OPTIC NERVE SWELLING
Neuro-ophthalmic Evaluation and Management

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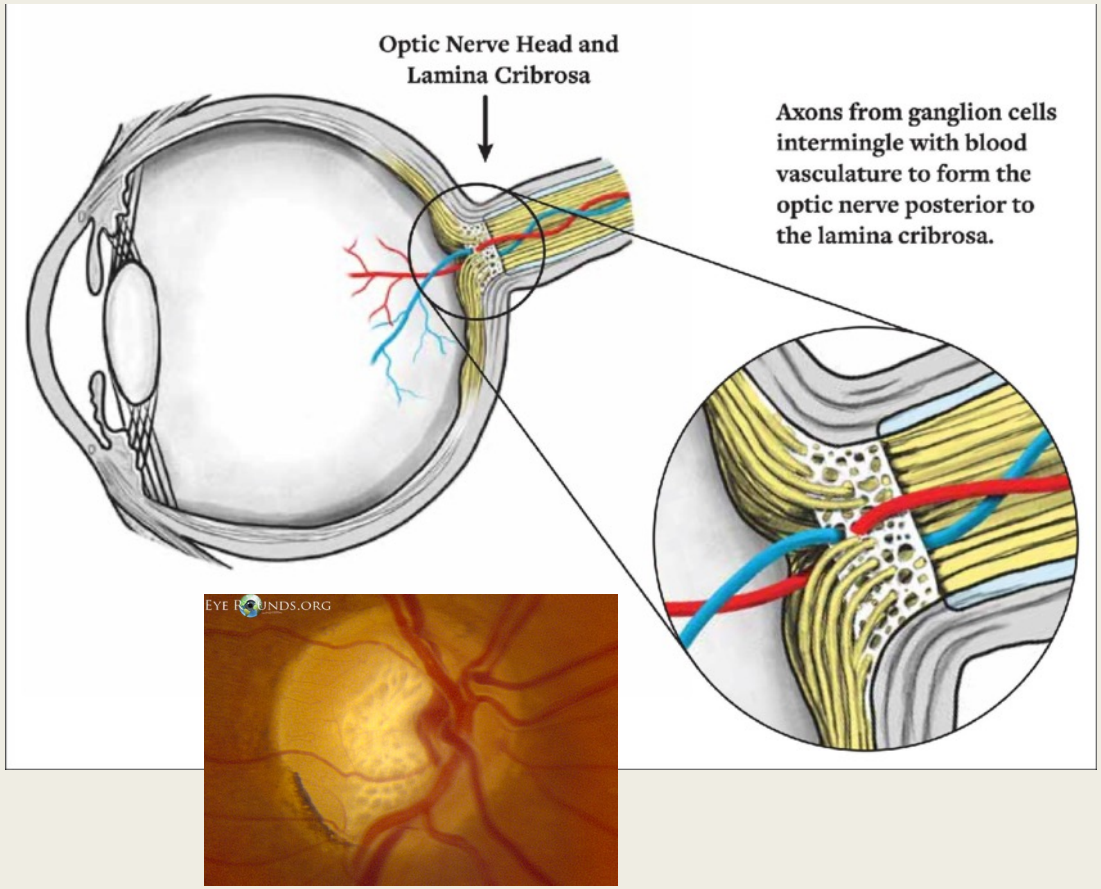
UCONN Health

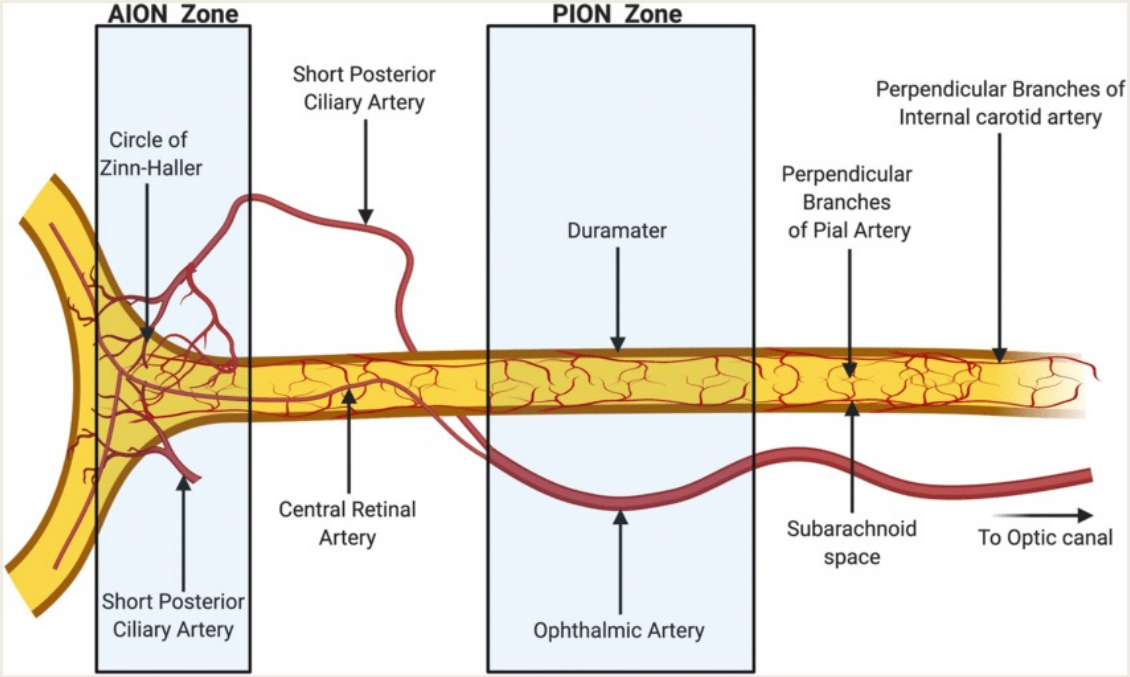
Disc Edema

- *A nonspecific term describing localized swelling anterior to the lamina cribrosa*
- *Papilledema -optic disc edema resulting from raised intracranial pressure*
- *All other optic disc edema is termed disc edema or swollen optic nerve*



Mechanisms of Optic Nerve Edema

1. *Local optic nerve injury:*
   - *Inflammation (anterior optic neuritis)*
   - *Papillitis*
   - *Ischemia (anterior ischemic optic neuropathy)*
   - *Fluctuations in intraocular pressure (high, as in acute glaucoma, or low, as in ocular hypotony)*
   - *Toxicity*



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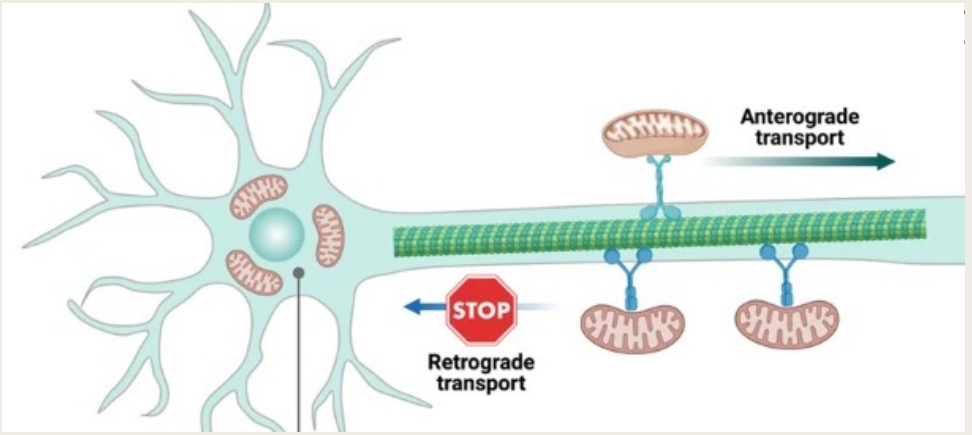
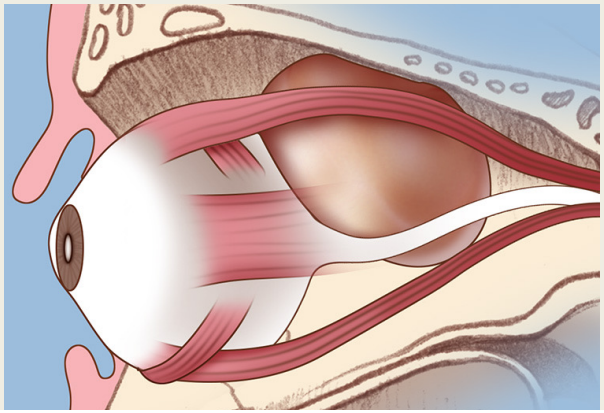
   - *Toxicity*

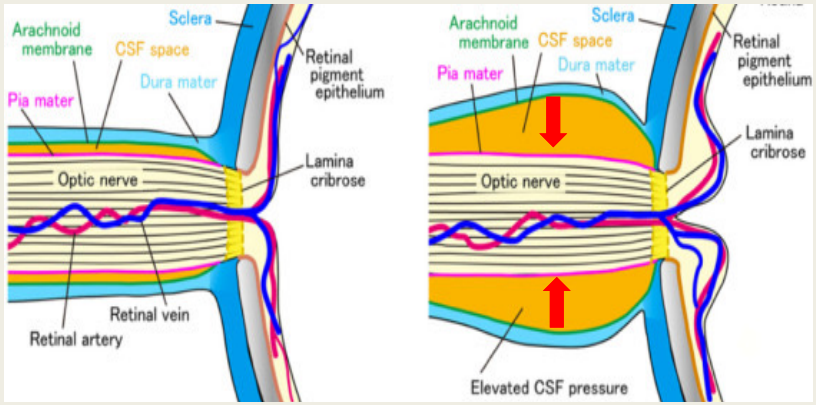
Mechanisms of Optic Nerve Edema

2. Blockage of retrograde axonal transport from optic nerve compression:

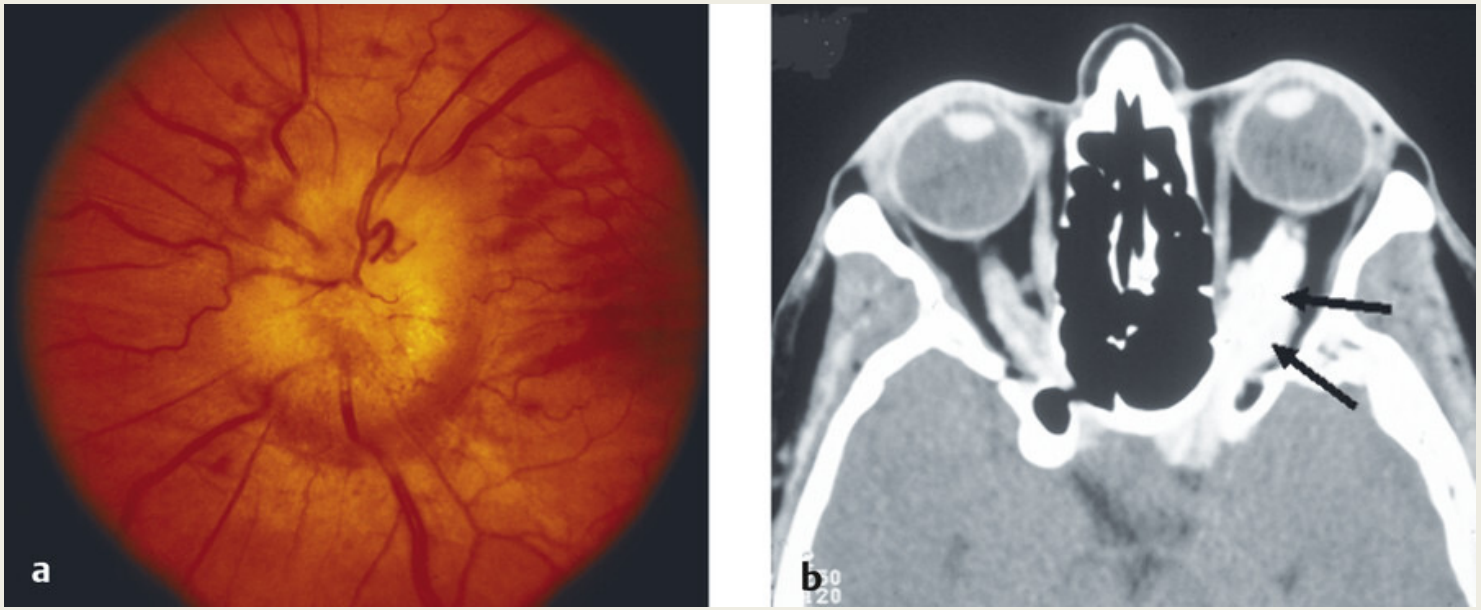
- *Optic nerve*
- *Tumor or orbital mass*
- *Raised intracranial pressure (papilledema)*





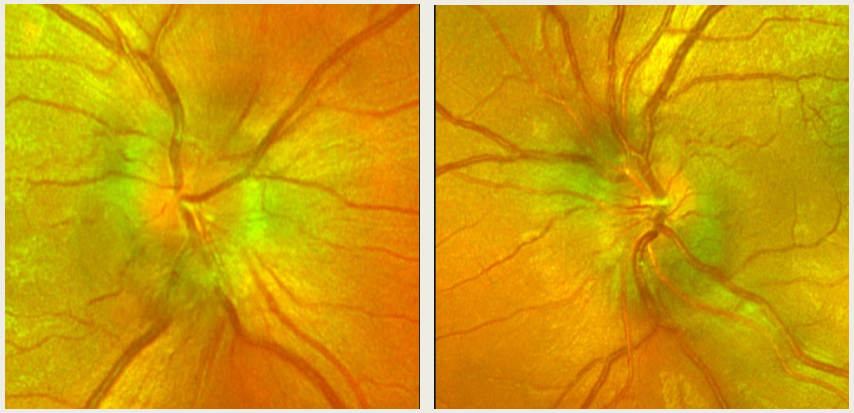


Left optic nerve sheath meningioma

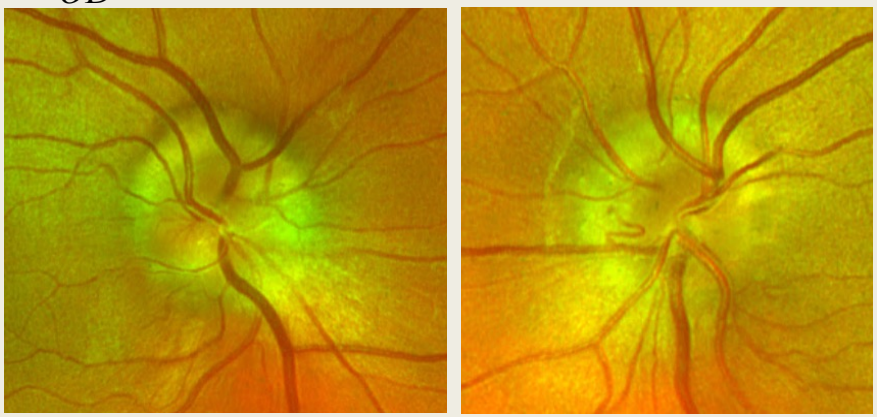


Differential Diagnosis of Disc Edema

OD *Patient A* *OS*



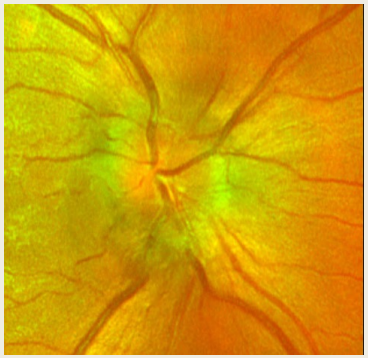
OD *Patient B* *OS*



Is it true edema or swelling? Or is it pseudo edema?

Characteristics of true disc edema versus pseudo-edema

True Disc Edema	Pseudo- edema
Elevated optic nerve	*Elevated optic nerve*
Margins blurry	*Sharp margins*
Vessels obscured	*Vessels not obscured*
Venous dilation and tortuosity	*Absence of central cup*
Peripapillary hemorrhages and exudates	*Anomalous retinal vasculature (arterial branching)*
Leakage on fluorescein angiogram	*No leakage on fluorescein angiogram*



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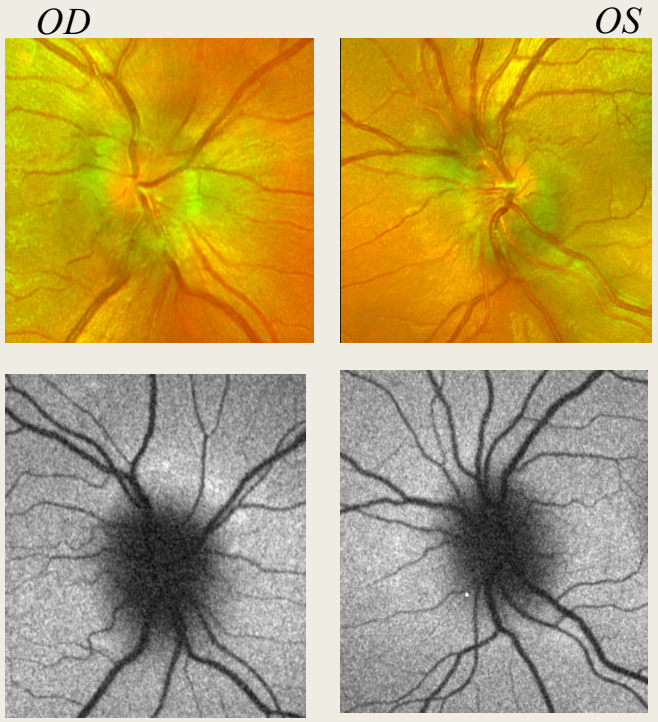
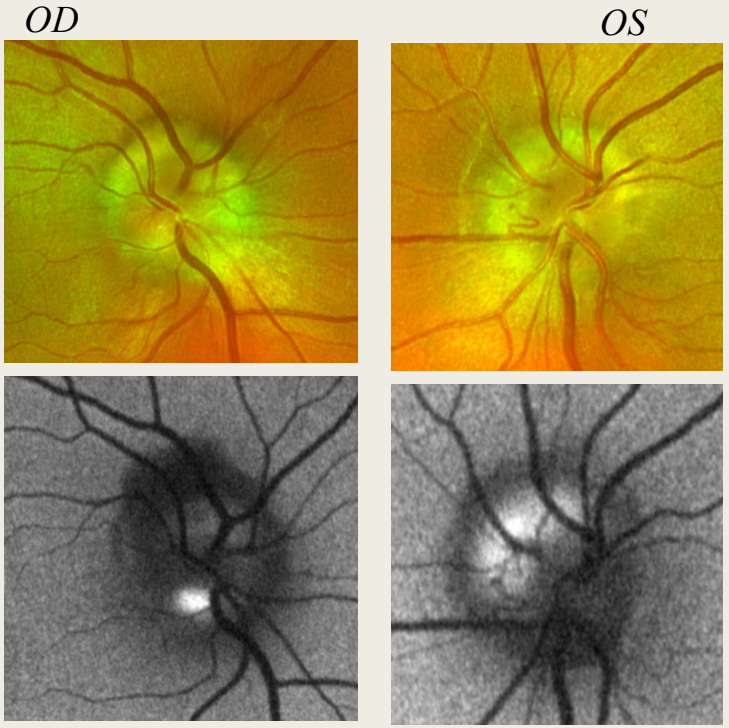
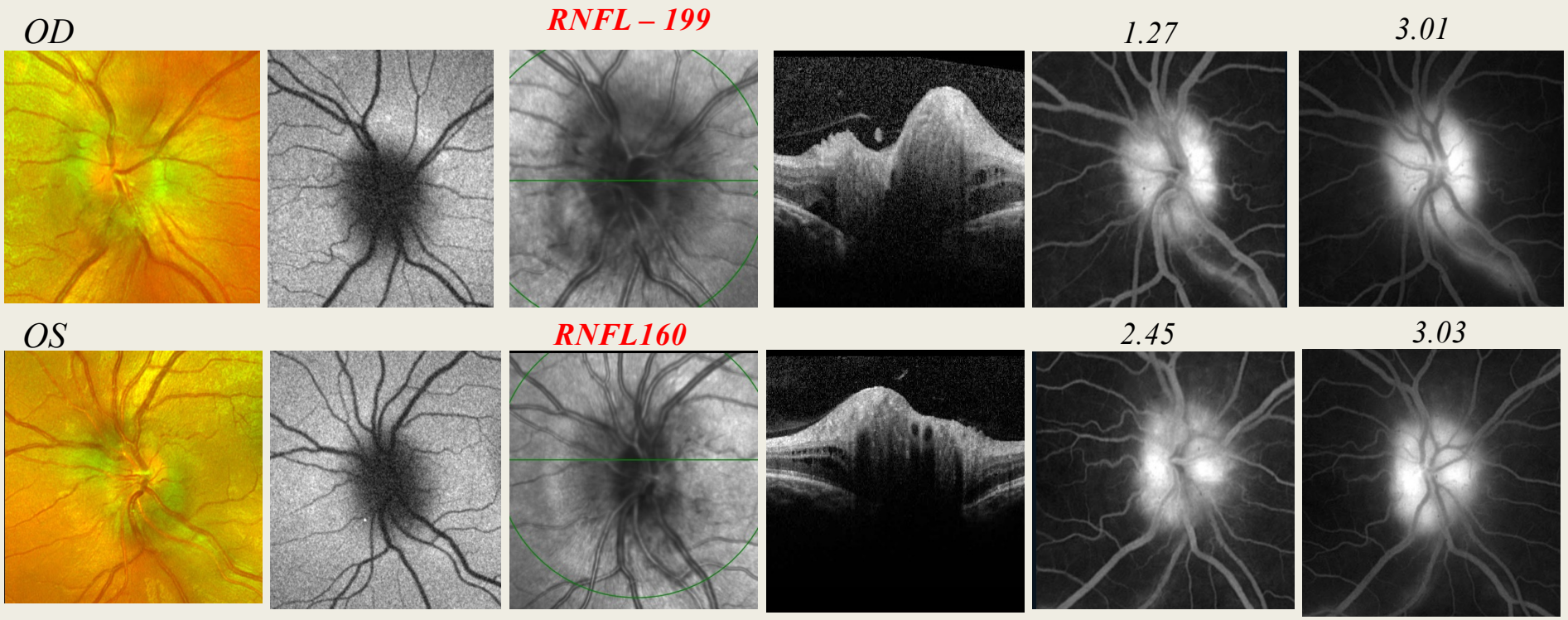


