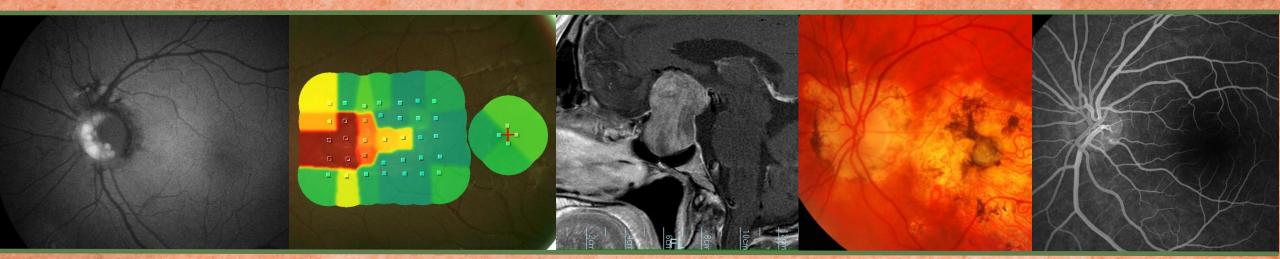
# Grand Rounds MULTIMODAL DIAGNOSTICS



#### **Richard Trevino, OD, FAAO** Indiana University School of Optometry

### **Multimodal Grand Rounds**

 Online notes -richardtrevino.net • Email us -rctrevin@iu.edu Disclosures -None

#### **Interactive Presentation**

- Vote on diagnosis and management options
  Enter below address in a web browser
  https://app.tophat.com/e/777538
  Click "Enter as a Guest"
  - Click "Mobile Site"

# **Battle of the Superheros!**

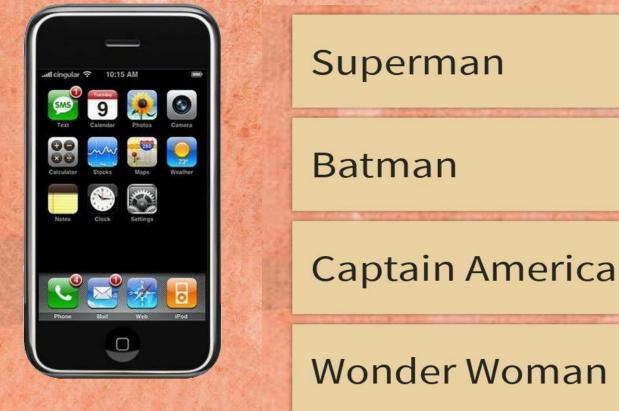
#### https://app.tophat.com/e/777538

Α

B

С

D





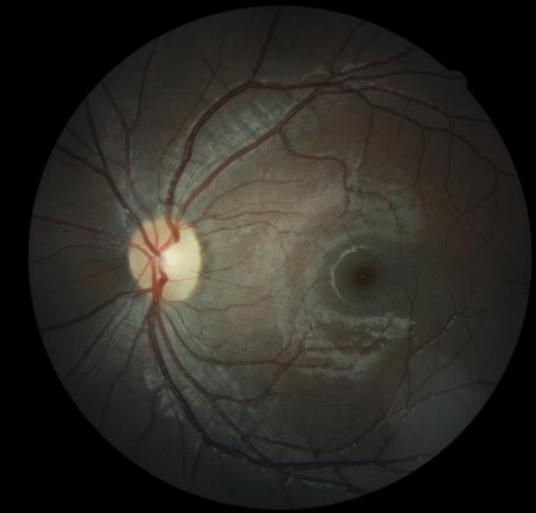
# Don't sink my battleship!

# CASE #1

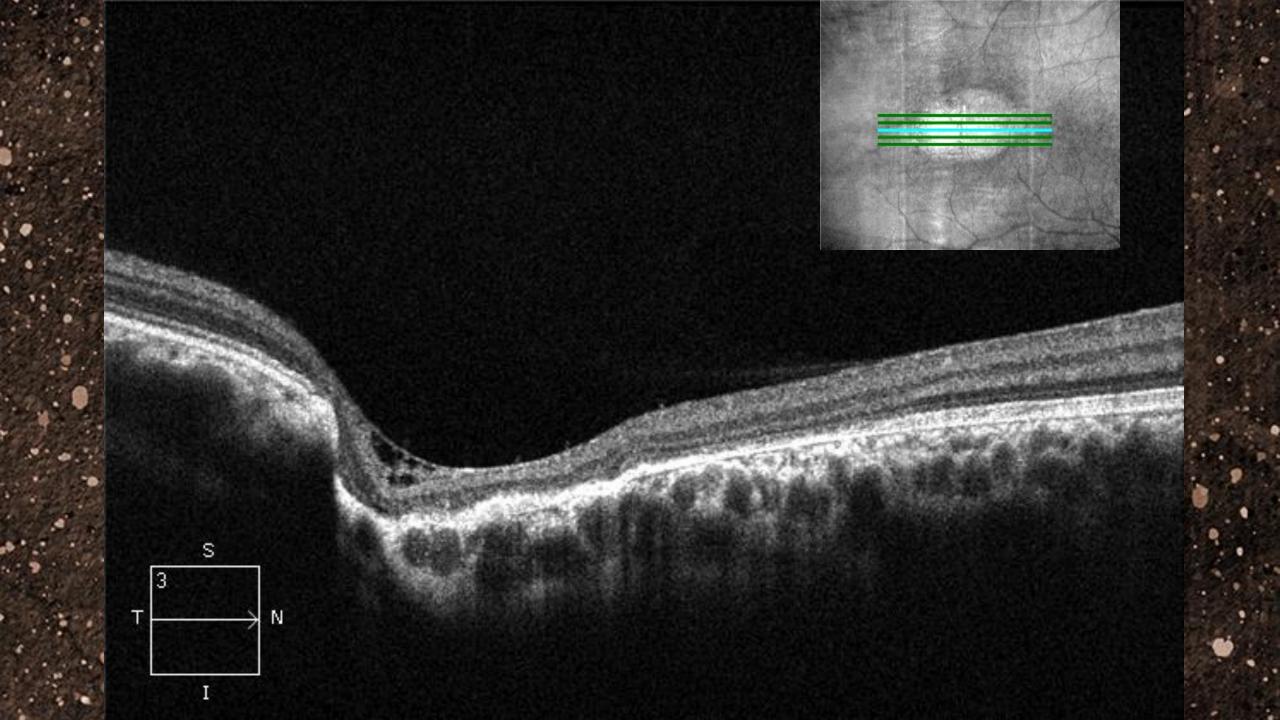
- 25yo East Indian woman presents without complaints for routine exam
- POH: Unremarkable. LEE: 1yr.
- MH: Good health. No medications
- Vision: 20/15 each eye without correction
- Entrance testing: Normal
- External exam: Normal OU
- Tonometry: 14/13 @11:00AM











Short-wavelength fundus autofluorescence (SW-FAF) uses blue light to stimulate lipofuscin in RPE cells to glow. Regions without viable RPE will appear dark.

Program: Macula screening	Stimulus: III, white	Pupil: Date of exam		Macula screening	Stimulus: III, white	Pupil: Date of exam.: 07/23/2012
Area: 10-2	Background: 10 cd/m <sup>2</sup> (31.8 asb)	Presentation time: 0.2 sec Time:	11:31:07 Area:		Background: 10 cd/m <sup>2</sup> (31.8 asb)	Presentation time: 0.2 sec Time: 11:40:27
Strategy: Threshold	Correction: No	Interval time: 0.6 sec Age:		Threshold	Correction: No	Interval time: 0.6 sec Age: 25
Fixation: Central	0 dB: 3180 cd/m² (simulated	d) Abs.loss:	0 Fixation:	Central	0 dB: 3180 cd/m <sup>2</sup> (simulated)	
Fixationcheck: 0/10 (0% Losses)		Rel.loss:	· ·	heck: 0/11 (0% Losses)		Rel.loss: 0
False positive: 1/26 (4% Error)			False pos	sitive: 0/28 (0% Error)		
Presented dots: 294				d dots: 310	T	T ADD
Duration: 05:19	35 35		ABS. Duration:		34 34	ABS.
Re-Examination: No			0000	ination: No		
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35	37 35 - 35 34 34			36	36 37 - 35 33 35	
33 37	35 34 35 35 37 36			34 33	33 35 36 35 35 35	
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		0 -2 -6 -4 -2 0 0 0	-0-0-ID	0 0 0 -2 -2 -2	0 0 -2 -2 0 0	0 -2 -2 -2 0 0 -2
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Deviation from age-	Corrected	-2 0	rolated no	from age- orm values 0 2	Corrected deviation	0 2 MS: 34.92 (35.05)
related norm values	deviation		MD: 0.67	· · ·	deviation	MD: -0.13
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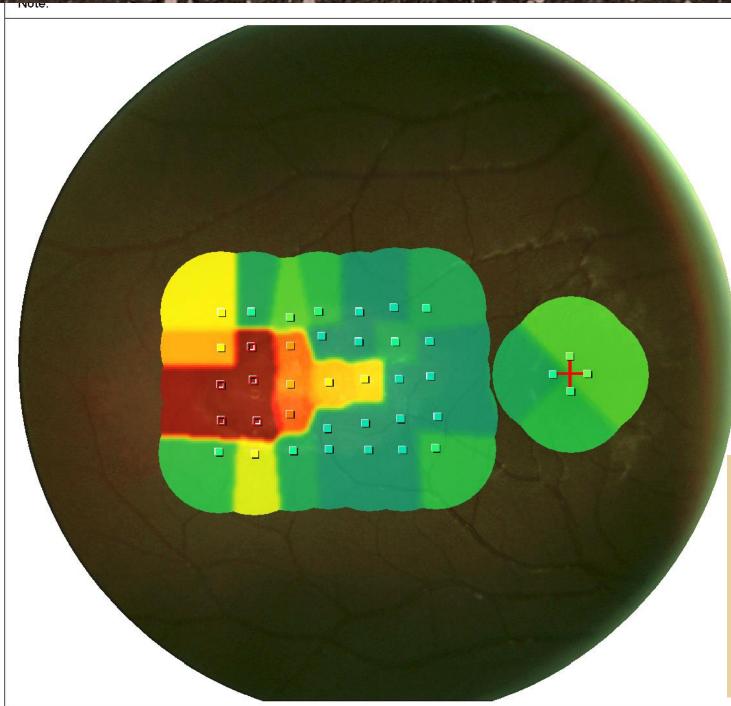
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6

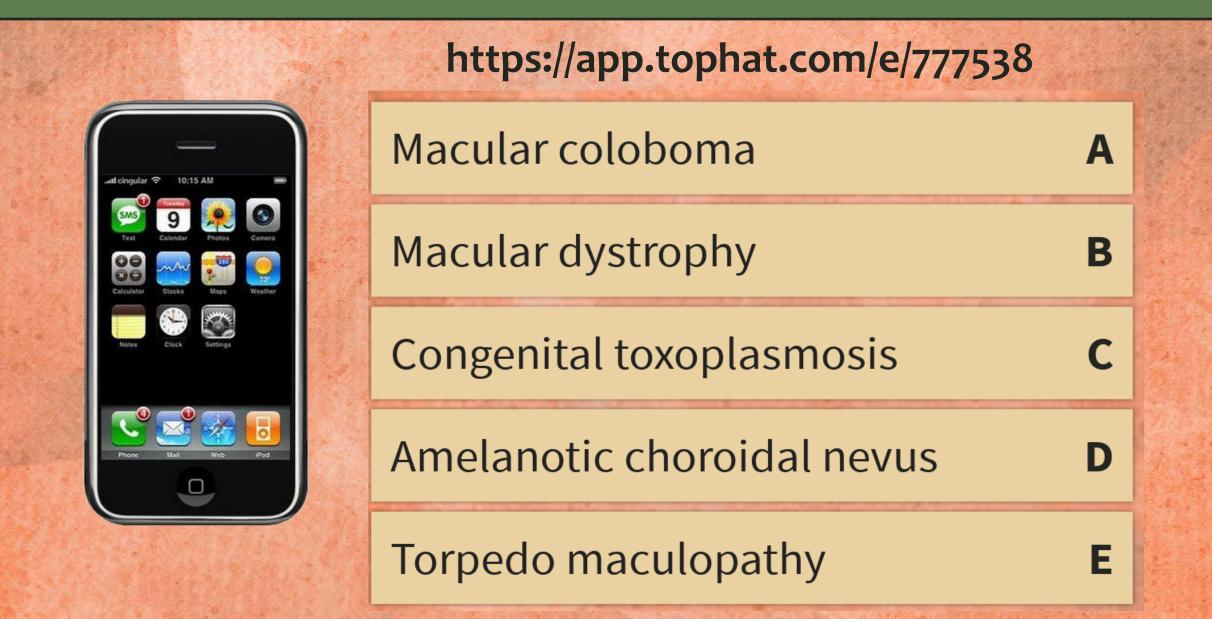
2



Microperimetry allows a retinal sensitivity map to be overlaid on a realtime fundus photograph



# What is going on here?



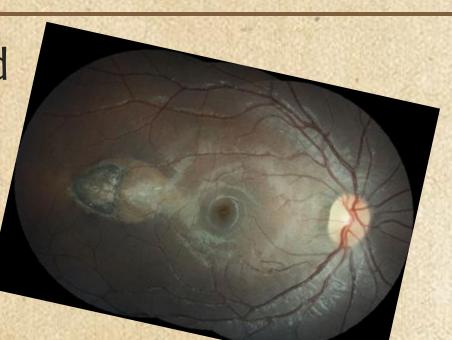
Macular coloboma (macular dysplasia)	A heterogenous group of developmental abnormalities. Often familial, frequently <b>bilateral</b> , systemic abnormalities not uncommon.
Macular dystrophy	Widespread retinal dysfunction, <b>bilateral</b> macular lesions, inheritance pattern, may be associated with features of Leber's amaurosis or RP. Electrophysiologic abnormalities common
Macular scars	Intrauterine infection with Toxoplasma gondii resulting in congenital toxoplasmic chorioretinitis.
Amelanotic choroidal nevus	Shares all the features of a typical choroidal nevus minus the melanin. Choroidal nevi are flat or slightly elevated – never excavated
Torpedo maculopathy	<u><b>Unilateral</b></u> congenital abnormality, characteristic "torpedo" shape, always located temporal to the fovea

#### Assessment

Torpedo maculopathy OD

## Management

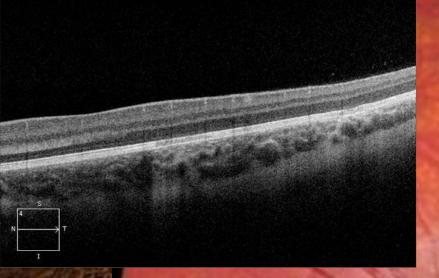
- No specific treatment indicated
- Patient education
- Routine eye care



## **Torpedo Maculopathy**

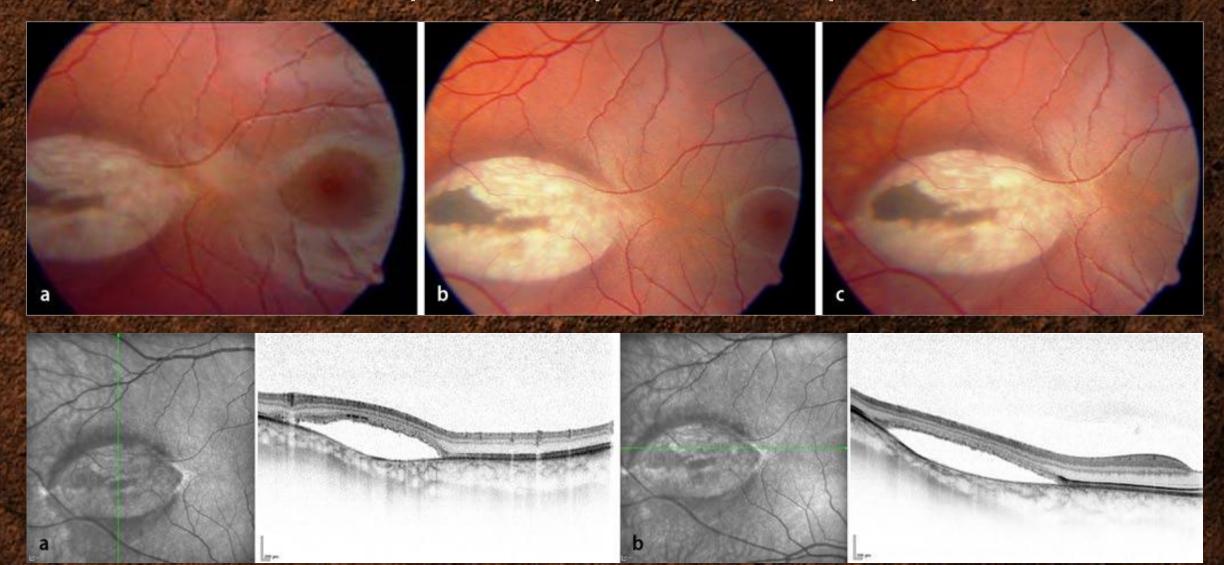
- Congenital, hypopigmented torpedo-shaped lesion in the temporal macula along the horizontal raphe
- Developmental defect of unknown etiology
- May encroach upon fovea, but rarely causes significant loss of vision
- Diagnosis based upon characteristic appearance and nonrandom location
- No treatment is required for this stable congenital lesion

Examples of torpedo maculopathy

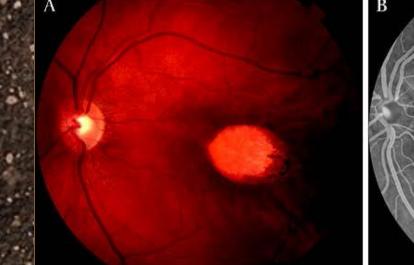


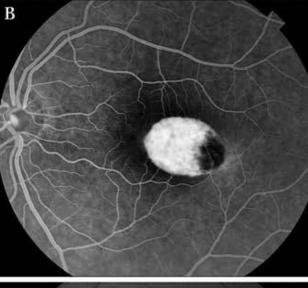
West Coast Retina, 2010

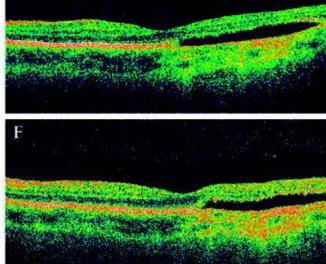
#### Examples of torpedo maculopathy



Bedar MS. Ophthalmologe, 2013;110:173

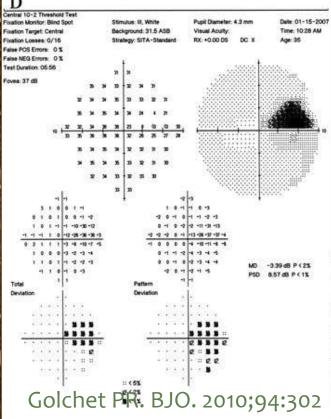


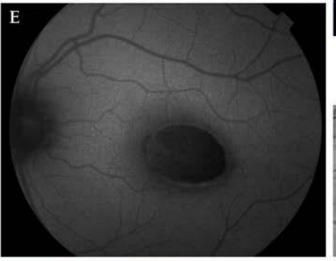


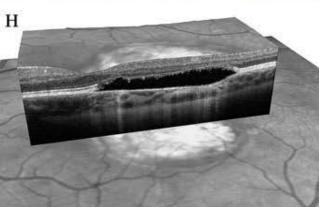


C













## **Torpedo Maculopathy**

#### **Range of clinical presentations**

#### **Constant features**

- Congenital
- Location temporal to fovea
- Horizontally oval shape
- Hypopigmentation

