The Secondary Glaucomas

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Financial Disclosure:

- Allergan
- Bausch & Lomb
- Carl Zeiss Meditec
- M&S Technologies
- Santen

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Differences with Secondary Glaucomas

- Diagnosis:
  - Laterality
  - Presentation (acute vs insidious)
  - Other clinical findings
- Management – treat the CAUSE, if possible
  - Medications: contraindications, effectiveness
  - Lasers: differences in response
- IMPORTANCE OF GONIOSCOPY
  - www.gonioscopy.org

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Secondary Glaucomas

Secondary <u>OPEN</u> Angle

- Exfoliation
- Pigmentary
- Lens-induced
  - Phacolytic
  - Lens Particle
  - Phakoanaphylaxis
- Inflammatory
  - Uveitic
  - Glaucomatocyclitic Crisis
  - Fuch's Heterochromic Iridocyclitis
- Steroid-induced
- Elevated EVP
- Traumatic
  - Hyphema
  - Angle Recession

Secondary <u>ANGLE CLOSURE</u>

- With Pupillary Block
  - Lens-induced
    - Phacomorphic
    - Ectopia Lentis
- Without Pupillary Block
  - Anterior Pulling
    - Neovascular
    - Iridocorneal Endothelial Syndrome
    - Inflammatory
  - Posterior Pushing
    - Aqueous Misdirection
    - Drug-Induced

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SECONDARY <u>OPEN</u> ANGLE GLAUCOMAS

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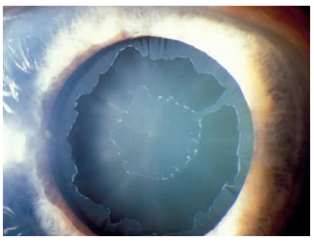
Exfoliation Syndrome (Pseudoexfoliation)

- Characterized by deposition of amyloid-like fibrillary material throughout anterior segment
  - Anterior lens capsule
  - Pupil margin
  - Lens zonules
  - Corneal endothelium
  - Inferior angle (gonioscopy)
- Iris transillumination at pupil margin
- Heavy dark pigment in TM, +/- Sampaolesi's line
- Weakened zonules -> phacodonesis, iridodonesis



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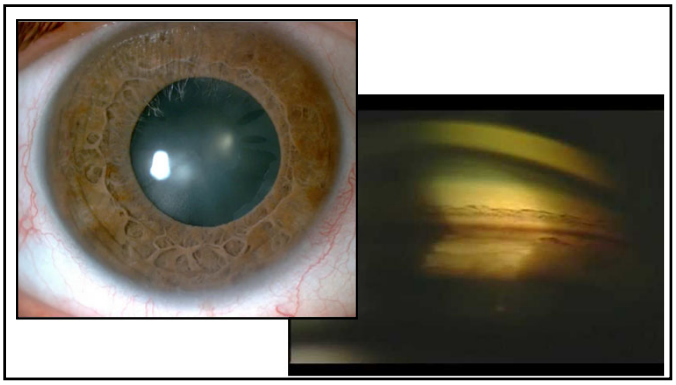

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Exfoliation Syndrome

- Strongly age-related; rare under 50 yo
- May be MONOCULAR or binocular with asymmetry in presentation and timing
- GLAUCOMA
  - Thought to be due to TM obstruction and subsequent damage/dysfunction
  - Odds ~ 40% over a 10-year time frame
  - Scandinavian countries: Exfoliation accounts for >50% of open angle glaucoma
- Differences between exfoliation glaucoma and POAG:
  - Often presents monocularly
  - Greater IOP fluctuations
  - Overall WORSE prognosis

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Exfoliation Syndrome

- Management:
  - Similar to POAG (meds)
  - SLT can be very effective
    - Power may be lessened, or treat only part of angle
    - Effect may not last as long as with POAG
- CATARACT SURGERY:
  - Lens extraction does NOT alleviate condition
  - Weakened zonules – higher risk of lens dislocation during/after surgery
  - Increased inflammation after all intraocular surgeries compared to POAG

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Mr. Garcia, 86 yo HM

- Followed as glaucoma suspect from 2008-2012 (pseudoexfoliation)
- Untreated IOP 14-18mmHg OU
- 2012: IOP began to be elevated, Tmax 25mmHg
  - Treatment initiated with Latanoprost 2012

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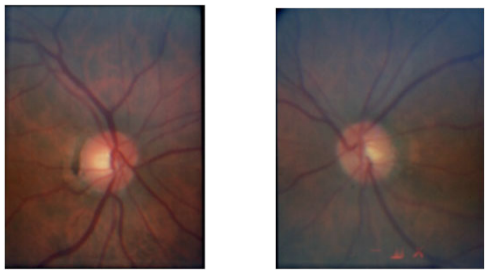

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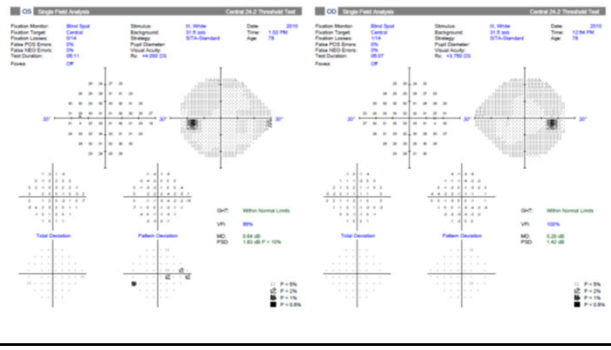

