Rapid Fire Grand Rounds

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Financial Disclosure

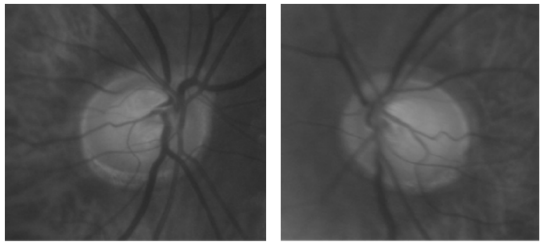
- Allergan
- Bausch & Lomb
- Carl Zeiss Meditec
- Ivantis
- Kala
- Santen

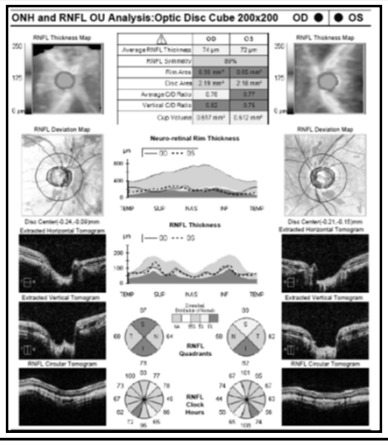
Case: Is This Glaucoma?

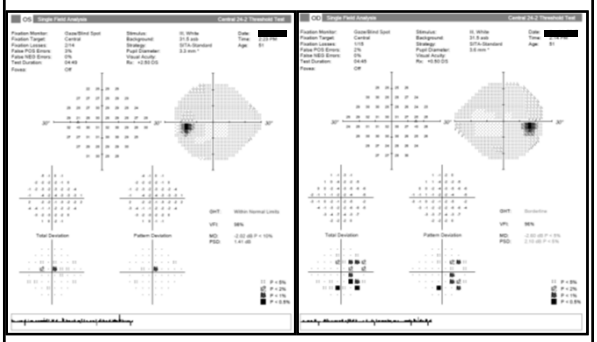
- Carlos, 51yo HM
- Referred for glaucoma suspicion due to ONH appearance
- POH:
  - LASIK OU (2000), PRK OS (2014)
- FOH:
  - (+) glaucoma (maternal gm)
- PMH: Unremarkable

Exam Findings

- Uncorrected VA: 20/20 OD, OS
- Pupils, motility, CVF: normal OD, OS
- SLE: LASIK flaps visible, otherwise normal OU
- Gonioscopy: open to CB 360° OD, OS
- Tmax: 18mmHg OU
- CCT: 523 OD 489 OS







Is This Glaucoma?

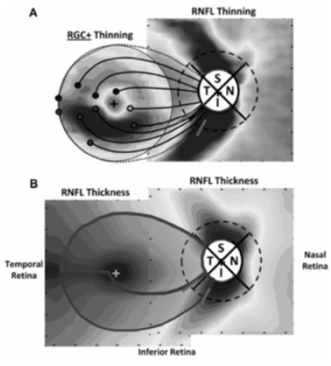
A. Yes
B. No
C. I need more information

Discussion Topics

- Macular Imaging in Glaucoma
- Imaging in the myopic eye

Newest Addition to Glaucoma Diagnosis Arsenal: Macular Imaging

- 1998: Zeimer et al reported on macular thickness loss in patients with known glaucomatous damage
- 2003: Greenfield reported correlation between total macular thickness and MD on VF in glaucoma patients (time domain OCT)
- 2013: Hood et al – extensive investigation of segmented "RGC+" (RGC + IPL) layer and description of the "Macular Vulnerability Zone" (MVZ)



TECHNOLOGY TODAY

Measuring Macular Thickness in Glaucoma

Clinicians must be aware of important differences in the macular imaging protocols of the commercially available optical coherence tomography systems.

BY AHMAD A. AREF, MD

TABLE. COMPARISON OF COMMERCIALLY AVAILABLE IMAGING DEVICES FOR MACULAR ANALYSIS IN GLAUCOMA

OCT Device	Macular Imaging Protocol	Macular Area of Analysis	Macular Layers Analyzed	Normative Database?
RTVue FD-OCT	Ganglion cell complex analysis	7 mm², centered 1 mm temporal to fovea	RNFL, RGC, IPL	Yes
Spectralis SD-OCT	Posterior pole asymmetry analysis	8 mm², centered on fovea	All macular layers	No
Cirrus HD-OCT	Ganglion cell analysis	Elliptical annulus (vertical radius of 2 mm, horizontal radius of 2.4 mm), centered on fovea	GC-IPL	Yes

Abbreviations: OCT, optical coherence tomography; RNFL, retinal nerve fiber layer; RGC, retinal ganglion cell; IPL, inner plexiform layer; GC-IPL, ganglion cell and inner plexiform layers.

RT-Vue

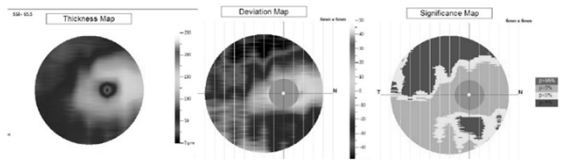


Figure 3. The Thickness, Deviation, and Significance Maps for a glaucoma patient. In the Deviation Map note the blue and black regions inferior and superior to the macula, corresponding to a 25% loss and 50% loss of GCC in those areas respectively. The center of the macula has a mask over the fovea because there are no ganglion cells in this area. The color scale to the right shows the percent loss associated with each color. Cooler colors such as blue and black represent areas with more loss.

RT-Vue

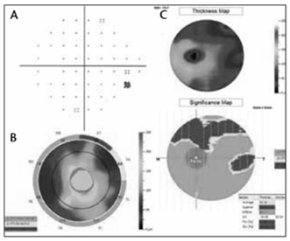
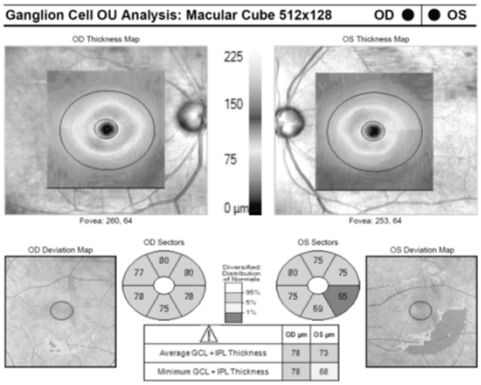
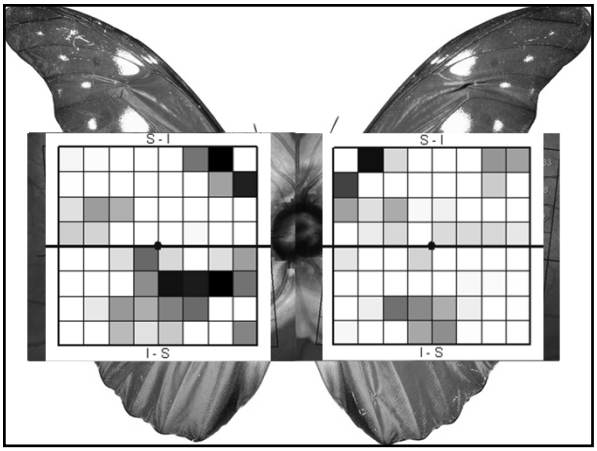


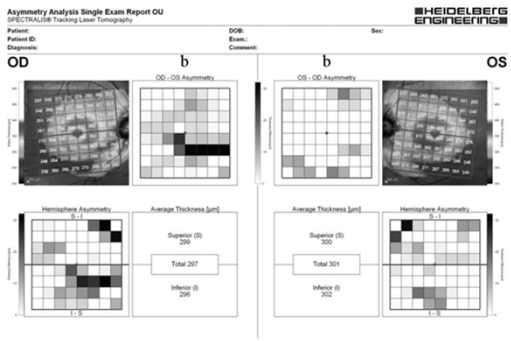
Figure 1. Automated visual field pattern deviation plot (A),

Cirrus OCT





Spectralis - macula



Advantages of Macular Analysis

- Macula contains ~50% of retinal ganglion cells
  - Glaucoma is a disease of these cells
  - Macular thinning/irregularity cannot be detected during clinical exam
- More reproducible measure (if not using retinal nerve fiber layer) than peripapillary RNFL
  - Fewer blood vessels an other cell components
  - Less anatomic variation compared to optic disc/peripapillary region
- Better superior/inferior symmetry and symmetry between eyes than peripapillary RNFL

