OCT CONNECT

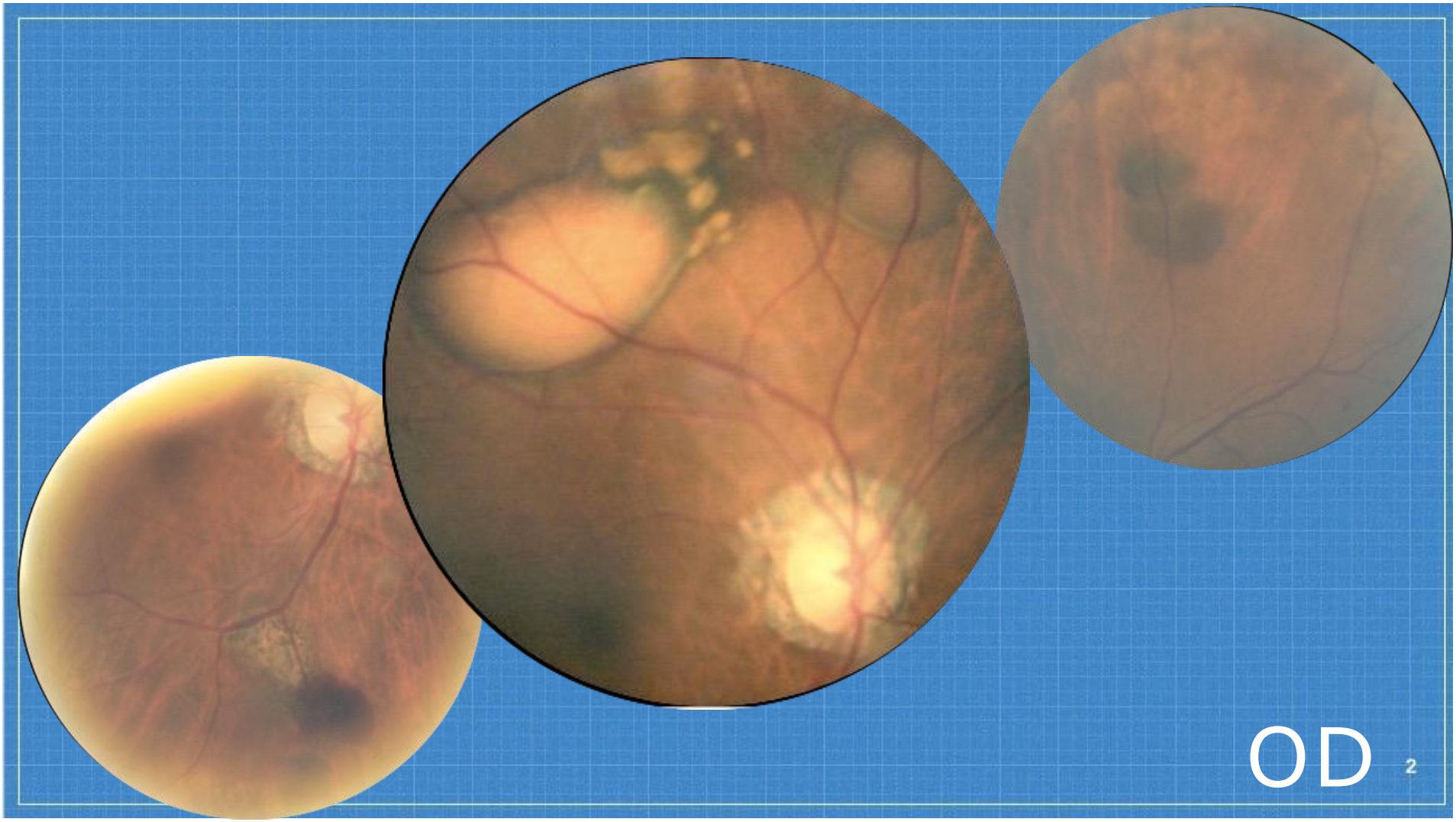
Julie Rodman OD, MSc, FAAO
Professor, Nova Southeastern University

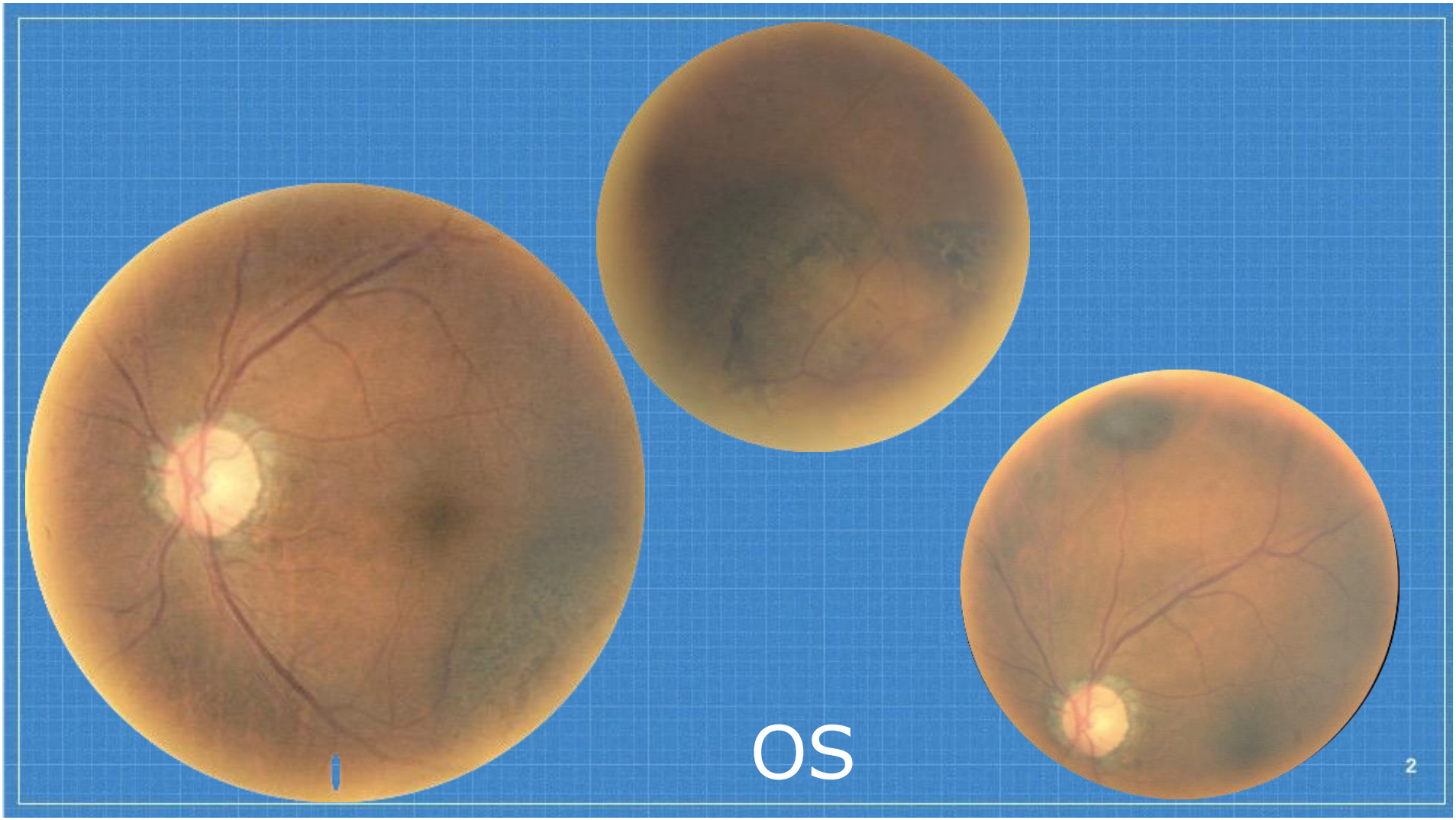
72-year-old Black Male

Presents with decreased vision bilaterally D and N

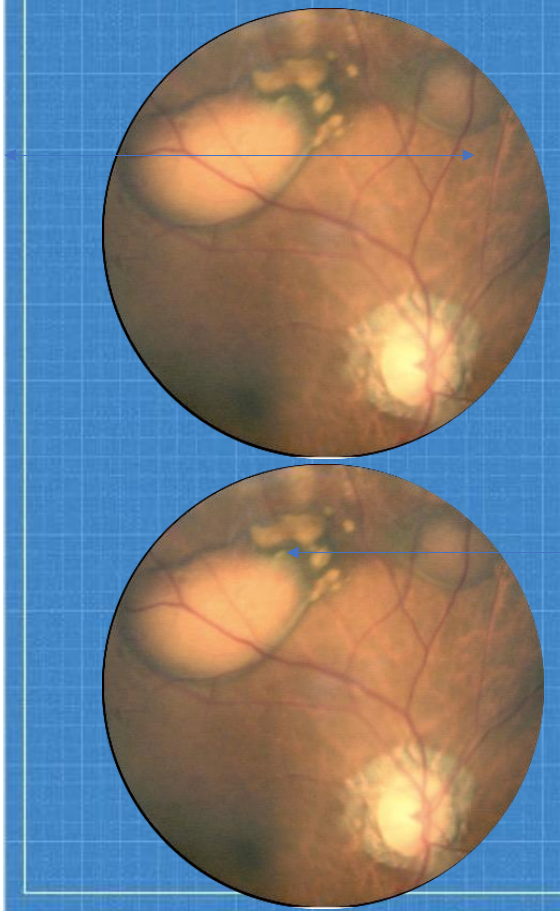
BCVA: 20/25+ OD, 20/25+ OS

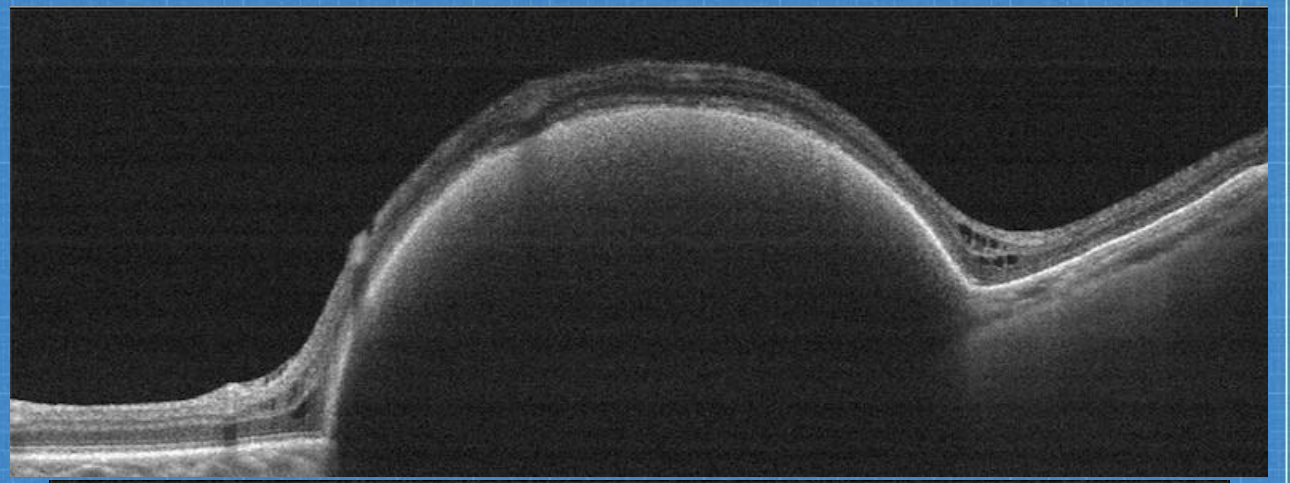
PMH:
(+) HIV; CD4 Count 336: Viral load 46
(+)Hypercholesterolemia
(+)Hypertension
(+)DM 2; poor BS control

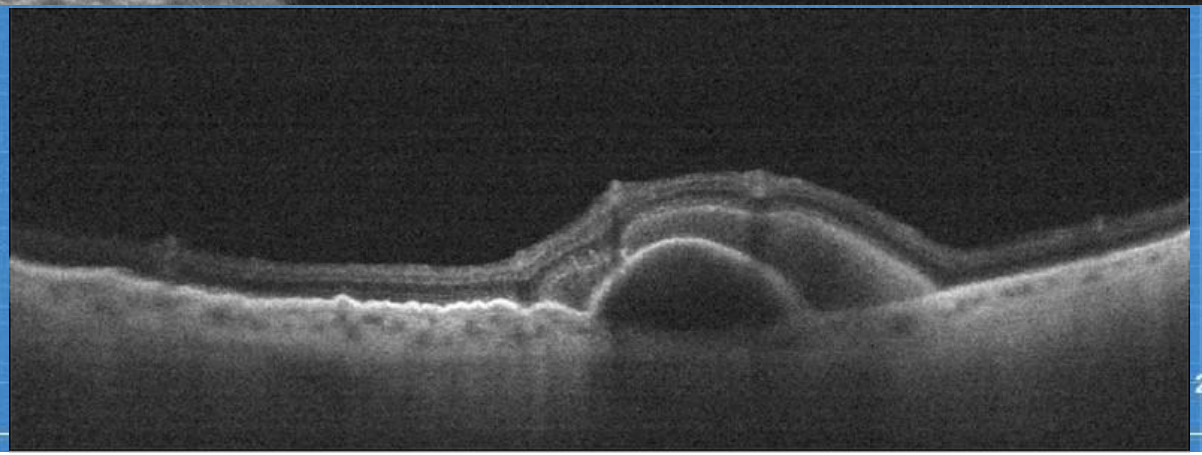

OD
2


OS
2

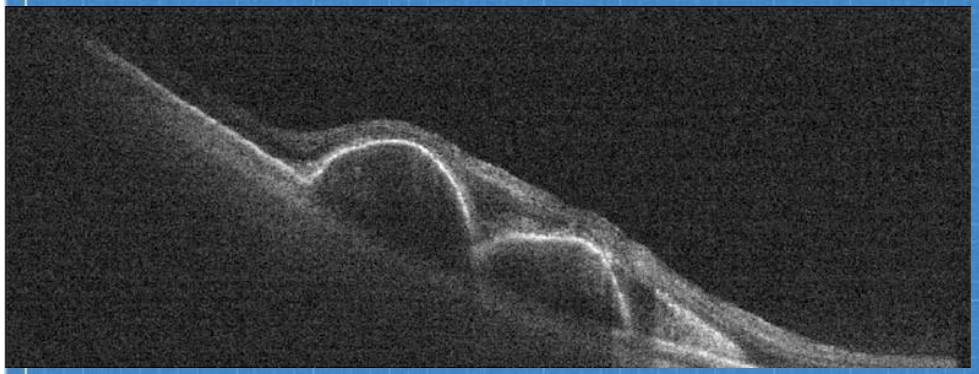
Let's look at the OCTs!

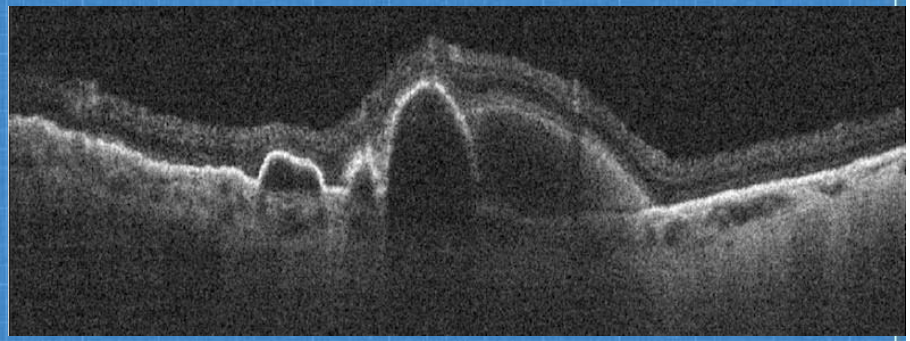


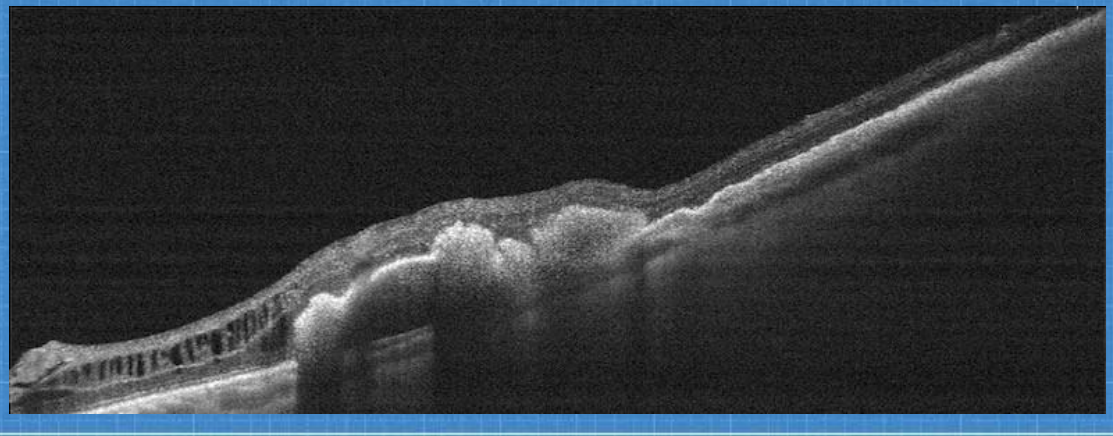




And more cuts….







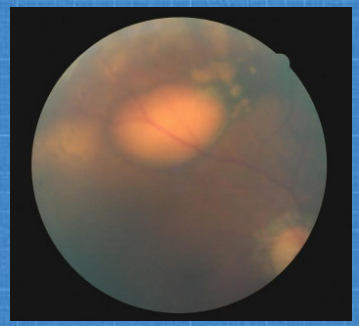
Polypoidal Choroidal Vasculopathy

- *Clinical Subtype with features of Neovascular AMD*

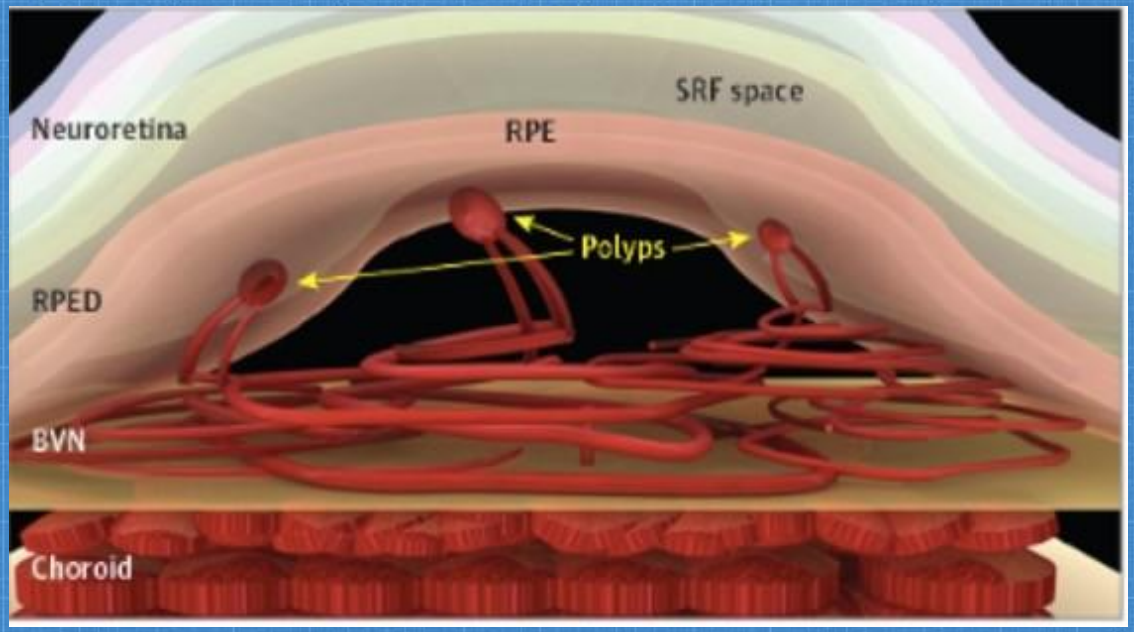
> *"Peculiar Hemorrhagic detachment of the RPE and choroid"*

Polypoidal Choroidal Vasculopathy

- Suspected in patient with: <u>sub-retinal orange polyp-like lesions</u>
  - Can be macular or peripapillary
  - Rarely in arcades as well
- Especially African or Asian descent (F>M)



Pathophysiology



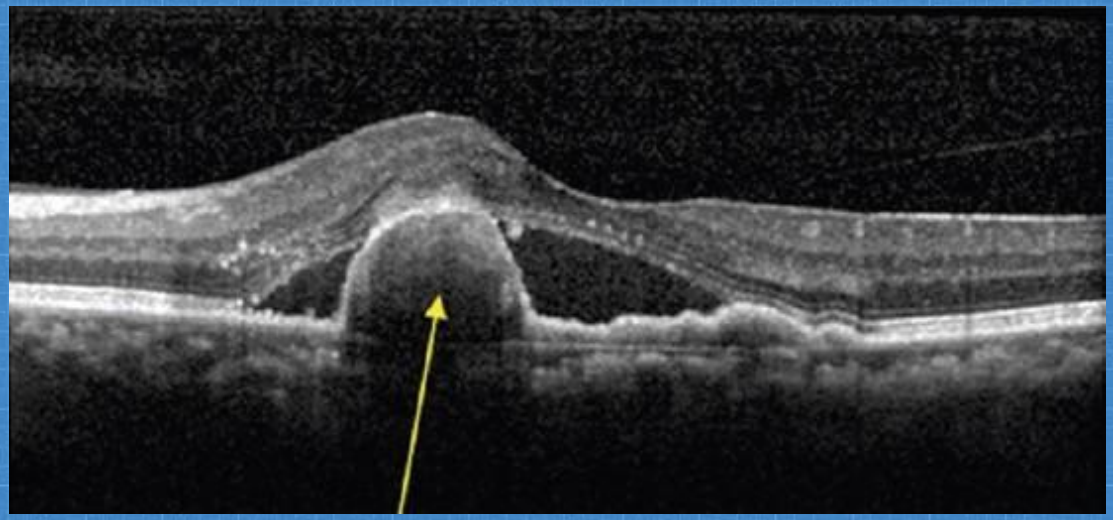
- Branching vascular network (BVN): originates in the choroid
- BVN may develop terminal, polyp-like aneurysmal dilatations

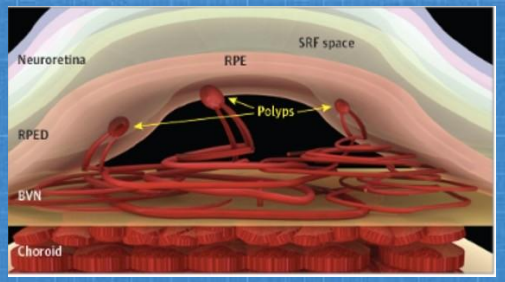
https://jamanetwork.com/journals/jamaophthalmology/article-abstract/2657209

Serosanguinous RPE detachments

Sero.....Sanguinous

Serous Fluid Blood

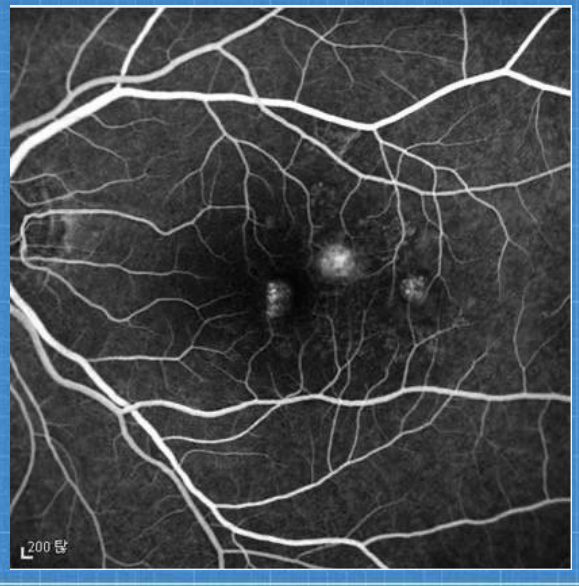


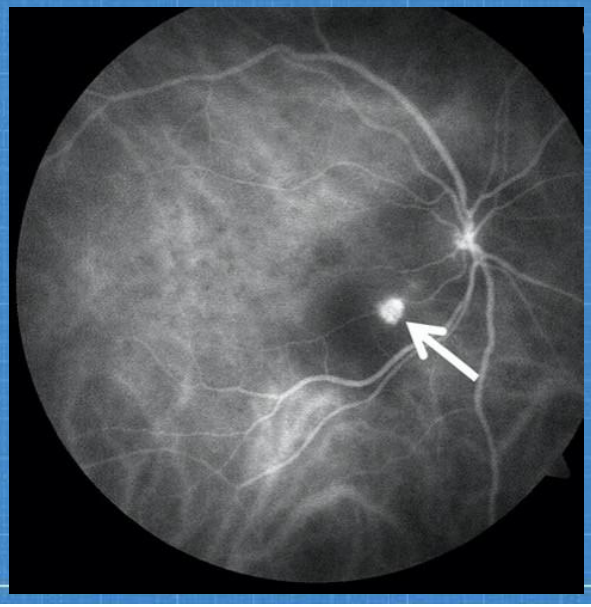


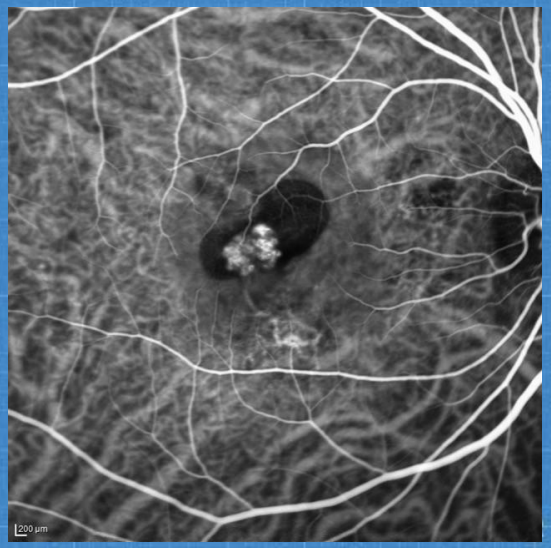
http://www.ijo.in/viewimage.asp?img=IndianJOphthalmol_2018_66_7_896_234954_f2.jpg

PCV and ICG

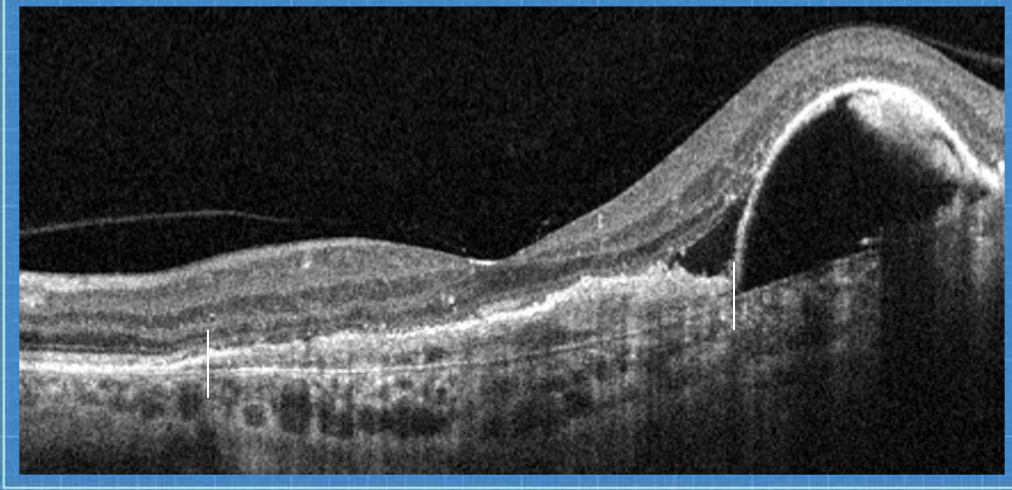
- Essential for detecting the choroidal network of polyps
  - Differentiation from AMD