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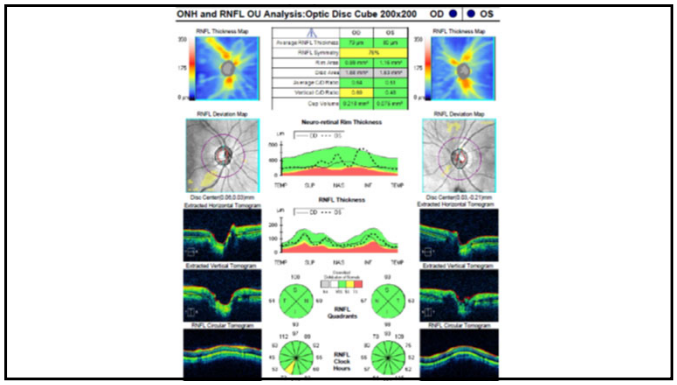

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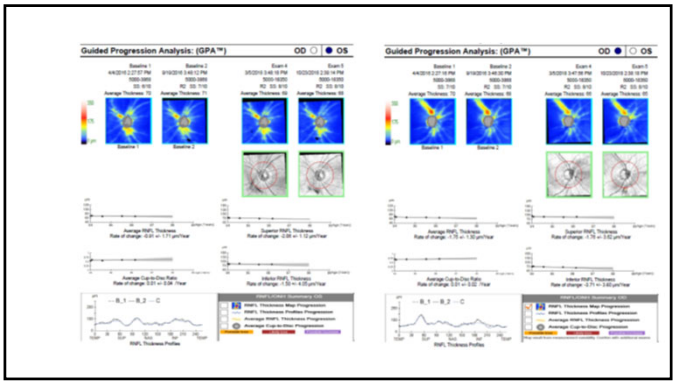

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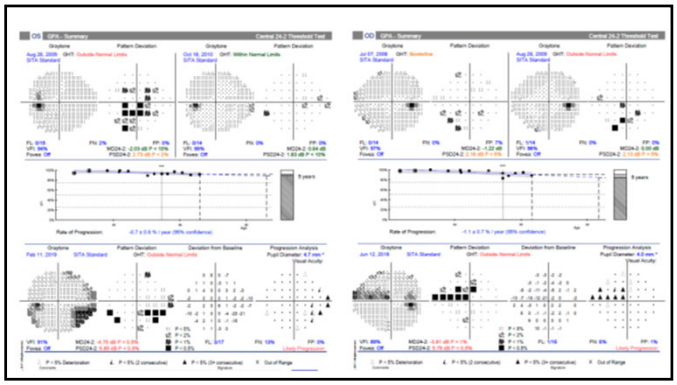

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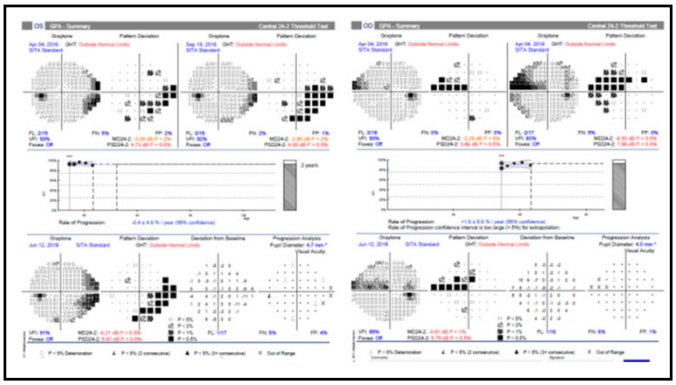

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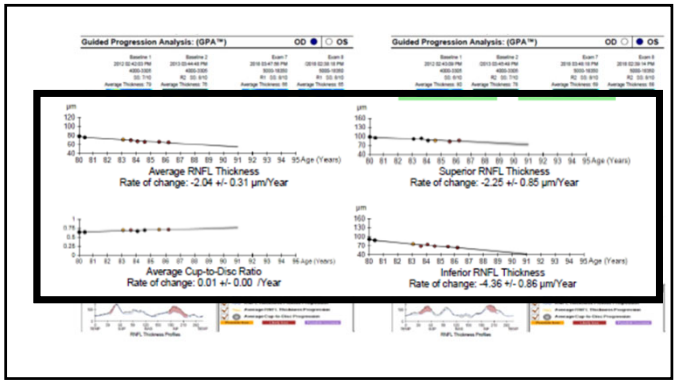

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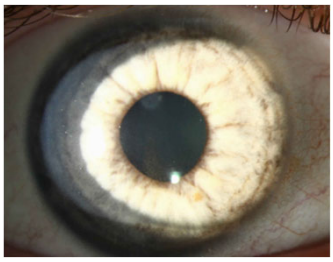
Pigmentary Glaucoma

- Pigment Dispersion Syndrome (PDS)
  - Pigment on corneal endothelium (Krukenberg Spindle)
  - Iris transillumination defects in spoke-like pattern (mid-periphery)
  - Homogenous heavy pigmentation in TM / speckled pigment ant to SL
  - Additional pigment: zonules, ant/post lens surface
  - Midperipheral iris is often concave (bowing toward zonules)



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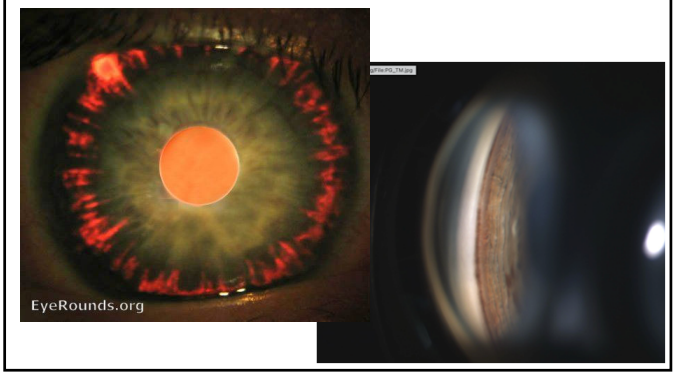

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Pigmentary Glaucoma

- PDS does not universally lead to glaucoma
  - 25-50% risk
  - Glaucoma develops most commonly in myopic males between 20-50 yo
- Characterized by WIDE fluctuations in IOP, often after pigment release
  - Exercise, pupillary dilation
  - May complain of blur or pain during episodes

