

POSTERIOR SEGMENT GRAND ROUNDS

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- No Financial Disclosures

Case: PT LM

- 72 yo hispanic female
- CC: burning sensation with with pressure in both eyes for 4 days
- Started with mild irritation and has progressively worsened
- Patient had no personal medical or ocular health history
- Hasn't been to PCP for 5+ years

Case: LM

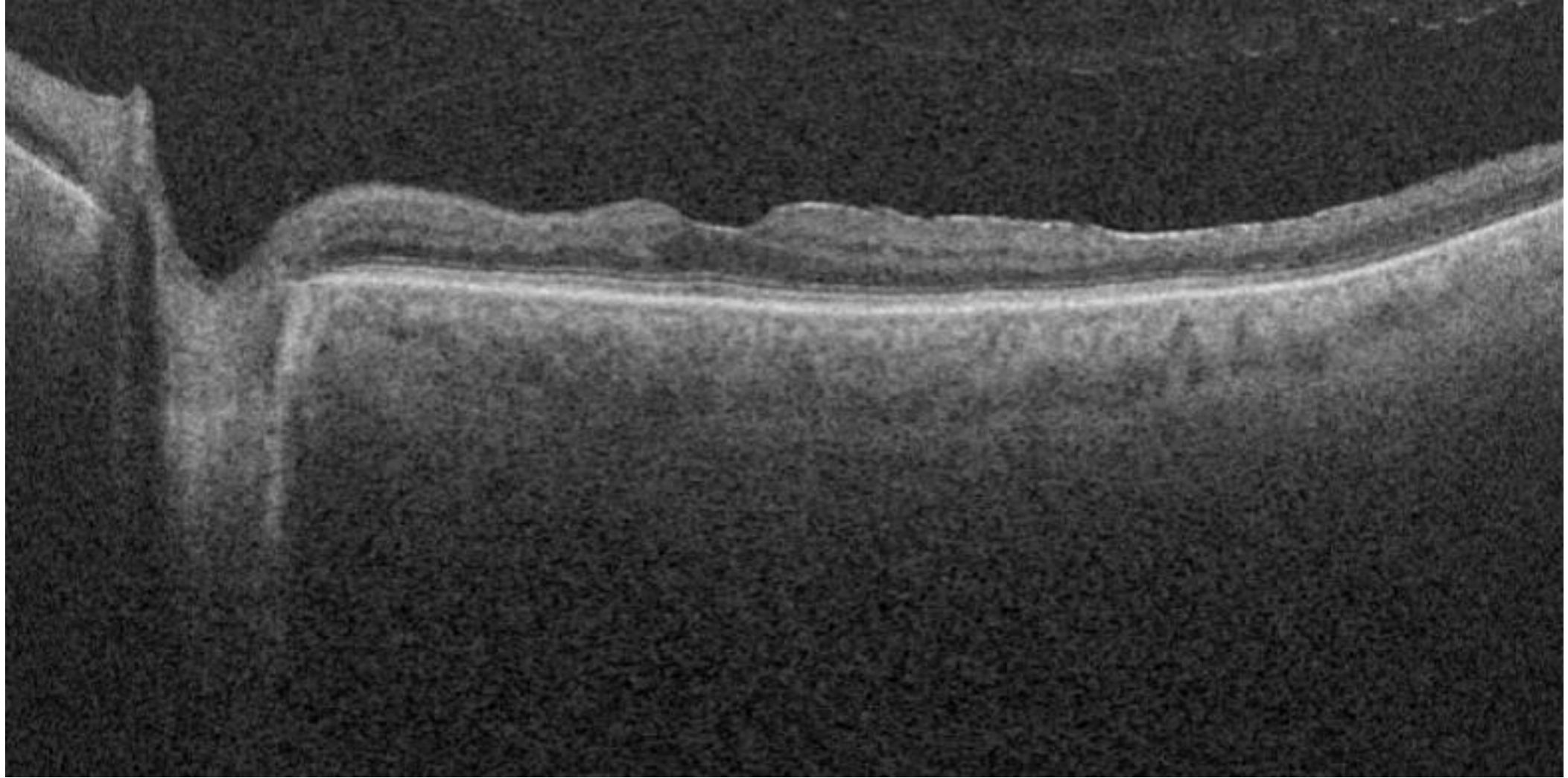
- Va with current RX: OD 20/50, OS 20/50, PH 20/40
- Pupils, EOMS, Matrix VF: normal
- IOP: OD 19, OS 20
- Anterior segment
 - 1+ bulbar injection OU
 - Trace to 1+ SPK
 - Reduced TBUT
 - 1+ NS
 - All other findings normal

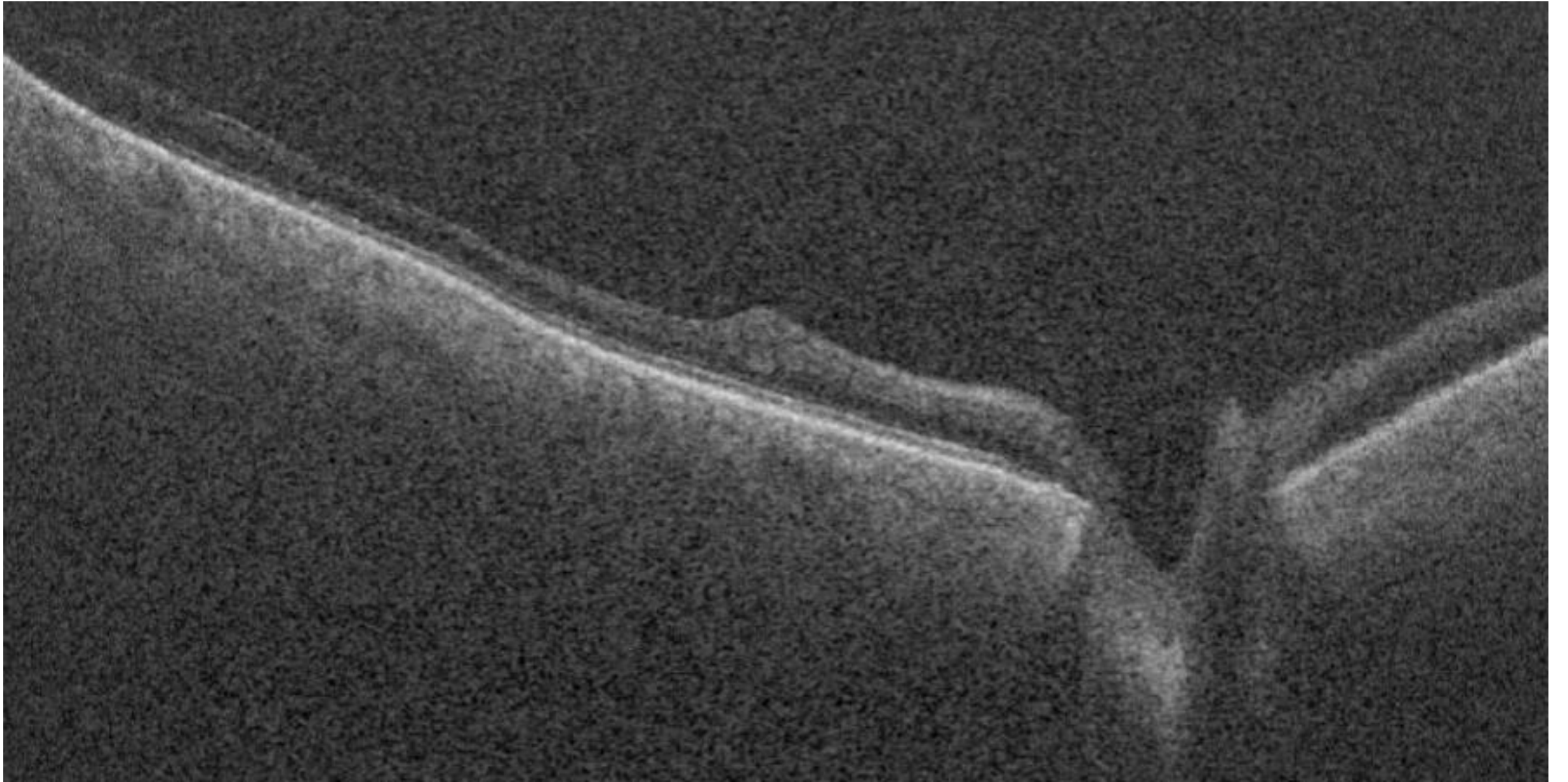
Poll: What is your next step?

- a) Preservative free artificial tears, warm compresses, omega 3's?
- b) Steroid and start cyclosporine treatment
- c) Tobradex
- d) View posterior segment?









Case: PT LM

- DDx of retinal vasculitis w multiple BRAO's
- Labs including:
 - CBC, ERS, CRP, ANA, RF, PRP, Quantiferon, CMP, Bartonella testing, VLDR, Lymes serology, Phospholipid AB
- Referral to retina ASAP for FA

Retinal Vasculitis

- Sight threatening inflammatory disease involving retinal vasculature
- May be idiopathic, infectious, neoplastic, or associated with systemic inflammatory disease.
- Incidence of retinal vasculitis is 1-2 per 10,000
- 55% of patients with RV have associated systemic inflammatory disease
- Large study shows approximately 15% patients will have a uveitis.
- More common <40, women

Retinal Vasculitis

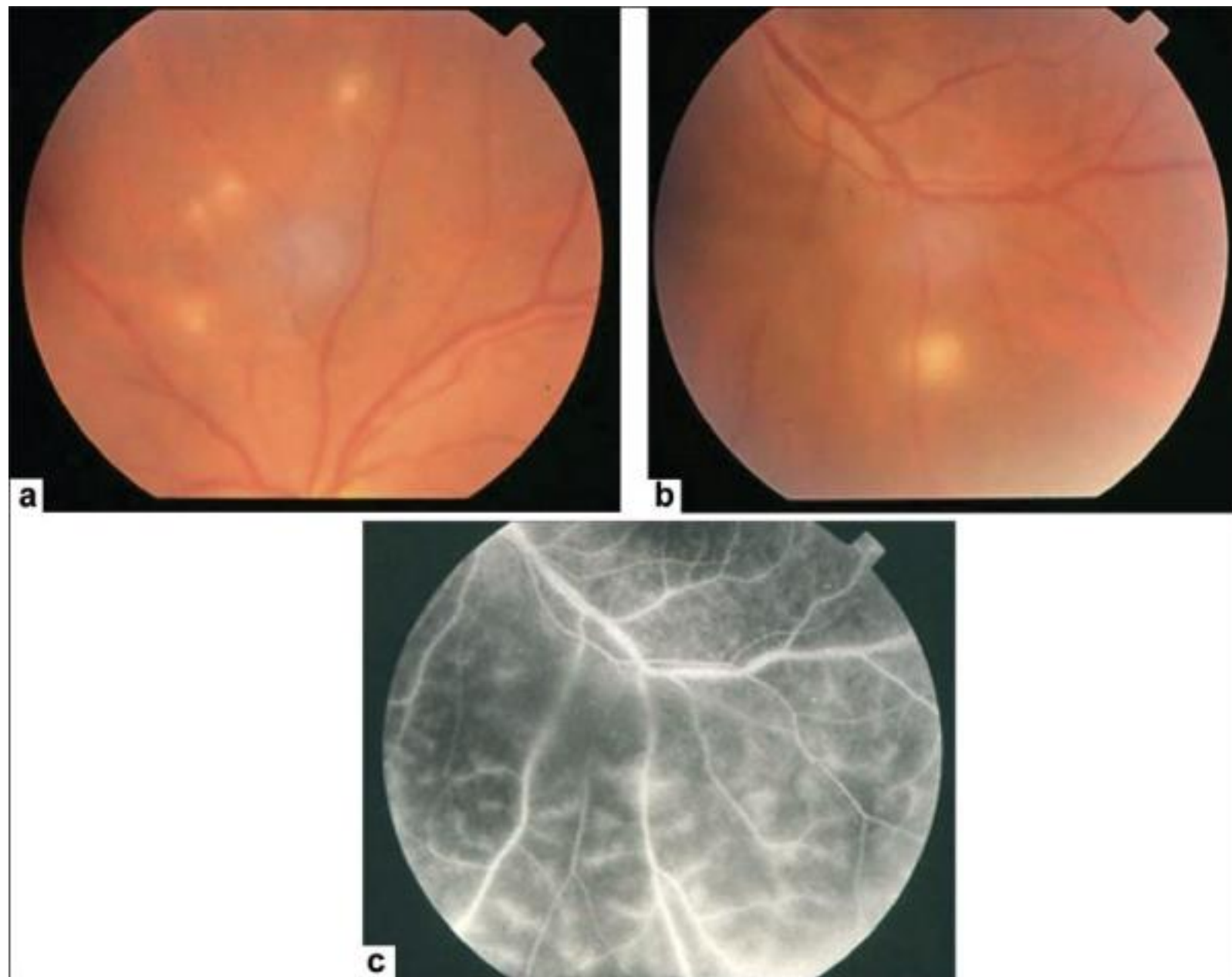
- Active disease characterized by:
 - Exudates surrounding vessels
 - **Sheathing or cuffing**
 - Leakage causing retinal swelling, macular edema
 - Occlusive disease leading to CWS and microinfarcts, CRAO. BRVO, periphlebitis
 - Poor visual outcomes occur with macular ischemia
- Late stage changes include:
 - Telangiectasias, microaneurysms, neovascularization, tractional detachment, recurrent vitreous hemorrhaging, glaucoma

Retinal Vasculitis

- Causes:
 - Bacterial: Tuberculosis, syphilis, lymes disease, whipple disease, cat scratch disease, endophthalmitis, post streptococcal syndrome,
 - Viral disorders: HIV, CMV, HSV, VZV, epstein-barr, west nile, dengue
 - Toxoplasmosis, rocky mountain spotted fever,
 - MS, Susac syndrome
 - Neoplasias
 - Behcet's, sarcoidosis, wergeners, RA, etc
 - Frosted branch angiitis, pars planitis, etc
 - Rarely isolated
- Testing
 - FA!!, CBC, ESR, CRP, serology tests for viral/parasitic, TB testing, VLDR, RA, ANA, ANCA

FA demonstrating focal leaking in FA in patient with vasculitis from Behcet's disease.