Red Free Image of optic disc – No disc autofluorescence – unlikely drusen



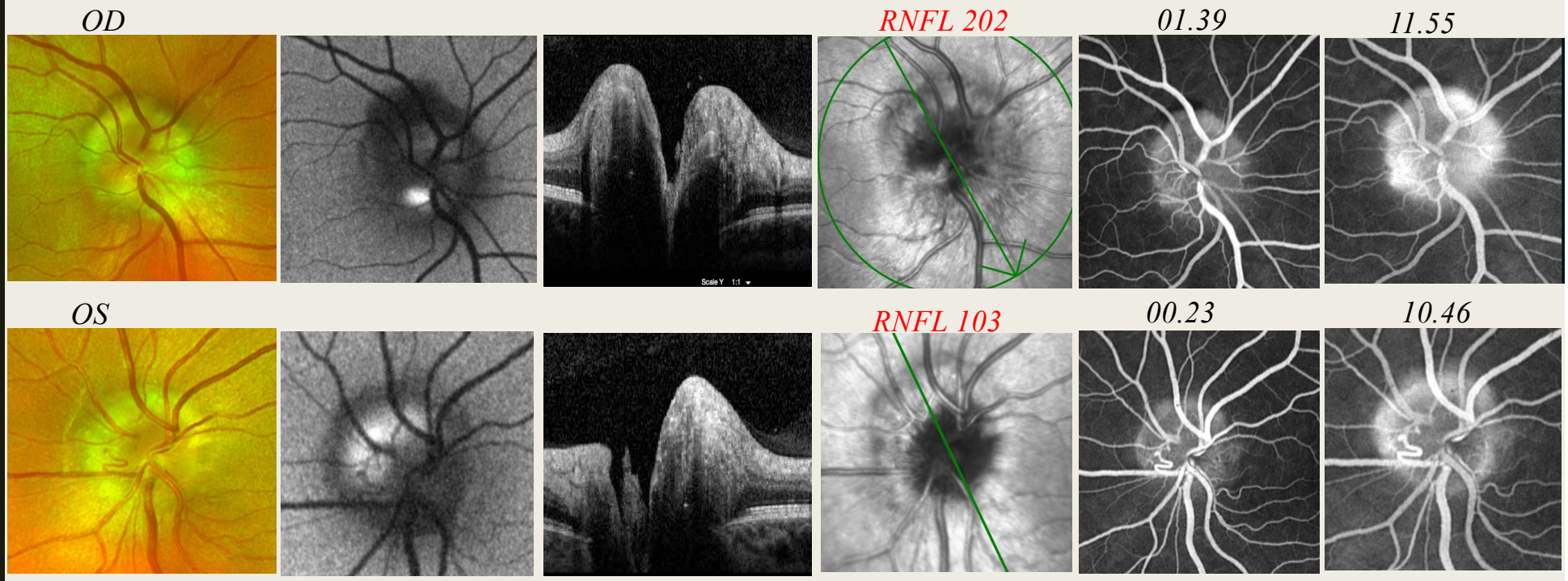
Red free image – Autofluorescence from Superficial optic disc drusen

Characteristics of true disc edema versus pseudo-edema



True disc edema with leakage on fluorescein angiography (late phase).

Characteristics of true disc edema versus pseudo-edema

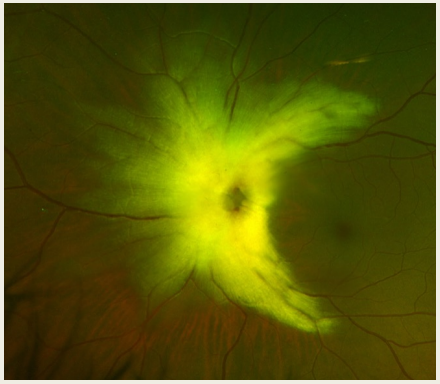


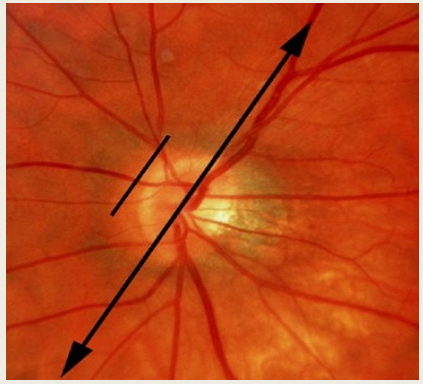
Pseudo-edema with no leakage on fluorescein angiography (there is late staining only).

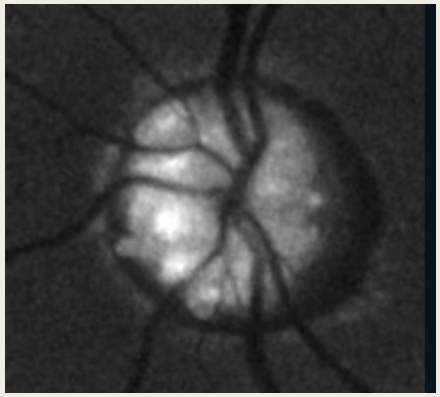
Differential Diagnosis of Disc Edema

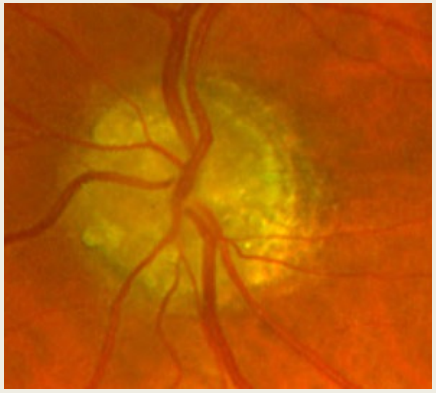
Disc elevation without true swelling

- *Optic disc anomalies*
  - ✓ *Myelinated nerve fibers*
  - ✓ *Tilted disc*
  - ✓ *Drusen*
  - ✓ *Crowded disc*
- *Optic disc infiltration*
- *Leber hereditary optic neuropathy*









Differential Diagnosis of Disc Edema

True disc swelling

- *Elevated intracranial pressure (papilledema)*
- *Inflammatory optic neuropathy*
  - ✓ *Demyelinating*
  - ✓ *Sarcoidosis or other inflammatory diseases*
  - ✓ *Infectious*
- *Neuro-retinitis*
- *Vascular optic neuropathy*
  - ✓ *Anterior ischemic optic neuropathy*
    - ❖ *Nonarteritic*
    - ❖ *Arteritic*
  - ✓ *Diabetic papillopathy*
  - ✓ *Central retinal vein occlusion*
  - ✓ *Carotid-cavernous fistula*
  - ✓ *Malignant systemic hypertension*

Differential Diagnosis of Disc Edema

True disc swelling

- *Compressive optic neuropathy*
  - *Neoplastic*
    - ✓ *Meningioma*
    - ✓ *Hemangioma*
    - ✓ *Lymphangioma*
  - *Non-neoplastic*
    - ✓ *Thyroid ophthalmopathy*
    - ✓ *Orbital inflammatory pseudotumor*
- *Infiltrative optic neuropathy*
  - *Neoplastic*
    - ✓ *Leukemia*
    - ✓ *Lymphoma*
    - ✓ *Glioma*
  - *Non-neoplastic*
    - ✓ *Sarcoidosis*
- *Intraocular hypotony (low intraocular pressure)*

Differential Diagnosis of Disc Edema

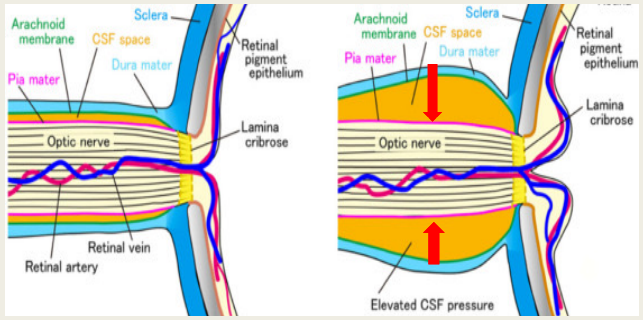
True disc swelling

- *Toxic*
- *Metabolic/nutritional deficiencies*
- *Traumatic optic neuropathy*
- *Intraocular hypotony (low intraocular pressure)*

Intraocular hypotony w/ hypotonic maculopathy

Mechanisms responsible for raised intracranial pressure and papilledema

- *Hydrocephalus*
- *Intracranial mass*
  - ✓ *Tumor, abscess*
  - ✓ *Intracerebral hemorrhage*
  - ✓ *Subdural/epidural hemorrhage*
  - ✓ *Large vascular malformation*
- *Idiopathic intracranial hypertension*
- *Meningeal process*
  - ✓ *Infectious*
  - ✓ *Inflammatory*
  - ✓ *Neoplastic*
- *Increased venous pressure*
- *Cerebral venous thrombosis*



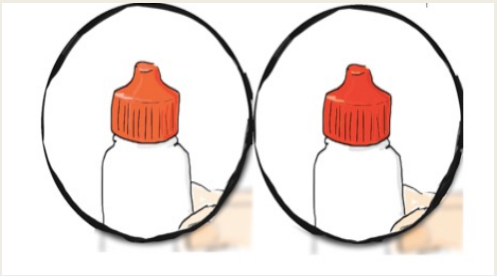
Disc Edema

- *Absence of disc edema does not rule out raised ICP in a patient presenting with headache.*
- *Look for an underlying neurologic process*
- *Careful evaluation of the visual function is needed*
- *High risk of permanent visual loss from secondary optic atrophy*
- *Check blood pressure*
- *Remember malignant hypertension mimics papilledema*

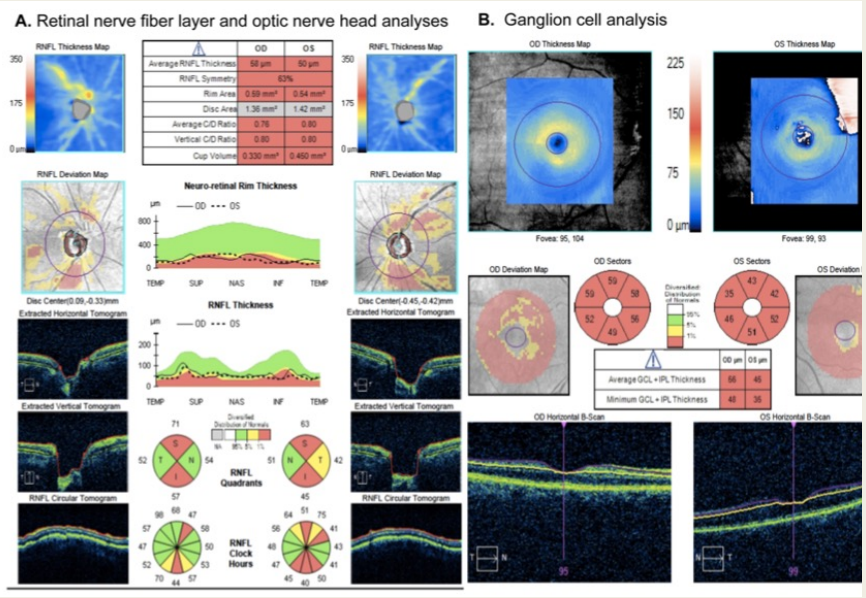
Evaluation of the Patient with Disc Edema

Clinical exam

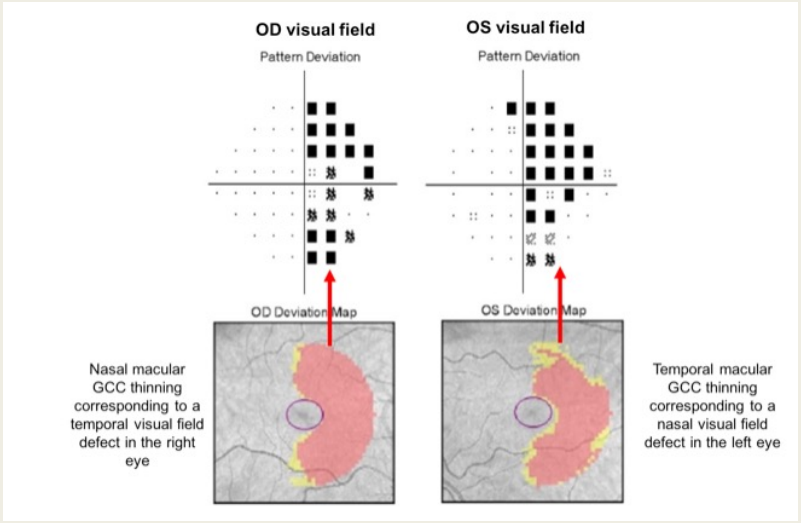
- *Visual acuity*
- *Pupillary Exam*
- *Confrontation*
- *Color/Stereo vision*
- *IOP*
- *EOM-Range of movement*
- *Fixation/Eye Alignment*
- *Saccades /Smooth pursuit*
- *Anterior segment*
- *Posterior segment – Dilated*
- *Neurology exam*



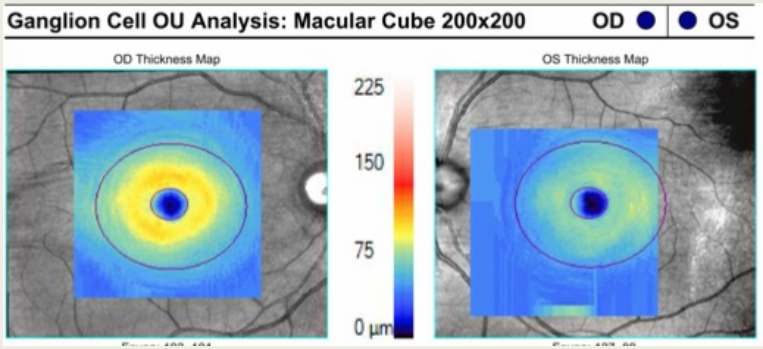
Dyschromatopsia / Red desaturation



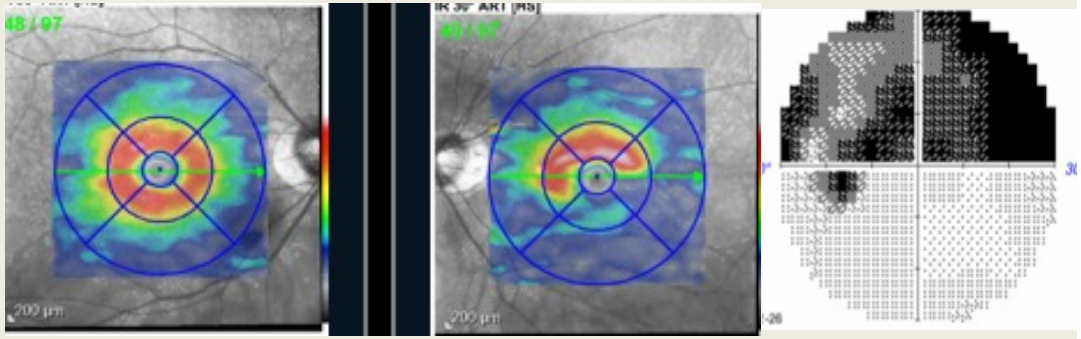
OCT –OCULAR COHERENCE TOMOGRAPHY



Optic Neuritis OS



Ischemia – NAION



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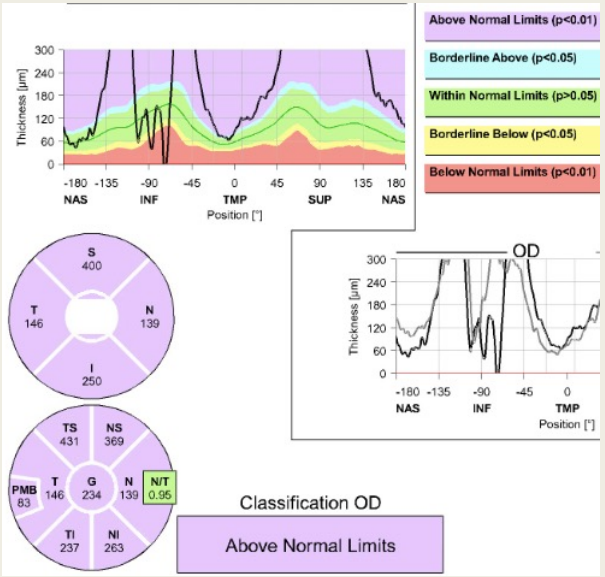
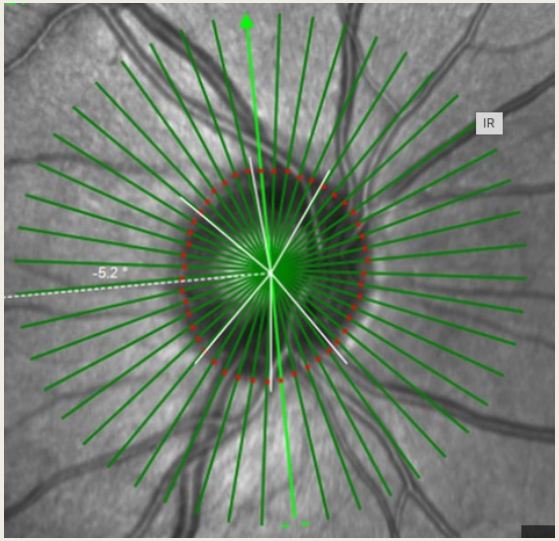
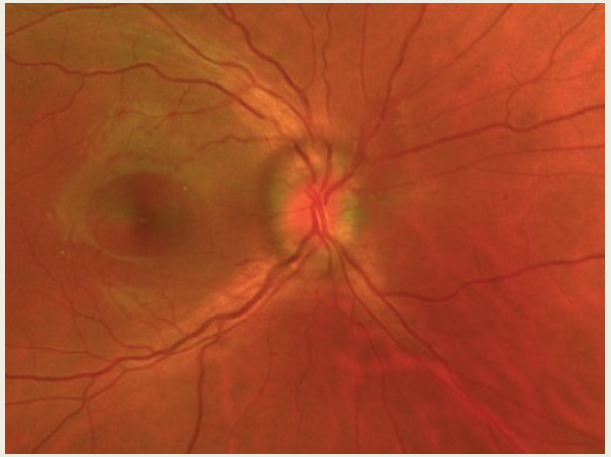
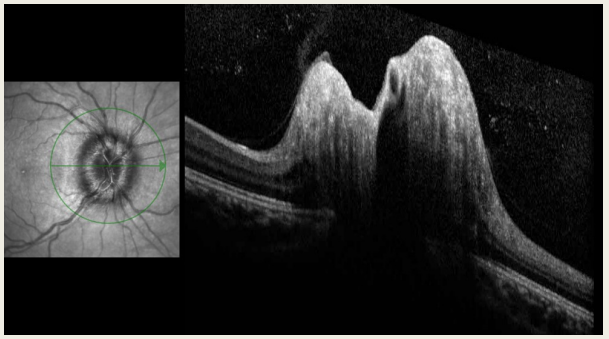


