GLAUCOMA GRAY AREAS

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- This presentation examines two areas that fall into the gray area of clinical glaucoma understanding. The first is the diagnosis and management of children who either have glaucoma or are suspected of having glaucoma. The second examines patients with glaucoma who follow a more ominous course with greater risk and likelihood of visual disability. Discussion will focus on features that may portend a poorer prognosis and alert practitioners to be mindful in follow up and aggressive in medical management and care.

PEDIATRIC GLAUCOMAS

PRIMARY PEDIATRIC GLAUCOMAS

- **Primary congenital/neonatal: birth – 1 mos**
- **Primary infantile: 1 mos – 2 yrs (infantile onset)**
  - Late onset > 2 years
  - Developmental anomaly of the trabecular meshwork – trabeculodysgenesis
- **JOAG does not have angle abnormality**

PRIMARY INFANTILE/ CONGENITAL GLAUCOMA

- **Aqueous outflow is impaired by an isolated trabeculodysgenesis**
- **Maldevelopment of the trabecular meshwork, including the iridotrabecular junction, with no other major ocular abnormalities**

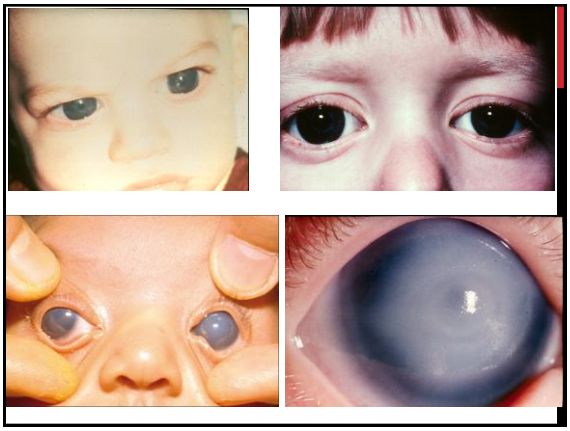
PRIMARY INFANTILE/ CONGENITAL GLAUCOMA

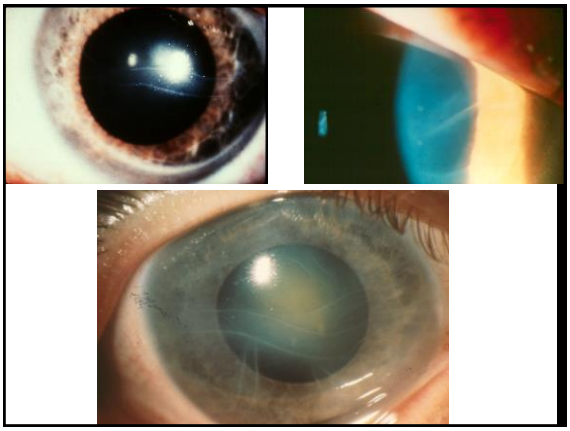
- **Absence of the angle recess**
- **Flat iris insertion: iris inserting flat and flush at or anterior to the scleral spur.**
- **Concave insertion: Superficial iris tissue sweeps over the iridotrabecular junction. The scleral spur and ciliary body are obscured**
- **Glaucoma: The iris and ciliary body have failed to recede posteriorly, and thus the iris insertion and anterior ciliary body overlap the posterior portion of the TM**

Loch Ness Monster Sign

PRIMARY INFANTILE/ CONGENITAL GLAUCOMA

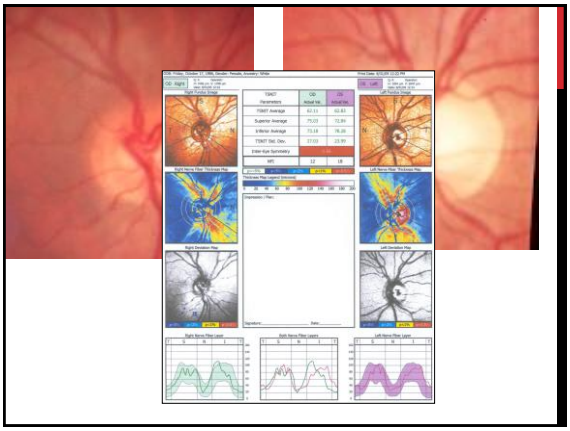
- The classic triad is epiphora, photophobia, and blepharospasm
- Corneal clouding from edema
- Descemet's tears-horizontal or vertical (Haab's striae)
- Known as buphthalmos
- Megalocornea-corneal enlargement (>12mm)