Disadvantages of Macular Imaging

- Macular imaging is not helpful in glaucoma cases in which patients have concurrent macular disease
  - ERM
  - CME
  - DME
  - AMD
  - Macular hole

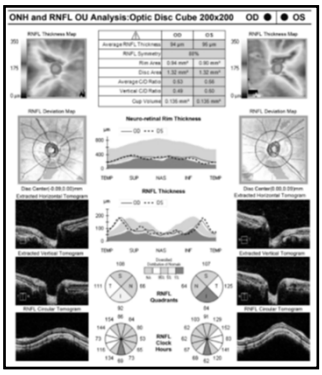
Glaucoma and Myopia
A Diagnostic Dilemma

- Myopia and Glaucoma:
  - Myopia epidemic: 5 Billion myopes by 2050
  - Myopia is a risk factor for glaucoma development
- Myopic nerves can be difficult to evaluate
  - Tilt, peripapillary changes, flattening of cup
- Challenges with OCT in myopic eyes:
  - Difficult to acquire image
  - Higher incidence of segmentation errors
  - RNFL database not typically adjusted for RE or AL
  - RNFL and macular thickness may be affected by increased AL

Glaucoma and Myopia – OCT Considerations

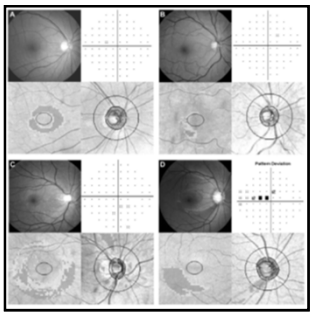
- RNFL:
  - Decreased RNFL thickness with increasing AL in S, I, N sectors
  - RNFL more temporally located (shifted peaks)
  - Increasing AL associated with false positives ("red disease")
- Macular Ganglion Cell:
  - Average thickness reduced in high myopia
  - Tend to have diffuse circular thinning with irregular inner margin
  - "GCIPL Hemifield Test" shown to have high sensitivity and specificity in high myopia (Kim YS, et al. IOVS 2016;57:5856-63)

Temporally displaced RNFL peaks



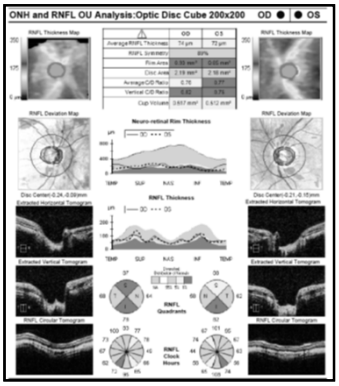
Tan, et al. Br J Ophthalmol 2019;103:1347-1355

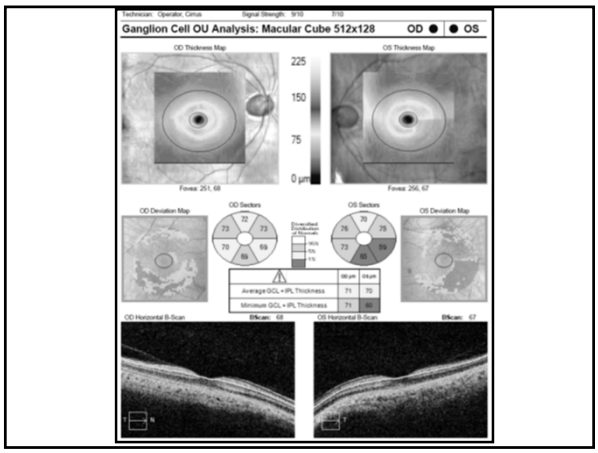
GCIPL False (+)



Kim, et al. Ophthalmology 2015;122(3):502-510

Back to Carlos



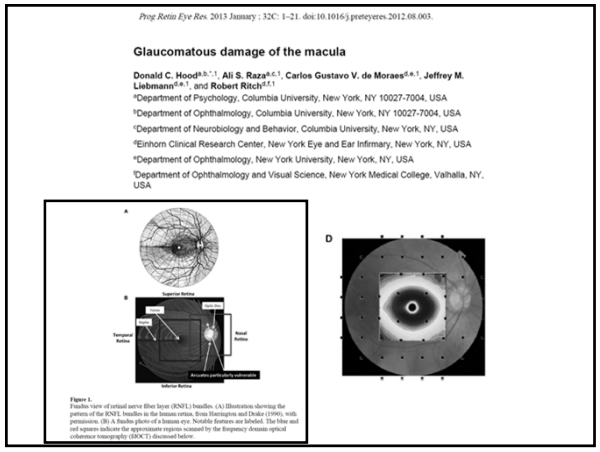


Is This Glaucoma?

A. Yes
B. No
C. I'm still not convinced

What about the 10-2 VF?

- Central 8 degrees from the center of the foveal contains more than 30% of retinal ganglion cells
- 24-2 and 30-2 test strategies use a 6 degree test grid pattern; these points fall outside of the densist region of ganglion cells
- 10-2 test strategy uses a 2 degree test grid
- Recent research has shown that in some patients with small regions of macular gangion cell loss, 10-2 testing may be better able to detect VF loss



Prog Retin Eye Res. 2013 January ; 32C: 1–21. doi:10.1016/j.preteyeres.2012.08.003.

Glaucomatous damage of the macula

Donald C. Hood[a,b,*,1], **Ali S. Raza**[a,c,1], **Carlos Gustavo V. de Moraes**[d,e,1], **Jeffrey M. Liebmann**[d,e,1], and **Robert Ritch**[d,f,1]

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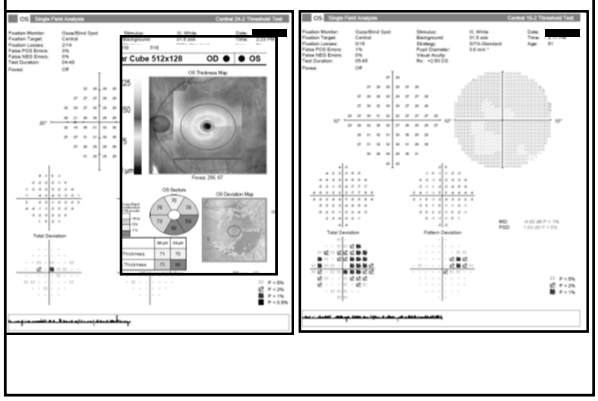
[e]Department of Ophthalmology, New York University, New York, NY, USA
[f]Department of Ophthalmology and Visual Science, New York Medical College, Valhalla, NY, USA



Is this glaucoma?

A. Yes
B. No
C. You'll never convince me

"I'd like to get my driver's license"

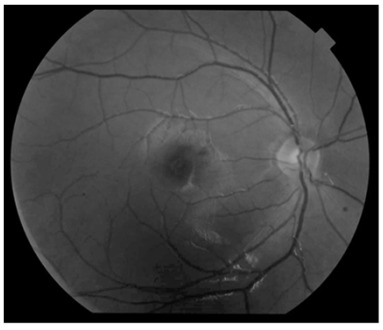
- Jacob,15 yo WM
- History:
  - Central vision loss noted OD>OS in 2014
  - Followed by retinal specialist who diagnosed "unspecified macular dystrophy" with differential of
    - Stargardts
    - Vitelliform
    - Cone Dystrophy
    - Rod-cone Dystrophy
    - **No genetic testing to date
  - Reports no changes in vision; retinal specialist has seen no/limited changes
  - Adaptations: large print/font; sits close to board in school; limited use of HHM and HHTS

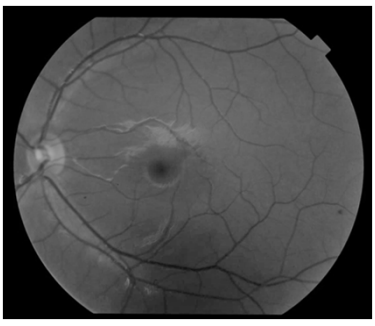
March 2019

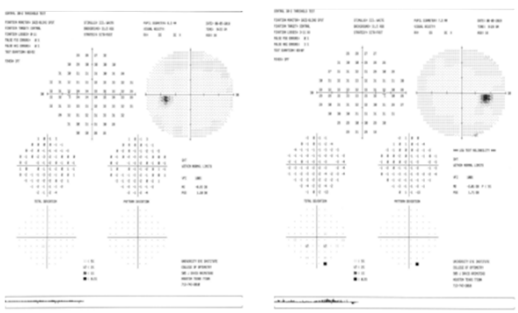
- Presented to Vision Rehabilitation Service with chief goal of driving (!)
- This visit prompted a referral for a multifocal ERG