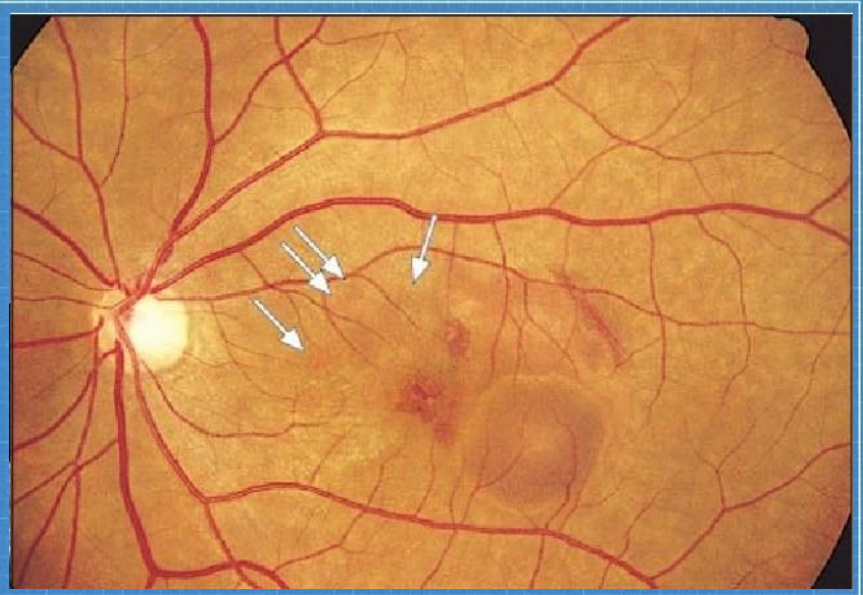






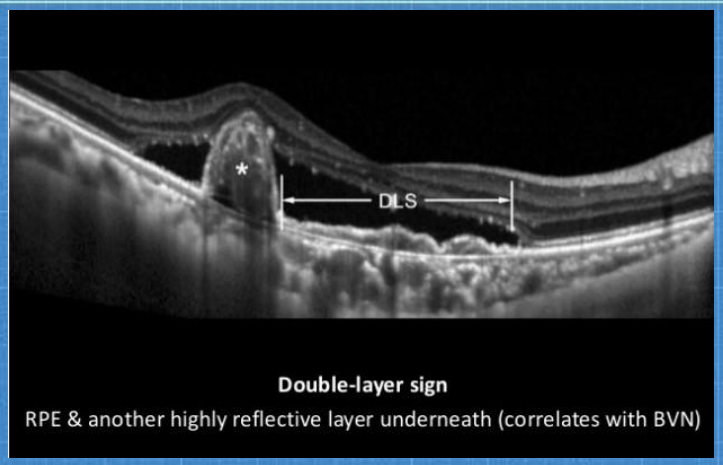
Double layer sign





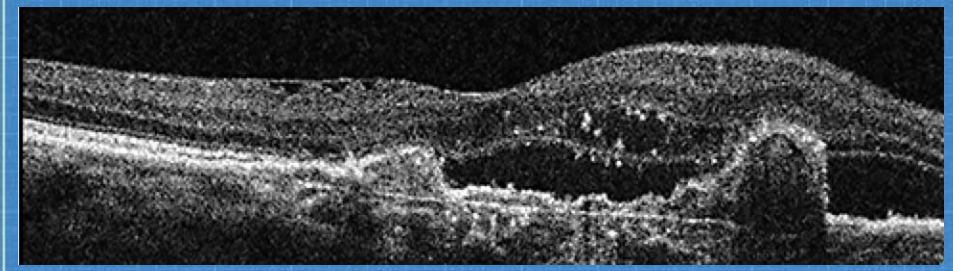
https://www.semanticscholar.org/paper/Idiopathic-polypoidal-choroidal-vasculopathy-in-Uyama-Matsubara/44eaddb3016b3bfe4d6ac09ee2f9861341ae8e1f

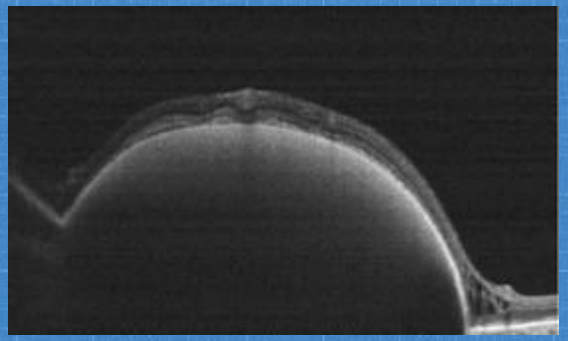
SO… how do we make the diagnosis??



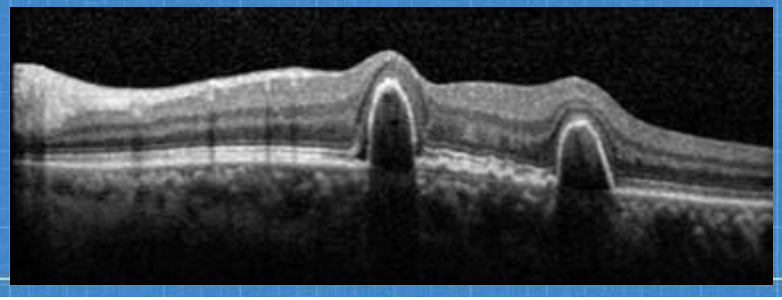
Double-layer sign
RPE & another highly reflective layer underneath (correlates with BVN)

Double Layer Sign





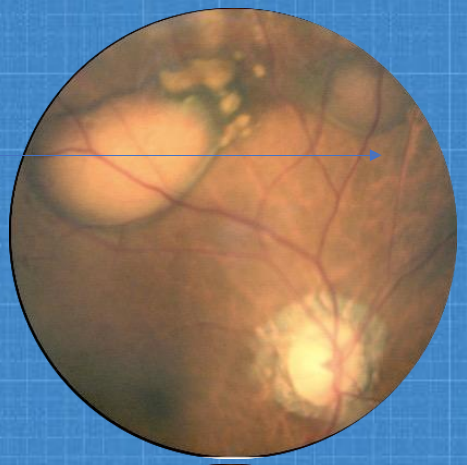
Large PEDs

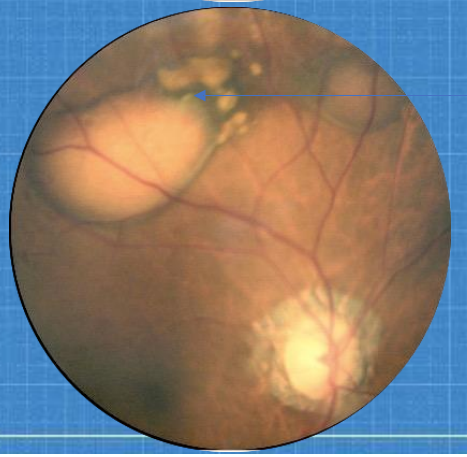


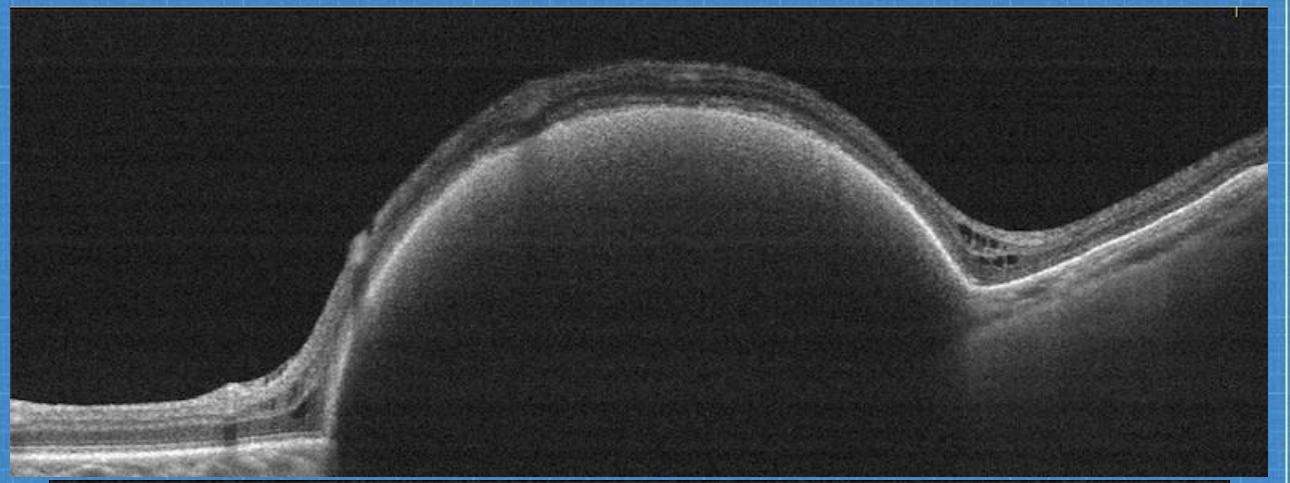
Sharp, multiple PEDs

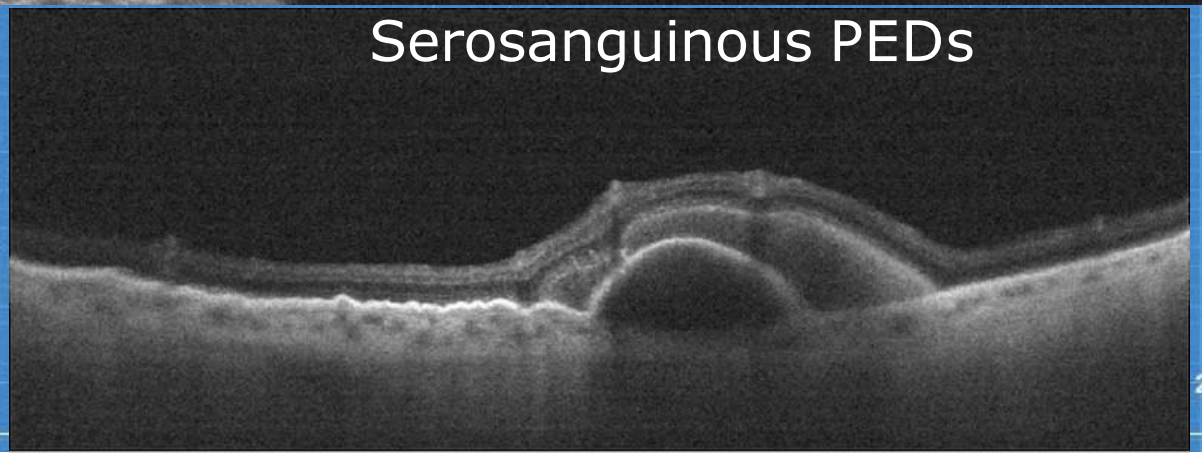
https://www.retinalphysician.com/issues/2015/april-2015/how-rap-and-pcv-can-affect-the-management-of-amd

Back to our patient:





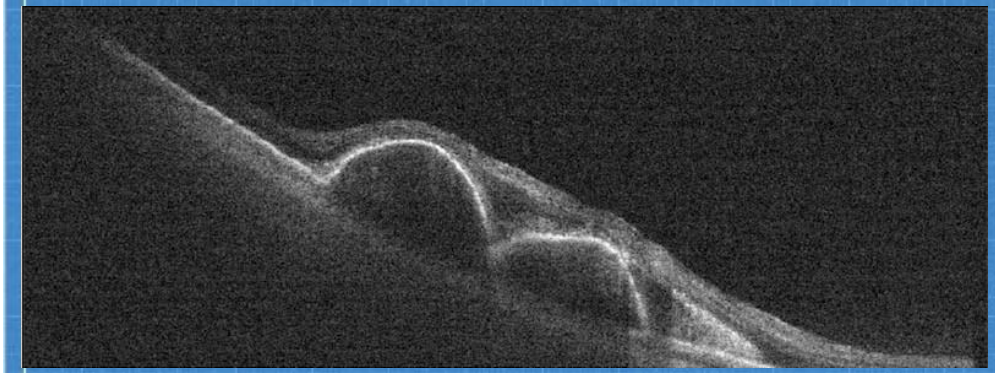


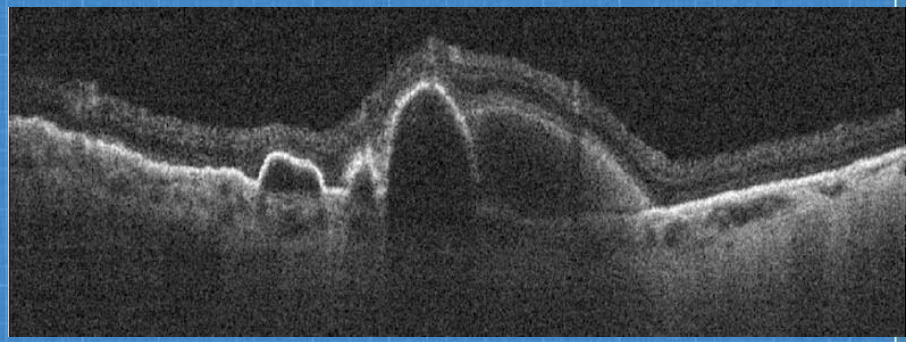


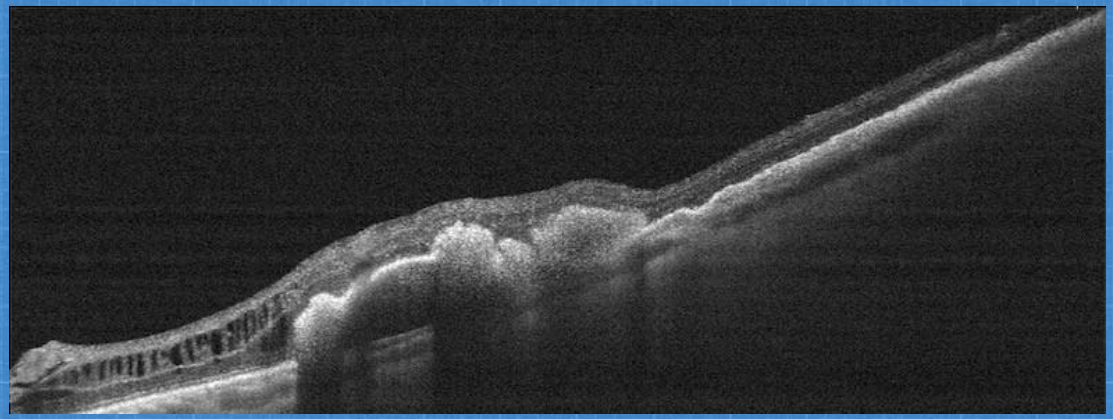
Serosanguinous PEDs

And more cuts….

Double layer sign; ?CNV





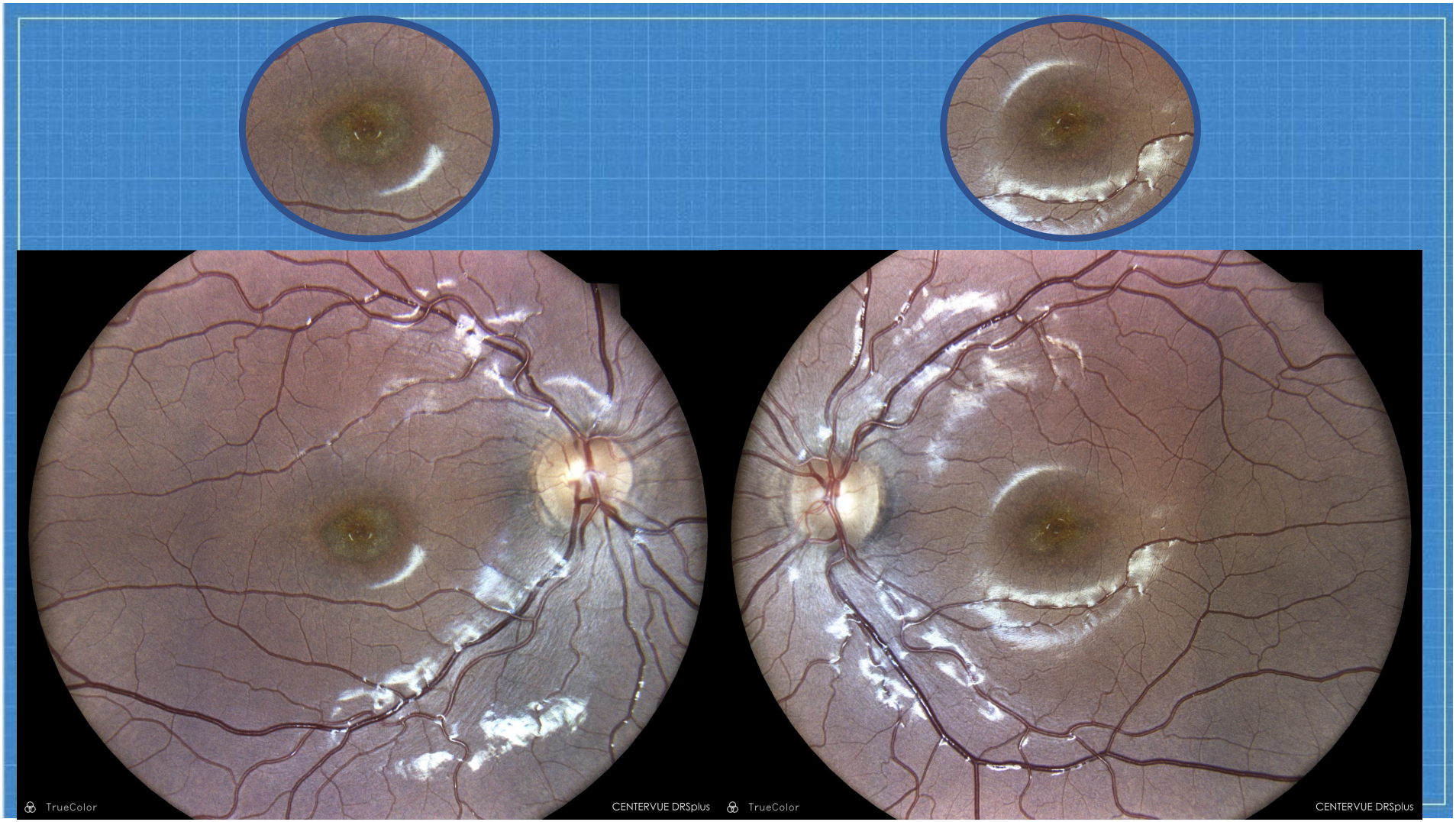


13-year-old Black Female

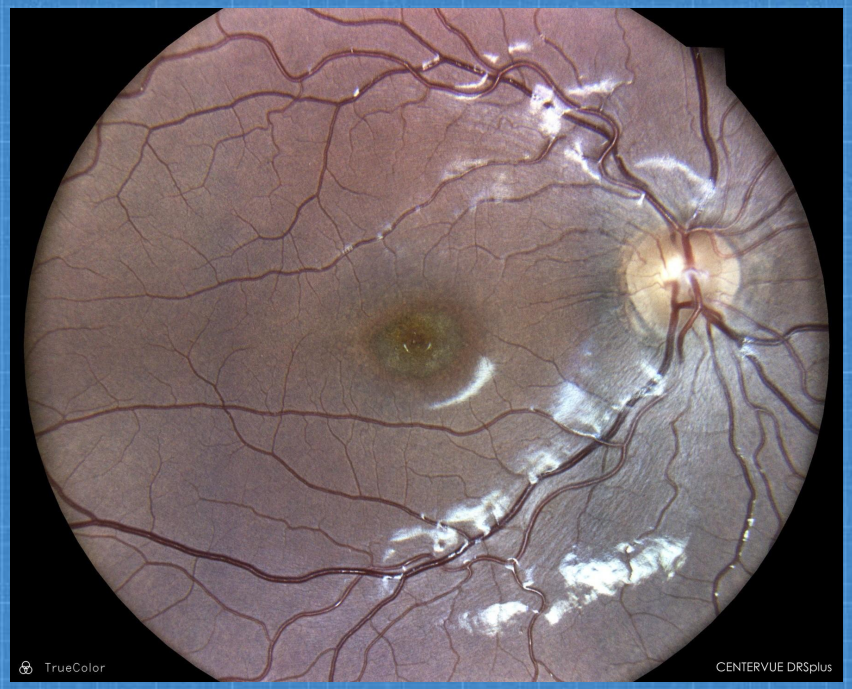
First eye exam ever!! Never had any visual problems
Mom reports that she is just NOT seeing right!

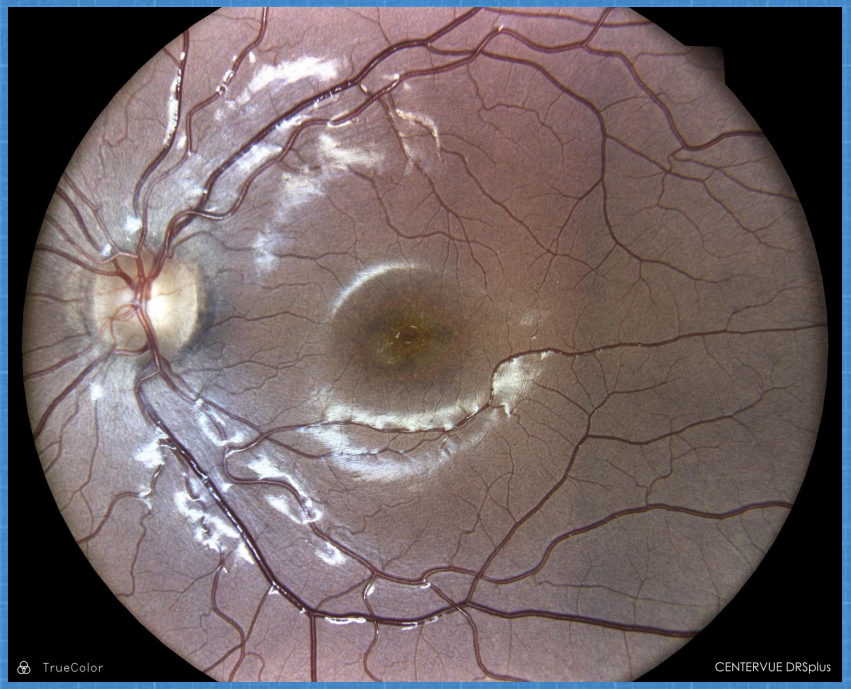
BCVA: 20/30 OD, 20/30 OS
Failed Color Vision OD and OS

"I can't see the blackboard at school and my grades are sinking!!!"


TrueColor
CENTERVUE DRSplus
TrueColor
CENTERVUE DRSplus

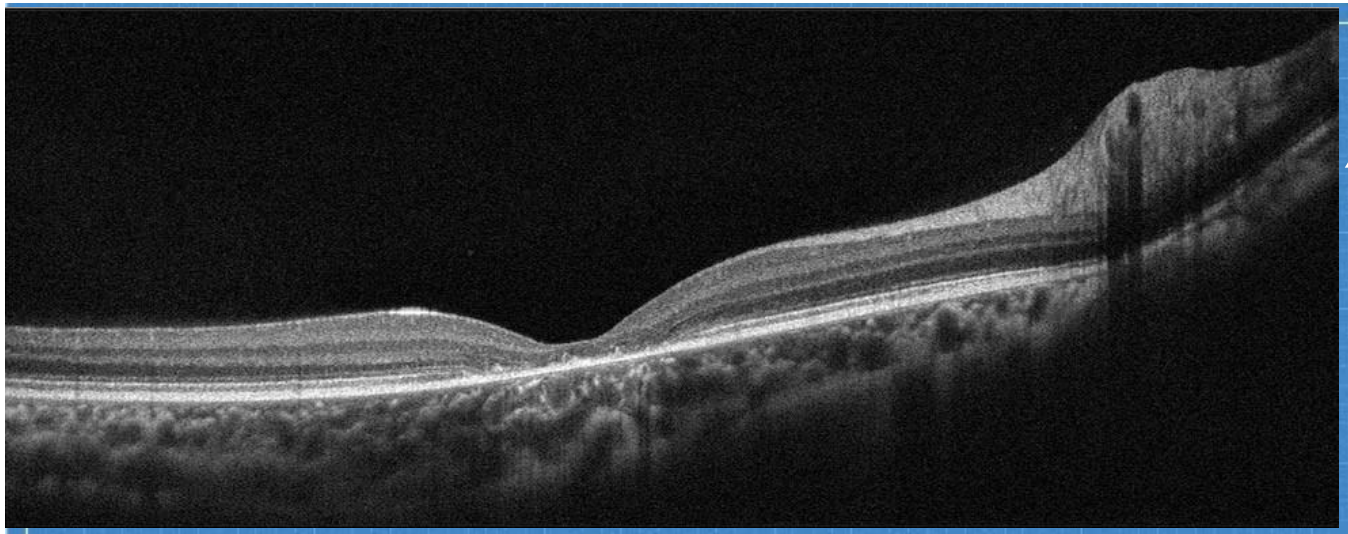
Bull's Eye Maculopathy???