#### **Variable features**

- Schisis cavity
- Excavation
- Degree of pigmentation
  Visual defect



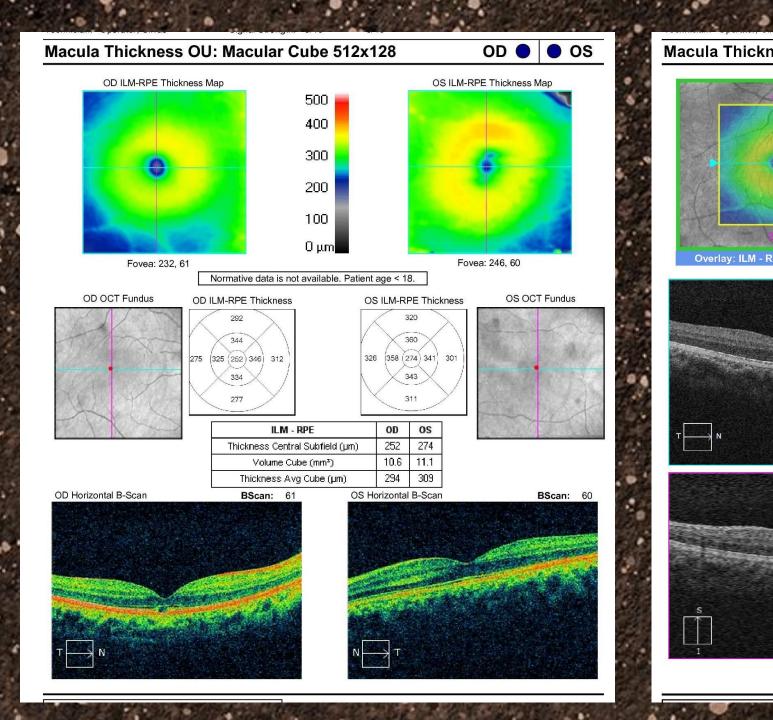
Torpedo maculopathy

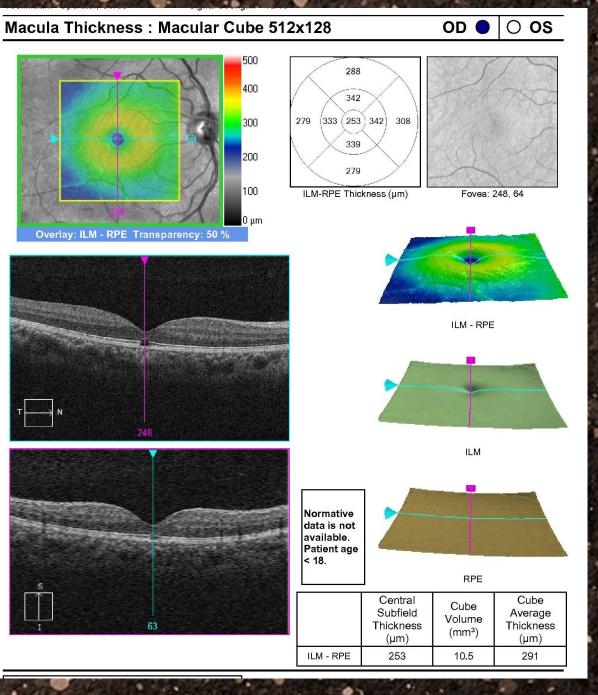
 Congenital perimacular lesion
 Distinctive appearance
 Usually asymptomatic
 Requires no special management



# CASE #2A

- 13yo Asian male referred for abnormal appearance of right macula on OCT
- POH: Refractive amblyopia OS
- PMH: Good health. No meds
- BCVA: 20/20 OD, 20/40 OS
- Ta: 12/13 @ 4PM; PERRL, NO APD
- SLE: W&Q OU

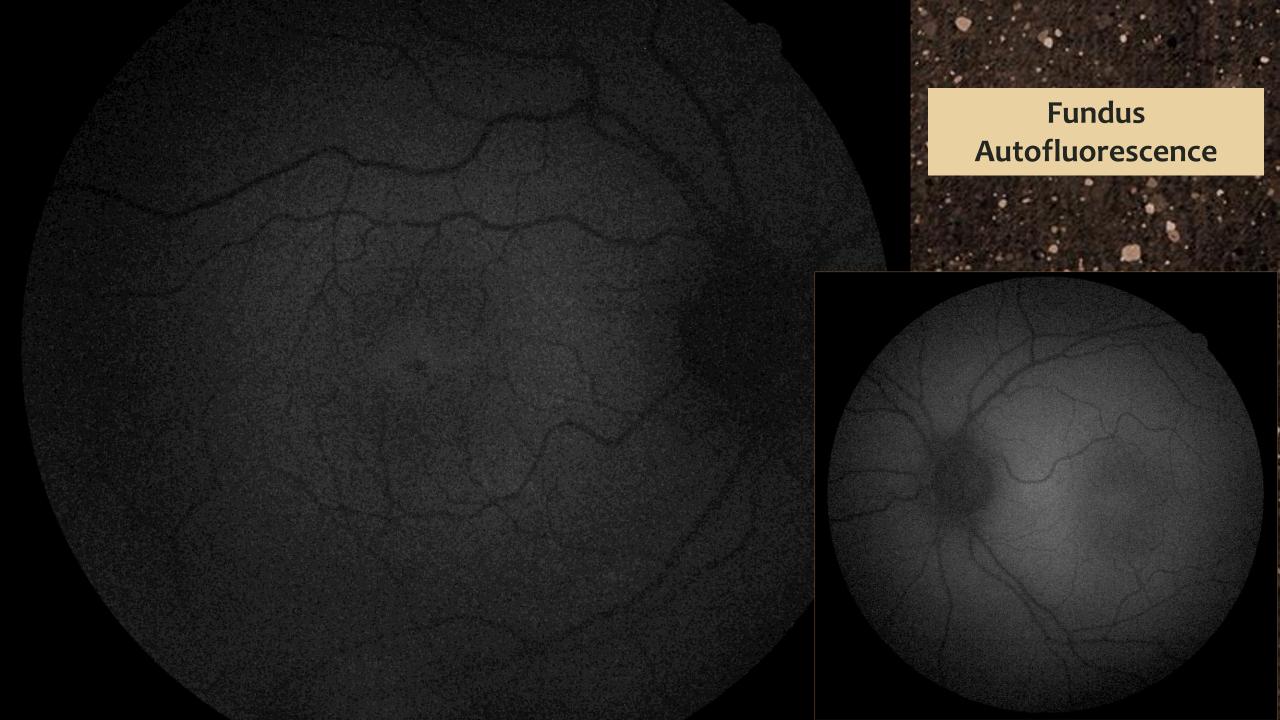




# CASE #2B

- 12yo Asian F (sibling of Case 3A) C/O "color difference" between eyes, with vision of right eye appearing yellowish x 1 month
- POH: LEE 2yrs
- PMH: Good health, no meds
- BCVA: 20/25 OD, 20/20 OS
- Ta 15/15 @11AM; PERRL NO APD
- SLE: W&Q OU





#### Macula Thickness OU: Macular Cube 512x128

#### I2x128 OD 🔵

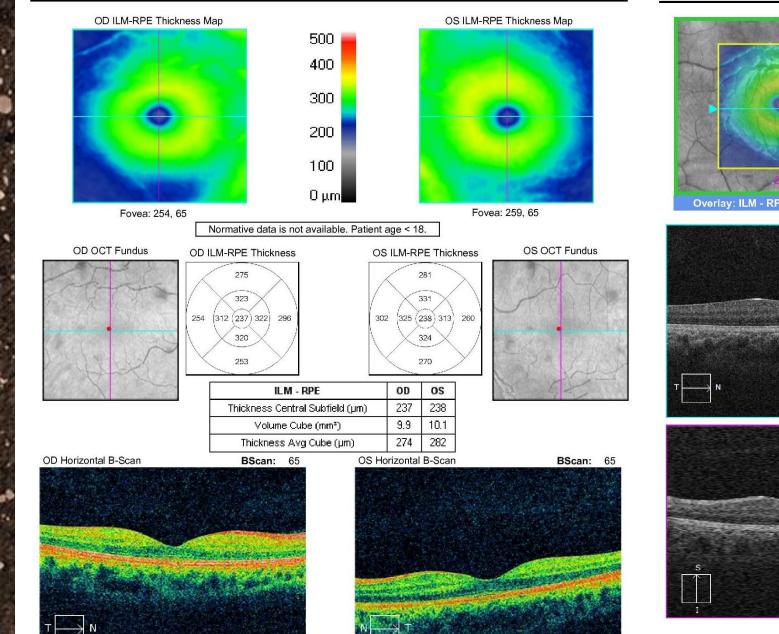
Macula Thickness : Macular Cube 512x128

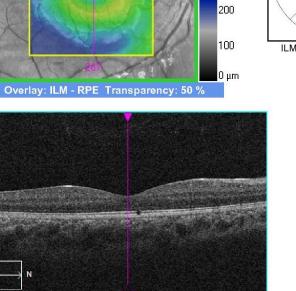
500

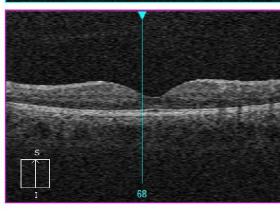
400

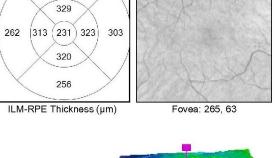
300

OD O OS

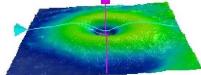




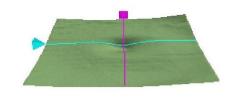




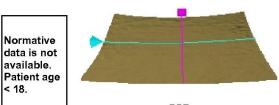
281



ILM - RPE







RPE

-		Central Subfield Thickness (µm)	Cube Volume (mm³)	Cube Average Thickness (µm)
	ILM - RPE	231	10.0	277

Dector's Signature

# What is going on here?



#### https://app.tophat.com/e/777538

Macular Coloboma

Macular Dystrophy

Macular Hole

Solar Maculopathy

Macular Edema

Macular coloboma (macular dysplasia)	A heterogenous group of developmental abnormalities. Often <u>familial,</u> frequently <u>bilateral</u> , systemic abnormalities not uncommon.		
Macular dystrophy	Widespread retinal dysfunction, <b>bilateral</b> macular lesions, <b>inheritance pattern</b> , may be associated with features of Leber's amaurosis or RP. Electrophysiologic abnormalities common		
Macular hole	<b>Full-thickness</b> retinal defect associated with major loss of visual acuity. Early stage or lamellar lesions associated with <b>inner retinal defects</b>		
Solar maculopathy	Bilateral or unilateral (dominant eye) <u>outer retinal OCT defect</u> with history of sun gazing		
Macular edema	Intraretinal fluid accumulation with a corresponding <b>increase in macular</b> thickness on OCT		

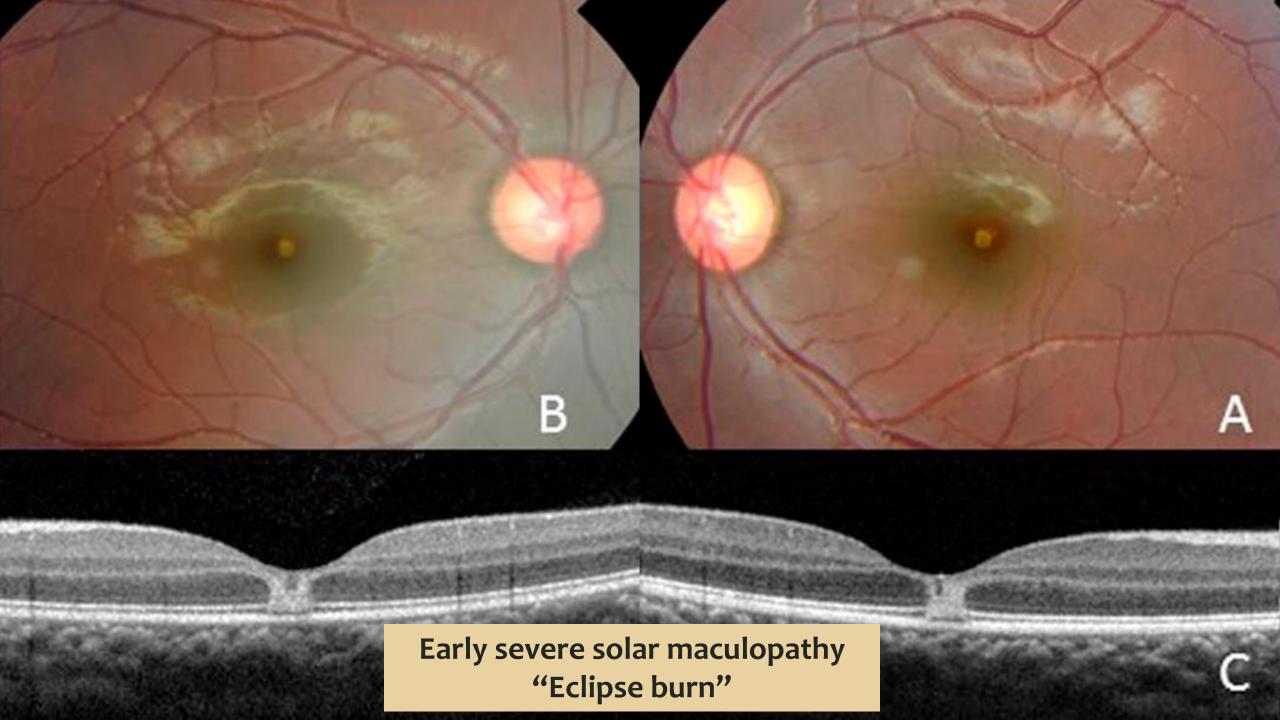
## **Solar Maculopathy**

- Thermal and photochemical damage to the RPE and photoreceptors
- Solar maculopathy: Prolonged direct sun gazing.
   Usually associated with eclipse viewing
- Photic retinopathy: Umbrella term that encompasses all retinal damage from light (laser, arc welding, solar retinopathy)



## **Solar Maculopathy**

- Clinical manifestations vary with severity of exposure
- Rapid VA decrease (20/30-20/100) following exposure
- Little or no ophthalmoscopic changes in mild cases (indistinct FLR, greyish macula) or a yellowish RPE lesion in more severe cases
- Vision gradually (months) returns to normal or near normal. There is less recovery in severe cases.
- An outer retinal cyst-like lesion develops and remains permanently ("virtually pathognomonic")

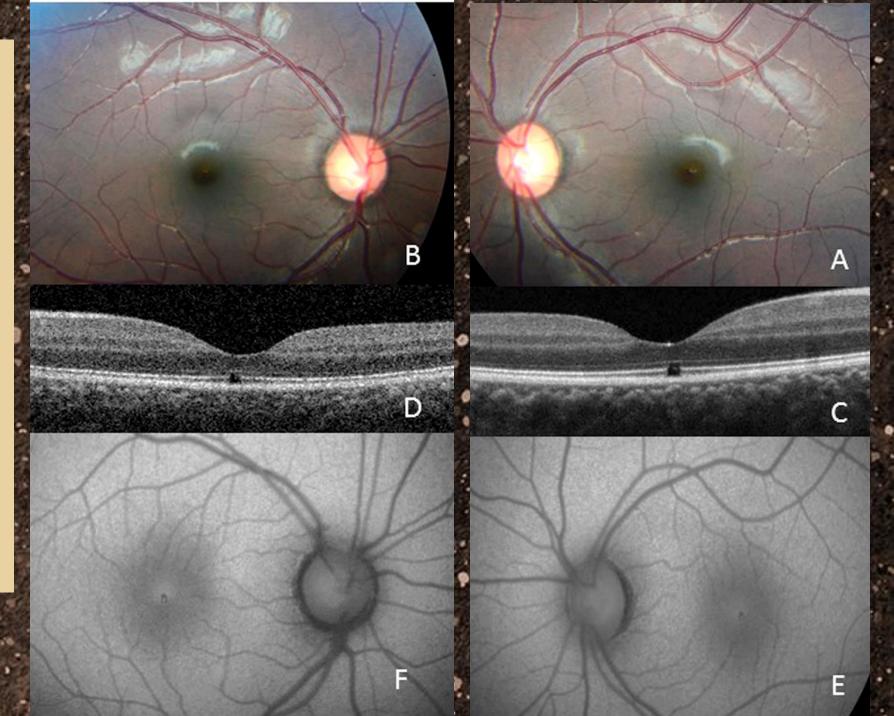


Severe solar maculopathy at 30-days

**TOP:** Resolving yellow lesions

**MIDDLE:** Square-shaped foveal photoreceptor defect

**BOTTOM:** FAF hypofluorescent spot surrounded by hyperfluorescence

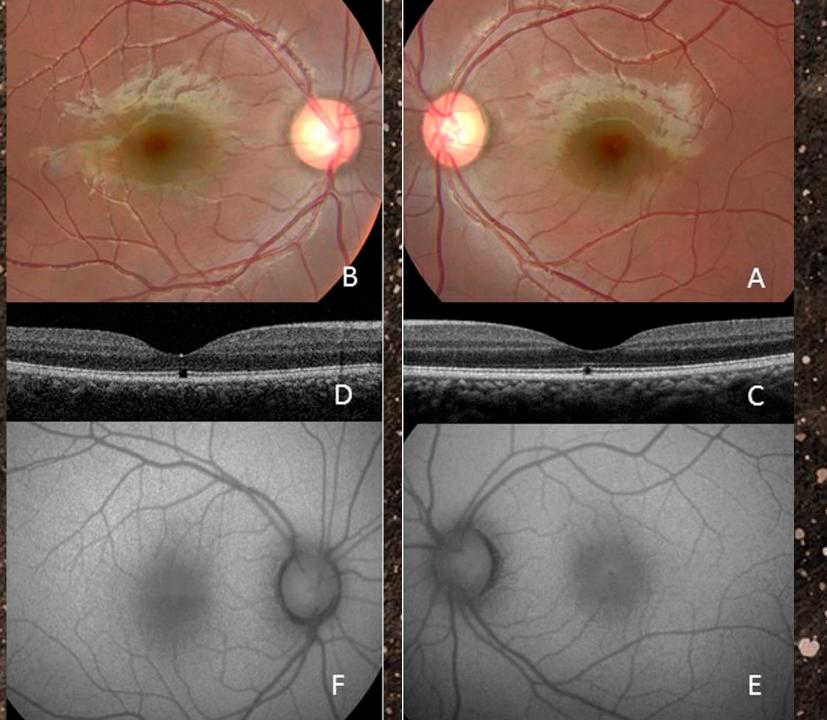


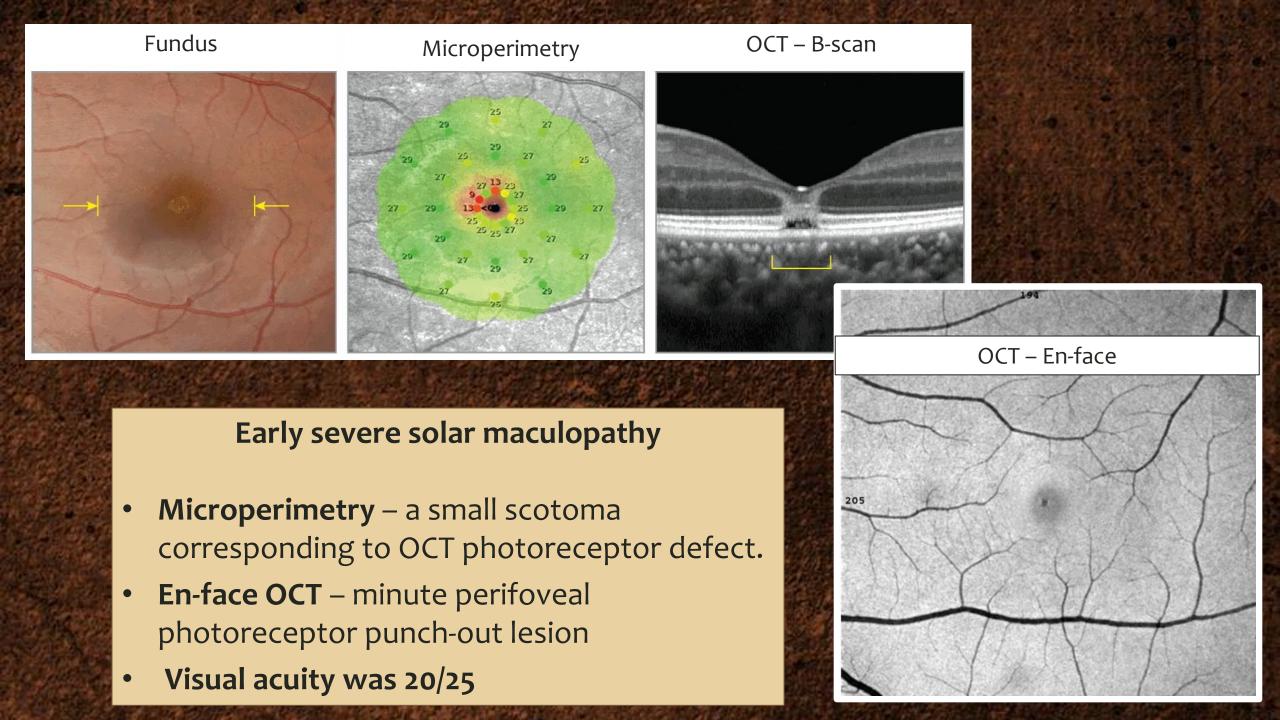
Typical findings of late-stage solar maculopathy