Courtesy NIH



A



B



C



D



AAO

Case: Pt LM

- 3 months later
- Confirmed retinal vasculitis with multiple BRAO's
- All imaging and blood labs negative
- Was initially started on oral Prednisone 60 mg then tapered to 10 mg x 2 weeks then 5 mg for 2 weeks
- Returned two weeks later and was treated with Ozurdex pellets OU 1 week apart
- Delayed long term systemic immunosuppression at this time due to risk of COVID

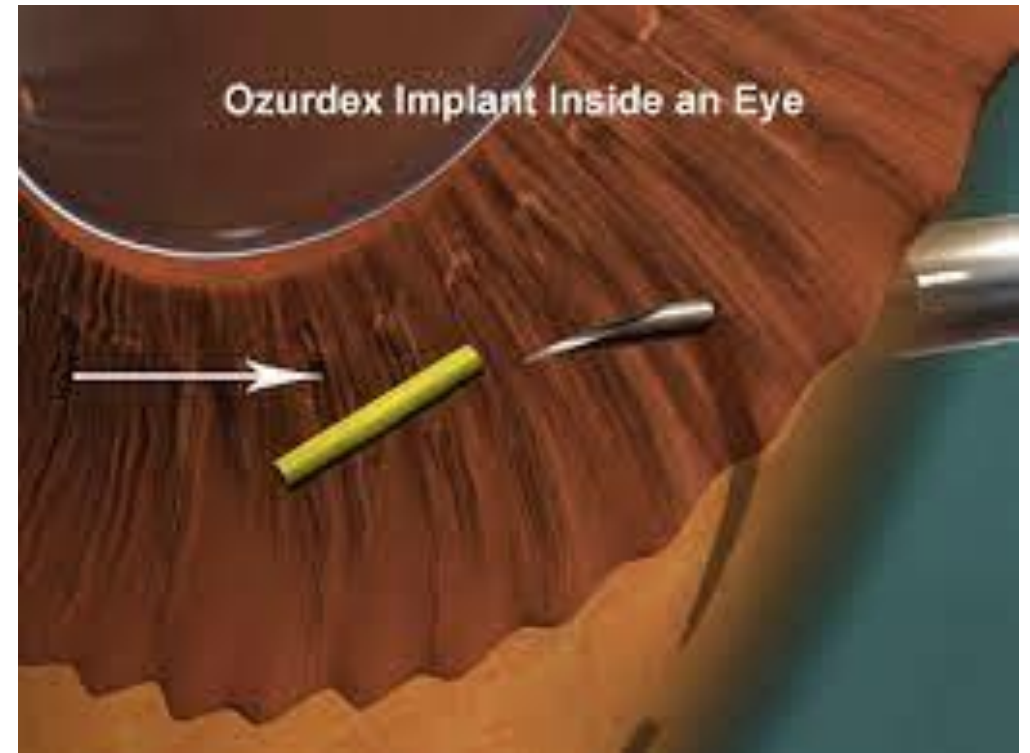


Retinal Vasculitis

- Treatment
 - Dependent upon etiology
 - Initial therapy consists of systemic corticosteroids
 - Local steroids
 - Secondary ischemic and neovascularization may be treated with bevacizumab or laser photocoagulation
 - Ladder approach to treatment necessary for long term steroid free remission
 - Anti metabolites (azathioprine, methotrexate) and tacrolimus can be utilized
 - Biologics (adalimumab, infliximab and rituximab) are effective treatments

Ozurdex

- Dexamethasone 0.7mg intravitreal implant
- Approved for 1) DME, 2) ME following BRVO/ CRVO, 3) noninfectious inflammation in choroid/ retina
- Contraindicated in cases of active infections, glaucoma that has progressed to a c/d ratio of >0.8 , or with a torn or ruptured posterior lens capsule
- Dissolves within 2-3 months

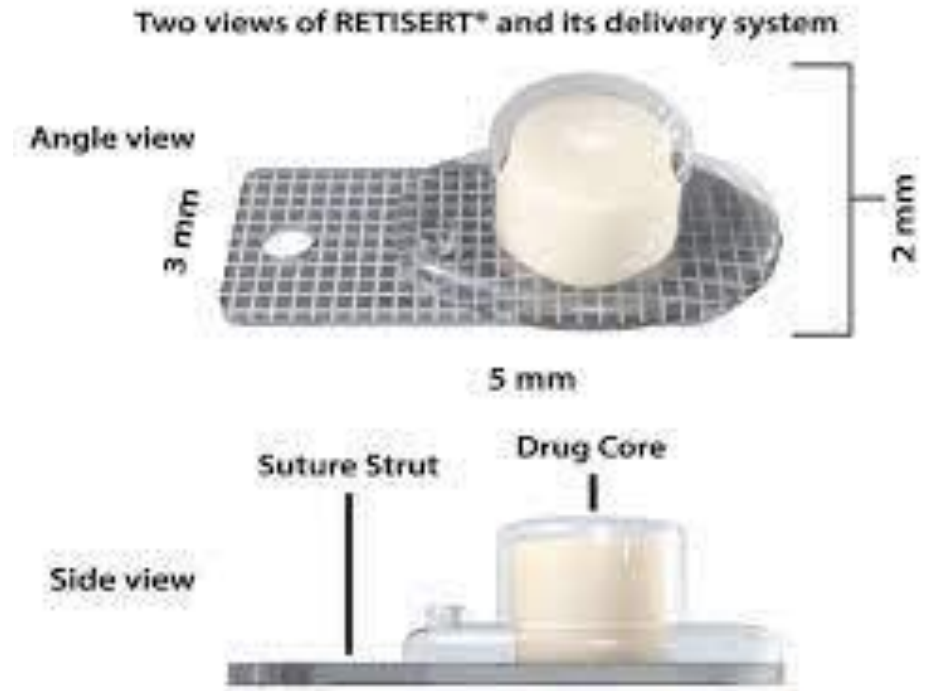


Case: PT LM

- Our patient underwent 2 more pellet injections OS and 1 OD over the next year.
- Patient developed cataracts and IOP in the mid 20's
- 13 months following initial visit patient had cataract surgery OS, with a concurrent PPV, Retisert implant and Ahmed valve implant
- 14 months following initial visit patient had cataract surgery OD with an Ahmed valve implant

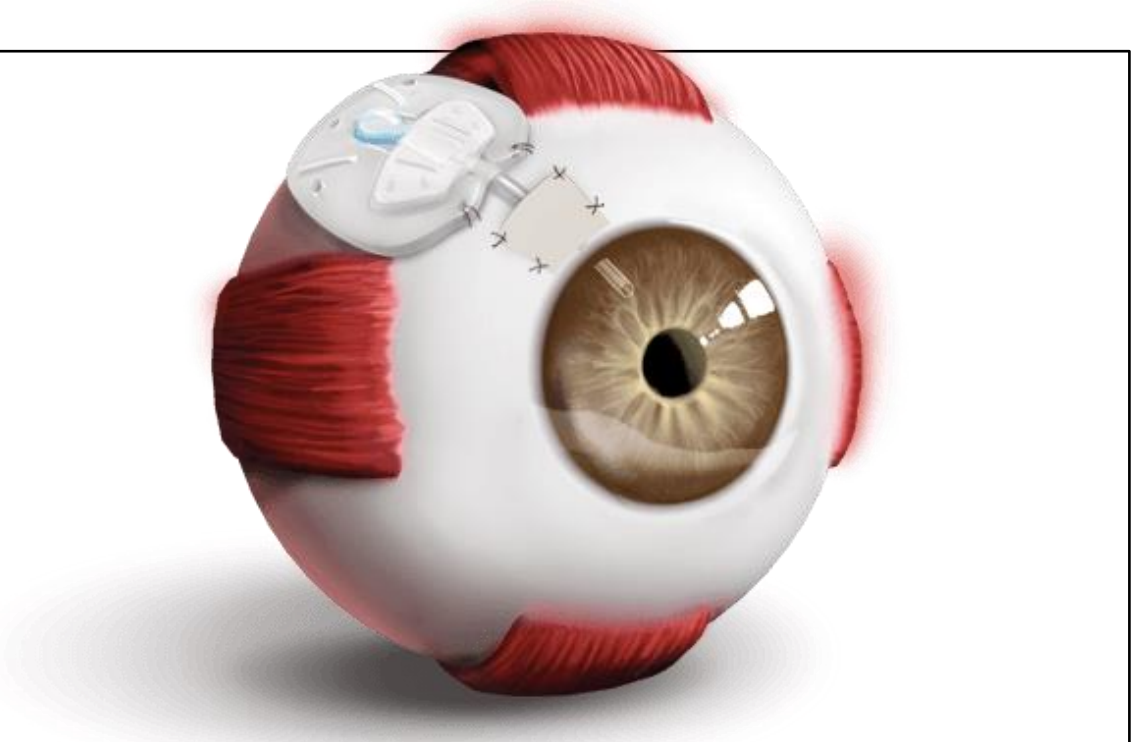
Retisert

- 0.59 mg fluocinolone acetonide intravitreal implant
- Contraindicated in active infectious processes
- Delivers corticosteroid therapy for ~2.5 years
- Nearly 100% phakic eyes at 3 years post implant develop visually significant cataracts
- Following implantation all patients experience immediate decrease in VA for up to 4 weeks
- Within 3 years post implantation 77% require IOP medications and 37% require filtering procedures



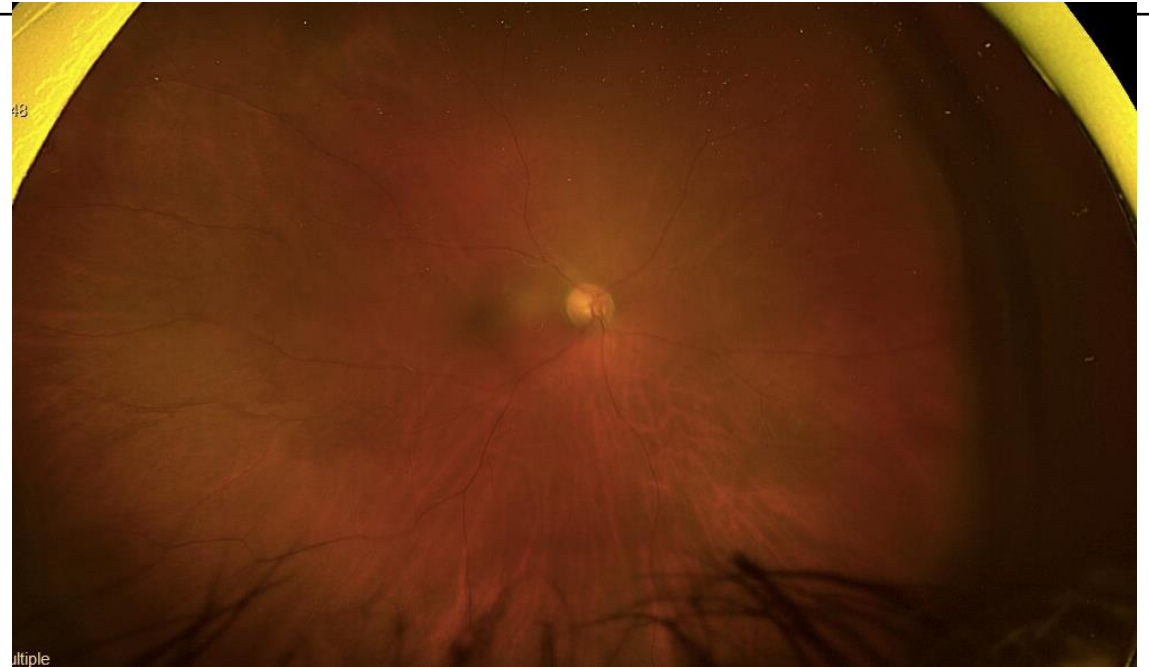
Ahmed Glaucoma Valve

- An implanted restrictive drainage valve consisting of thin silicone elastomer membranes
- Pressure valve that opens and closes according to IOP by which postoperative hypotony is reduced
- Inserted under the conjunctiva and joined to sclera typically between the superior and lateral rectus muscles
- Excellent in cases of secondary refractory glaucoma



Case: Pt LM

- Most recent visit 1 month ago
- BCVA 20/25 OD, 20/30 OS
- IOP 8 OD, 7 OS
- Currently going over options for biological immunosuppressives



Cases: Pt R

- 76 yo female
- Comes in to have a Yag Cap OS in September- notices vision has gotten significantly worse in last 2 weeks
- Had cataract surgery in march
- Was seen for consultation visit for Yag Cap in July
 - 20/30 OD, 20/30
 - 1+ PCO

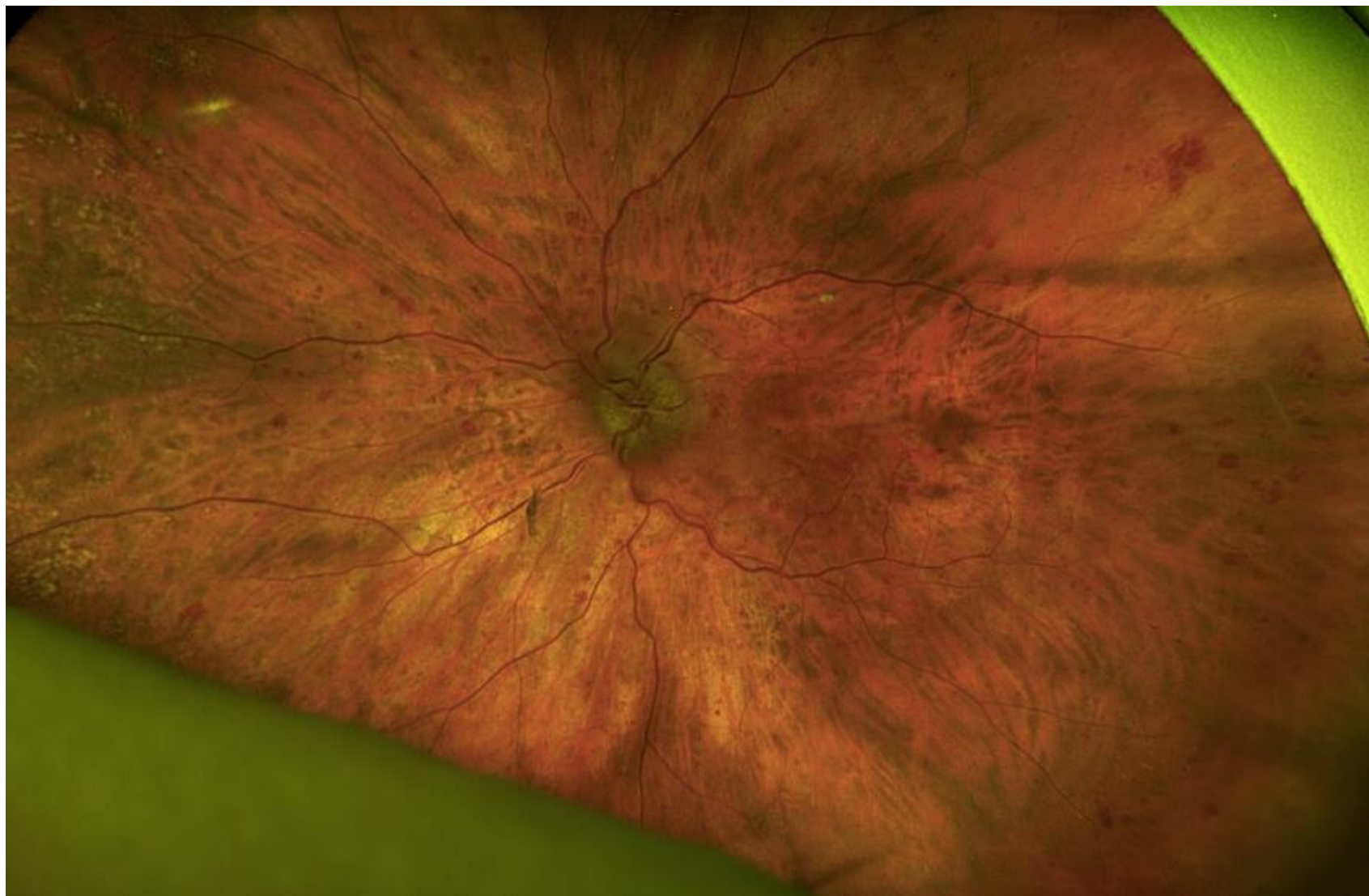
- On Pre op work up
 - 20/30 OD, 20/800 OS

Poll Question

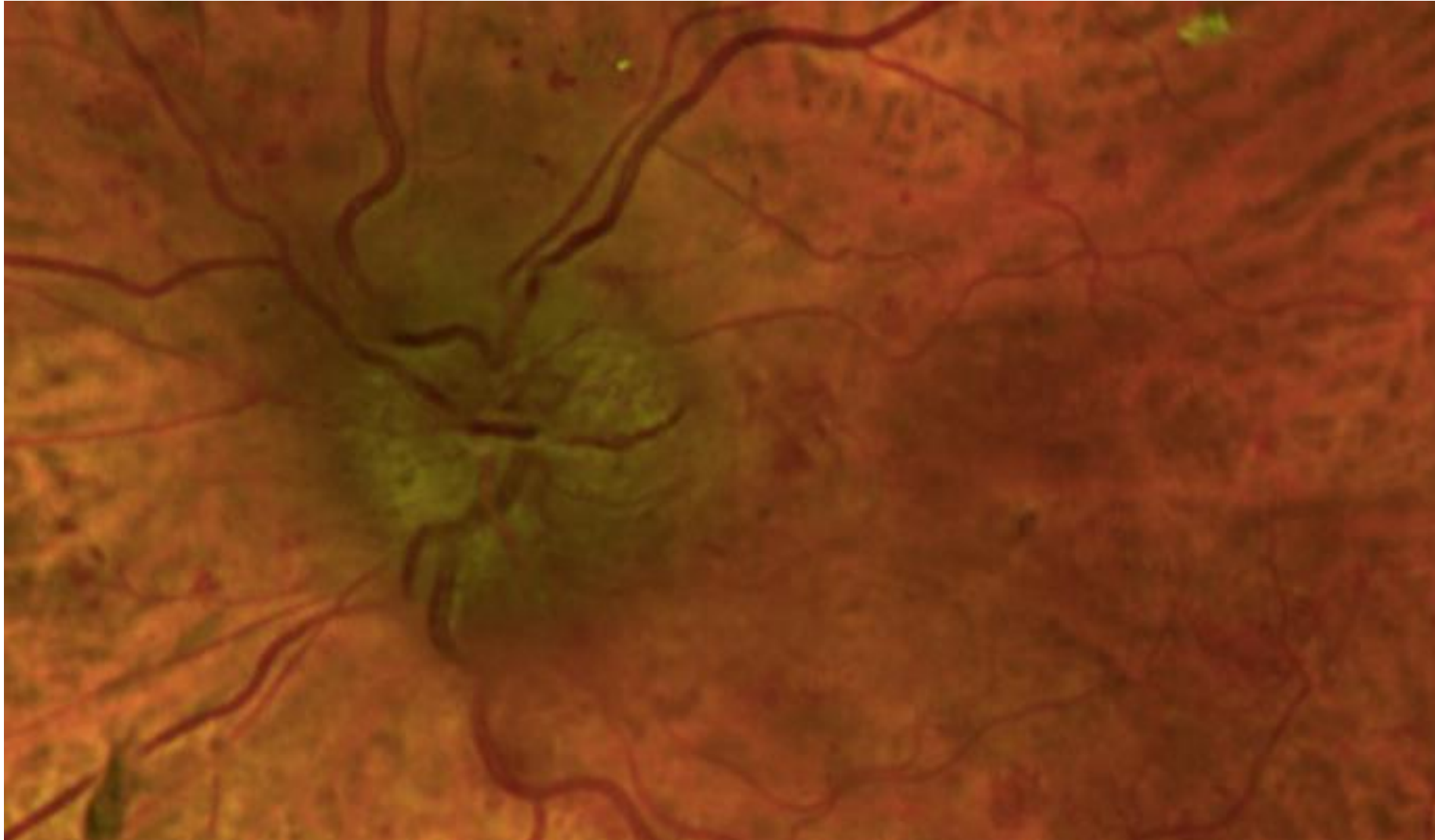
Is that reduction in vision expected from a PCO?