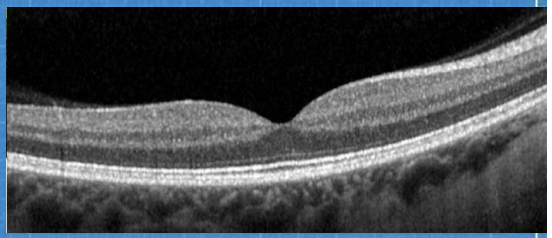


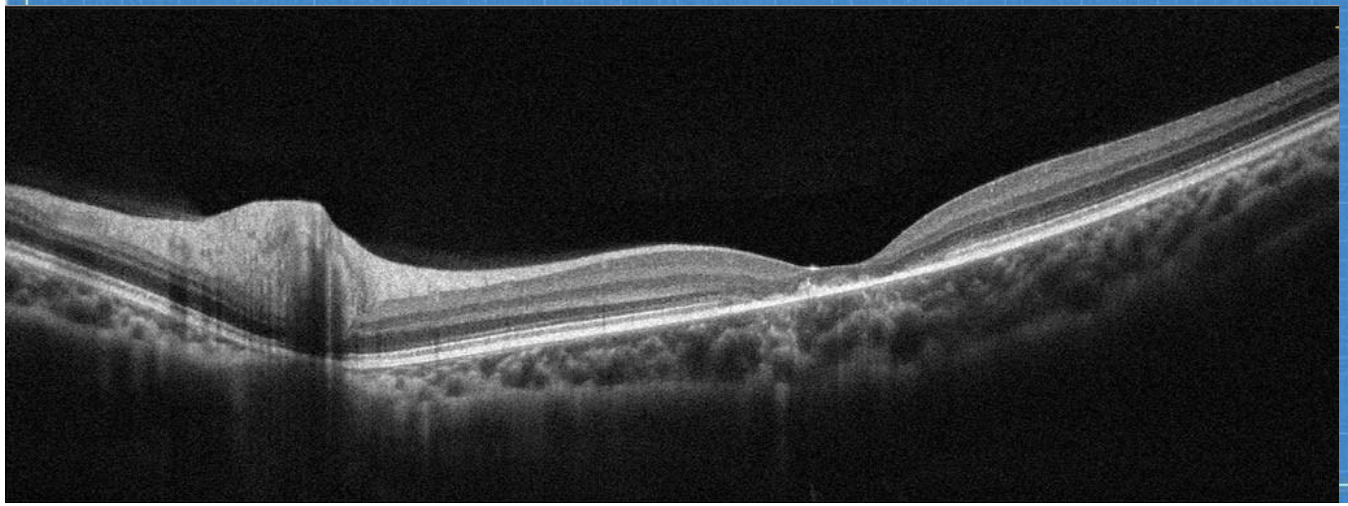
Differential Diagnosis:

1. Stargardt's Disease
2. Cone Dystrophy
3. Chloroquine Toxicity
4. Retinitis Pigmentosa?

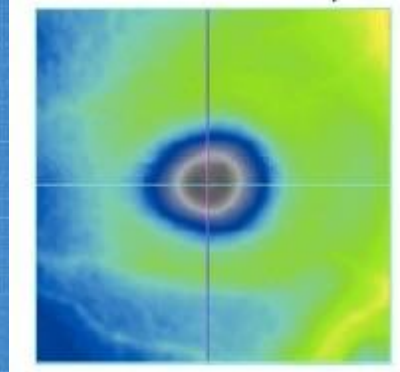


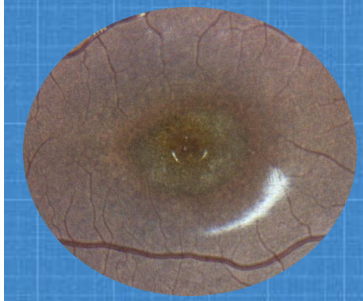
Ancillary Testing: OCT



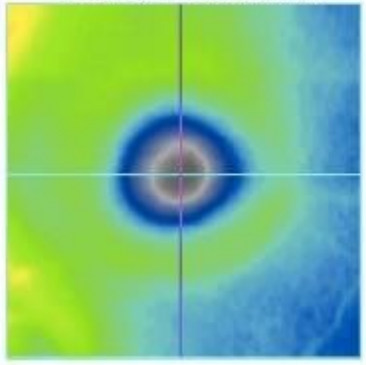


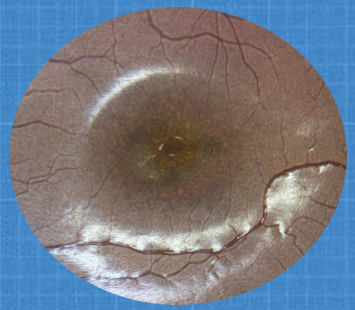
OD ILM-RPE Thickness Map



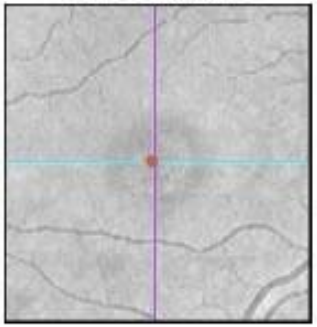


OS ILM-RPE Thickness Map

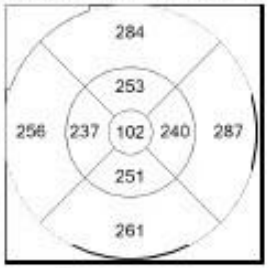




OD OCT Fundus



OD ILM-RPE Thickness

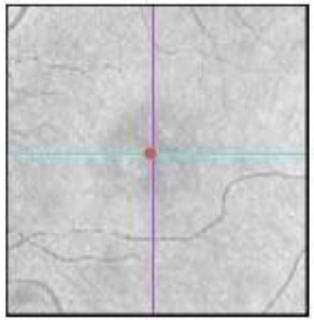


ILM - H

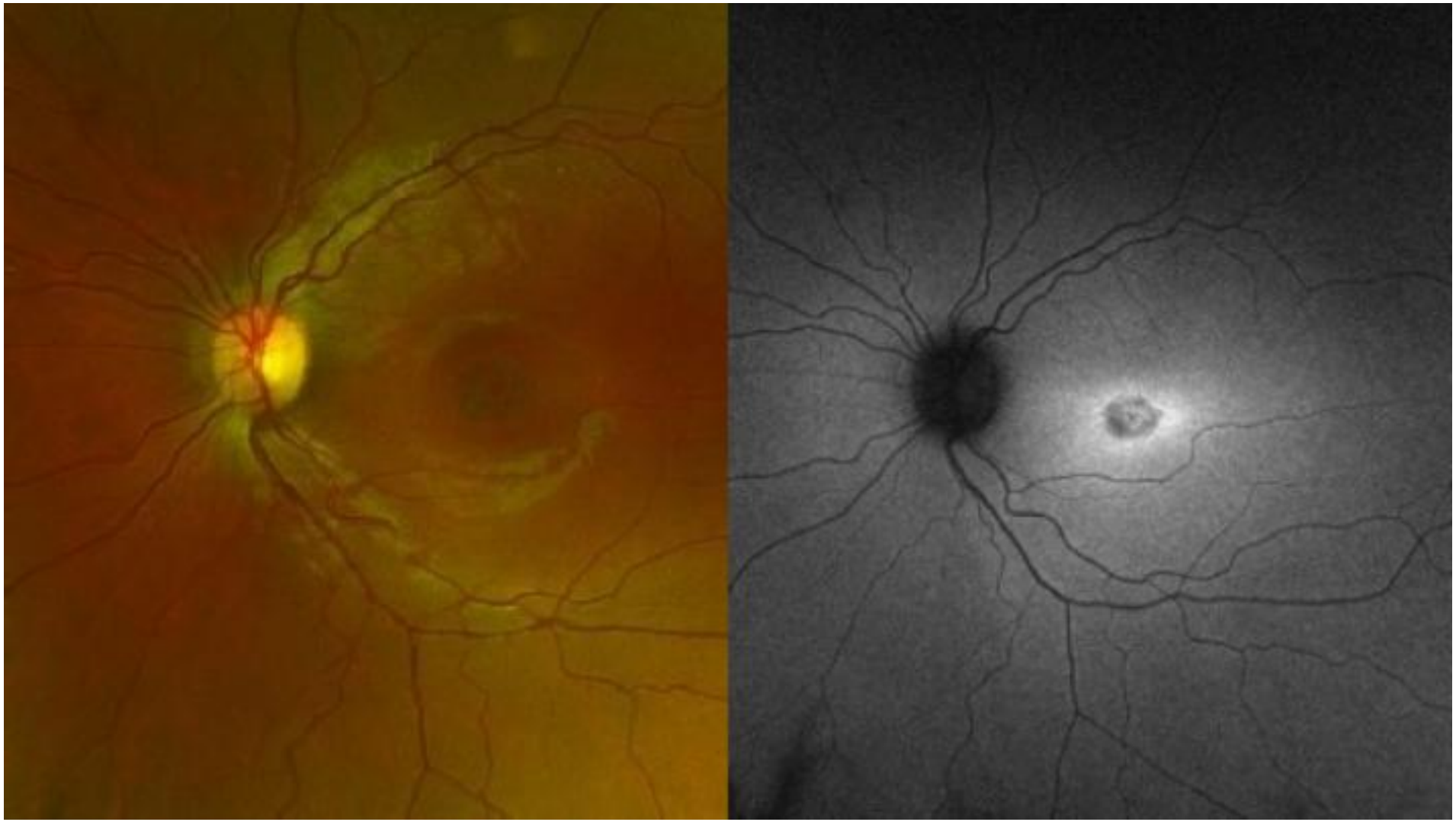
OS ILM-RPE Thickness

	OD	OS

OS OCT Fundus

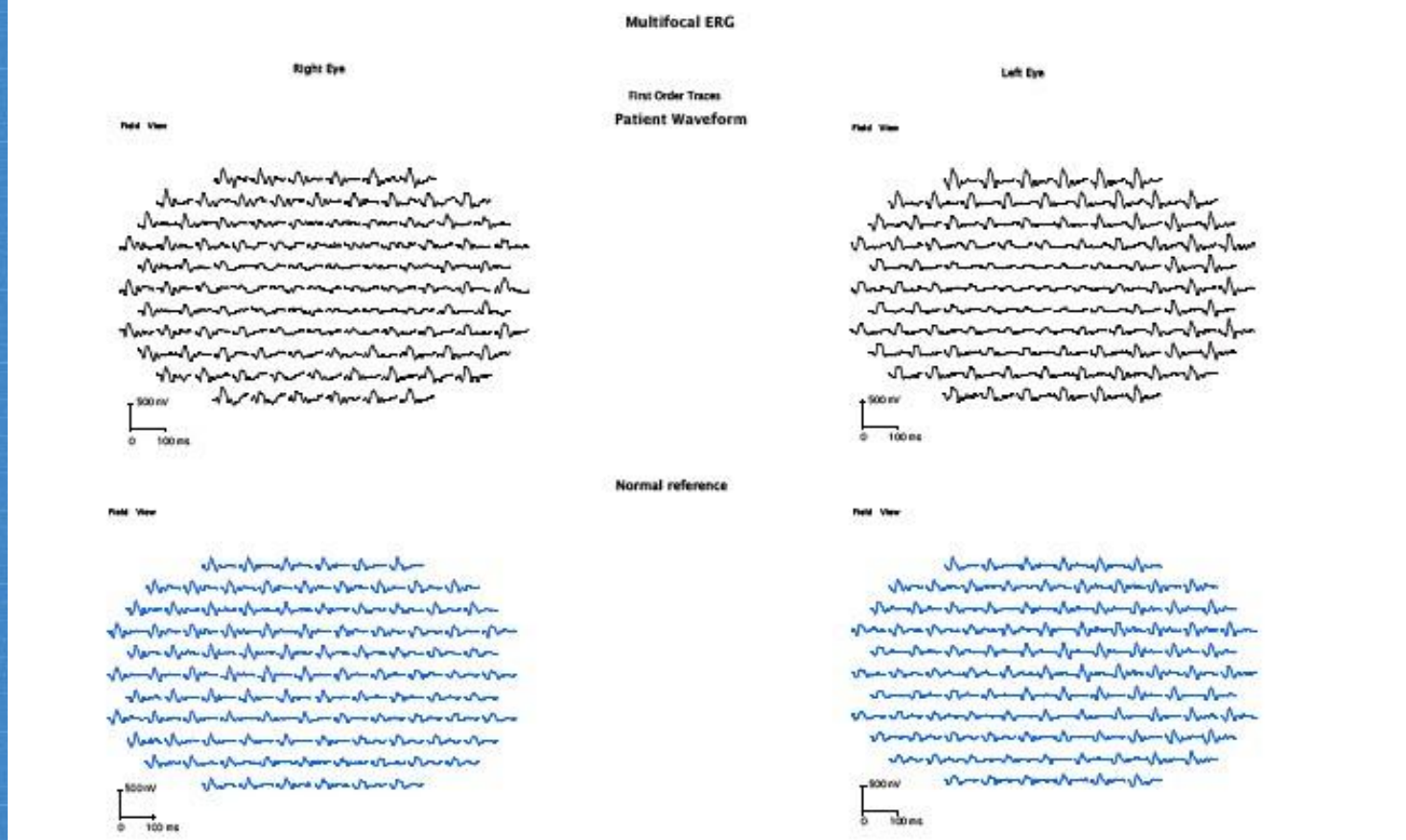


Ancillary Testing:
Fundus Autofluorescence



Electrodiagnostics: mfERG

The mfERG findings show moderate diffuse cone dysfunction in the macula area of the right and left eyes.



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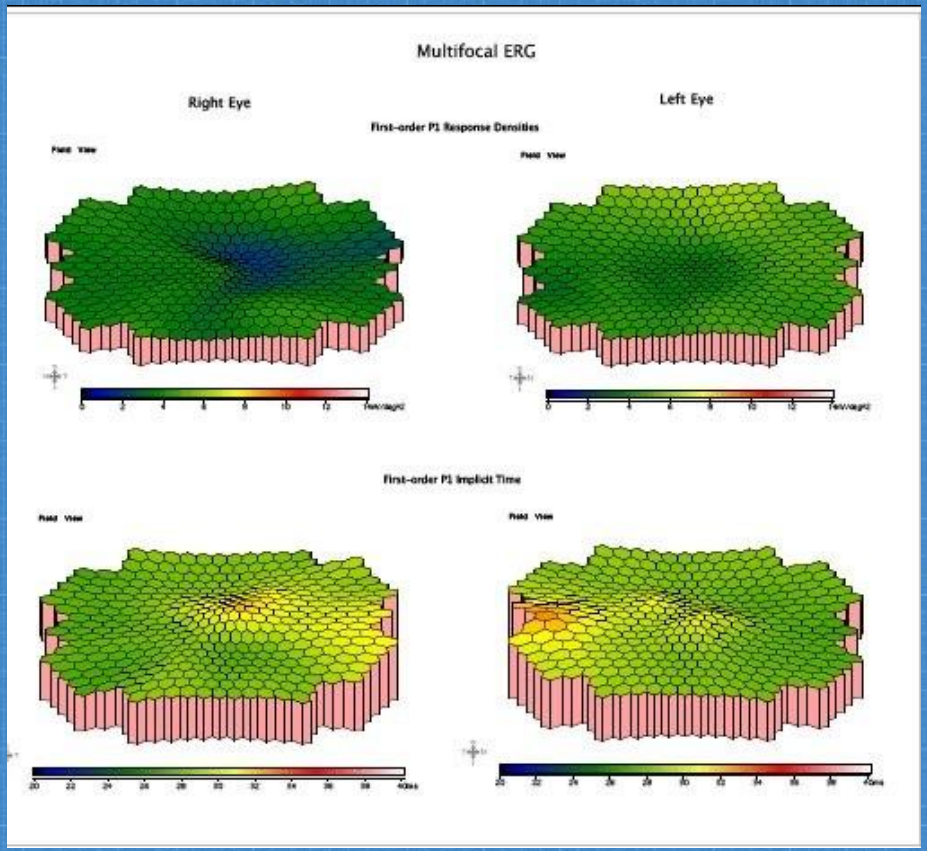


What is this??...13 y/o... we need help!!

INVITAE

INVITAE DIAGNOSTIC TESTING RESULTS

Patient name:	[redacted]	**Sample type:**	Saliva	**Report date:**	11/08/2021
DOB:	09/29/2008	**Sample collection date:**	10/12/2021	**Invitae #:**	RQ2834631
Sex assigned at birth:	Female	**Sample accession date:**	10/23/2021	**Clinical team:**	Julie Rodman
Gender:		**MRN:**			

Reason for testing

Test performed

Sequence analysis and deletion/duplication testing of the 328 genes listed in the Genes Analyzed section.

- Invitae Inherited Retinal Disorders Panel

RESULT: CARRIER

One Pathogenic variant identified in EYS. EYS is associated with autosomal recessive retinitis pigmentosa.

Additional Variant(s) of Uncertain Significance identified.

GENE	VARIANT	ZYGOSITY	VARIANT CLASSIFICATION
EYS	c.6794del (p.Pro2265Glnfs*46)	heterozygous	PATHOGENIC
ABCA4	c.2161-6T>C (Intronic)	heterozygous	Uncertain Significance
BBS1	c.1076G>A (p.Arg359His)	heterozygous	Uncertain Significance
COL11A2	c.2682G>A (Silent)	heterozygous	Uncertain Significance
PDE6A	c.916A>G (p.Arg306Gly)	heterozygous	Uncertain Significance
PDZD7	c.244G>A (p.Asp82Asn)	heterozygous	Uncertain Significance
PEX6	c.1081A>G (p.Thr361Ala)	heterozygous	Uncertain Significance
RP1	c.4397A>T (p.Glu1466Val)	heterozygous	Uncertain Significance

About this test

This diagnostic test evaluates 328 gene(s) for variants (genetic changes) that are associated with genetic disorders. Diagnostic genetic testing, when combined with family history and other medical results, may provide information to clarify individual risk, support a clinical diagnosis, and assist with the development of a personalized treatment and management strategy.

Clinical summary

A Pathogenic variant, c.6794del (p.Pro2265Glnfs*46), was identified in EYS.

autosomal recessive retinitis pigmentosa (RP)

- The EYS gene is associated with autosomal recessive retinitis pigmentosa (RP) (MedGen UID: 350427).
- This individual is a carrier for autosomal recessive EYS-related conditions. This result is insufficient to cause autosomal recessive EYS-related conditions; however, carrier status does impact reproductive risk.
- Retinitis pigmentosa (RP) is a genetically heterogeneous group of inherited eye disorders characterized by progressive degeneration of the retina, typically beginning in the midperiphery and advancing toward the macula and fovea (PMID: 17296890). Abnormalities of the photoreceptors (rods and cones) or the retinal pigment epithelium (RPE) lead to progressive visual loss (PMID: 26835369, 26106463). Typical symptoms include night blindness followed by constriction of peripheral visual fields, which leads to tunnel vision and eventually loss of central vision (PMID: 17296890). RP is highly variable in regards to severity, clinical symptoms, and age of onset (PMID: 17296890).
- Biological relatives have a chance of being a carrier for or being at risk for autosomal recessive EYS-related conditions. Testing should be considered if clinically appropriate. The chance of having a child with autosomal recessive EYS-related conditions depends on the carrier state of this individual's partner.

A Variant of Uncertain Significance, c.2161-6T>C (Intronic), was identified in ABCA4.

autosomal recessive cone-rod dystrophy

- The ABCA4 gene is associated with autosomal recessive cone-rod dystrophy (CRD) (MedGen UID: 349030), Stargardt disease (STGD) (MedGen UID: 383691), and retinitis pigmentosa (RP) (MedGen UID: 400996). Additionally, there is preliminary evidence supporting a correlation with autosomal dominant age-related macular degeneration (ARMD) (PMID: 10880298).
- Not all variants present in a gene cause disease. The clinical significance of the variant(s) identified in this gene is uncertain. Until this uncertainty can be resolved, caution should be exercised before using this result to inform clinical management decisions.
- Familial VUS testing is not offered. Testing family members for this variant will not contribute evidence to allow variant reclassification. Details on our VUS Resolution and Family Variant Testing Programs can be found at https://www.invitae.com/family.

A Variant of Uncertain Significance, c.1076G>A (p.Arg359His), was identified in BBS1.

- The BBS1 gene is associated with autosomal recessive Bardet-Biedl syndrome (MedGen UID: 422452) and non-syndromic retinitis pigmentosa (PMID: 23143442, 27032803, 21520335).
- Not all variants present in a gene cause disease. The clinical significance of the variant(s) identified in this gene is uncertain. Until this uncertainty can be resolved, caution should be exercised before using this result to inform clinical management decisions.
- Familial VUS testing is not offered. Testing family members for this variant will not contribute evidence to allow variant reclassification. Details on our VUS Resolution and Family Variant Testing Programs can be found at https://www.invitae.com/family.

A Variant of Uncertain Significance, c.2682G>A (Silent), was identified in COL11A2.

autosomal recessive Bardet-Biedl syndrome

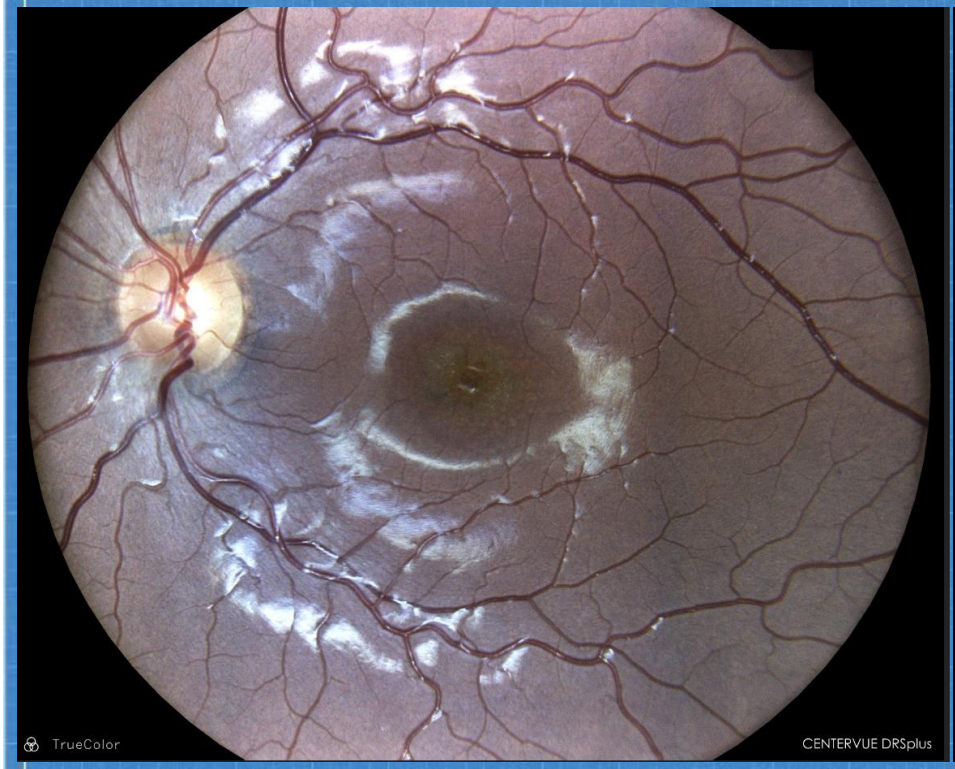
- The COL11A2 gene is associated with a spectrum of related autosomal recessive conditions including nonsyndromic deafness (MedGen UID: 400602), otospondylomegaepiphyseal dysplasia (OSMED) (MedGen UID: 1617409), and fibrochondrogenesis (MedGen UID: 479768). COL11A2 is also associated with a spectrum of related autosomal dominant conditions including Stickler syndrome III (MedGen UID: 349293 and 120521), OSMED (also known as Weissenbacher-Zweymüller syndrome; MedGen UID: 341234) and nonsyndromic deafness (MedGen UID: 400917).
- Not all variants present in a gene cause disease. The clinical significance of the variant(s) identified in this gene is uncertain. Until this uncertainty can be resolved, caution should be exercised before using this result to inform clinical management decisions.
- Familial VUS testing is not offered. Testing family members for this variant will not contribute evidence to allow variant reclassification. Details on our VUS Resolution and Family Variant Testing Programs can be found at https://www.invitae.com/family.

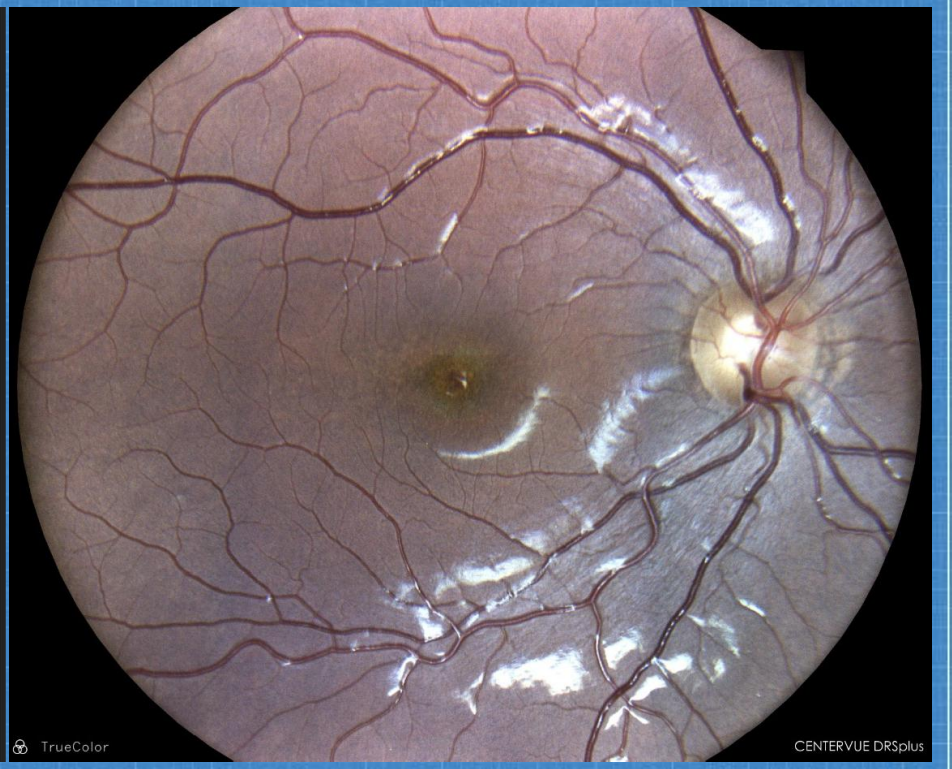
A Variant of Uncertain Significance, c.916A>G (p.Arg306Gly), was identified in PDE6A.

- The PDE6A gene is associated with autosomal recessive retinitis pigmentosa (MedGen UID: 462489). Additionally, the PDE6A gene has preliminary evidence supporting a correlation with autosomal dominant periventricular nodular heterotopia (PMID: 29738522).

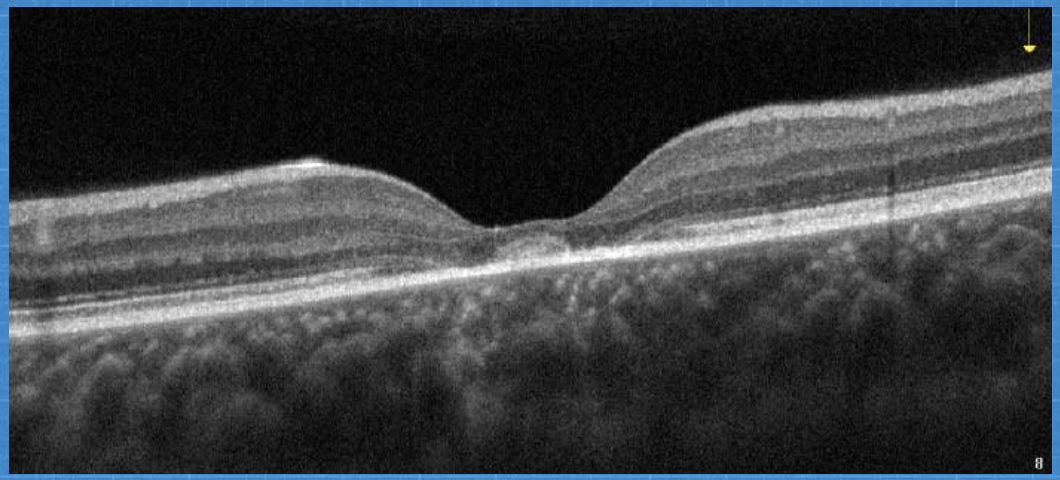
But that's not all!!!!!