PEDIATRIC GLAUCOMA SUSPECT: MANAGEMENT STRATEGIES

- **Infant: Descemet's tears, megalocornea, classic triad, corneal edema**
  - Obvious referral to pediatric glaucoma surgeon:
- **Large c/d ratio w/o IOP rise:**
  - Photos and imaging
- **Elevated IOP with normal angle and disc**
  - Photos and imaging
  - Give to parents



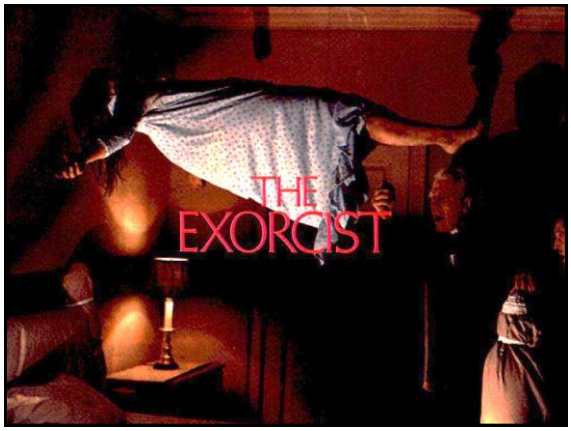
CASE

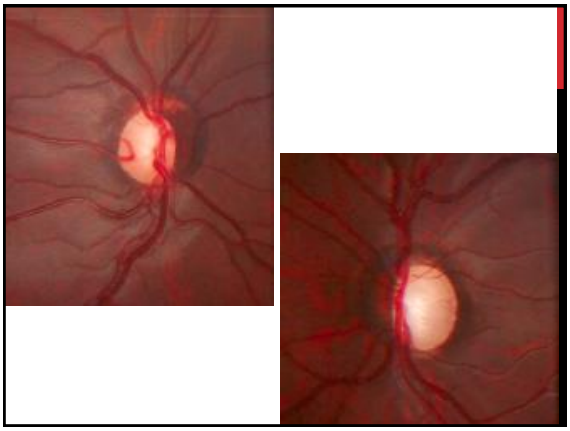
- 8-year-old Haitian female
- Blur @ distance OS – 1st eye exam
- Healthy; normal development; born in Haiti
- (+) maternal uncle with glaucoma- Age 40
- BVA 20/20 OD, 20/60 OS
- (+) RAPD OS
- IOP 21 mm Hg OD, 24 mm Hg OS
- Large cupping OU; disc pallor OS
- Referred for evaluation of OS optic neuropathy
  - MRI recommended

EXAM RESULTS

- **MRI not performed**
- **Confrontation fields**
  - Could not perform
- **Threshold perimetry**
  - Are you kidding?
- **OCT**
  - No luck there either

Now, I'm not saying the kid was hard to work with…





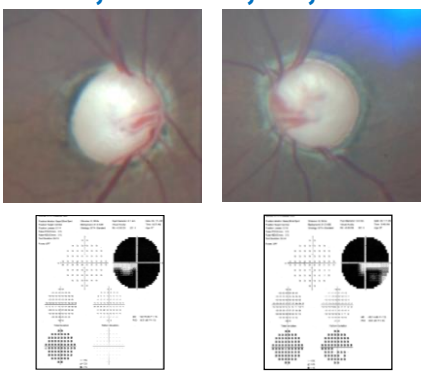
EXAM RESULTS

- **IOP: 30 mm Hg OD, 36 mm Hg OS**
- **CCT: 561 OD, 551 OS**
- **No pigment dispersion**
- **Gonioscopy:**
  - Irregular pigmentation
  - Open
  - No developmental abnormalities; no trabeculodysgenesis
- **Dx: Juvenile open angle glaucoma (JOAG)**