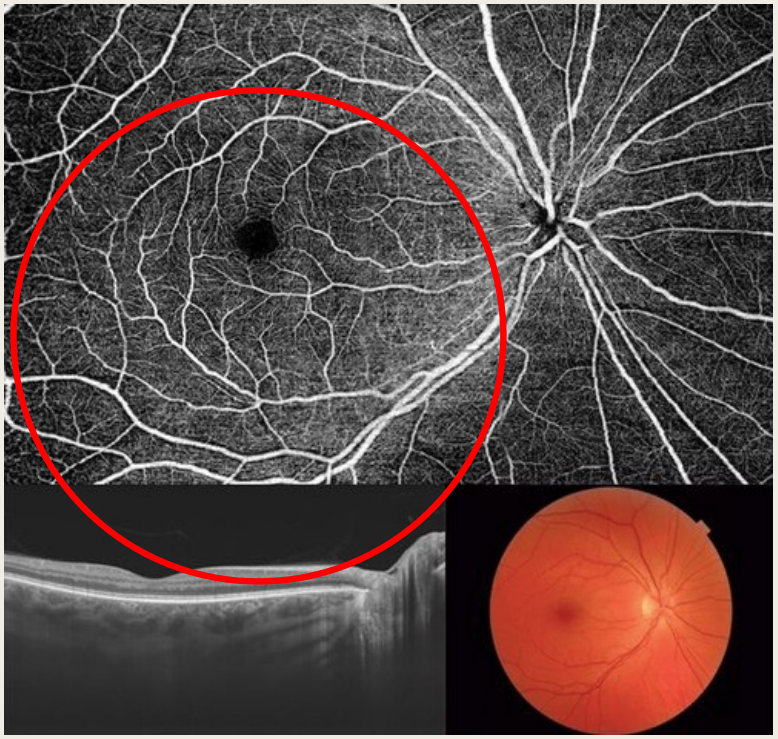


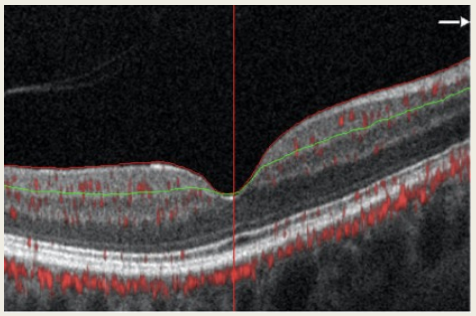


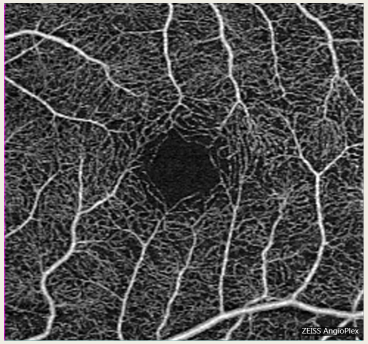


OCULAR TESTING
OCULAR COHERENCE TOMOGRAPHY

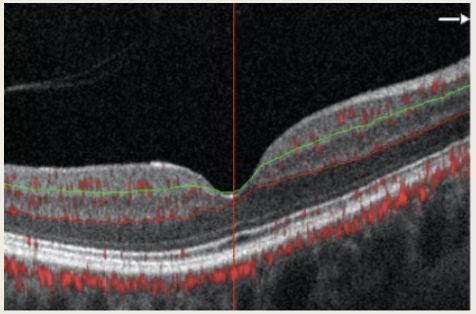
OCTA

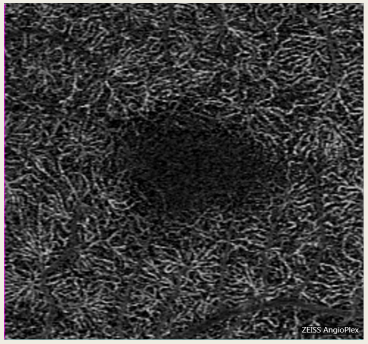




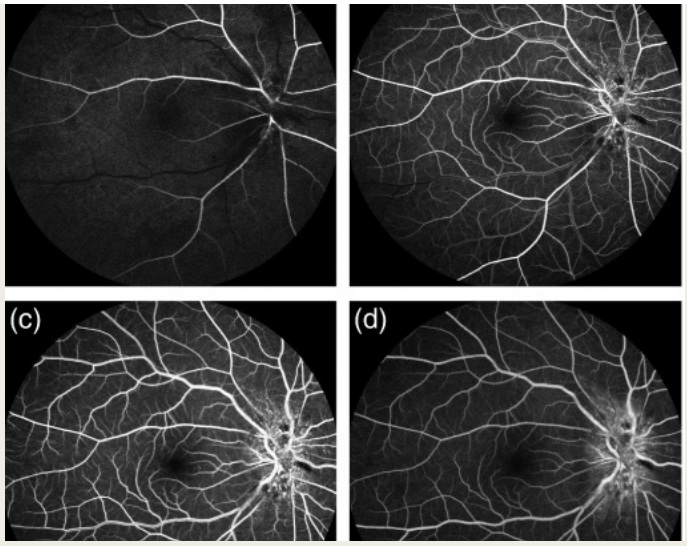


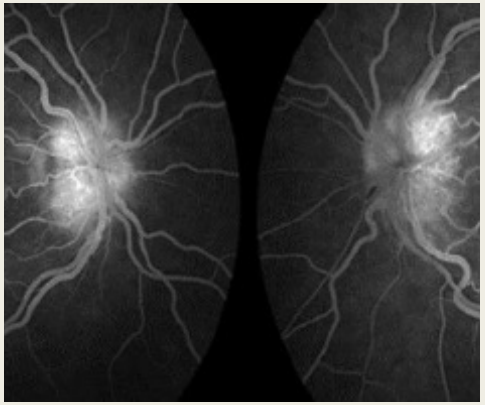
Superficial layer





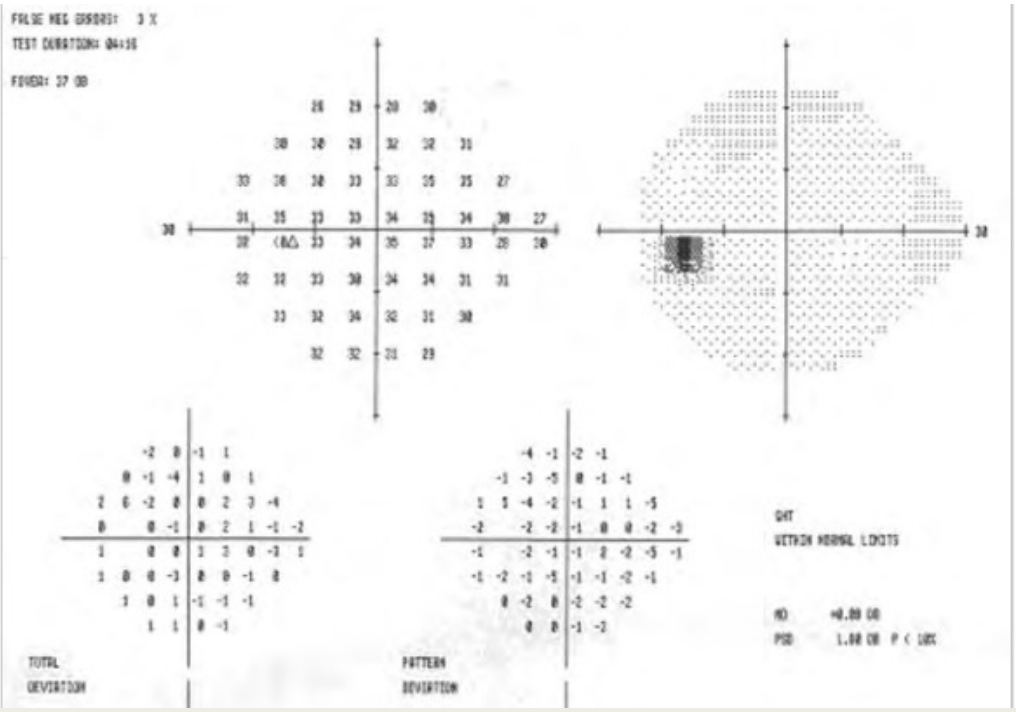
Deep layer





Fundus Fluorescein Angiogram

Ref: Neuro-Ophthalmology Illustrated book by Valerie Biousse



VISUAL FIELD- HUMPHREY

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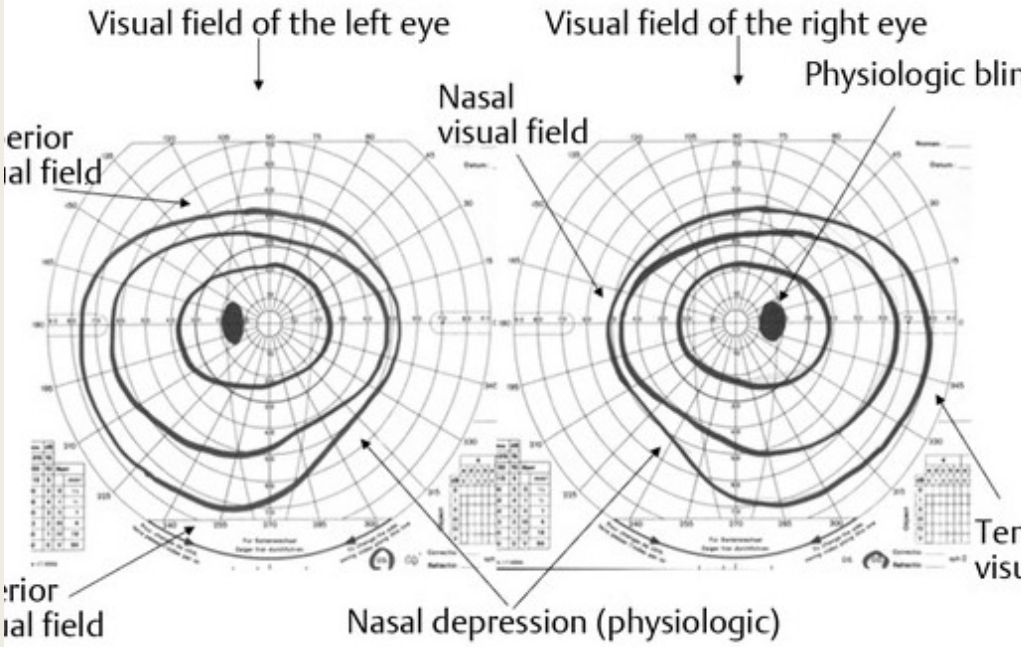


GOLDMAN VISUAL FIELD

Neurological history and exam

- *Headaches: Positional , worse on lying down or bending down*
- *Tinnitus : Pulsatile/Buzzing*
- *Transient loss of vision*
- *Diplopia: Horizontal with 6^{th} nerve palsy (aka False localizing signs)*
- *Recent weight gain or loss, fever, chills*
- *Recent infections*
- *Photophobia /Phonophobia- non-specific*

Work up -Neuro Imaging /Investigations

MRI Brain /Orbits w/wo gadolinium *CT Head*	*Consider MRV* *MRA*	*Lumbar puncture with opening pressure and chemical study*
Serologies for inflammatory conditions	*Selected cases :* *CT angiography* *CT Chest /Abdomen/Pelvis*	*Beware of mimickers – Hyperension , diabetes, Hypercholesterolemia*

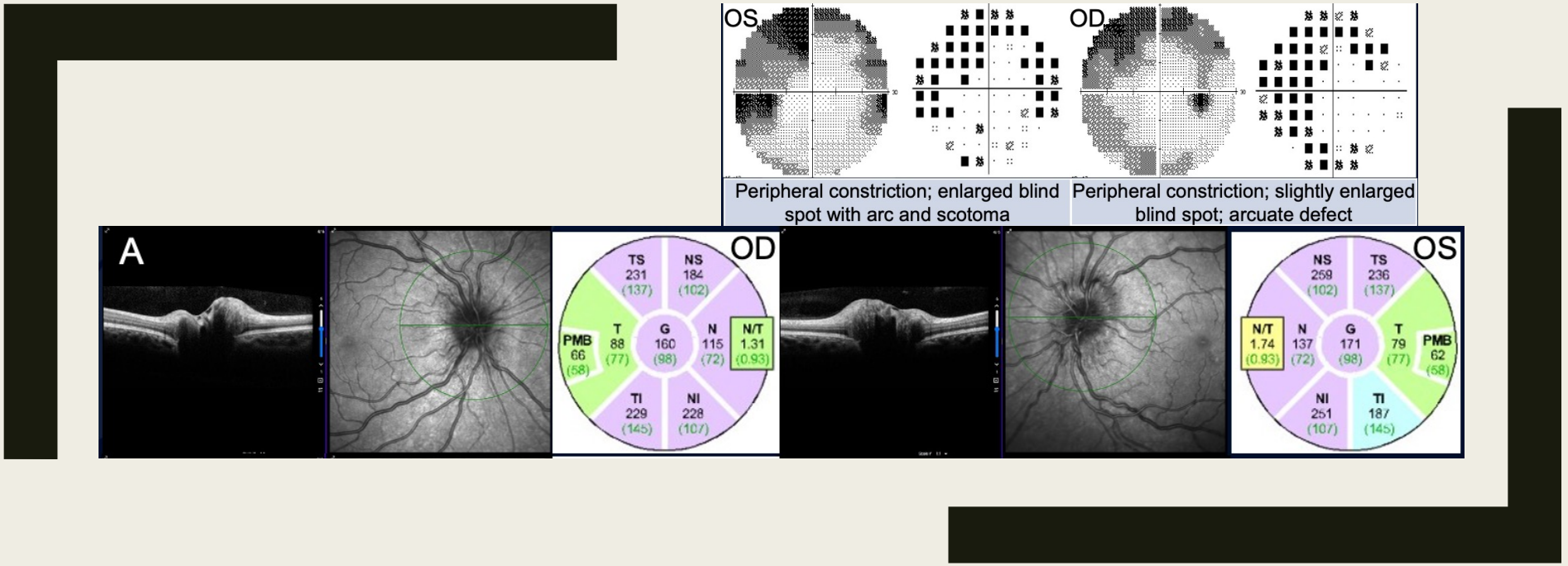
Treatment

- *Treat the underlying cause - increased Intracranial pressure or optic nerve inflammation or retinal/systemic etiology*
- *Medical , surgical treatment to prevent visual loss*
- *Treat blood pressure , diabetes, underlying inflammatory condition*

CASES

29-year-old Hispanic female with new-onset headaches.

- *Vitals normal*
- *BMI 21.22 kg/m²*
- *10 lbs weight gain*
- *VA 20/20 -1 OU*
- *Night sweats*



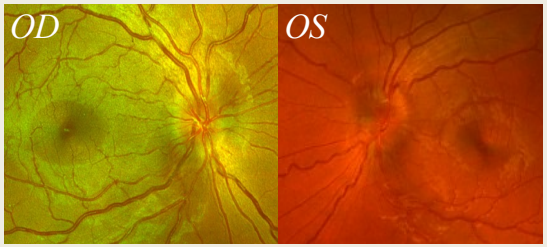
VISUAL FIELD AND OCT

IS THIS IIH?

IIH-Modified Dandy Criteria

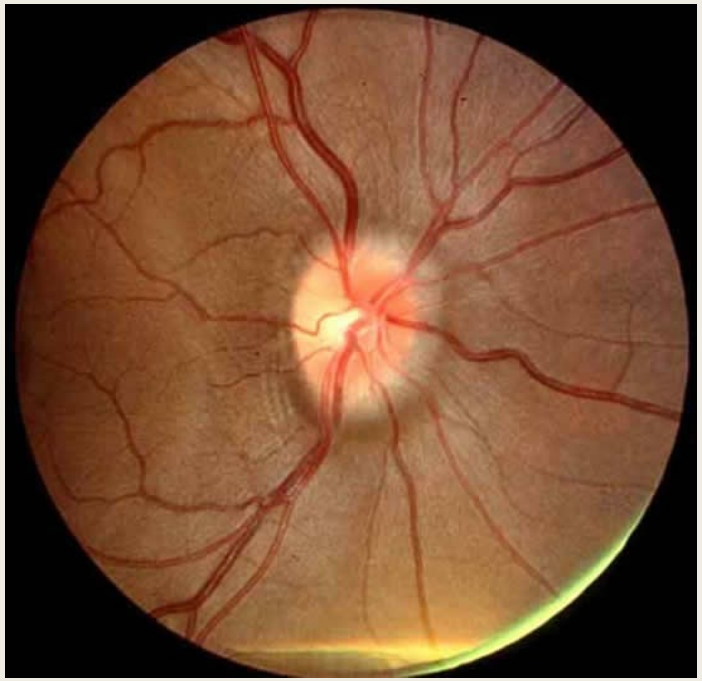
- *Raised intracranial pressure (ICP): headache, nausea, vomiting, transient visual obscurations, or papilledema*
- *No localizing signs with the exception of abducens (sixth) nerve palsy*
- *The patient is awake and alert*
- *Normal CT/MRI findings without evidence of thrombosis*
- *LP opening pressure of >25 cmH2O and normal biochemical and cytological composition of CSF*
- *No other explanation for the raised intracranial pressure*

Identifying Papilledema



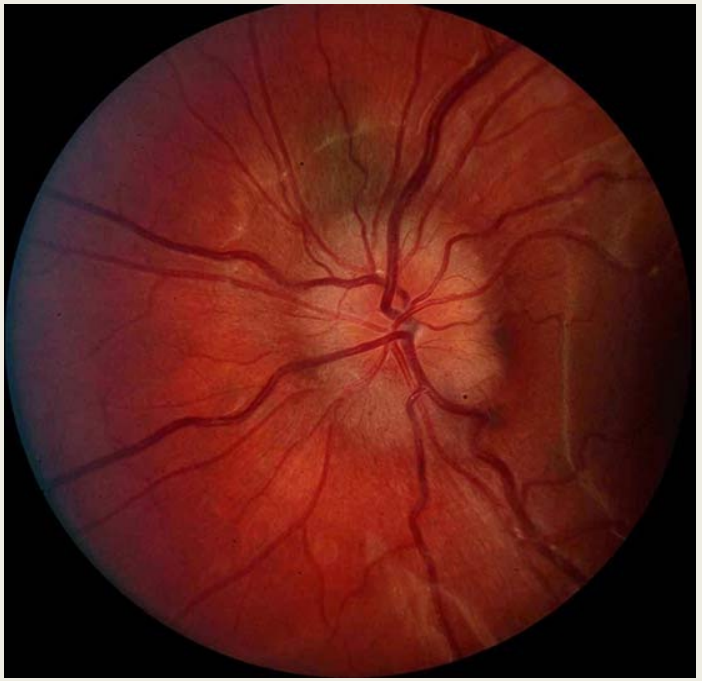
Grade I

C-shaped halo with a temporal gap- normal temporal disc margins



Grade II

the halo becomes circumferential, temporal margins blurr

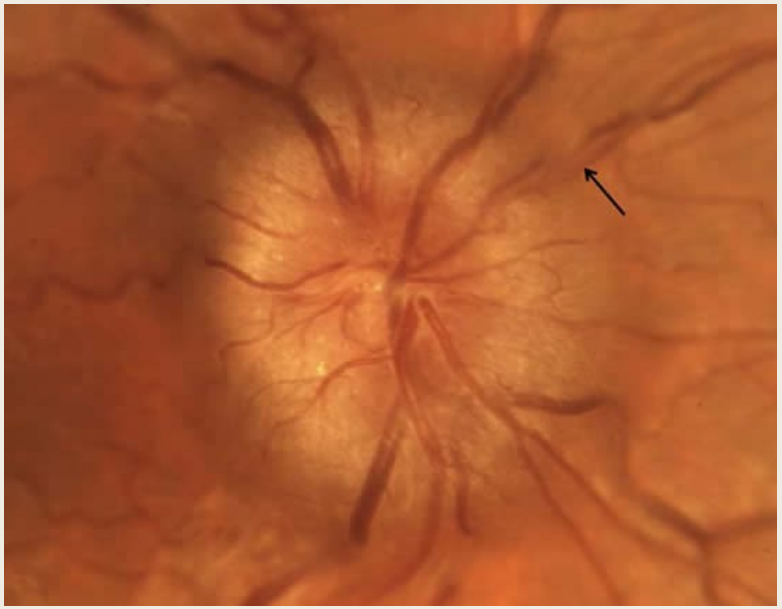


http://webeye.ophth.uiowa.edu/eyeforum/cases/papilledema-grading.htm

Identifying Papilledema

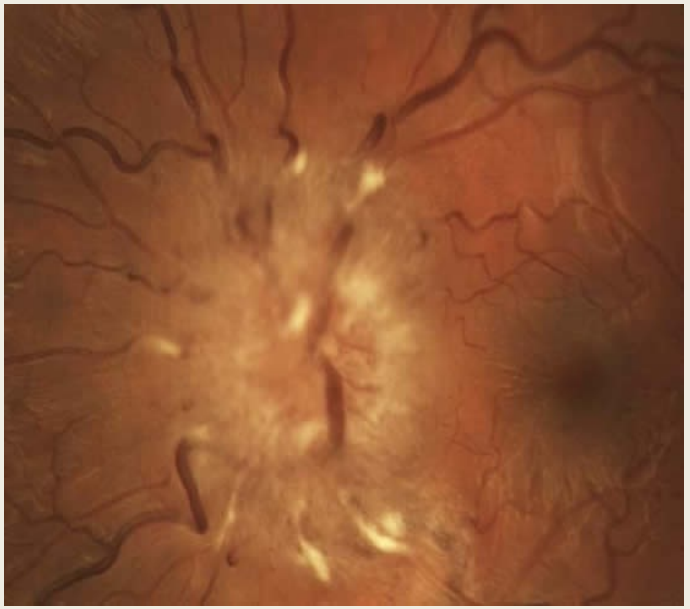
Grade III

Loss of major vessels AS THEY LEAVE the disc (arrow)



Grade IV

Loss of major vessels ON THE DISC.