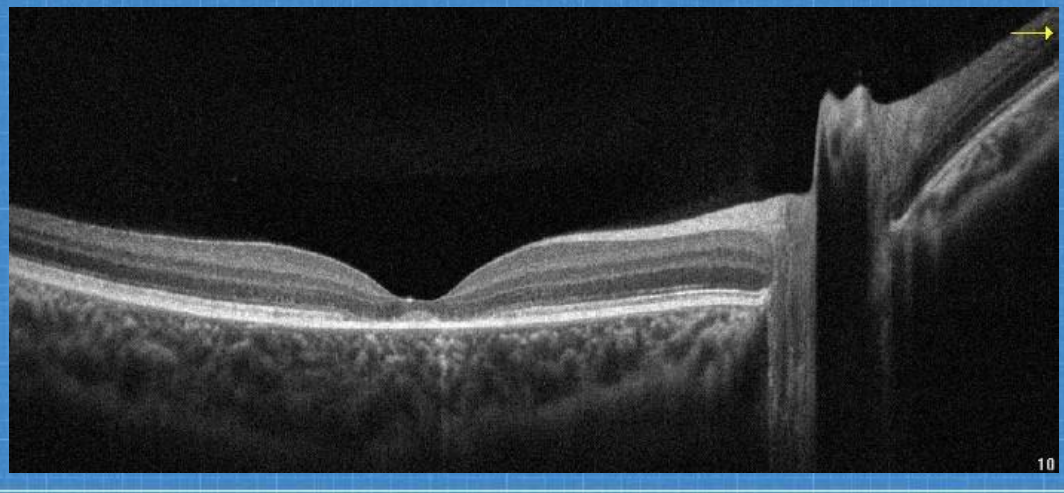
13-year-old twin sister with NORMAL vision came in for an exam also



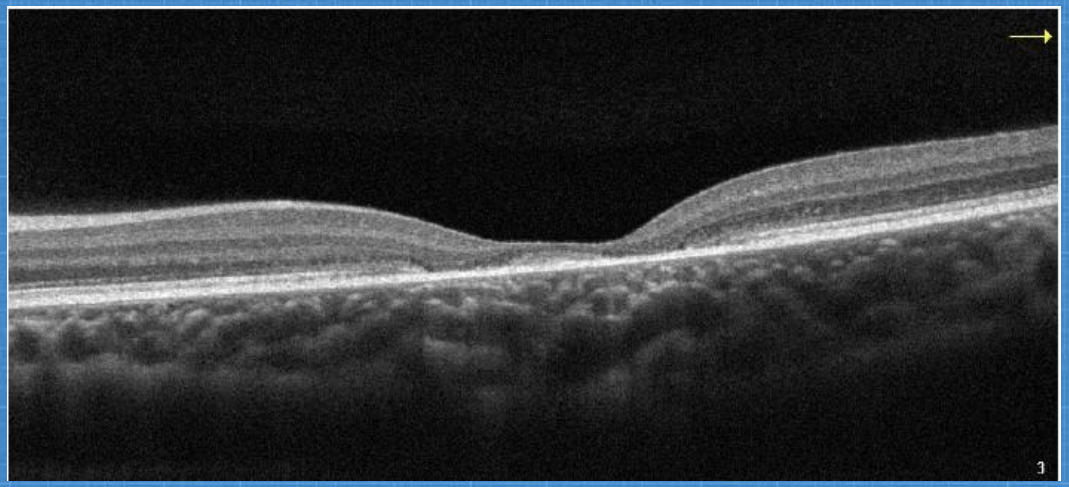


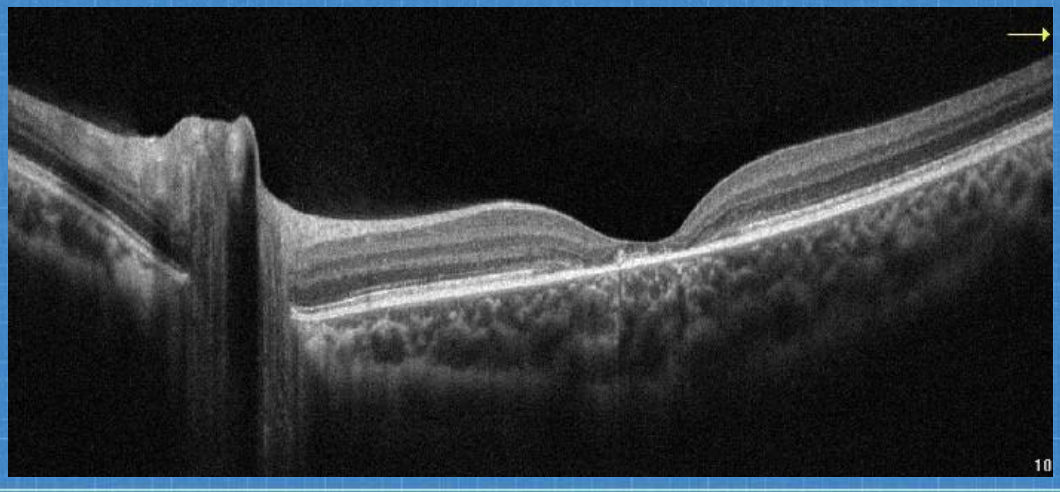
Right Eye

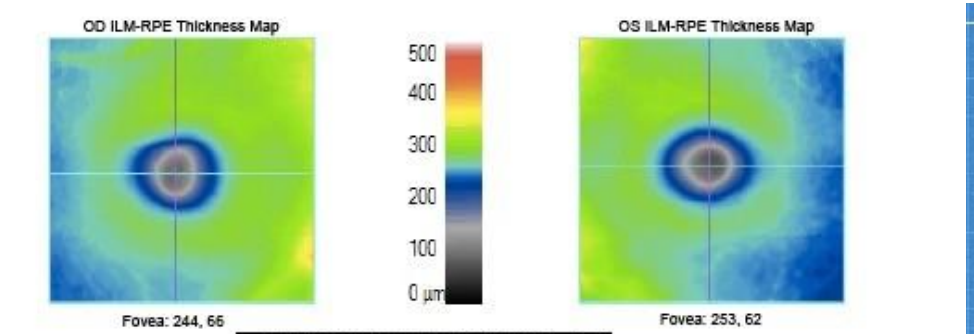




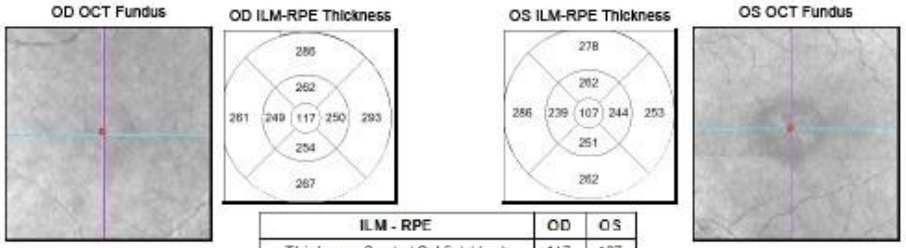
Left Eye



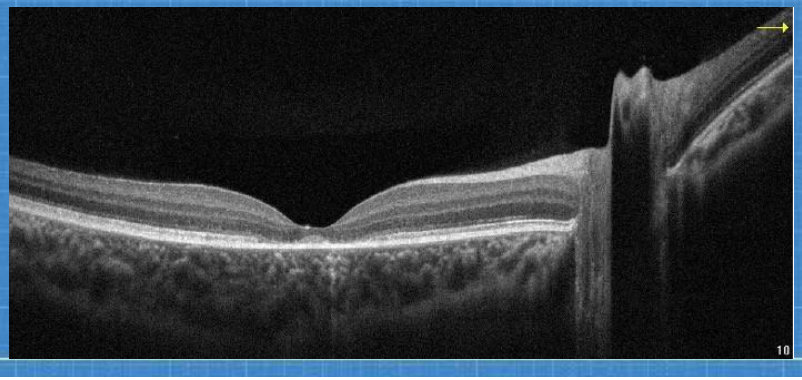


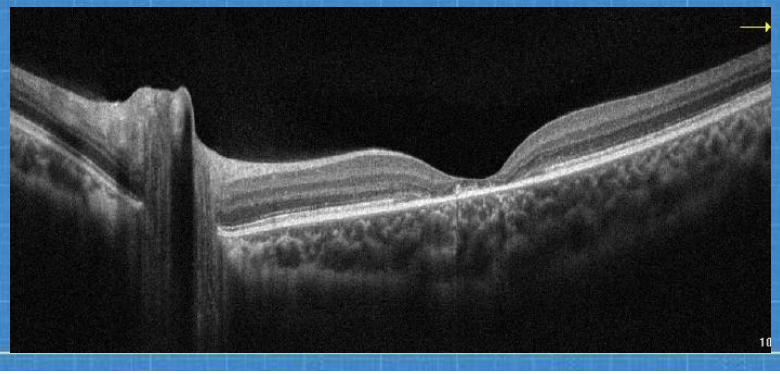


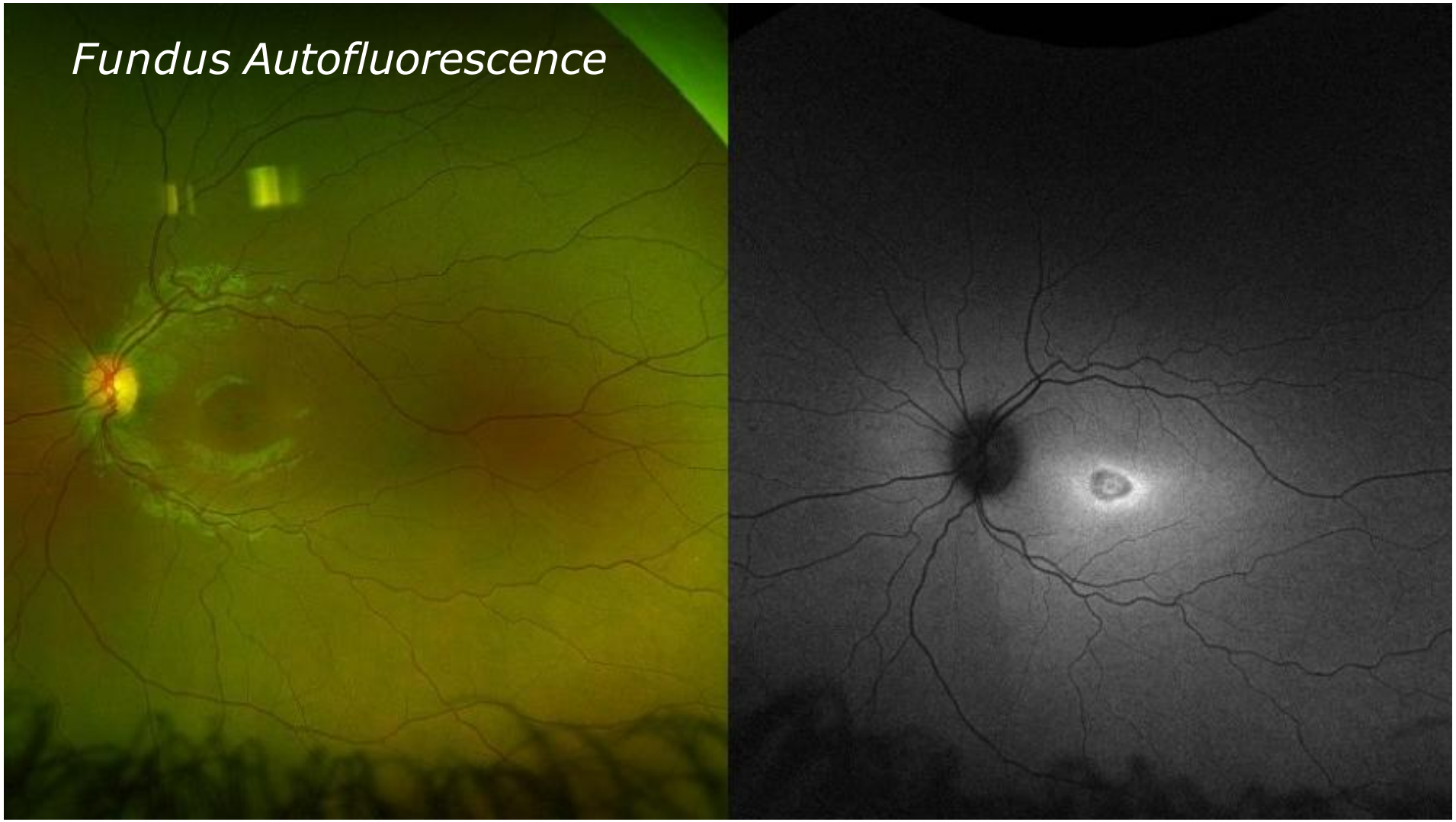
Normative data is not available. Patient age < 18.



ILM - RPE	OD	OS
Thickness Central Subfield (µm)	117	107
Volume Cube (mm³)	9.8	9.5
Thickness Avg Cube (µm)	272	283

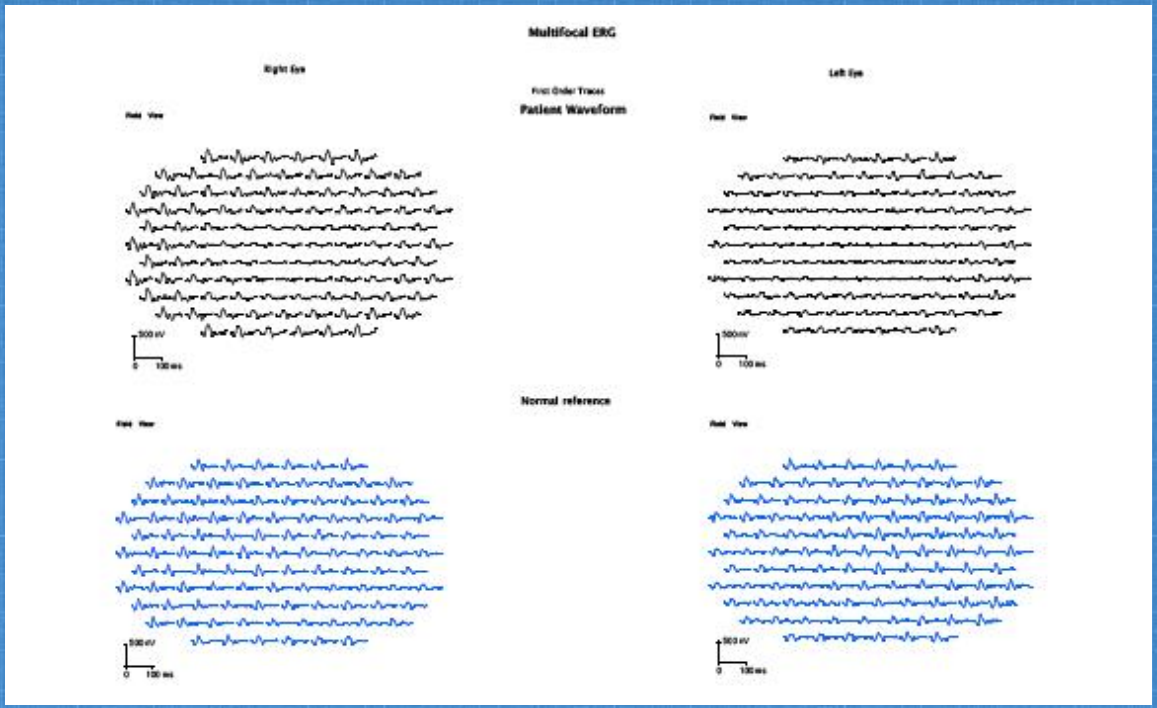





Fundus Autofluorescence

Electrodiagnostics: mfERG

The mfERG findings show moderate diffuse cone dysfunction in the macula area of the right and left eyes.



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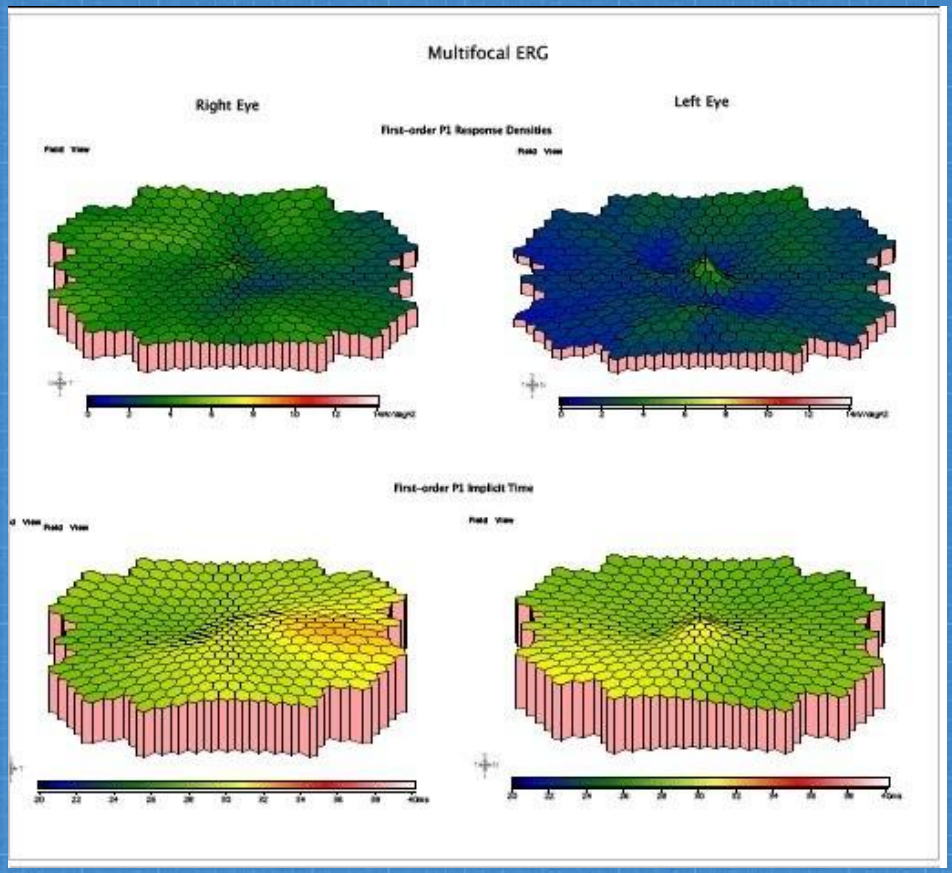


One Pathogenic variant identified in EYS. EYS is associated with autosomal recessive retinitis pigmentosa.

Additional **Variant(s) of Uncertain Significance identified.**

EYS	c6794del (p.Pro2263GInfs*46)	heterozygous	PATHOGENIC
ABCA4	cZJ 61-6T>C (Intronic)	heteraz/gous	Uncertain Significance
BBS1	c 1D76G>A (p.Arg359HJs)	hetc•rcrzygOuS	Uncertain Sfgnificsnce
COLI1AI	c I682G>A (5ileM)	heterazygous	UncertainSignificance
PDE6A	cdl € (pAg306[Iy)	heiernfygous	Uncertain Sfgnificsnce
PDZD7	cI44G>A (pAsp$ZAsn)	heteraz/gous	Uncertain Significance
PEX6	**c1D81A>G (p.Thr361Ala)**	heter ous	Uncertain Sfgnificsnce
RP1	c‹397A>T (p.Glui‹66Val)	heterazygous	UncertainSignificance

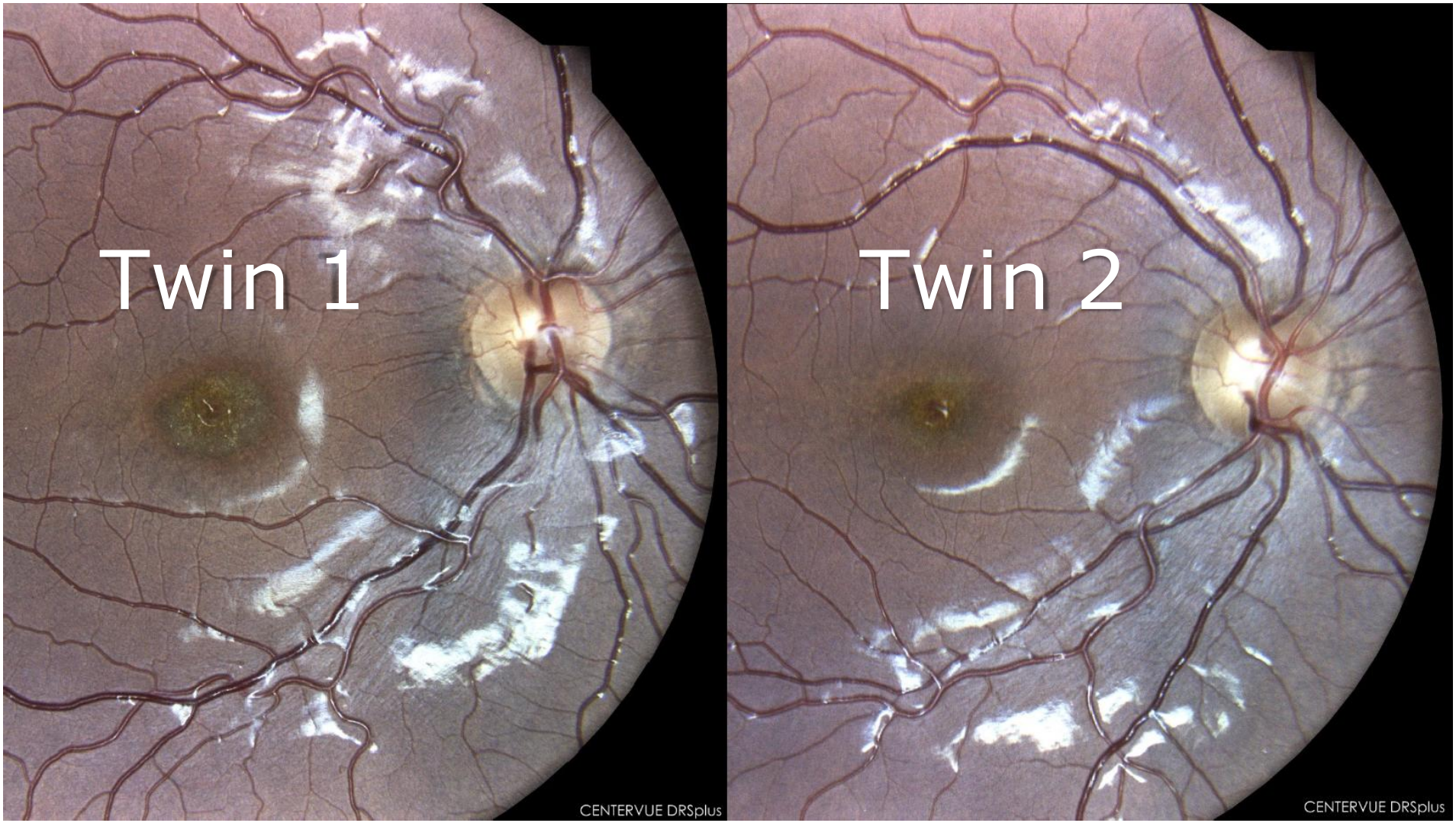
About this test

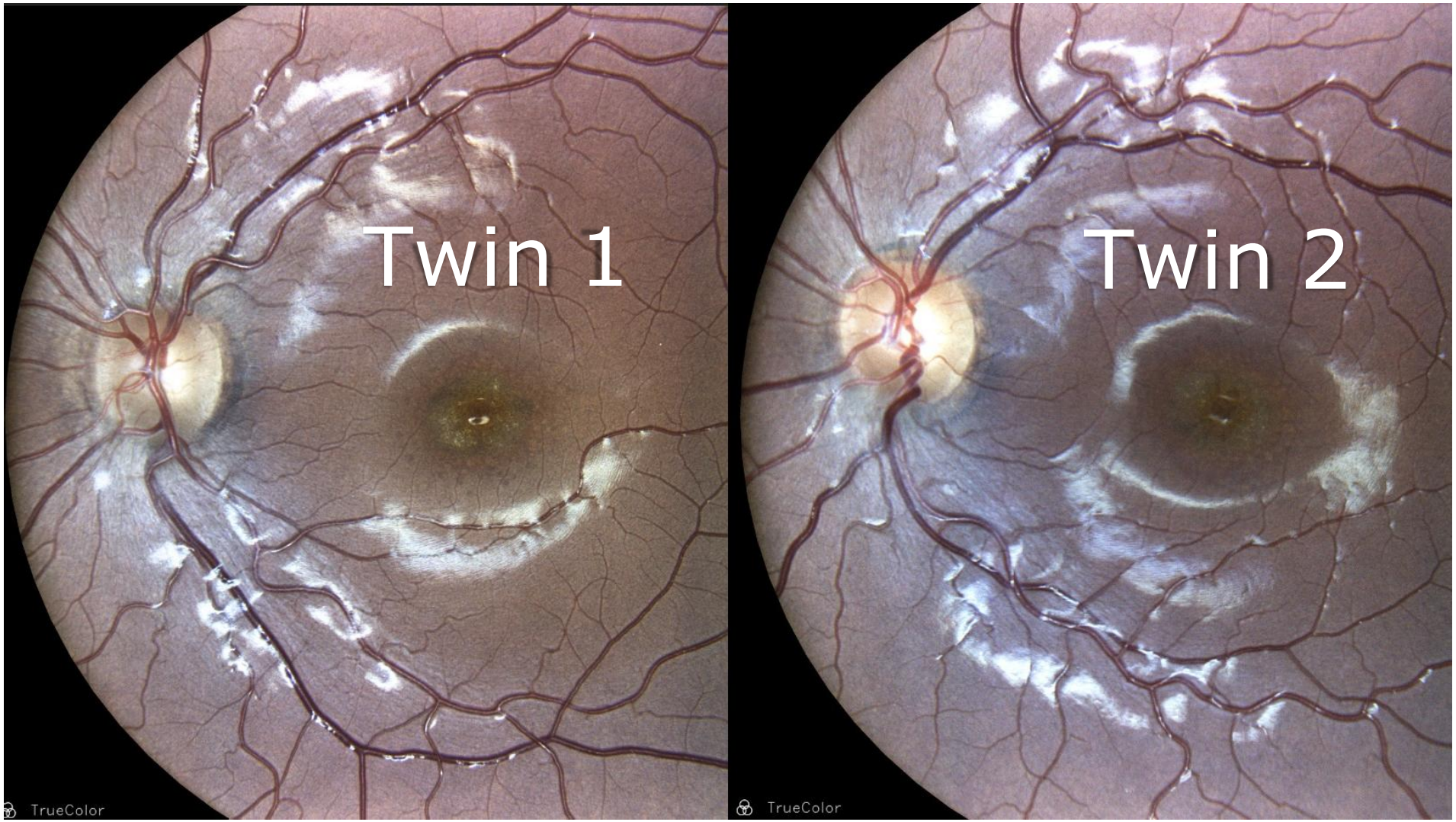
This diagnostic test evaluates 330 gene(s) for variants (genetic changes) that are associated with genetic disorders. Diagnostic genetic testing, when combined with family histoiy and other medical results, may provide information to clarify individual risk, support a clinical diagnosis, and assist with the development ofa personalized treatment and management strategy.

EYS mutations are one of the most common causes of autosomal recessive retinitis pigmentosa in Asia and Europe. Novel findings included the presence of homozygous *EYS* mutations in Cone Rod Dystrophy patients and compound **heterozygous** *EYS* mutations in patients with macular dystrophy.

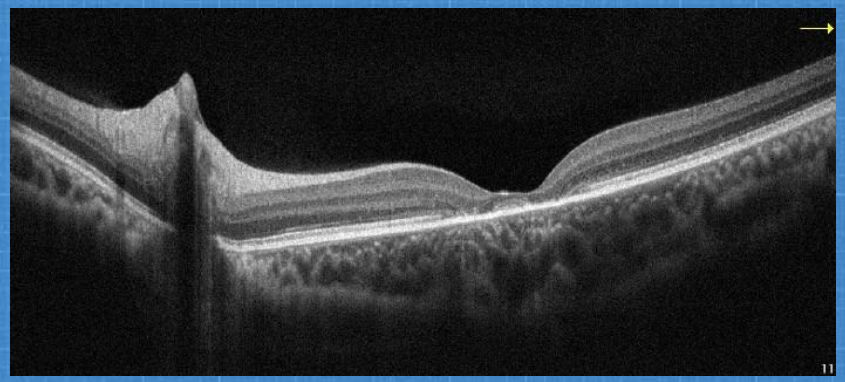
Disease progression of EYS-associated macular dystrophy

Comparison Between Twins

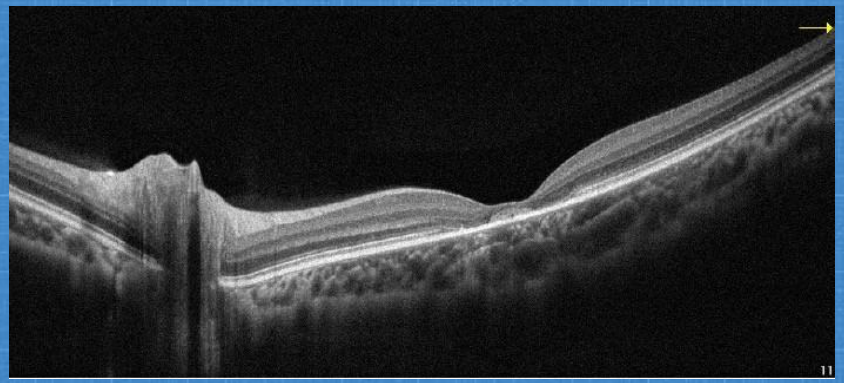

Twin 1
CENTERVUE DRSplus
Twin 2
CENTERVUE DRSplus


Twin 1
Twin 2
TrueColor
TrueColor

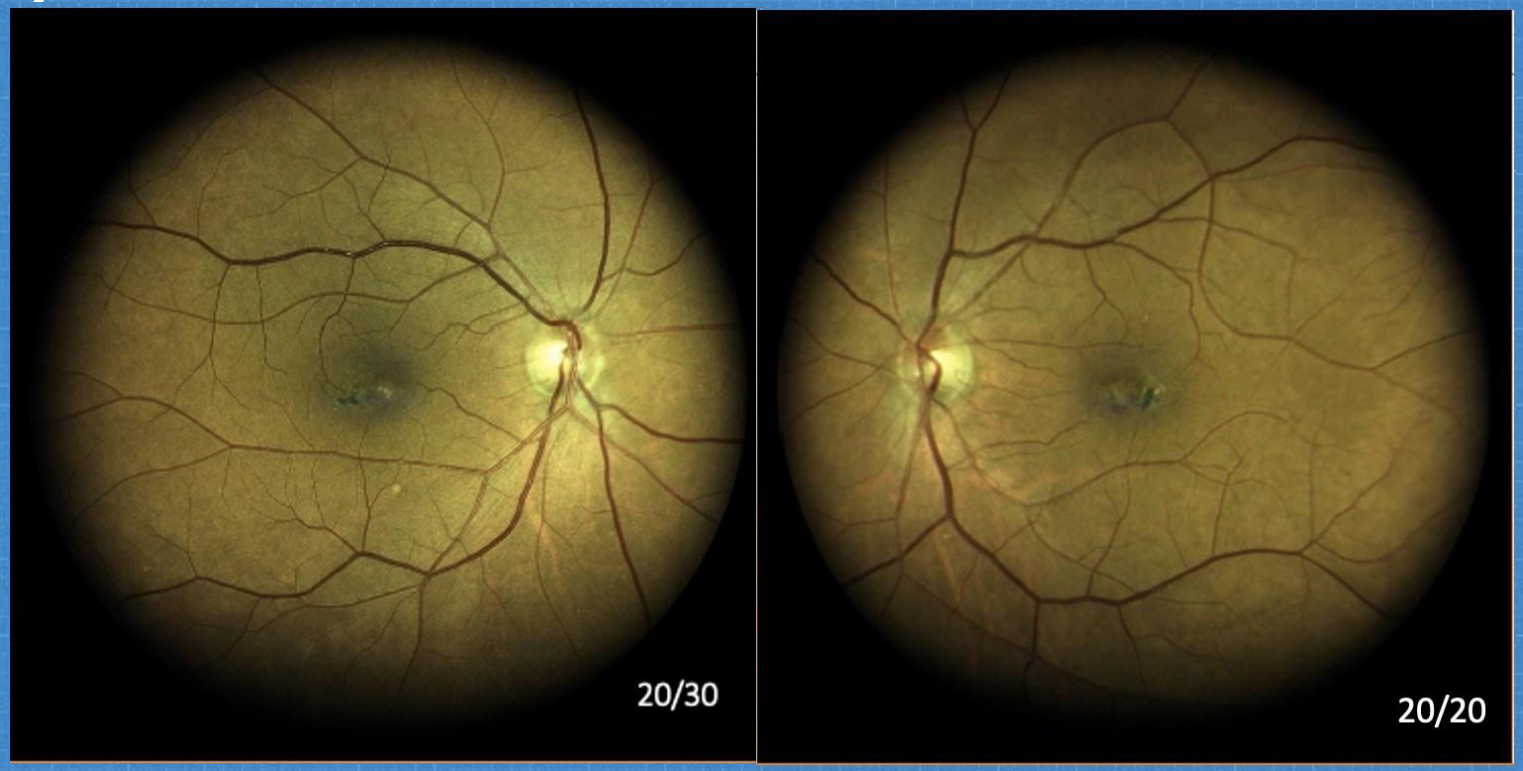
Twin 1



Twin 2



70-year-old Caucasian female



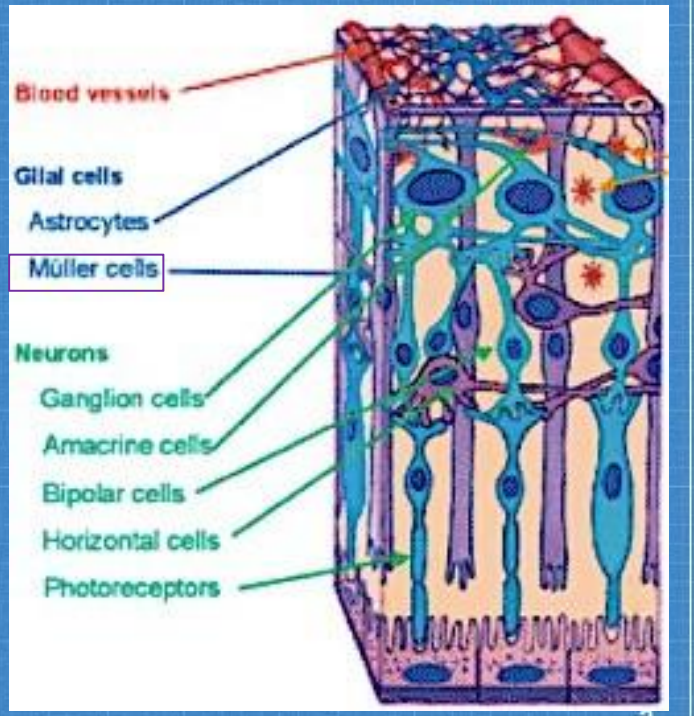
"I have started to see waviness of surfaces and while using HAG; taking AREDS for AMD"

What is Macular Telangiectasia 2?