JOAG

- **OAG diagnosed during childhood**
- **Occurring between age 3 years and early adulthood (40 yrs?)**
  - Pressure rise occurs after 3rd birthday, but before 16th birthday
- **More aggressive course in JOAG than POAG.**
- **Anterior segment and anterior chamber angle normal**
- **Appears to be autosomal dominant**

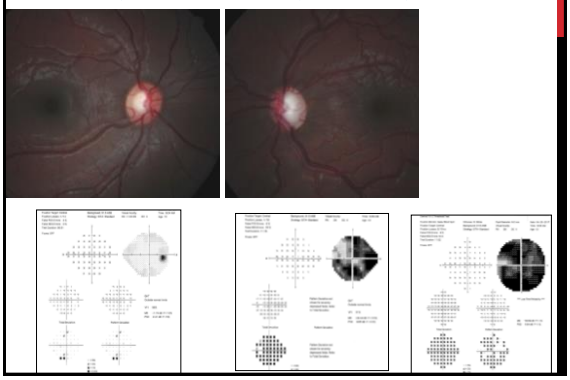
EDNA, 20/20 OD, OS, AGE 37



JOAG

- **Developmental immaturity of the trabecular meshwork**
  - Endothelial cells lining the inner wall of Schlemm's Canal lack giant vacuoles
  - Thick, compact tissue on the anterior chamber side of Schlemm's canal
  - Abnormal deposition of ground substances.
- **Essentially normal appearance**
- **Note: There are no 'normal tension' JOAG pts**
  - IOP *will* be high

13 YOF; IOP: 28 OD, 43 OS, APD OS

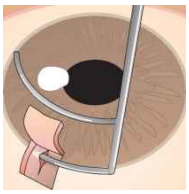


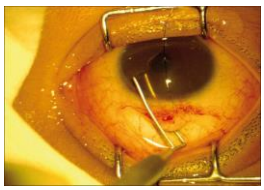
OTHER PEDIATRIC GLAUCOMAS

- **Glaucoma associated with congenital cataract surgery**
  - In aphakic or pseudophakic children following congenital cataract surgery.
  - Mechanism in aphakic glaucoma is unclear, but gonioscopy may reveal a blockage of the trabecular meshwork secondary to an acquired repositioning of the iris against the posterior trabecular meshwork. Also, prolapsed vitreous may block meshwork
  - There is often associated abnormal pigmentation and synechiae formation within the meshwork.
- **Glaucoma associated with non-acquired systemic disease or syndrome**
  - Sturge-Weber
- **Glaucoma associated with non-acquired ocular anomalies**
  - Axenfeld syndrome
- **Glaucoma associated with acquired conditions**
  - Glaucoma can occur in a pediatric patient from a number of other causes including but not limited to trauma, inflammation, episcleral venous pressure elevation as seen in Sturge-Weber syndrome, tumor, pupil block from subluxation, retinopathy of prematurity and infectious disease

MANAGEMENT IN CHILDREN

- **Primary infantile/ congenital glaucoma**
  - Goniotomy or trabeculotomy
  - Primary medical therapy for infantile glaucoma inappropriate
  - Medicines only adjunctively with surgery
- **JOAG, Aphakic, Secondary Glaucoma**
  - Medicine or surgery (trabeculectomy, tube, goniotomy, trabeculotomy)
  - Case dependent





MEDICAL MANAGEMENT IN CHILDREN

- **Topical beta blockers: Safe and effective in children.**
- **Prostaglandin analogs: Safe, well tolerated, but not very effective.**
  - Best for older children with JOAG.
- **Topical CAI: Safe and effective**
  - Probably the best option.
- **Brimonidine: Effective, but unacceptable side effects. Should not be used**