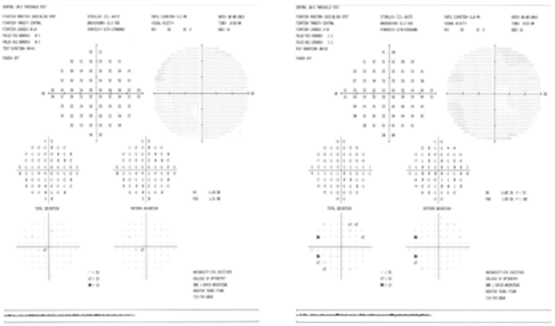
June 2019

- BCVA: 20/160 OD 20/60- OS
- Pupils, Motility, CVF: Normal OU
- SLE: Normal OU
- IOP: 20mmHg OU
- DFE: Normal disc, vessels, periphery OU
  - Macular pigment changes OD without FLR
  - Very mild pigment changes OS with FLR



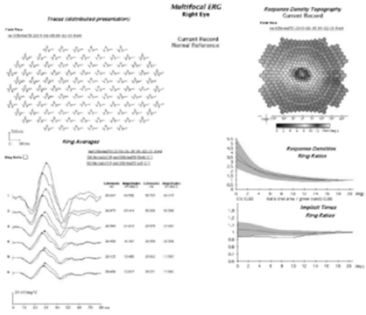


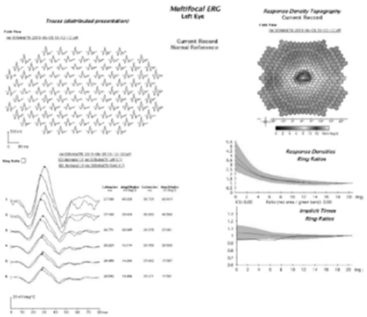


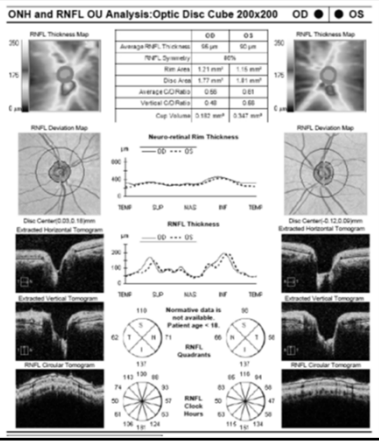


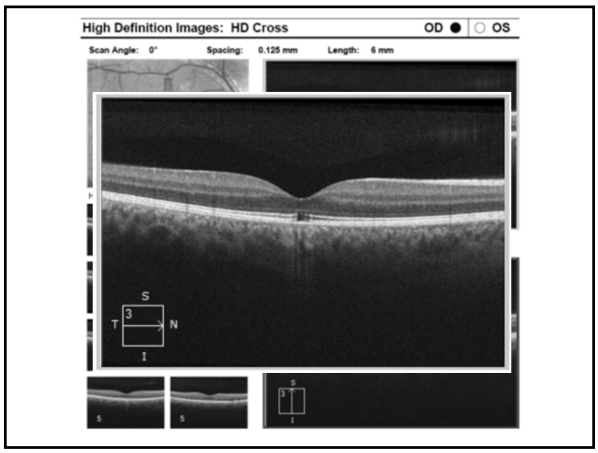
What is your diagnosis?

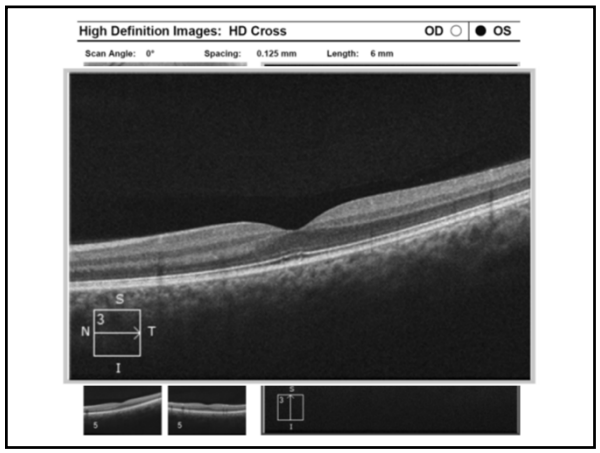
A. Stargardts
B. Cone dystrophy
C. Traumatic maculopathy
D. Other
E. I need more information











A few more questions…

A few more questions…

- Have you ever looked directly at the sun?
  - Answer: Nope
- Have you ever looked directly at, or played with, a laser pointer?
  - Answer: Ummm…

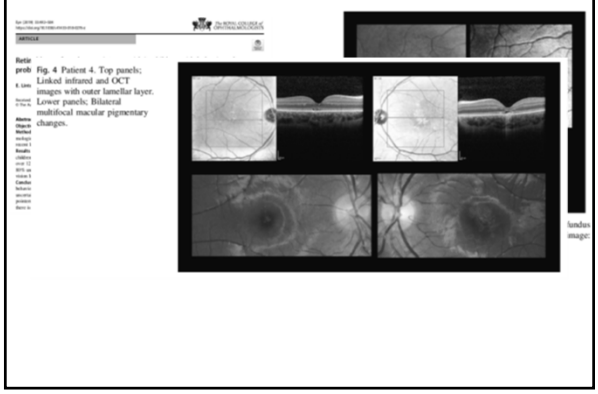
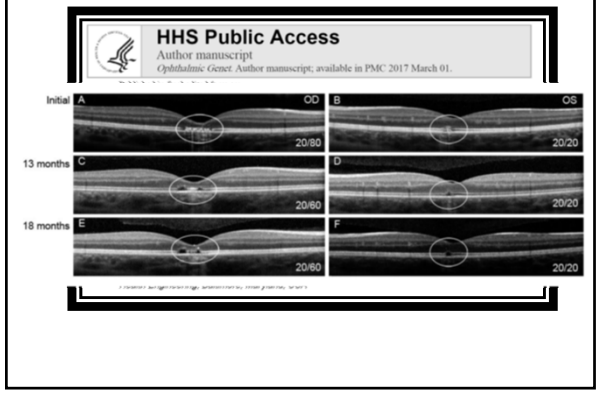


Fig. 4 Patient 4. Top panels; Linked infrared and OCT images with outer lamellar layer. Lower panels; Bilateral multifocal macular pigmentary changes.



Finally…

Sex: Male
MRN:
Sample accession date: 07/27/2019
Clinical te

Reason for testing
Diagnostic test for a personal history of disease

Test performed
Sequence analysis and deletion/duplication
listed in the results section below.
▪ Invitae Inherited Retinal Disorders Pan

RESULT: UNCERTAIN

Variant(s) of Uncertain Significance identified.

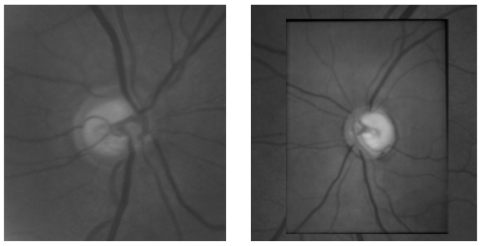
GENE	VARIANT	ZYGOSITY	VARIANT CLASSIFICATI
CEP250	c.6515A>G (p.Lys2172Arg)	heterozygous	Uncertain Significance
GNAT1	c.278A>G (p.Asp93Gly)	heterozygous	Uncertain Significance
NR2E3	c.1149G>A (Silent)	heterozygous	Uncertain Significance

What Did I Learn From This Case?