Proposed hypothesis: Neuro-degenerative disorder

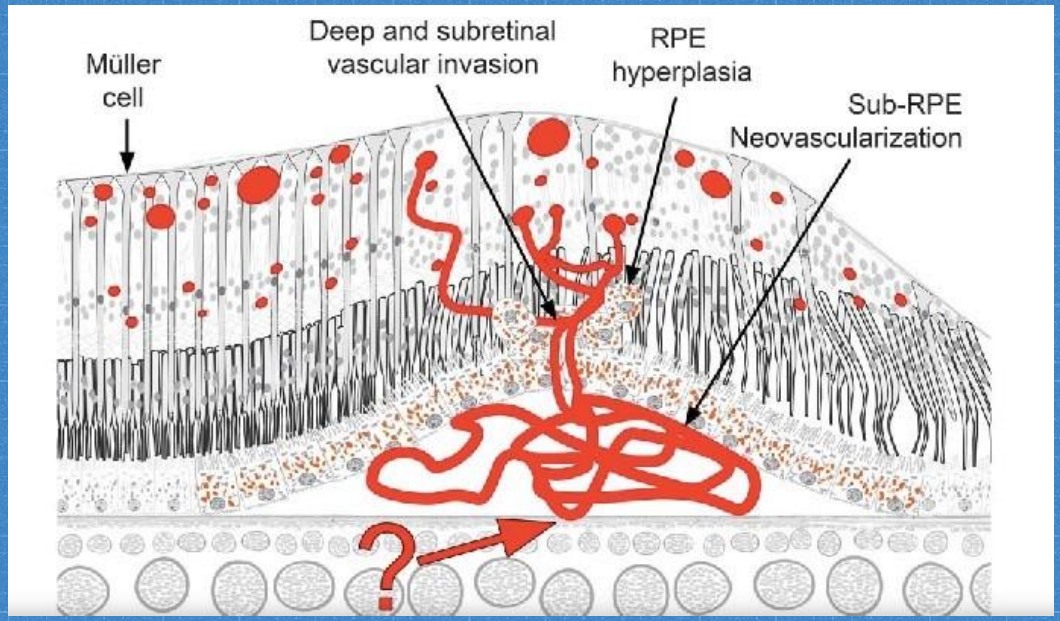
1. *Originates from abnormality in the Muller Cells*

2. *Integrity of retinal vasculature affected*



What is Macular Telangiectasia 2?

Proposed hypothesis: Neurodegenerative disorder

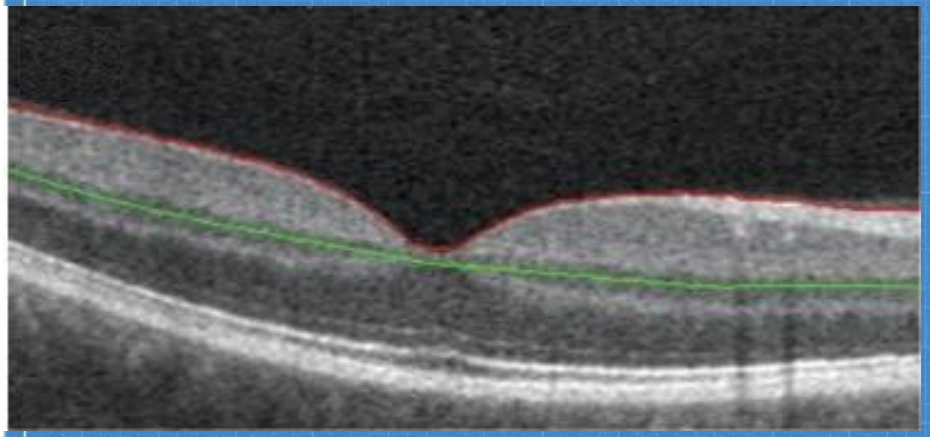


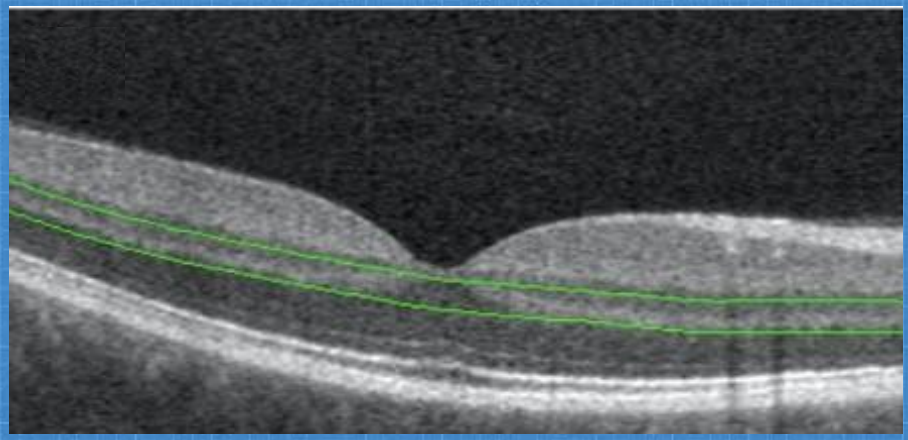
3 *Muller cell depletion: telangiectasia and vessel dilation*

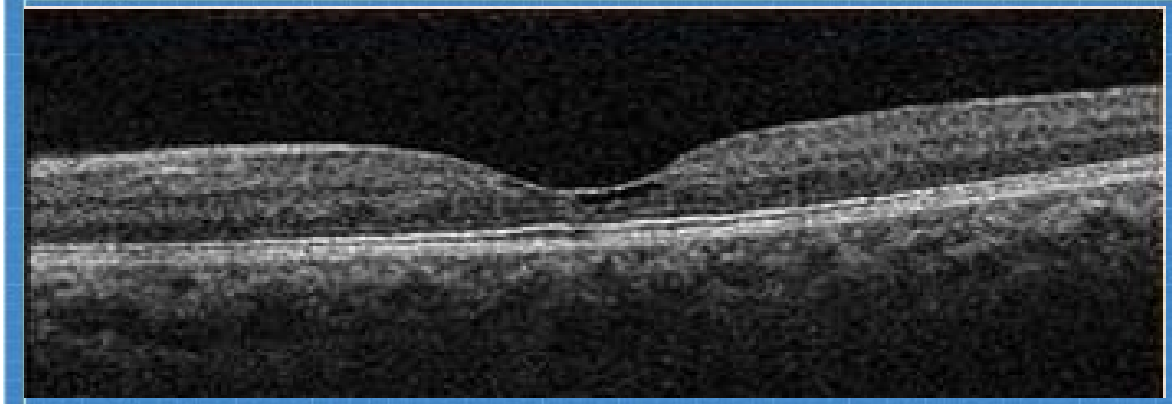
4 *Photoreceptor death*

5 *Weakening of blood retinal barrier*

OCT: Normal

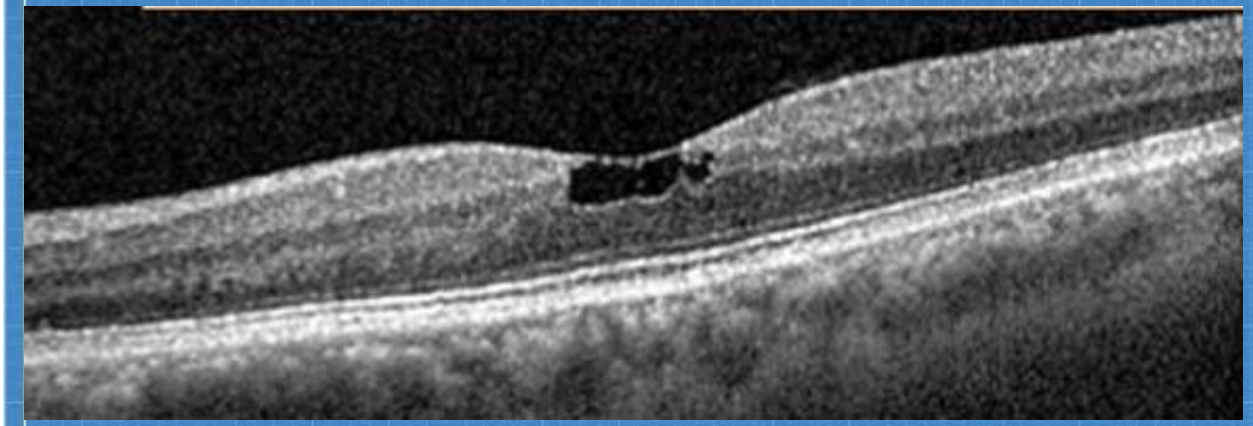




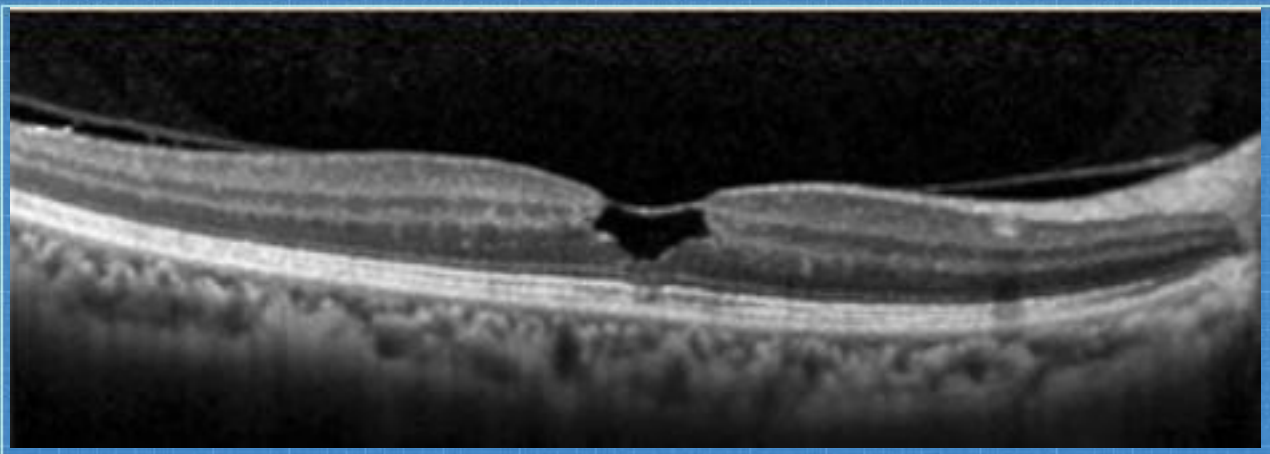


"ILM DRAPE"

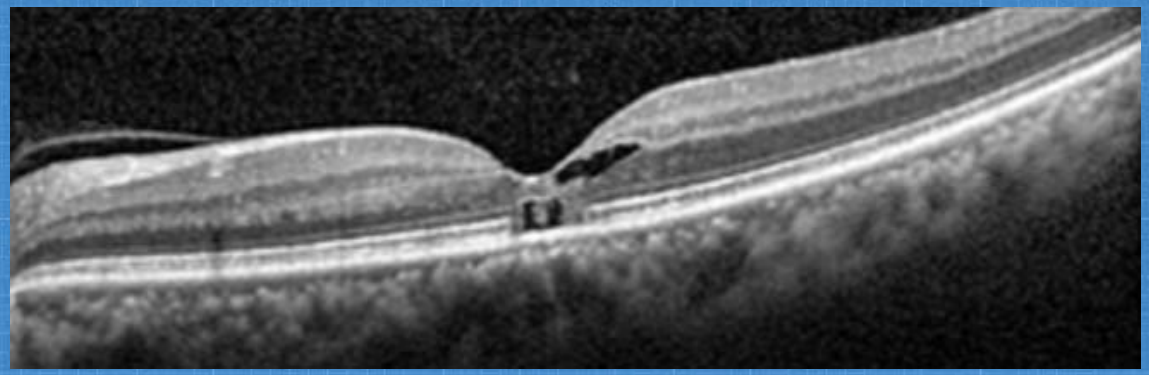
Hypo-reflective cavities in inner retina



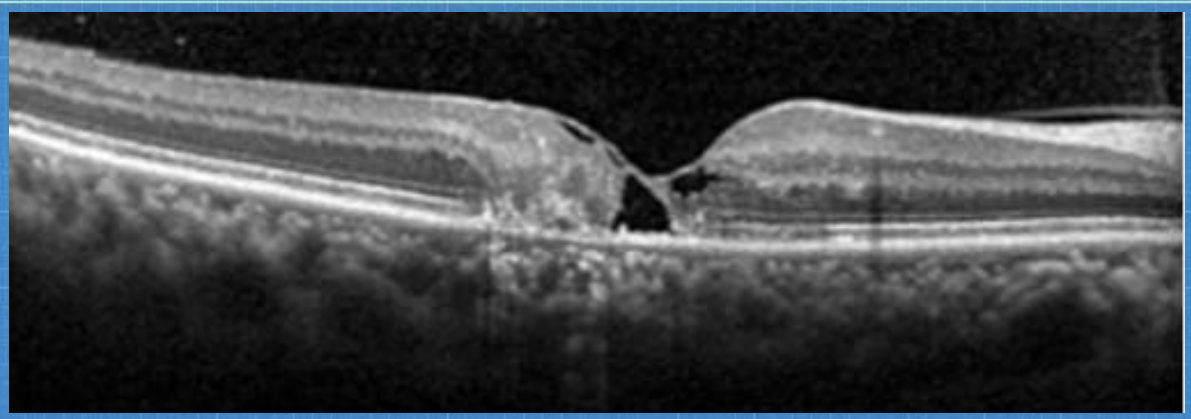
https://www.nature.com/articles/s41598-020-73403-5



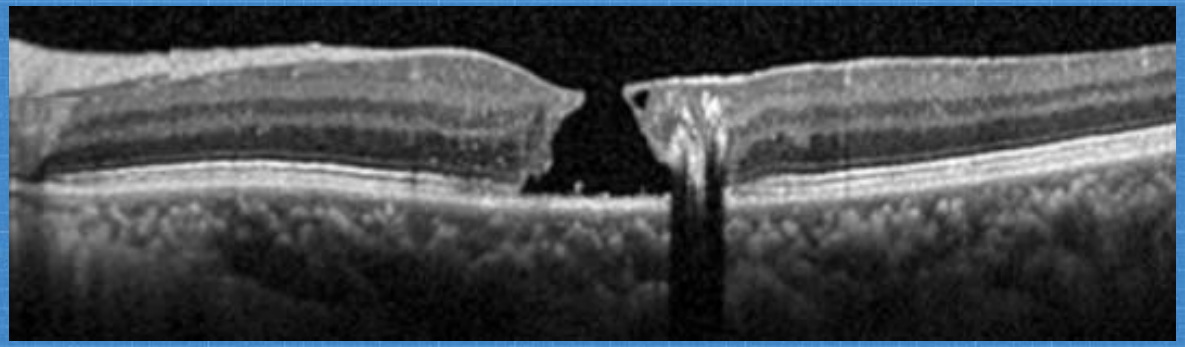
Enlargement of ILM drape



Involvement of outer retina; IS/OS

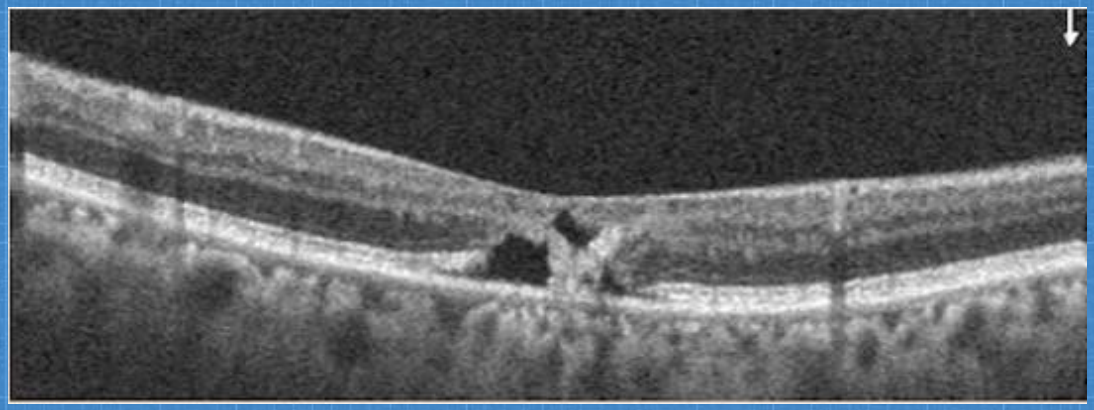


Cavity spans to outer retina



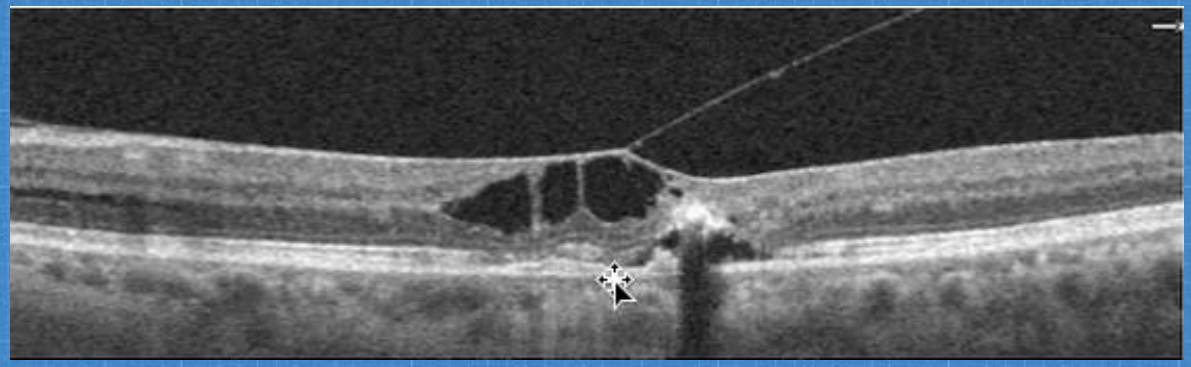
Macular hole formation

https://www.nature.com/articles/s41598-020-73803-9

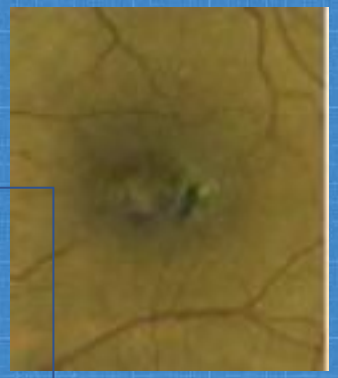


Our patient…..OD

Disruption of EZ
Collapsing of layers
Hyper-reflective lesions in outer retina
Intra and sub-retinal cavitations

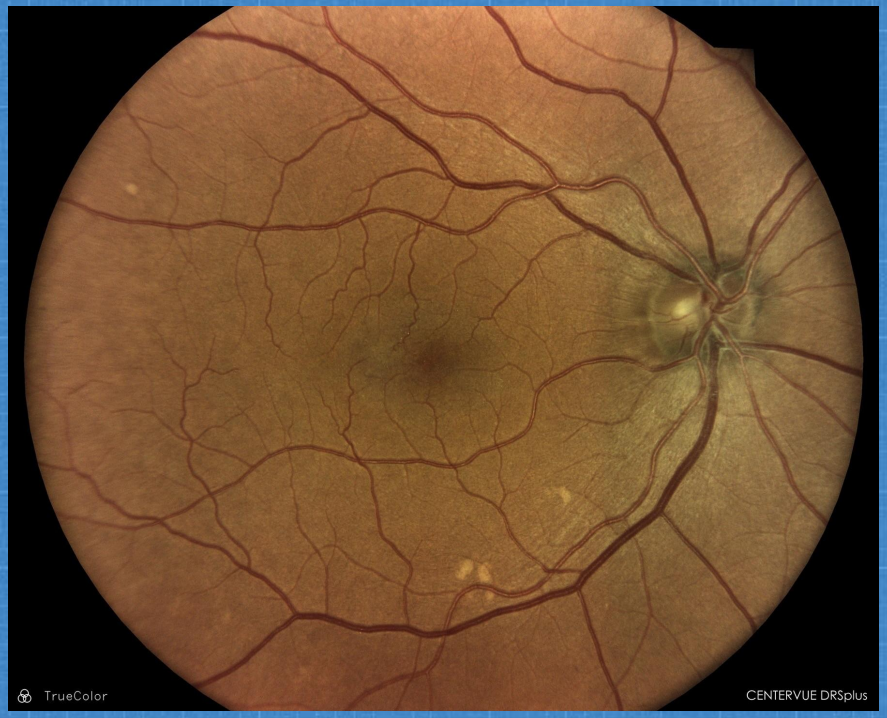


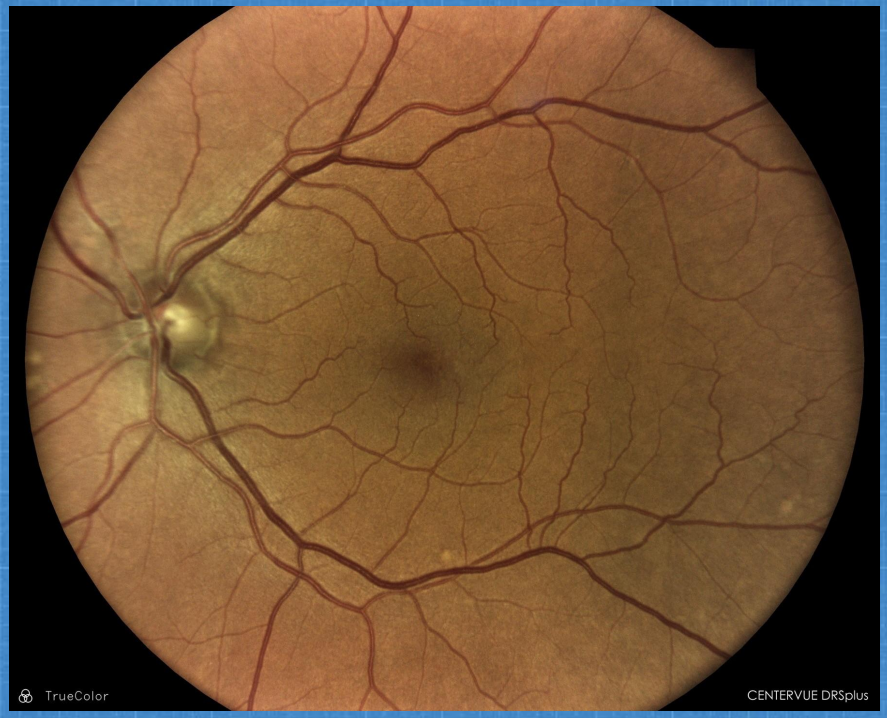
Our patient…..OS



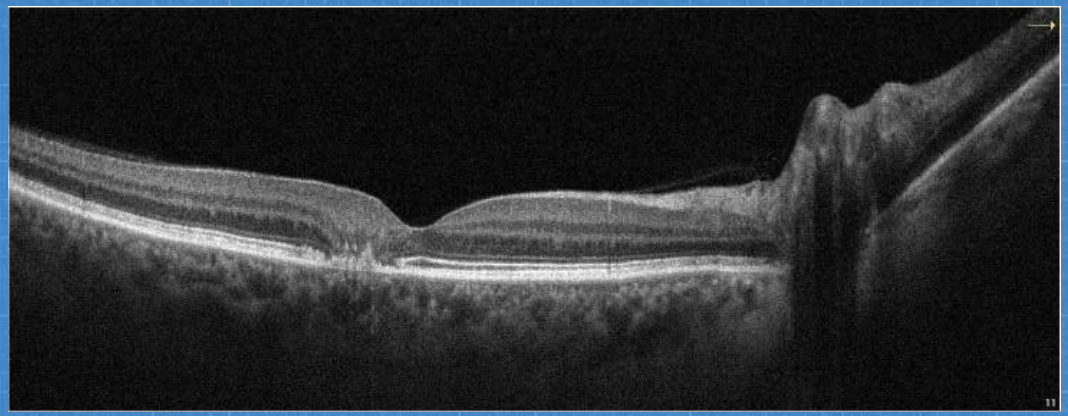
Disruption of EZ
Collapsing of layers
Hyper-reflective lesions in outer retina
Intra and sub-retinal cavitations

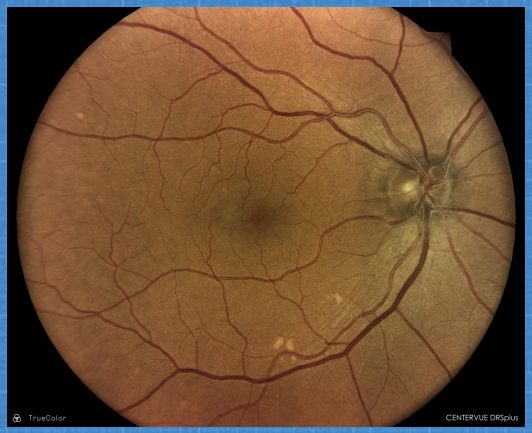
And other…. Not so obvious!!!