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Pigmentary Glaucoma

- TREATMENT:
  - Standard medical therapy
  - Peripheral laser iridotomy or iridoplasty – proposed to alter iris configuration and decrease peripheral iris rubbing on zonules (not well established as a treatment)
  - SLT: higher risk of IOP spike than POAG
- With age, TM pigment may fade away as mechanism of pigment dispersion decreases ("burnout")

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Case: Kevin, 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

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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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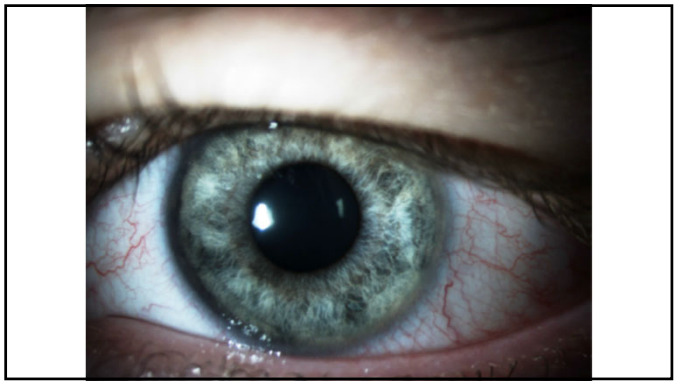

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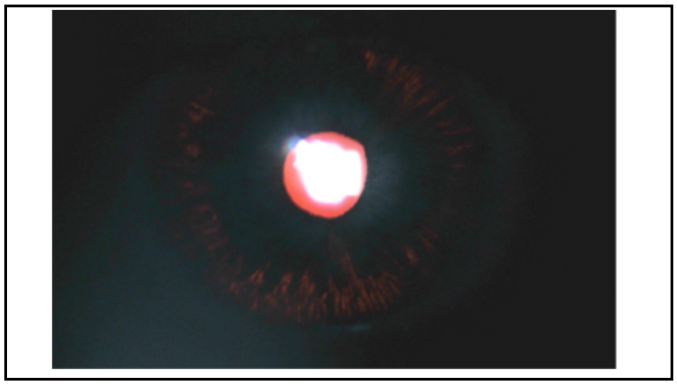

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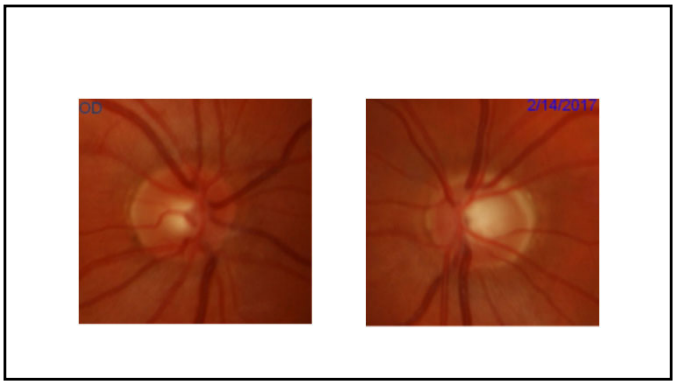

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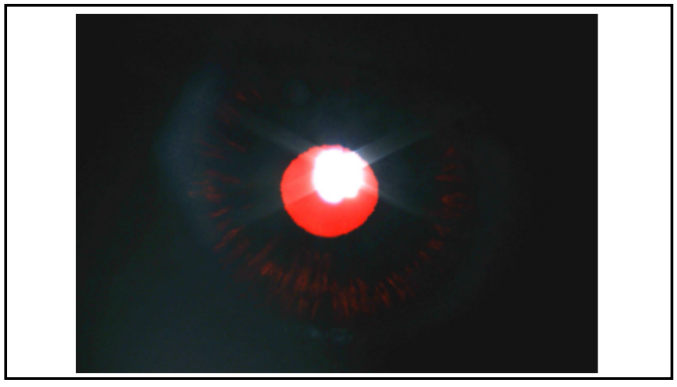

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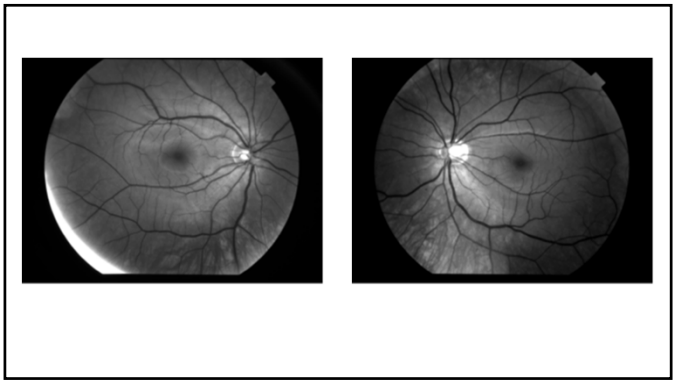

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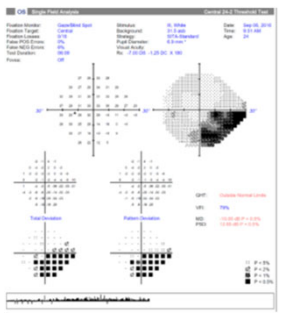

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How Do We Treat?

- PGA?
- Beta Blocker?
- Brimonidine?
- CAI?
- Laser?