TOP: Essentially normal ophthalmoscopic appearance

MIDDLE: Rectangular photoreceptor defect in fovea on OCT

BOTTOM: FAF hypofluorescent spot corresponding to OCT photoreceptor defect



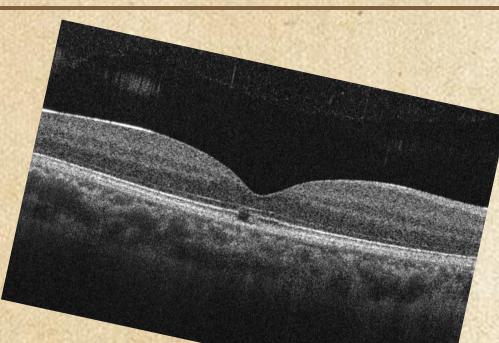


#### Assessment

Solar maculopathy secondary to sun gazing

## Management

- No specific treatment indicated
- Patient education
- Routine eye care



# Sun. Moon. You!

ECI

-02

APRIL 8:2024 ECLIPSE

https://eclipse.aas.org

A AMERICAN ASTRONOMICAL SOCIETY

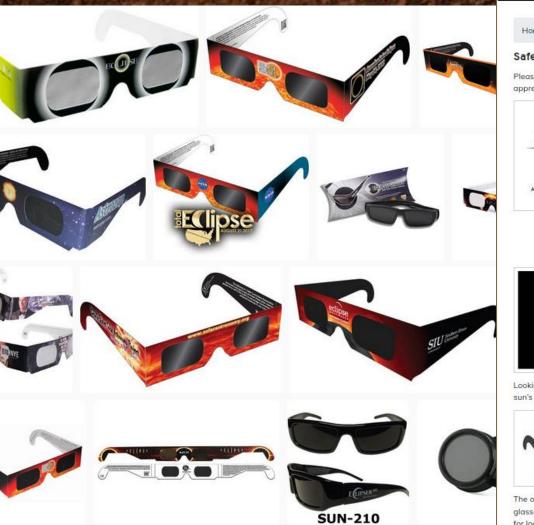
ANNULAR 50











Home / Eclipse 101 / Safety

#### Safety

Please feel free to download maps, posters, fact sheet, safety bulletin and other materials for use in your communities and events. We appreciate it if you credit NASA.



#### How to View the 2017 Solar Eclipse Safely



Looking directly at the sun is unsafe except during the brief total phase of a solar eclipse ("totality"), when the moon entirely blocks the sun's bright face, which will happen only within the narrow path of totality.



The only safe way to look directly at the uneclipsed or partially eclipsed sun is through special-purpose solar filters, such as "eclipse glasses" (example shown at left) or hand-held solar viewers. Homemade filters or ordinary sunglasses, even very dark ones, are not safe for looking at the sun; they transmit thousands of times too much sunlight. Refer to the American Astronomical Society (AAS) Reputable







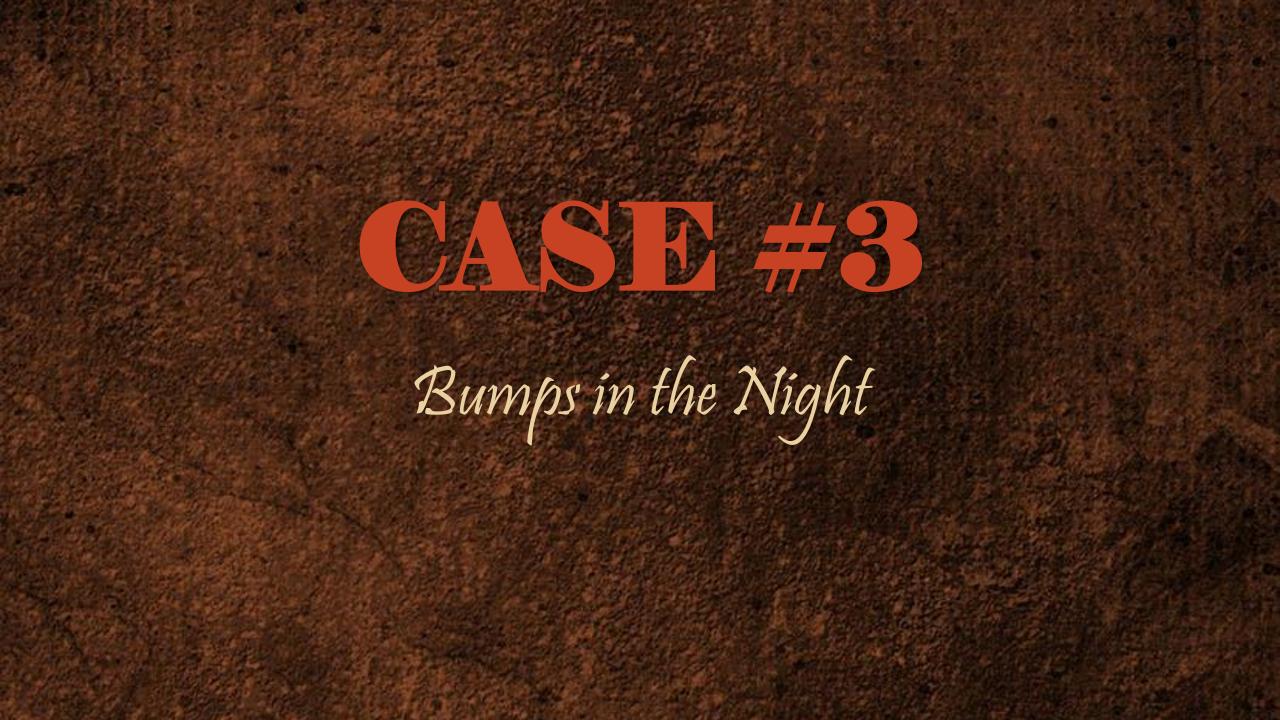
**TOTAL**SolarEclipse WHO? WHAT? WHERE? WHEN? and HOW?

NASA'sEyes ON THE 2017 ECLIPSE





 Solar Maculopathy -Mild visual acuity loss -Outer retinal OCT defect -History of sun gazing -No specific treatment -Patient education regarding safe solar eclipse viewing



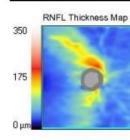
## CASE #3

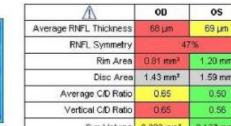
- 66yo WM presents for routine exam
- POH: S/P localized RD OS (3yrs). S/P ECCE OU (5yrs) LEE: 3yr.
- MH: T2DM x 6yrs (HbA1c: 9.4), OSA, HTN
- Vision: 20/20 OD, 20/25 OS
- Ta: 14/22 @10:00AM; PERRL, NO APD
- SLE: W&Q OU

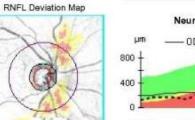


Name:			OD	OS
ID:		Exam Date:	7/6/2018	7/6/2018
DOB:	2/18/1952	Exam Time:	11:32 AM	11:33 AM
Gender:	Unknown	Serial Number:	4000-6813	4000-6813
Technician:	Operator, Cirrus	Signal Strength:	10/10	10/10

#### ONH and RNFL OU Analysis:Optic Disc Cube 200x200 OD O OS







200 -100

TEMP

85

72

51

54

77

117 88

84

SUP

57

58

NAS

Diversified: Distribution of Normals

NA 95% 5% 1%

RNFL Quadrants

RNFL

Clock Hours 70

74

63

73

TEMP

71

68

75 66

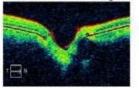
63 61

72

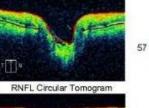
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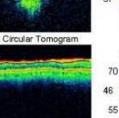
65

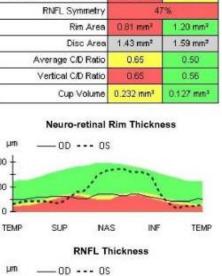
Disc Center(-0.09,-0.09)mm Extracted Horizontal Tomogram

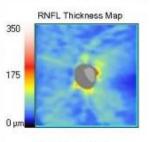


Extracted Vertical Tomogram



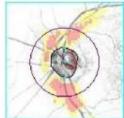




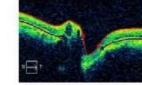


ZEISS

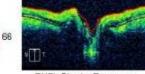
**RNFL Deviation Map** 



Disc Center(-0.06,0.00)mm Extracted Horizontal Tomogram

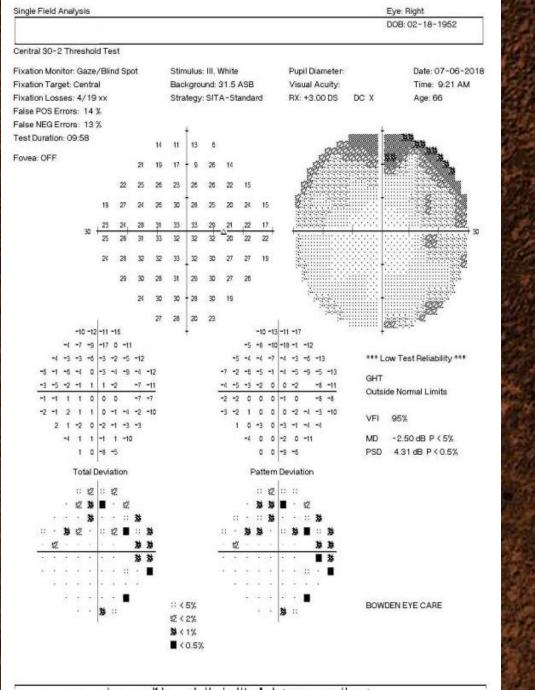


Extracted Vertical Tomogram



RNFL Circular Tomogram

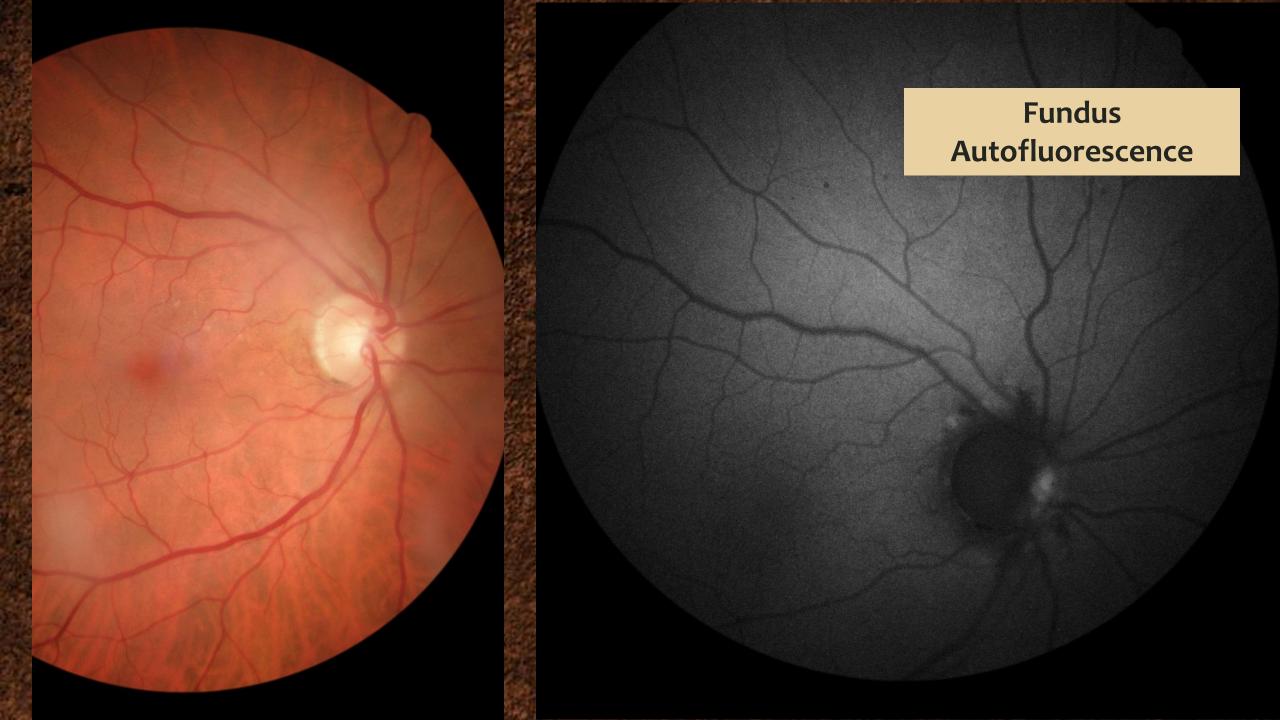
Name: ID: DOB: Gender: Technician:	2/18/1952 Unknown Operator, Cirrus	Exam Date: Exam Time: Serial Number: Signal Strength:	OD 4/25/2018 5:15 PM 4000-6813 8/10	OS 4/25/2018 5:15 PM 4000-6813 8/10		ZEISS
Gangli	on Cell OU A	nalysis: Macu	lar Cub	e 512x128	OD 🌒	o os
	OD Thickness Ma	ар		OS	Thickness Map	
			150	1		i i
K.					•	No. of Street
X			75 Ο μm			2
	Fovea: 238, 67	Contraster of the	o pan	F	ovea: 241, 66	121.2.1.1.1.1.1.1.1.1.1.1.1.1.1.1.1.1.1
00.0	eviation Map	OD Sectors		OS Sectors	OS Deviat	on Man
	Martin W N	69 72 76 70 71 75	Diversified: Distribution of Normals 95% 5% 1%	63 64 64 60 55 58		Pert High
1:00	a start	A		OD µm OS µm		1
	A THE R	Average GCL + IP	L Thickness	72 61	Salata and	-18
		Minimum GCL + IP	L Thickness	72 56	A Part Raine	
OD Horizon	ntal B-Scan	BScan: 67		Horizontal B-Scan	65	Scan: 66



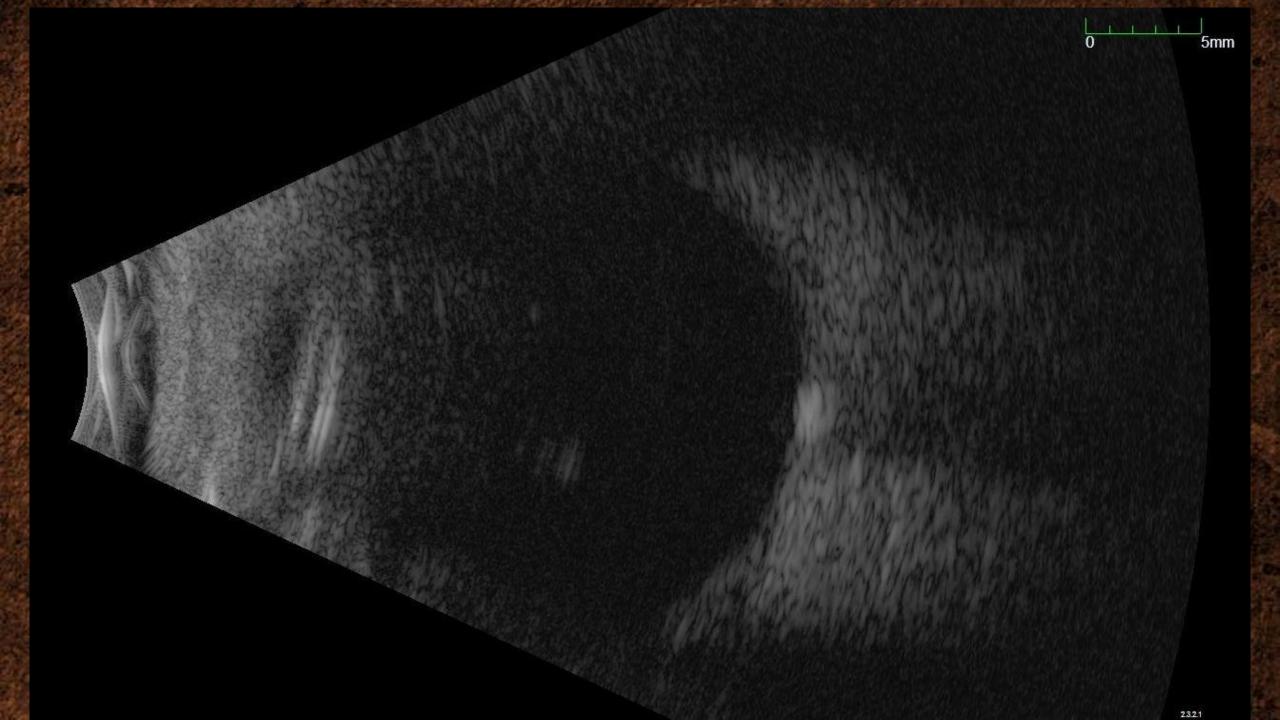
Fixation Monitor: Gaze/Blind Spot Stimulus: III, White Pupil Diameter: Fixation Target: Central Background: 31.5 ASB Visual Acuity: Fixation Losses: 1/23 Strategy: SITA-Standard RX: +1.50 DS DC X False POS Errors: 14 % False NEG Errors: 13 %	DOB: 02-18-1952 Date: 07-06-2011 Time: 9:34 AM Age: 66
Fixation Target: Central     Background: 31.5 ASB     Visual Acuity:       Fixation Losses: 1/23     Strategy: SITA-Standard     RX: +1.50 DS     DC X       False POS Errors: 14 %     False NEG Errors: 13 %     Fest Duration: 14:07     Fest Duration: 14:07	Time: 9:34 AM
Fixation Target: Central         Background: 31.5 ASB         Visual Acuity:           Fixation Losses: 1/23         Strategy: SITA-Standard         RX: +1.50 DS         DC X           False POS Errors: 14 %         False NEG Errors: 13 %         False NEG Errors: 13 %         False NEG Errors: 14.07         False NEG Errors: 14.0	Time: 9:34 AM
Fixation Losses: 1/23 Strategy: SITA-Standard RX: +1.50 DS DC X False POS Errors: 14 % False NEG Errors: 13 % fest Duration: 14:07	
False POS Errors: 14 %	Age: 66
ralse NEG Errors: 13 %	
Fest Duration: 14:07	and the second s
Test Duration: 14:07 2 6 0 0 00000000	
Fovea: OFF	
(0 6 13 - 22 19 11	
12 20 9 11 22 28 25 2	
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فسمع لعربته وفقاؤه الماقة كسن الشريسة كالرجا كالتربي والساؤب العالي والمراجع والمستريب المستحد والمسترك والمراجع والم