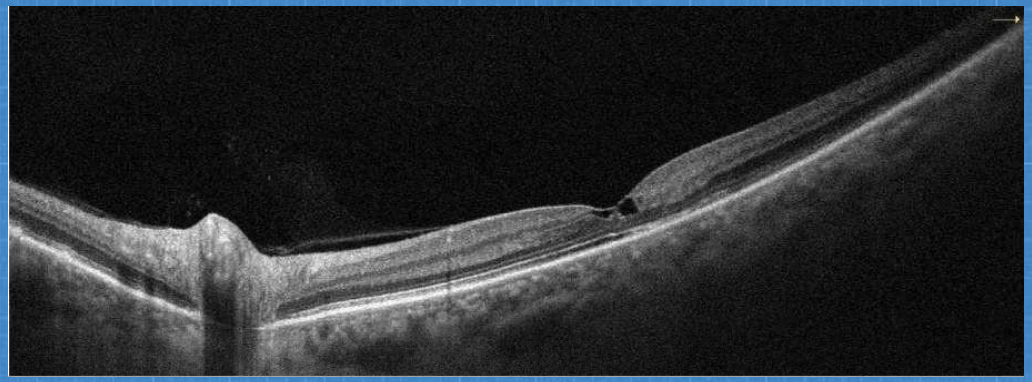


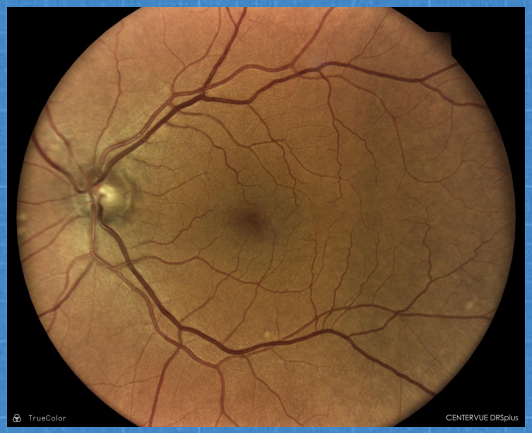


And other…. Not so obvious!!!





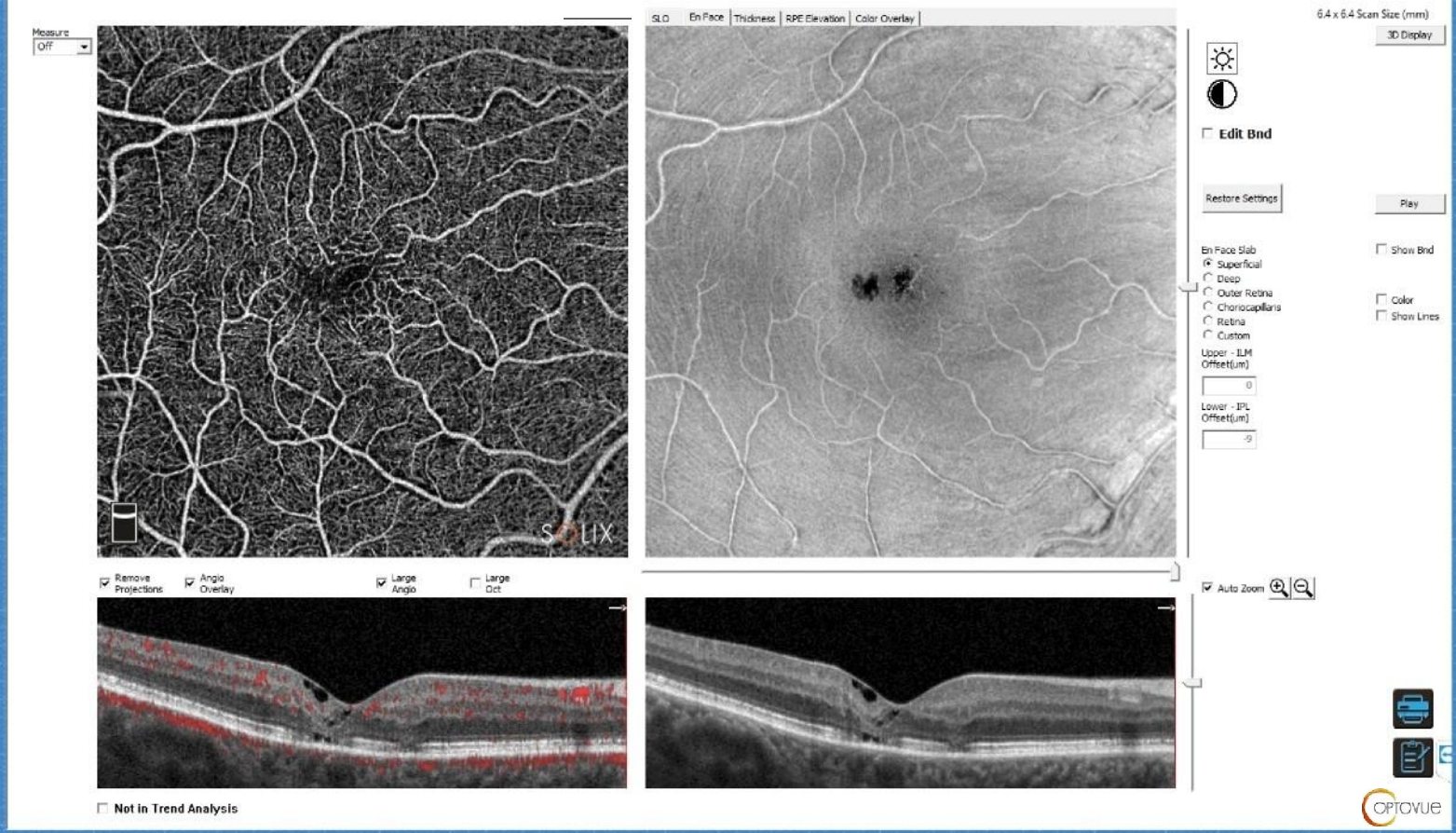


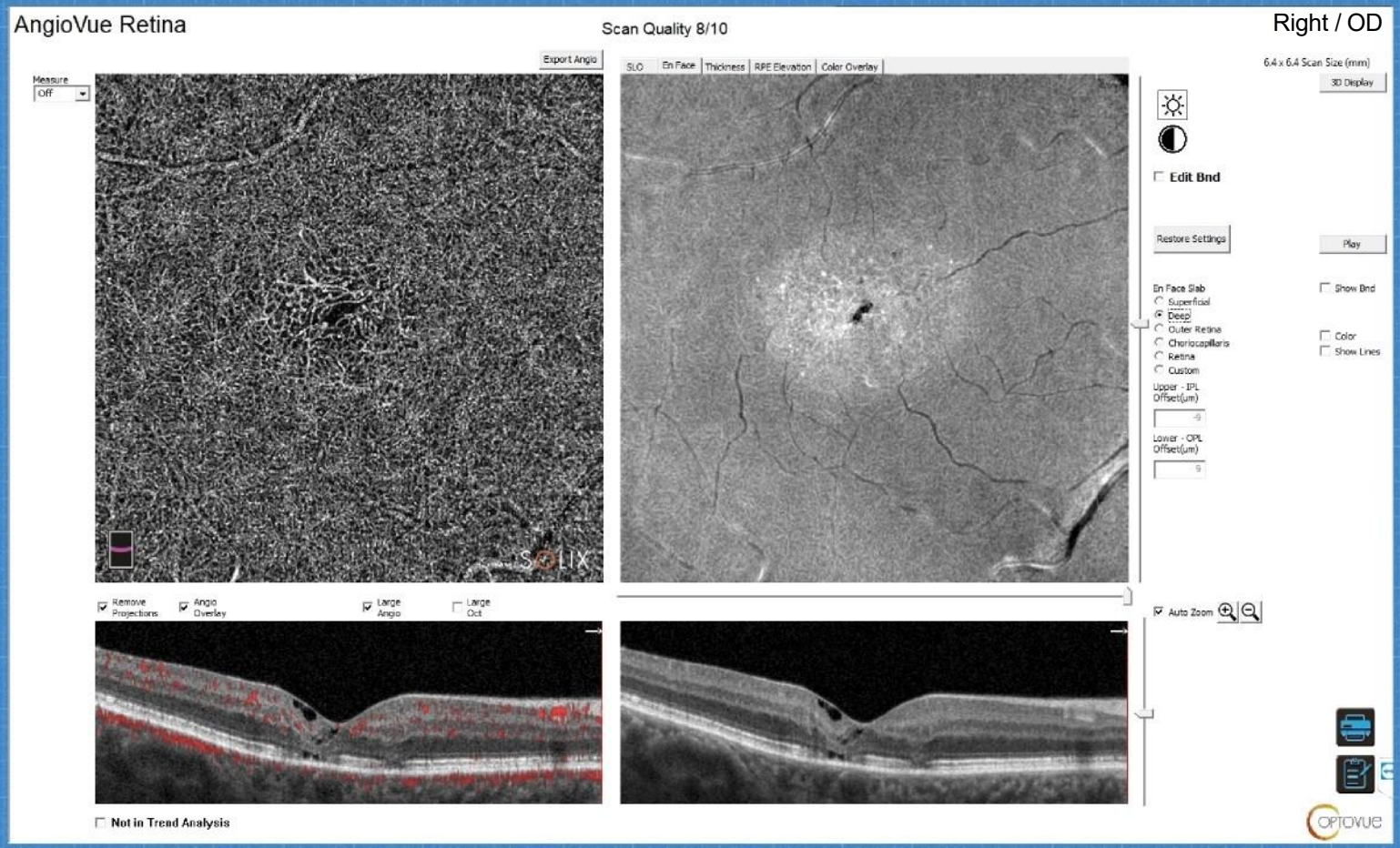


AngioVue Retina

Scan Quality 8/10

Right / OD




AngioVue Retina
Scan Quality 8/10
Right / OD
Export Angio
Measure
Off
SLO
En Face
Thickness
RPE Elevation
Color Overlay
6.4 x 6.4 Scan Size (mm)
3D Display
Edit Bnd
Restore Settings
Play
En Face Slab
Superficial
Deep
Outer Retina
Choriocapillaris
Retina
Custom
Upper - IPL Offset(um)
Lower - OPL Offset(um)
Show Bnd
Color
Show Lines
Remove Projections
Angio Overlay
Large Angio
Large Oct
Auto Zoom
Not in Trend Analysis
OPTOVUE

65-year-old Caucasian Female

- Complaints of "central darkening" OU
  - Progressive worsening
- History of rheumatoid arthritis (20+ years)

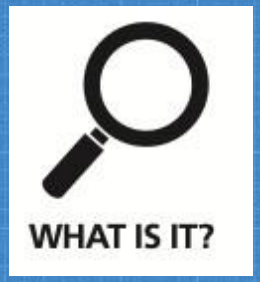
- BCVA:
  - OD 20/40; OS 20/40-

Medications:

- *Methotrexate*
- *Plaquenil: 400 mg x 20 years*

What is Hydroxychloroquine (Plaquenil)?

- Disease-modifying anti-rheumatic drug (DMARD)
- Originally anti-malarial

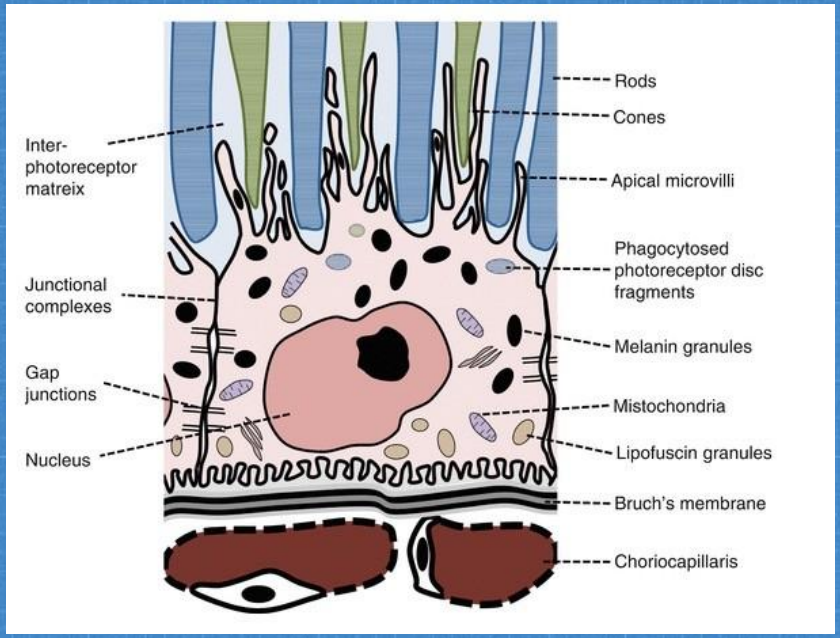


- Used to treat rheumatoid arthritis, lupus, and other inflammatory and dermatologic conditions

What is Hydroxychloroquine (Plaquenil)?

- Metabolite of chloroquine
- Longer half life
  - Less drug needed for efficacy

- Binds to melanin in RPE
  - Results in Bulls-Eye Maculopathy



Recommendations on Screening for Chloroquine and Hydroxychloroquine Retinopathy (2016 Revision)

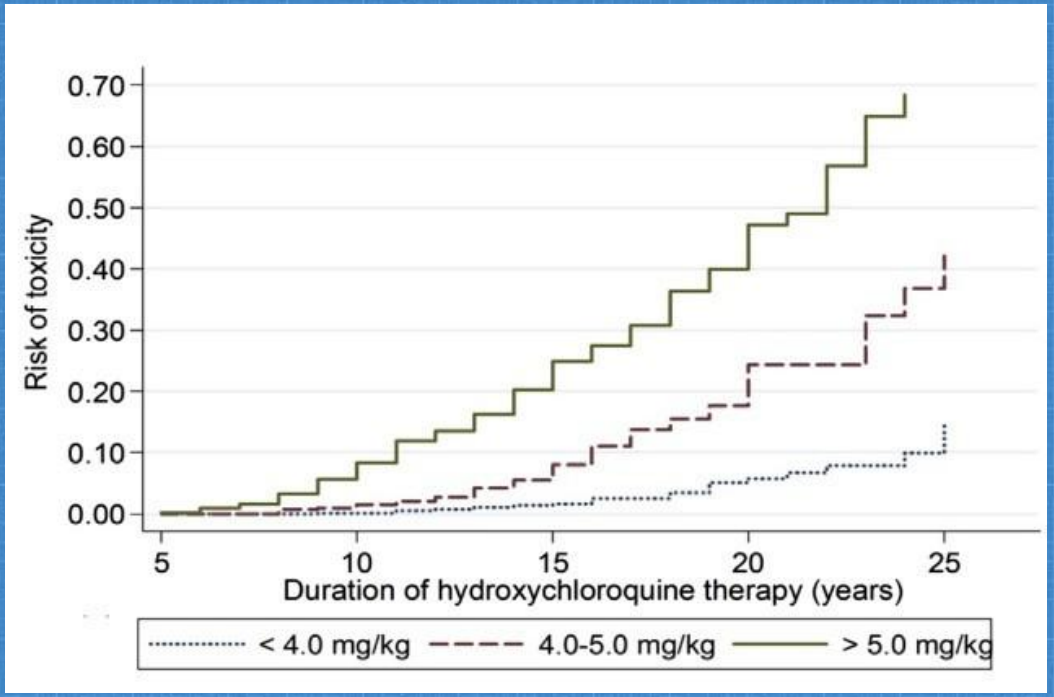
AMERICAN ACADEMY™ OF OPHTHALMOLOGY

- ***Dose:***
  - Maximum daily HCQ use of ≤5.0 mg/kg real weight
- ***Duration:***
  - At recommended dosage, risk of toxicity up to 5 years is under 1% and up to 10 years is under 2%... **BUT 20% AFTER 20 YEARS!!!**

High dose and long duration of use are most significant risk factors

Recommendations on Screening for Chloroquine and Hydroxychloroquine Retinopathy (2016 Revision)

AMERICAN ACADEMY™ OF OPHTHALMOLOGY



JAMA Ophthalmol 2014;132:1453e60

Recommendations on Screening for Chloroquine and Hydroxychloroquine Retinopathy (2016 Revision)

AMERICAN ACADEMY™ OF OPHTHALMOLOGY

- ***Major Risk Factors:***
  - Concomitant renal disease
    - Subnormal glomerular filtration rate
  - Concomitant Drugs
    - Tamoxifen Use

Retinopathy is not reversible!!

Recommendations on Screening for Chloroquine and Hydroxychloroquine Retinopathy (2016 Revision)

AMERICAN ACADEMY™ OF OPHTHALMOLOGY

Screening Schedule

- Baseline Screening
  - Fundus examination within first year of use
  - Add VFs and OCT if maculopathy is present
- Annual Screening
  - Begin after 5 years of use
  - Sooner in the presence of major risk factors

Sunday	Monday	Tuesday	Wednesday	Thursday	Friday	Saturday
						1
2	3	4	5	6	7	8
9	10	11	12	13	14	15
16	17	18	19	20	21	22
23	24	25	26	27	28	29

JAMA Ophthalmol 2014;132:1453e60

Risk of Plaquenil Maculopathy

Step 1: Evaluate the dosage:

- ***Dose:***
  - Maximum daily HCQ use of $\leq$ 5.0 mg/kg real weight

150 lb. converts to 68 kg

400mg/68 kg =5.88 mg/kg

Risk of Plaquenil Maculopathy

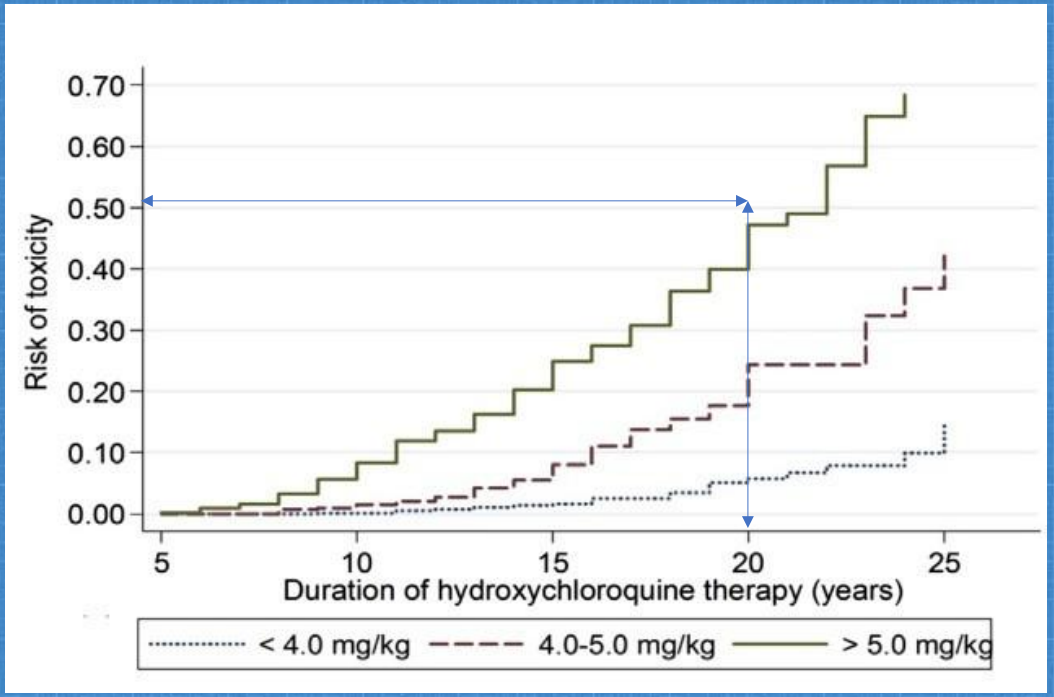
Step 2: Evaluate the duration:

- ***Duration:***
  - At recommended dosage, risk of toxicity up to 5 years is under 1% and up to 10 years is under 2%... **BUT 20% AFTER 20 YEARS!!!**

20 years of use and dosage higher than recommended

Recommendations on Screening for Chloroquine and Hydroxychloroquine Retinopathy (2016 Revision)

AMERICAN ACADEMY™ OF OPHTHALMOLOGY



JAMA Ophthalmol 2014;132:1453e60

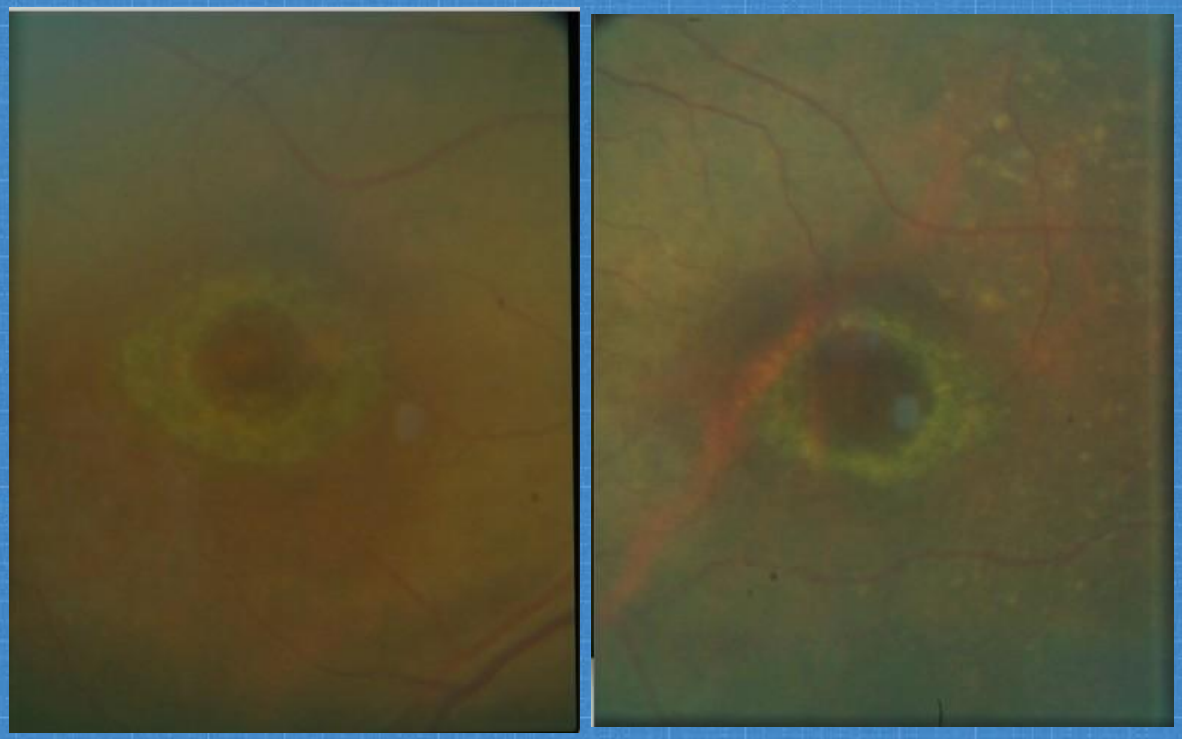
Risk of Plaquenil Maculopathy

Step 3: Assess other major risk factors:

- ***Major Risk Factors:***
  - Concomitant renal disease
    - Subnormal glomerular filtration rate
  - Concomitant Drugs
    - Tamoxifen Use

This patient does not have any other major risk factors

Dilated Fundus Examination

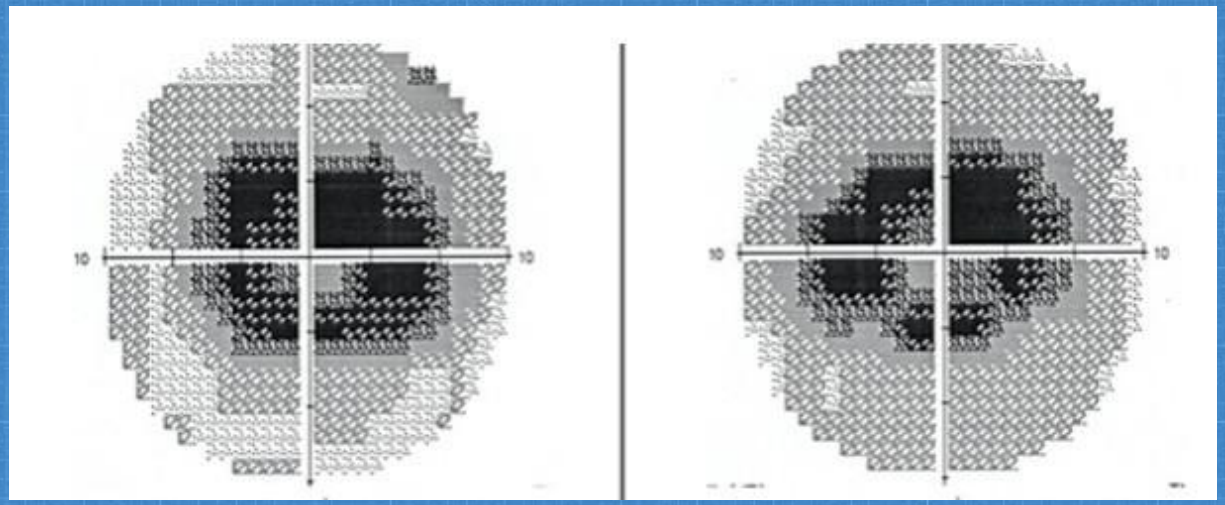


Ring of parafoveal RPE depigmentation sparing fovea

"Bull's Eye Maculopathy"