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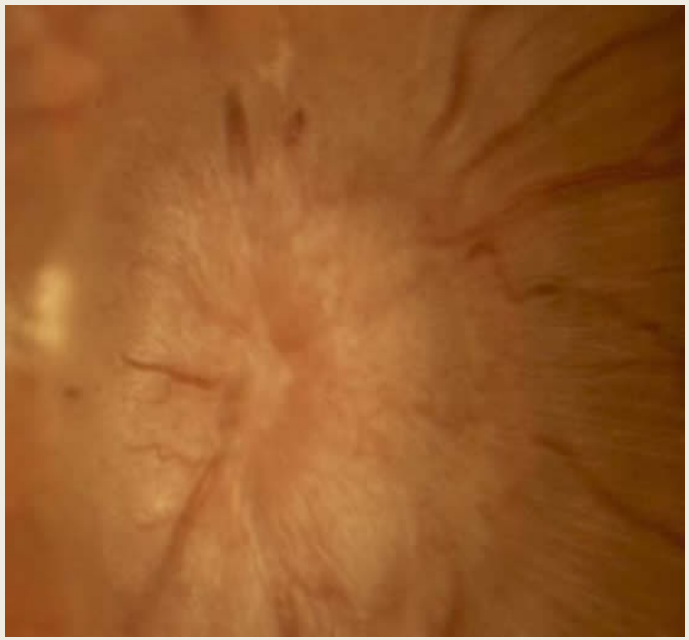
Identifying Papilledema

Grade V

Grade IV plus partial or total obscuration of all vessels of the disc

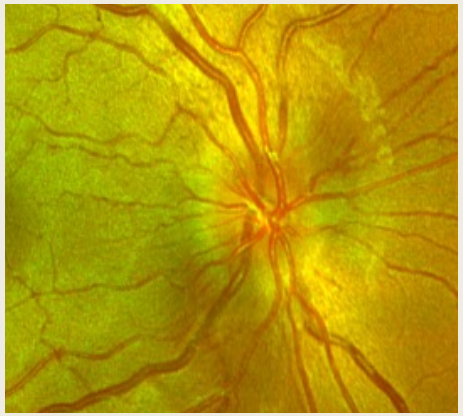
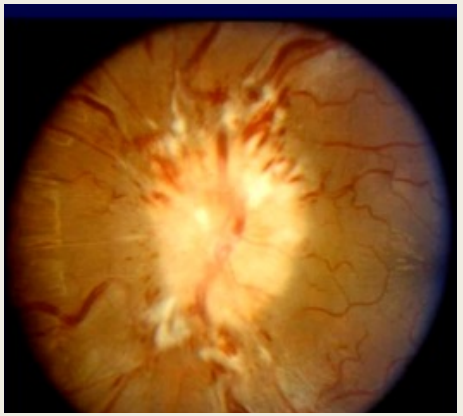
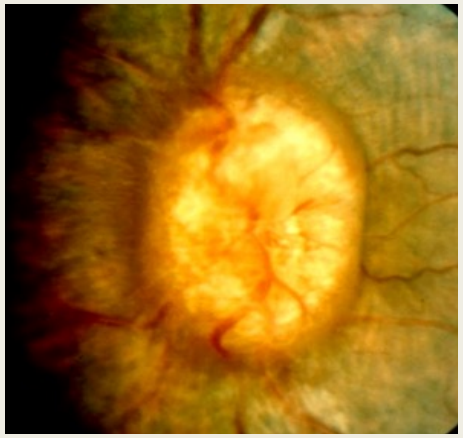


Optic Atrophy

http://webeye.ophth.uiowa.edu/eyeforum/cases/papilledema-grading.htm

Identifying Papilledema -A different way

- *Early : Hyperemia, superior /inferior edema*
- *Acute: Hemorrhages, infarcts, retinal folds*
- *Chronic: Champagne cork appearance*
- *Atrophic: Pallor, arteriolar narrowing*

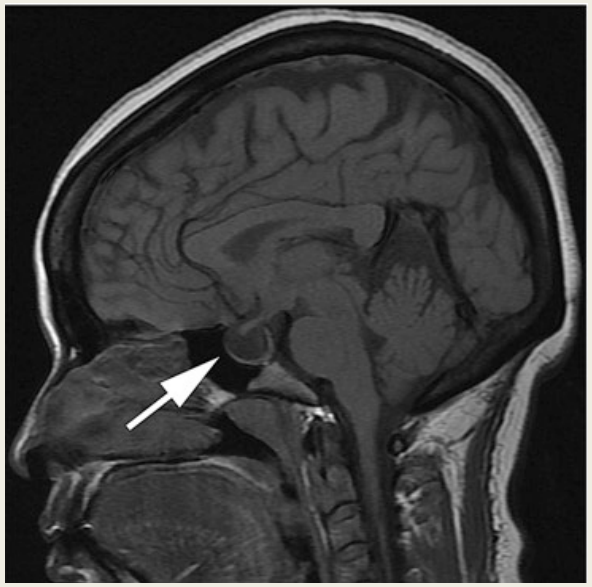





MRI Findings of increased ICP

1. Flattening of the posterior pole of the eyes

2. Dilation and tortuosity of the optic nerve sheaths,

3. Empty sella turcica



4. Stenosis of one or both transverse cerebral venous sinuses

Accuracy of MRI Findings

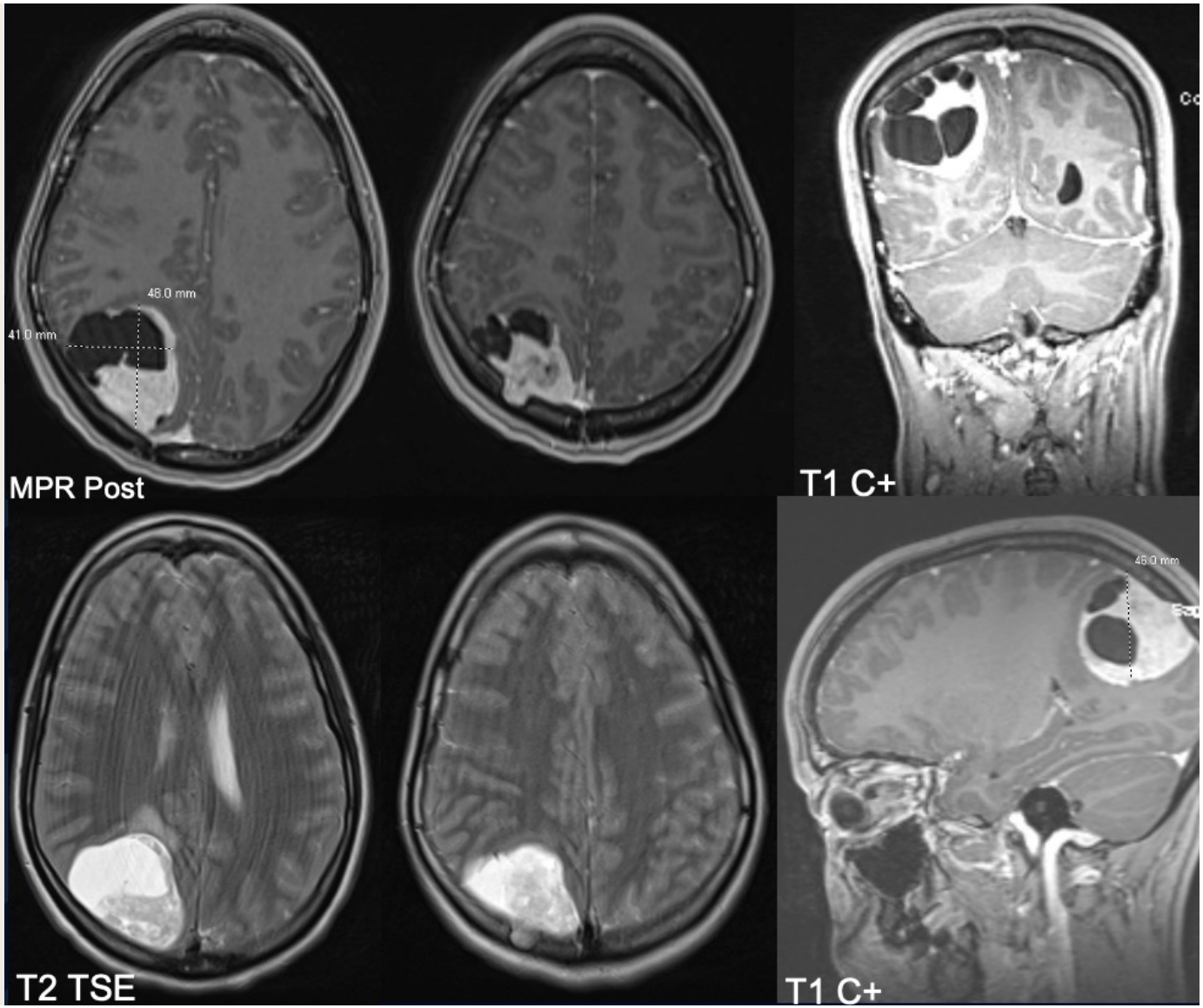
- *Each neuroimaging finding differs in its sensitivity and specificity when reported individually and when found in combination with other signs*
- *None of the findings discussed have both high sensitivity and specificity when isolated*
- *It is important to remember that the positive or negative predictive value, which is most relevant for diagnosis, will depend on the prevalence in the population being studied*

MRI finding	References	Sensitivity (%)	Specificity (%)
Empty sella	[18]	80[a]	83[a]
	[9]	26.7	94.6
	[25]	70	95
Partially empty sella	[10]	65.1	95.3
	[9]	53.3	75
Optic nerve tortuosity	[9]	43[a]	90[a]
	[10]	34.9	86
	[9]	40	91.1
	[25]	40	95
Distended optic nerve sheath	[9]	58[a]	89[a]
	[10]	48.8	88.4
	[9]	66.7	82.1
	[25]	45	95
Optic nerve head protrusion	[10]	37.2	100
	[9]	3.3	100
	[25]	30	95
Posterior globe flattening	[10]	53.5	100
	[9]	43.3	100
	[25]	80	95
Slit-like ventricles	[10]	39.5	79.1
	[9]	3.3	100
Transverse sinus stenosis	[20]	93	–
CSS < 4	[10]	62.8	100

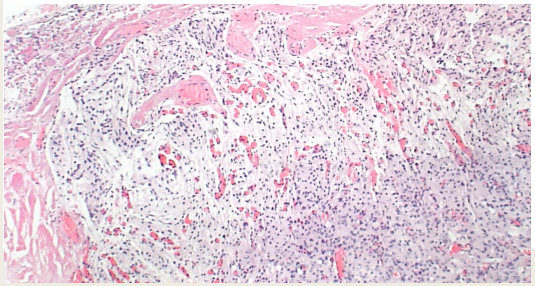
CSS, Combined transverse sinus stenosis score.
[a]Pooled data from multiple studies.

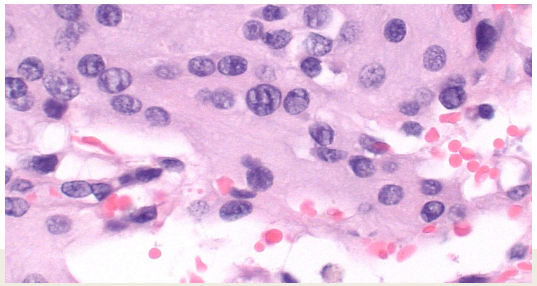
MRI brain w/ w/o contrast

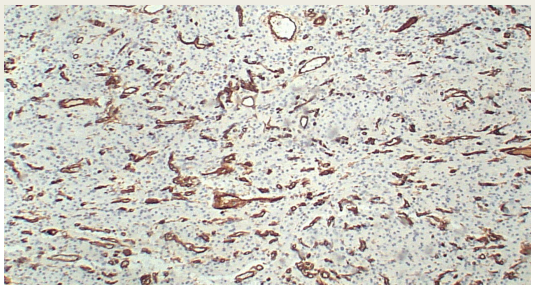
Coming back to the case ….

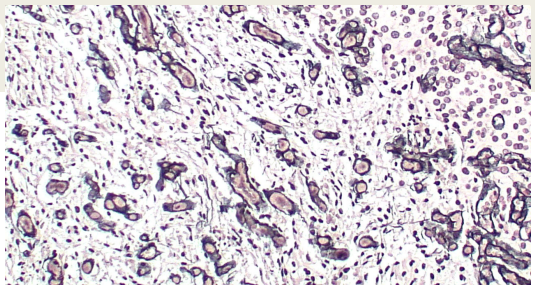


PATHOLOGY









Grade I meningioma with features of two rare variants, angiomatous and microcystic meningioma

Treatment

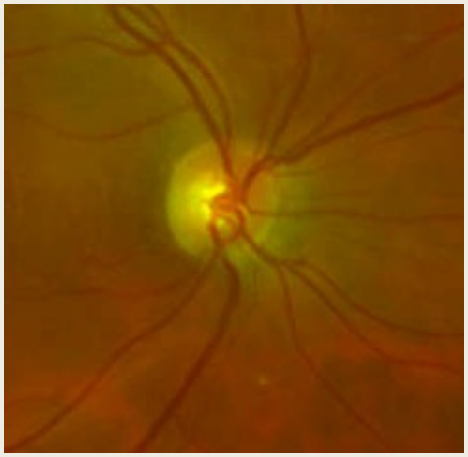
- *The patient underwent craniotomy 1 month after presentation*
- *The tumor was highly vascular and had invaded pial boundaries*
- *En bloc resection was performed without complications*

Pearls

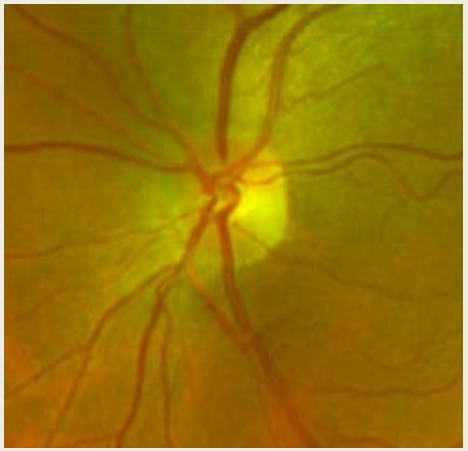
This case highlights the importance of ruling out structural causes of elevated intracranial pressure prior to lumbar puncture, as well as maintaining vigilance for red flag symptoms.

35 YOF, Caucasian, with right eye blurry vision w/ headaches

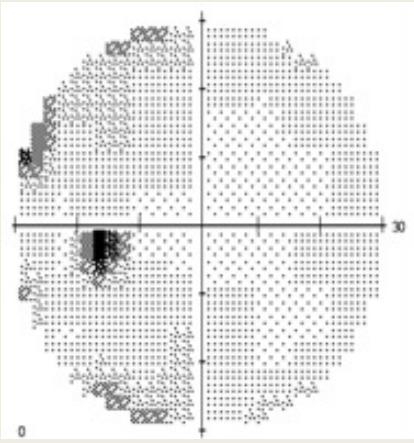
OD RNFL 88
VA 20/40 1+APD



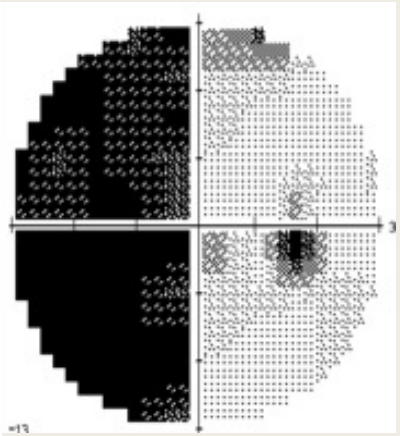
OS RNFL 94
VA 20/20 No APD



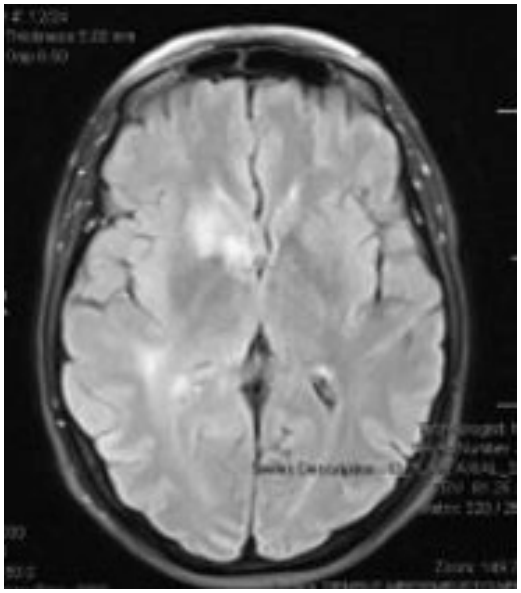
OS



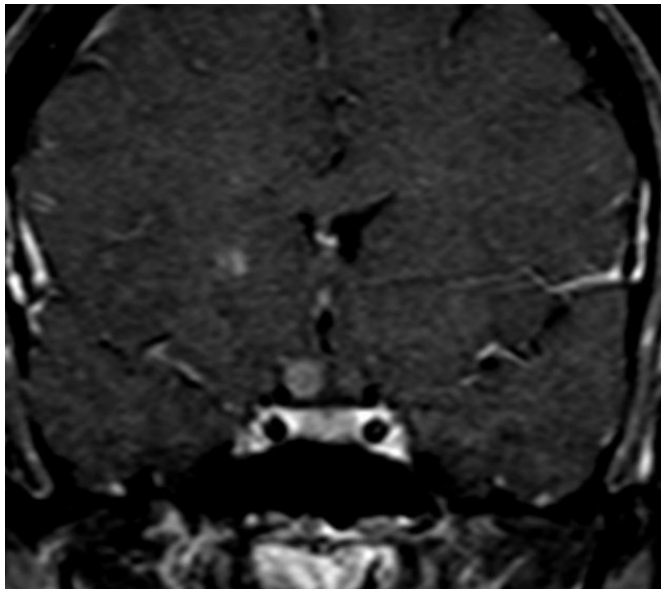
OD



T 2 FLAIR



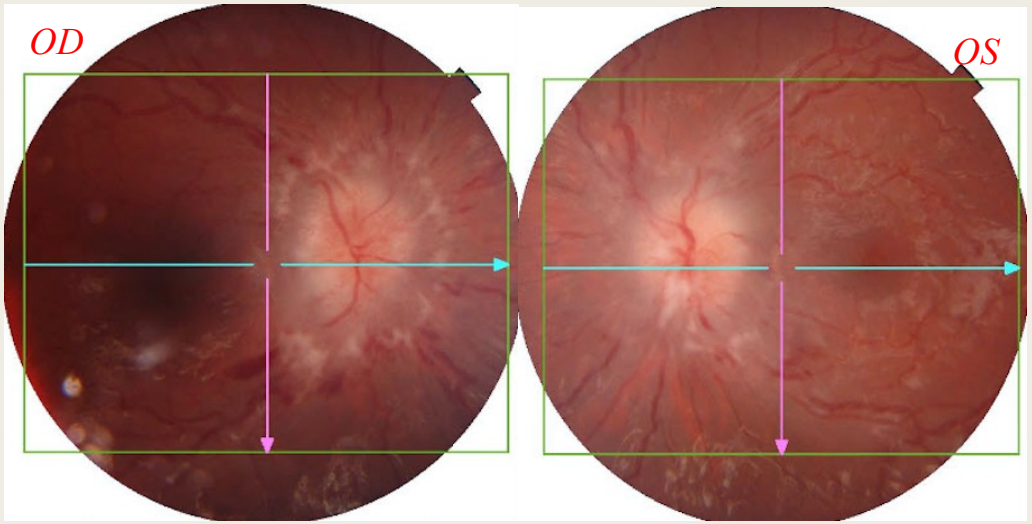
T1 Coronal w/ Contrast

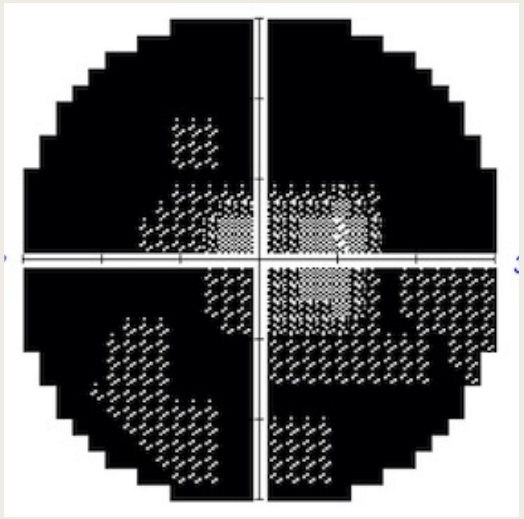


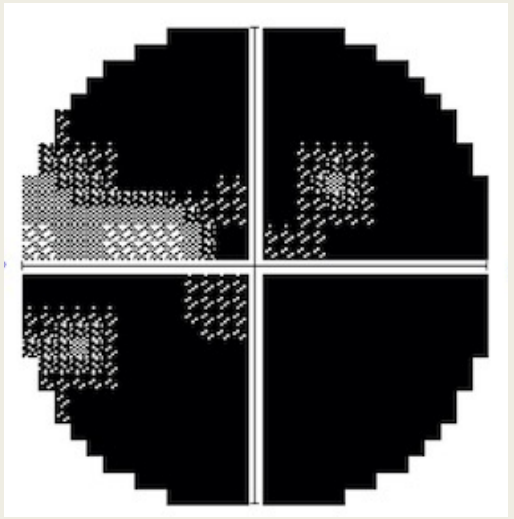
Strongly Recommend Obtaining Visual Fields when patients present with visual loss

Brain Imaging

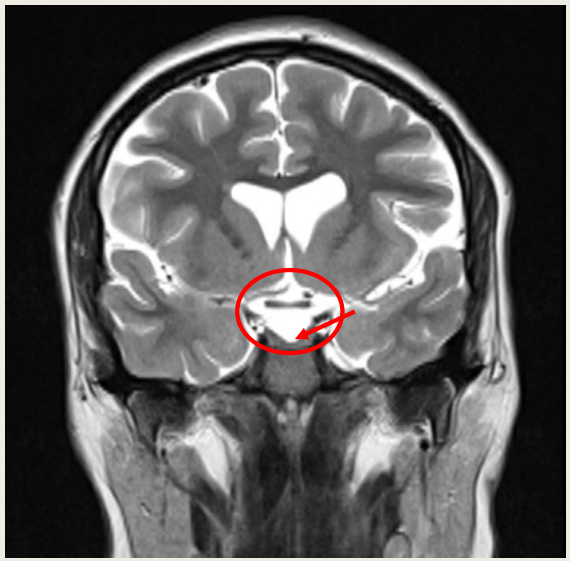
15-year-old girl with new onset vision loss and headaches presented to the ED







MRI Brain W/o Contrast



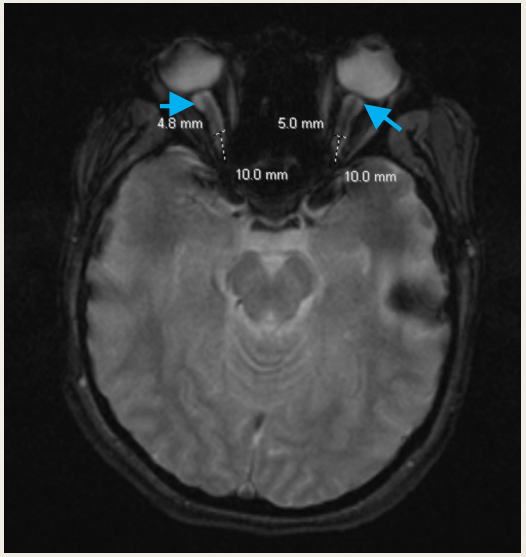


Figure 4: *(1) Coronal T2-weighted MRI demonstrating partially empty sella turcica (red circle) and pituitary gland (red arrow); (2) Mid-sagittal T1-weighted MRI demonstrating partially empty sella turcica (red circle) and pituitary gland (red arrow); (3) Axial fluid-attenuated inversion recovery (FLAIR) demonstrating posterior globe flattening (blue arrows) and optic nerve sheath diameter (ONSD) widening (measured 10 mm anterior to optic foramen) – right ONSD 4.8 mm, left ONSD 5.0 mm*

Impression: *Partially empty sella with no ventriculomegaly and bilateral optic nerve head fullness is consistent with IIH*

Previous studies looking at the optic nerve sheath diameter (ONSD) as an auxiliary tool for diagnosing IIH