الفارحية أستعمل ويجازها فالبناء والماعف أوروا للمرافع حريان أربي مريون أربيت وحريب ويعتر والمراجع







## What is going on here?



#### https://app.tophat.com/e/777538

Glaucoma

Papilledema

Foster-Kennedy Syndrome

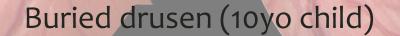
**Optic Nerve Head Drusen** 

Ischemic Optic Neuropathy

Glaucoma	Abnormal <u>IOP asymmetry</u> (14/22). Large optic cups with <u>rim area</u> < 1 mm². Characteristic <u>VF defects</u>
Papilledema	Bilateral <u>disc edema</u> secondary to increased intracranial pressure.
Foster-Kennedy Syndrome	<b>Unilateral papilledema</b> with optic atrophy in fellow eye. Associated with sphenoidal ridge meningiomas.
Optic Nerve Head Drusen	<u>Calcific deposits</u> in the prelaminar optic nerve head. Causes scalloping and obscuration of the disc margin without edema. Will <u>hyperfluoresce</u> on FAF
Ischemic Optic Neuropathy	Acute onset <b>pallor and edema</b> of ONH associated with APD, color vision loss, and nerve fiber bundle VF defects. <b>Disc-at-risk</b> . Systemic risk factors: <b>DM, OSA</b>

### **Optic Nerve Head Drusen**

- Acellular calcific deposits of the ONH; 66%-80% bilateral
- Idiopathic; Familial clustering suggests genetic factors
- Early childhood onset of lesions deep in ONH ("buried drusen")
- Lesions usually increase in visibility and size over time
- 25%-75% of adults have VF defect (enlarged BS, arcuate)
- Visual acuity rarely affected



#### Partially exposed drusen (12yo child)

#### Superficial ONH drusen

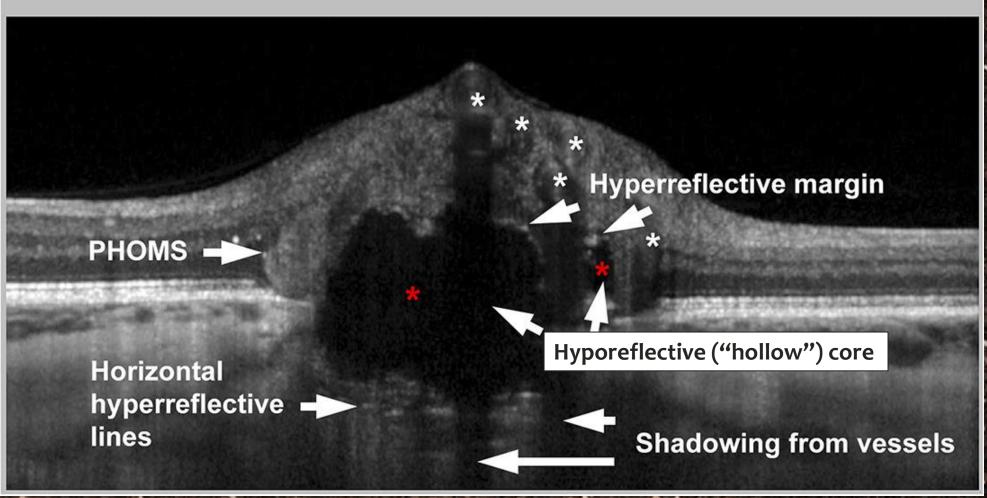
#### Superficial ONH drusen

#### Absence of FAF signal in a pt with deep buried ONH drusen FAF is more likely to detect ONH drusen that are larger and more superficial

Δ

Loft, J Neuroophth, 2019

- ODD are always located above lamina cribrosa
- ODD always have a signal-poor core
- ODD are often seen with a hyperreflective margin, most prominent superiorly
- ODD are sometimes seen as conglomerates of smaller ODD with internal reflectivity within the signalpoor core
- Hyperreflective horizontal lines might represent early ODD but should not be diagnosed as ODD
- Peripapillary hyperreflective ovoid mass-like structures (PHOMS) should not be diagnosed as ODD



#### **ODD on EDI-OCT**

ODD are "hollow" structures with a hyperreflective margin ("capsule")

Internal reflectivity represents the fusion of multiple ODD

Hyperreflective horizontal lines might be early ODD

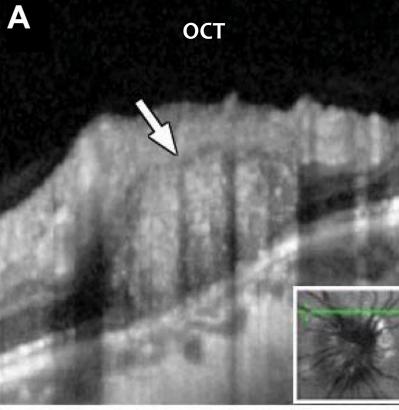
PHOMS circumscribe the disc and are not ODD.

White asterisks: vessels. Red asterisks: ODD.

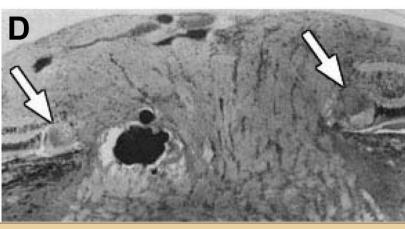
ODD: optic disc drusen

**PHOMS:** peripapillary hyperreflective ovoid mass-like structures.

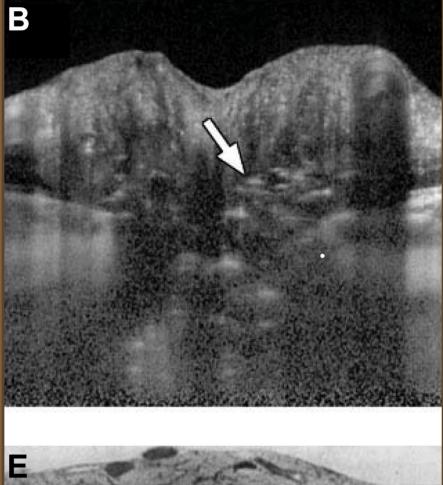
Malmqvist, 2017

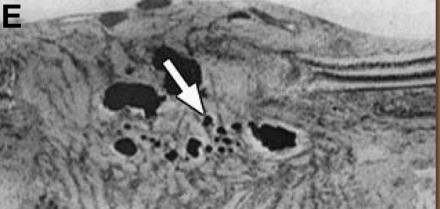


HISTOLOGY



Peripapillary waste (not hollow)





Small drusen (horizontal lines)





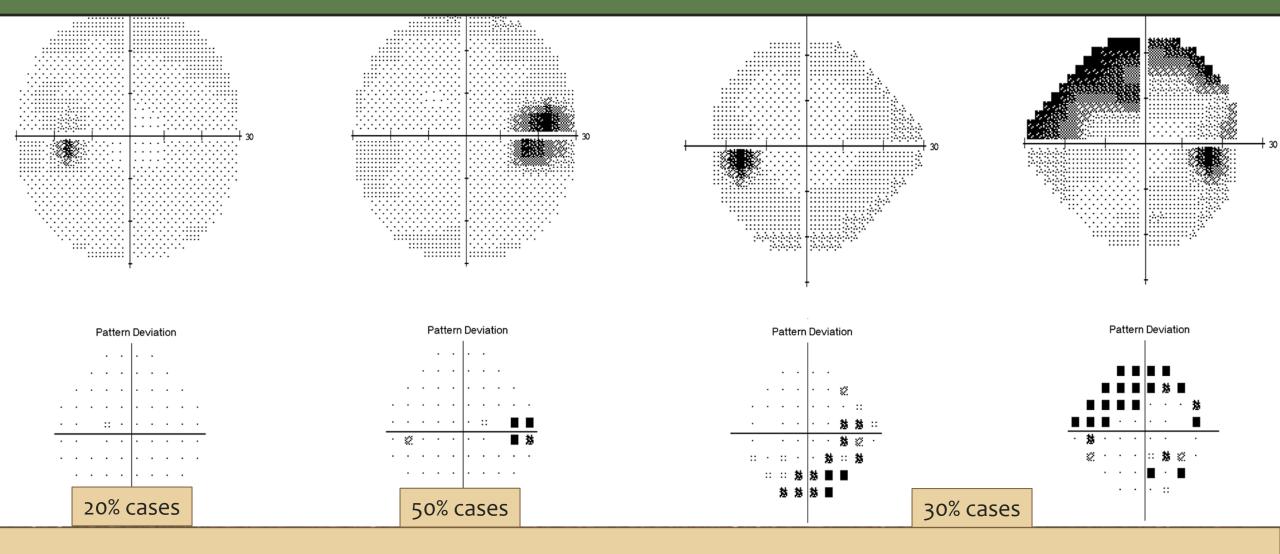
Confluent drusen (septa)

Ultrasonography has high sensitivity (90%) and specificity (80%) in diagnosis of ONH drusen

Represents "gold standard" for diagnosis

Carter, 2014

### **VF Defects with ONH Drusen**



Normal

Enlarged Blind Spot

Early Arcuate

Adv. Arcuate

## **VF Defects with ONH Drusen**

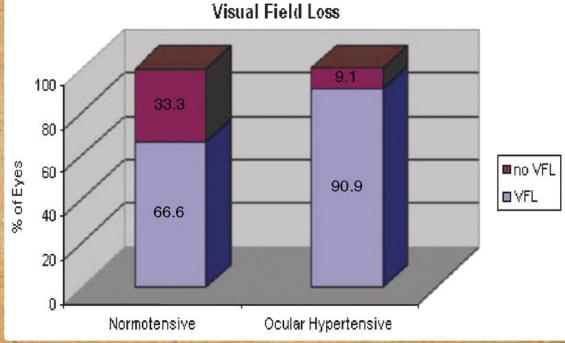
- Over 80% of adult cases will have VF defects
- Enlarged BS, arcuate defects and constricted VF are most common
- VF loss tends to progress slowly over time
- Eyes with larger and more superficial drusen tend to have greater VF damage
- Arcuate VF defects have associated RNFL thinning
- Visual acuity and color vision usually remain normal
- No effective treatment for ONH drusen

### **ONH Drusen & Glaucoma**

- Challenge of managing glaucoma in patients with concurrent ONH drusen
- Eyes with ONH drusen are more susceptible to glaucomatous VF loss

  Visual Field Log

 IOP-lowering treatment should be considered in all patients with ONH drusen and elevated IOP
 Grippo, 2008





#### **Glaucoma with ONH Drusen**

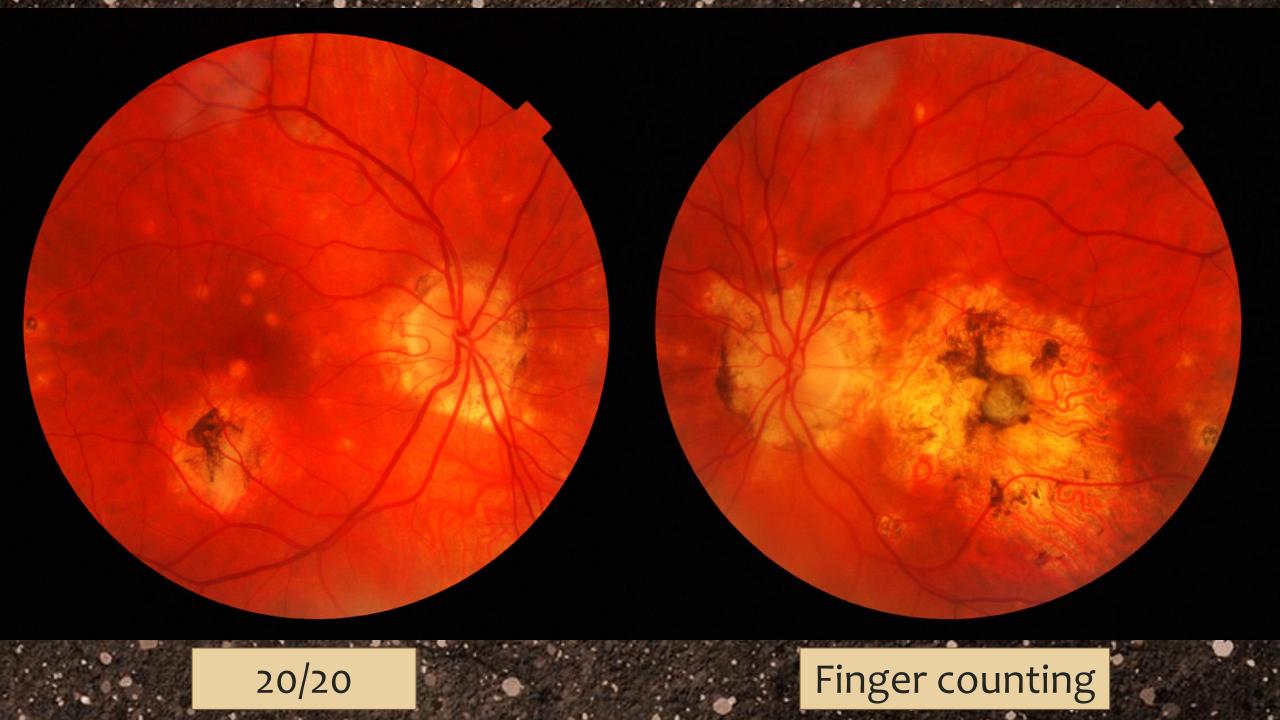
- Perform OCT and FAF to confirm presence of ONH drusen
- Obtain ultrasound in difficult cases
- VF loss is common and can mimic glaucomatous loss
- Patients with ONH drusen and elevated IOP are more susceptible to VF loss and may benefit from IOP lowering therapy



## The bird man cometh

## Case #4

- 56yo WM presents for routine eye exam
- Occupation: Maintenance man and farmer
- POH: H/O vision loss OS due to "bleeding" 5 years ago. Treated with laser
- MH: NIDDM x 4yrs (HbA1c: 6.5)
- BVA: 20/20 OD, FC @ 10ft OS
- Pupils and motility: Normal
- IOP: 14/16 mmHg @ 2pm
- External: Normal OU



#### Assessment

- POHS OU
- Histoplasmic maculopathy OS with severe vision loss
- Evidence of prior reactivation OD
- No diabetic eye disease

## What is the plan?



#### https://app.tophat.com/e/777538

Referral for laser photocoagulation

**Referral for Avastin injection** 

Mask or ventilator use at work

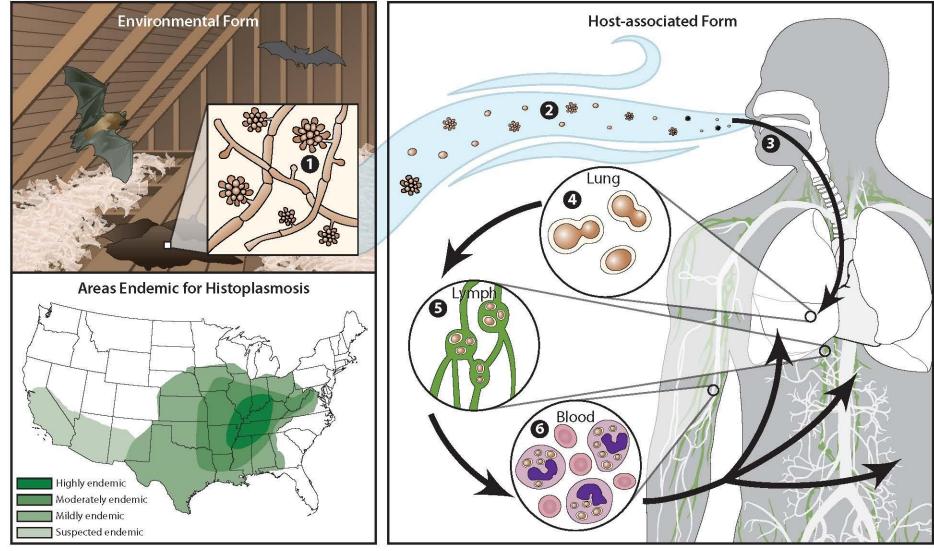
Referral for low vision care

Routine annual eye exams

## Management

- Recommend use of protective devices because pt is frequently exposed to soil and bird droppings as a farmer and maintenance man.
- Daily Amsler grid and environmental Amsler
- Safety issues for monocular patients
- Patient education diabetic eye disease.
- RTC 6 months, sooner PRN

#### **Biology of Histoplasmosis**



In the environment, *Histoplasm capsulatum* exists as a mold (1) with aerial hyphae. The hyphae produce macroconidia and microconidia (2) spores that are aerosolized and dispersed. Microconidia are inhaled into the lungs by a susceptible host (3). The warmer temperature inside the host signals a transformation to an oval, budding yeast (4). The yeast are phagocytized by immune cells and transported to regional lymph nodes (5). From there they travel in the blood to other parts of the body (6).