- Final disposition

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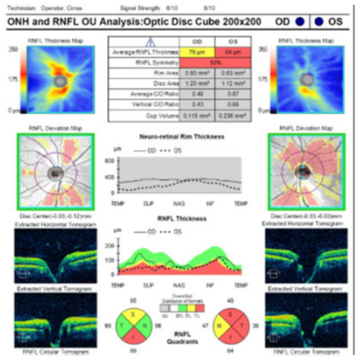

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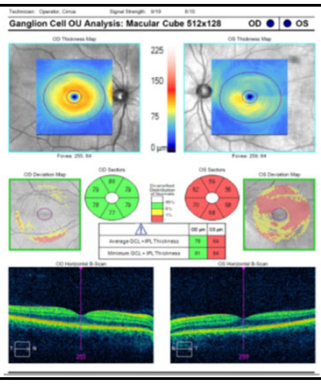

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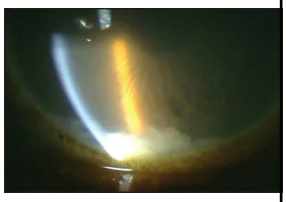
Lens-induced <u>OPEN ANGLE</u> Glaucoma

- <u>Phacolytic</u>: Inflammatory open angle glaucoma caused by leaking proteins through capsule of mature cataract
  - Sudden onset pain, conjunctival hyperemia, vision loss
  - Markedly elevated IOP
  - Corneal edema
  - Anterior chamber reaction <u>WITHOUT KP</u> (*** helps to differentiate from phacoanaphylaxis); may layer into hypopyon
  - Wrinkled capsule of hypermature cataract
  - TREATMENT: Acute management to lower IOP, then surgery



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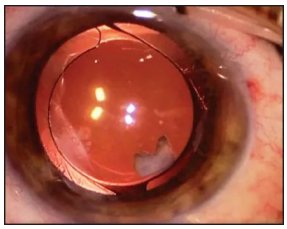
Lens-induced OPEN ANGLE Glaucoma



- Lens Particle:
  - Cortical material left in AC after surgery
  - Present with visible cortical material in AC, AC reaction, posterior synechiae
  - TREATMENT: mydriatic, corticosteroid, aqueous suppressants, occ. surgery
- Phacoanaphylaxis: RARE
  - Patient becomes sensitized to their own lens protein following penetrating injury or surgery
  - *GRANULOMATOUS* inflammation with KP, vitritis, synechiae
  - Glaucoma is uncommon
  - Treat with corticosteroid and aqueous suppressants

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Inflammatory OPEN ANGLE Glaucoma

- Inflammation can cause secondary glaucoma that has both open- and closed-angle components
- Uveitis: Elevated IOP can occur due to a variety of mechanisms
  - Trabeculitis
  - TM endothelial dysfunction
  - TM blocked by fibrin and cellular debris
  - Steroid-induced resistance to outflow

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Inflammatory Open-Angle Glaucoma: The DILEMMA

- Often difficult to establish cause of elevated IOP
- Inadequately controlled inflammation with elevated IOP is often mistaken for steroid-induced glaucoma
  - Presence of AC reaction: presume that inflammation is cause (increase steroid)
  - Timing may help (steroid-induced IOP elevation takes time)
- TREATMENT:
  - Control inflammation
  - Aqueous suppressants
  - NO PILOCARPINE, no SLT
  - ? Use of prostaglandin analogs

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The Use of Prostaglandin Analogs in the Uveitic Patient

Michael B. Horsley and Teresa C. Chen

Glaucoma Service, Massachusetts Eye and Ear Infirmary, Harvard Medical School, Boston, MA, USA

Seminars in Ophthalmology, 26(4-5), 285–289, 2011

SUMMARY

The use of prostaglandin analogs in uveitic patients remains controversial. A causal relationship has yet to be established between prostaglandins and the reactivation of anterior uveitis, the development of cystoid macular edema, or the reactivation of HSK.

Due to the efficacy of prostaglandins in lowering IOP in patients with uveitis and the small likelihood of developing these rare complications, prostaglandin analogs should remain in the treatment algorithm of uveitic glaucoma patients.

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Inflammatory: Glaucomatocyclitic Crisis

- AKA Posner-Schlossman Syndrome
- Uncommon, episodic, unilateral open angle inflammation with markedly elevated IOP and low grade AC reaction
- Pt presents with mild symptoms
- Exam reveals mild AC reaction, few small discrete KP
- IOP 40-50mmHg, may have corneal edema
- In between attacks, inflammation resolves and IOP returns to normal
- Differential: intermittent angle closure (GONIOSCOPY!!!)

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Glaucomatocyclitic Crisis

- TREAMENT:
  - No evidence that chronic suppressive treatment helps
  - Treat each episode with corticosteroid and aqueous suppressants
  - Consider oral antiviral (HZV, HSV have been implicated)
    - ?anterior chamber tap with PCR
  - Secondary glaucoma may develop with repeated attacks
    - BASELINE VF, OCT are helpful

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Jackie, 54 year old Asian Female

- H/O initial presentation of glaucomatocyclitic crisis in 1994 OS
  - Blur and mild discomfort OS x 2 days
  - IOP 47mmHg OS, trace cell and single small KP
  - Treated with corticosteroid and timolol, complete resolution
- Initially had episode about 2x/year, but became more frequent in early 2000s
  - Sent to uveitis specialist who performed AC tap, (+) HSV
  - Oral acyclovir at therapeutic dose followed by prophylactic dose
  - Despite acyclovir, continued to have multiple episodes per year, sometimes as often as every 2 months

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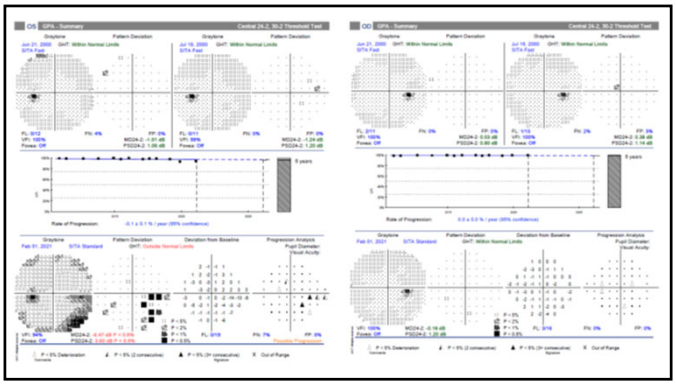