EN, 80 year old Hispanic female

- Presented to clinic with chief complaint of SEVERE loss of vision OD>OS, worsening over the past 2 weeks; moderate photophobia, moderate pain/headache (only upon asking)
- POH:
  - Severe NTG OS, mild OD
  - Surgeries:
    - cataract surgery with KDB goniotomy OU 2017
    - SLT x2 OS
    - Yag Capsulotomy OS

A Glimpse at the Glaucoma



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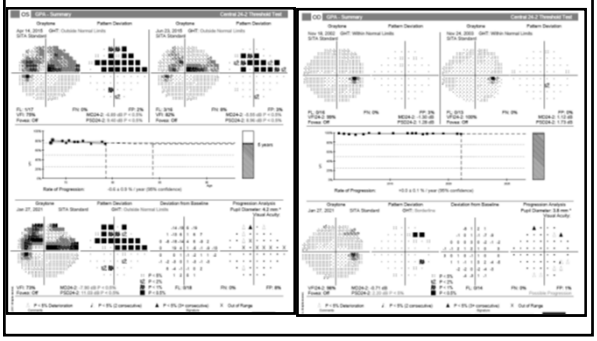
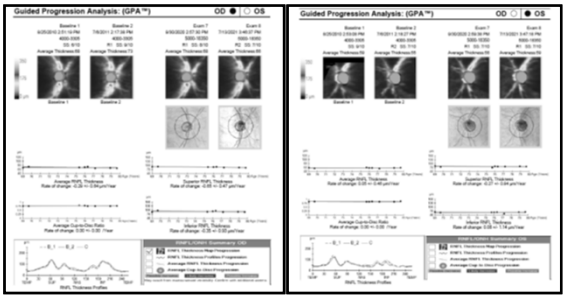


A Glimpse at the Glaucoma



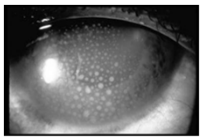
EN, 80 yo Hispanic female

- PMH:
  - Systemic hypertension
  - Benign kidney tumor removed 2019
  - Lung cancer dx 2020
  - Thyroid dysfunction x 2022
- Meds:
  - Sutent
  - Metoprolol
  - Losartan
  - Levothryroxine
  - Lumigan OU

EXAM FINDINGS

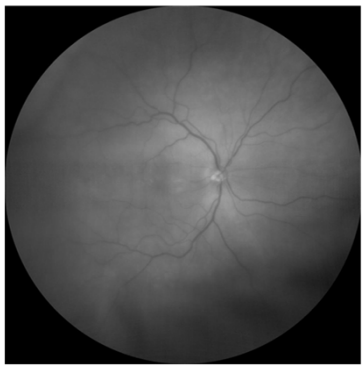
- BCVA: 20/200 OD 20/50- OS
- Pupils: 3mm OD, OS; sluggish OU; (+) RAPD OS (longstanding)
- Motility: Full OD, OS
- CVF: FTFC OD, OS
- BP: 127/68

Exam Findings:

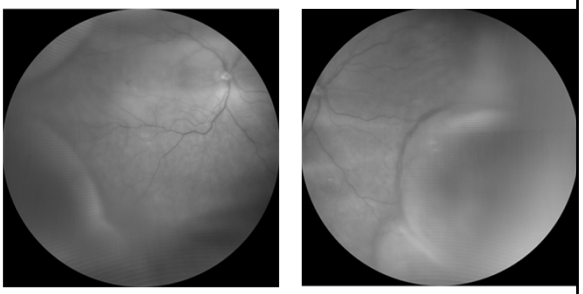


- SLE:
  - Lids/Lashes: MGD, thickened, red OU
  - Conj: 1+ diffuse injection
  - Cornea: trace striae OU, 3+ mutton fat KP OD, 2+ OS
  - AC: 3+ Cells/flare OD, OS
  - Iris: no nodules, no neovascularization, no posterior synechiae
  - PCIOL in place OU, capsule open OS
  - Vitreous: 2+ anterior cells
- IOP: 10mmHg OD 12mmHg OS

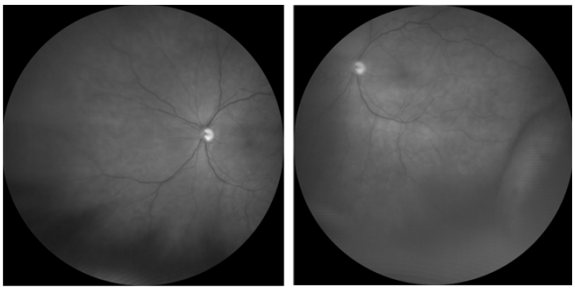
Fundus OD

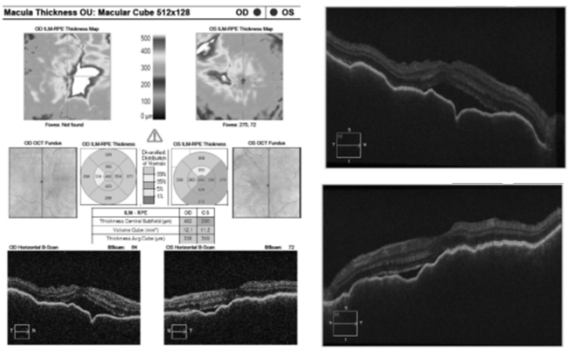


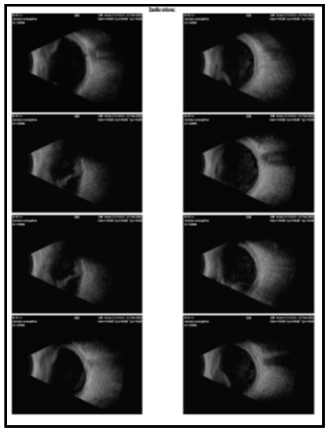
Fundus - OD



Fundus OS







What the heck is going on?

What the heck is going on?

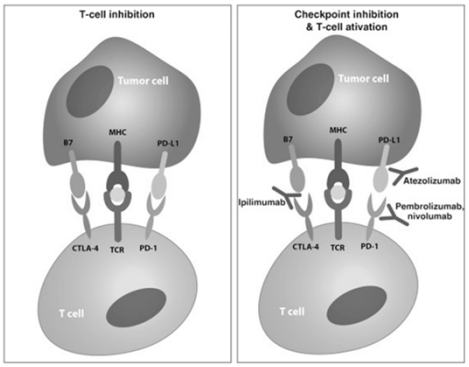
- ANYTHING NEW going on with you? New medications?
- "Oh, yes, I just started this new cancer medication."
  - pembrolizumab (Keytruda®)

Immune Checkpoints – Normal immune system

- Immune checkpoints are endogenous inhibitory receptor-mediated regulators of the immune system
  - CENTRAL to controlling the duration and extent of the normal immune response
  - Self-regulation allows for appropriate T-cell activity (attack foreign antigen) when needed, but then deactivation once the T-cell has served its purpose
  - Two targets (CTLA-4 and PD-1) are receptors directly on T-cell membrane and play a role in the downregulation of T-cell activity

Immune Checkpoint Inhibitors (ICIs, ICPIs)

- A growing class of cancer immunotherapy drugs (monoclonal antibodies) designed to BLOCK the deactivation of T-cells (and therefore allow the T cells to kill cancer cells)
- Three classes:
  - CTLA-4 inhibitors: ipilimumab (Yervoy)
  - PD-1 inhibitors: pembrolizumab (Keytruda) and nivolumab (Opdivo)
  - PD-L1 inhibitors: atezolizumab (Tecentriq), avelumab (Bavencio), and durvalumab (Imfinzi)
- Approved for a variety of cancers (melanoma, non-small cell lung cancer, Hodgkins lymphoma, some gastric renal cancers)



Immune-related Adverse Events (IRAEs)

- Immune-related Adverse Events occur in at least 1/3 of patients receiving ICPI therapy; most are mild, but can be very severe
  - Skin
  - Heart
  - Lung
  - Liver
  - Kidneys
  - CNS
  - GI
  - GU
  - Hormonal regulation
- Most common: immune-related skin rash (50%), itching (40%), diarrhea (45%)
- Side effects typically start within a few weeks to months after initiation

Ocular IRAEs

- Occur in approximately 1% of ICPI patients
  - Ipilimumab > pembrolizumab > nivolumab
- Most can be managed with topical, periocular, or systemic corticosteroid therapy
- May have to delay or discontinue ICPI therapy, though often can continue