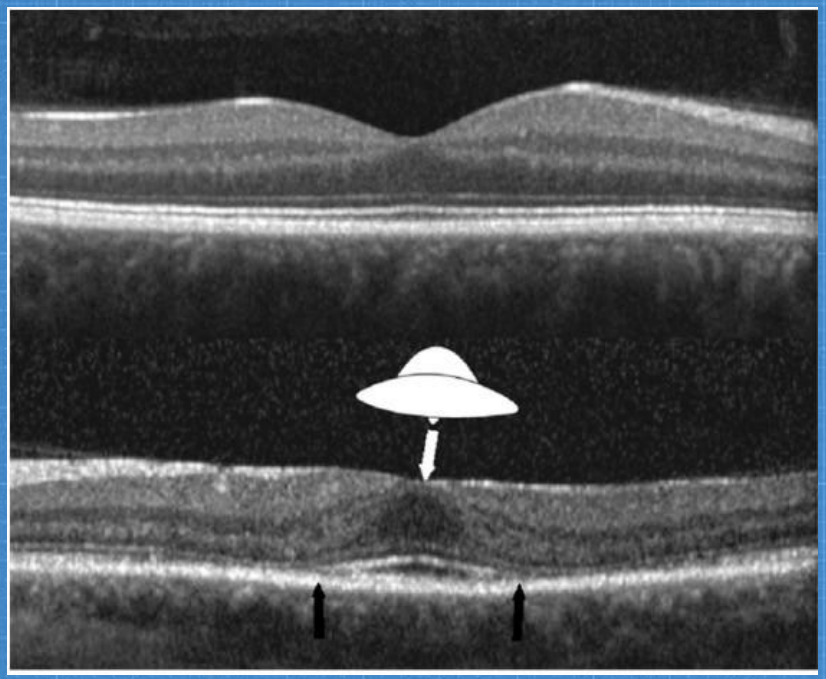
Ancillary Testing

10-2 Humphrey Visual Field



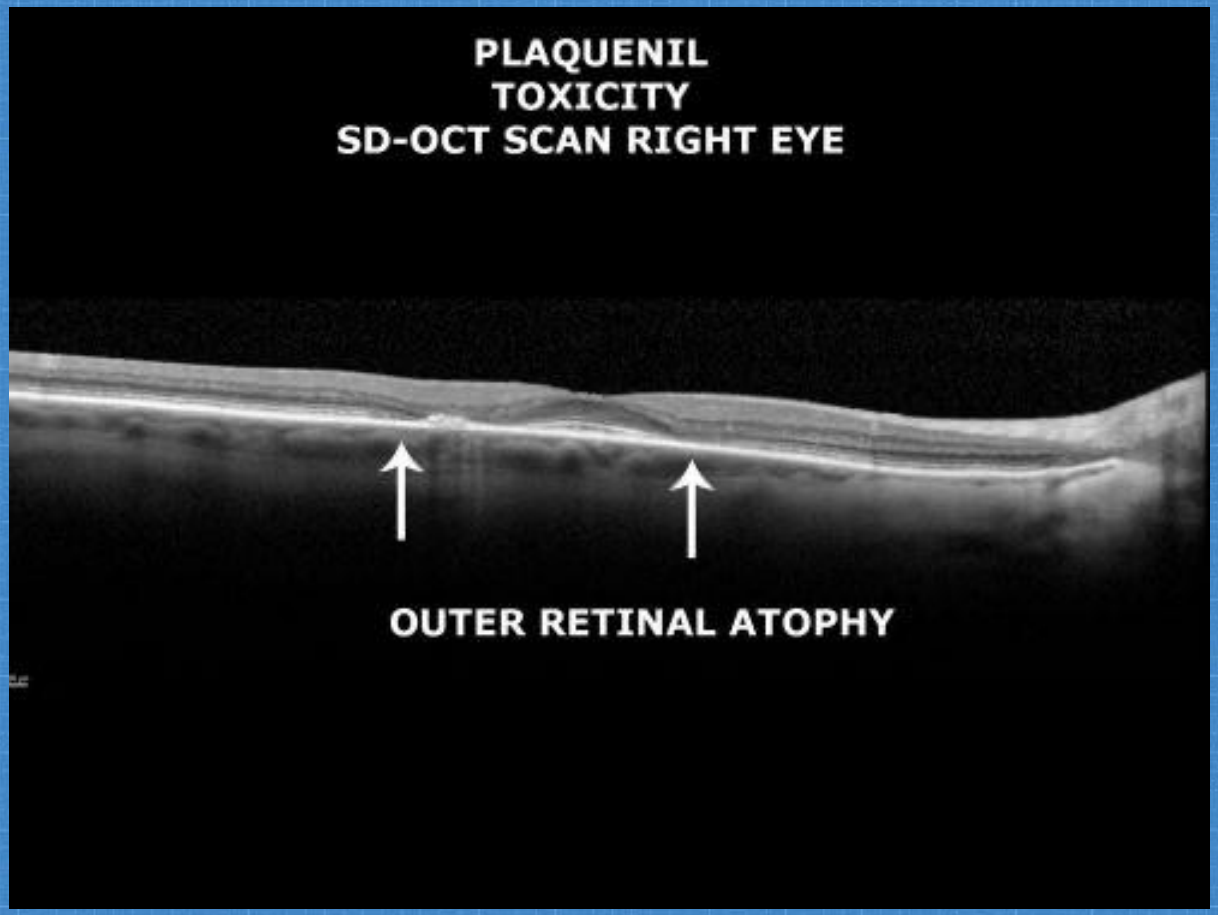
Dense ring scotoma bilaterally

Ancillary Testing: Optical Coherence Tomography

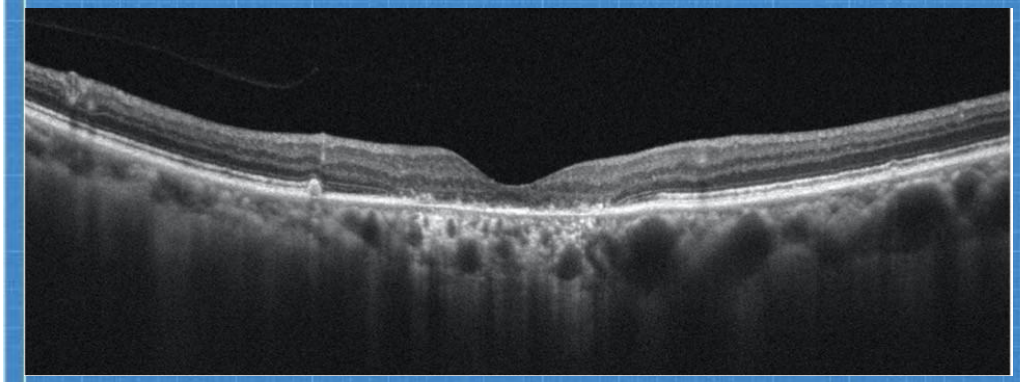


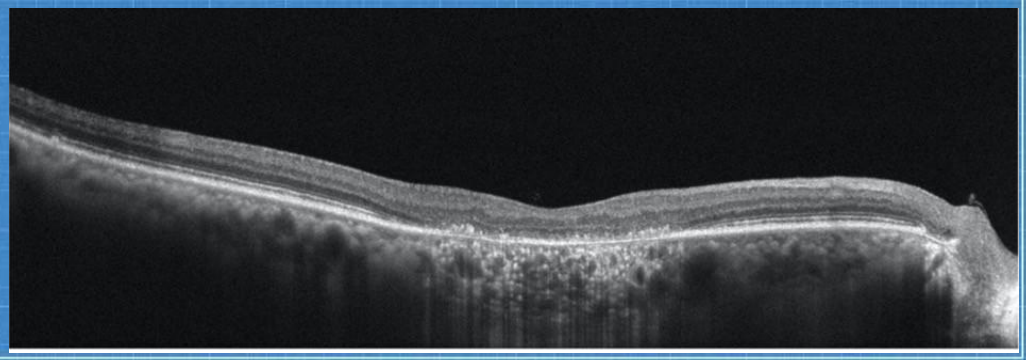
https://www.researchgate.net/figure/Top-Normal-Spectralis-spectral-domain-optical-coherence-tomography-SD-OCT-image-with_fig3_49602243

Recommendations on Screening for Chloroquine and Hydroxychloroquine Retinopathy (2016 Revision)

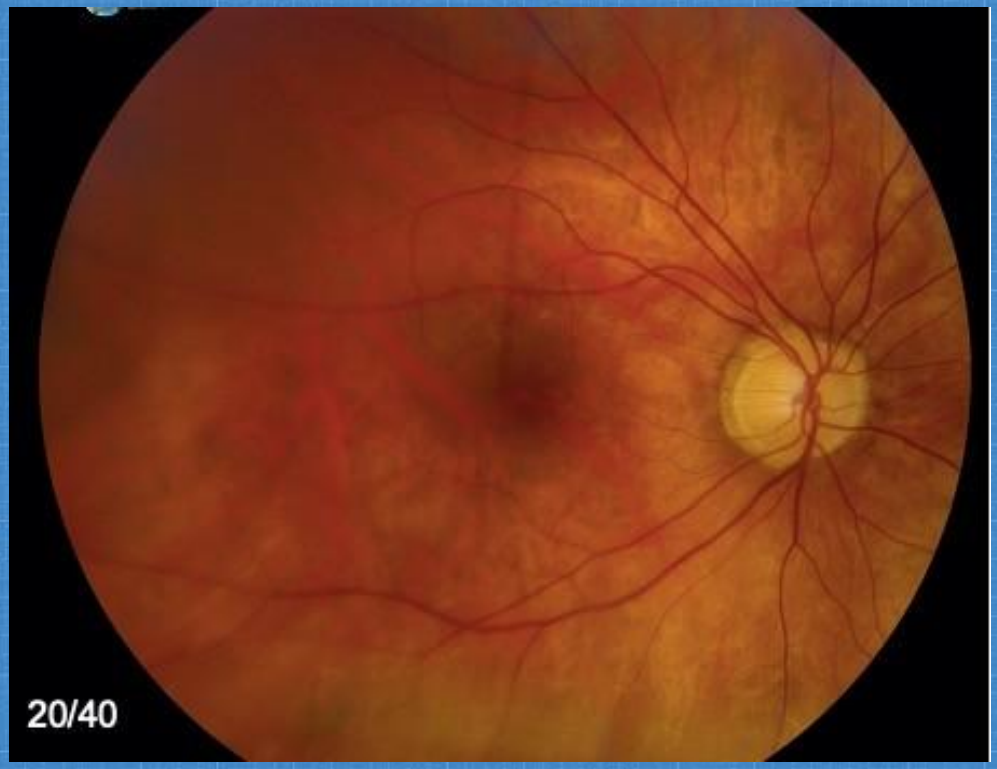

PLAQUENIL
TOXICITY
SD-OCT SCAN RIGHT EYE
OUTER RETINAL ATOPHY

Ancillary Testing: This patient



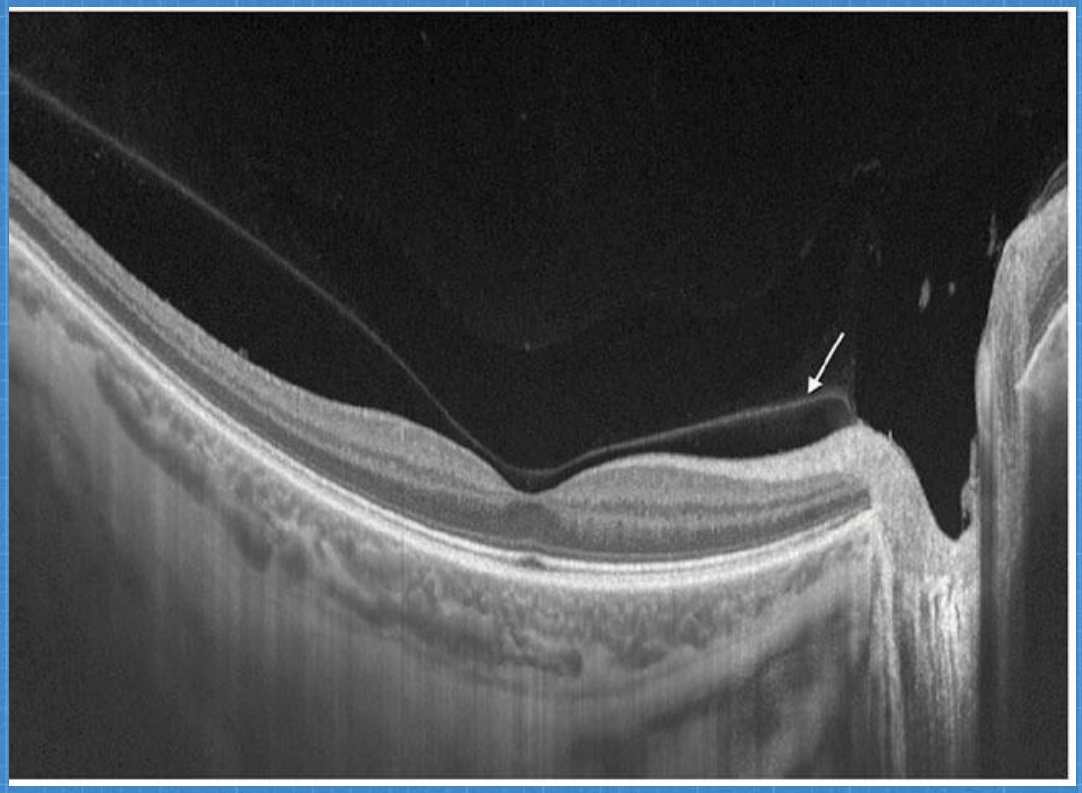


73 year-old female



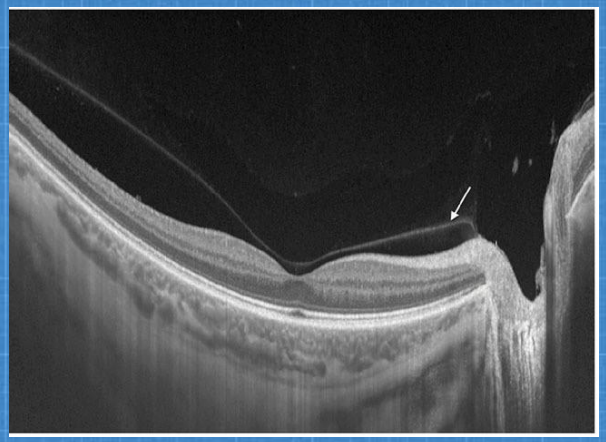
"I feel a film of cobwebs over my right eye. It started a year ago but is progressively getting worse. I had cataract surgery in both eyes a few years ago.. Is it related??"

73 year-old female



20/40

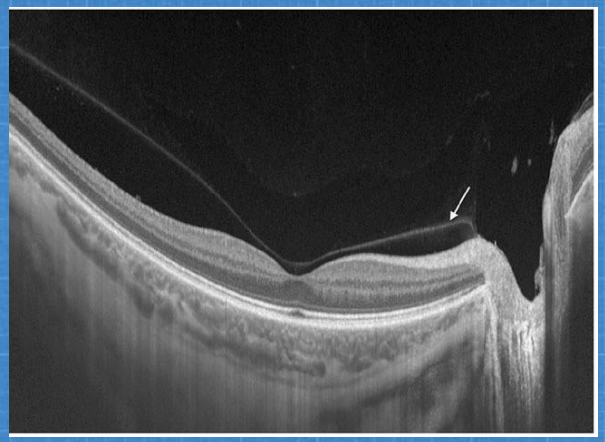
73 year-old female



The above OCT illustrates the following:

1. Complete PVD
2. There is partial detachment of the posterior hyaloid from the central fovea with persistent attachment at the ONH
3. There is vitreomacular traction resulting in obscuration of the foveal contour

73 year-old female



The above OCT illustrates the following:

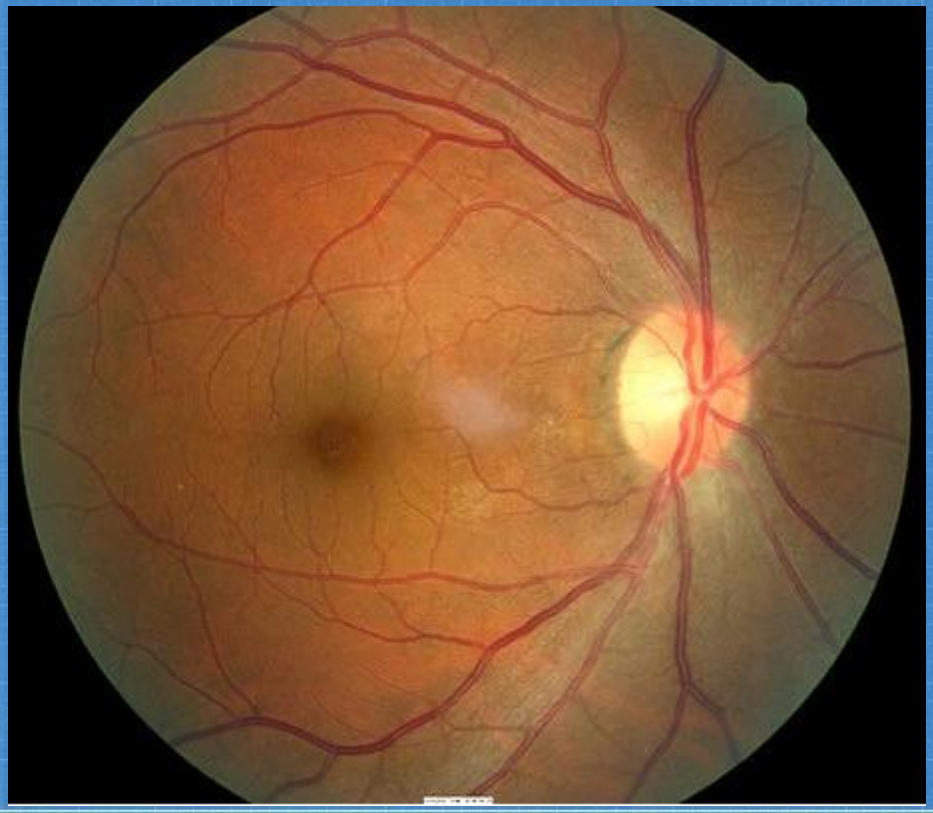
1. Complete PVD
2. **There is partial detachment of the posterior hyaloid from the central fovea with persistent attachment at the ONH**
3. There is vitreomacular traction resulting in obscuration of the foveal contour

Case: Management

Patients with *symptomatic* (floaters and/or photopsia) PVD without vitreous hemorrhage or peripheral retinal breaks *require no immediate treatment* but may be re-examined in *one to two weeks*, since some retinal breaks appear to develop days to weeks after the onset of symptoms.

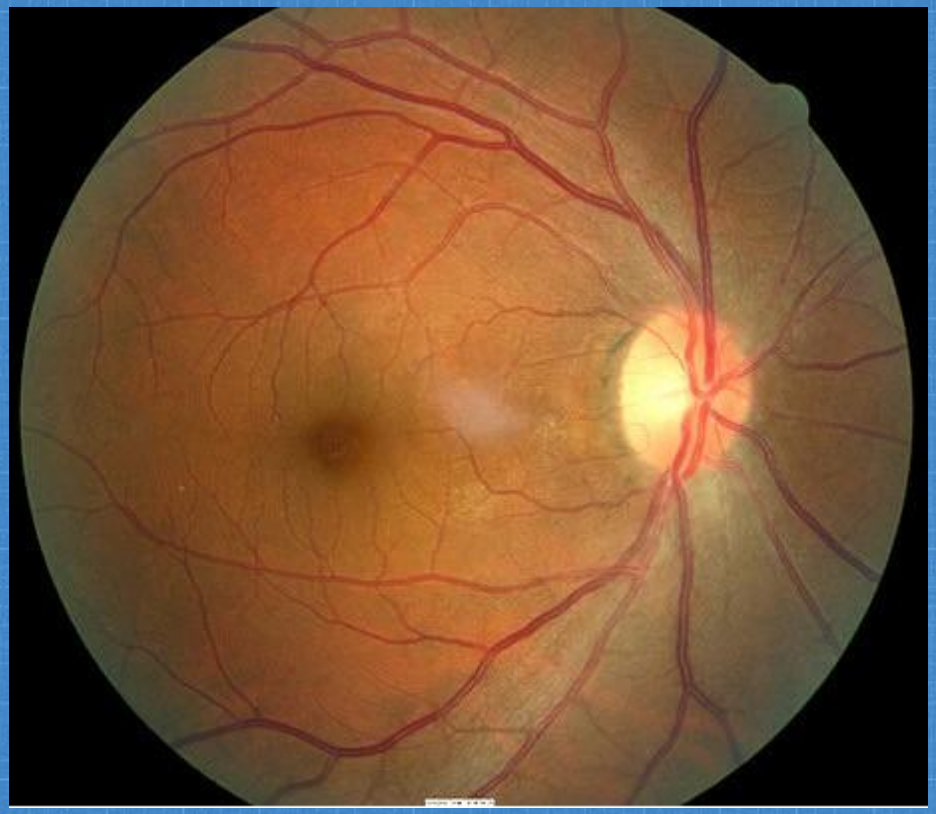
Incomplete PVD…..

71 year-old female

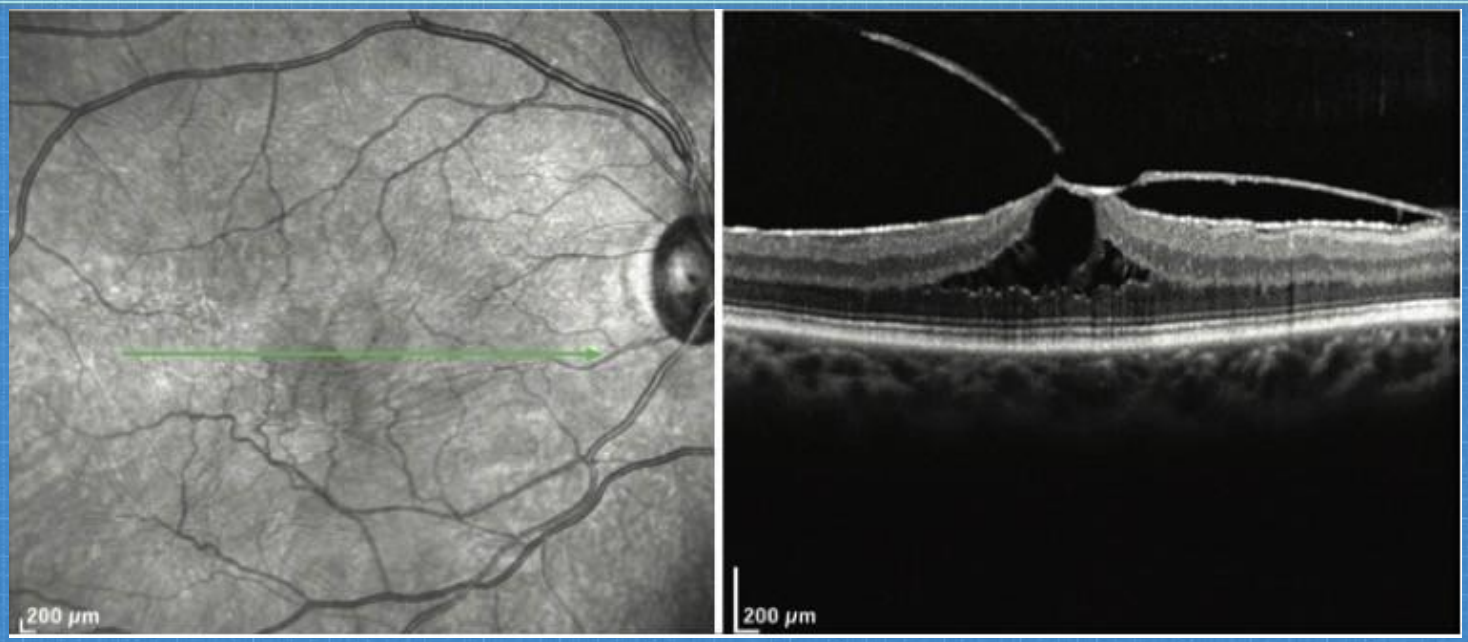


"The vision in my right eye has gotten progressively worse over the last three months and everything looks wavy!!"

Pertinent Findings

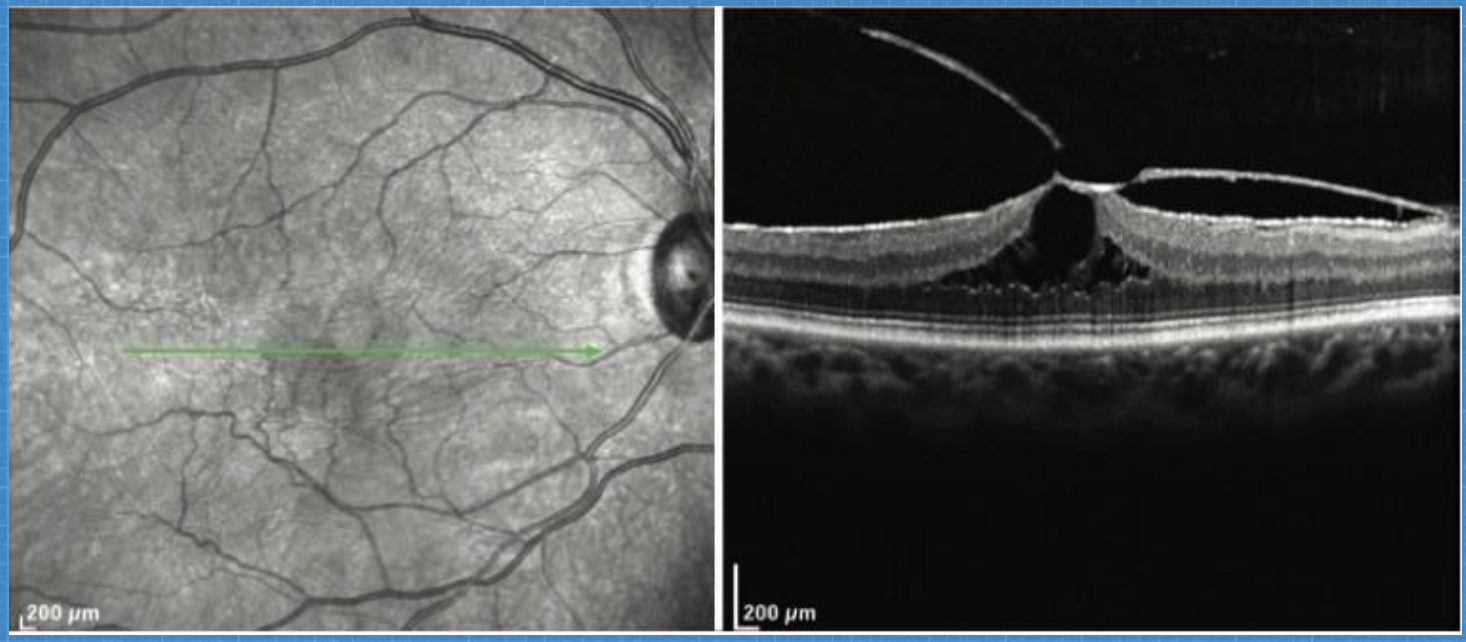


- BCVA *OD: 20/80*, OS: 20/20
- Fundoscopy: Vitreous condensation in fovea OD



The following is not true of the above OCT:

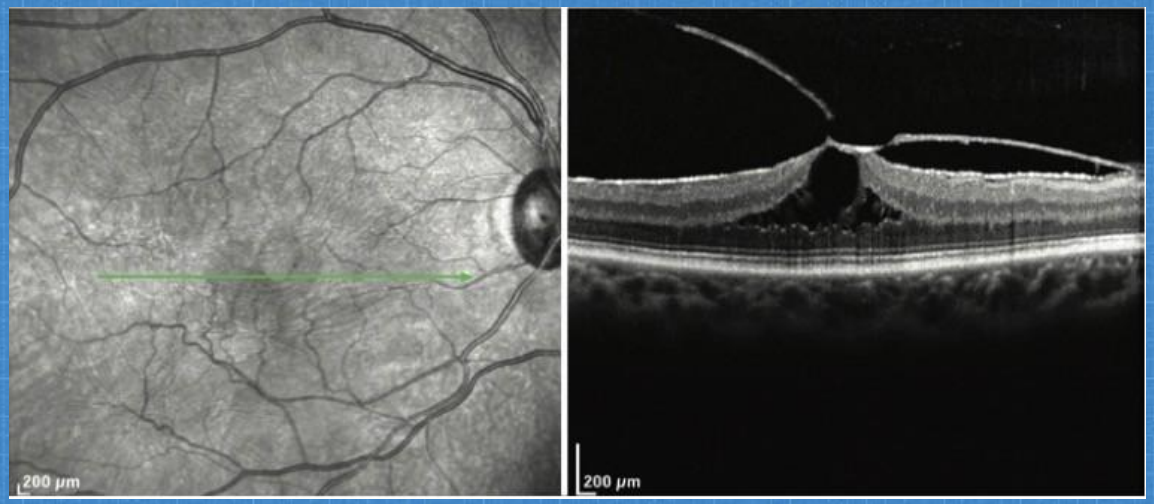
1. VMT due to anomalous PVD that disorganized the foveal structure
2. ERM causing traction on the retinal surface
3. Broad vitreomacular traction (>1500 um)



The following is not true of the above OCT:

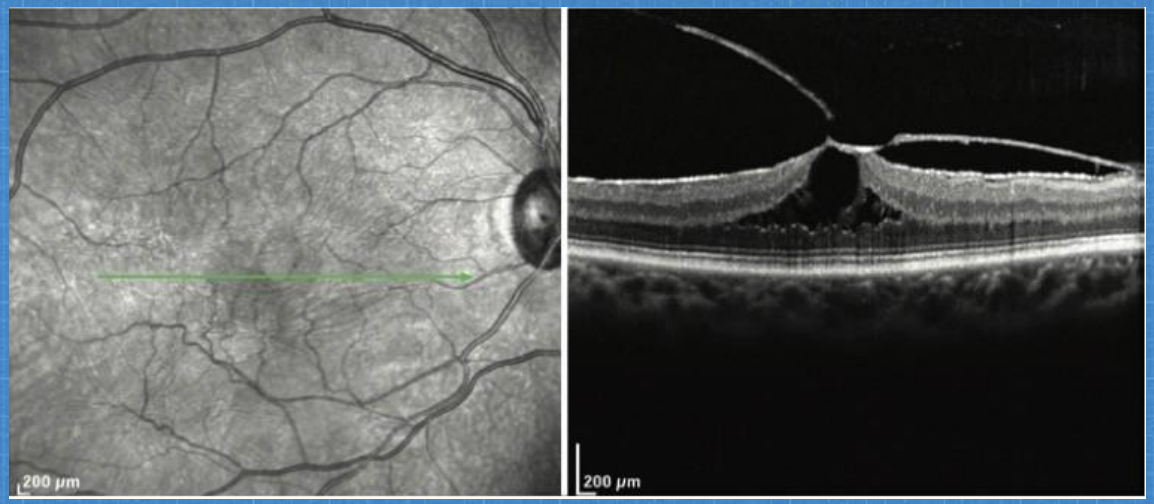
1. VMT due to anomalous PVD that disorganized the foveal structure
2. ERM causing traction on the retinal surface
3. **Broad vitreomacular traction (>1500 um)**

Management Decisions



1. Is patient symptomatic?
2. What is the size of the VMT?
3. Is there an associated ERM?

Management Decisions



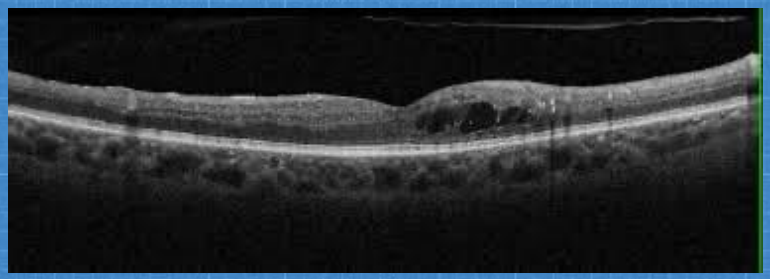
1. Is patient symptomatic? Yes
2. What is the size of the VMT? <1500 um (focal)
3. Is there an associated ERM? Yes

This patient:

A vitrectomy and ERM peel were performed

https://www.youtube.com/watch?v=DDzPUAdQGpo

49-year Hispanic female; Type 2 DM x 10 years

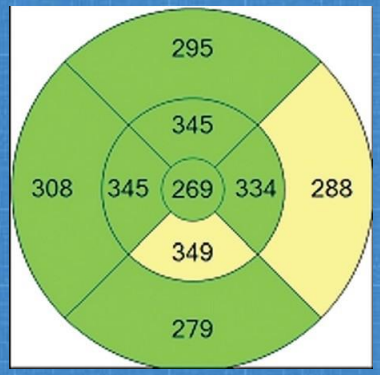


What else?