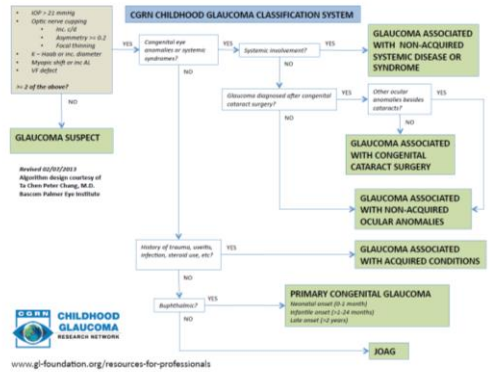
UNDERSTANDING PEDIATRIC GLAUCOMA

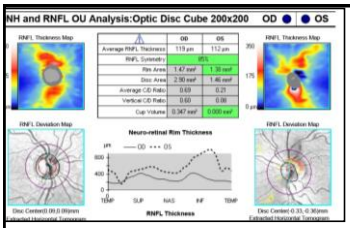
PRIMARY INFANTILE/ CONGENITAL

- Abnormal angle
- Globe enlargement
- Corneal edema
- Onset near birth
- Megalocornea
- Symptomatic
  - Blepharospasm, photophobia, lacrimation

OPEN ANGLE/ SECONDARY

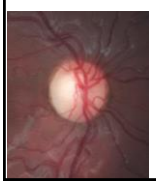
- Normal angle
- Normal axial length
- Clear cornea
- Onset later
- Normal corneal size
- Asymptomatic

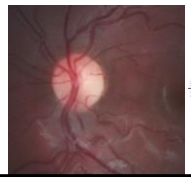


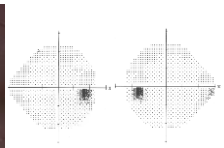


15 YOF diagnosed with JOAG OU
IOP 16 mm Hg OU

Clinical Pearl: There is no 'normotensive' JOAG; IOP is always very high.



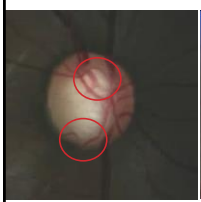


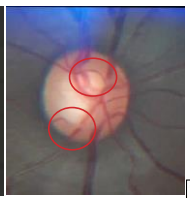


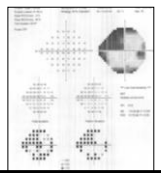
THAT BEING SAID...

- **17 YOF- glaucoma suspect at age 10 based upon disc appearance**
  - Disc normal; OCT normal
- **Peak IOP: 19 mm OD, 17 mm OS (2010)**
  - 14 mm OD, 17 mm OS (2017)
- **CCT 564 OU**
- **20/15 OD, OS**
- **Color vision normal OU**

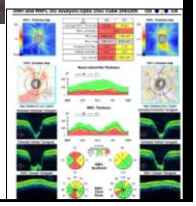
THAT BEING SAID...



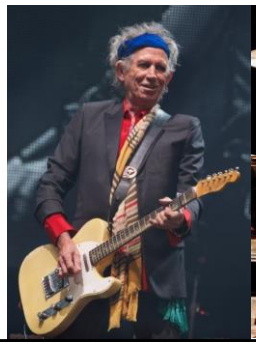


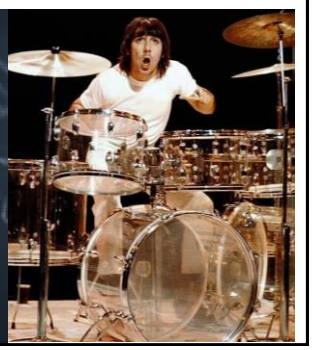


- Young age
- Asymmetric disease
- Definitive change
- Normal IOP
- Is this glaucoma?



GLAUCOMA GRAY AREAS





GLAUCOMA GUESSING GAME

- **Open angle glaucoma is typically (slowly) progressive and predictable**
- **Some patients will never become visually disabled**
- **Some patients will be blind regardless of treatment**
- **Who is who?**
- **Intermediate subset living in 'the gray area'**
  - Most likely to show progression with a poor prognosis/ outcome
  - *Often not the ones that you think*

RISK FACTORS FOR PROGRESSION:

- **IOP level**
  - The most significant modifiable risk factor for glaucoma development and progression
- **Exfoliation**
  - Higher IOP, worse disease, more difficult to control, more likely to reach visual disability- noted in numerous studies in association with progression

RISK FACTORS FOR PROGRESSION:

- **Disc hemorrhages**
  - Inferior temporal most commonly
  - Best evidence indicates mechanical shifting of tissue
  - Recurrent
    - Recurrent disc hemorrhages no more associated with progression risk than single hemorrhage, unless recurs in different part of disc
- **Time:**
  - Glaucoma is by nature a progressive disease and treatment likely only slows the progression
  - Given enough time, most will demonstrate progression and this is NOT a sign of treatment failure
    - Approximately 10 years

PROGRESSION FACTORS FROM THE MAJOR STUDIES

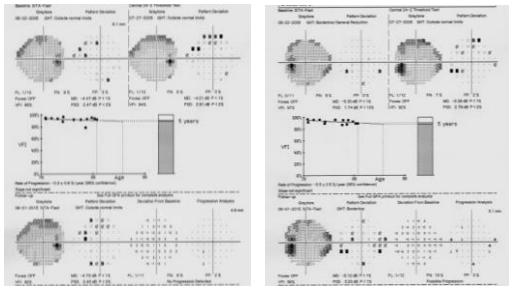
- **Disc hemorrhage (CNTGS, OHTS, EMGT)**
  - NTGS, EMGT saw no difference with IOP reduction
- **Thin cornea (EMGT, OHTS)**
- **Higher baseline IOP (EMGT, OHTS, AGIS)**
- **Exfoliation (EMGT)**
- **Cardiovascular disease (EMGT, CNTGS)**
- **Lower OPP (EMGT)**
- **Older age (EMGT, AGIS, *not* CNTGS)**

Early manifest Glaucoma Trial (EMGT)
Ocular Hypertension Treatment Study (OHTS)
Advanced Glaucoma Intervention Study (AGIS)
Collaborative Normal Tension Glaucoma Study (CNTGS)

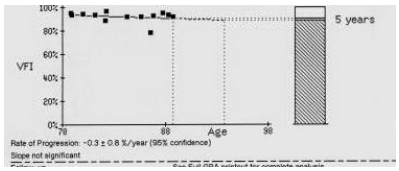
78 YOF

- **Exfoliative glaucoma OU**
- **Peak IOP: *34 mm Hg OD, 37 mm Hg OS***
  - Most typical IOP: upper 20s
- **CCT: 517 OD, 527 OS**
- **Pt declines all treatments**
  - more afraid of treatment than glaucoma
  - Wants to see change or other conclusive proof of need for treatment.
- **However, everything says she will do poorly**

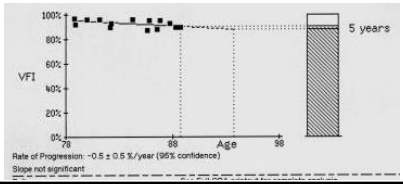
2005-2015



2005-2015



OD

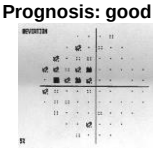


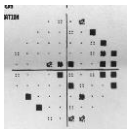
OS

Risk factors: PXE, High IOP, older age, thin CCT: yet no progression?! Ultimate outcome?

71 YOF

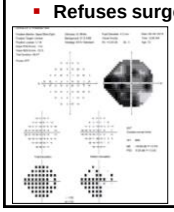
- 71 YOF; 6/6 acuity OD, OS
- Diagnosed POAG OU 2009- treated with travoprost with good response (IOP drops to 18 from 28)
- CCT: 579, 583
- Angles open- no evidence of secondary glaucoma
- Prognosis: good

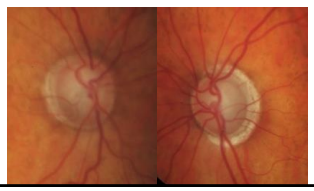




71 YOF

- Transfers care but returns in 2012: 6/9 OD, 6/120 OS
- SLT OU x2
- Meds: bimatoprost, brimonidine/timolol, brinzolamide
- Intolerant to Diamox and pilocarpine
- IOP- 22 mm OD and 38 mm OS
- Refuses surgery





71 YOF

- Uses meds- requests refills consistently; visits sporadic
- Feb 2014: 6/15 OD, LP OS; IOP 36 mm OD, 30 mm OS
  - Declines surgery again
- Feb 2015: 6/19 OD, NLP OS; Using brim/timolol only; IOP 46 mm OD and 72 mm OS; restart all meds
  - Declines surgery again
- April 2016: 6/120 OD, NLP OS; IOP: 32 mm OD and 48 mm OS
  - New views on surgery
- Both patients had an unexpected course
  - atypical