Ocular IRAEs

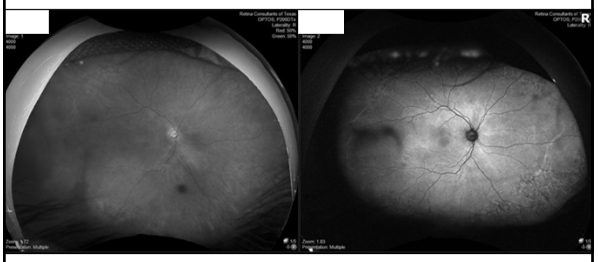
- Orbit/adnexa:
  - Ocular Myasthenia Gravis
  - Inflammatory orbitopathy
  - CN palsies
- Ocular surface
  - Dry eye
  - Inflammatory keratitis
  - Conjunctivitis (mild)
- Optic nerve
  - Optic neuritis

Ocular IRAEs

- Uveitis
  - Anterior (most common)
  - Posterior
  - Panuveitis
    - With or without granulomatous features
    - Specifically, Vogt-Koyanagi-Harada (VKH)-like panuveitis with choroiditis and serous retinal detachments (pembrolizumab***)

Back to Patient

- Sent to uveitis specialist for consultation
- Initially started on 60mg prednisone per day
  - In consultation with oncologist, continued on Keytruda therapy
  - Currently on 5mg per day
    - BCVA 20/50 OD 20/25 OS



BCVA: 20/25 OD 20/20 OS

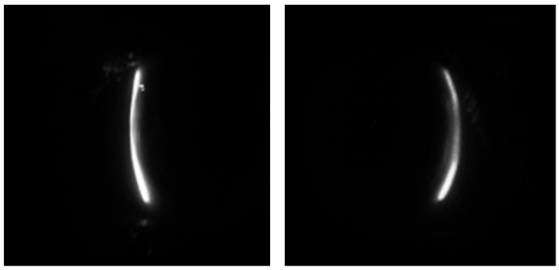
"My Eye Hurts & I Can't See"

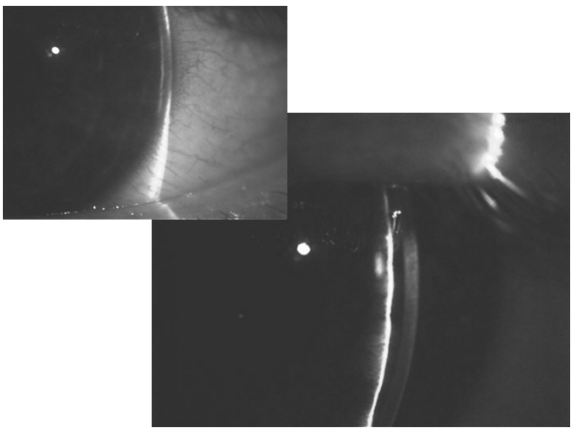
- CC: 28 YO WF presented with blurry vision OU, seeing rainbows around lights, severe frontal HA, and nausea for one day
- Ocular History: unremarkable, 5D Myope OU (DWSCL)
- Medical History: (+) HA, Tremors, Dizziness – currently under care of neurologist for evaluation/management
- Family Ocular/Medical History: unremarkable
- Medications: new med for neurologic symptoms (unknown name) x 8 days; acetaminophen prn for HA (no relief)
- Allergies: Codeine
- Social History: unremarkable

Clinical Exam

- VA w/glasses: 20/100 OD and OS, PH – 20/40 OD, OS
- Pupils: 4mm OU, sluggish reaction OU
- Motility normal OU
- SLE:
  - 1+ diffuse Corneal Edema OU
  - Closed angles OU (Van Herrick)
  - Shallow anterior chambers OU
- IOP: 34 OD, 35 OS @ 2:15 pm

Due to nausea & vomiting, unable to perform gonioscopy at initial visit



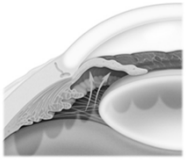


What's Your Diagnosis?

A. Posner-Schlossman Syndrome
B. Ocular Hypertension
C. Bilateral acute primary angle closure
D. Bilateral acute secondary angle closure

Classification of Angle Closure (Primary versus Secondary)

- **Primary Angle Closure**
  - With Pupillary Block
    - Acute/Subacute/Chronic
  - Without Pupillary Block (Iris Plateau)
- **Secondary Angle Closure**
  - With Pupillary Block
    - Lens-induced
    - Complete posterior synechiae
  - Without Pupillary Block
    - Anterior Pulling (NVG, ICE syndrome)
    - Posterior Pushing (Drug-induced/Choroidal Expansion, malignant glaucoma/aqueous misdirection) ****



Angle Closure (Anatomical Consideration)

- **Anterior to Lens**
  - Pupil block (major contributor)
  - Non-pupillary block (ciliary body)
    - Plateau iris configuration
    - Plateau iris syndrome
    - Pseudo-plateau iris
- **Lens-induced**
  - Phacomorphic
  - Subluxation of lens
- **Retro-lenticular forces**
  - Malignant glaucoma
  - Choroidal effusion/ciliary body rotation

Topiramate-induced Angle Closure

- May cause myopic shift and acute angle closure – occurs in 3/100,000
- Usually occurs within the first two weeks – one case was after only two doses at 25mg/day
- Pathophysiology:
  - Unknown what triggers reaction:
    - Possible blood-eye barrier disruption?
    - Hypersensitivity reaction?
    - Change in membrane potential?

Topiramate-induced Angle Closure

Choroidal effusion

2) Anterior displacement of Iris/CB/Lens diaphragm

3) Zonules relax

4) Lens thickens

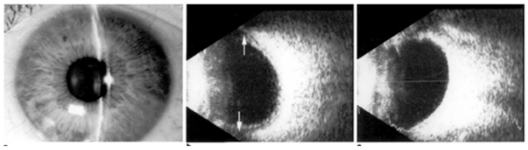
5) Induced Myopia

6) Acute angle closure

IOP: usually below 40, can be in 60s

- Some degree of CB shutdown
- Carbonic Anhydrase inhibition

Topiramate-Induced Angle Closure



TOPIRAMATE (TOPAMAX®, TROKENDI XR®)

- FDA approved for:
  - Various Epileptic Disorders
  - Migraines
  - Pain
  - Weight loss
    - phentermine with topiramate (Qsymia®)
- Sulfa-based with carbonic anhydrase inhibition

TOPAMAX® (topiramate)

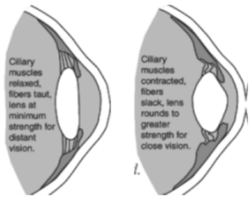
POLL: What Is Your Treatment Plan?

A. Urgent referral to ophthalmology
B. Topical aqueous suppressants, pilocarpine, oral acetazolamide
C. Topical aqueous suppressants, cycloplegic agent, steroid
D. Laser peripheral iridotomy (LPI)

Treatment – DIFFERENT THAN PRIMARY ANGLE CLOSURE!!!

- Discontinuation of Topamax
- Strong, short course of cycloplegic:
  - 1 or 2 doses generally sufficient

  1) Relaxes ciliary muscles
  2) Iris/Lens/CB diaphragm displace posteriorly
  3) Zonules tighten
  4) Angle opens/Myopia reduced
- Pilocarpine contraindicated:
  - Causes ciliary contraction, exacerbating forward movement of lens-iris diaphragm
  - Slightly pro-inflammatory

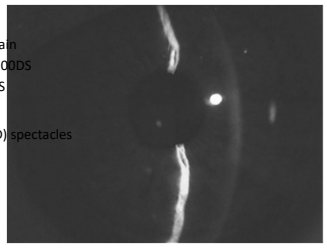


Treatment continued

- IOP – lowering agents:
  - Beta-blockers and Alpha-agonists typically first choice
  - Prostaglandins effective but not first choice due to delayed onset of effect
  - Topical CAIs also effective but not commonly used since they are sulfa-based, and thus chemically related to Topiramate (although no incidences of angle closure have been reported with *topical* CAIs)
- Steroids:
  - Tighten capillary junctions as well as decrease CB swelling
- Surgical:
  - LPI is not effective because mechanism is not pupillary block
  - Drainage of suprachoroidal fluid – very rarely done (usually medical therapy is sufficient)
  - Trabeculectomy/Filtering surgery - only if PAS formed after resolution (rare)

Back to Our Patient…

- Immediate Therapy (In Office)
  - Two doses of scopolamine OU, topical steroid, and Combigan®
  - IOP reduced to 20mmHg OD and 26mmHg OS
  - Discharged with Combigan® and steroid
  - D/C Topamax®
- Follow-up (24 h):
  - VA still blurry (no haloes), no pain
  - VA: 20/25 OD, OS through -10.00DS
  - IOP: 10mmHg OD , 12mmHg OS
- Follow-up (Day 4):
  - VA 20/20 through habitual (-5D) spectacles
  - IOP 10mmHg OU
  - D/C all topical meds



Clinical Pearls

- Angle closure is not always pupillary block mechanism
- Bilateral angle closure is *nearly* ALWAYS secondary angle closure – think medications!
- Clinical management of choroidal effusion/ciliary detachment angle closure is different than that of pupillary block (no pilo, no LPI, no acetazolamide) – CYCLOPLEGIA is key.