## **POHS and Vision Loss**

- Vision loss occurs secondary to exudative maculopathy caused by CNV &/or inflammation
- Reactivation of histo spots in the macular region is believed to play a role in triggering maculopathy
- Patients with perimacular histo spots, especially near the fovea, are at risk for vision loss

5 years



Perimacular histo spot



Active maculopathy

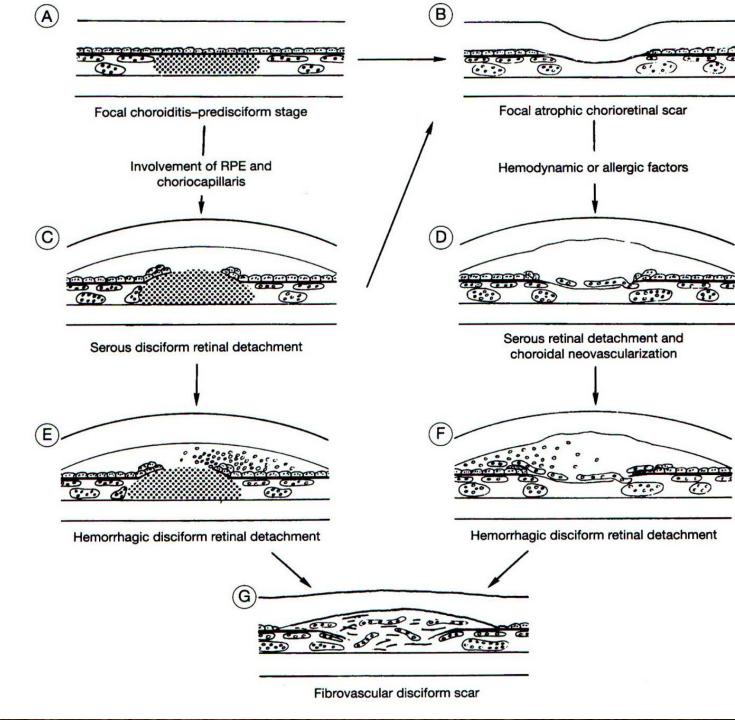
#### Inflammatory maculopathy pathway

Focal choroiditis

Serous detachment

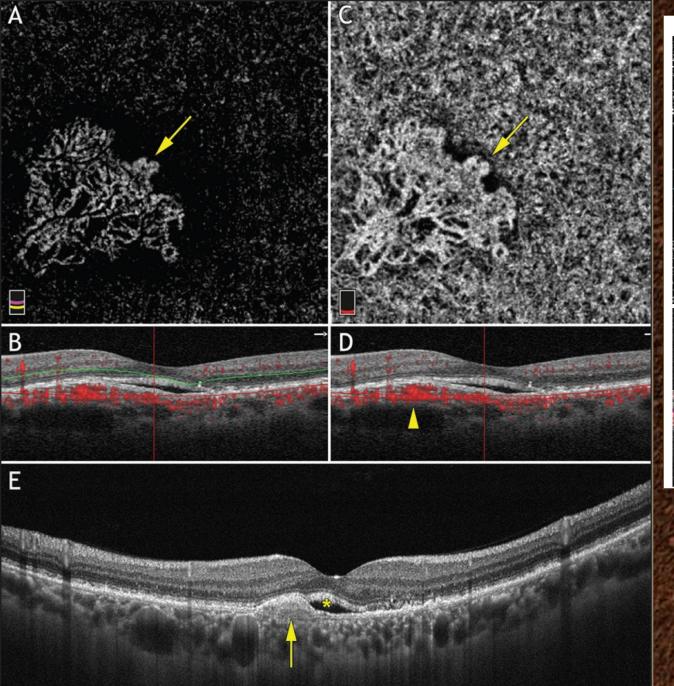
Subretinal hemorrhage

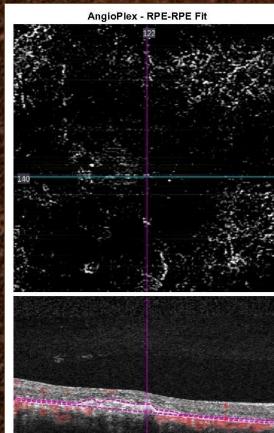
Scar



# Neovascular maculopathy pathway Histo spot CNV Subretinal hemorrhage

Scar





T

# Structure - RPE-RPE Fit

Overlays Structure - None AngioPlex - None

OCT angiography can differentiate neovascular from inflammatory maculopathy in patients with POHS

## **Preventing Reactivation of POHS**

- Re-exposure to histoplasmosis may play a role in reactivating retinal lesions and promoting development of maculopathy
- Avoid high risk areas where histoplasmosis levels tend to be highest
  - Caves, chicken coops, old buildings
- Protect yourself or avoid high risk activities
  - Construction and demolition, working with poultry, HVAC installation or service, farming, gardening

## **Preventing Reactivation of POHS**

- Personal protective equipment
  - Masks and respirators
- Dust control
  - High efficiency air filters
  - Vacuum cleaning
  - Wetting contaminated soil
- Endogenous factors

   Chronic fungal infections
   LASIK



- Patients with histo spots near the central macula are at risk for vision loss due to maculopathy
- OCTA can differentiate neovascular from inflammatory maculopathy
- Take steps to decrease the risk of histo spot reactivation in at-risk patients





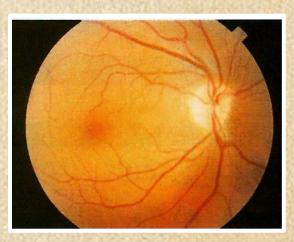
# Hear no evil, see no evil!

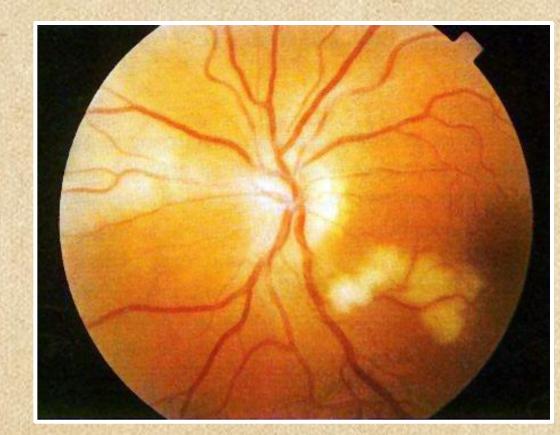
# Visit 1 (1993)

- 36yo WF presents with c/o "gray haze" OS x 2 weeks
  - 1 week prior had suffered left-sided HA and photopsias OS that sent her to the ER. Exam was normal and Fiorinal with codeine was prescribed
- POH: Normal.
- MH: Head and neck aches and parestheias in her arms following MVA x 2yrs
- FOH: Father with glaucoma

# Visit 1

- Vision 20/20 in each eye
- Pupils: Normal
- Motility: Normal
- IOP: 13 mmHg OU
- External: Normal





#### Assessment

- Acute BRAO OS
- Evidence of older resolving BRAO OS

### Management

- Carotid duplex scan Normal
- Echocardiography Normal
- Labs: CBC, rheumatoid factor, RPR, fasting glucose, ANA all normal
- Start ASA
- Retinal lesions resolved @ 1 month F/U visit

# Visit 2 – 3yrs Later

- C/O constant "flash bulb glare" OS x 1 day
- Vision: 20/20 OD, 20/25 OS
- Pupils normal, Motility normal, IOP 14/12 mmHg
- External: Normal OU
- DFE: Normal OD, BRAO of superior-temporal artery OS
  - No visible embolus
  - Occlusion does not appear to occur at bifurcation

#### Assessment

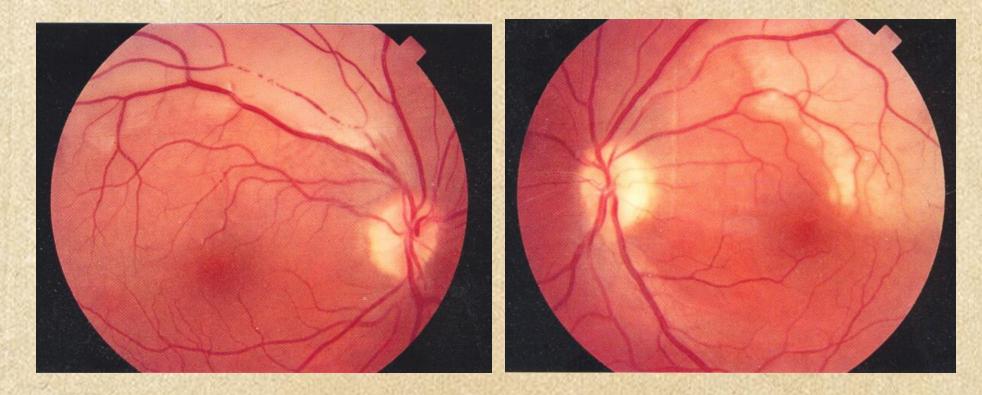
• BRAO OS

#### Management

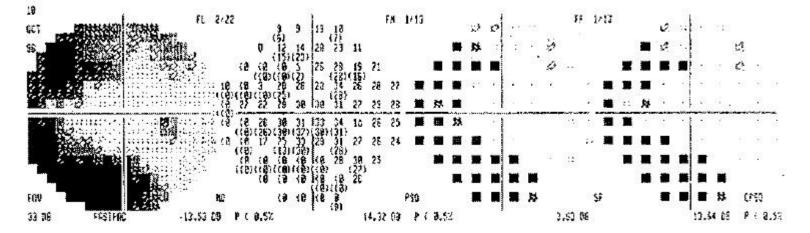
- Retinal consult Diagnosis of "idiopathic recurrent BRAO." No known cause or tx for condition
- IM consult Obese (238lbs) but otherwise in good health. No BCPs, no vasculitis, synovitis, diabetes, or HTN
- Continue ASA, start low fat diet

# Visit 3

- Presented 2 weeks later with photopsia OD
- Examination was remarkable for the presence of new BRAO OD

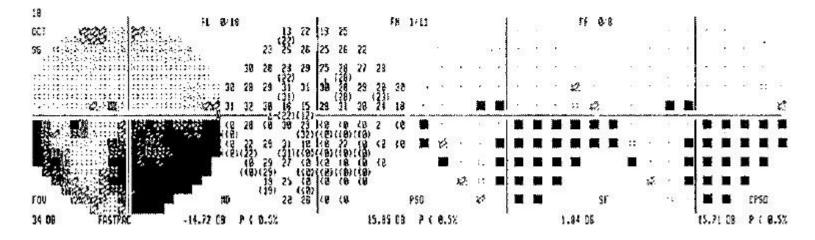


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RGE	48	STERNER	f Øçi,	1HRESHOLD



LEFT

REE 48 STRATECY FULL INRESHOLD



#### Assessment

Idiopathic recurrent BRAO, OU

#### Management

- IM consult: Negative evaluation except mildly elevated ESR (27 mm/h, normal: 0-20)
- Rheumatology consult: Negative evaluation. Normal temporal artery biopsy
- Audiometry and otolaryngology consult: moderately severe sensorineural hearing loss. Referred for hearing aid fitting

## What is going on here?



#### https://app.tophat.com/e/777538

Multiple sclerosis

Systemic lupus erythematosis

Sarcoid

Susac's syndrome

Lyme disease

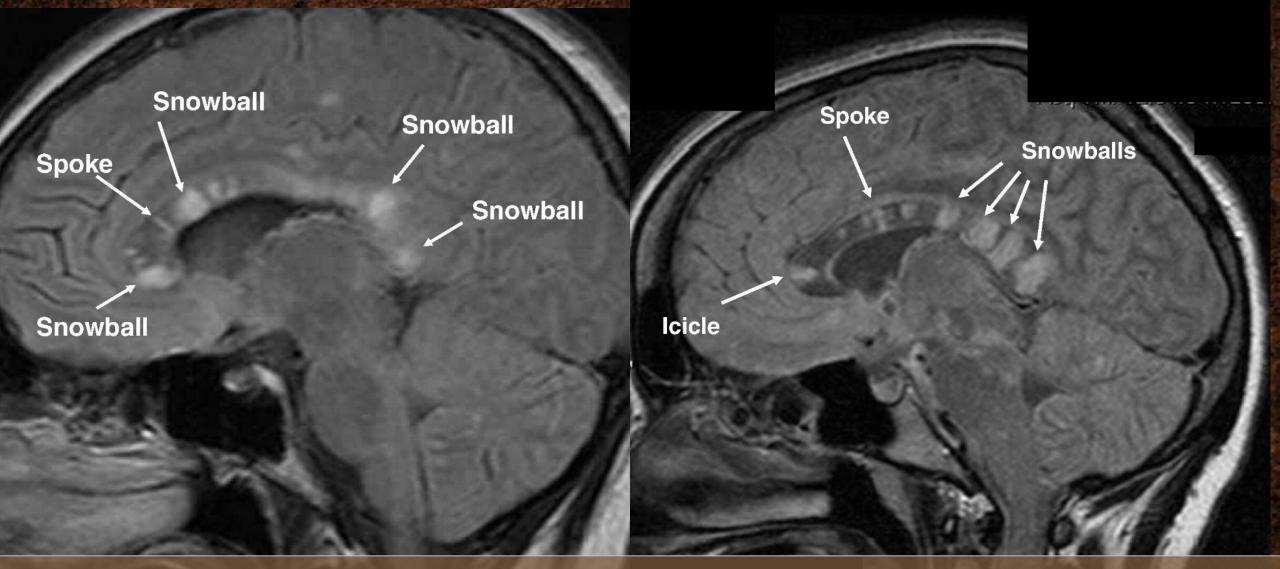
Multiple sclerosis	Uveitis and retinal phlebitis are common posterior segment manifestations of MS. <b>BRAO is not associated with MS</b>	
Systemic lupus erythematosis	Retinopathy may include CWS, <b>BRAO</b> , neovascularization and VH. Diagnosis made on clinical and lab grounds.	
Sarcoid	Antiphospholipid antibody syndrome may be associated with sarcoidosis and can lead to <b>retinal artery occlusion</b>	
Susac's syndrome	Clinical triad of <b>encephalopathy, BRAO and hearing loss</b>	
Lyme disease	<b>BRAO</b> has been reported as an uncommon complication of ocular Lyme borreliosis. Negative RPR test	

# **BRAO** in the Young

Emboli	Cardiac disease, IV drug abuse	
Thrombosis	Pregnancy, BCP use, Coagulopathy	
Arteritis	Lupus, Lyme Susac's Syndrome	
Arterial spasm	Migraine, Drug abuse (cocaine, meth)	
Vascular compromise	Orbital, optic nerve, retinal disease; Trauma	

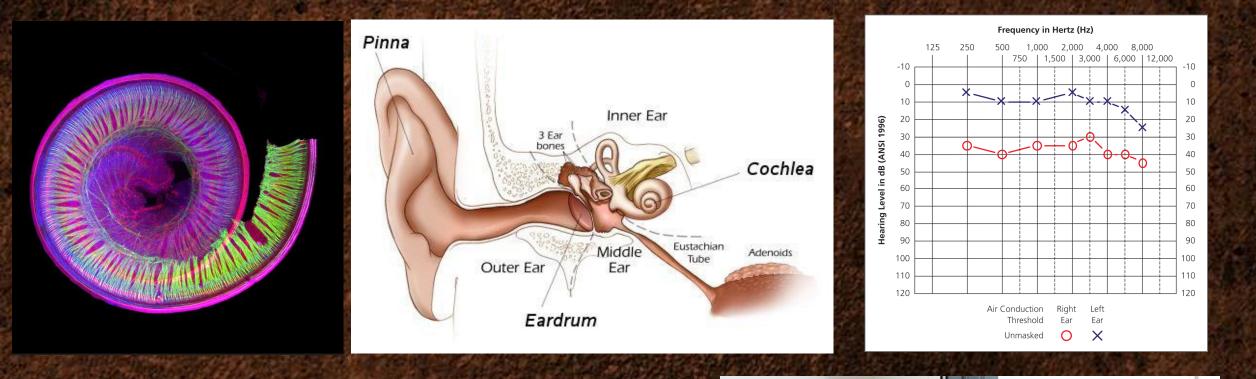
#### **Susac's Syndrome**

- Clinical triad of (1) encepholopathy, (2) BRAO, and (3) hearing loss that typically occurs in young adult women
- First described by Susac in 1994
- Immune mediated microangiopathy affecting blood vessels in the retina, chochlea and brain
- MRI findings of lesions in the corpus callosum
- Treatment with steroids and immune suppressants can slow progression of the disease



MRI findings in the corpus callosum of patients with Susac's syndrome

Rennebohm R. J Neurol Sci. 2010;299:86-91

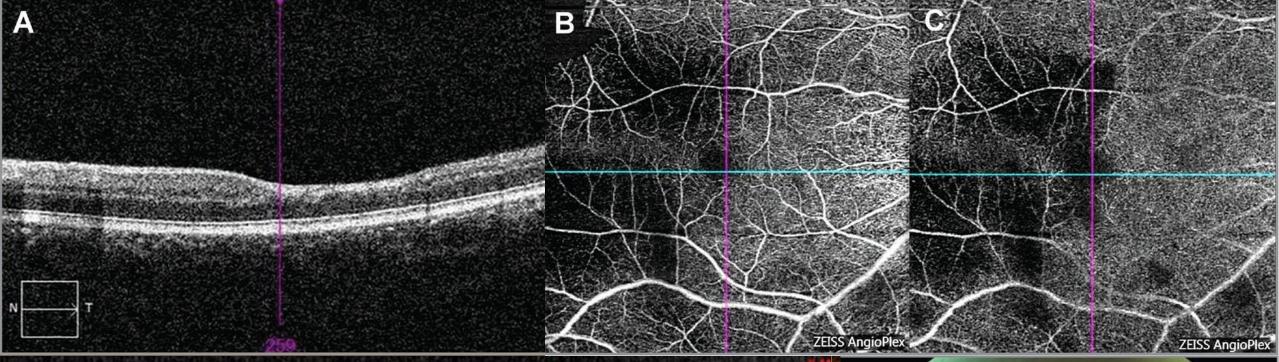


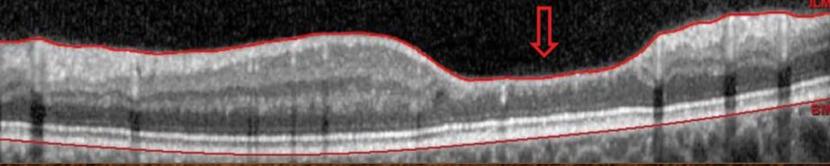
Damage to the cochlea (inner ear) results in sensorineural hearing loss, and is a key diagnostic finding in Susac's syndrome

Egan RA. J Neurol Sci. 2010;299:97-100

Acute retinal findings include arterial wall hyperfluorescence on FA and "Gass plaques" – yellow deposits believed to represent focal damage to the arterial wall

Egan RA. J Neurol Sci. 2010;299:97-100





Late retinal findings include arterial attenuation, inner retinal atrophy and regions of nonperfusion

Egan RA. J Neurol Sci. 2010;299:97-100



- BRAO in the young is less likely to be embolic
  - Look for coagulopathies and inflammatory disease
- Susac's syndrome is one cause of recurrent BRAO in the young
  - Check for hearing loss and MRI lesions



# Glaucoma Plus!

### CASE #6

- 57yo HM presents c/o distance blur x 2yrs
- POH: Diagnosed with glaucoma at age 20.
  - LEE: 10yrs ago
  - S/P unspecified glaucoma laser procedure 15yrs ago.
  - Glaucoma is not currently treated
- MH: HTN, Depression, Anxiety, OCD

### CASE #6

#### Vcc

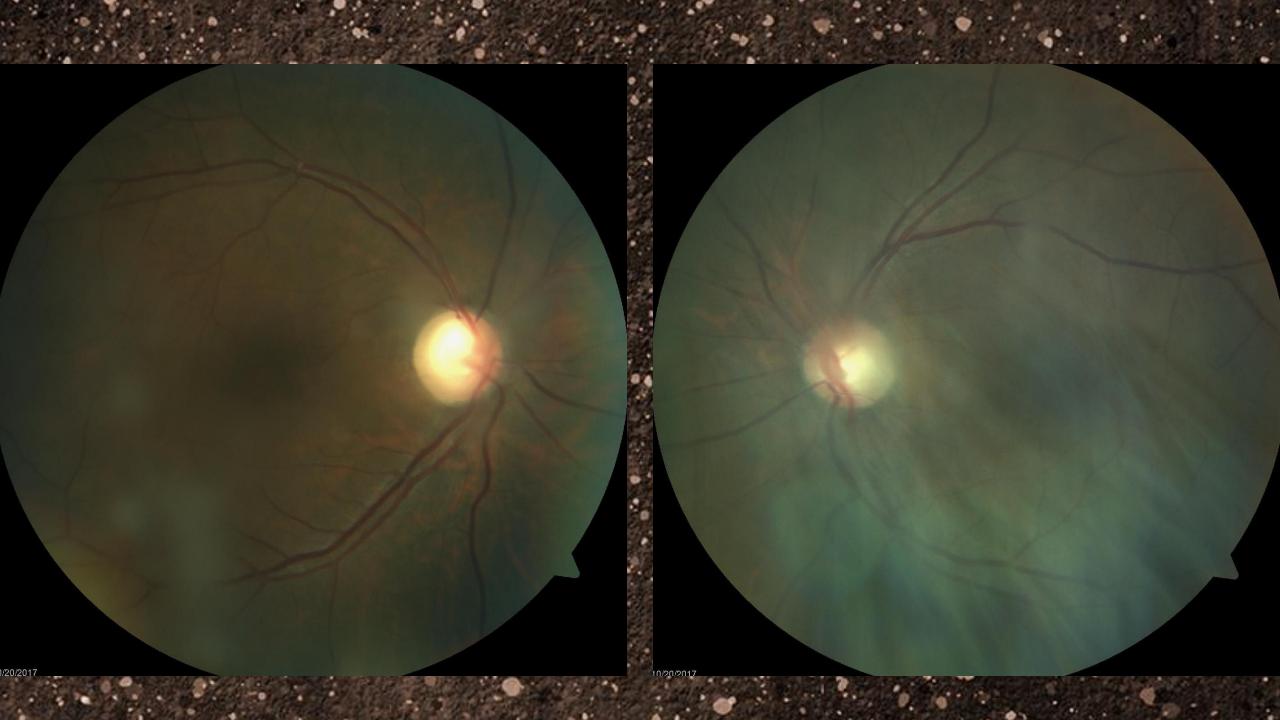
- OD: 20/40
- OS: 20/150

Ta 28/27 @ 2:00PM PERRL, (-) APD FCCF: Constricted OU Color: 2/0 (HRR)