- a) Yes
- b) No
- c) Maybe

YAG Cap Cases: PT R

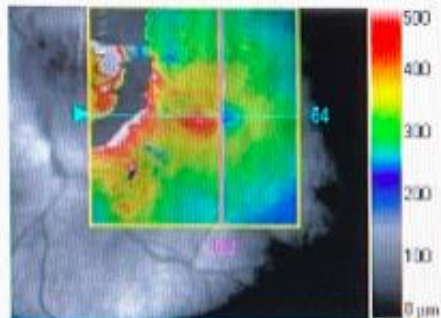


YAG Cap PT: RK

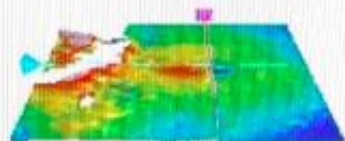
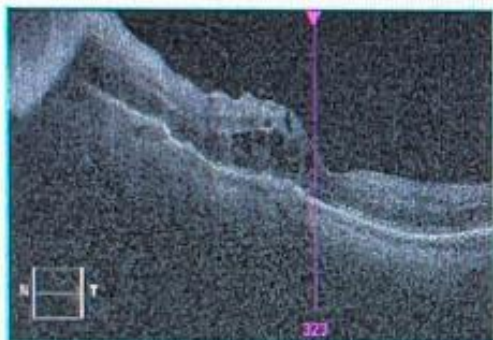
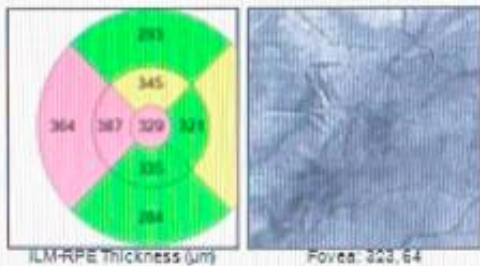


Macula Thickness : Macular Cube 512x128

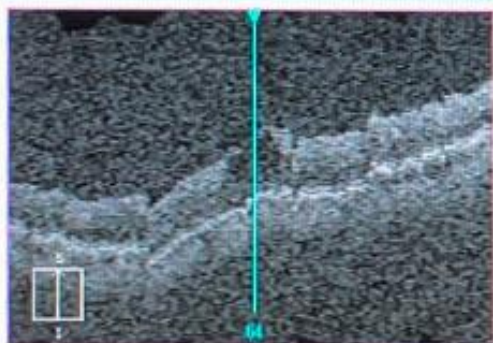
OD OS



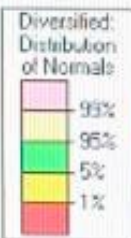
Overlay: ILM - RPE Transparency 32%



ILM - RPE



ILM

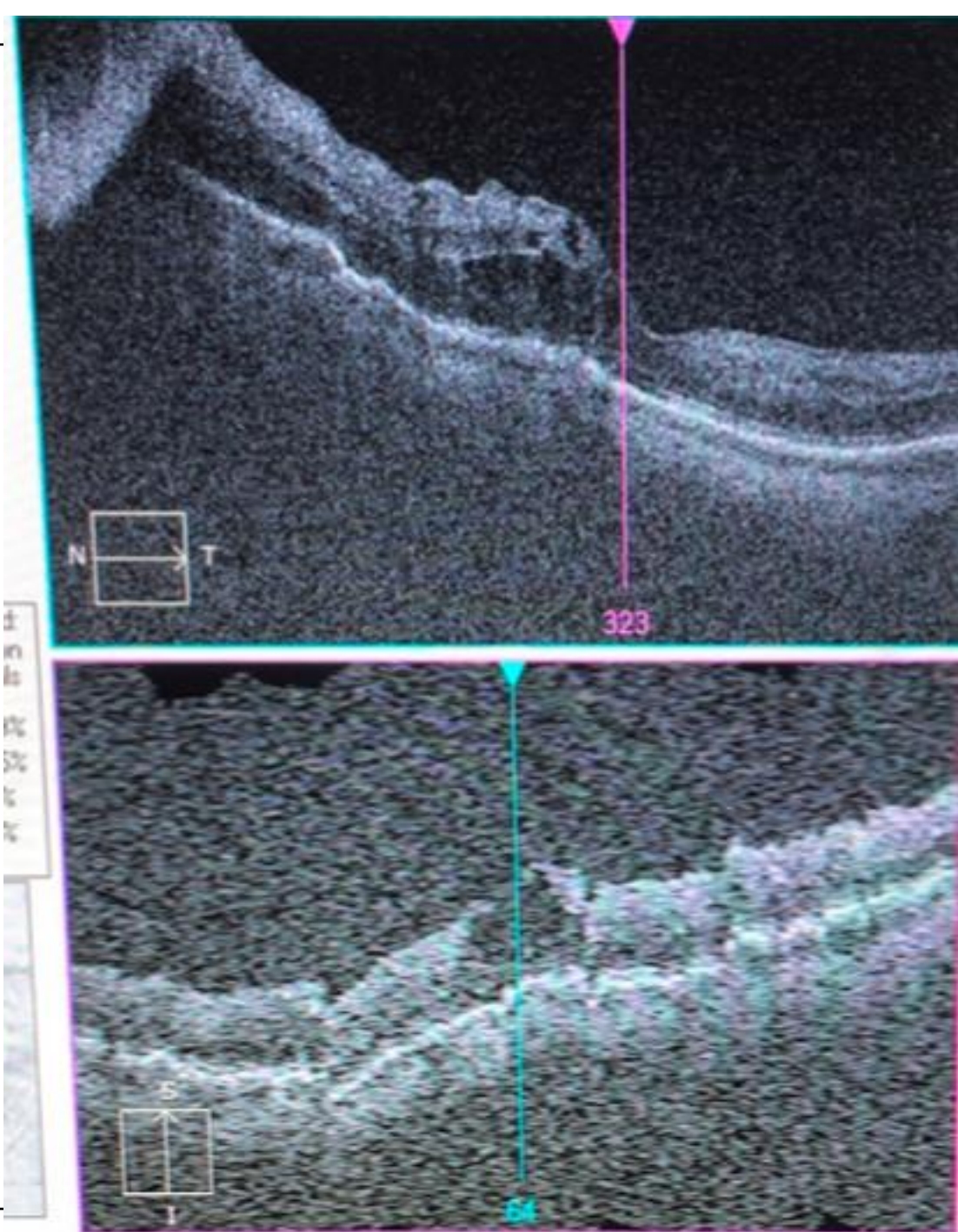


RPE

	Central Subfield Thickness (µm)	Cube Volume (mm³)	Cube Average Thickness (µm)
ILM - RPE	329	10.4	290

Comments

Doctor's Signature

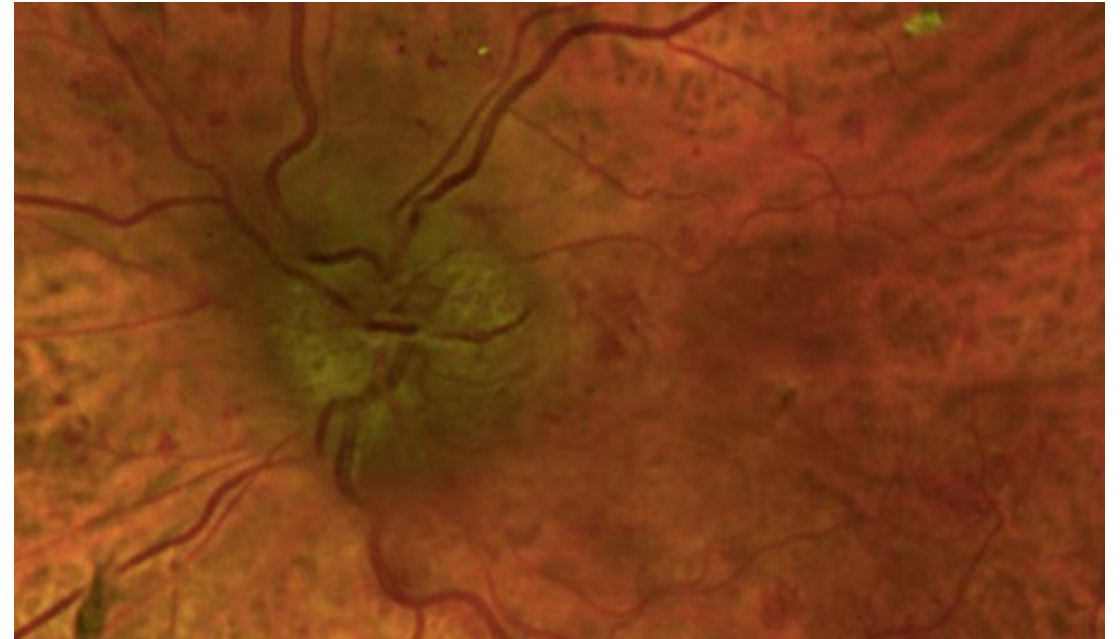


Pt R

- Ischemic central retinal vein occlusion with macular edema, Disc edema
- Referred to retina specialist for treatment
- Referred for GCA work up, cbc, and MRI
 - Normal levels, MRI showed mild enhancement of left optic nerve
 - CBC results not available
- Working Diagnosis

Central Retinal Vein Occlusion

- New evidence indicates thrombus is typically located in ON posterior to lamina Cribrosa
- >50
- Painless vision loss ranging from near normal to NLP
- Signs: dilated and tortuous vessel, hemorrhages, CWS, disc edema in severe cases



Central Retinal Vein Occlusion

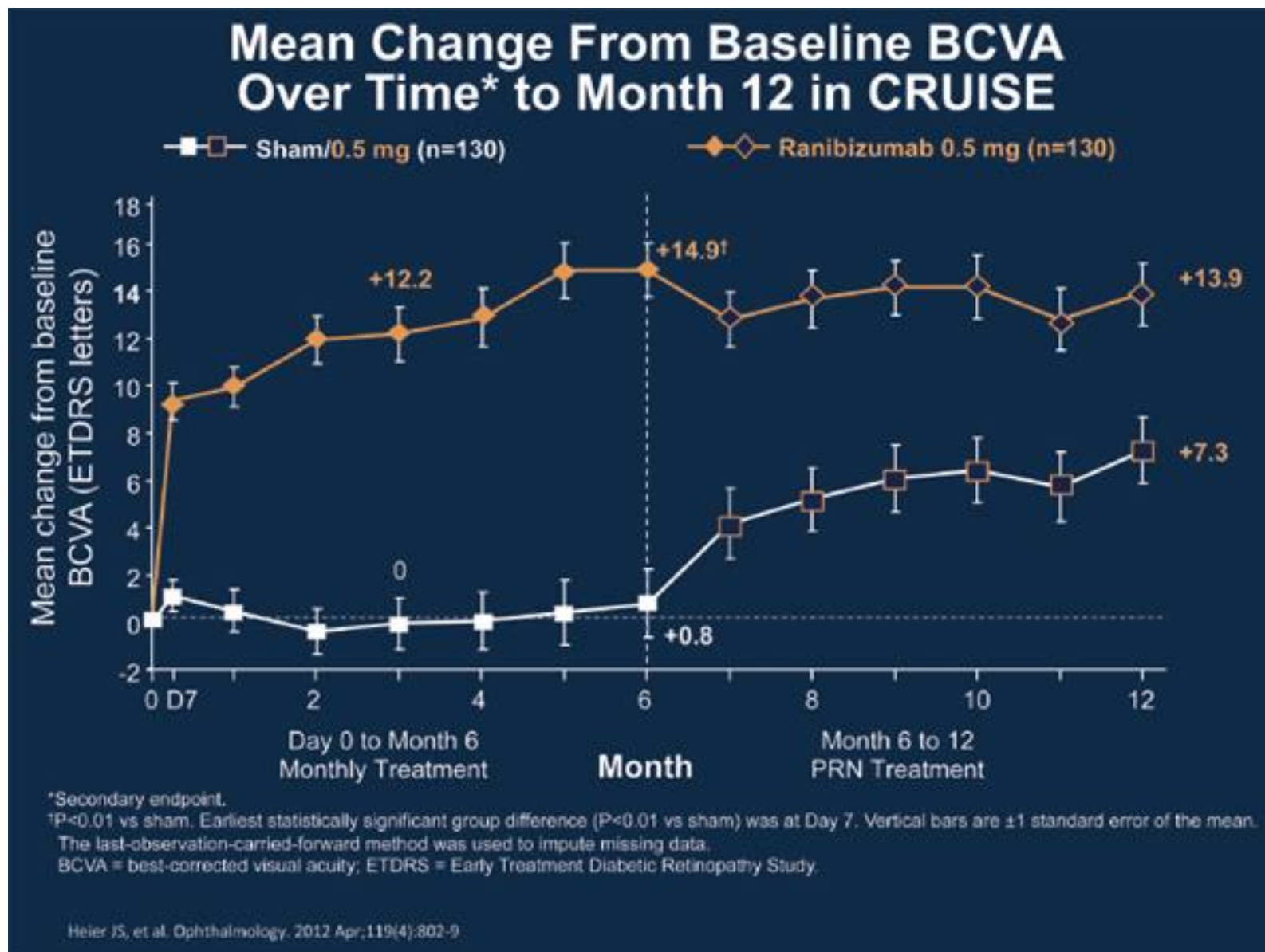
	Non-ischemic CRVO	Ischemic CRVO
Visual acuity	>20/200	<20/200
RAPD (relative afferent papillary defect)	Mild or absent	Present (>0.7 log units of neutral density filter)
Visual field defect	Rare	Common (use of Goldmann perimeter is suggested, as 30 degree field misses peripheral changes)
Fundus appearance	less disc/macular edema, hemorrhage, cotton-wool spots Mild venous tortuosity and dilation	More disc/macular edema, hemorrhage, cotton-wool spots Severe venous tortuosity and dilation
Fundus fluorescein angiogram	Less area of nonperfusion	Retinal capillary nonperfusion more than 10 disc areas
ERG/electroretinogram	Normal	Reduced b wave amplitude ($\leq 60\%$ of the normal mean value of both photopic and scotopic ERG), and reduced b/a
Prognosis	Good, less chance of anterior segment neovascularization/neovascular glaucoma	Poor, high chance of anterior segment neovascularization/neovascular glaucoma The visual prognosis may be worse than central retinal arterial occlusion