- ✓ *ONSD > 4mm suggestive of increase intracranial pressure*

MRV

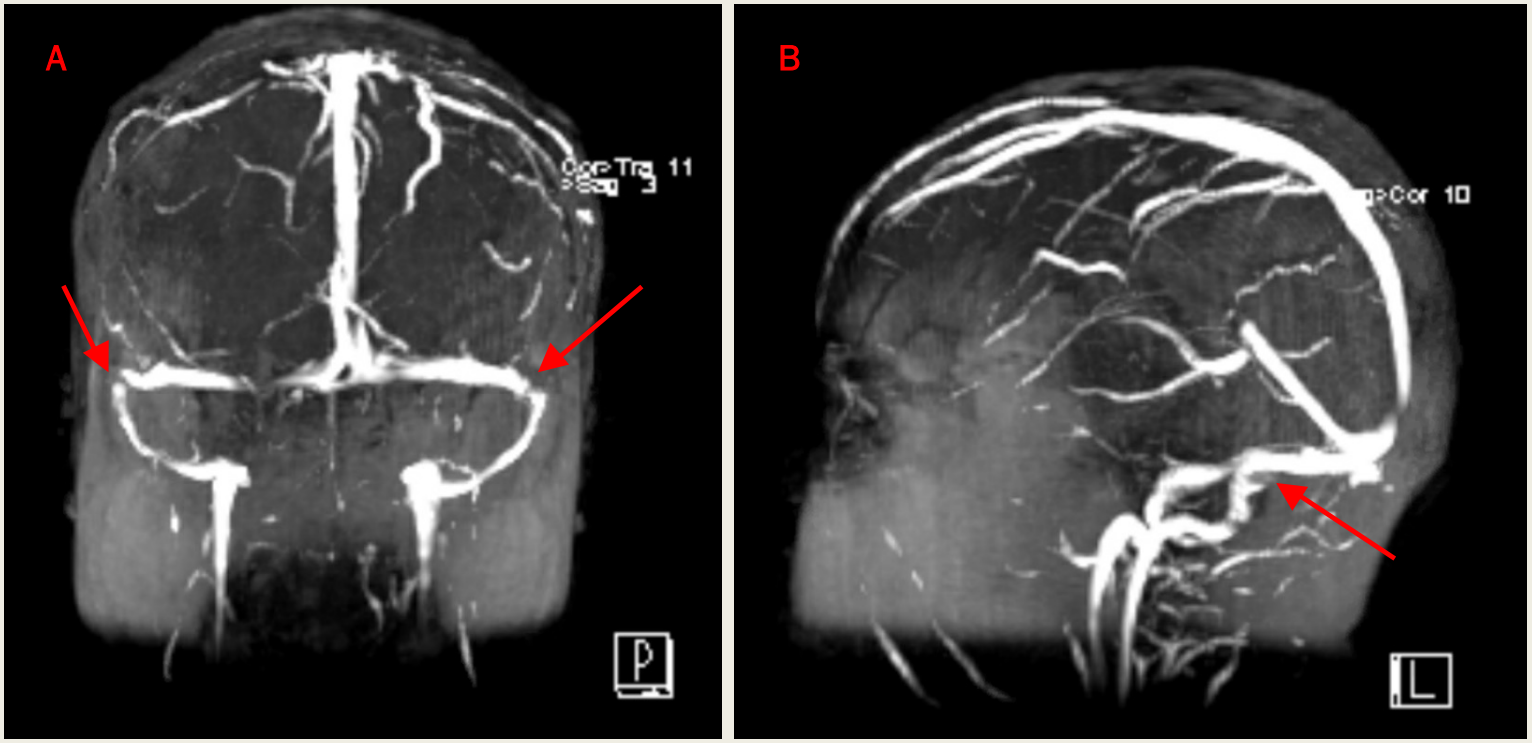


Figure 5: *(A) Magnetic resonance venography (MRV) posterior view demonstrating bilateral transverse sinus stenosis (arrow); (B) MRV lateral view demonstrating left transverse sinus stenosis.*

Impression: *Bilateral transverse sinus stenosis is consistent with IIH*

LP OPENING PRESSURE

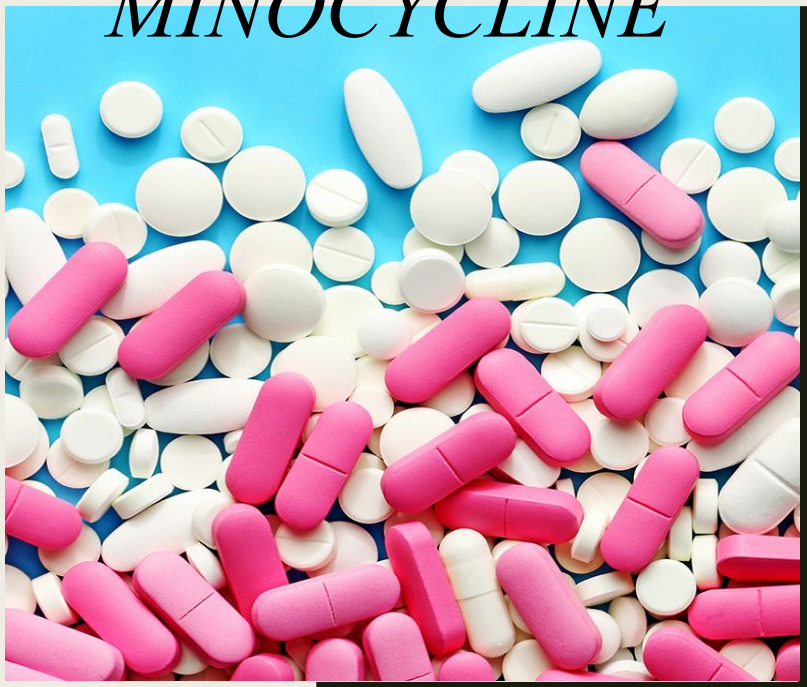
>55CM H2O

Diagnosis : IIH

Normal BMI

History ?

MINOCYCLINE



Treatment

- *Acetazolamide 500 → 750 mg by mouth (PO) twice per day (BID)*
- *Fulminant Papilledema w/ no improvement in vision with Acetazolamide → warranted surgical LPS*
- *Presented 4 months later for worsening vision*
- *Found to have shunt migration - shunt revision*

Lumboperitoneal Shunt (LPS)

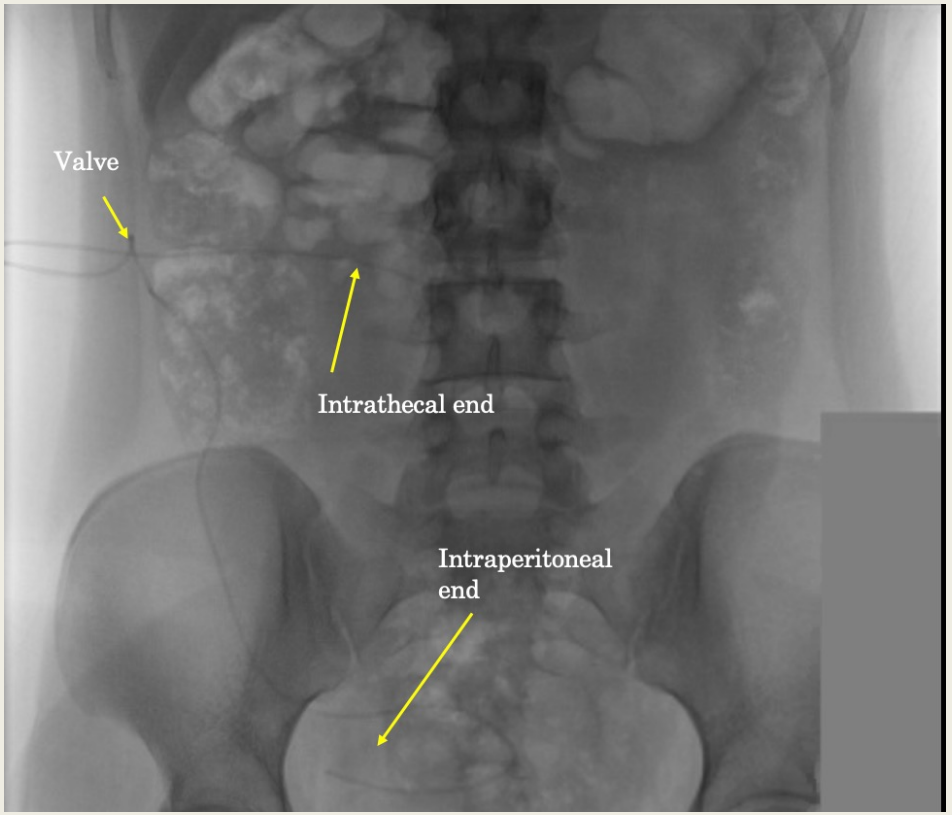
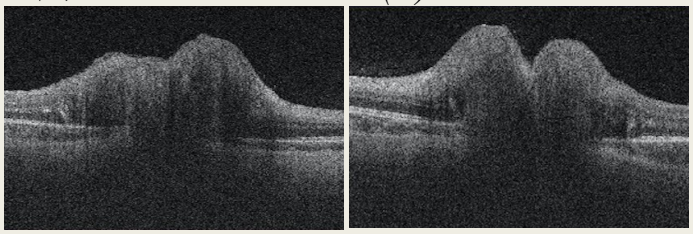


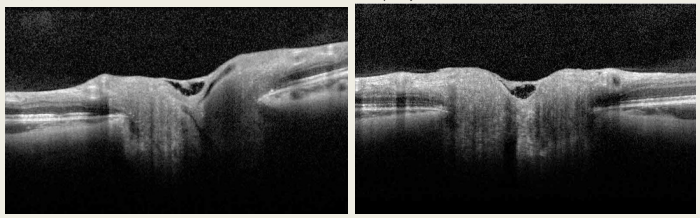
Figure 6: *Anterior-posterior view of abdominal X-ray demonstrating lumbar catheter, valve, and peritoneal catheter (arrows).*

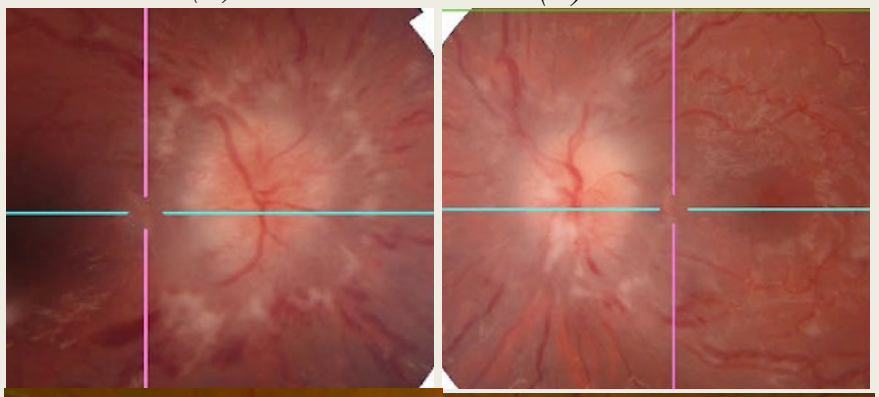
Optical coherence Tomography (OCT)

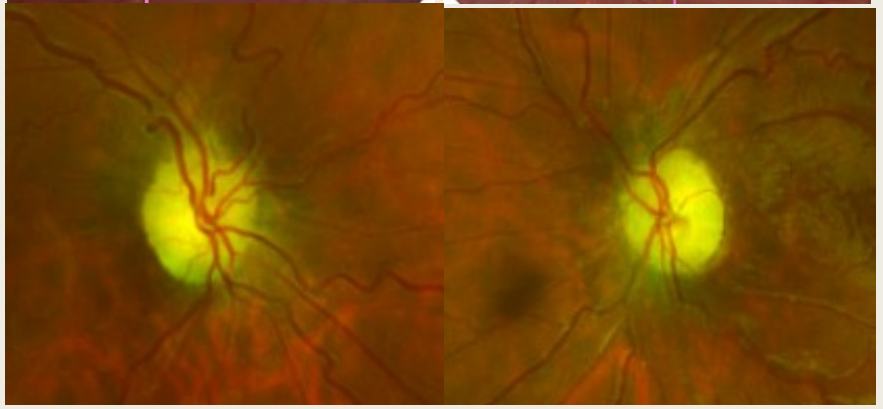
(A) OD *(A) OS*



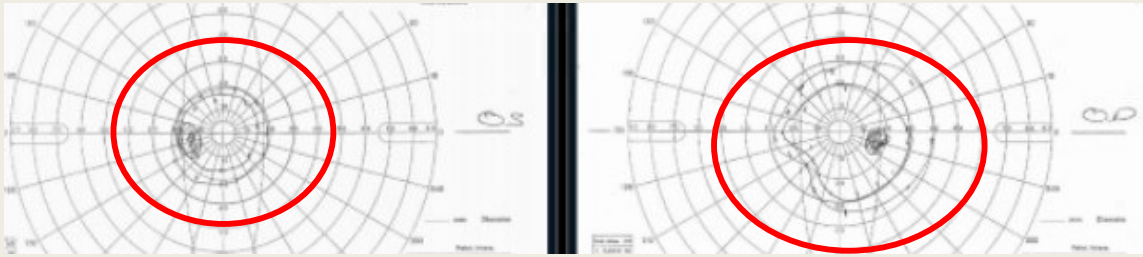
(B) OD *(B) OS*







Average RNFL thickness	OD	OS
(A) At presentation	364	377
(B) 5 months post-treatment	63	61



Case Reports > BMJ Case Rep. 2023 Mar 28;16(3):e252731. doi: 10.1136/bcr-2022-252731.

Optic atrophy secondary to minocycline–induced idiopathic intracranial hypertension

Ricky Paramo [1], Lakshmi Leishangthem [2]

Affiliations + expand

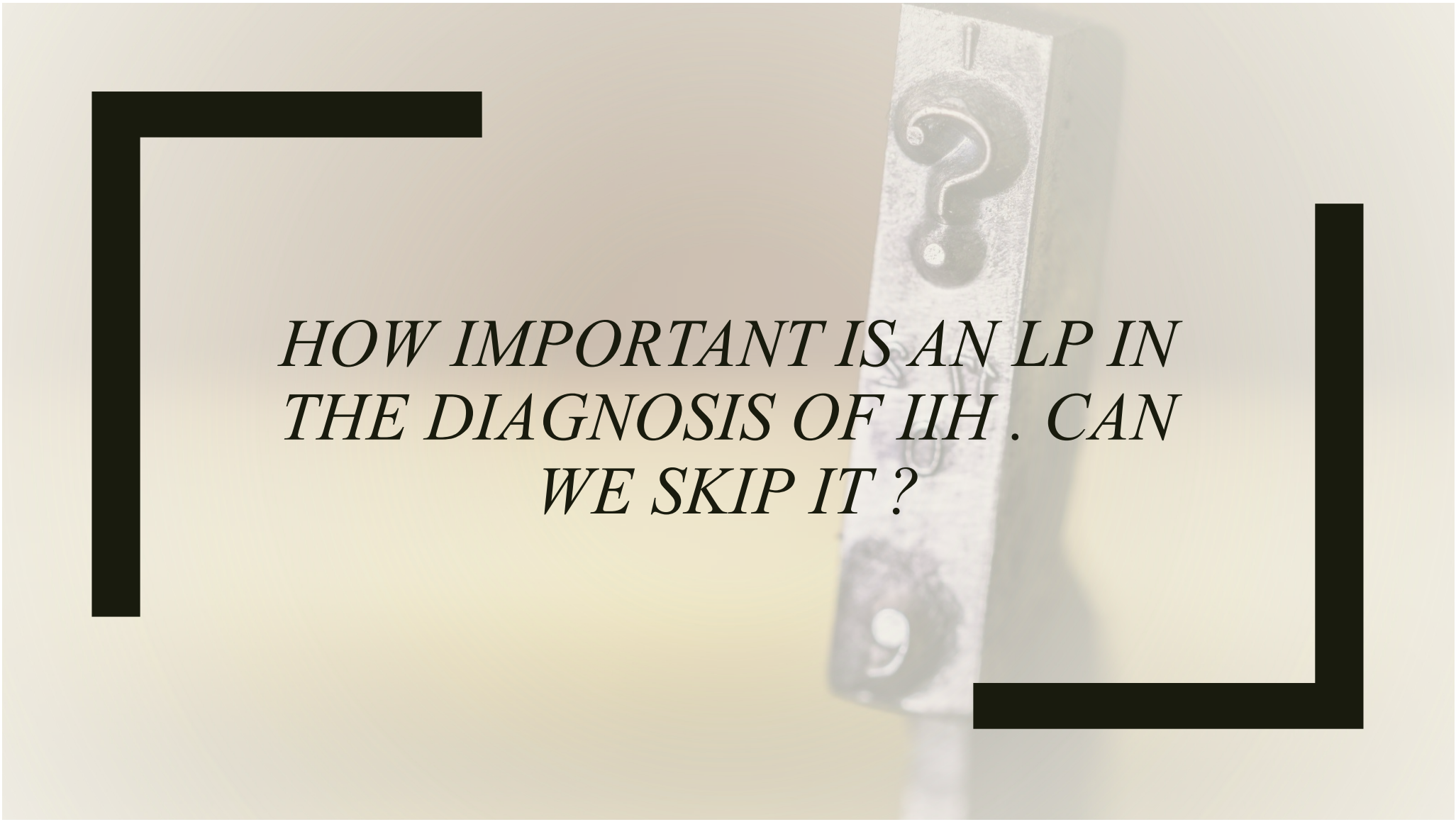
PMID: 36977510 PMCID: PMC10069554 (available on 2025-03-28)
DOI: 10.1136/bcr-2022-252731

Abstract

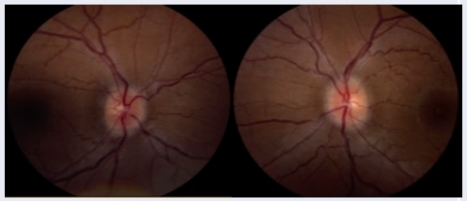
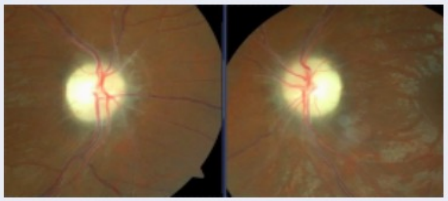
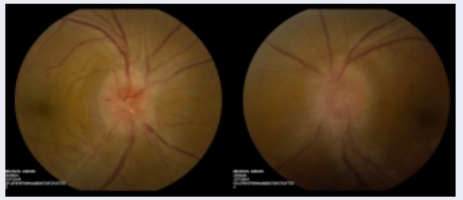
An early adolescent female presented with blurry vision, ocular 'fullness', pulsatile tinnitus and gait difficulty due to poor vision. She was found to have florid grade V papilloedema, 2 months after the use of minocycline for the treatment of confluent and reticulated papillomatosis for 2 months. MRI of the brain without contrast showed fullness of the optic nerve heads concerning for increased intracranial pressure, which was confirmed on lumbar puncture with an opening pressure greater than 55 cm H_2O. She was initially started on acetazolamide, but due to high opening pressure and severity of visual loss, a lumboperitoneal shunt was placed in 3 days. This was complicated by a shunt tubal migration 4 months later, leading to worsening vision of 20/400 in both eyes for which she underwent shunt revision. By the time she presented to the neuro-ophthalmology clinic, she was legally blind with her exam consistent with bilateral optic atrophy.

Pearls

- *Minocycline-induced IIH has been reported in the literature many times. It has a half-life of 24 hours and symptoms related to IIH are often reported during use.*
- *However, prior minocycline use can present with elevated intracranial pressures (ICP) up to 5 weeks after cessation.*
- *Visual and hearing symptoms did not present until 4-5 weeks after last minocycline dose.*



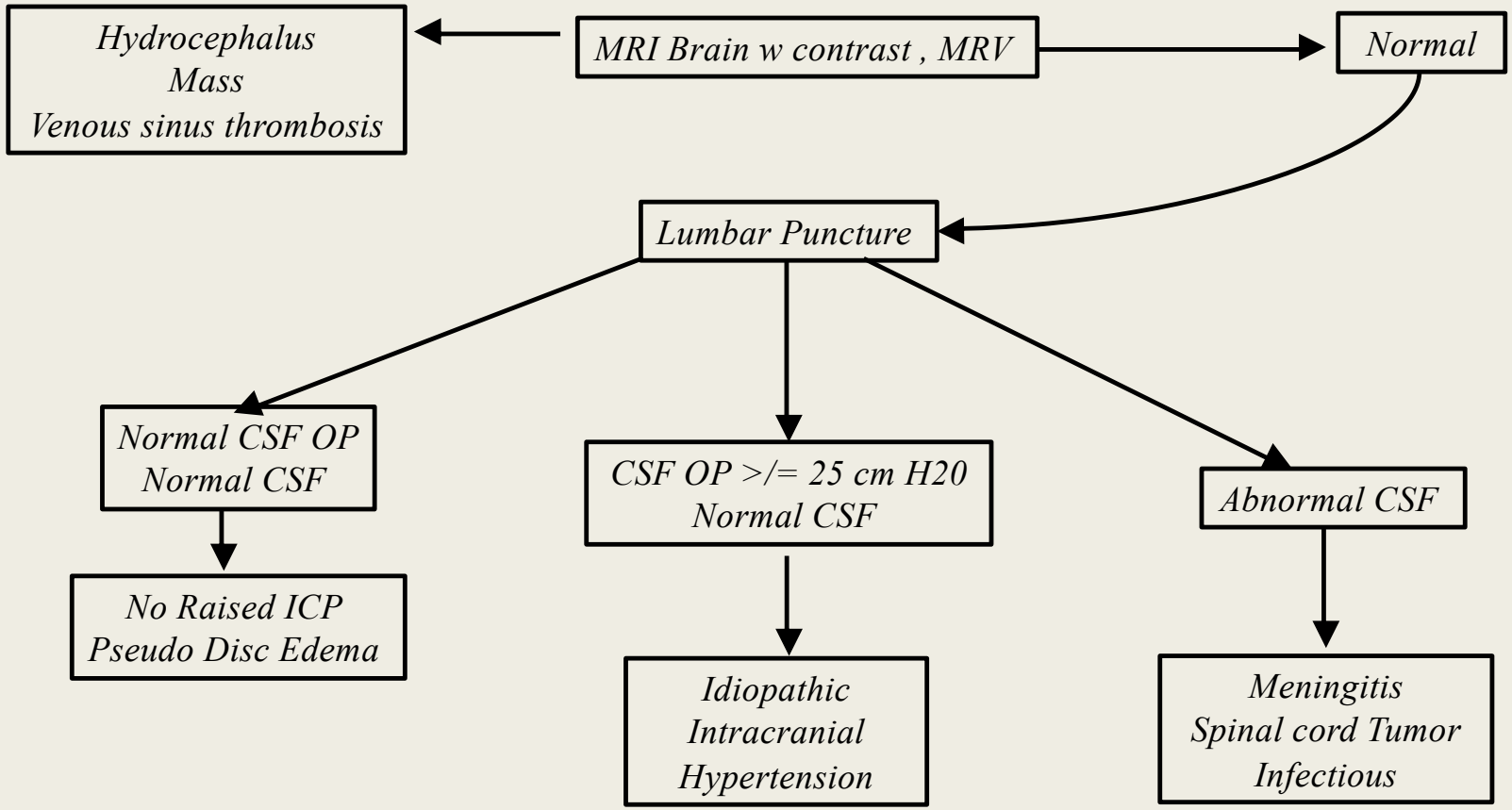
HOW IMPORTANT IS AN LP IN THE DIAGNOSIS OF IIH . CAN WE SKIP IT ?