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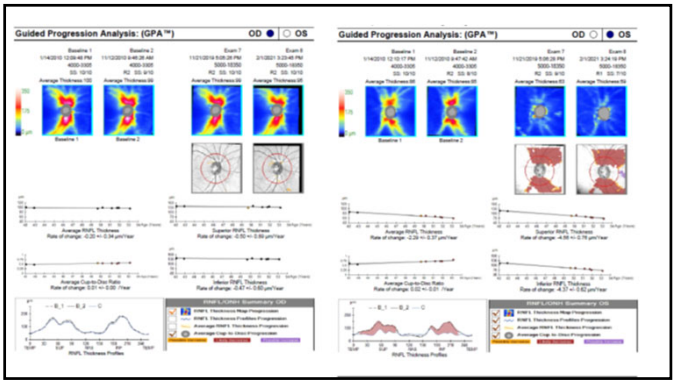

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Inflammatory: Fuch's Heterochromic Iridocyclitis

- Rare, UNILATERAL low grade chronic iridocyclitis with iris heterochromia (lighter typically on involved side)
- Clinically:
  - Low grade AC reaction
  - Small stellate KP
  - PSC**
  - Fine blood vessels on gonioscopy that do not lead to PAS
- Insidious onset in middle age, females > males
  - Often associated with HSV, CMV, RV
- OAG in 15% of cases
  - Glaucoma does not correlate with degree of inflammation
  - Can be difficult to control
    - Inflammation often does not respond to steroid
    - IOP can be very difficult to control medically
    - Laser trabeculoplasty is not indicated

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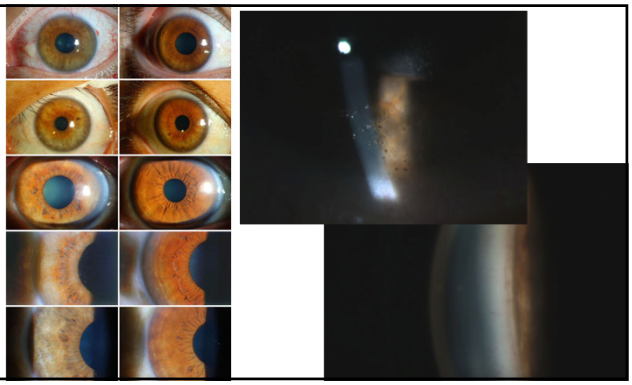

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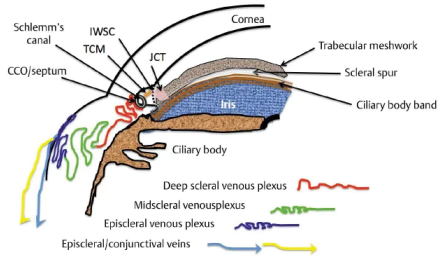

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Case: Why Are My Eyes STILL Red?

- 60yo HF presents complaining of worsening red eyes
- HPI:
  - (-) photophobia or eye pain
  - (+) itching & tearing
  - Diplopia "periodically" x 2 months
  - Most recent diagnosis was unspecified conjunctivitis WITH CNVI palsy due to DM

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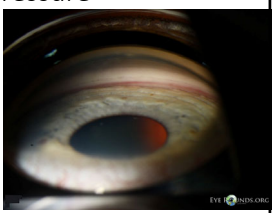
Elevated Episcleral Venous Pressure

- Normal EVP is 8-10mmHg; can be elevated by a variety of conditions that block venous outflow (unilateral or bilater)
  - Carotid cavernous fistula
  - Orbital varix
  - Sturge-Weber syndrome
  - Retrobulbar mass
  - Thyroid Ophthalmopathy

****Key clinical finding is dilated tortuous episcleral veins***

- Gonioscopy: blood in Schlemm's canal
- Treatment:
  - aqueous suppressants, PGAs (?)
  - SLT ineffective
  - Surgical complication risk: ciliochoroidal effusion, suprachoroidal hemorrhage



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Patient History

- POH: Unremarkable
- PMH: DM2 x 4 years; HTN
  - Recent hospitalization due to severe HA
- FHx: non-contributory
- Meds: Benicar®, metformin
- All: None
- Social: No tobacco/alcohol

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Examination:

- VA: 20/20 OD, OS
- Pupils: equal, round, 3+D/C, (-) RAPD
- Motility: abduction deficit OU
- Slit lamp:
  - L/L/L – normal
  - Conj – 4+ hyperemia with corkscrew vessels, chemosis
  - Clear cornea, AC
- IOP: 32mmHg OD 24mmHg OS
- DFE: Normal DMVP OU

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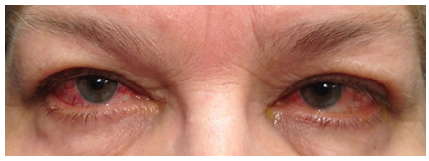

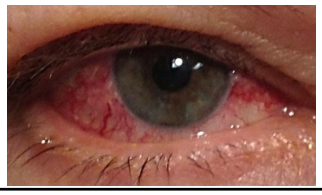

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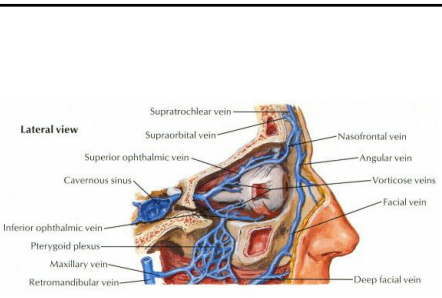

