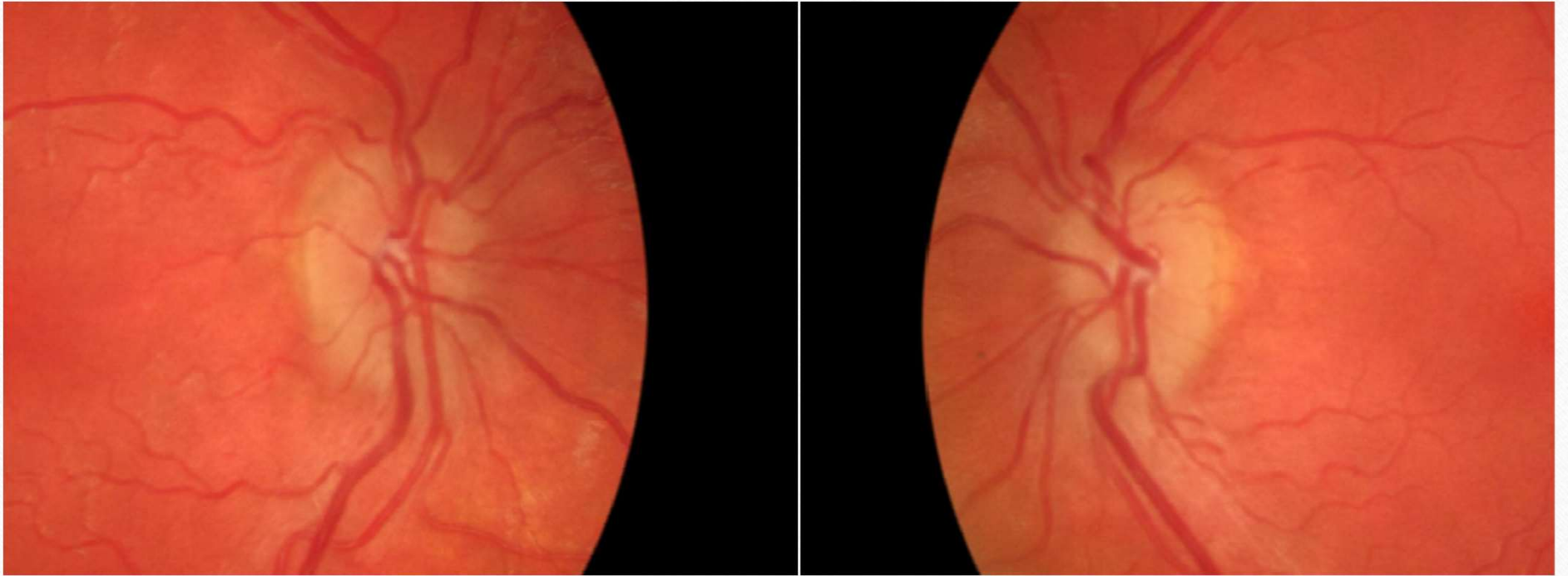
Evaluation: Nasal disc elevation OU, Stable vision and 24-2 visual fields w normal fixation losses