Central Retinal Vein Occlusion

- If macular edema present:
 - First Line: Anti Vegf with bevacizumab, ranibizumab, or aflibercept
 - 2nd Line: intravitreal triamcinolone or dexamethasone implant
- CRUISE study (2010)
 - 0.3mg ranibizumab vs 0.5mg vs sham
 - 46.2-47.7% gained more than 15 letters compared to 16.9%
 - 43.9- 46.9% better than 20/40 compared to 20.8%

CRUISE Study



Retinal Physician

LEAVO Study (2018)

	Ranibizumab	Aflibercept	Bevacizumab
Mean (SE) BCVA at baseline	53.6 (1.2)	54.1 (1.2)	54.4 (1.1)
Mean (SE) BCVA at 52 weeks	65.4 (1.6)	67.2 (1.5)	66.4 (1.6)
Mean (SE) BCVA at 100 weeks	65.6 (1.7)	68.4 (1.6)	64.6 (1.8)
Mean (SD) gain in BCVA at 100 weeks	12.5 (21.1)	15.1 (18.7)	9.8 (21.1)
≥15 letter gain in BCVA from baseline at week 100	47%	52%	45%
≥30 letter loss in BCVA from baseline at week 100	5%	2%	6%
OCT CST <320 μm at week 100	66%	81%	59%
Number of injections at week 52	8.1	7.1	8.1
Number of injections at week 100	11.8	9.8	11.5

Via Eye
News

TRUCKEE Study- 2022

- Faricimab (Vabysmo)
- Study consisted of patients treated with Eylea, Lucentis, Beovu, and treatment naive
- Results
 - +0.5 letters
 - Decrease in CST by 32 microns
 - PED height decreased by 14 microns

Case: PT RK

- WBC count came back severely elevated
- LP showed malignant lymphocytes- pt diagnosed with leukemia
 - Infiltrative optic neuropathy causing secondary CRVO
 - Due to poor prognosis of visual recovery with ischemic CRVO retinal specialist decided to forgo injections
- Patient started therapy for cancer

Case: 29 year old male

- CC: New floaters in my left eye for 3 days.
- No flashes
- No curtain or veil
- No trauma
- No recent illnesses
- Progressive cloudy haze
- Mild ache
- No recent sicknesses

Case

- **Visual acuity:**
 - OD 20/20
 - OS 20/25
- **IOP:**
 - 18/18
- **Pupils:**
 - No RAPD
- **Anterior segment:**
 - Mild trace cells in AC OS; no flare; cornea clear

Case

- **Dilated Fundus Exam:**
 - 2+ vitreous cells centrally
 - Inferior peripheral snowballs
 - Mild vitreous haze
 - No retinal breaks
 - No hemorrhages
 - No vascular sheathing

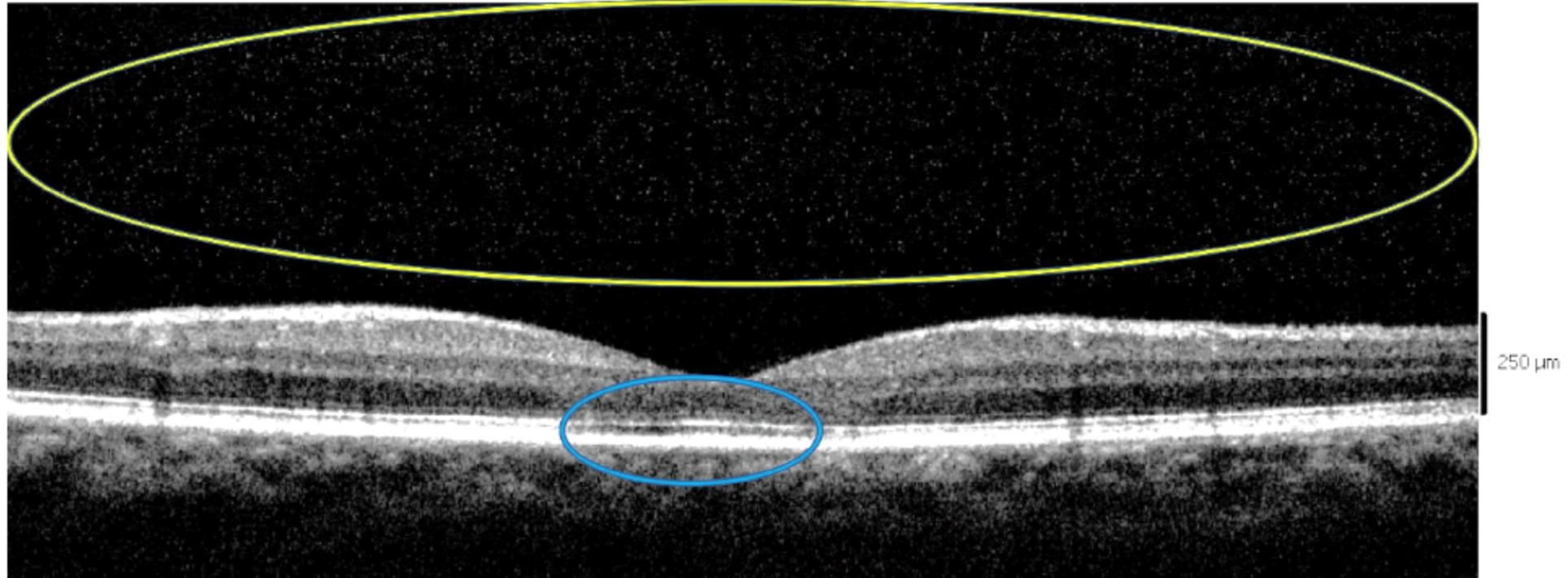


Figure 3: OCT is the hyper reflective spots in the vitreous cavity, suggestive of vitritis (yellow circle) and rupture at the level of the photoreceptor layer (blue circle).

- **OCT Macula:**

- Vitreous hyperreflective dots
- No CME (but up to 50% of intermediate uveitis develops CME)

Case: DDx

- High-likelihood diagnoses
 - Intermediate uveitis (pars planitis)
 - Posterior uveitis (viral, toxoplasmosis, TB, syphilis, Lyme)
- Moderate likelihood
 - Sarcoidosis-associated uveitis
 - Multiple sclerosis–associated uveitis

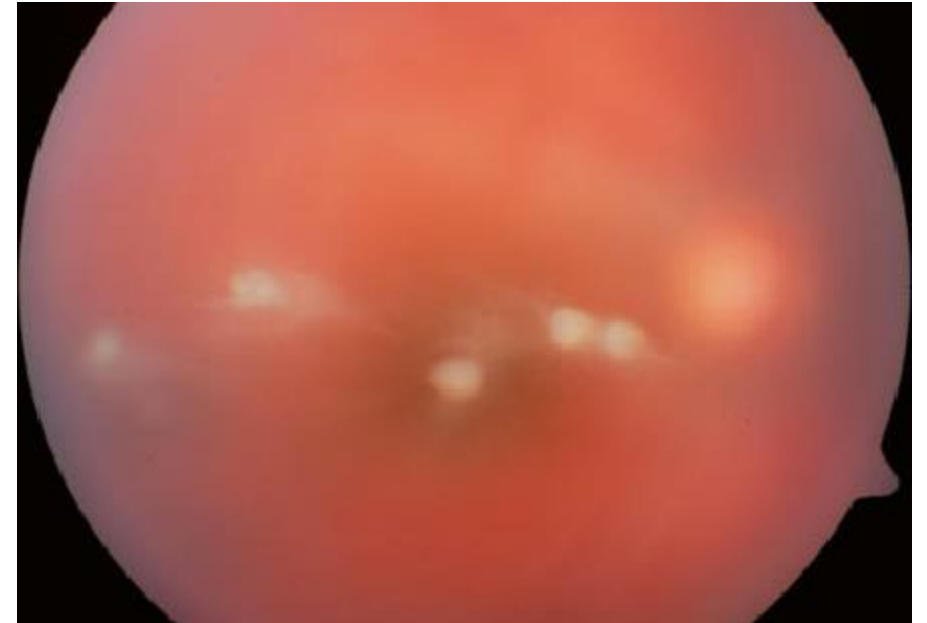
Posterior Uveitis

- Ocular lymphoma (Primary Vitreoretinal Lymphoma)
 - Average delay in diagnosis: 12–18 months
 - Looks EXACTLY like mild vitritis
 - Vitritis with NO pain + NO photophobia + older age or immune compromise
- Syphilitic posterior uveitis
 - Incidence rising by over 300% in last decade (CDC Surveillance Report, 2022)
- Drug-induced uveitis — emerging cause
 - Newer immune checkpoint inhibitors
 - Bisphosphonates
- Tuberculosis
 - TB-related posterior uveitis represents 8–13% of all posterior uveitis in endemic areas
- COVID / Post-viral Vascular Uveitis
 - Emerging literature shows:
 - Microangiopathic outer retinal changes
 - Increased cases of AMN
 - And inflammatory vitreous reactions

Posterior Uveitis Pearls

- **Age is your biggest diagnostic clue**
 - Floaters in a 29-year-old: uveitis until proven otherwise.
- **Snowballs = intermediate uveitis**
 - Classic, pathognomonic aggregates in the inferior vitreous.
- **Use a 78D or 90D lens**
 - Vascular sheathing is often subtle in early posterior inflammation.
- **OCT: Look for CME**
 - The most common complication and cause of vision loss.
- **Ask neurologic questions**
 - Paresthesia
 - Limb numbness
 - Balance issues

These screen for MS-related uveitis.



NIH Vitreous Haze Grading Scale

- 0- Normal findings of no haze
- 0.5- trace haze is defined as slight blurring of the optic disc margin.
- 1- Obscured view with definition to the optic nerve head and retinal vessels
- 2- Obscured view with definition to the retinal vessels.
- 3- the optic nerve head is visualized, but with blurry borders.
- 4- An obscured fundal view

Posterior Uveitis Pearls

- **30–45° off-axis illumination**
 - Reveals *faint* vitreous cells better than standard optic section.
- **Dynamic examination trick:**
 - Ask patient to look quickly side-to-side: watch for “snowball lag.”
This is **pathognomonic** for vitreous aggregates.
- **Green (“red-free”) channel on widefield imaging**
 - Highlights vascular sheathing even when subtle.
- **OCT biomarkers of inflammation**
 - **Vitreous reflectivity index (VRI)** objectively quantifies vitritis
 - **Hyperreflective dots** are correlated with disease activity
- Order a brain MRI if:
 - Young patient
 - Bilateral intermediate uveitis
 - *Intermediate uveitis is the presenting sign in up to 1% of MS patients.*

Posterior Uveitis Work Up

- Infectious Panel:
 - RPR + FTA-ABS
 - Quantiferon or PPD
 - IgG/ IgM (Toxoplasmosis)
 - Lyme Titers
- Autoimmune Panel
 - ACE/ Chest Xray
 - HLA-B27
 - ANA/ RF
 - Potential: IL-10 : IL-6 Ratio
- Imaging
 - OCT of nerve and macula
 - Widefield FA

Posterior Uveitis Treatment

- Topical Steroids
- Posterior subtenon triamcinolone
- Prednisone .5-1mg/kg/day
 - Taper on improvement

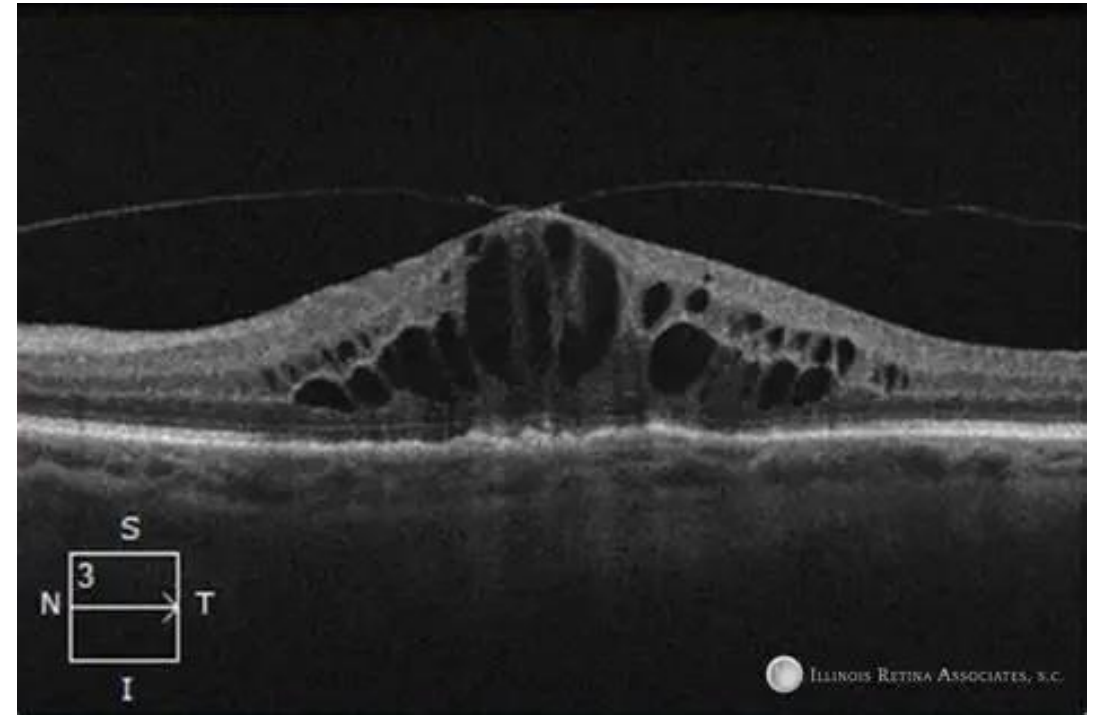
- Immunomodulators
 - Methotrexate
 - Mycophenolate
 - Azathioprine

Posterior Uveitis Advanced Treatments

- **Biologic Agents**
 - **Adalimumab (Humira)**
 - FDA-approved for non-infectious intermediate/posterior uveitis
 - Reduces recurrence and steroid dependency
 - Reduced recurrence 50-70%
 - NJEM RCT shows high potential for steroid sparing resolution
 - **Infliximab**
 - Effective in refractory cases
 - JAK Inhibitors: block intracellular inflammatory signaling- testing
- **Long-acting steroid implants**
 - **Fluocinolone acetonide implant (Iluvien)**
 - 36-month duration
 - Significant reduction in recurrence
 - **Dexamethasone implant (Ozurdex)**
 - Works for ~3 months
 - Useful for CME or recurrent uveitis
- **Intravitreal Biologics**

Complications

- Cystoid macular edema (CME) (50%)
- Cataract from chronic steroids
- Glaucoma
- Retinal vasculitis leading to ischemia
- Epiretinal membrane formation
- **Follow-Up & Monitoring**
 - **1–2 weeks** after initiating treatment
 - Monthly once stable
 - OCT every 4–8 weeks if CME present
 - FA every 3–6 months in vasculitic cases
 - Regular IOP checks