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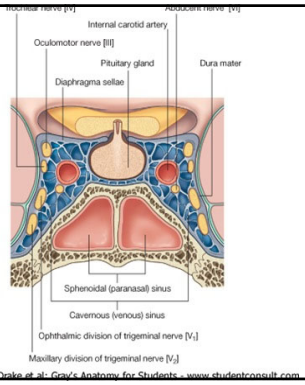
Cavernous Sinus:

- ICA
- CNIII
- CNIV
- CNV1
- CNV2
- CNVI



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Carotid Cavernous Fistula

- Pathophysiology: Abnormal communication between the ICA (and its branches) and the cavernous sinus
- Causes reversal of blood flow from the superior ophthalmic vein resulting in venous congestion

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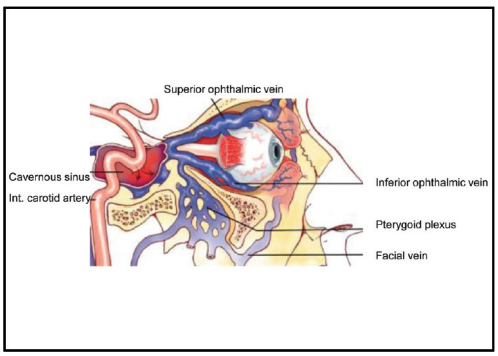

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Angle Recession Glaucoma

- Angle recession: cleavage/tear between circular and longitudinal muscles of CB
- Often associated with direct TM damage, as well
- Gonioscopy:
  - Broad angle recession
  - Absent or torn iris processes
  - White glistening scleral spur
  - Depression of overlying TM
  - Localized PAS at edge of recession

Traumatic Glaucoma

- Elevated IOP can occur due to inflammation, blood and RBCs, and direct injury to TM
- HYPHEMA:
- Several mechanisms for elevated IOP
  - Re-bleed
  - RBCs obstructing TM
  - Sickle cell hemoglobinopathies: RBCs tend to sickle in AC, making it harder to pass through TM
- Uncomplicated hyphema: manage conservatively
- If increased IOP: aqueous suppressants (avoid CAI in sickle cell)
- AC washout may be needed if IOP elevated too high or too long

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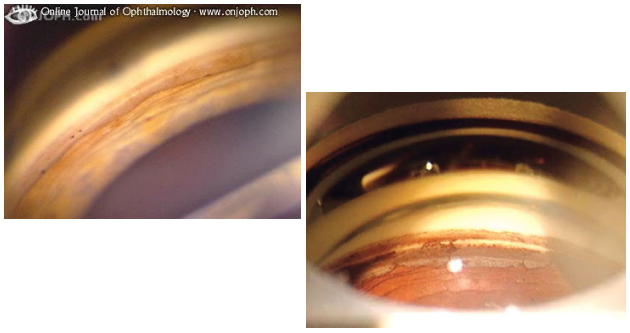

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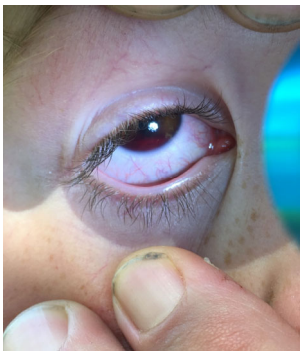
James, 9 year old male

- Sudden pain and poor vision OD after being struck with nerf bullet
- Exam findings:
  - VA: 20/60 OD, 20/20 OS
  - Pupils, motility, CVF: normal OD, OS
  - Layered hyphema 20% height OD
  - IOP 20mmHg OD, 16mmHg OS
  - DFE: Normal DMVP OD, OS
- Management?
  - Short term
  - Long term



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Angle Recession Glaucoma

- Chronic UNILATERAL glaucoma that can occur years to months after injury
- A significant portion of fellow eyes (50%) will develop increased IOP, suggesting predisposition to develop glaucoma
- Risk of glaucoma increases with degree of angle recession
- Risk if LIFELONG
  - Baseline OCT, VF, photos helpful after injury
- Treatment:
  - Medical: TM outflow agents not likely effective
  - Laser trabeculoplasty of limited value/success

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SECONDARY ANGLE CLOSURE GLAUCOMAS

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Secondary Angle Closure WITH Pupillary Block

- Lens-induced
- Phacomorphic: much more rapid development than primary angle closure with pupillary block
  - Lens swelling in eye not pre-disposed to angle closure
  - Difference in AC depth
  - Difference in cataract
- Ectopia Lentis:
  - Trauma
  - Marfan Syndrome
  - Homcystinuria
  - Microspherophakia
  - *Exfoliation syndrome

TREATMENT: REMOVE LENS

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Secondary Angle Closure WITHOUT Pupillary Block

- Two mechanisms for Secondary Angle Closure without pupillary block:
  - "Anterior Pulling": Contraction of inflammatory, hemorrhagic, or vascular membrane in angle -> PAS
    - NVG
    - ICE Syndrome
  - "Posterior Pushing": Forward displacement of lens-iris diaphragm, often accompanied by swelling and anterior rotation of ciliary body
    - Aqueous misdirection
    - Drug-induced

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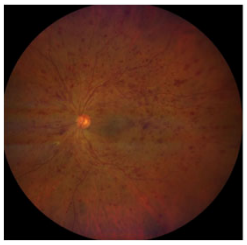
Neovascular Glaucoma (Open or Closed angle)

- Most commonly associated with proliferative diabetic retinopathy, CRVO, and ocular ischemic syndrome
- Clinical presentation:
  - Often acute onset of pain and decreased vision
  - Conjunctival injection, microcystic corneal edema, high IOP
  - GONIOSCOPY: neovascularization in TM, PAS that stop at SL**
  - Typically have visible retinal cause
    - If no obvious cause, consider carotid occlusive disease

Angle Neovascularization



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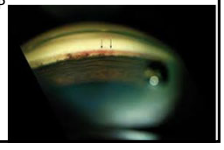
Neovascular Glaucoma