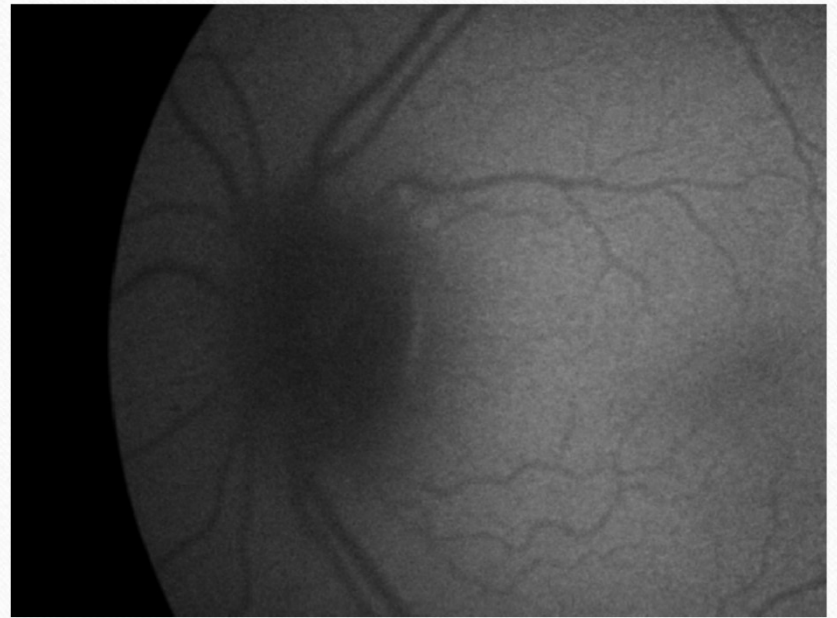
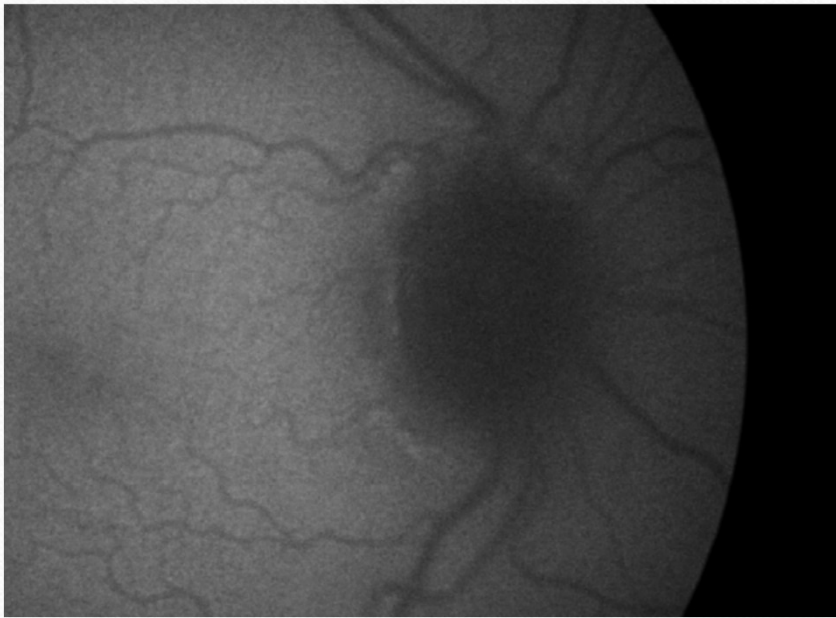
Diagnosis: Pseudopapilledema of the optic discs OU
Buried disc drusen OU

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

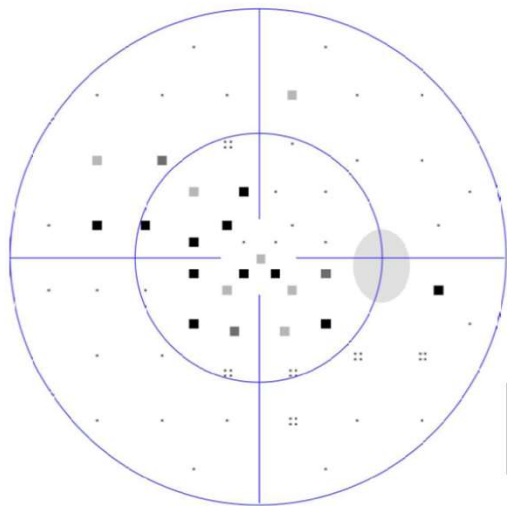
Case #6 21yo WF

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





Right eye (OD) / 08/05/2020 / 13:58:57
Corrected probabilities



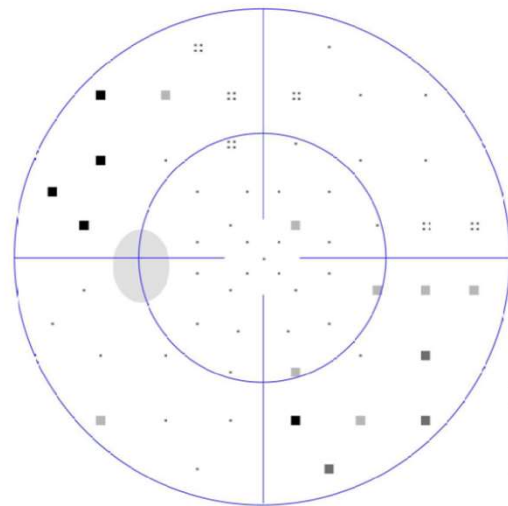
- [.] P > 5
- [.] P < 5
- [■] P < 2
- [■] P < 1
- [■] P < 0.5

30°	
MS [dB]:	18.8
MD [< 2.0 dB]:	10.1
sLV [< 2.5 dB]:	5.8

Programs: G Standard White/White / TOP Questions / repetitions: 69 / 0
 Parameters: 31.4 / 4000 asb III 100 ms Duration: 03:01
 Catch trials: 0/7 (0%) +, 1/7 (14%) - RF: 7.1
 Trial lens: \otimes VA [m]:
 Pupil [mm]: 5.0 IOP [mmHg]:
 NV: T53 V2.1