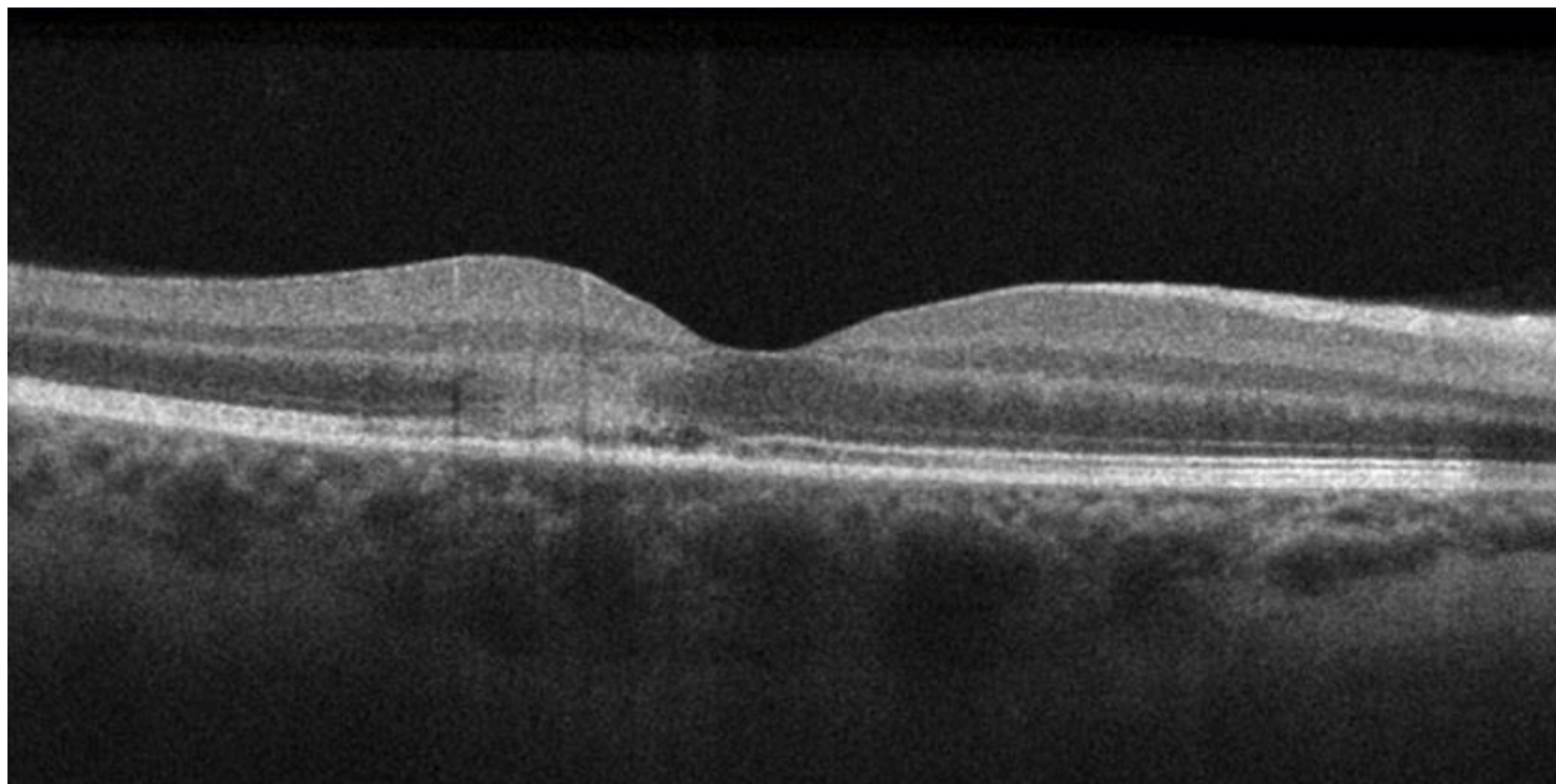


32 year old female

- **CC:** "I woke up with a dark smudge near the center of my vision."
- Describes small paracentral scotoma OS
- No flashes, no floaters
- No pain
- Says, "It's like a thumbprint, but only in certain lighting."
- Had a bad flu 2 weeks ago, took decongestants
- She went to Urgent Care and was told it's "probably ocular migraine."

Case

- **Visual Acuity**
 - 20/20 OU
- **Pupils**
 - No RAPD
- **Anterior Segment**
 - Normal
- **Dilated Fundus Exam**
 - Normal

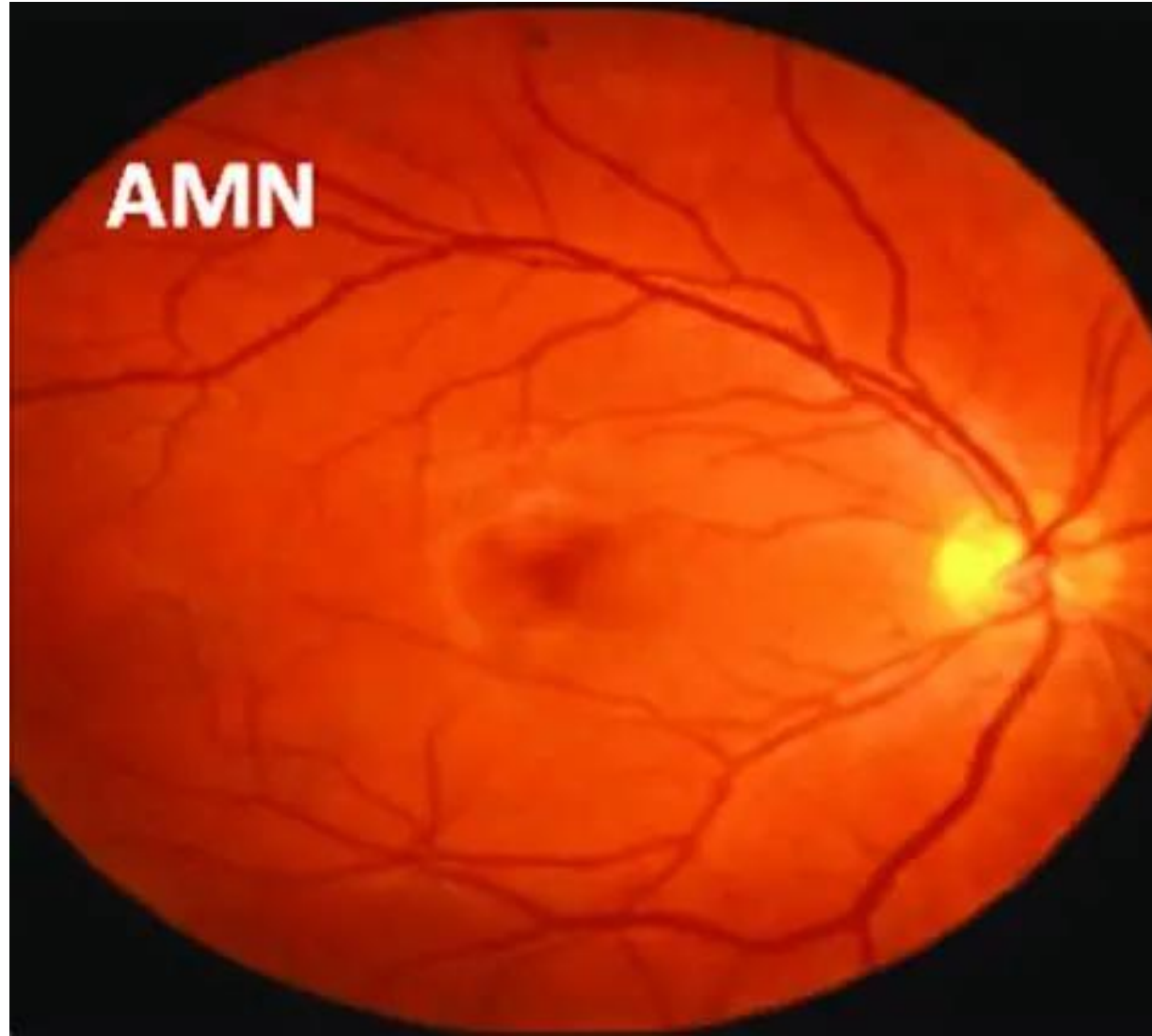


Case: DDX

- **Layer Model**
 - **Outer Retina (AMN, PAMM, early macular dystrophy)**
 - AMN = OPL/ONL lesion
 - PAMM = middle retinal layers (INL + intermediate plexus)
 - Early dystrophy = photoreceptor loss but no hyperreflective plaque
 - **Inner Retina (retinal vasospasm, BRAO)**
 - Would have whitening, RNFL hyperreflectivity
 - OCT pattern is very different
 - **Choroidal Causes (CSC, early MEWDS)**
 - CSC = detachment
 - MEWDS = white dots + disrupted EZ but no OPL plaque

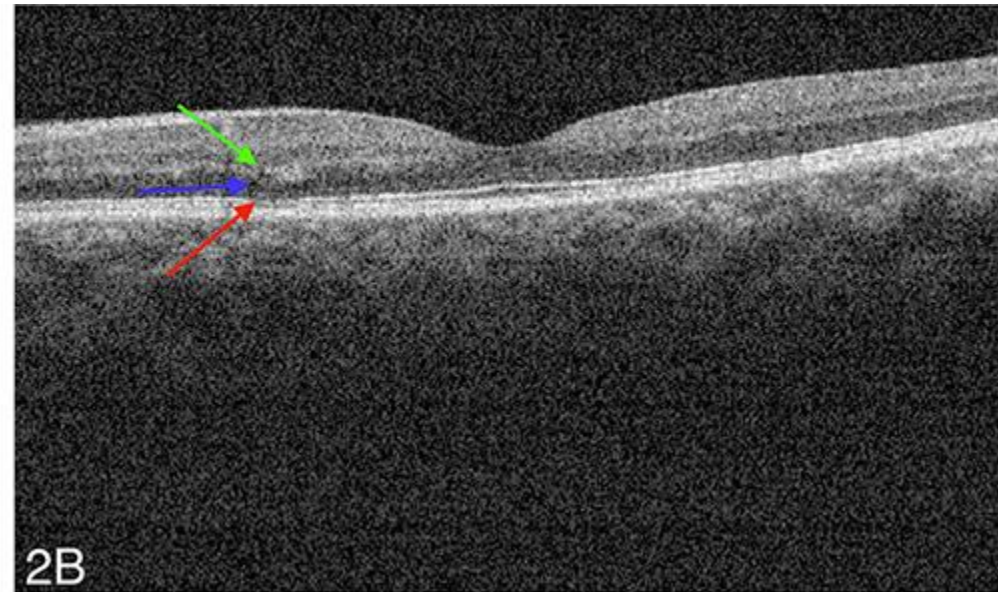
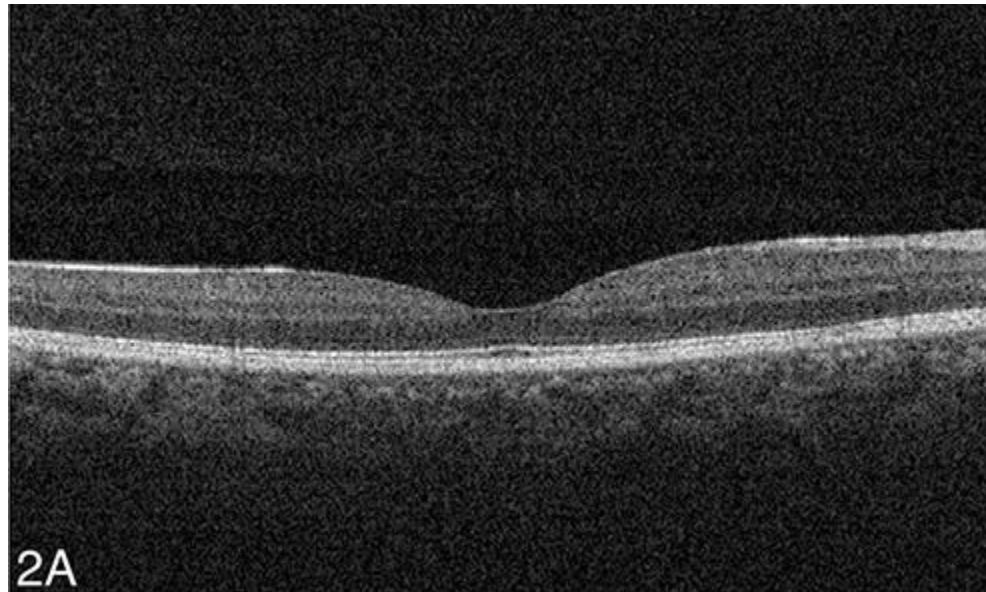
Acute Macular Neuroretinopathy (AMN)

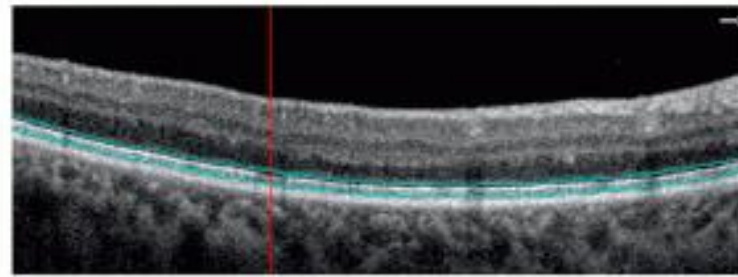
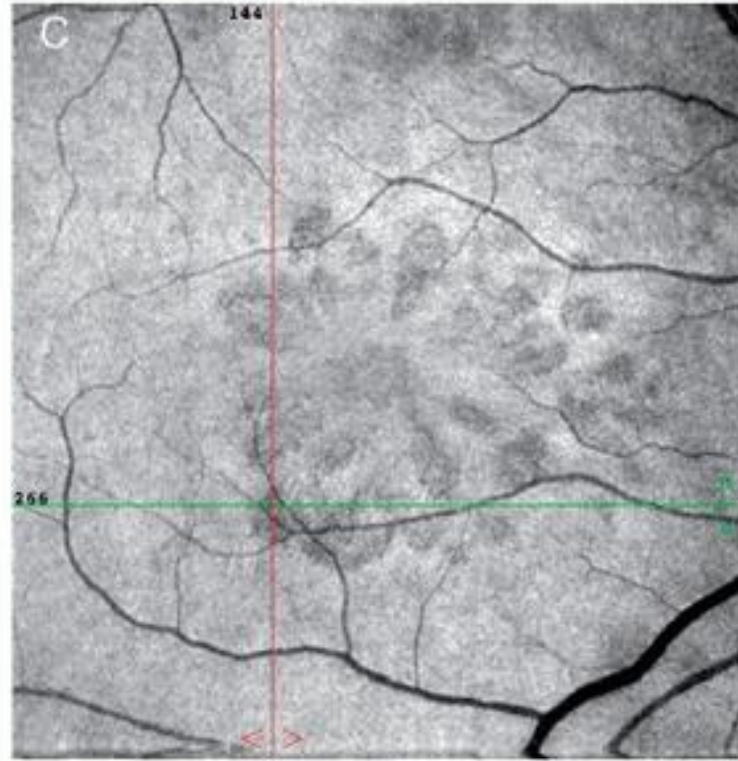
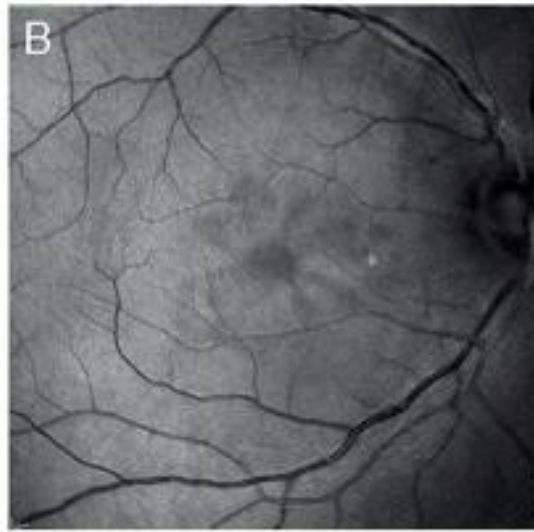
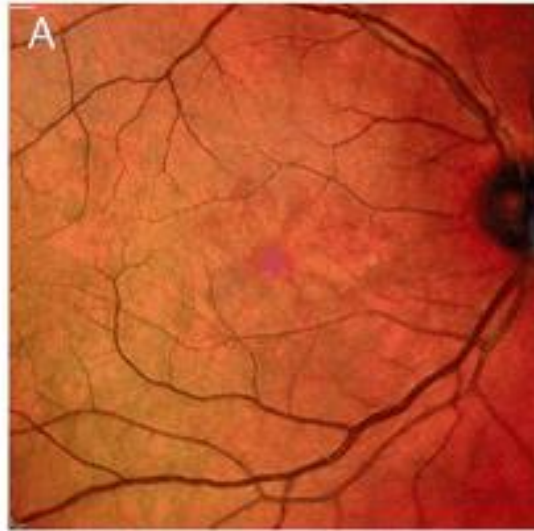
- **Epidemiology**
- Incidence estimated at **<1 per million** (rare).
- But **post-viral and vaccine-associated cases increased significantly from 2020–2023**.
- **90% of patients are young women** (often on hormonal contraceptives).