"I Almost Hit a Car!"

- Pedro, 31yo HM
- CC: Pt feels that vision is blurry or "incomplete" to the right
- HPI:
  - First noticed a few weeks ago when he failed to see a car merging into his lane from the right
  - No other sensory changes; no weakness/paresthesia; no other neurologic symptoms
  - (+) HA – says he has "migraines" which are worse in past few months
  - Fall while ice skating approximately 3 months ago; fell onto his back, broke his arm, hit the back of his head. No LOC

Pedro, 31yo HM

- POH: Unremarkable
- PMH: recent elevated cholesterol
- Social: Unremarkable; college student, recently married; no health insurance

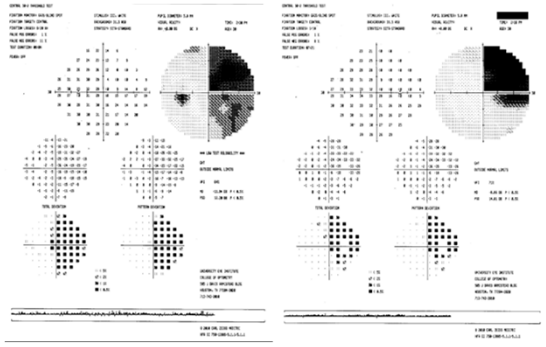
Pedro, 31yo HM

- Examination:
  - BCVA: 20/20 OD, OS
  - Pupils: 5mm OU; brisk D/C OU; (-) RAPD
  - Motility: Full OU
  - CVF (finger counting):
    - OD: Right superior restriction
    - OS: Right superior and slightly inferior restriction
  - Color vision: Normal OD, OS

Pedro, 31yo HM

- Slit lamp: Normal OU
- IOP: 14mmHg OD 15mmHg OS
- DFE: Normal DMVP OU

- See visual fields:



Pedro, 31yo HM

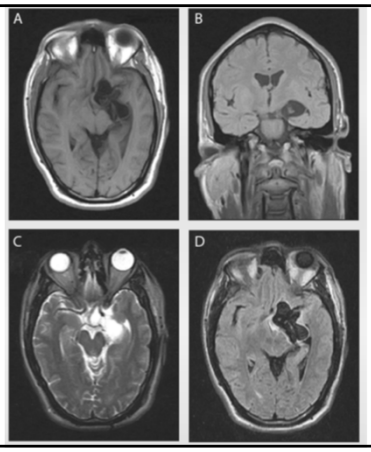
- Thoughts?
  - Could the fall have anything to do with this?
  - What do we do next?

  - PROBLEM: Uninsured

Pedro, 31yo HM

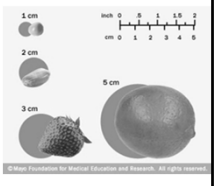
- Sent through community health clinic for MRI of brain with and without contrast.

- See MRI



MRI Report

- Sizable multi-lobulated partially cystic mass lesion in medial/superior temporal lobe (5cm x 4.2 cm x 2.5 cm)
  - Considerable mass effect to surrounding brain, with deviation of the left part of the optic chiasm and shifting of the midline to the right
  - Considerable mass effect to left lateral ventricle
  - Most consistent with glioblastoma multiforme (GBM)



Glioblastoma Multiforme

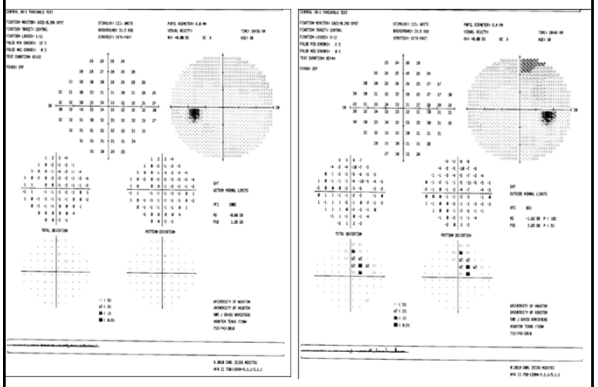
- GBM is the most common and most malignant of glial tumors.
- Hx is usually short with progressive neurologic deficit, often motor weakness, and headache
- Symptoms may also include general symptoms of increased ICP, including nausea/vomiting, cognitive impairment, seizures

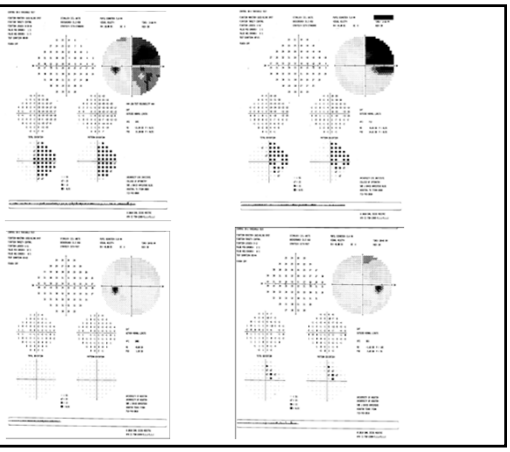
Glioblastoma Multiforme

- MRI with/without contrast is study of choice for diagnosis
- Tumors do not have clearly defined margins, tend to appear multicentric
- NO CURATIVE TREATMENT
  - Maximum surgical resection followed by radiotherapy and concomitant chemotherapy
  - Primary surgical goal is to relieve mass effect
- Prognosis: Without therapy, patients with GBM uniformly die within 3 months. With therapy, the mean survival is 12-18 months

Pedro, 31yo HM

- But wait…
- In interim (waiting for insurance to go into effect)…
  - Reports back to clinic ONE WEEK BEFORE SURGICAL RESECTION IS SCHEDULED, stating that he feels that his vision has gone "back to normal"





Pedro, 31yo HM

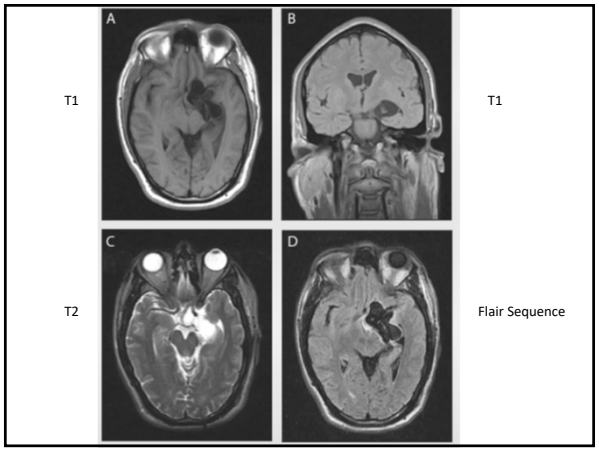
- IMMEDIATE repeat imaging shows NO CHANGE in lesion appearance; however, a second opinion radiology report indicates the lesion may be an INTRACRANIAL EPIDERMOID CYST.

Intracranial Epidermoid Cyst (IEC)

- Rare, slow-growing BENIGN congenital tumor.
- Predominantly located in the cisternal spaces
- Because they are so slow growing, symptoms take many years to manifest and are typically due to mass effect or from encasing neurovascular structures (CN)
- Predominant symptom is headache, but otherwise dependent on location and structures encased.