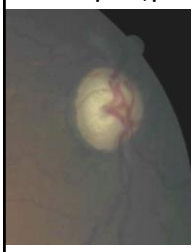
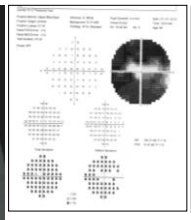
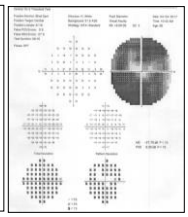
CLINICAL PEARL

- You can only call a glaucoma patient "well controlled" in retrospect
- Some patients progress slowly without treatment and some progress rapidly, even with treatment
  - You don't know who is who until you follow up over time

PATIENTS I WORRY LESS ABOUT

66 YOM: 3 YEARS

- Starting IOP 58 mm Hg; CCT 536
  - 20/25 OD; OS not seeing due to vascular occlusion
- Stepped regimen: Brimonidine, dorzolamide/timolol, latanoprost, pilocarpine (GlaucAll)- IOP 14 mm Hg

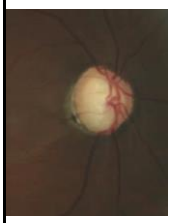
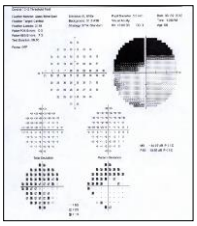
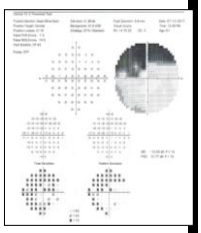




2015 2017

- Very high peak IOP
- Exceptional IOP reduction (75%) ☺

61YOM

- IOP 30 mm; CCT 545
- Latanoprost, dorzolamide/timolol – 12 mm

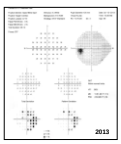




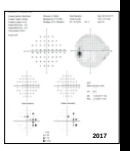
2012 2017

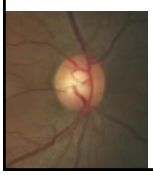
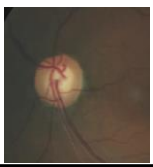
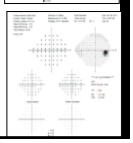
- High peak IOP
- Excellent IOP reduction (60%) ☺

53 YOM- FOLLOWED 4 YEARS

- Peak IOP: 32 mm OD, 43 mm OS; CCT 453 OD, 446 OS
- Latanoprost: 15-18 mm OD, 18-22 mm OS
  - Recently added dorzolamide



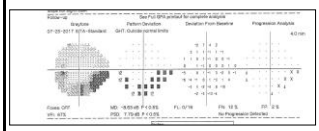
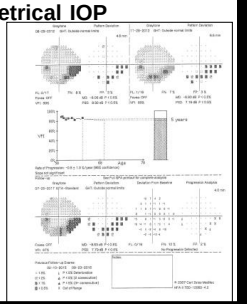


- High peak IOP
- Significant initial IOP reduction with 1 med
- Low med load ☺

63 YOF: GLAUCOMA OS X 5 YEARS

- IOP typical range: 14-18 OD; 15-18 OS; CCT: 556 OD; 543 OS
- Unilateral disease; symmetrical IOP
- Pt chooses observation.

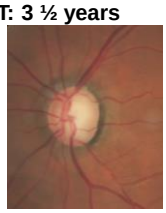
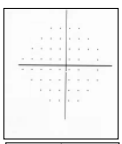
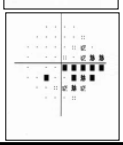



- True "normotensive" range
- Moderate disease not threatening fixation
- Stable ☺

WHICH PATIENTS REPRESENT UNSUSPECTING DANGER?