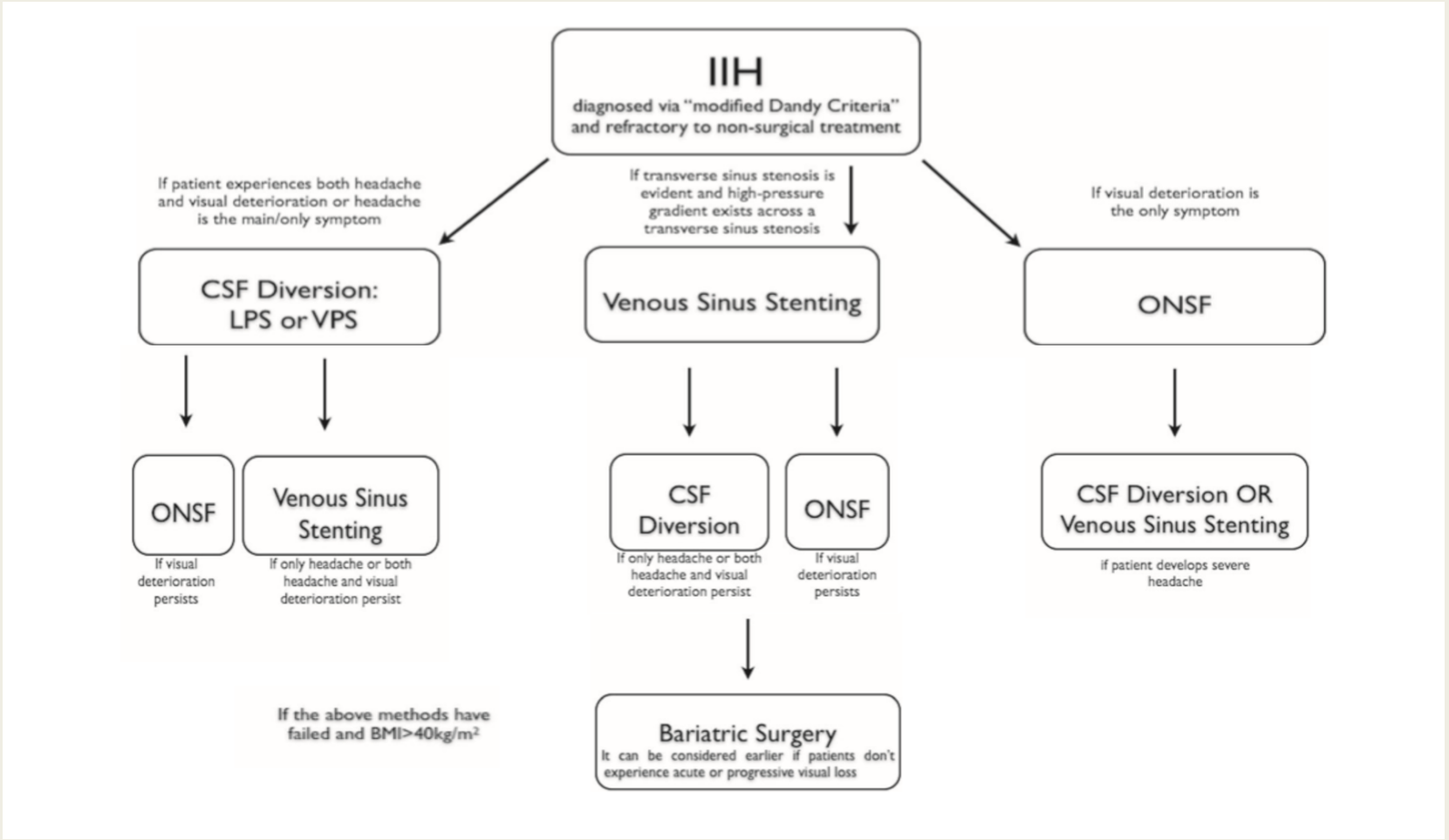
	CASE 1	CASE 2	CASE 3
Patient Description	36 year-old woman (BMI 58) c/o headaches, blurry vision x 1 month	27 year-old woman (BMI 24) c/o headaches, blurry vision, pulsatile tinnitus	55 year-old male (BMI 35) c/o headaches, blurry vision x 2 months
Fundus Photos	 Grade 2 bilateral disc edema	 Bilateral disc edema with atrophic changes	 Grade 4 bilateral disc edema
MRI/MRV	Empty sella, bilateral transverse sinus stenosis	Normal	Flattening of pituitary. Right transverse sinus stenosis
Lumbar Puncture Opening Pressure and CSF Studies	**Opening pressure**-33cm H_2O **RBC**-28 **WBC**-240 (75% lymphocytes) **Glucose**-43 **Protein**-163 VZV negative HSV positive	**Opening pressure**-55cm H_2O **RBC**-0 **WBC**-55 (30% atypical cells) **Glucose**-46 **Protein**-55 **Cytosin prep w/ papanicolou stain:** small blue cell tumor	**Opening pressure**-32cm H_2O **RBC**-0 **WBC**-6 **Glucose**-normal **Protein**-56 VDRL, CMV, EBC, HSV negative RPR titer 1:64, + syphilis IgG/IgM
Other studies	None	Immunohistochemistry + for CD56, synaptophysin and chromogranin. Negative for CKAE1/AE3 (carcinoma stain), GFAP, Olig2, melanin-A, HMB 45 and SOX10 (melanoma stain)	Serum HIV positive, CD4 count 387
Final Diagnosis	**HSV meningitis**	**Primary CNS neuroendocrine tumor**	**Neurosyphilis in the setting of new HIV diagnosis**

Pearls

- *In the work-up of bilateral disc edema, a lumbar puncture with opening pressure and cerebrospinal studies (CSF) should always been obtained*
- *Forgoing the lumbar puncture can lead to devastating consequences if a deadly diagnosis is missed and treatment is delayed*

Bilateral Disc edema +/- Headache

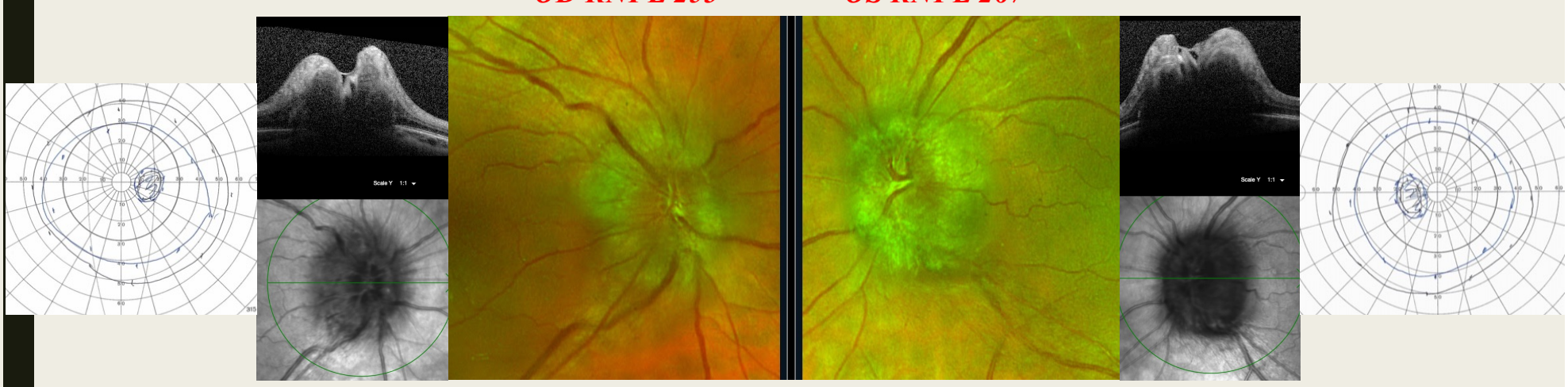



IIH
diagnosed via "modified Dandy Criteria" and refractory to non-surgical treatment
If patient experiences both headache and visual deterioration or headache is the main/only symptom
If transverse sinus stenosis is evident and high-pressure gradient exists across a transverse sinus stenosis
If visual deterioration is the only symptom
CSF Diversion: LPS or VPS
Venous Sinus Stenting
ONSF
ONSF
If visual deterioration persists
Venous Sinus Stenting
If only headache or both headache and visual deterioration persist
CSF Diversion
If only headache or both headache and visual deterioration persist
ONSF
If visual deterioration persists
CSF Diversion OR Venous Sinus Stenting
If patient develops severe headache
If the above methods have failed and BMI>40kg/m²
Bariatric Surgery
It can be considered earlier if patients don't experience acute or progressive visual loss

Cases .. IIH and Pregnancy

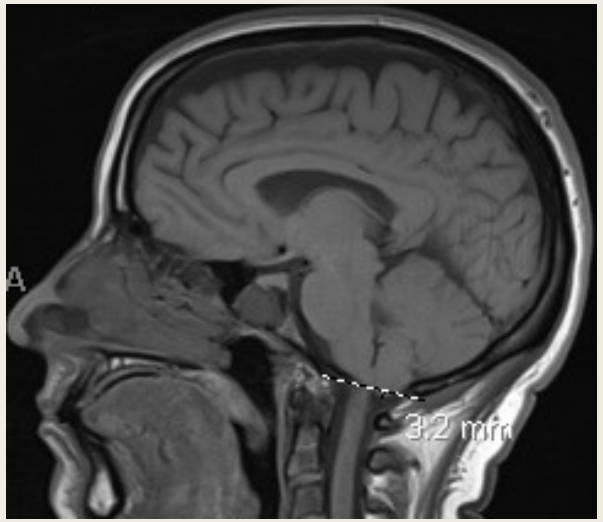
- *41 YOF pregnant 22 weeks with severe positional headaches and tinnitus*
- *Bedside OP: 29 cm of water*
- *Treated with Acetazolamide 500 mg BID*

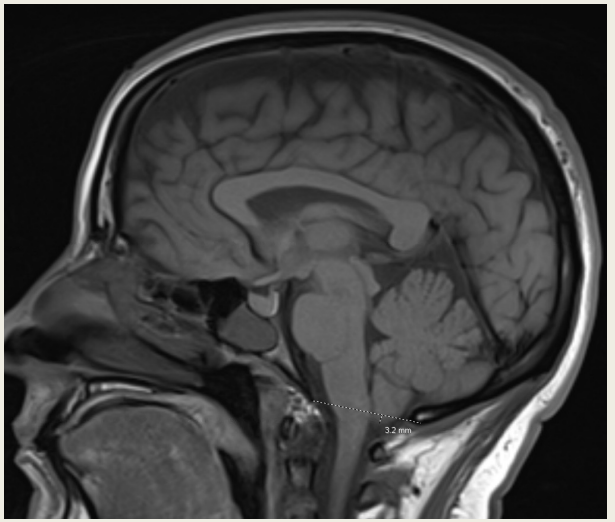
OD RNFL 253 ***OS RNFL 267***

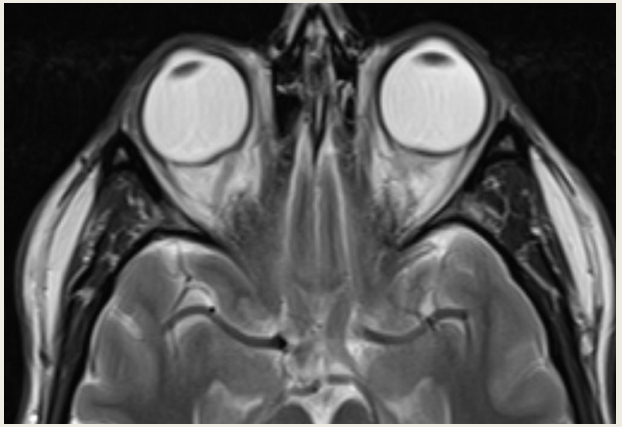


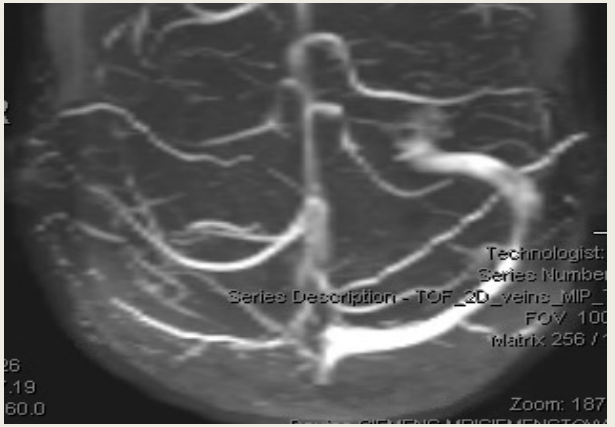
MRI Brain : Low lying cerebellar tonsil

MRV – B/L Transverse Sinus stenosis









IIH and Pregnancy

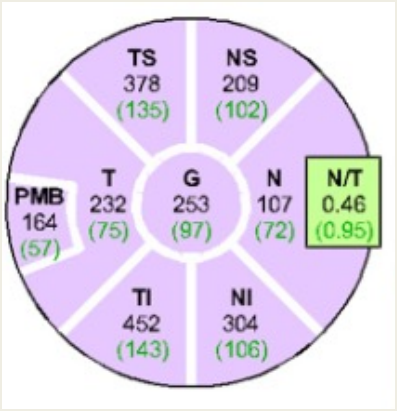
- *CTV Neg for Clots, No aneurysm on CTA*
- *DVT : Positive clot right distal femoral vein.*
- *Shortness of breath - PE studies : Positive for Pulmonary embolism. Started on therapeutic Neutropenia after starting Enoxaparin*
- *Acetazolamide discontinued.*
- *In two months , she had transient loss of vision, worsening papilledema and other symptoms.*
- *Acetazolamide resumed at 500 mg and delivery done at 32 wks.*
- *LP just before C-section was 17 cm of water.*

IIH and Pregnancy

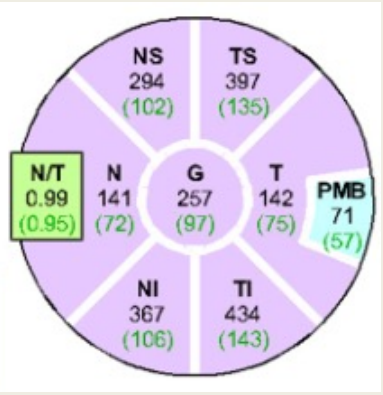
- *MRI Brain : Low lying cerebellar tonsils, MRV – B/L Transverse Sinus stenosis*
- *CTV Neg for Clots, No aneurysm on CTA*
- *DVT : Positive clot right distal femoral vein.*
- *Shortness of breath - PE studies : Positive for Pulmonary embolism. Started on therapeutic Neutropenia after starting Enoxaparin*
- *Acetazolamide discontinued.*
- *In two months , she had transient loss of vision, worsening papilledema and other symptoms.*
- *Acetazolamide resumed at 500 mg and delivery done at 32 wks.*
- *LP just before C-section was 17 cm of water.*

At presentation

OD RNFL 253

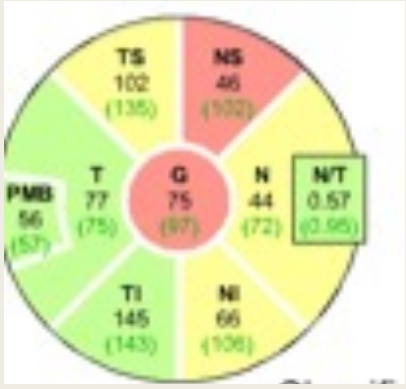


OS RNFL 267



3 months later

OD RNFL 75



OS RNFL 73

IIH and Pregnancy

Idiopathic Intracranial Hypertension in Pregnancy

Daniel S.J. Park, MSc;[1] John S.Y. Park, MD;[2,3] Sapna Sharma, MD;[4] Rahul A. Sharma, MD, MPH[5]

[1]Faculty of Medicine, University of Toronto, Toronto, ON

[2]Department of Ophthalmology, Ottawa Eye Institute, The Ottawa Hospital, Ottawa, ON

[3]Faculty of Medicine, University of Ottawa, Ottawa, ON

[4]Department of Obstetrics and Gynecology, McMaster University, Hamilton, ON

[5]Department of Ophthalmology & Vision Sciences, University of Toronto, Toronto, ON

- *IIH in pregnant patients : Same rate similar to that in the general population of similar sex and age.*
- *Subsequent pregnancies: Not a risk factor for IIH recurrence.*
- *Coexistence of pregnancy and IIH : Not associated with poorer visual prognosis*
- *Workup : Similar to that of non-pregnant patients.*
- *Lumbar puncture: Not contraindicated*
- *CT imaging may have harmful effects on the fetus and should be avoided where possible.*
- *MRI/MRV : Rule out space-occupying lesions or mimics, such as CVT*
- *Contrast agents : Case by case – Only if benefits clearly outweigh the risks.*
- *Cerebral venous sinus thrombosis should be considered as a cause of intracranial hypertension given the hyper coagulability that occurs during pregnancy.*

IIH and Pregnancy

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[3]Faculty of Medicine, University of Ottawa, Ottawa, ON

[4]Department of Obstetrics and Gynecology, McMaster University, Hamilton, ON

[5]Department of Ophthalmology & Vision Sciences, University of Toronto, Toronto, ON

- *Active labour in patients with IIH :Not associated with a higher risk of vision loss.*
- *Vaginal delivery in patients with IIH: Not associated with worse maternal outcomes.*
- *Coexistence of IIH and pregnancy: Not associated with worse maternal or fetal outcomes.*
- *Current evidence suggests that the mode of delivery should be determined by obstetrical indications only, regardless of IIH diagnosis.*

Teratogenic effects of Acetazolamide?

C	Animal studies have shown an adverse effect, and there are no adequate and well-controlled studies in pregnant women, *or* no animal studies have been conducted, and there are no adequate and well-controlled studies in pregnant women.

The Use of Acetazolamide in Idiopathic Intracranial Hypertension During Pregnancy

ANDREW G. LEE, MD, MISHA PLESS, MD, JULIE FALARDEAU, MD,
TAMMY CAPOZZOLI, COPT, MICHAEL WALL, MD, AND RANDY H. KARDON, PHD, MD

CONCLUSIONS: Acetazolamide at high doses may produce birth defects in animals, but there is little clinical or experimental evidence to support any adverse effect of the drug on pregnancy outcomes in humans.

If the clinical situation warrants the use of acetazolamide in IIH, then the drug probably can be offered after appropriate informed consent.

TABLE 3. Pregnancy Outcomes of Acetazolamide Treatment in Pregnancy in Patients From Selected Studies in the Literature

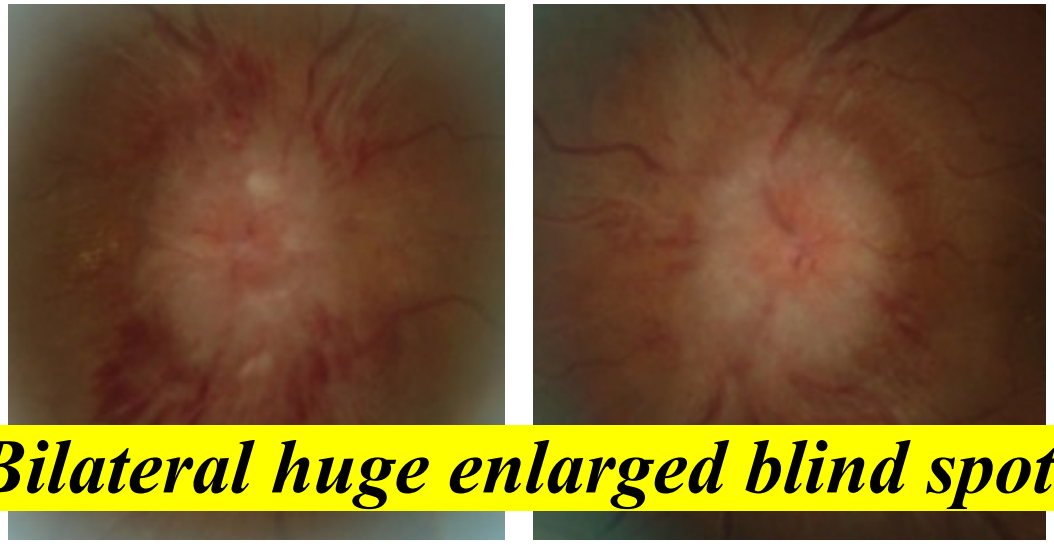
Author, Year of Study	Age (years)	Pregnancy Outcome	Trimester of Treatment With Acetazolamide	Comment
Dieckmann et al, 1957	N/A	No increase in fetal mortality and no congenital anomalies	Clinical trials of acetazolamide for preeclampsia	Therapy started after first trimester
Crane, 1957		Transient dehydration in infant	Third trimester	
Alderson, 1983	23	Normal	First	
Kassam et al, 1983	27	Normal	Second	
Ibid	32	Normal	Second	
Digre et al, 1984	23	Normal	First	Acetazolamide stopped after pregnancy discovered
Ibid	20	Normal	Second	
Ibid	24	Spontaneous abortion at 12 weeks	Second	
Worsham et al, 1978	22	Sacrococcygeal teratoma	Discontinued at 19 weeks	Treated for glaucoma
Merlob et al, 1989	29	Metabolic acidosis reversed with treatment	Treated throughout pregnancy	Treated for glaucoma
Ibid	Not reported	Two normal births	Treated throughout pregnancy	Treated for seizures
Katz et al, 1989	33	Normal		
Ibid	24	Normal	24 weeks	
Ibid	30	Normal		
Ibid	31	Normal		
Ibid	30	Normal		
Ozawa et al, 2000	Not reported	Transient renal tubular acidosis	3 days before delivery	Treated for glaucoma
Huna-Baron and Kupersmith, 2002	22	Normal		

35 YO Transgender Female, receiving Testosterone shots.

> Ophthalmic Plast Reconstr Surg. 2023 Sep-Oct;39(5):449-453.
doi: 10.1097/IOP.0000000000002344. Epub 2023 Feb 21.