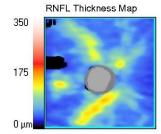
#### SLE

- W&Q OU
- Patent LPI OD
- Closed LPI OS

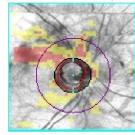
Gonio: D4or OU Pachs: 616/611



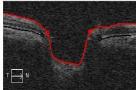
#### ONH and RNFL OU Analysis:Optic Disc Cube 200x200 OD O OS



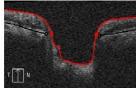




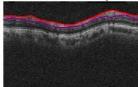
Disc Center(0.00,-0.36)mm Extracted Horizontal Tomogram



Extracted Vertical Tomogram

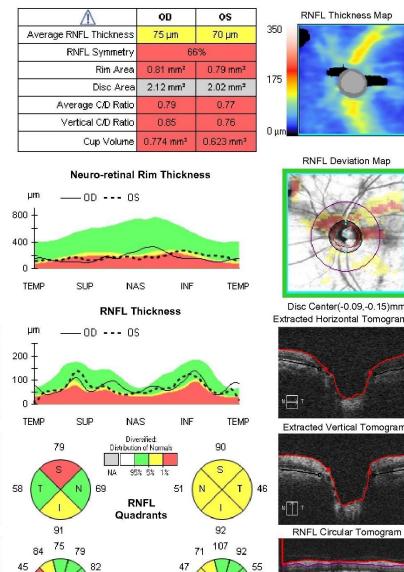






78

97 113 64

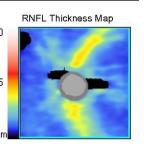


RNFL

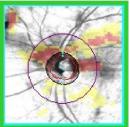
Clock Hours 46

59

75 117 85



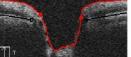
**RNFL** Deviation Map



Disc Center(-0.09,-0.15)mm Extracted Horizontal Tomogram

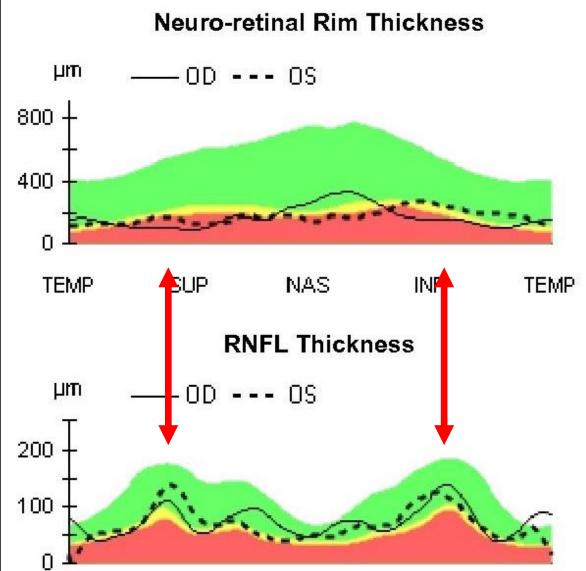


Extracted Vertical Tomogram



TEMP

SUP



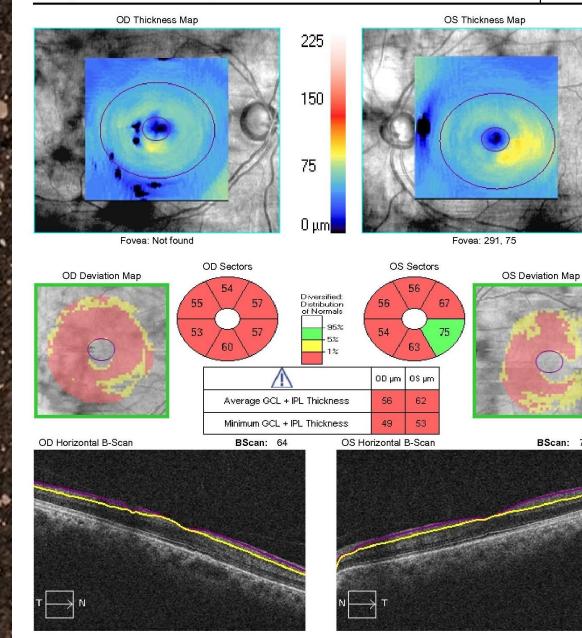
NAS

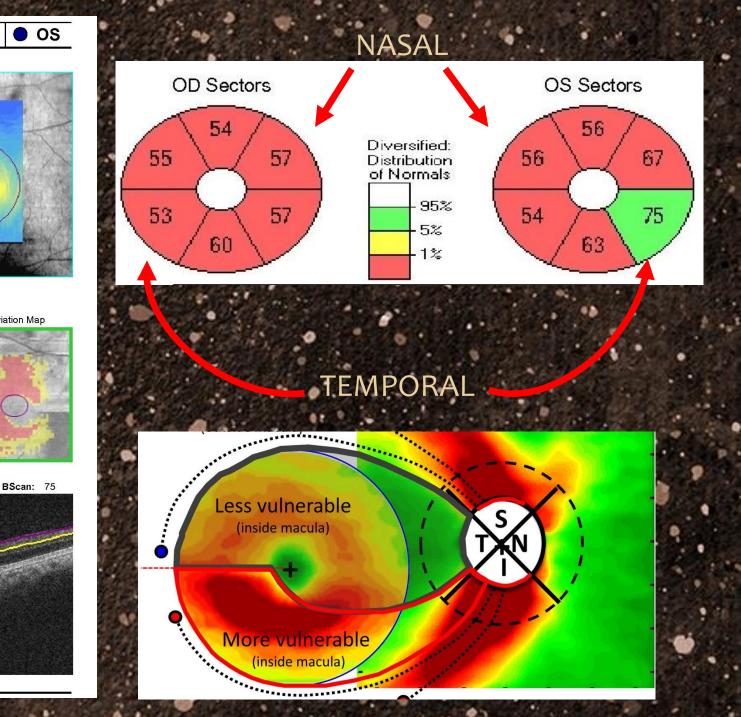
INF

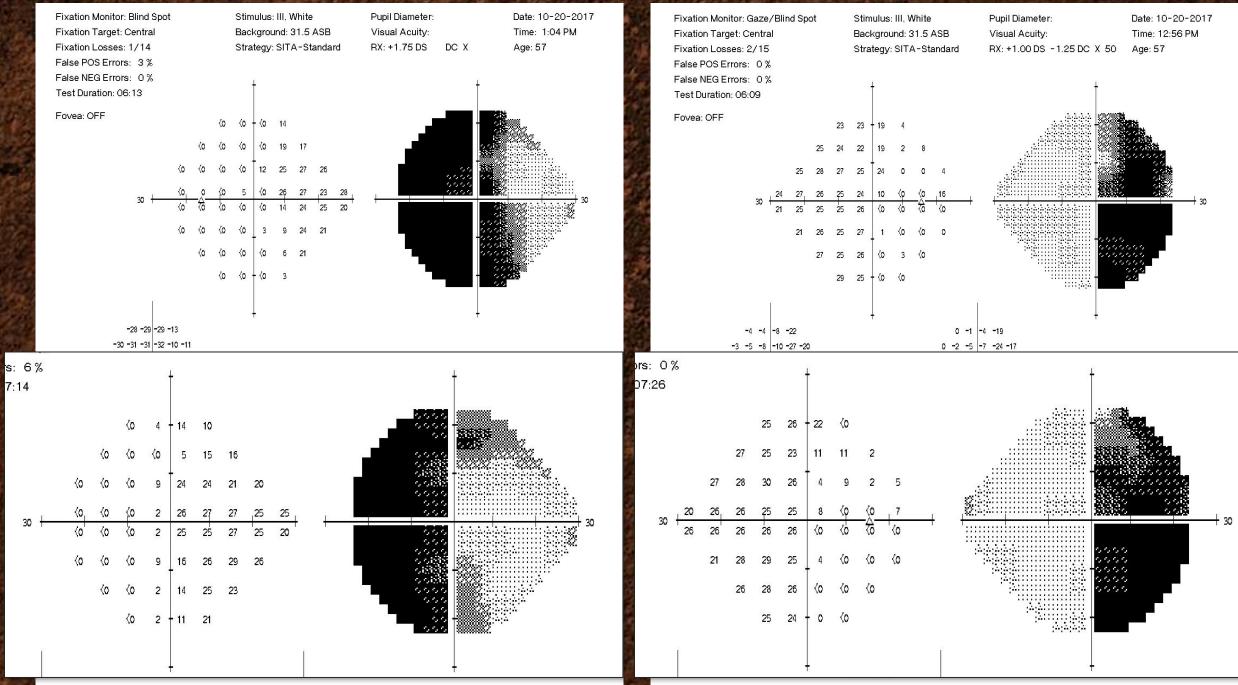
TEMP

#### Ganglion Cell OU Analysis: Macular Cube 512x128

OD 🔵





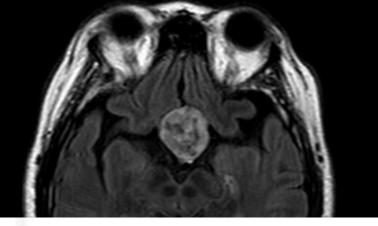


### Now what?



#### https://app.tophat.com/e/777538 Send patient to ER stat Α Refer patient to ophthalmology B Order MRI of head С Call patient's PCP D Prescribe PGA and RTC in 4 weeks Ε

W: 1805 - L: 1039 A# 33880217 SE: 501 IM: 15 3042.81064224243



Acq Tm: 9:11 PM Pat Pos: HFS

10cm

#### TE: 1 TR: 1 |

EC: 1 4 Thk

SP

Sorna

A# 3

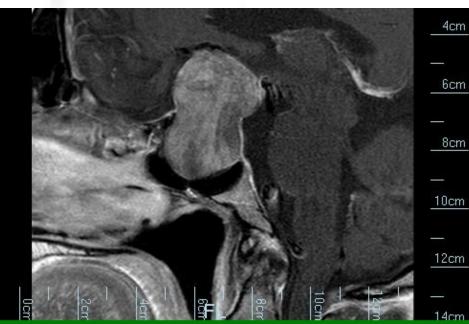
AF

IMPRESSION: A 2.6 x 3 x 4.4 cm T1 isointense, T2 iso/hyperintense, heterogeneously enhancing sellar lesion with suprasellar extension, mass effect on the optic chiasm and less than 50% encasement of bilateral cavernous ICAs, compatible with macroadenoma.

SE: 401 IM: 7 -3053.6687332479

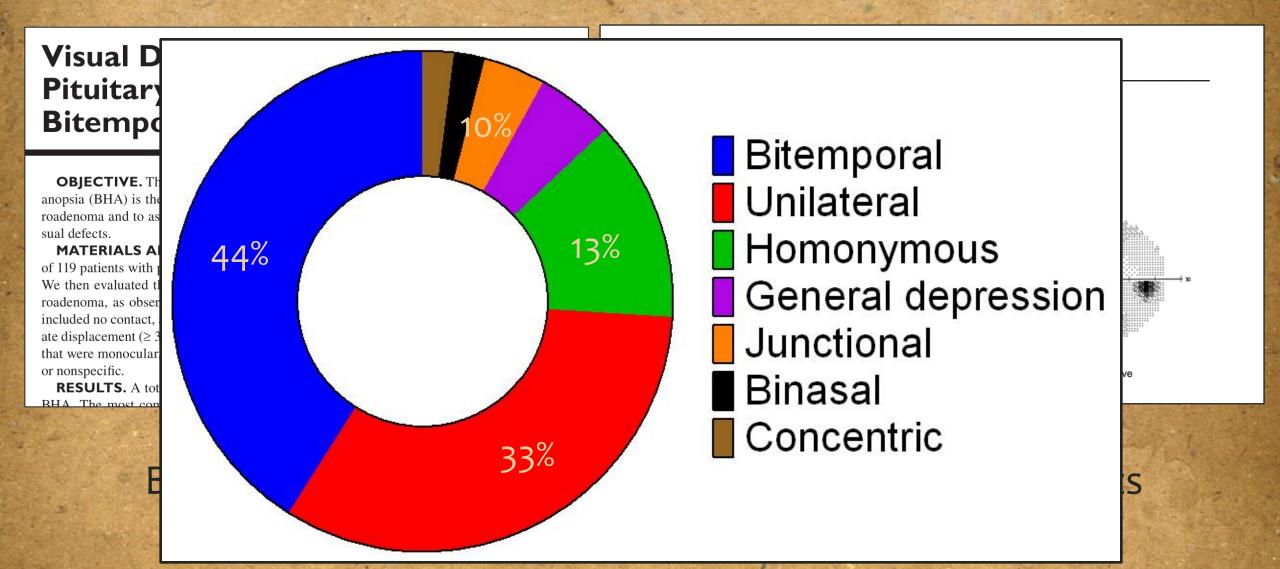
TE: 8.2 ET: 3 TR: 460.651092529296 EC: 1 3 Thk 3.5 SP FC

Sorna Corporation

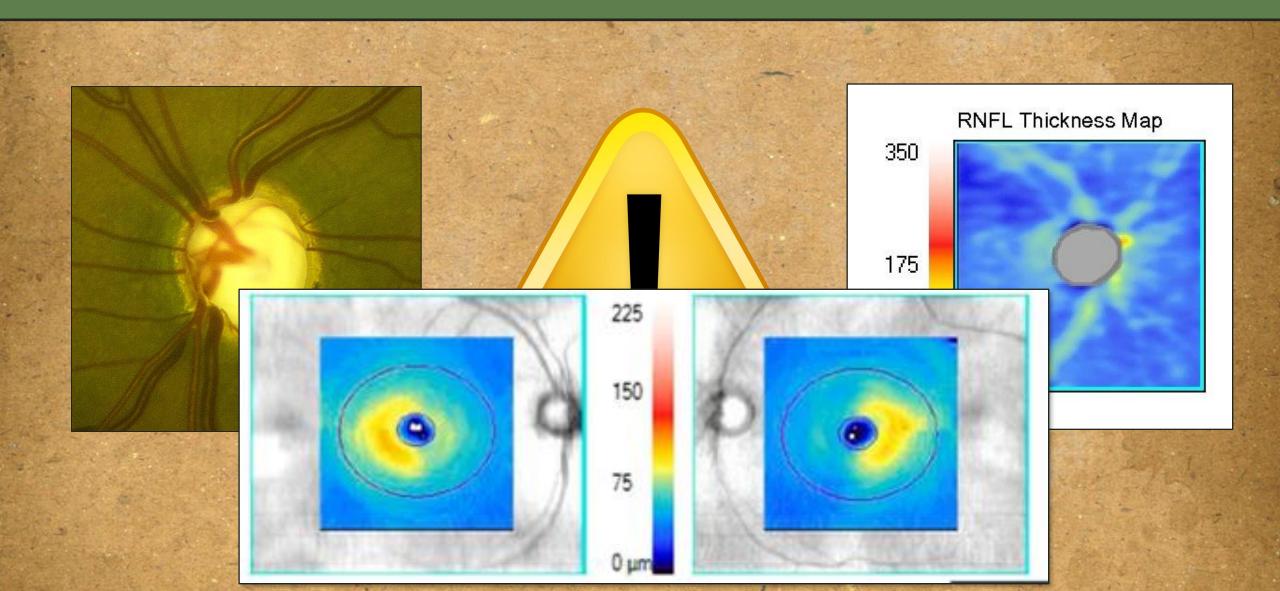


EAR

#### **Pituitary Adenoma Visual Field Defects**



#### Pituitary Adenomas + Glaucoma



Take Home Message

- Glaucoma + Pituitary Adenoma
  - Chiasmal compression is an important cause of non-glaucomatous cupping
  - Binasal ganglion cell loss is a sensitive early indicator of chiasmal compression
  - Beware of glaucoma suspects with atypical findings
  - Patients can have more than one active disease process

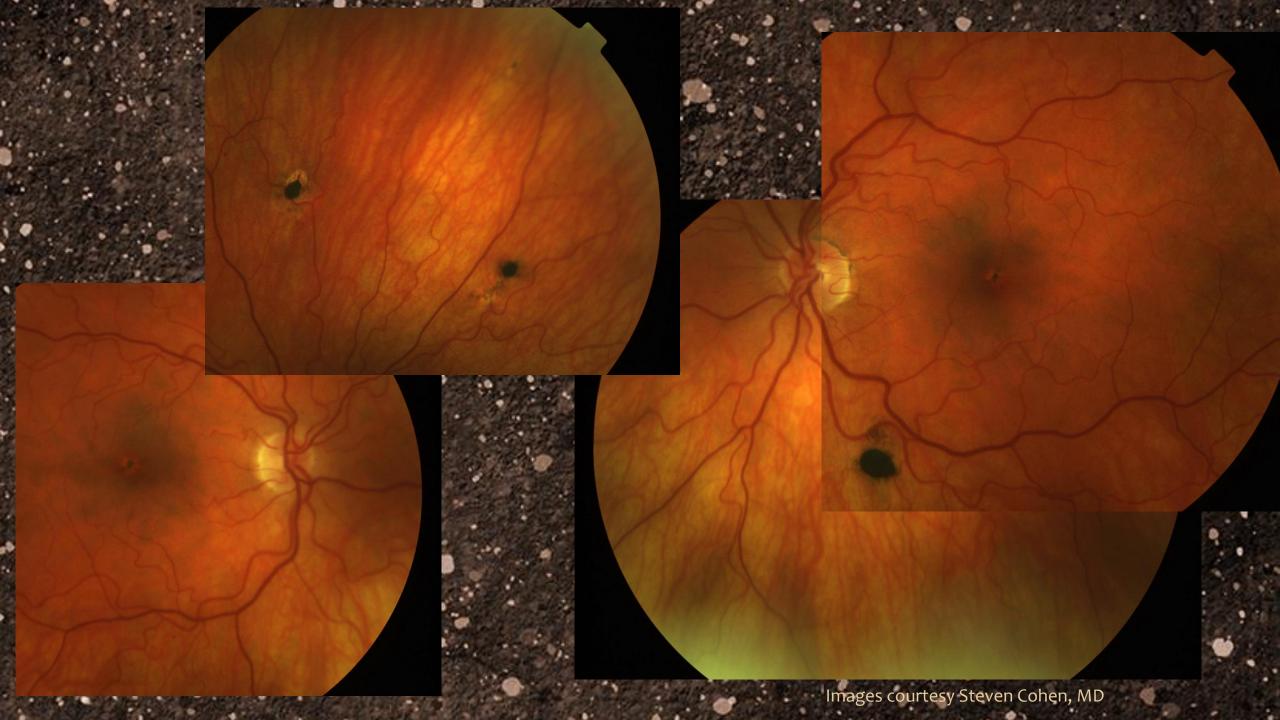




# The spots are revealing!

#### **Case #7**

- Parents bring 7yo child in for his first eye exam. No complaints
- POH: No h/o any eye problems
- MH: Good health. No meds
- FOH: Unremarkable
- Vision: 20/20 each eye without correction
- Pupils and motility: Normal.
- IOP: 12/13 mmHg @ 9am
- External: Normal



# What is going on here?



#### https://app.tophat.com/e/777538

Choroidal nevi

Congenital hypertrophy of RPE

**Chorioretinal scars** 

Gardner's Syndrome

Retinoblastoma

Choroidal nevi	Slate gray mass with indistinct margins
Congenital hypertrophy of RPE	Flat jet-black retinal lesion with sharp margins. Multifocal CHRPE ("bear tracks") are typically <u>unilateral</u> and clustered in a single quadrant
Chorioretinal scars	Composed of RPE hyperplasia (black) and fibrosis (white), sharp margins, often irregular in shape
Gardner's syndrome	Familial adenomatous polyposis with CHRPE-like lesions. Retinal lesions are bilateral & may appear in >1 quadrant
Retinoblastoma	Yellow-white retinal mass frequently associated with subretinal and vitreous seeding