65 YOM

- Peak IOP 22 mm OD, 29 mm OS; CCT 560
  - Followed 5 ½ yrs so far
- Latanoprost, dorzolamide/timolol, brimonidine- 15 mm OD, 14 mm OS
- Time to MMT: 3 ½ years

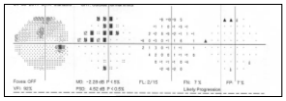





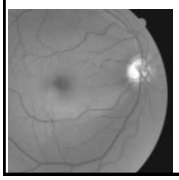
- Peak IOP not terribly high
- Short duration to MMT
- High med load for modest reduction ☹

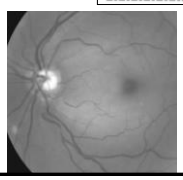
Fields unchanged; possible disc change OS

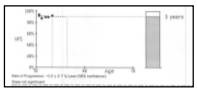
55 YOF

- **Initial: Peak IOP??- treated since age 35**
  - Timolol; CCT 472 OD, 497 OS; Disc change OD 2010
  - Currently: latanoprost, dorzolamide/timolol, brimonidine; 15 mm OU
    - Followed 11 yrs so far





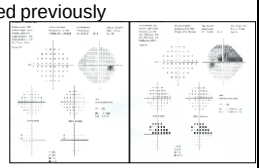




- Young age
- Long duration of treatment
- MMT
- Thin cornea ⊗

53 YOF

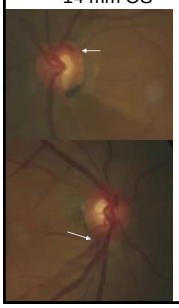
- **Peak IOP: 20 mm OD, 22 mm OS; CCT: 510 OD, 508 OS**
- **Treated IOP: 12-15 mm OD, 12-16 mm OS**
  - Brimonidine, latanoprost, dorzolamide/timolol
  - Field progression documented previously

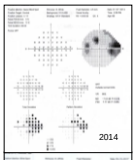


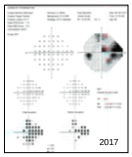
Low baseline IOP (low 20s), MMT to achieve 'modest IOP reduction, bilateral recurrent disc hemorrhages ⊗

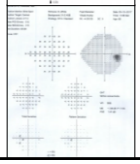
69 YOF: POAG OU X 11 YEARS

- **Peak IOP: 20 mm OU; CCT: 540 OD, 532 OS**
  - Dorzolamide/timolol; latanoprost OU; IOP- 15 mm OD, 14 mm OS



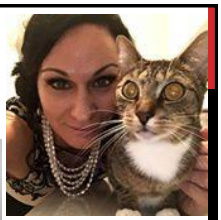


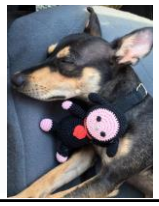




- Bilateral disc hemorrhages
- Low peak/baseline IOP
- Modest pressure reduction
- High med load
- Field/ disc change ⊗

THINGS THAT GIVE ME COMFORT





OTHER THINGS THAT GIVE ME COMFORT

- **High initial peak IOP**
  - 30s and 40s better than low 20s
- **Significant IOP reduction**
  - Regardless of disc/ field status
- **Good initial response to one medication**
- **Minimal medications**
- **High peak IOP and significant medical response**

THINGS THAT MAKE ME UNCOMFORTABLE

OTHER THINGS THAT MAKE ME UNCOMFORTABLE

- **Exfoliation**
- **Disc hemorrhages**
- **Rapid escalation in therapy**
  - Adding 2 meds w/i 3 years
- **Low peak IOP**
  - Low to mid 20s bad
  - Mid teens- not so bad
- **Poor initial IOP reduction**
- **Low peak IOP and poor initial IOP reduction**

Portends worse prognosis

ODE TO GLAUCOMA TREATMENT

When the pressure starts high and the treated drop great,

Likely a good outcome is to be the fate.

Compliance, exfoliation and disc hemorrhage must be watched,

So the case doesn't get botched.

Most patients can be predicted,

And your Zen won't be afflicted

But some patients will surprise,

And cause your blood pressure to rise.

Lowering 22 down to 18 is not enough,

Go for 50% so they don't snuff.

Joseph Sowka, OD

BE AWARE OF THE GRAY AREA WHERE DANGER LURKS…

…MAY YOU HAVE NOTHING BUT KITTENS AND BUNNIES