Idiopathic Intracranial Hypertension in Female-to-Male Transgender Patients on Exogenous Testosterone Therapy

Naomi E Gutkind [1], David T Tse, Thomas E Johnson, Brian C Tse



Bilateral huge enlarged blind spots

Testosterone therapy plays an essential role in FTM hormonal transitioning and may play a role in IIH.

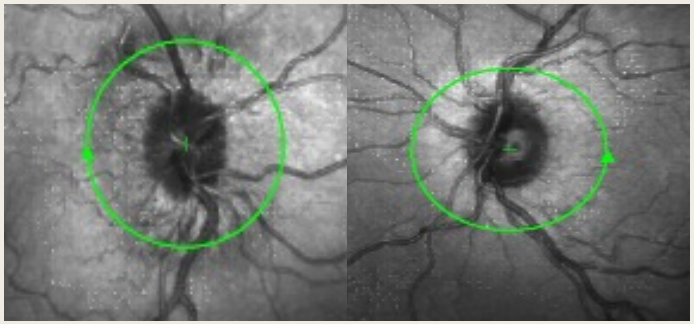
Patients undergoing testosterone therapy for gender transition should be informed of the possibility of developing IIH while on treatment, with obesity possibly increasing this risk.

Comprehensive eye examinations should be considered in these patients before initiating hormone therapy.

35 YO Hispanic F, with right sided visual loss

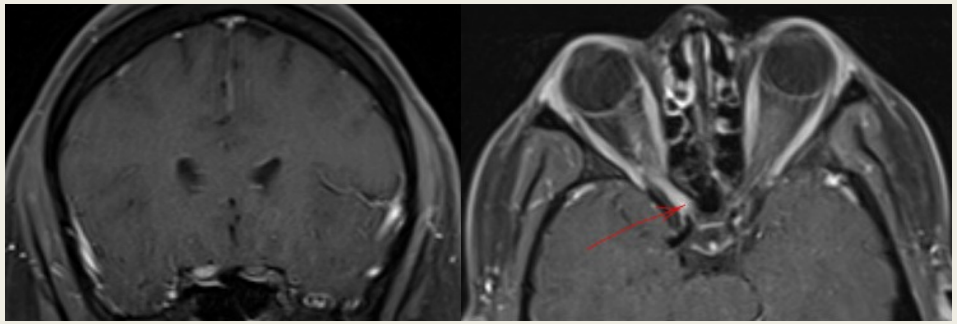
OD RNFL 218
HM, RAPD

OS RNFL 120
20/20 HVF Normal



T1 coronal w/ contrast 2019

T1 axial w/ contrast 2019

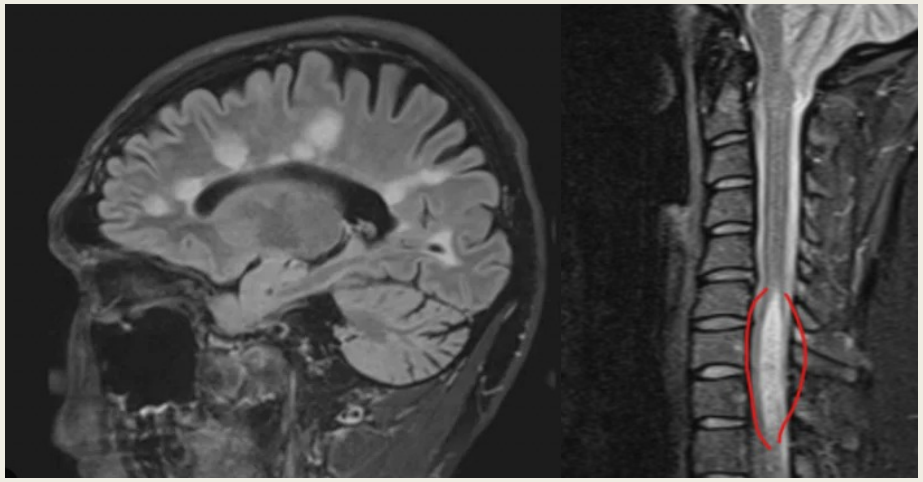


2019: MRI Brain w/c: Periventricular lesion
MRI spine: Positive for longitudinally extensive lesion >2 segments

LP OP – 17, CSF : Positive for 5 Oligoclonal bands

NMO and MOG antibodies normal

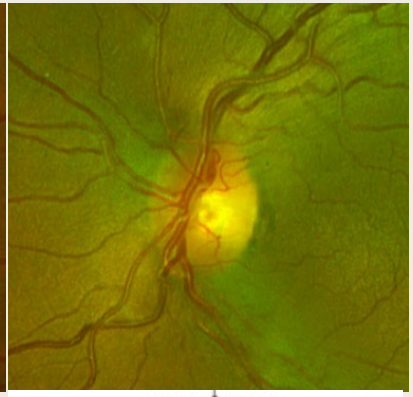
Diagnosis : Multiple sclerosis

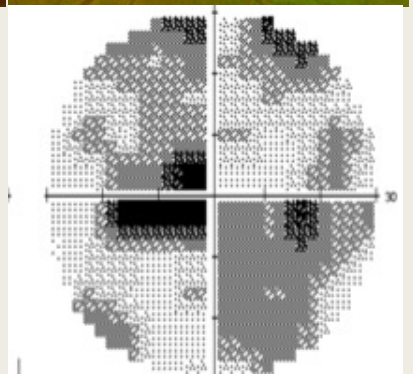


A year later .. New visual loss left eye

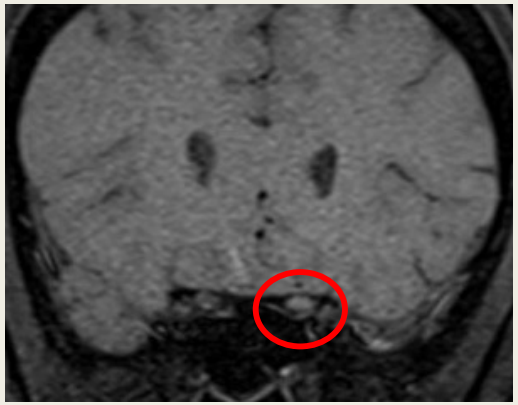
OD RNFL 52
HM, RAPD

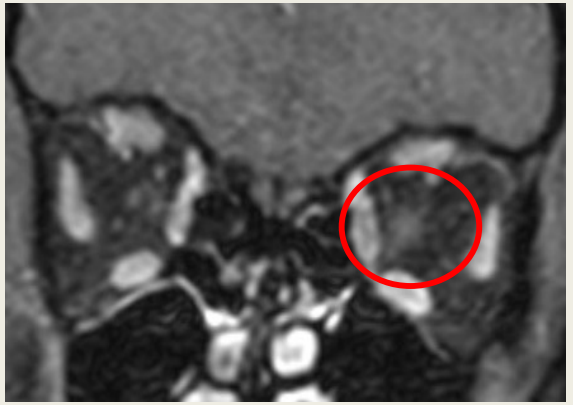
OS RNFL 140
20/50





Coronal T1 w/ Contrast FS 2020





Headaches: Multifactorial

IIH

Chiari Malformation

Migraine

Repeat LP – OP 35 cm of H2O

Repeat MOG and NMO Negative

Recent weight gain : 20 lbs in 1 yr

Additional Diagnosis : IIH

Steroid use is not benign
Can lead to weight gain and increased BP that adds to more headaches

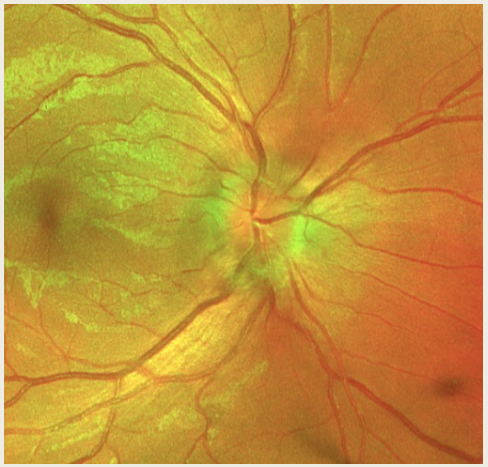
Differences between MOG/NMO and MS

Demographics and characteristics	MOG-IgG	AQP4-IgG	Multiple sclerosis
Median age	30's + children	40's	20's
Sex	Female~male	Female>>male	Female>male
Optic neuritis characteristics			
Bilateral ON	Frequent	Frequent	Infrequent
Severe vision loss at nadir	Very frequent	Very frequent	Frequent
Risk of recurrent ON	Very frequent	Very frequent	Frequent
Steroid dependent	Frequent	Rare	Rare
Risk of blindness (< 20/200)	Infrequent	Very frequent	Infrequent
MRI optic nerve enhancement			
Length and location	Long and anterior	Long and posterior	Short
Perineural enhancement	Frequent	Rare	Rare
Optic chiasm involvement	Infrequent	Frequent	Rare

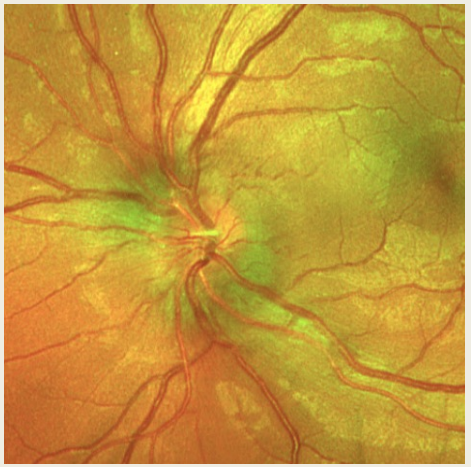
Ref: Deena et al. Clinical Characteristics and Treatment of MOG-IgG–Associated Optic Neuritis Current Neurology and Neuroscience Reports (2019) 19:100

25 YO , very thin built, Caucasian, man, with a new onset blurry vision bilaterally

OD RNFL 193
20/20 No APD

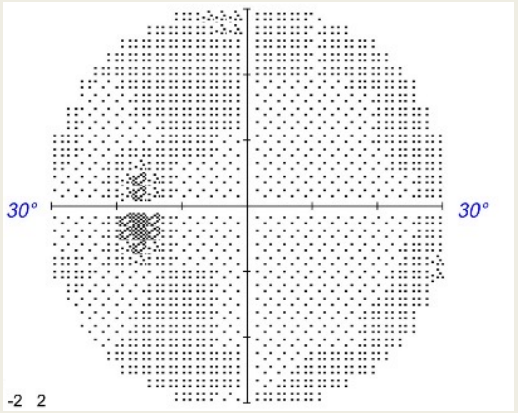


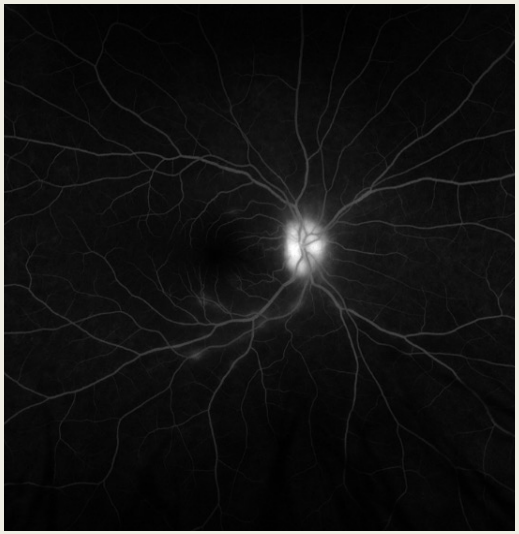
OS RNFL 160
20/80 No APD



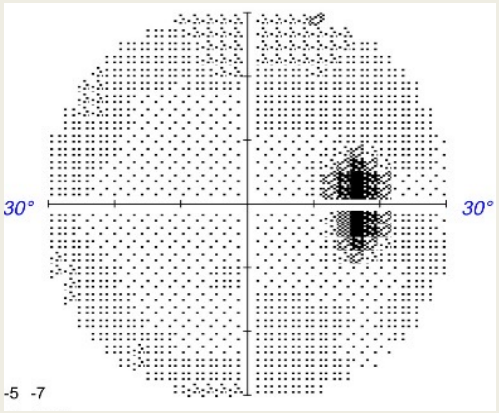
Hx of left ulnar neuropathy ,Generalized fatigue, Night sweats for almost a year
Significant hair loss since last 1 year, Intermittent arthralgias

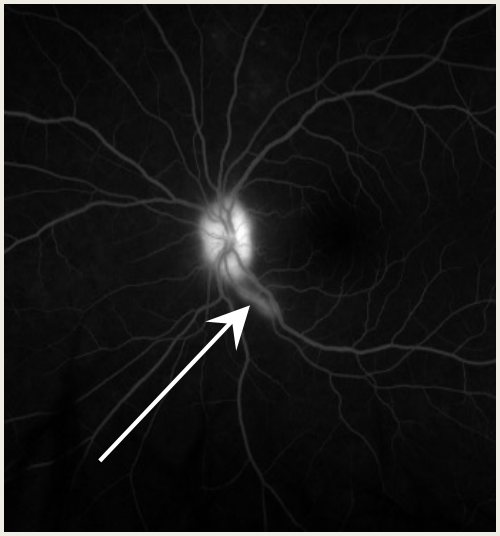
OD





OS





- *MRI Brian/MRV was normal*
- *VDRL Reactive (1:512)*
- *T. Pallidum (FTA-ABS), serum IgG Reactive*
- *LP opening pressure: 15 cm of water*
- *Social history !!!!*

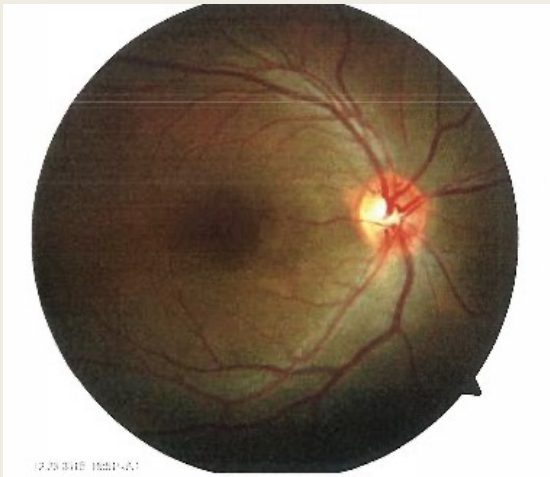
Neuro-Syphilis – The great imitator strikes back !

Pearls

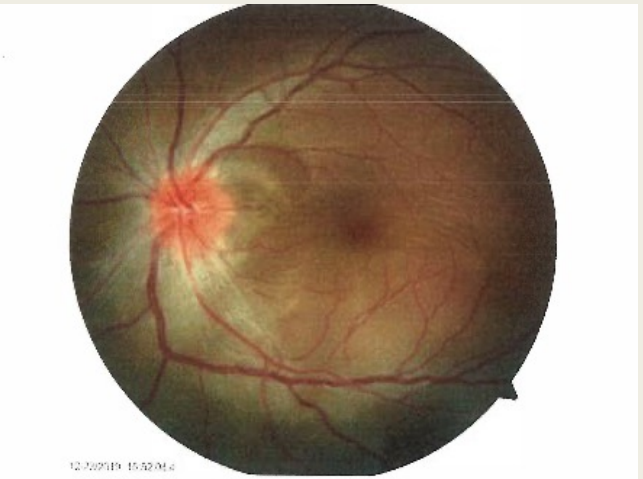
- *Symptomatic neurosyphilis can occur at any stage of infection*
- *High prevalence in MSM, Important to know sexual history well*
- *The diagnosis of neurosyphilis requires interpretation of testing from both serum and CSF in the appropriate clinical and epidemiologic context*
- *In a patient with a concerns for sexually transmitted disease , alopecia may be an important associated finding and can provide clues to diagnosis*

30 Y Indian female new Left sided visual loss

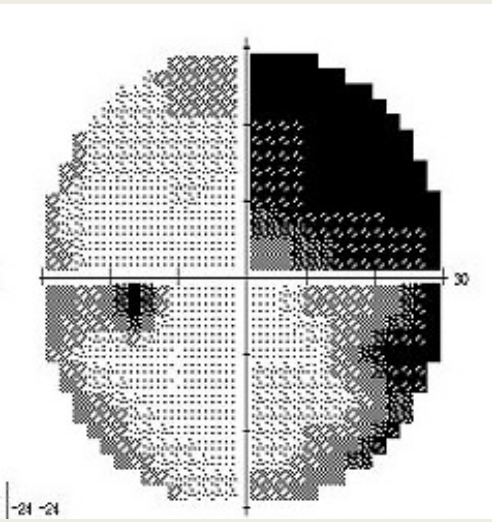
OD RNFL 82
20/20 HVF Normal



OS RNFL 326
20/80 Trace APD

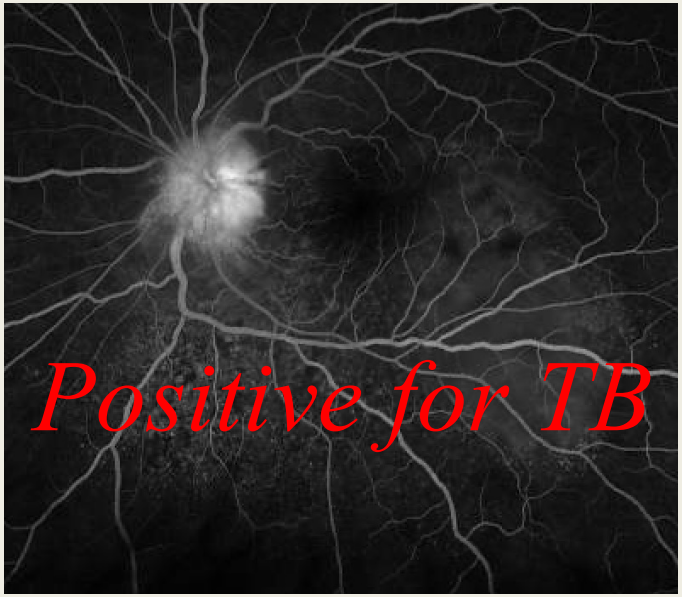


HVF
Supranasal deficits

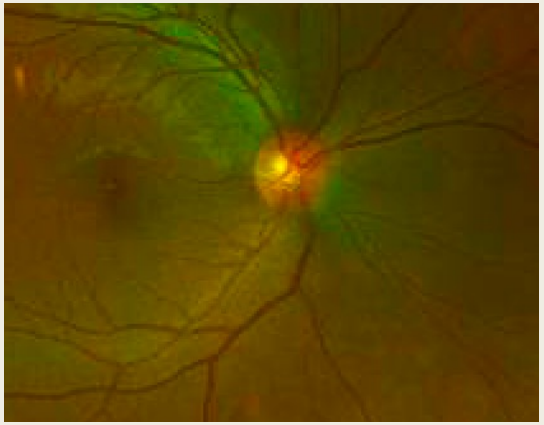


MRI brain, orbits was normal. Patient deferred an LP, and steroids.