### Assessment

- Multiple, bilateral CHRPE-like lesions
- Suspect familial adenomatous polyposis

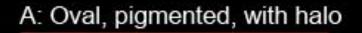
### Management

- Gastroenterology consult negative colonoscopy of child and parents
- Genetic testing offered declined by parents
- Medical surveillance for onset of polyposis

# **Familial Adenomatous Polyposis**

- An uncommon hereditary form of colon cancer (autosomal dominant)
  - About 1% of all colon cancer in US annually
- 20% of cases have no FH of FAP, suggesting a spontaneous mutation
- Some FAP patients have congenital CHRPE-like retinal lesions (hamartomas = benign RPE tumors)
  - Retinal lesions are a reliable clinical marker for FAP in these patients

# Examples of RPE lesions in FAP http://retinagallery.com



#### 62% FAP patients

B: Round, small, pigmented, no halo

#### Most common lesion

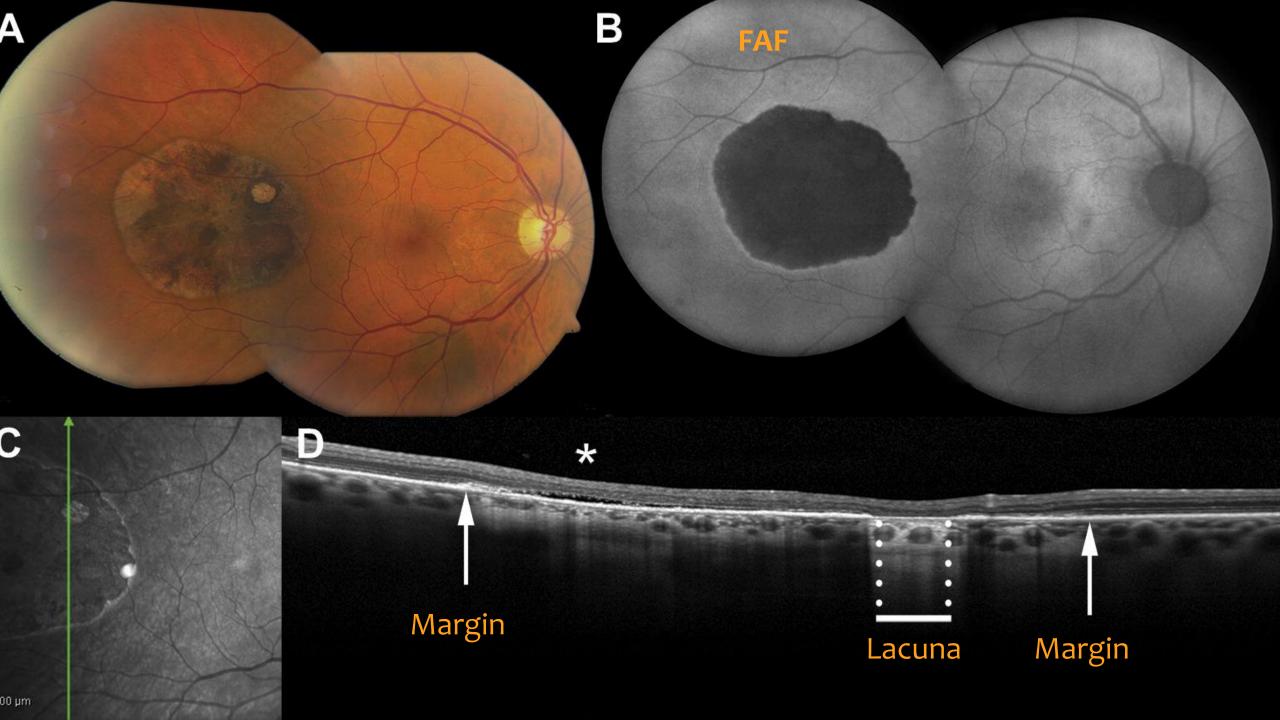


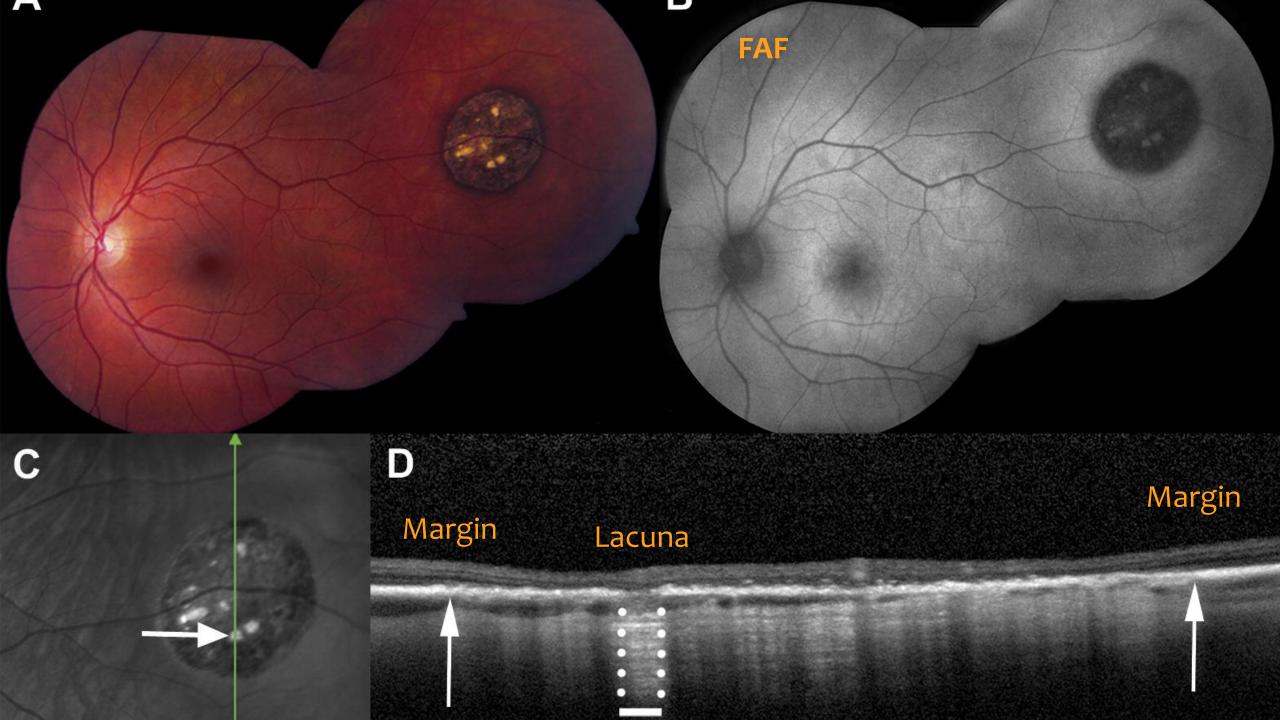
C: Round, large, pigmented D: Round, large, depigmented Chen CS. Fam Cancer. 2006;5:397 & Polkinghorne PJ. Eye 1990;4:216

# **CHRPE-like Lesions in FAP**

	Gardner's Syndrome	CHRPE
Appearance	Small: Identical to CHRPE Large: Oval with tail	Flat, round-oval, jet black
Bilaterality	Common (86% cases)	Very rare (5% cases)
Multiple quadrants	Common	Rare

#### Examples of "Bear Tracks"





#### **Retinal Lesions as a Genetic Marker**

- Retinal lesions are a sensitive and specific marker for FAP mutation carrier status
- Congenital retinal lesions may serve as an early marker for those patients destined to develop polyposis later in life
  - Onset of retinal lesions: Birth
  - Onset of polyposis: Age 25 yrs



CHRPE-like lesions may signal risk of colon cancer
How to spot suspicious CHRPE-like lesions **1.** Bilaterality!
2. > 1 quadrant per eye
3. Lesions associated with a depigmented streak

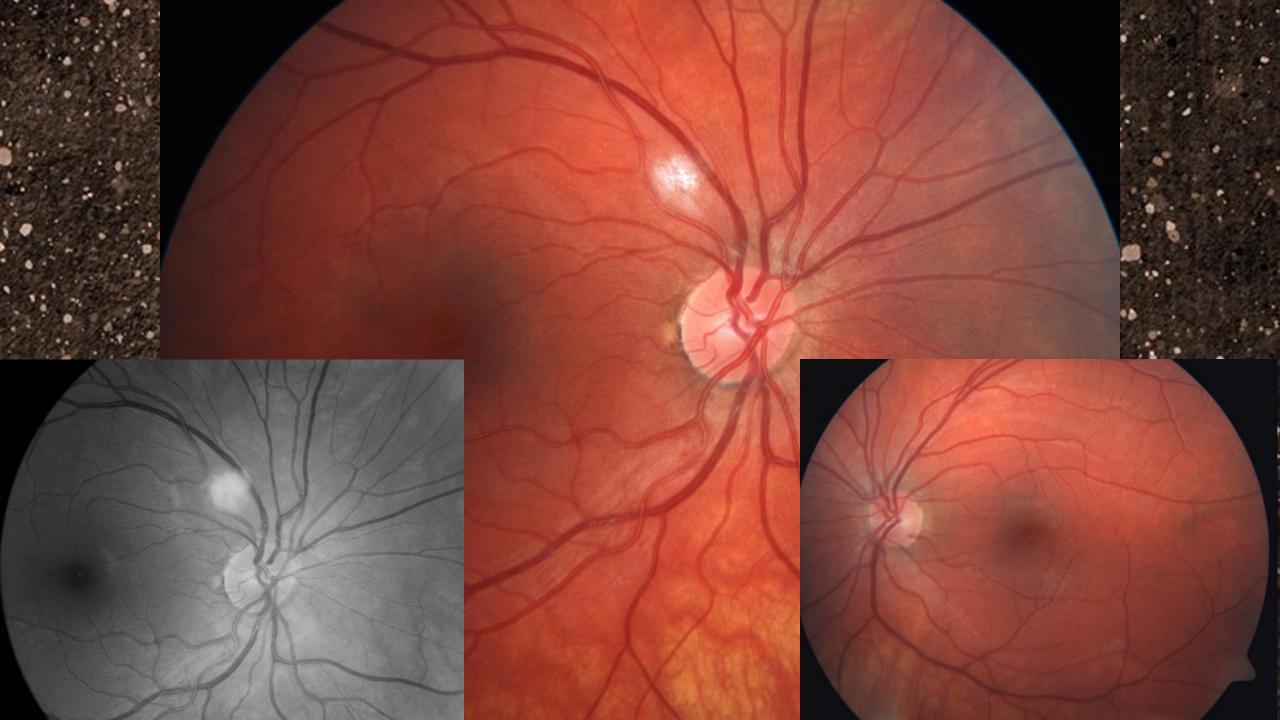


# **Case #8**

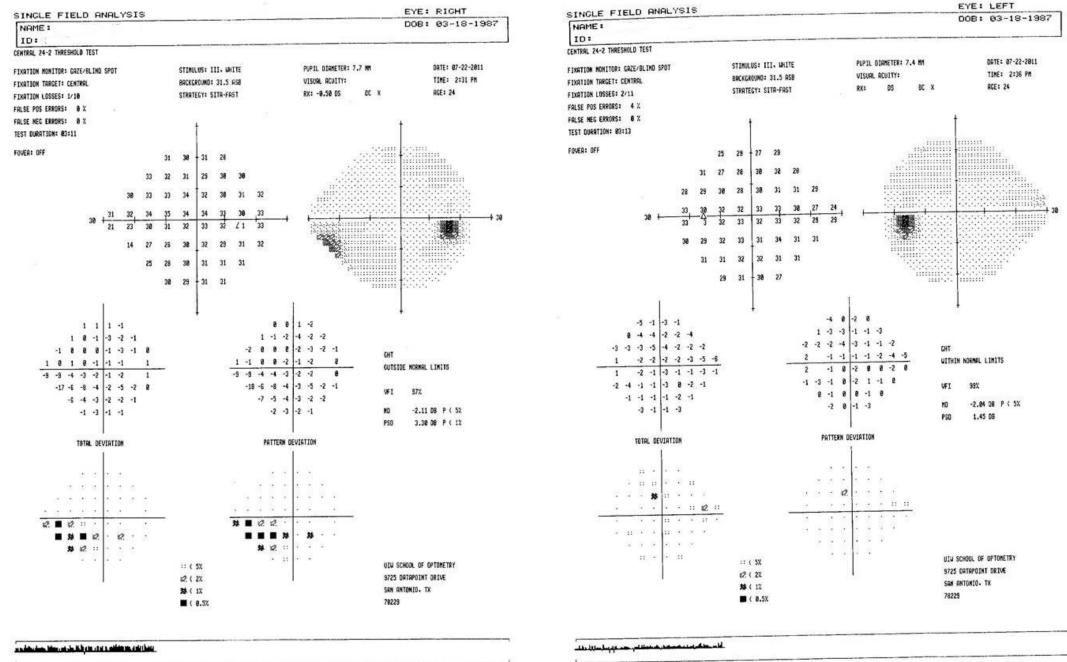
- 24yo WF presents with c/o "blurry light spot" in the inferior-nasal paracentral vision of the right eye x 1 week. Symptoms are made worse while exercising.
- POH: LEE 2yrs. Negative for any prior eye dx
- MH: Good health. Nonsmoker. Meds: BCP
- FH: MGM with diabetes

# Case #8

- BVA: 20/20 in each eye
- Pupils and motility: Normal
- BP: 113/71 RAS, Pulse: 81 bpm
- IOP: 13/12 mmHg @ 4pm
- Amsler: Blurred region *inferior-temporal* to fixation OD, Normal OS
- Color: Normal OU (HRR)
- External: Normal OU







■ 2007 CARL ZEISS MEDITEC HER II 750-13293-4.2.2/4.2.2

### Assessment

 Isolated cotton wool spot in an apparently healthy young woman

# Management

- Medical evaluation to identify cause for CWS
- Recommend D/C BCP and start ASA
- Follow-up in 2 weeks

# What is going on here?



#### https://app.tophat.com/e/777538

Idiopathic cotton wool spot

Undiagnosed diabetes

**Undiagnosed HIV** 

Undiagnosed NTG

BRAO

Idiopathic CWS	It has been reported that an underlying disorder can be found in 95% of patients with isolated CWS	
Diabetes	Undiagnosed diabetes is the <u>most common</u> cause of isolated CWS in an apparently normal patient	
HIV	CWS are a prominent feature of HIV noninfectious retinopathy, the prevalence of which is inversely related to the patient's CD4+ count	
NTG	Cotton wool spots are not associated with NTG	
BRAO	CWS are a universal feature of BRAO	

# **Medical Evaluation**

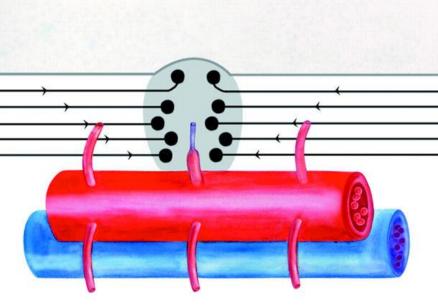
- Physical exam by PCP was normal
- Normal laboratory testing: Fasting glucose, CBC, ANA, Rheumatoid factor, C-reactive protein, HIV screen
- Normal carotid Doppler and echocardiogram
- FTA-ABS was minimally reactive
  - Serologic ELISA testing for Lyme disease recommended
- Follow-up: Photopsia persisted x 4-6 wks before abating. VF defect remained unchanged

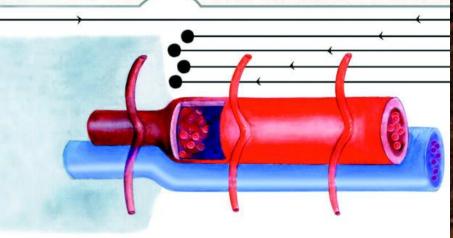
# What is a CWS?

- CWS is a localized accumulations of axoplasmic material within adjacent bundles of ganglion cell axons.
- Two clinical presentations:
  - 1. Focal ischemia from terminal arteriolar occlusion.
  - 2. Appearance at the boundary of an ischemic region of the retina

Focal ischemia: Occlusion of a terminal branch of a retinal arteriole results in a small area of infarction (grey) in the RNFL where axoplasmic transport is obstructed.

Sentinal lesion: Occlusion of a retinal arteriole results in retinal infarction







# Vision Loss Associated with CWS

- CWS are almost always asymptomatic
- Localized or arcuate scotomas common
- OCT studies reveal permanent loss of the inner retinal layers at the site of resolved CWS lesions
- Some visual recovery may occur following resolution of CWS lesions

# **Evaluation of Idiopathic CWS**

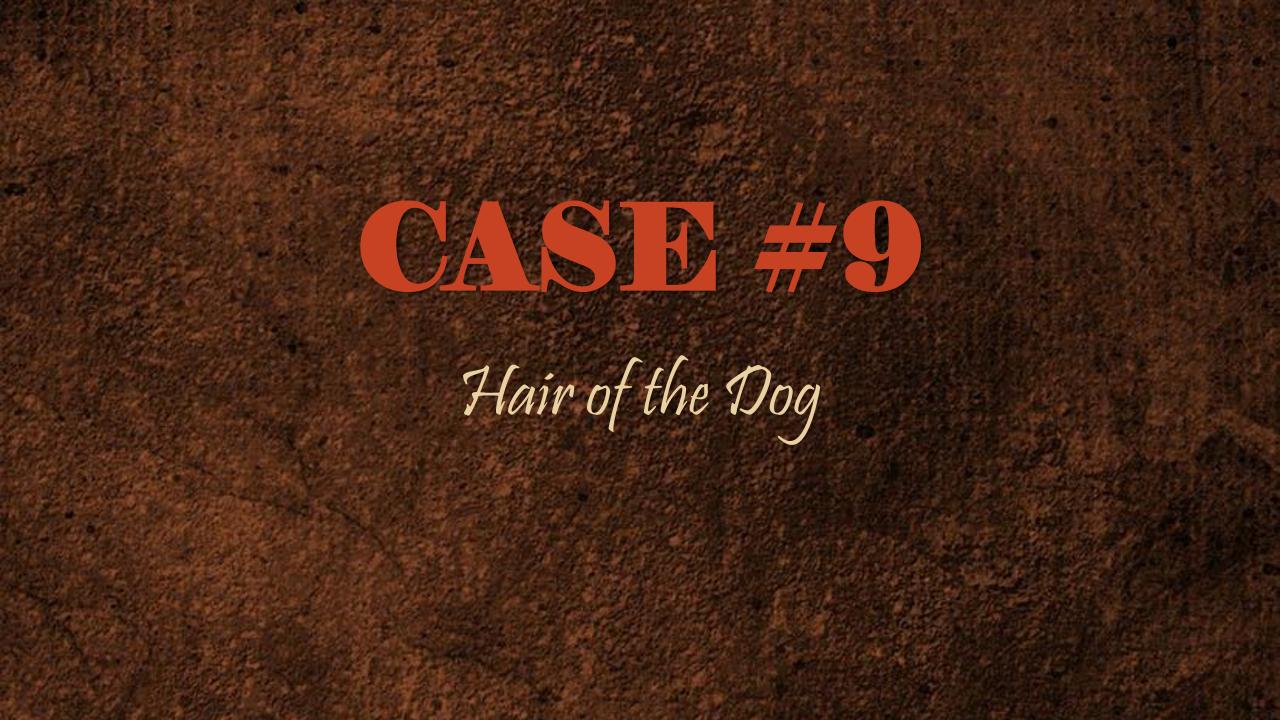
- Search for conditions that predispose the patient toward embolism and thrombosis
- Common: Diabetes, hypertension, and collagen vascular disease
- Less common: HIV and other infections, hematologic disease and coagulopathies, pancreatitis, embolic disease, trauma, and pregnancy