- Management:
  - Acute: aqueous suppressants (often need oral CAI) will not be sufficient
  - Urgent referral for anti-VEGF and panretinal photocoagulation or other retinal ablation
  - Retinal ablation markedly decreases neovascularization, but IOP may remain elevated depending on PAS
  - Trabeculectomy or tube shunt often needed to control IOP
  - Cryoablation if tube/trab fails
- VERY POOR PROGNOSIS – early detection is key



Case: Gloria, 57yo HF

- CC: Pain and blurred vision OD for several ***months***
- Ocular History: Unremarkable
- Med Hx:
  - Type 2 DM (A1C 6.5%)
  - Systemic hypertension (108/68)
  - Rheumatoid arthritis
- Exam:
  - BCVA: 20/50- OD 20/20 OS
  - Pupils: 4mm OU, sluggish reaction, (+) RAPD OD
  - SLE: microcystic edema OD, NVI OU
  - IOP: 51mmHg OD, 26mmHg OS
  - Gonioscopy: difficult due to K edema; multiple areas large PAS OD only (open to SS OS without neovascularization or PAS

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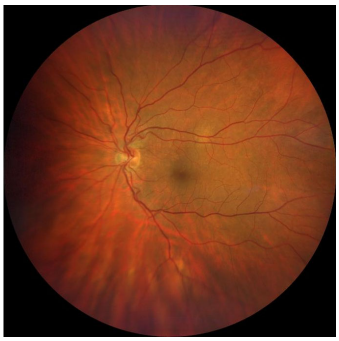

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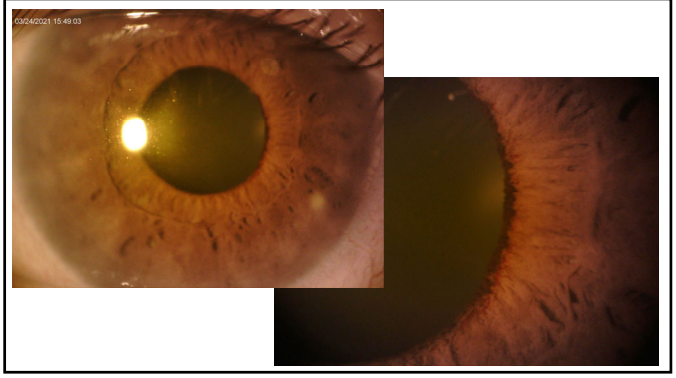

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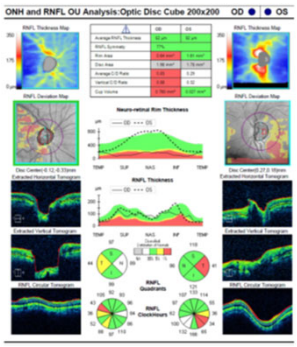

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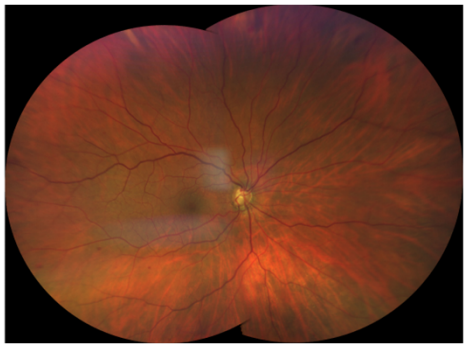

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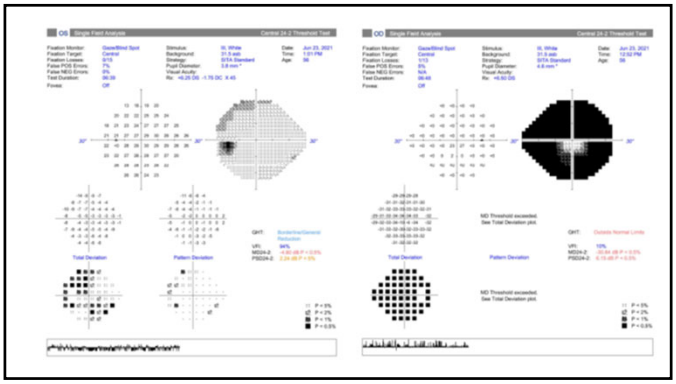

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Final disposition…

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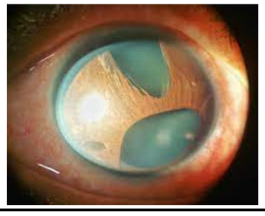
Iridocorneal Endothelial (ICE) Syndromes

- Progressive Iris Atrophy:
  - Severe iris atrophy with heterochromia and ectropion uveae, iris stromal and pigment epithelial atrophy, iris holes



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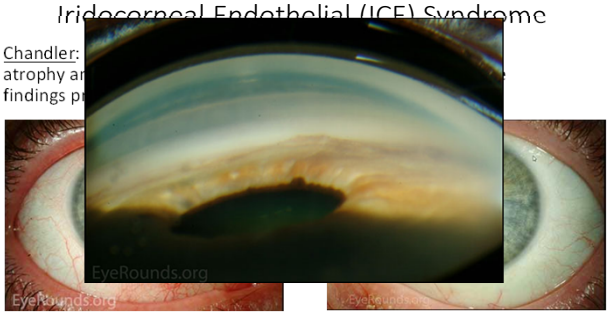
Iridocorneal Endothelial (ICE) Syndrome

- Chandler:
  atrophy ar
  findings p



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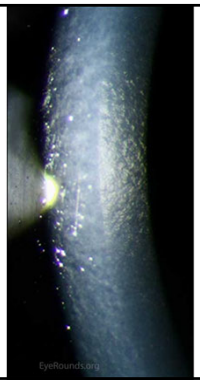
Iridocorneal Endothelial (ICE) Syndromes