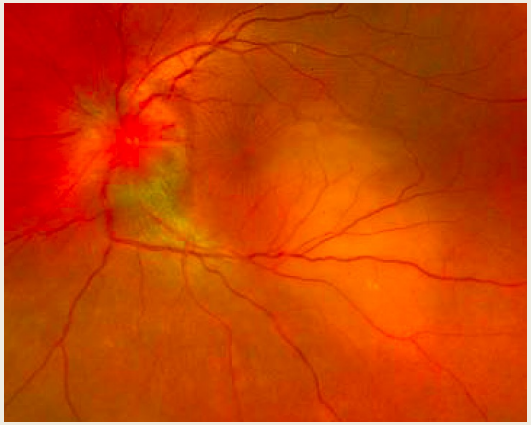
A week later , she presents for repeat HVF

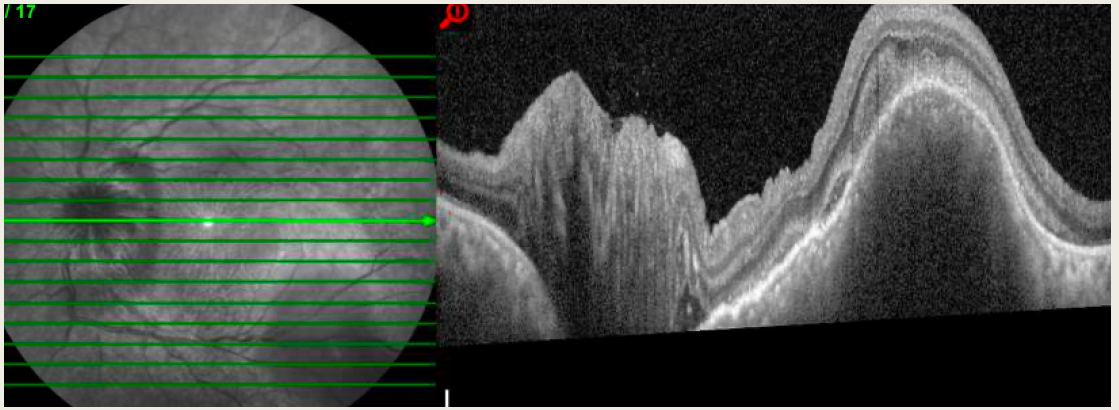


OD

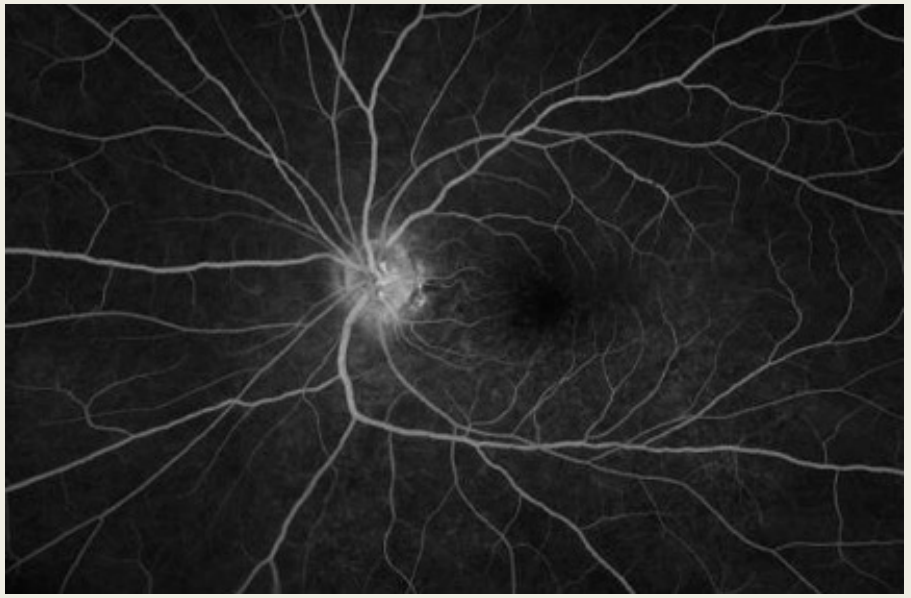


OS





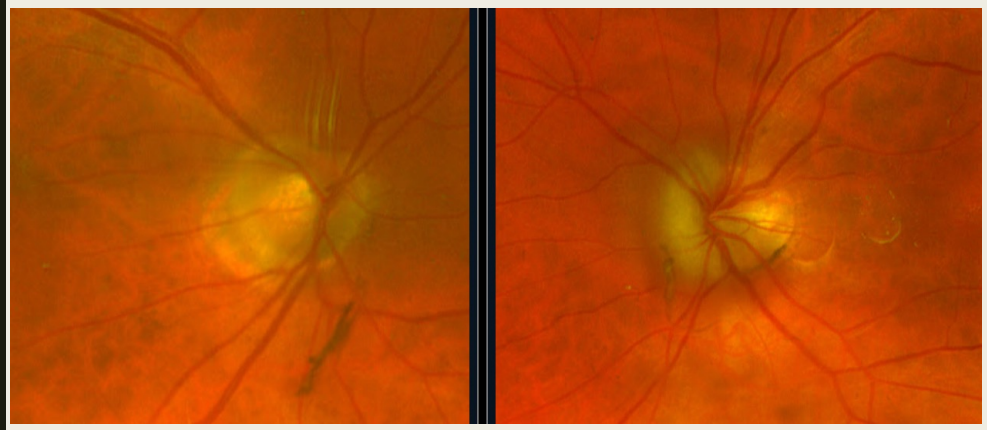
After Treatment



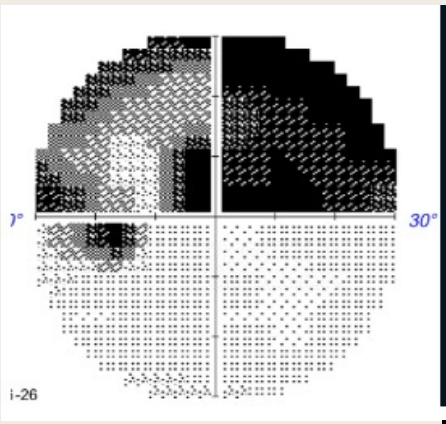
78 YOM with newly diagnosed DM , with diplopia and headaches

OD RNFL 82
20/20

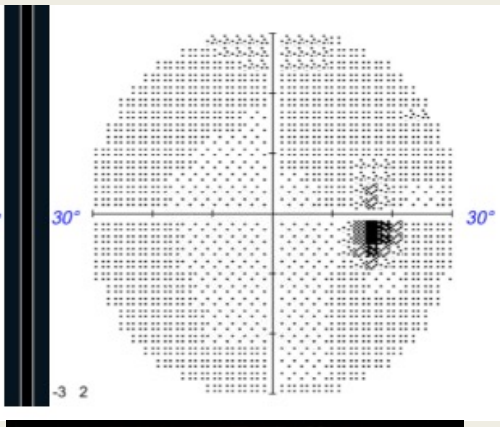
OS RNFL
20/40 2 + APD



OS

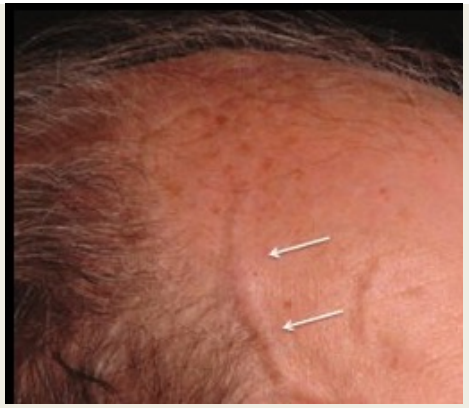


OD



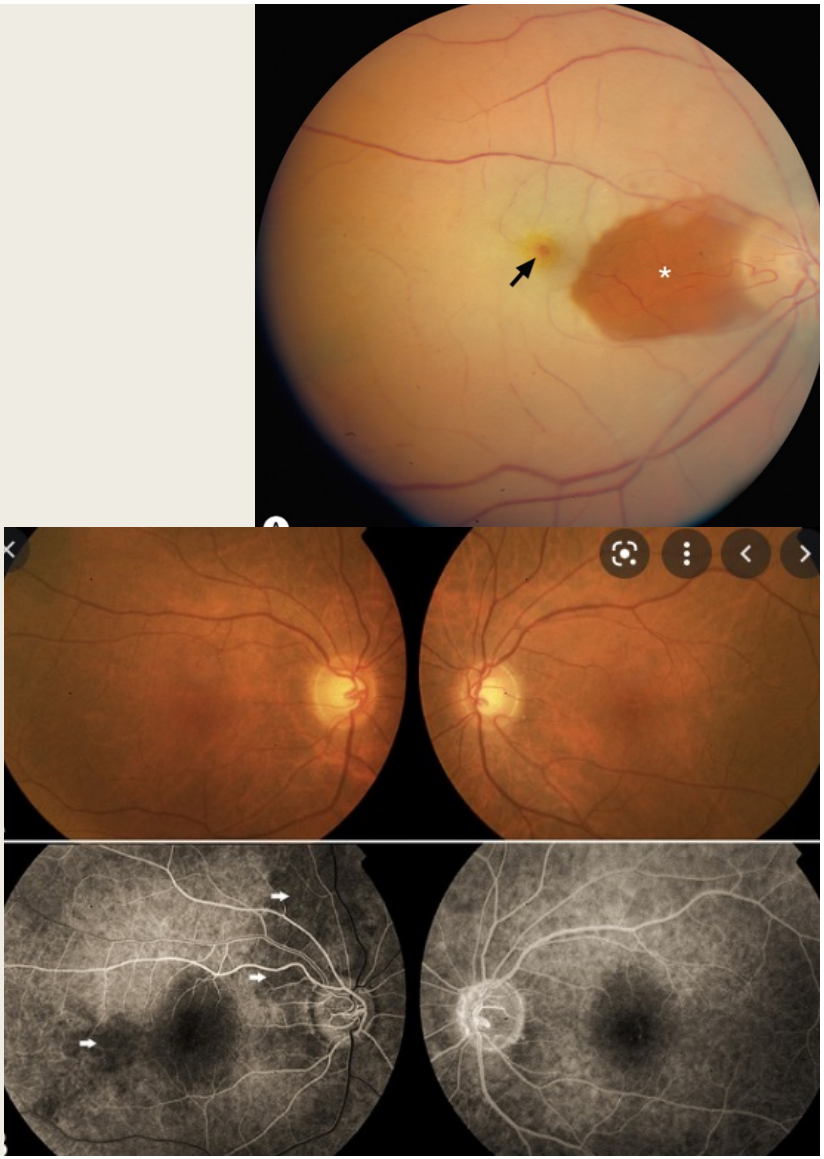
- Temporal tenderness
- Jaw pain
- General fatigue
- Weight loss and fever

Temporal artery biopsy Positive



Other ophthalmic features

- *Retinal artery occlusion, cotton wool spots*
- *Choroidal ischemia*
- *Anterior chamber flare, lens opacity, ocular hypotony ("ocular ischemic syndrome")*
- *Diplopia from extraocular muscle ischemia or ocular motor palsy*



GCA

Diagnosis

- *Prodrome, GCA symptoms*
- *Elevated ESR/CRP (combination of the two gives high specificity [97%])*
- *Elevated platelet count (acute phase reactant)*
- *Ophthalmoscopy*
- *Fluorescein angiography*
- *Temporal artery biopsy*

Treatment

- *Prompt steroids and hydration*
- *Consider IV when vision loss present*
- *Effective in prevention of second eye*
- *Occasionally restores vision*

Beware of starting steroids on the patient without a biopsy OR follow up .
Hypertensive side effects can be dangerous
Hypertension from steroids can cause more headaches and risk of stroke itself

ORIGINAL ARTICLE

Trial of Tocilizumab in Giant-Cell Arteritis

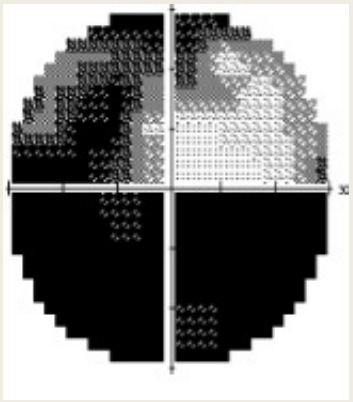
Authors: John H. Stone, M.D., M.P.H., Katie Tuckwell, Ph.D., Sophie Dimonaco, M.Sc., Micki Klearman, M.D., Martin Aringer, M.D., Daniel Blockmans, M.D., Ph.D., Elisabeth Brouwer, M.D., Ph.D., +8, and Neil Collinson, Ph.D. Author Info & Affiliations

Published July 27, 2017 | N Engl J Med 2017;377:317-328 | DOI: 10.1056/NEJMoa1613849 | VOL. 377 NO. 4

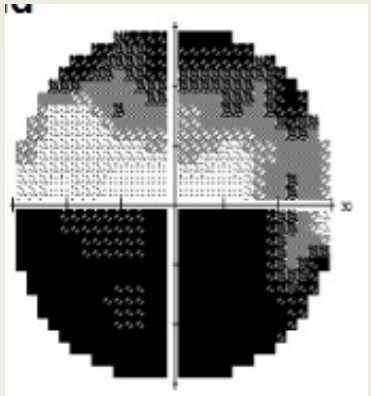
Tocilizumab, received weekly or every other week, combined with a 26-week prednisone taper was superior to either 26-week or 52-week prednisone tapering plus placebo with regard to sustained glucocorticoid-free remission in patients with giant-cell arteritis.

62-YOM, w/ PMH of HLD, HTN , DM with right eye visual loss.

OS
VA: 20/80

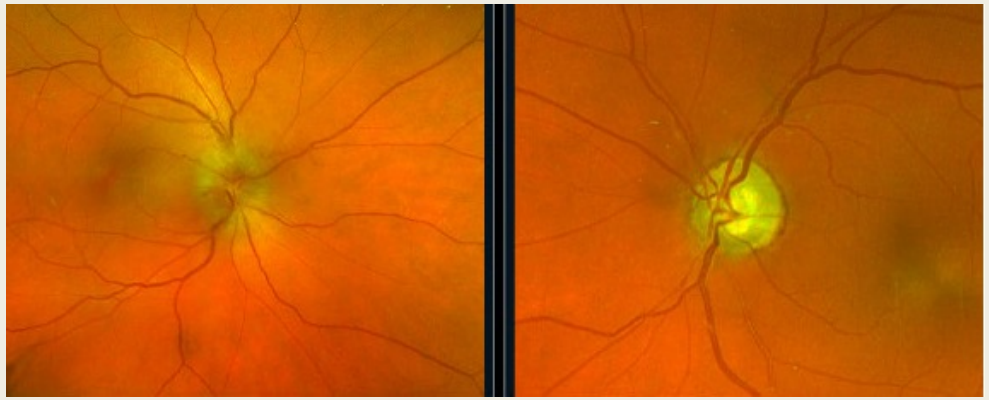


OD
VA: 20/60



OD RNFL 190

OS RNFL 52



Ischemic Optic Neuropathy – Non Arteritic

Consecutive NAION

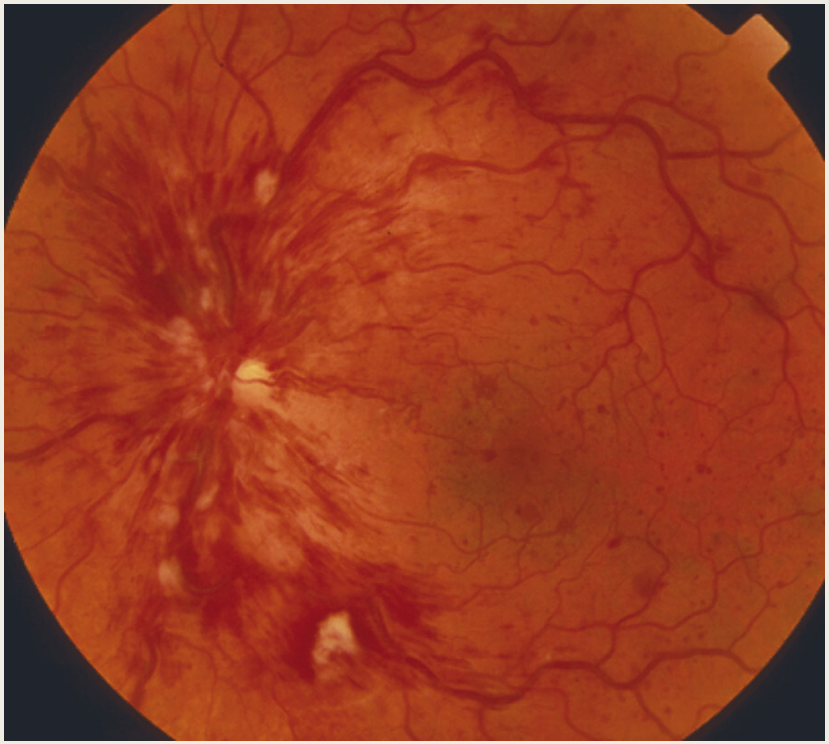
Clinical distinction between optic neuritis and ischemic optic neuropathy

- *Age group : Can overlap significantly- ages of 30 and 50 years*
- *Both can have disc swelling – although rare with optic neuritis*
- *Important to consider patient's age, associated symptoms, and examination findings.*
- *Presence of pain with eye movement is common in optic neuritis*
- *But Can be present in less than 10% of ION patients, and generally is not exacerbated by eye movements.*
- *Normal-appearing nerve is common in ON and by definition not seen in AION.*
- *APD will be present in both but not in Acute papilledema*

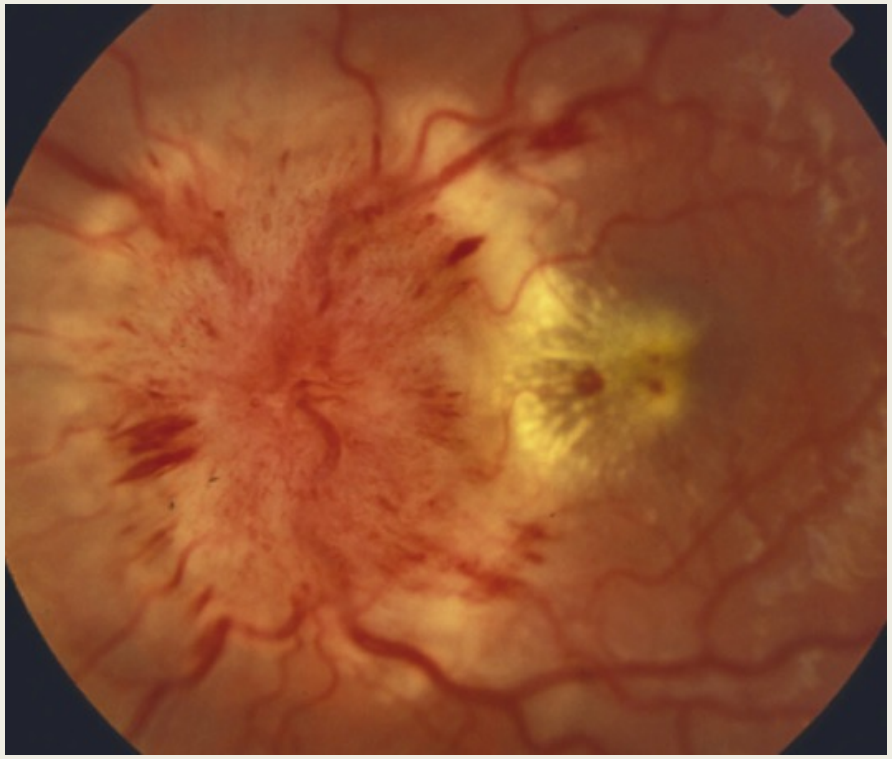
Clinical Distinction Between and Non-Arteritic and Arteritic ION

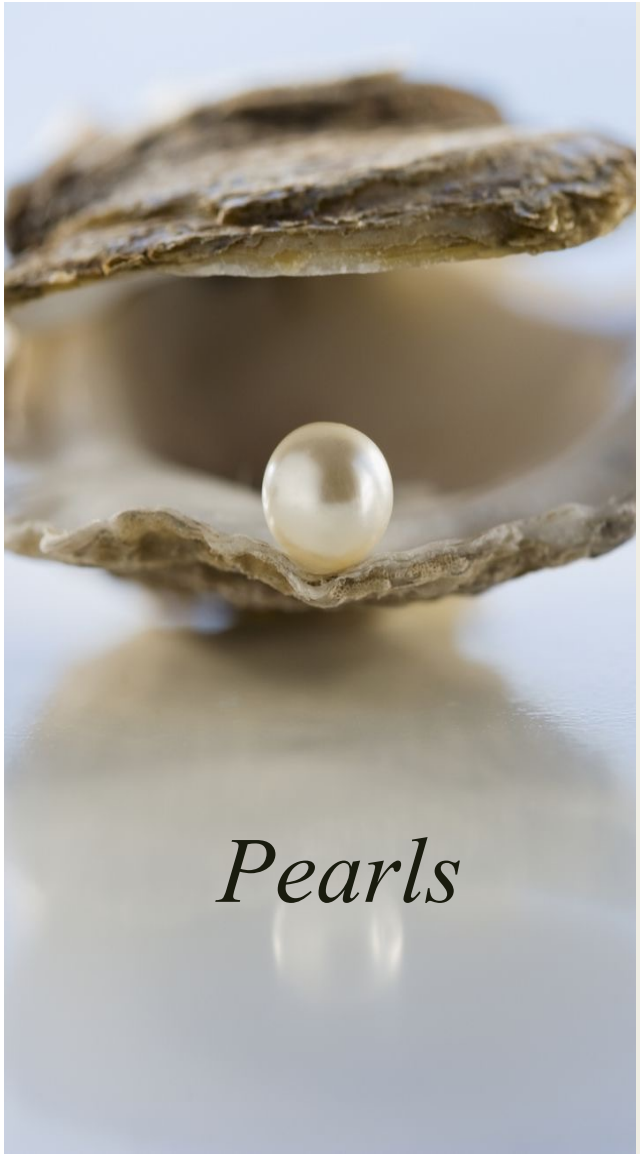
Criteria	*Non Arteritic*	*Arteriitic*
Age	*60-70 yrs*	*Most > 70 , Rare <50*
Race	*No difference*	*MC in Caucasian*
Sex	*No difference*	*MS in Females*
Pain	*Rare*	*Common*
Preceding symptoms		*HA, TLOV, Diplopia*
Second eye Involvement	*15–30% in mths or yrs*	*75% within days or weeks*
Disc appearance	*Sectoral edema*	*Normal or chalky white swelling*
ESR	*<40*	*Any: usually > 90*
FA	*Normal; can have delayed optic nerve head filling*	*Choroidal filling defects*
Response to steroids	*None*	*Systemically: definite* *Vision: sometimes*

Central retinal vein occlusion



Malignant systemic hypertension





Pearls

- *New onset headaches + swollen optic nerve - get neuro imaging!*
- *Painful visual loss in a young woman with an afferent pupillary defect - think about optic neuritis!*
- *Painless visual loss +altitudinal field loss + swollen optic nerve and headaches - get ESR , CRP!*
- *IIH has a spectrum - from mild to severe swelling - Triage them well !*
- *Dont forget to check blood pressure.*
- *History taking if of utmost importance. Dont forget to ask the medications they are taking.*
- *Make sure you check for an afferent pupillary defect before dilating them if patient is coming of severe visual loss.*
- *Ischemia and demyelination causes an APD, not early papilledema from increased ICP.*
- *A visual field and OCT is must !! You can't get it that day , bring the patient in next day or within a week.*

THANK YOU

QUESTIONS?