# **OCP** and Thromboembolism

- Birth control pills increase the risk of venous thromboembolism (VTE) by about 5x
  - Additional risk factors: smoking, obesity, HTN, coagulopathies, and a FH of thromboembolic disease
- Third generation OCPs
  - Lower risk of MI, CVA, and other side effects (weight gain, acne, headaches and unwanted hair growth)
  - Higher risk of VTE



- Isolated CWS are not truly isolated
- Diabetes, hypertension, and collagen vascular disease are common causes of CWS
- CWS are frequently associated with permanent VF defects
- Birth control pills are a significant risk factor for vascular occlusions in young healthy women



# Case #9

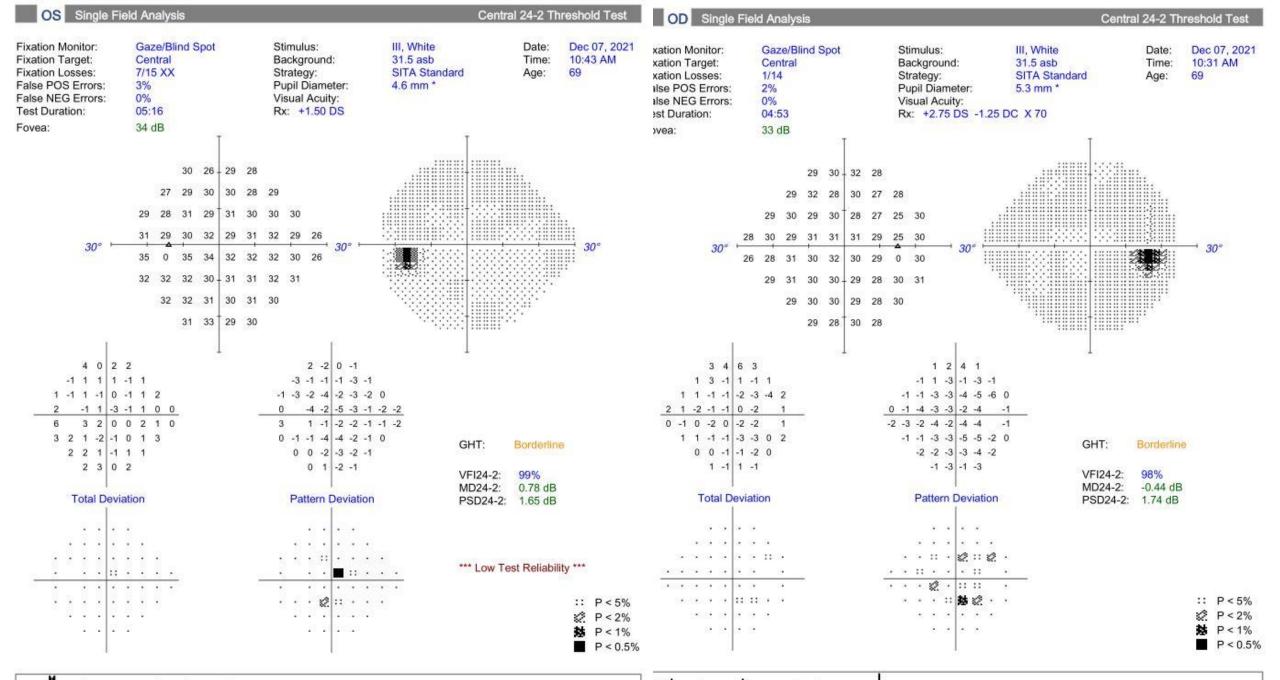
- 69yo WF presents for routine eye exam.
- POH: LEE 1yr. Negative for any prior eye dx
- MH: Good health
- VA: 20/25+ OD, 20/20 OS
- PERRL, (-)APD
- GAT: 27/28 @ 12pm
- SLE: White & Quiet

↑10 mmHg from 1yr ago

### Case #9

- C/D: 0.6 OD, 0.5 OS
- Mild ERM OD
- Gonio: D4of OU
- CCT: 572 OD, 576 OS
- IMP: Glaucoma suspect
- PLAN: Schedule OCT & VF

# Fundus photos taken with OPTOS camera

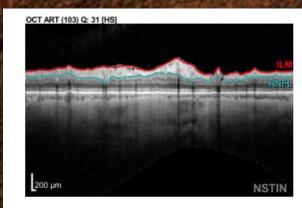


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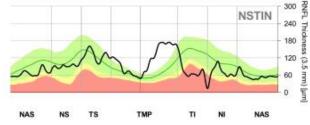
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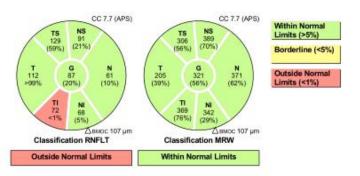
#### Hood report reveals effect of ERM OD

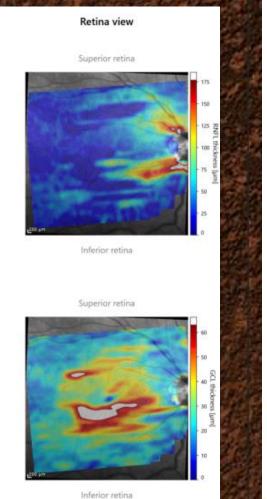
#### OS



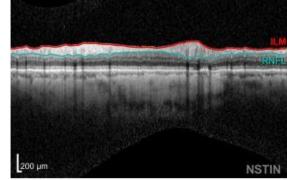
OD

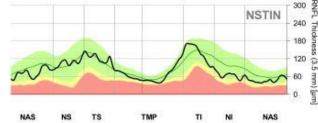


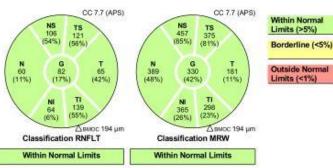




OCT ART (26) Q: 26 [HS]

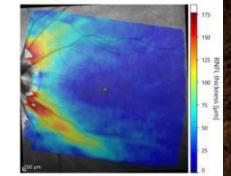






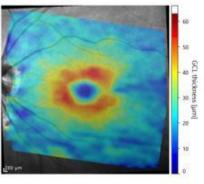
**Retina view** 

Superior retina

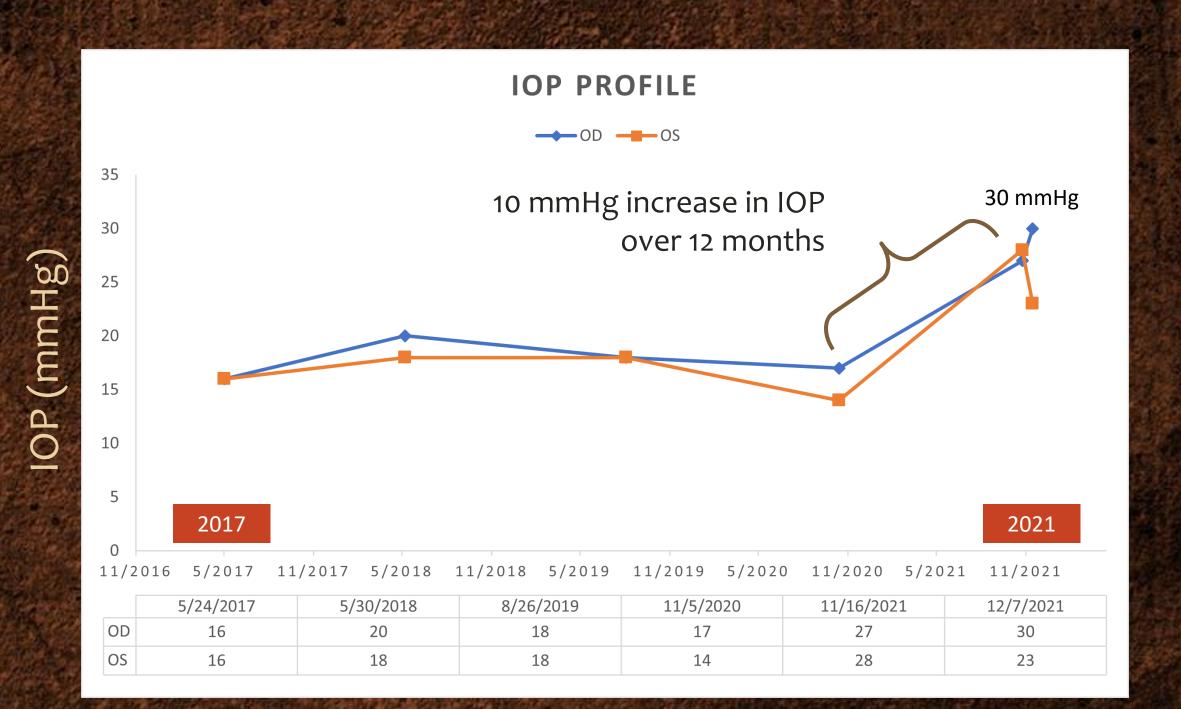


Inferior retina

Superior retina



Inferior retina



# What is going on here?



#### https://app.tophat.com/e/777538

Angle Closure

Start on steroid medication

Previously undetected large diurnal variation

Discontinuation of systemic beta blocker

Influence of other drugs or activities

Angle closure	Always suspect angle closure in patients with sudden changes in IOP	
Steroid	Any steroid by any route of administration can elevate IOP	
Diurnal variation	Normal: 2-6 mmHg. Checking IOP on another day, Water drinking test or iCare HOME to investigate	
Beta blocker	Systemic beta blockers can affect IOP same as topical	
Other factors	Caffeine, ethanol, marijuana, exercise	

#### BEFORE



### **AFTER 16 WEEKS**

Source: lattise.com

#### Latisse

- Latisse == Generic Lumigan == bimatoprost 0.03%
- Latisse has all the same clinical effects as Lumigan
- Adverse effects with Latisse: conjunctival hyperemia and irritation, increase in iris pigmentation, periocular skin pigmentation, and periorbital fat atrophy



Latisse is applied to the upper lid only. Excess solution is immediately removed to minimize risk of periorbitopathy



Once nightly, start by ensuring your face is clean, makeup and contact lenses are removed.



Remove an applicator from its tray. Then, holding the sterile applicator horizontally, place one drop of LATISSE® on the area of the applicator closest to the tip but not on the tip.



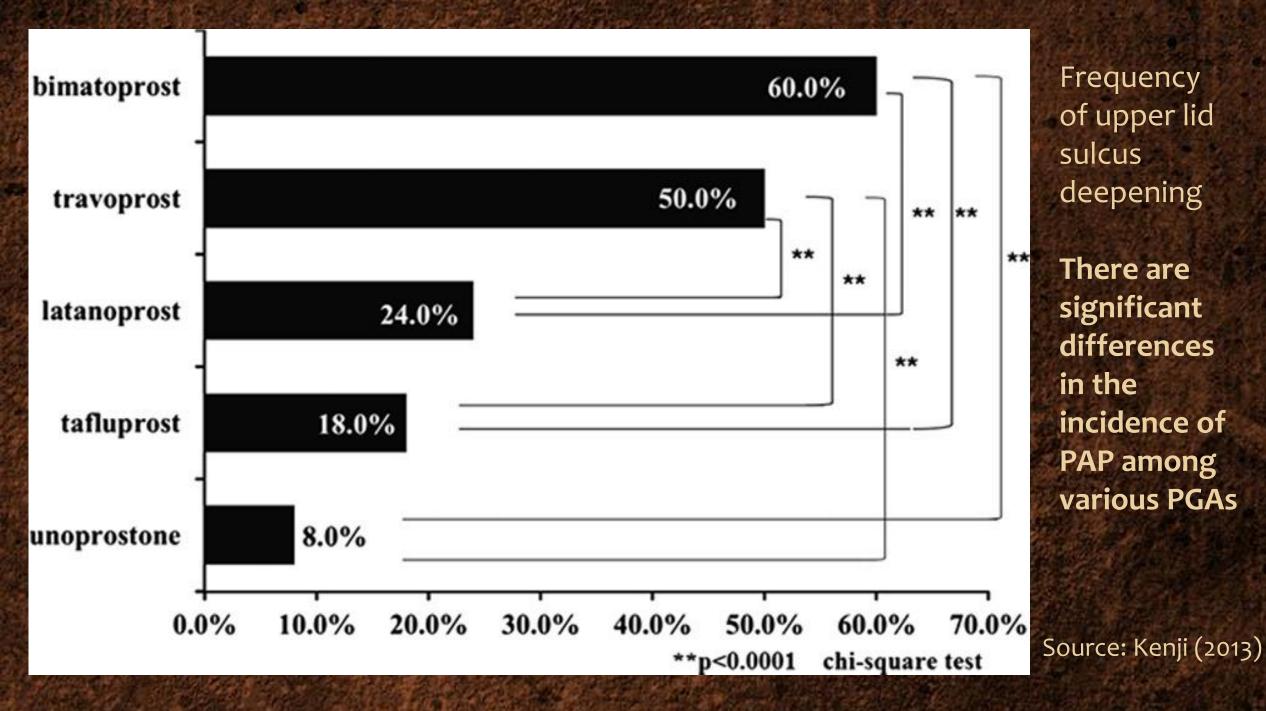
Then immediately draw the applicator carefully across the skin of the upper eyelid margin at the base of the eyelashes (where the eyelashes meet the skin) going from the inner part of your lash line to the outer part.



Blot any excess solution beyond the eyelid margin. If the solution gets into the eye, it is not expected to cause harm. The eye should not be rinsed.



Dispose of the applicator after one use. Repeat for the opposite upper eyelid margin using a new sterile applicator. This helps minimize any potential for contamination from one eyelid to another.



#### Persistence of periorbitopathy after D/C bimatoprost

#### Initial presentation

#### 4mos after d/c bimatoprost

30 mos

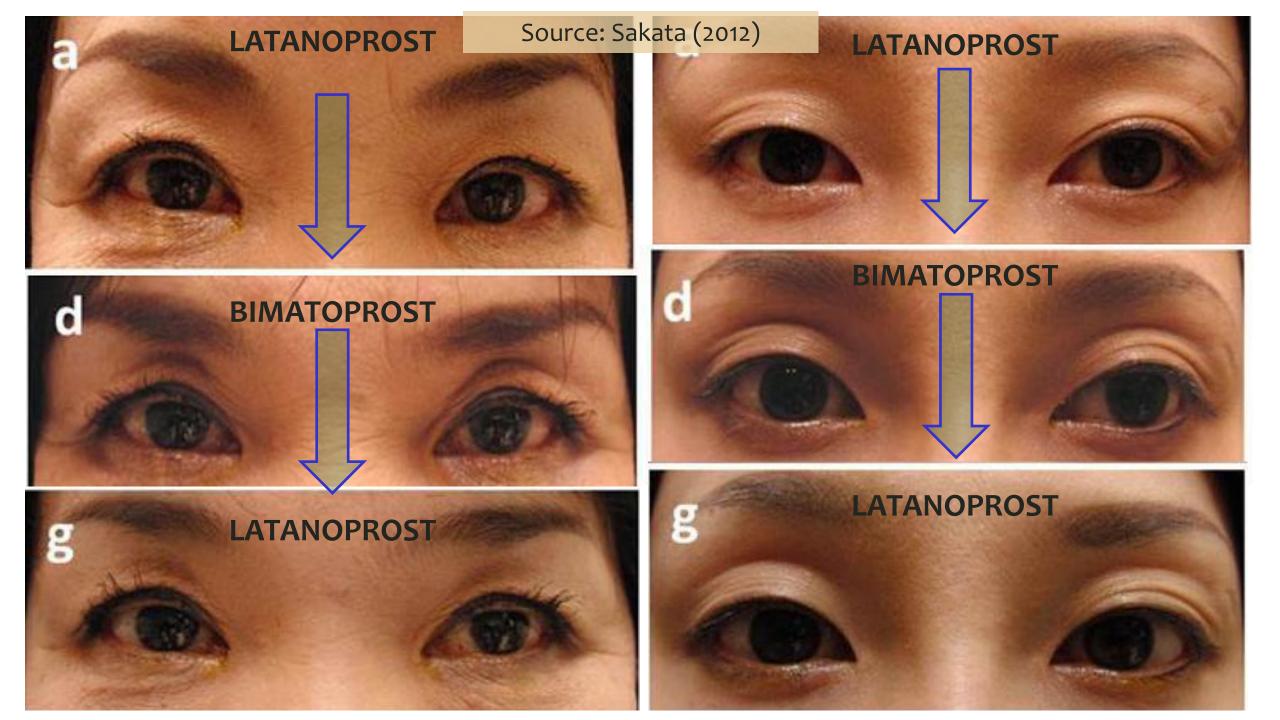


#### Initial presentation

5mos after d/c bimatoprost

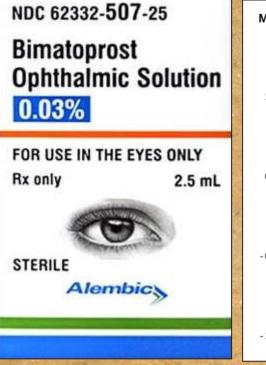
24 mos

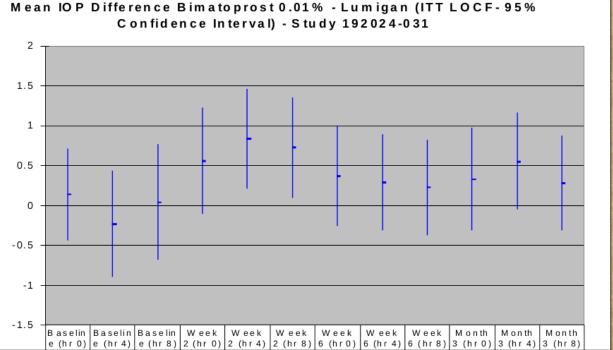
Source: Aydin (2010)



#### Generic

#### **Name-Brand**







Greater IOP lowering More side effects Less BAK (0.005%)

Less IOP lowering Fewer side effects More BAK (0.05%) Periocular discoloration after using a prostaglandin analog for eyelash enhancement: evaluation with reflectance confocal microscopy

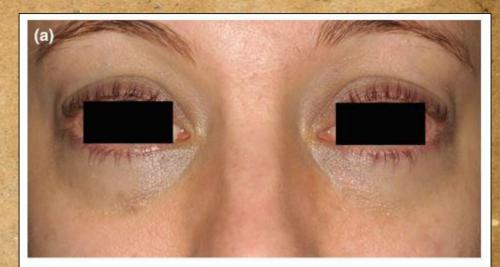
Orsolya N Horváth, MD, Valerie Letulé, MD, Thomas Ruzicka, MD, Thomas Herzinger, MD, Ilana Goldscheider, MD, & Tanja von Braunmühl, MD

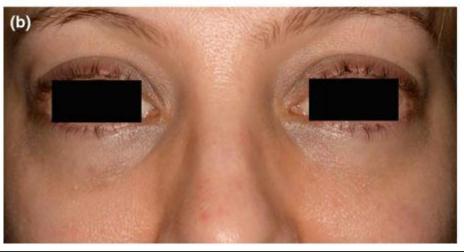
Department of Dermatology and Allergology, Ludwig Maximilian University, Munich, Germany

OTC eyelash serums may contain isopropyl cloprostenate, a PGA that can trigger the same periorbital changes seen with bimatoprost.

All have been withdrawn by FDA(?)

Source: Horvath, 2017







- Latisse can significantly affect IOP
- Bimatoprost is more frequently associated with periorbitopathy than latanoprost
- Periorbitopathy improves following D/C of PGA, but may not fully resolve
- OTC products containing isopropyl cloprostenate can induce periorbitopathy