Comment:

Left eye (OS) / 08/05/2020 / 14:02:55
Corrected probabilities



- [.] F > 5
- [.] F < 5
- [■] F < 2
- [■] F < 1
- [■] F < 0.5

30°	
MS [dB]:	20.5
MD [< 2.0 dB]:	8.4
sLV [< 2.5 dB]:	6.5

Programs: G Standard White/White / TOP Questions / repetitions: 69 / 0
 Parameters: 31.4 / 4000 asb III 100 ms Duration: 02:51
 Catch trials: 0/7 (0%) +, 2/7 (29%) - RF: 14.2
 Trial lens: \otimes VA [m]:
 Pupil [mm]: 5.0 IOP [mmHg]:
 NV: T53 V2.1

Comment:

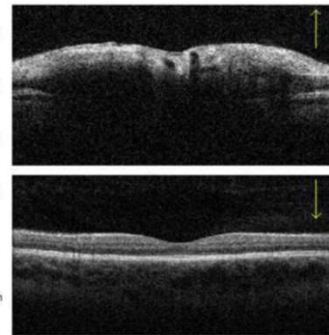
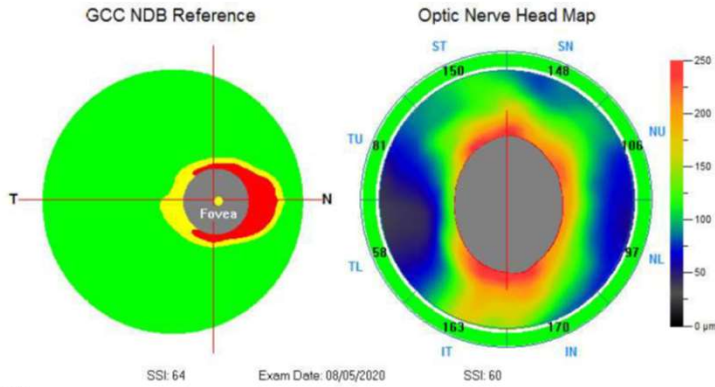
OCTOPUS®

EyeSuite™ Static perimetry
OCTOPUS 600, SN 860, V 2.2.1 / 3.5.0

HS HAAG-STREIT
DIAGNOSTICS

ONH/GCC OU Report

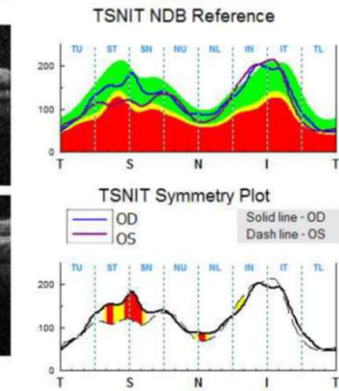
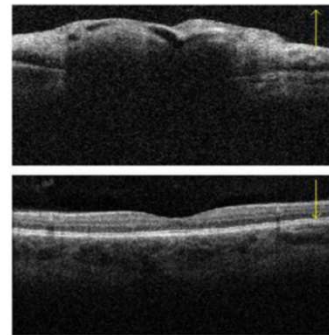
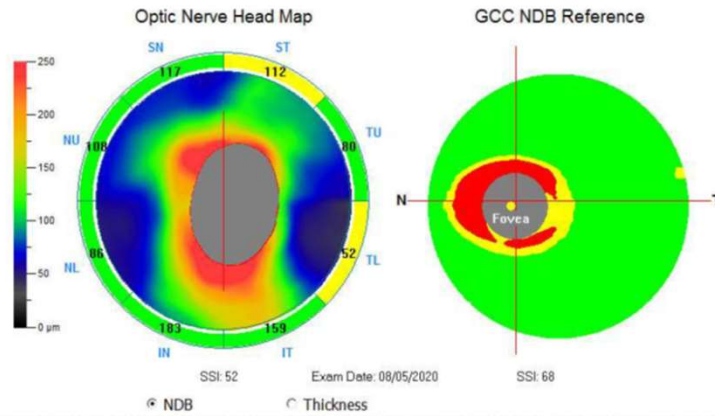
Right / OD