Intracranial Epidermoid Cyst

- Diagnosis: MRI is considered the best imaging modality
  - Iso- to hypo-intense to CSF in T1-weighted images
  - Hyper-intense in T2-weighted images
  - Little to no enhancement with contrast
  - Difficult to definitively diagnose based solely on imaging
  - Differential: arachnoid cyst, dermoid cyst, neurocysticercosis, astrocytoma, glioma



Intracranial Epidermoid Cyst

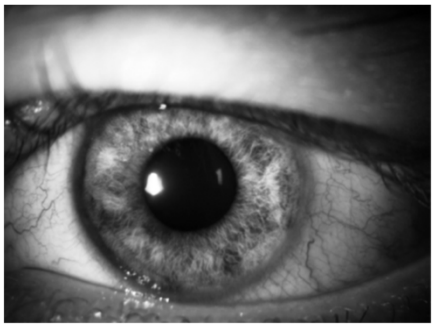
- Very rare malignant potential
- Complete surgical excision is advised
  - Relieve mass effect
  - Reduce risk of chemical meningitis that can occur when cyst ruptures
- Final Disposition

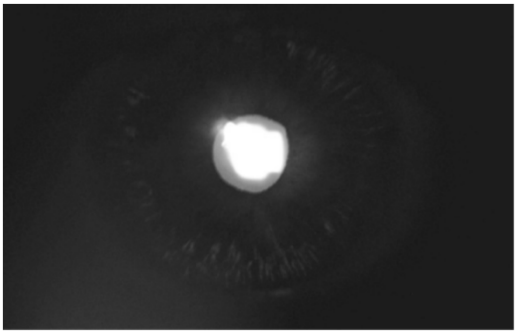
What's the Deal With YOUNG glaucoma patients?

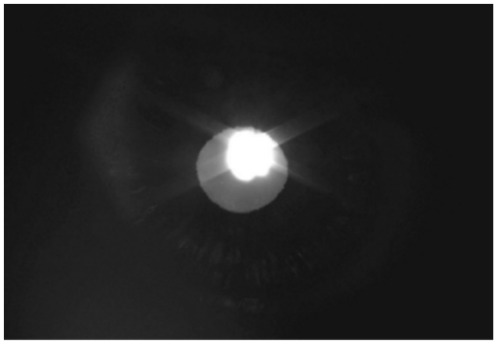
- KC, 25 year old white male
- CC: Wants some new soft CLs, time for exam
- HPI: High myopia (-7.50 OU), wears daily wear SCL, monthly disposable lenses, no problems
- POH: Unremarkable, (-) surg/trauma
- PMH: "Very healthy", (-) chronic illnesses, surgeries, hospitalizations
- FH: Unremarkable
- Meds: None
- Allergies: NKDA
- Social: Occasional alcohol, no smoking

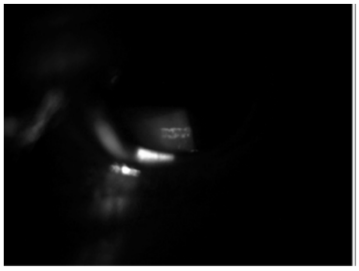
Exam Data

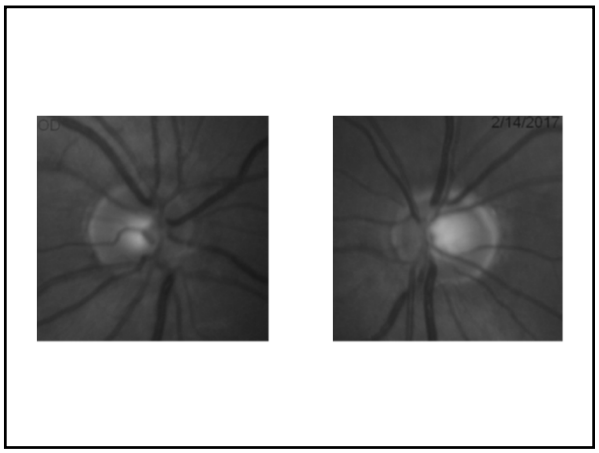
- BCVA: 20/20 OD, OS
- Motility: Full OU
- Pupils: 5mm OD/OS, 3+ D/C, (-) RAPD
- SLE: See photos
- IOP: 21mmHg OD 38mmHg OS
- Gonioscopy: See photos
- DFE/VF







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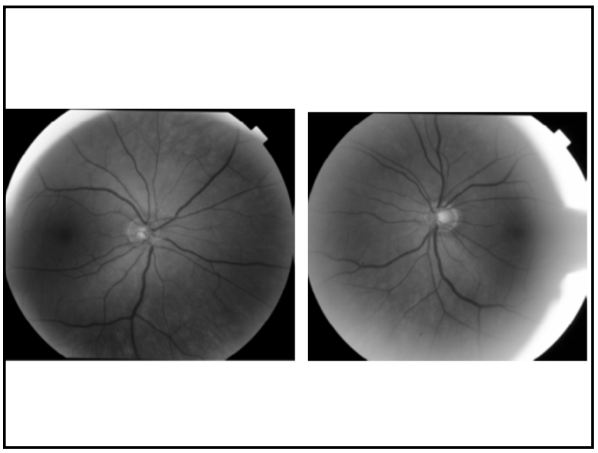
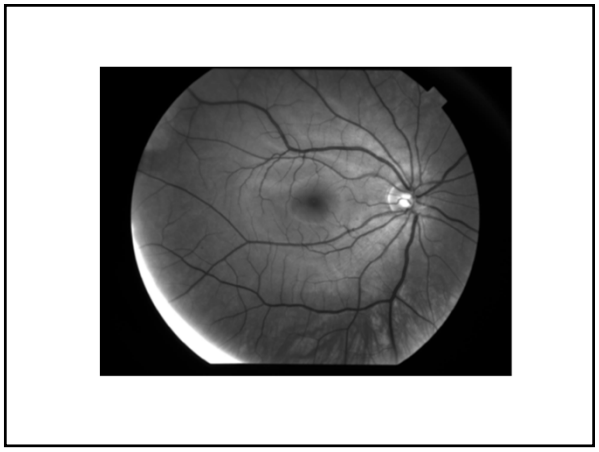
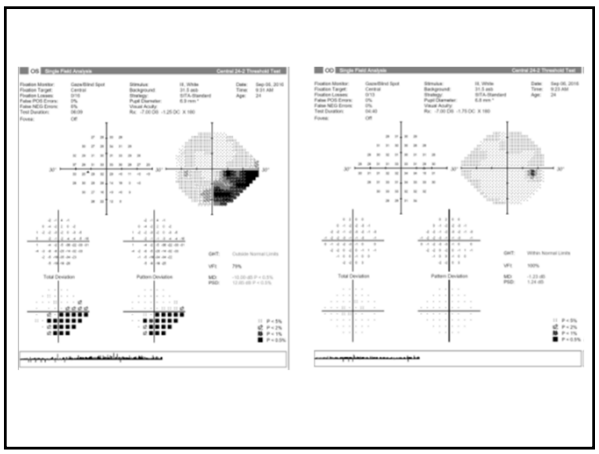
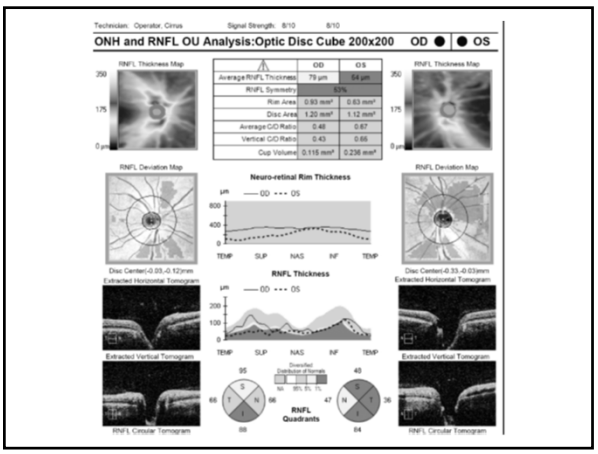
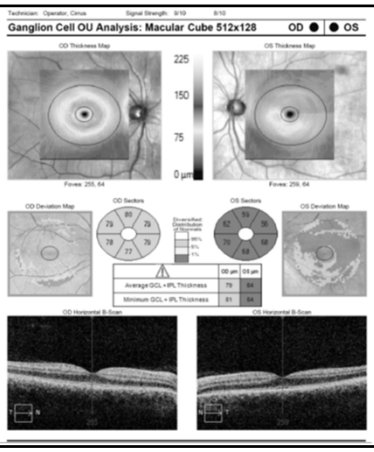