Acute Macular Neuroretinopathy (AMN)

- OCT:
 - Hyperreflective plaque in the **outer plexiform layer (OPL) + outer nuclear layer (ONL)**
 - Disruption of the **ellipsoid zone (EZ)**
 - Possible thinning of outer retina over time
- **OCT-Angiography**
 - **Deep capillary plexus (DCP) dropout**





AMN Diagnostic Pearls

- Recent viral illness
- Fever, dehydration, caffeine intake
- Intense exercise
- Hormonal contraception
- Vasoconstrictive medications (Sudafed, caffeine pills, energy drinks)

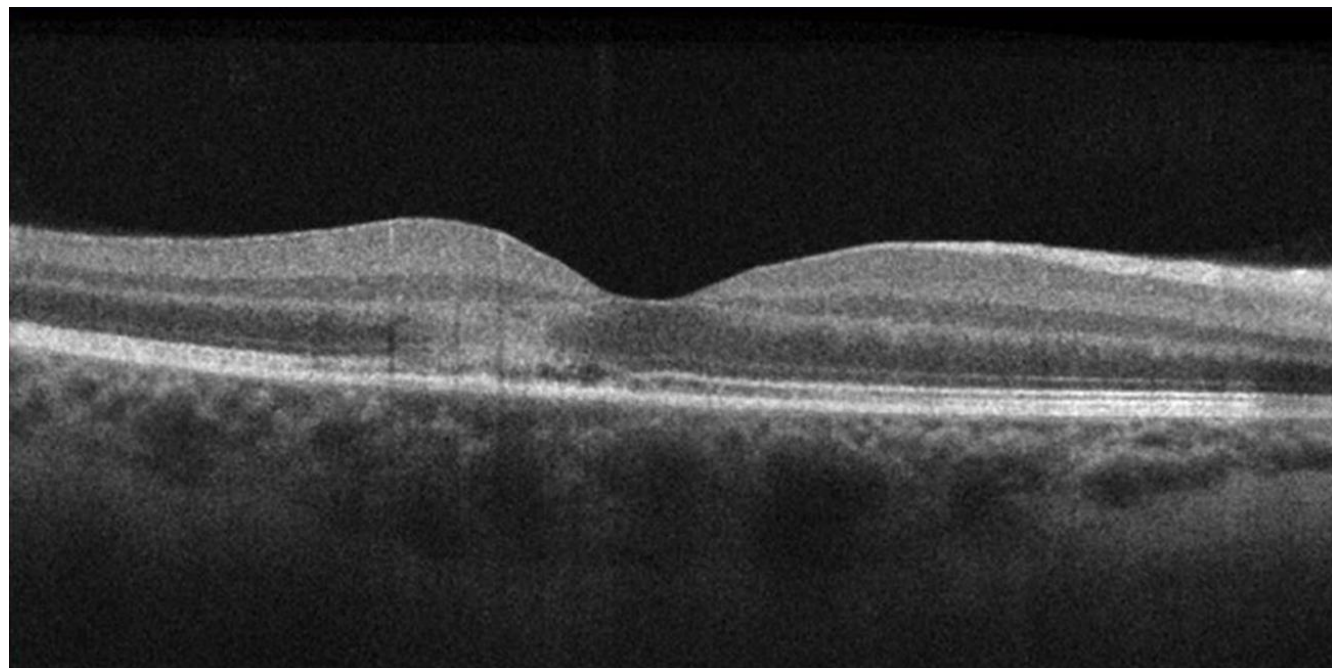
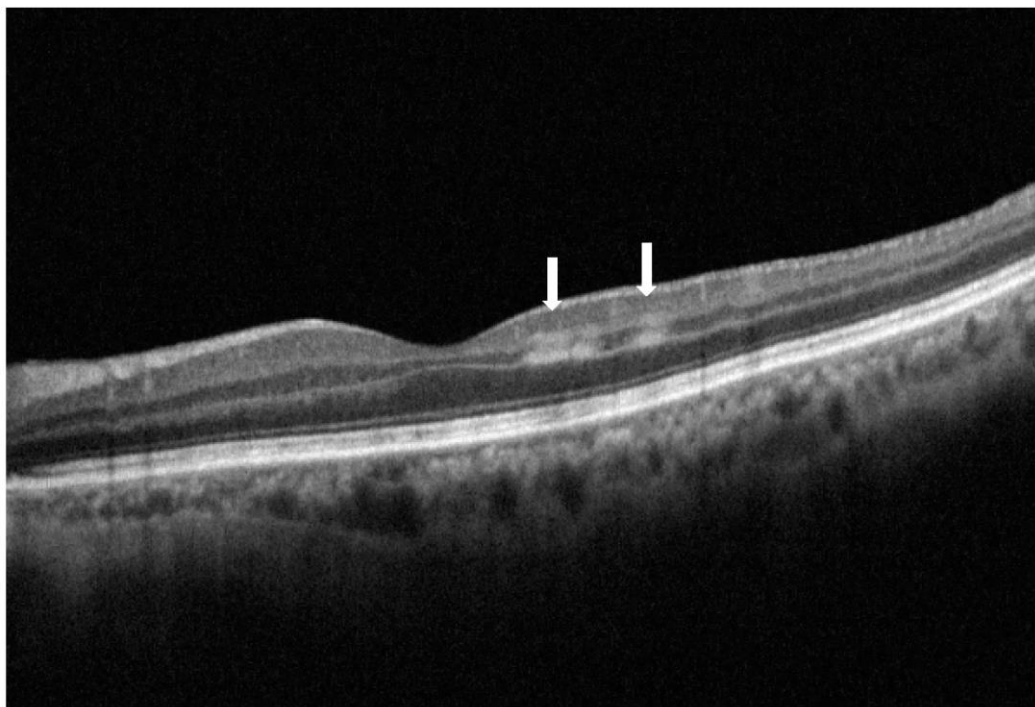
- Use infrared (IR) imaging
 - IR is the single most sensitive clinical tool for detecting AMN lesions.

- Paracentral scotoma with normal fundus = do OCT immediately

- Look at the outer retina CLOSELY
 - AMN lives at the OPL/ONL junction — many clinicians focus only on the inner retina when scotomas are reported.

- Ask about COVID exposure
 - AMN incidence spiked after both COVID infection and vaccination.

AMN vs PAMM



AMN Treatment

- Traditionally no treatment
- Emerging Evidence
 - **Steroids may accelerate recovery**
 - Case series show quicker improvement in some AMN cases.
 - **NSAIDs may help reduce deep capillary plexus ischemia**
 - Evidence limited but plausible.
 - **Avoid vasoconstriction**
 - Stop caffeine, Sudafed, smoking, energy drinks.
 - Potential Neuroprotection

AMN Treatment

- Trials
 - Pentoxifylline
 - Improves red blood cell deformability
 - Potentially increases capillary plexus perfusion (ophthalmology Retina 2019)
 - Rho-kinase inhibitors
 - Improves choroidal blood flow
 - Hyperbaric O₂ Therapy
 - Case report suggesting improvement in AMN
 - Subthreshold micropulse laser
 - Limited studies

Prognosis

- 50–75% of patients recover partially
- 25–30% have permanent small scotomas

Ellipsoid zone disruption may persist

- **Prognosis worsens if:**
 - Lesions are multiple
 - Patient has vascular risk factors
 - Delayed diagnosis (>7 days)
 - Deep capillary plexus non-perfusion is extensive

Case 45 year old male

- **CC:** "My eye hurts and lights hurt."
- Photophobia developing rapidly
- "Foggy" central vision
- New floaters
- Deep ache around the orbit
- Slightly decreased color saturation
- Had shingles on his torso 3 months ago
- Works outside, no known trauma
- No recent travel or systemic illness

Case

- **Visual Acuity**
 - OD 20/20
 - OS 20/60
- **Anterior Segment**
 - Mild anterior chamber cell
 - No keratic precipitates
- **Posterior Segment Findings —**

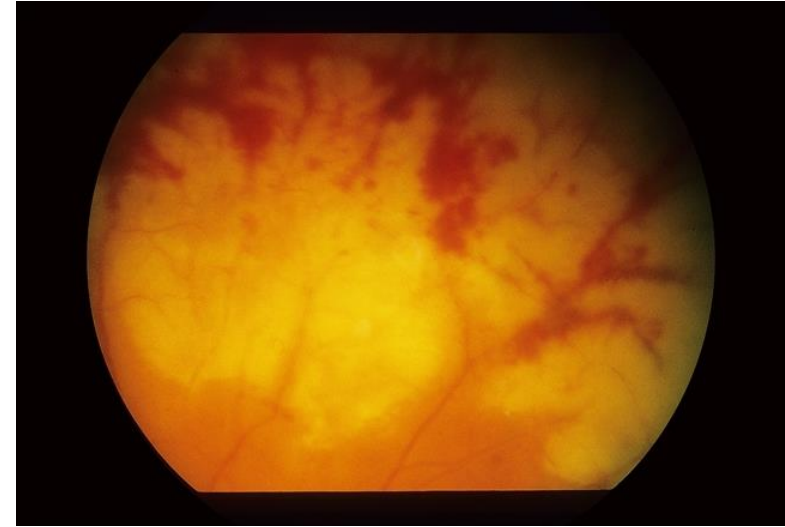


Case

- **Ultra-Widefield Imaging:**
 - 3+ vitritis
 - Peripheral yellow-white patches of necrosis
 - Arteriolar vasculitis — arteries more affected than veins
 - “Granular” whitening
 - Full peripheral necrosis
 - Segmental artery occlusion
 - Peripheral hemorrhages
 - Areas of impending retinal tear formation
- **OCT:**
 - Hyperreflective necrotic retina
 - Disruption of the outer retinal layers
 - Choroidal thickening

DDX

- **Acute Retinal Necrosis (ARN) — VZV/HSV**
 - Symptoms: pain + photophobia + vitritis + peripheral necrosis
- **Progressive Outer Retinal Necrosis (PORN)**
 - Occurs in immunocompromised
 - Minimal vitritis
 - Very rapid progression
 - CMV/HSV/VZV possible
(Usually “necrosis WITHOUT inflammation” = PORN)
- **Toxoplasmosis**
 - Focal lesion + dense vitritis
 - But toxo is usually adjacent to a pigmented scar
- **Syphilitic Necrotizing Retinitis**
 - Elevated RPR
 - Multifocal but often symmetrical
 - Less “marching” pattern
- **Cytomegalovirus Retinitis**
 - Immunocompromised
 - “Cheese and ketchup” pattern
 - Venules more involved than arterioles



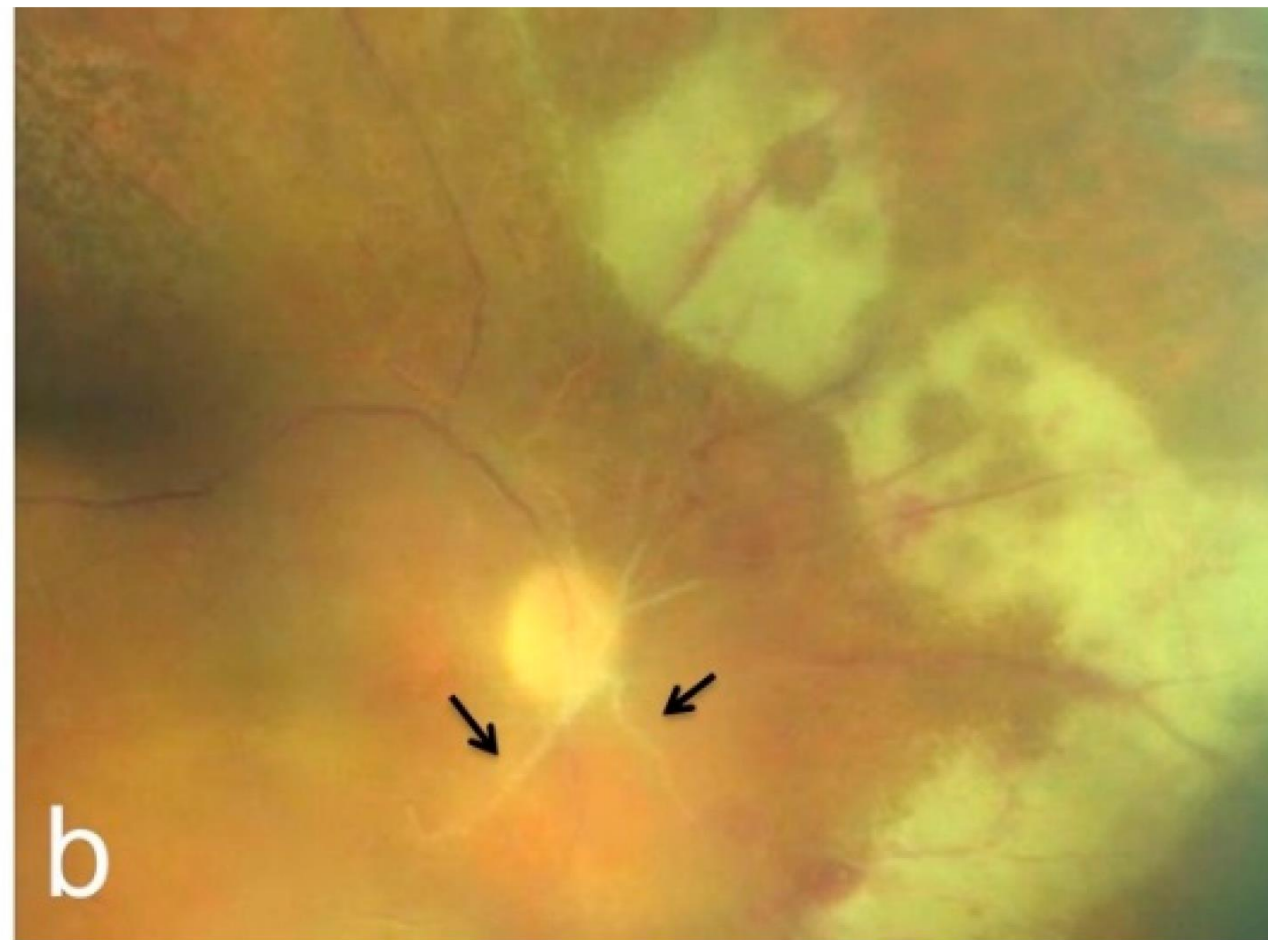
Acute Retinal Necrosis (ARN)

- **Key Statistics**
- ARN leads to **retinal detachment** in **47–75%** of cases if untreated
- Vision worse than 20/200 occurs in **~50%** of cases
- VZV causes **68–74%** of ARN in adults
- Fellow-eye involvement occurs in **30–35%** without systemic antivirals

ARN Diagnostic Pearls

- **Pain + heavy vitritis = infectious until proven otherwise.**
 - Non-infectious uveitis almost never hurts this much.
- **Extreme Light Sensitivity**
- **Examine the ENTIRE periphery**
 - ARN begins in the periphery.
You MUST depress the peripheral retina — unless necrosis is severe (danger of tear).
- **Look at arteries first**
 - Arteries become “white sticks” due to occlusion.
- **The progression pattern is diagnostic**
 - Necrosis = starts as patches - merges - spreads circumferentially.
- **OCT-A shows deep capillary non-perfusion**
 - New research shows OCT-A can identify ARN before necrosis fully forms.