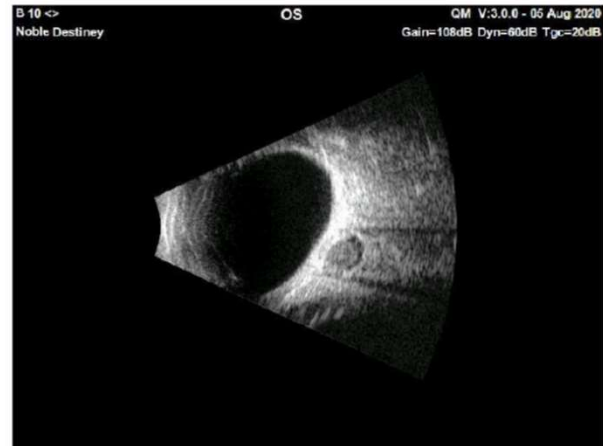
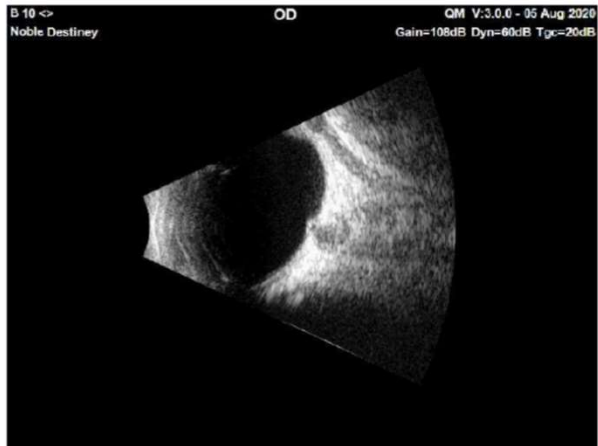
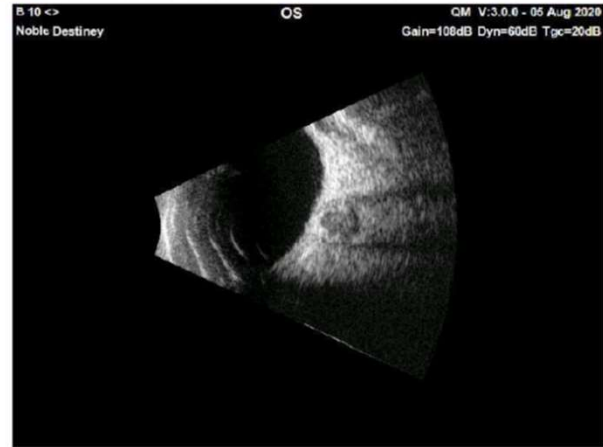
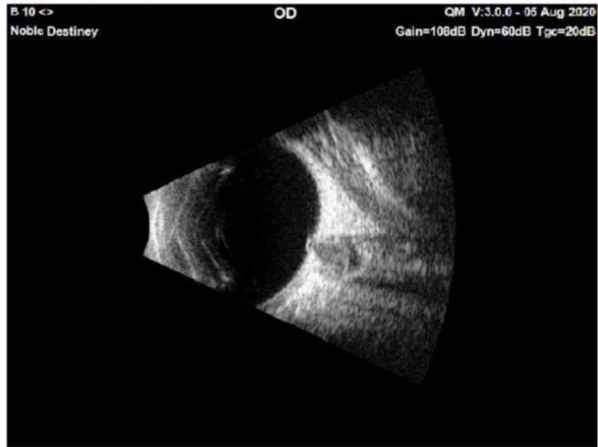


Summary Parameters			
RNFL Analysis			
Average RNFL (µm)	OD	OS	Inter Eye (OD-OS)
	122	112	10
Superior RNFL (µm)	121	104	17
Inferior RNFL (µm)	122	120	2
Intra Eye (S-I) (µm)	-1	-16	N/A
ONH Analysis			
Cup/Disc Area Ratio	OD	OS	Inter Eye (OD-OS)
	0.00	0.00	0.00
Cup/Disc V. Ratio	0.00	0.00	0.00
Cup/Disc H. Ratio	0.00	0.00	0.00
Rim Area (mm ²)	4.22	3.08	1.14
Disc Area (mm ²)	4.22	3.08	1.14
Cup Volume (mm ³)	0.000	0.000	0.000
GCC Analysis			
Average GCC (µm)	OD	OS	Inter Eye (OD-OS)
	92	91	1
Superior GCC (µm)	92	90	2
Inferior GCC (µm)	92	92	1
Intra Eye (S-I) (µm)	-1	-2	N/A
FLV (%)	2.44	0.28	-0.84
GLV (%)	4.31	5.34	-1.03

■ p<5% Within Normal
■ p<5% Borderline
■ p<1% Outside Normal

Left / OS





Case #6 21yo WF

- Diagnosis: Probable Idiopathic Intracranial Hypertension
Consistent profile and symptoms?
- Plan: Immediate MRI imaging to r/o intracranial lesion
Neuro consultation
If MRI negative, initiate weight loss and Acetylzolamide

Summary