Technician: Operator, Cirrus
Signal Strength: 8/10 8/10
ONH and RNFL OU Analysis:Optic Disc Cube 200x200 OD ● ● OS
RNFL Thickness Map
RNFL Deviation Map
Neuro-retinal Rim Thickness
RNFL Thickness
Extracted Horizontal Tomogram
Extracted Vertical Tomogram
RNFL Circular Tomogram
RNFL Quadrants



Pigmentary Glaucoma

- Pigment Dispersion Syndrome
  - Classic triad:
    - Krukenberg spindle
    - Midperipheral iris transillumination defects
    - Heavy uniform pigmentation of TM
      - Often with posteriorly bowed iris on gonioscopy
  - Other signs:
    - Deposition of pigment on anterior iris
    - Pigment on zonules
    - Pigment on posterior lens surface

Pigmentary Glaucoma

- If increased resistance to aqueous outflow occurs and IOP is elevated, pigmentary glaucoma can occur
- Risk somewhere between 10% and 50%
- Bilateral
- Young, myopic males
- May have h/o episodic eye pain/blurred vision

Pigmentary Glaucoma

- Management:
  - Secondary open angle glaucomas: Direct treatment to the CAUSE, if possible
    - Laser peripheral iridotomy (controversial)
    - Laser iridoplasty (controversial)
    - Pilocarpine (not well tolerated)
  - Medical Management
    - Prostaglandin analog
    - Beta blocker
    - Brimonidine
    - CAI
    - netarsudil
  - Laser trabeculoplasty
  - Target IOP???

What is Your Initial Treatment Plan?

- PGA
  - NO-releasing PGA
- Beta Blocker
- Brimonidine
- CAI
- Netarsudil
- Fixed dose combination
- Laser trabeculoplasty

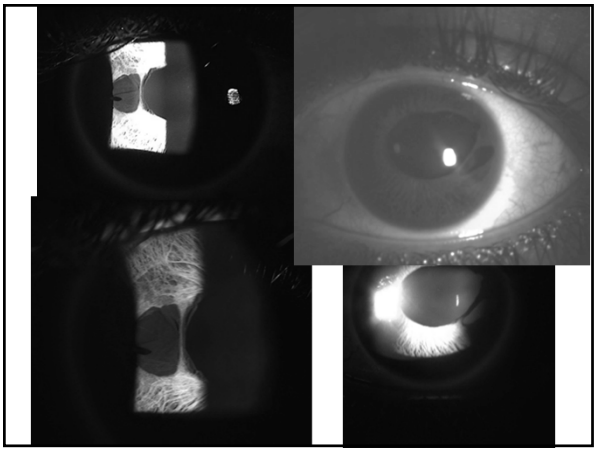
Final Disposition

Well, I Got Shot With a BB Gun

- 18 year old Jordanian male presented for routine exam, c/o blurred vision with glasses
  - POH: Left eye has multiple pupils because he got shot with a BB gun by his cousin at a young age
  - PMH: Unremarkable
  - FOH/FMH: Non-contributory

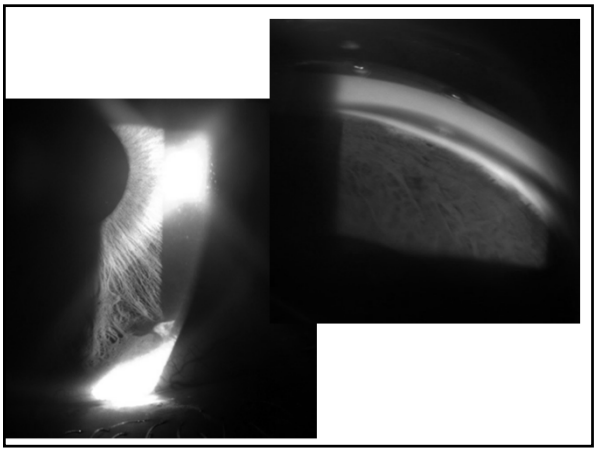
Well, I Got Shot With a BB Gun

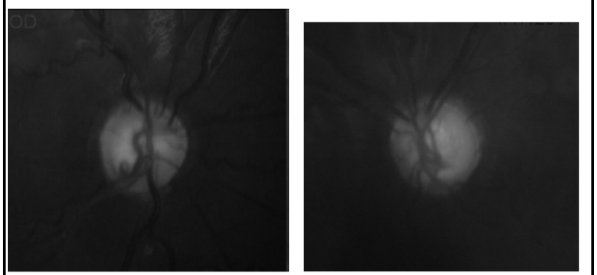
- BCVA: 20/20 OD 20/25 OS
- Pupils: 6mm round OD; correctopia/polycoria OS; (-) RAPD
- EOMs: Unrestricted
- CVF: FTFC OD Significant inferior restriction OS
- Slit lamp:

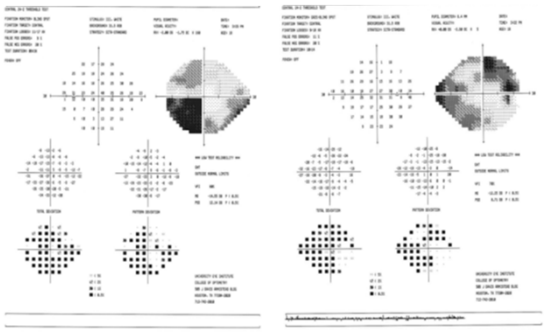


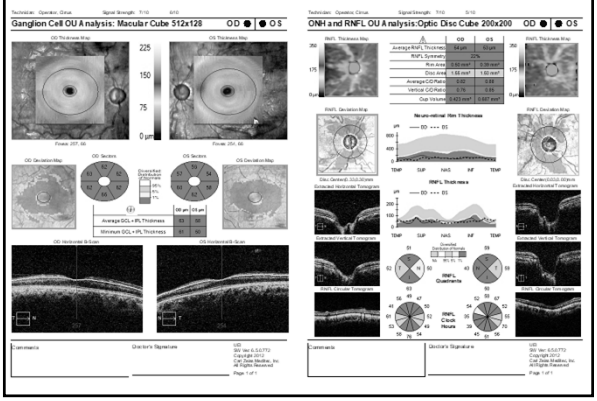
Well, I Got Shot With a BB Gun

- IOP: 37mmHg OD 39mmHg OS
- DFE:
  - C/D 0.9v OD 0.95 v OS
  - M, V, P normal OU
- See gonio, disc and VF

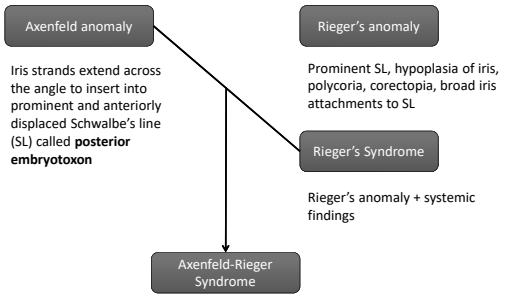








Anterior Segment Dysgenesis (ASD)

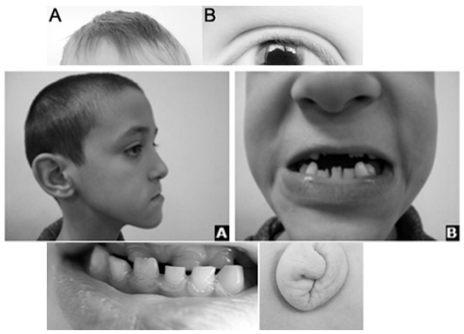


Axenfeld-Rieger Syndrome

- Autosomal dominant genetic condition
- Clinical characteristics:
  - Posterior embryotoxon
  - Iris strands/PAS
  - Iris hypoplasia
  - Correctopia
  - Polycoria
  - Ectropion uveae

Axenfeld-Rieger Syndrome

- Systemic Characteristics:
  - Mild craniofacial abnormalities
    - Hypertelorism/telecanthus
    - Maxillary hypoplasia
    - Broad, flat nasal bridge
  - Dental abnormalities
    - Microdontia
    - Oligodontia, hypodontia



Differential Diagnosis

- ICE (unilateral, female, corneal changes, lack of systemic anomalies)
- Peter's Anomaly (central corneal opacity)
- Aniridia
- Congenital Ectropion Uveae

Glaucoma in ARS

- Approximately 50% of ARS patients will develop glaucoma
- May occur in early infancy, but more common in adolescence or early adulthood
- Medical management
- Surgical management

Thank You For Your Attention!

Questions?

Email me: dmarrelli@uh.edu