Posterior Segment Triad



ARN Work Up

- **PCR of aqueous/vitreous** for:
 - VZV
 - HSV-1, HSV-2
 - CMV
- **RPR/TP-PA** — syphilis is a great masquerader
- **HIV test** — helps distinguish ARN vs PORN vs CMV
- **CBC, CMP** — baseline for systemic antivirals
- **Imaging tests:**
 - **Ultra-widefield fundus photography**
 - **OCT** of necrotic regions
- **OCT-A** (if available)
- **MRI brain + orbits** if corticosteroid-responsive optic neuropathy suspected or neurologic symptoms present

Treatment- ARN

- **Acyclovir 10 mg/kg IV every 8 hours for 7–10 days**
- Transition to **valacyclovir 2 g TID** for 4–6 weeks
- **Alternate:**
- Famciclovir 500 mg TID
- **Intravitreal Antivirals**
 - Foscarnet 2.4 mg/0.1 mL
 - Ganciclovir 2 mg/0.05 mL
 - Intravitreal antivirals reduce retinal detachment risk by **up to 32%**.

Treatment- ARN

- Steroids
 - 48-72 hours after antivirals
 - 40-60mg daily
- Emerging Therapies
- **Prophylactic Laser Retinopexy**
 - Dramatic reduction in RD rates in multiple studies
 - Still controversial
- **Early Pars Plana Vitrectomy (PPV)**
- PPV reduces RD risk in cases with severe vitritis

Treatment- ARN

- **Combination Intravitreal Therapy**
 - Dual antiviral injections (ganciclovir + foscarnet)
 - Increased viral suppression
(Tian et al., Retina, 2019)
- **Anti-IL-6 therapies (experimental)**
 - Used in severe necrotizing retinitis
(very early data)
- **Genetic Susceptibility Studies**
 - HLA haplotypes associated with severe ARN
(Ongoing research — Johnson et al., 2022)

ARN

- **Without treatment:**
- 70–80% → severe vision loss
- 35% → fellow eye involvement
- 50–75% → retinal detachment
- **With aggressive early treatment:**
- RD rates drop
- Fellow eye involvement drops to <10%
- **Follow up**
 - **Every 24–48 hours initially**
 - evaluate new necrosis or RD risk
- **Weekly**- until stable
- **Monthly** - for first 6 months
- **Long term** - may need lifelong antiviral suppression in severe HSV/VZV cases

23 year old female

CC: "My vision is blurry when I bend over."

- Transient visual obscurations lasting 5–10 seconds
- Headaches worse in the morning
- Pulsatile tinnitus
- Occasionally sees "sparkles"
- Pain behind eyes when lying down
- On minocycline for acne

Case

- **Visual acuity:**

- 20/20 OU

Pupils:

- No RAPD

- **Color vision:**

- Slight red desaturation

- **Visual fields (HVF 30-2):**

- Enlarged blind spot OU



Case

- Elevated blurred margins
- Hyperemic nerve head
- Peripapillary wrinkles (Paton's lines)
- No spontaneous venous pulsation (SVP)

Case DDX

- **TRUE Papilledema (raised ICP)**
 - IIH
 - Tumor
 - Venous sinus thrombosis
 - Meningitis
 - Chiari malformation
- **Pseudopapilledema**
 - Optic disc drusen (buried or visible)
 - Small crowded hyperopic discs
 - Myelinated nerve fibers
 - Tilted discs
- **Other optic nerve edema**
 - Optic neuritis
 - Ischemic optic neuropathy
 - Toxic/nutritional neuropathy

Idiopathic Intracranial Hypertension

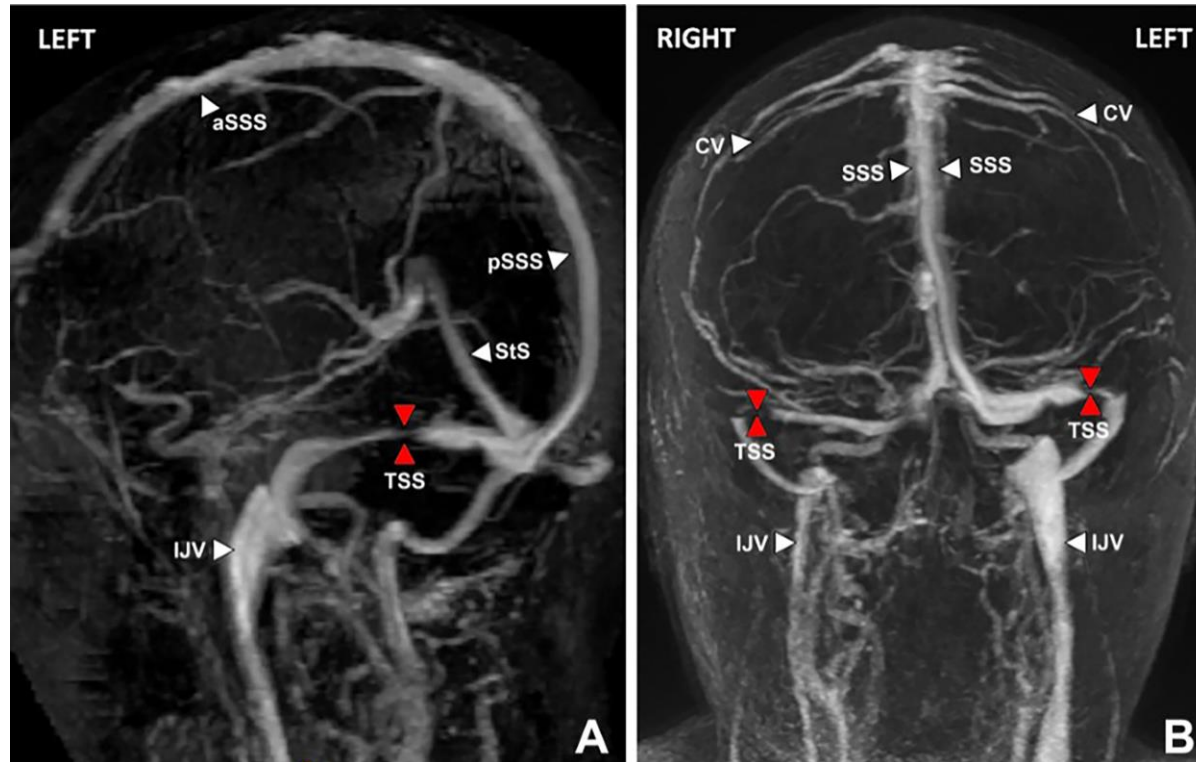
- Incidence: **1–3 per 100,000**, but **up to 20 per 100,000 in obese women**
- **25%** of patients develop some degree of permanent vision loss
- Up to **9–10%** risk of severe permanent vision loss
- Diagnostic delay = worse visual outcomes

Idiopathic Intracranial Hypertension IIG

- **Known associations:**
- Female gender
- Obesity
- Rapid weight gain (~5–15%)
- Tetracycline antibiotics
- Vitamin A derivatives
- Growth hormone
- Estrogen-containing contraceptives

IIH

- New research (2020–2023):
- Cerebral venous outflow obstruction
- Transverse sinus stenosis is present in **>90%** of IIH patients
(Studies show stenosis may be **cause**, not effect.)

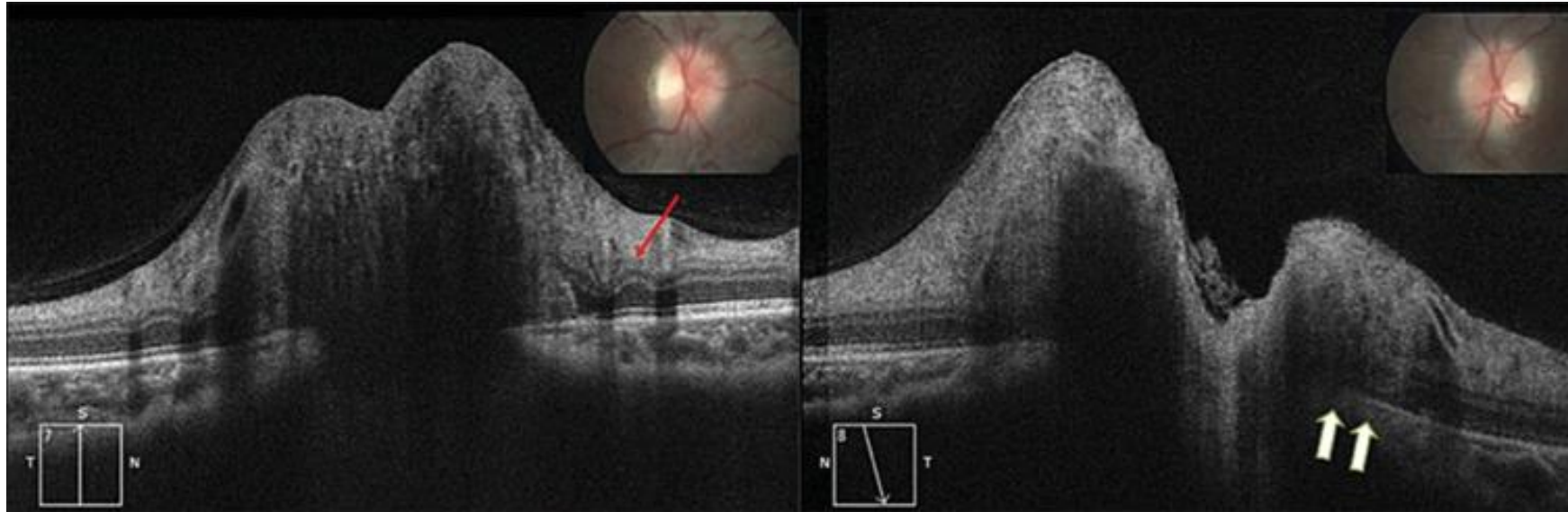


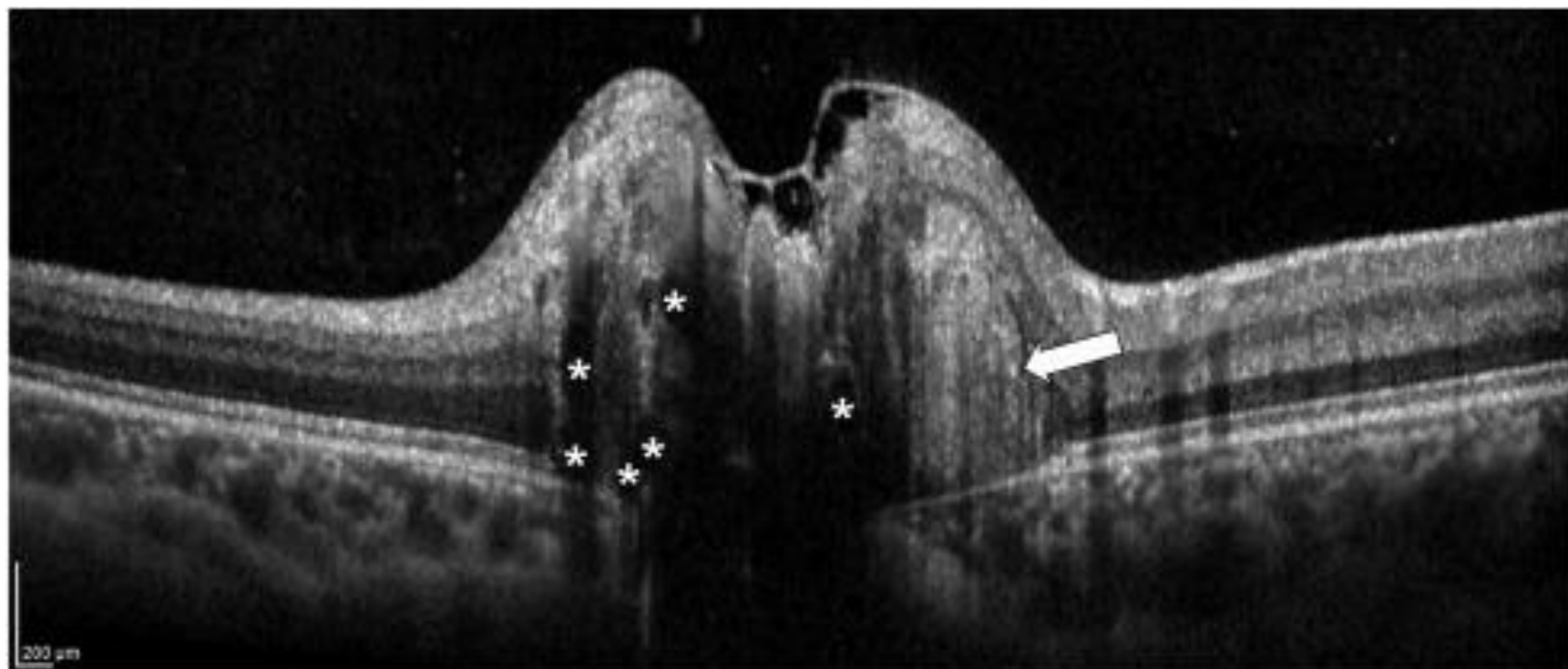
IIH

- Hyperemia
- Paton's lines (peripapillary folds)
- Peripapillary hemorrhages
- Loss of SVP
- SVP is present in **90%** of normal eyes.

OCT in papilledema vs pseudo

- Papilledema = **global RNFL thickening**, especially temporally.
Pseudopapilledema = **nasal elevation** with normal RNFL globally.





IIIH

- **Autofluorescence**

- ODD = bright autofluorescence spots

Papilledema = no autofluorescence

- **B-scan Ultrasound**

- ODD = highly reflective calcified bodies with **posterior shadowing**.

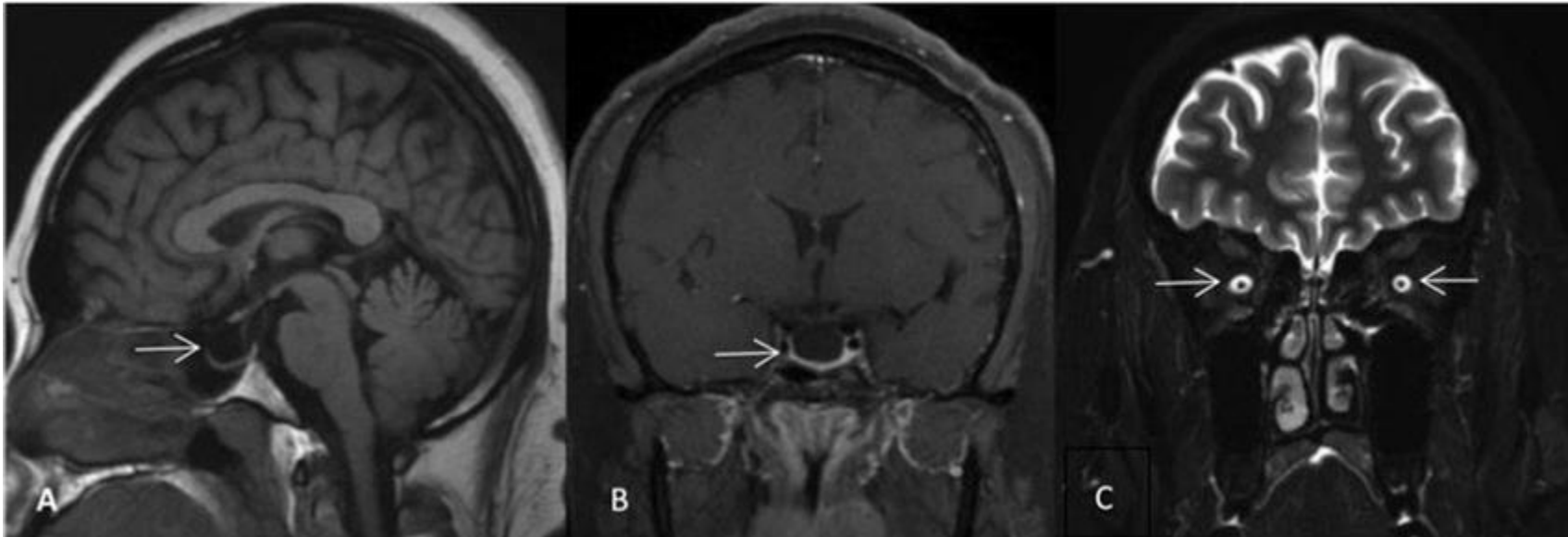
Papilledema = NO shadowing.