- Characterized by triad: abnormal corneal endothelium, varying degrees of iris atrophy, and secondary glaucoma
- UNILATERAL, found in 20-50 year old age range, women>men
- In EACH syndrome, corneal endothelium has "beaten metal" appearance similar to Fuch's endothelial dystrophy AND "high" PAS (anterior to SL) – the PAS can help to separate from uveitic and NV
- Degree of PAS does not correlate with elevated IOP
- Varying degrees of iris atrophy and corneal changes separate the 3 types



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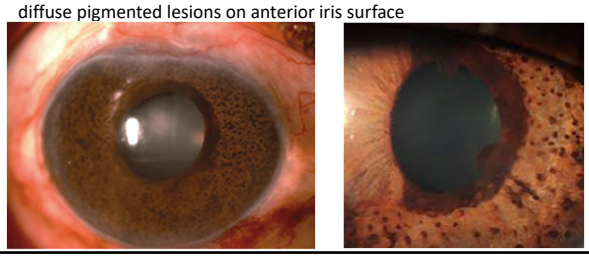
Iridocorneal Endothelial Syndrome

- Cogan-Reese: less severe iris atrophy; tan pedunculated nodules or diffuse pigmented lesions on anterior iris surface



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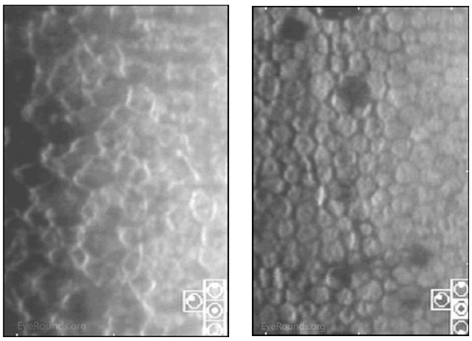

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ICE Syndromes - Glaucoma

- Glaucoma occurs in 50% of ICE syndrome patients
  - Glaucoma tends to be worse in Progressive Iris Atrophy and Cogan-Reese
- MUST ALWAYS THINK OF ICE in young to middle-aged patient with UNILATERAL angle closure glaucoma
  - Specular microscopy can reveal asymmetric endothelial loss and highly atypical morphology of involved eye's corneal endothelium
- Treatment geared toward managing corneal edema and IOP
  - Trabecular outflow drugs not effective
  - Laser trabeculoplasty not effective
  - Surgical therapy has low long-term success

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Aqueous Misdirection

- AKA "malignant glaucoma" and "ciliary block" glaucoma
- Usually presents following intraocular surgery in patient with h/o angle closure glaucoma or PAS, -OR- in open angle glaucoma patient after cataract surgery
- ACUTE presentation with UNIFORM flattening/shallowing of anterior chamber and elevated IOP
- Thought to result from anterior rotation of CB and misdirection of aqueous in association with relative block of aqueous movement at the level of the lens equator, vitreous face, and CB processes

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Aqueous Misdirection

- Diagnosis of Exclusion
- Differential Diagnosis:
  - Pupillary block
    - Moderately deep *central* anterior chamber depth
    - Anteriorly bowed iris
    - Unlikely if patent PI
  - Choroidal detachment
    - Usually low IOP
  - Suprachoroidal hemorrhage*
    - DFE/b-scan ultrasound

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Aqueous Misdirection

- Management:
  - Medical:
    - Intensive cycloplegic (NO MIOTICS)
    - Aqueous suppressants
    - Hyperosmotics (?available)
  - Laser:
    - YAG laser to vitreous face (if pseudophakic)
    - LPI (if not already present)
  - May require return to OR
    - 5-day limit for conservative management
  - Fellow eye: At high risk after intraocular surgery

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Drug-Induced Secondary Angle Closure

- Most common: topiramate (Topamax, Trokendi, Qsymia)
  - Has also been reported with acetazolamide
- Clinical Presentation:
  - Acute myopic shift >6D
  - BILATERAL angle closure with uniformly shallow/flat AC
  - Usually occurs within 1 month of initiation of topiramate therapy
- MANAGEMENT: DIFFERENT THAN PRIMARY ANGLE CLOSURE!!!!
  - Immediate d/c topiramate
  - Strong cycloplege (NOT pilocarpine!!!)
  - Aqueous suppressants (avoid oral CAI)
  - Corticosteroid
  - Resolves in 24-48 hours

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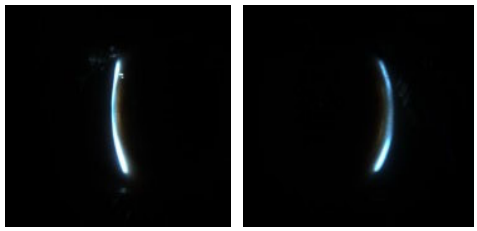

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"My Eye Hurts & I Can't See"

- 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

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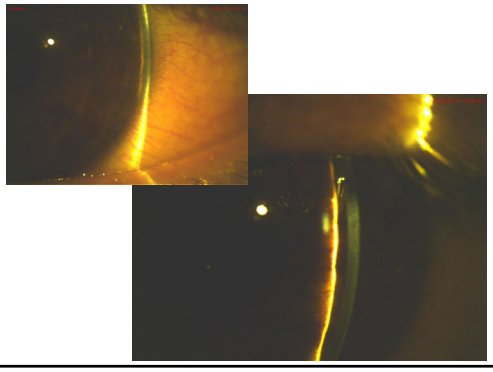

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Clinical Exam

- 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

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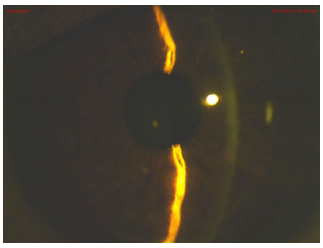
Treatment:

- 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 pga
  - 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



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Thank You For Your Attention!

Questions?
Email me: dmarrelli@uh.edu

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