- Tell the patient to squat & stand quickly -- increased ICP transiently --

TVOs appear immediately in papilledema.

IIH Testing

- **MRI brain + MRV**
 - Rules out mass, hemorrhage, venous sinus thrombosis
 - Must be done **before** lumbar puncture



IIH Testing

- **Lumbar puncture (LP)**

- Opening pressure > **25 cm H₂O** = diagnostic
- CSF contents normal

(Friedman et al.)

- **Blood tests (optional but useful):**

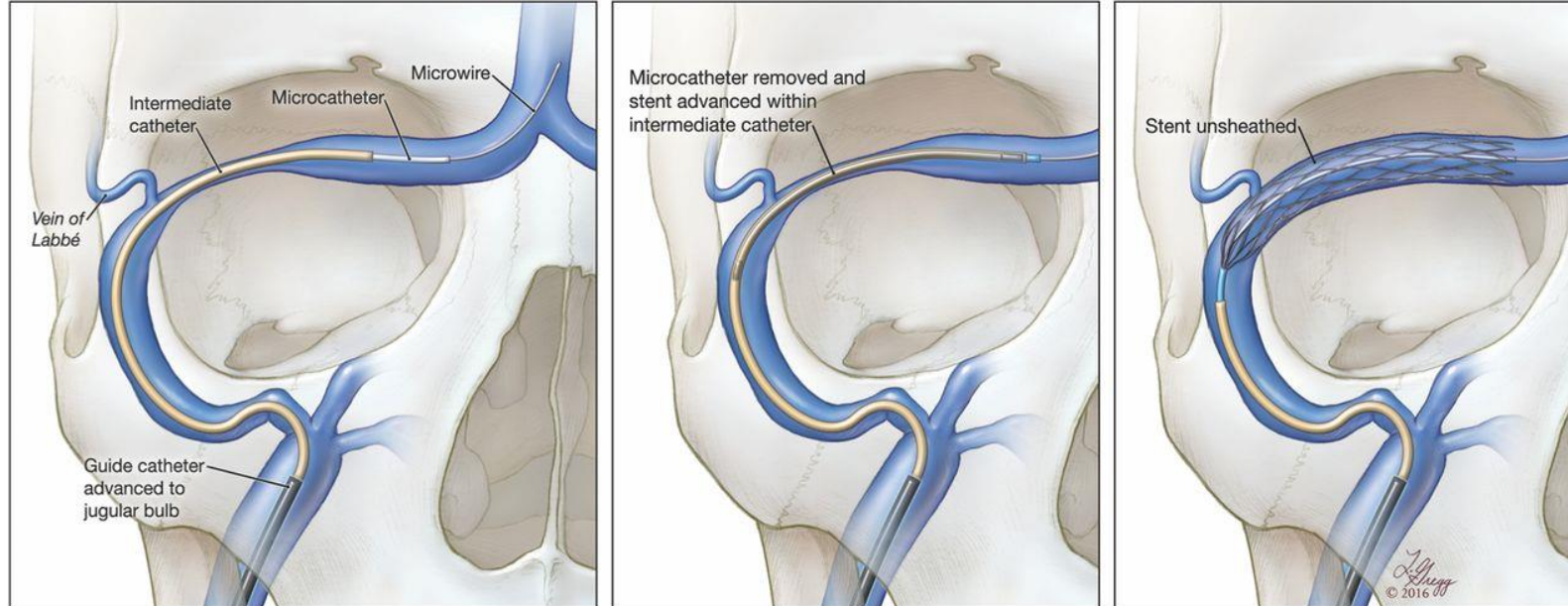
- CBC, CMP
- Thyroid panel
- Pregnancy test
- Hypercoagulable panel if sinus thrombosis suspected

IIH Treatment

- **Weight Loss**
 - 5–10% body weight loss can induce remission
- **Acetazolamide**
 - First-line medication
 - Starting dose: 500 mg ER BID
 - IIH Treatment Trial:
 - **Significant improvement in VF mean deviation**
- **Topiramate**
 - Bonus: migraine prophylaxis
 - Causes mild weight loss
 - Effective alternative
- **Corticosteroids**

IIH Treatment

- **Venous Sinus Stenting ("Future standard of care")**
 - Restores normal cerebrospinal fluid dynamics
 - Symptom improvement in **>90%**
 - VF improvement in **70%**
 - Lower recurrence rate than surgery



IIH Treatment

- **GLP-1 Agonists (Obesity-targeted therapy)**
 - Semaglutide & tirzepatide show dramatic weight loss
 - Early IIH case series show rapid improvement
 - A Phase II trial is underway specifically for IIH reduction
- **Telemetric ICP Monitoring**
 - Implantable devices continuously monitor intracranial pressure.

Monday Morning Pearls

- **If you see an elevated disc — prove it's real.**
- **No SVP? Check the RNFL NOW.**
- IR + EDI-OCT = ODD detection > 90%.
- TVOs are *hugely* predictive of true papilledema.
- Papilledema = MRI/MRV → LP → aggressive management.
- Acetazolamide works — but 5–10% weight loss works better.
- Transverse sinus stenosis likely plays a causal role — stenting is coming.
- Beware of minocycline, tetracyclines, retinoids.
- Follow fields closely — progression can be silent.
- Missing IIH is one of the top medico-legal risks in optometry.

Case 56 year old female

- **CC:** "My new glasses still don't make things sharp."
- Blurred and "washed out" central vision
- Difficulty reading
- No flashes or floaters
- No pain
- Symptoms worsened slowly over 18 months
- Past ocular hx: none

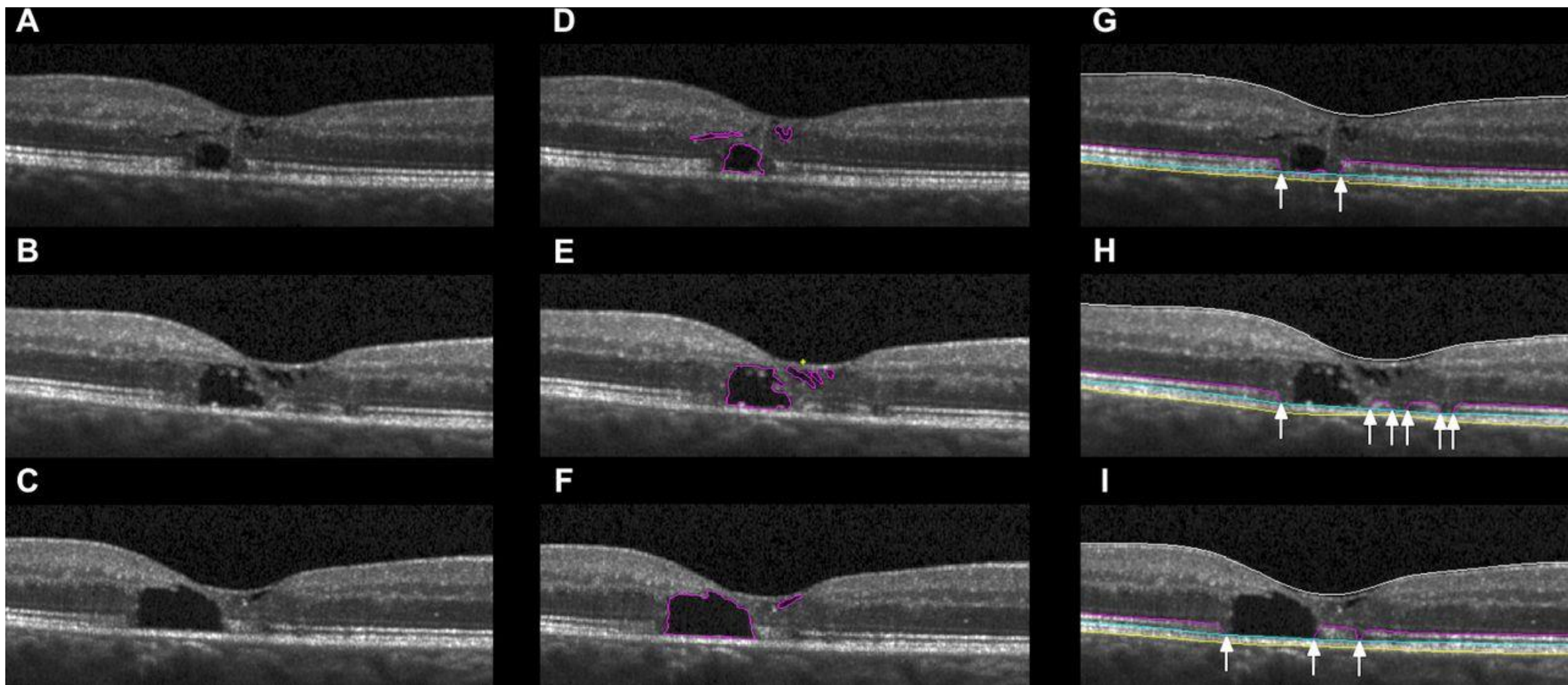
Case

- **Visual Acuity:**
 - 20/25 OU
- **Amsler Grid:**
 - Subtle paracentral metamorphopsia temporally.
- **Fundus Exam:**
 - Temporal parafoveal grayish discoloration
 - Loss of retinal transparency

Case

- **Autofluorescence (FAF):**
 - FAF shows a trilateral zone of hyperautofluorescence in the temporal macula
- **OCT**
 - Temporal foveal cavitations
 - Thinning of temporal inner retina
 - Breaks in the ellipsoid zone (EZ)
 - Loss of outer retinal layers ("square sign")
 - No CME
- **OCT-Angiography (OCT-A):**
 - Reduced flow in deep capillary plexus temporally
 - Enlargement of the FAZ
 - Early remodeling of capillaries





DDX

- **Dry AMD**
 - Has drusen
 - Has RPE mottling
- **Diabetic macular edema**
 - Has intraretinal fluid
 - DME is not always bilateral
- **Macular hole / lamellar hole**
 - OCT cavitations may be confused
 - MacTel2 doesn't create full-thickness defects early on
- **Paracentral Acute Middle Maculopathy (PAMM)**
 - More acute and painful
 - OCT shows INL hyperreflectivity
 - MacTel2 is a chronic degenerative disease

Macular Telangiectasia Type 2

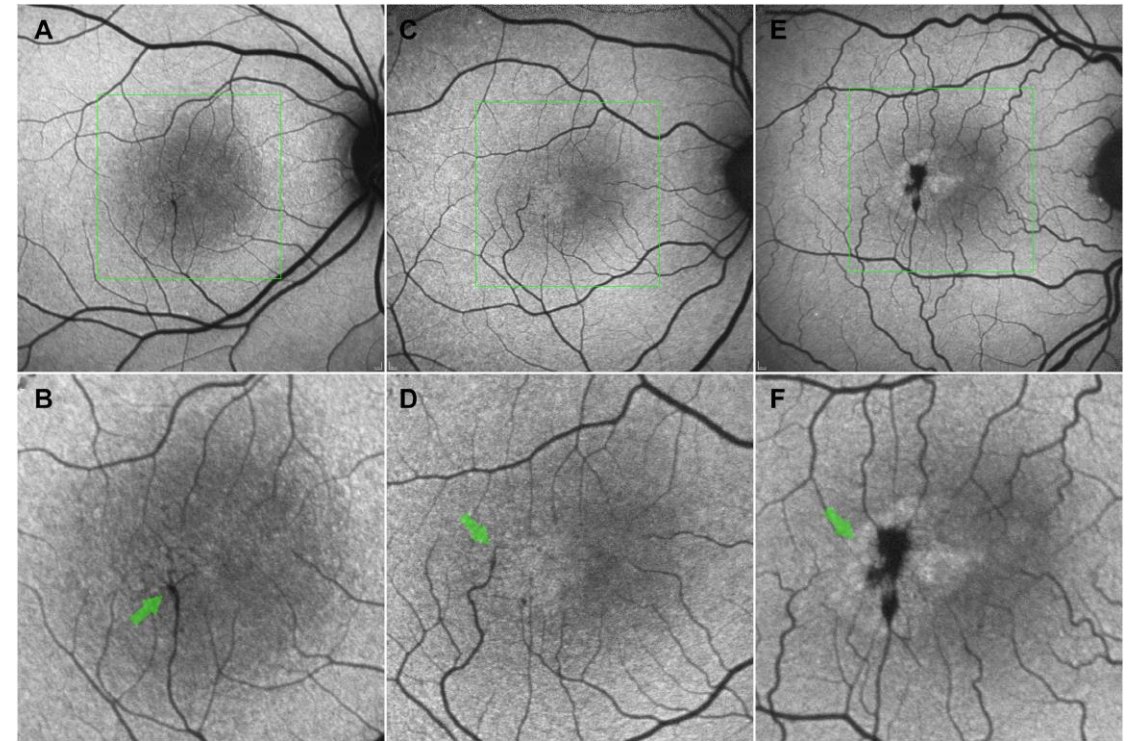
- Prevalence: approx. **0.1%–0.5%** in population studies
Bilateral in **almost 100%** of cases
- Strong association with **metabolic dysfunction** (diabetes, obesity, metabolic syndrome)
- Complications
 - Neovascularization (5–10%)
 - Requires anti-VEGF therapy.
 - Progressive photoreceptor loss
 - EZ breaks correlate strongly with vision loss.
 - Macular holes
 - Occasionally occur due to structural weakness.

MacTel Stages

- 1: Fovela reflex loss, OPL cavitation, peatoid IR
- 2: Crystals in ILM, right angle venules, ELM/EZ/IZ disruption, oval foveal hyper FAF
- 3-4: RPE plaque, Outer retinal atrophy, SRNV leakage, Hyper FAF

MacTel Diagnostic Pearls

- **Cavitation, Not Cyst**
 - MacTel cavitations are not fluid — they are **degenerative loss of tissue**
- They **do not respond** to anti-VEGF.
- **Look TEMPORALLY**
 - MacTel2 almost always begins temporal to the fovea.
- **Search for RIGHT-ANGLE VENULES**
 - A pathognomonic but subtle finding.
- **Autofluorescence is gold**
 - Hyper-autofluorescent patches temporally = high suspicion.
- **Think metabolic syndrome**
 - Strong association with systemic metabolic dysfunction.



MacTel 1 vs MacTel2

- Unilateral vs Bilateral
- Congenital vs acquired
- Systemic Correlations
- Symptoms

MacTel 2 Testing

- A1c
- Lipid panel
- Liver function tests
- Microalbumin/creatinine ratio
- May reveal underlying systemic contributors.
- **Imaging**
 - OCT
 - FAF
 - OCT-A (
 - Fluorescein angiography (FA)
 - Reveals temporal leakage in later disease stages
 - Shows telangiectatic changes

MacTel 2 Treatment

- **There is NO cure, but stabilization is possible.**
- **Anti-VEGF — NOT EFFECTIVE**
- **Low Vision Rehabilitation**
 - Early referral is helpful
 - Contrast sensitivity deficits are profound
- **Nutritional support**
 - Lutein/zeaxanthin may improve visual function modestly

Emerging Treatments

- **Revakinagene Taroretcel (Encelto) — First Gene/Cell Therapy**
 - **FDA-approved in March 2025 (U.S.)**
 - A combination **gene therapy + cell therapy**
 - Injected into the vitreous
 - Replenishes Müller cells affected in MacTel2
 - Clinical trials showed:
 - **Improved retinal sensitivity**
 - **Reduced photoreceptor loss**
 - Slowed disease progression

Emerging Treatments

- **Metabolic-targeted therapies**

- New studies show MacTel2 may be a **serine biosynthesis disorder** mediated by PHGDH gene variants.
- Treatments under investigation:
- Serine supplementation
- Dietary modification
- Glycemic control strategies

MacTel 2

- **Prognosis- good**
 - Early detection
 - Good systemic metabolic control
 - Small cavitations
 - Minimal EZ disruption
- **Prognosis- poor**
 - Extensive EZ loss
 - Neovascular complications
 - Severe metabolic dysfunction
 - Long-standing undiagnosed disease
- **Follow-Up**
 - Every 6–12 months for stable disease
 - Every 3–6 months if:
 - EZ disruption advancing
 - New symptoms
 - Considering emerging therapy enrollment