Moderate NPDR

More than just microaneurysms but less than severe

DME: Center Involving (CI-DME) or Non-Center Involving (NCI-DME)



ETDRS grid map with numerical data for central subfield retinal thickness within innermost circle (1mm diameter)

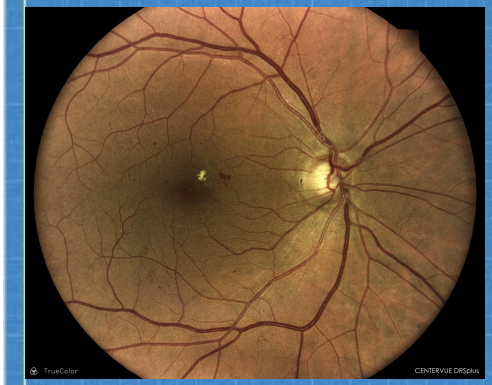
CI-DME: OCT demonstrating foveal involvement of intraretinal or subretinal fluid with concurrent thickening affecting the 1mm diameter **central** subfield thickness

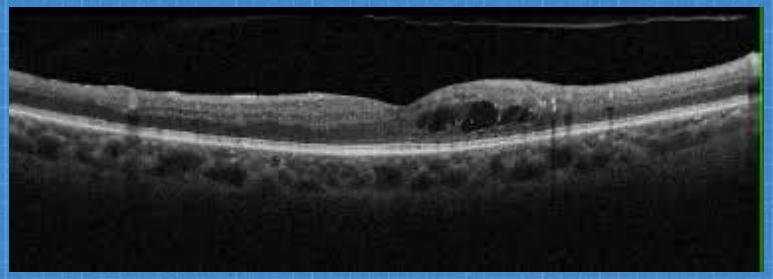
https://www.asrs.org/content/documents/evidence-based-guidelines-for-management-of-diabetic-macular-edema.pdf

Why does it matter? (ETDRS)

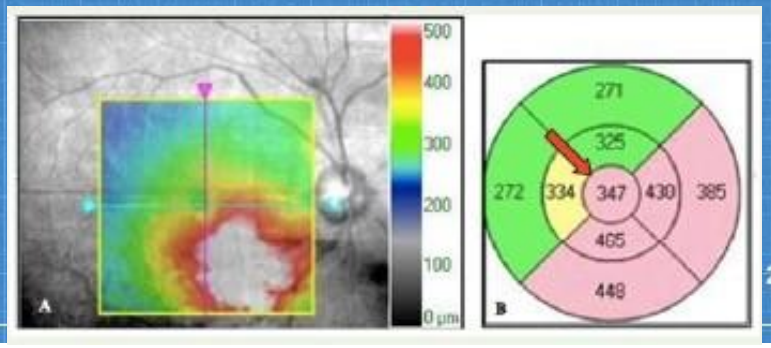
Eyes with CI-DME have a 10-fold greater risk of moderate vision loss compared to eyes without center involvement!

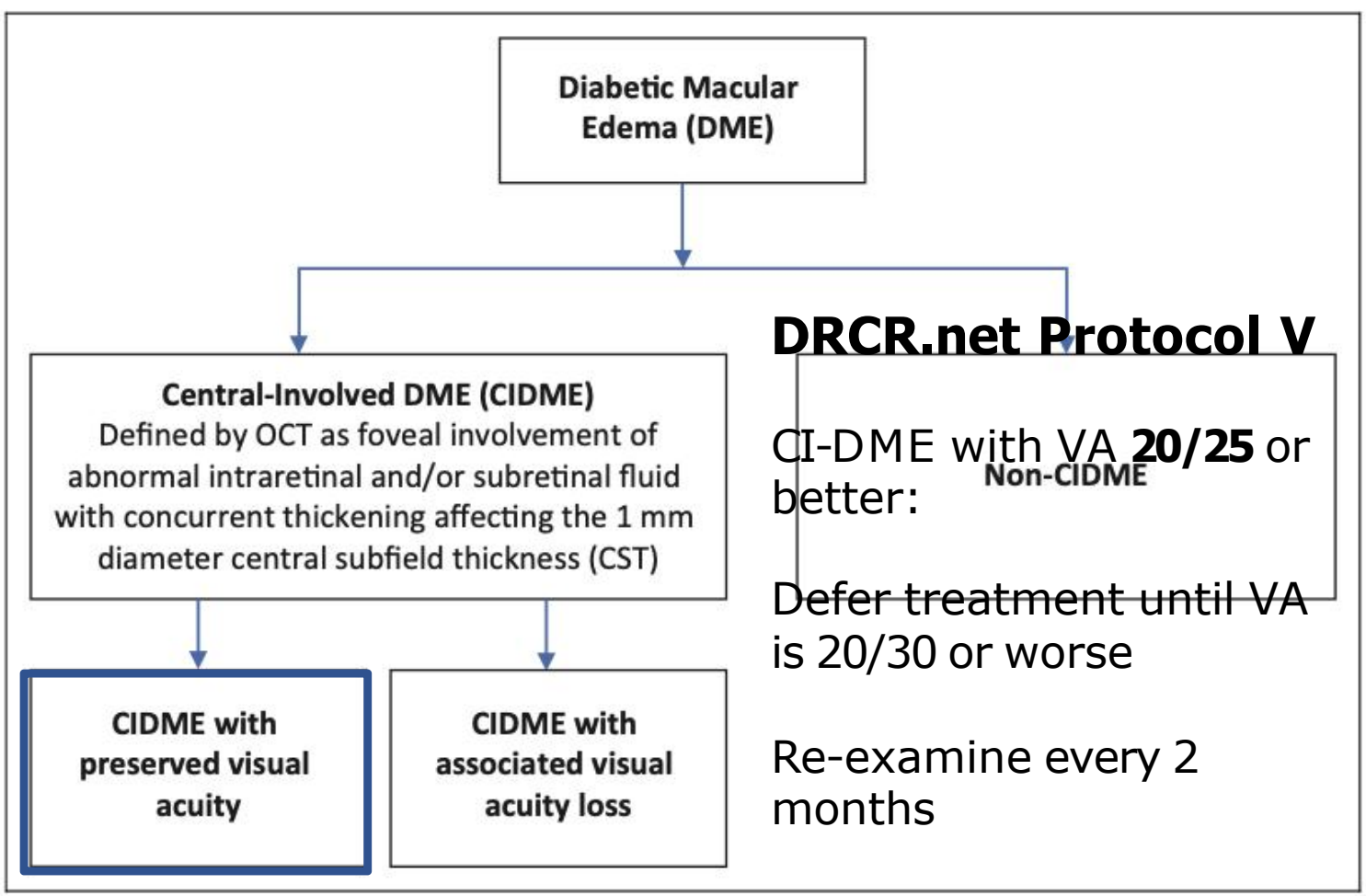
Severity of Retinopathy	Presence of Macular Edema	Follow-up (Months)	Panretinal Photocoagulation (Scatter) Laser	Focal and/or Grid Laser*	Intravitreal Anti-VEGF Therapy
Moderate NPDR	No	6-12[‡]	No	No	No
	NCI-DME	3–6	No	Sometimes	Rarely
	CI-DME[†]	1*	No	Rarely	Usually





How do we define the CME?





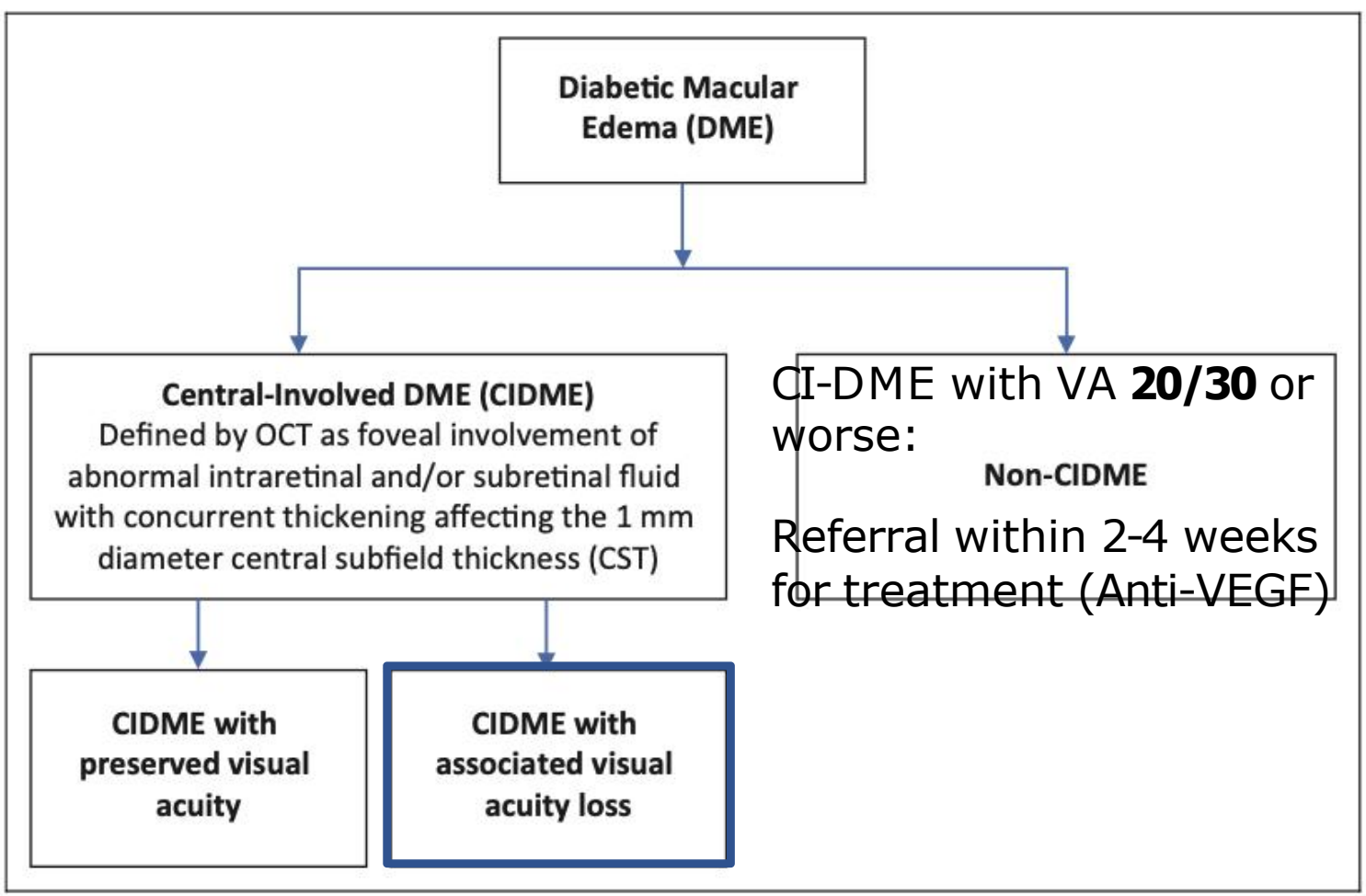
DRCR.net Protocol V

CI-DME with VA **20/25** or better:

~~Defer treatment until VA~~ is 20/30 or worse

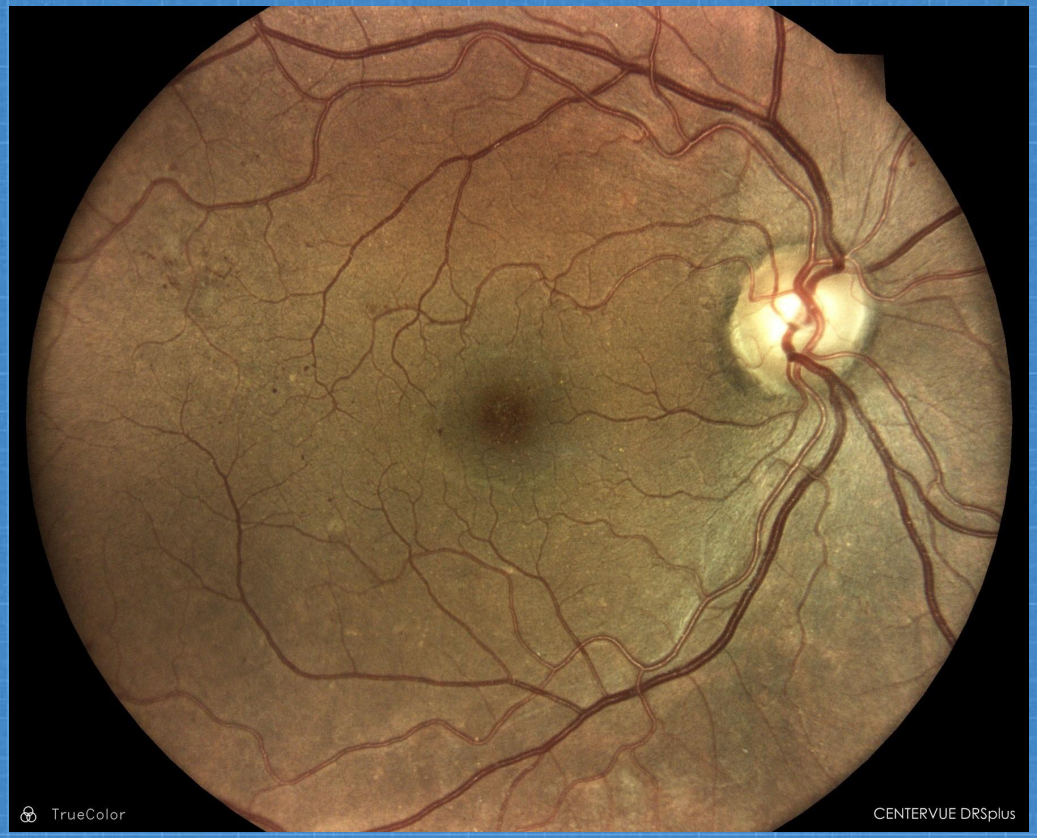
Re-examine every 2 months

https://www.asrs.org/content/documents/evidence-based-guidelines-for-management-of-diabetic-macular-edema.pdf



https://www.asrs.org/content/documents/evidence-based-guidelines-for-management-of-diabetic-macular-edema.pdf

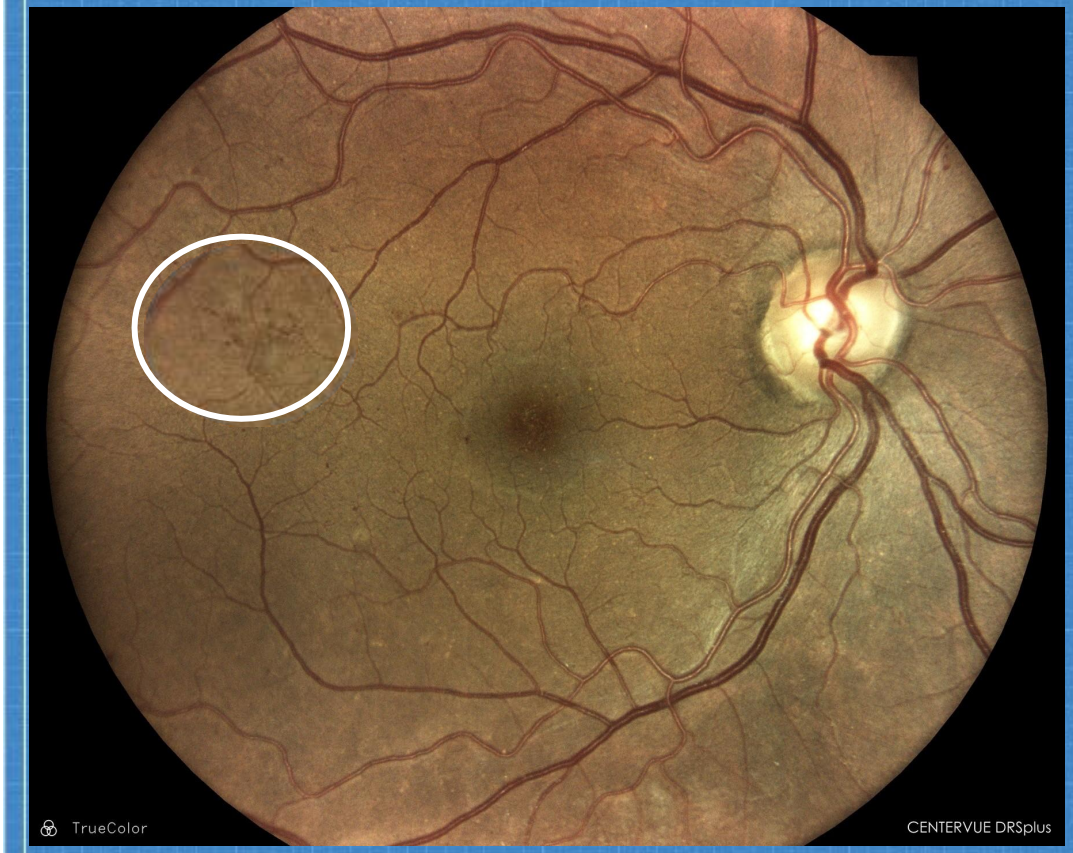
60-year-old Hispanic male



"The vision in my right eye is not good… it has been getting progressively worse. Could it be my diabetes?"

20/40

Ancillary Testing



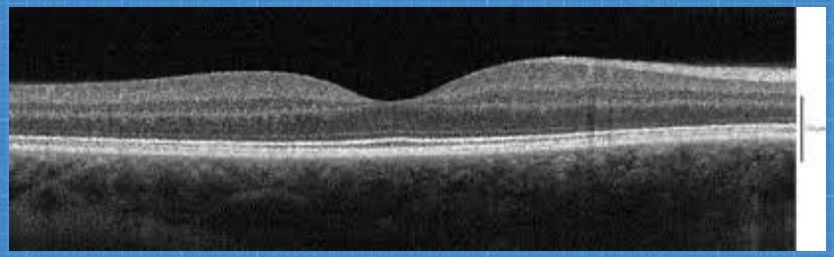
- Fundus Photography: Grade the retinopathy

Hemorrhages, CWS

Moderate NPDR	More than just microaneurysms but less than severe

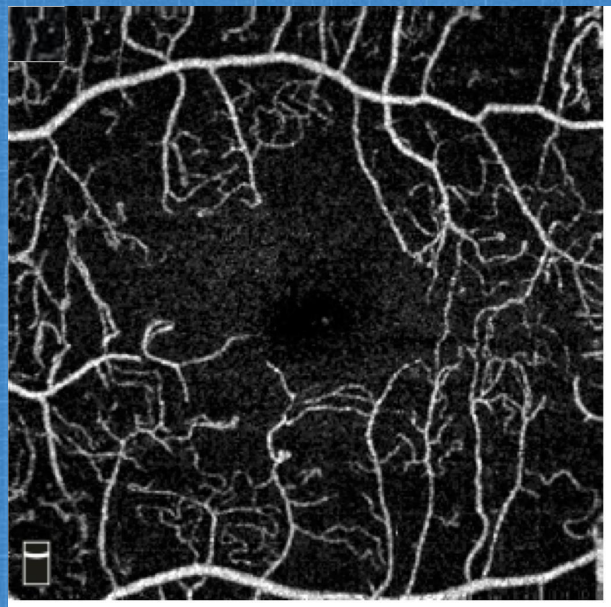
Ancillary Testing

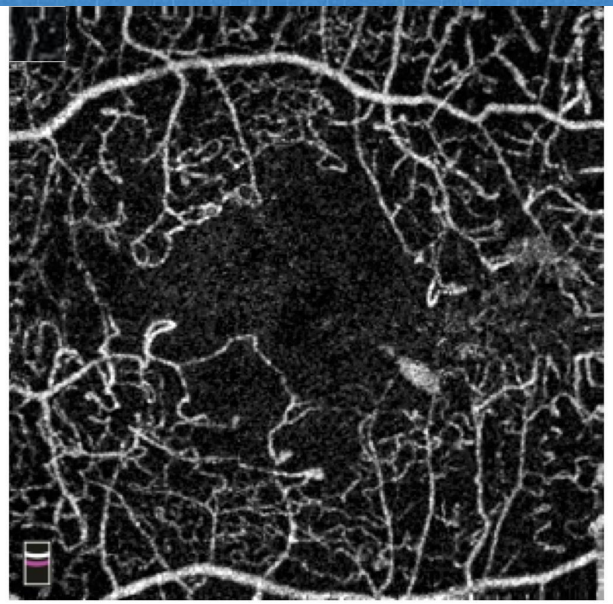
- OCT: Is there Macular Edema?

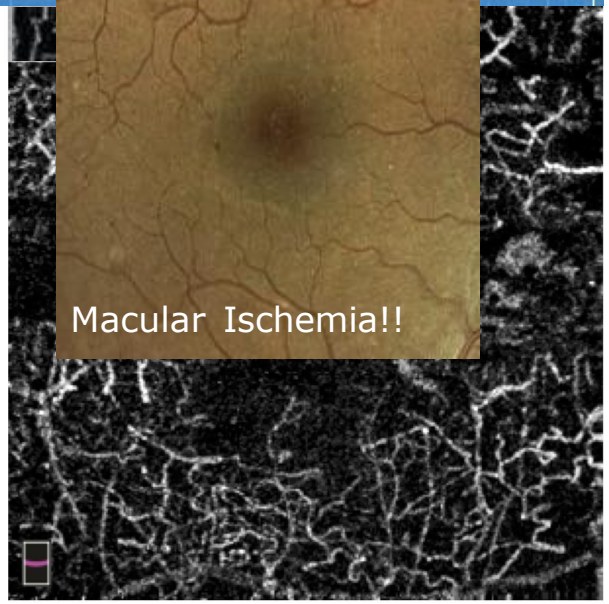


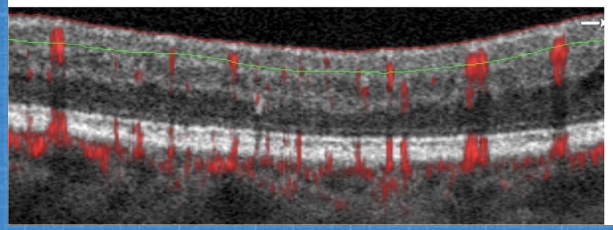
No!! So why the reduced vision??

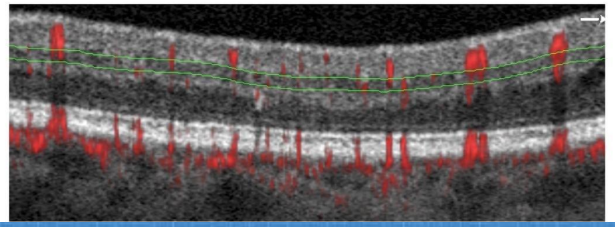
Ancillary Testing: OCTA

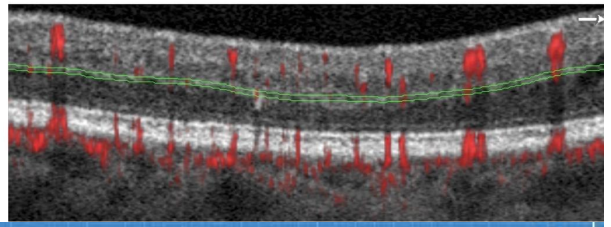












Management?

Severity of Retinopathy	Presence of Macular Edema	Follow-up (Months)	Panretinal Photocoagulation (Scatter) Laser	Focal and/or Grid Laser*	Intravitreal Anti-VEGF Therapy
Normal or minimal NPDR	No	12	No	No	No
Mild NPDR	No	12	No	No	No
	NCI-DME	3–6	No	Sometimes	No
	CI-DME†	1*	No	Rarely	Usually
Moderate NPDR	No	6–12‡	No	No	No
	NCI-DME	3–6	No	Sometimes	Rarely
	CI-DME†	1*	No	Rarely	Usually

What about the macular ischemia???

Guarded prognosis!
Anti-VEGF NOT EFFECTIVE

A Triad of Conundrums…

Case Report #1

24-year-old female

"I have been noticing colored spots in my vision in my right eye for the past 8 months."

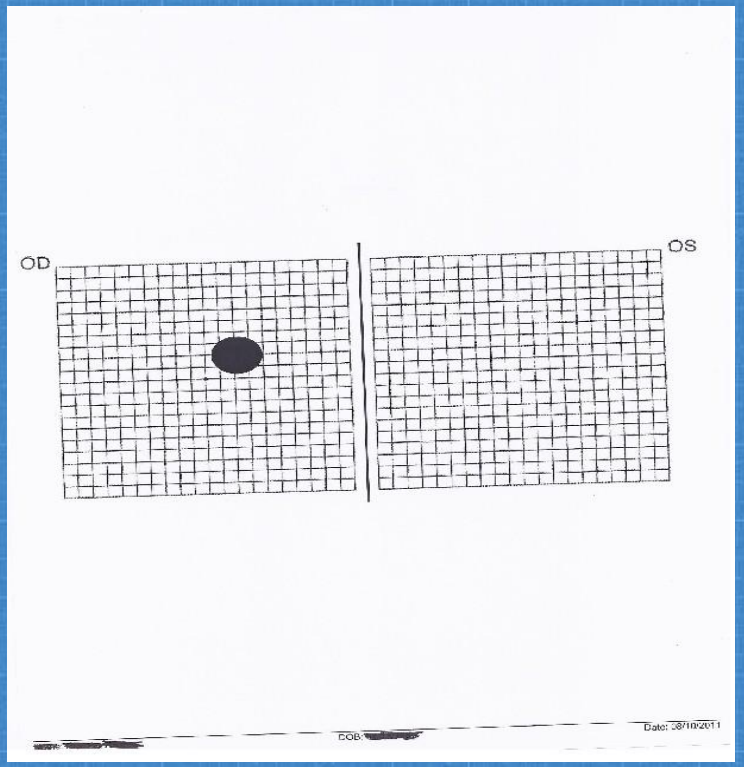
- Ocular disturbances coincided with onset of a coughing fit 8 months prior to this exam and patient stated she had some type of cold or virus.
- Systemic history of anemia and patient reports only taking oral contraceptives and vitamins.

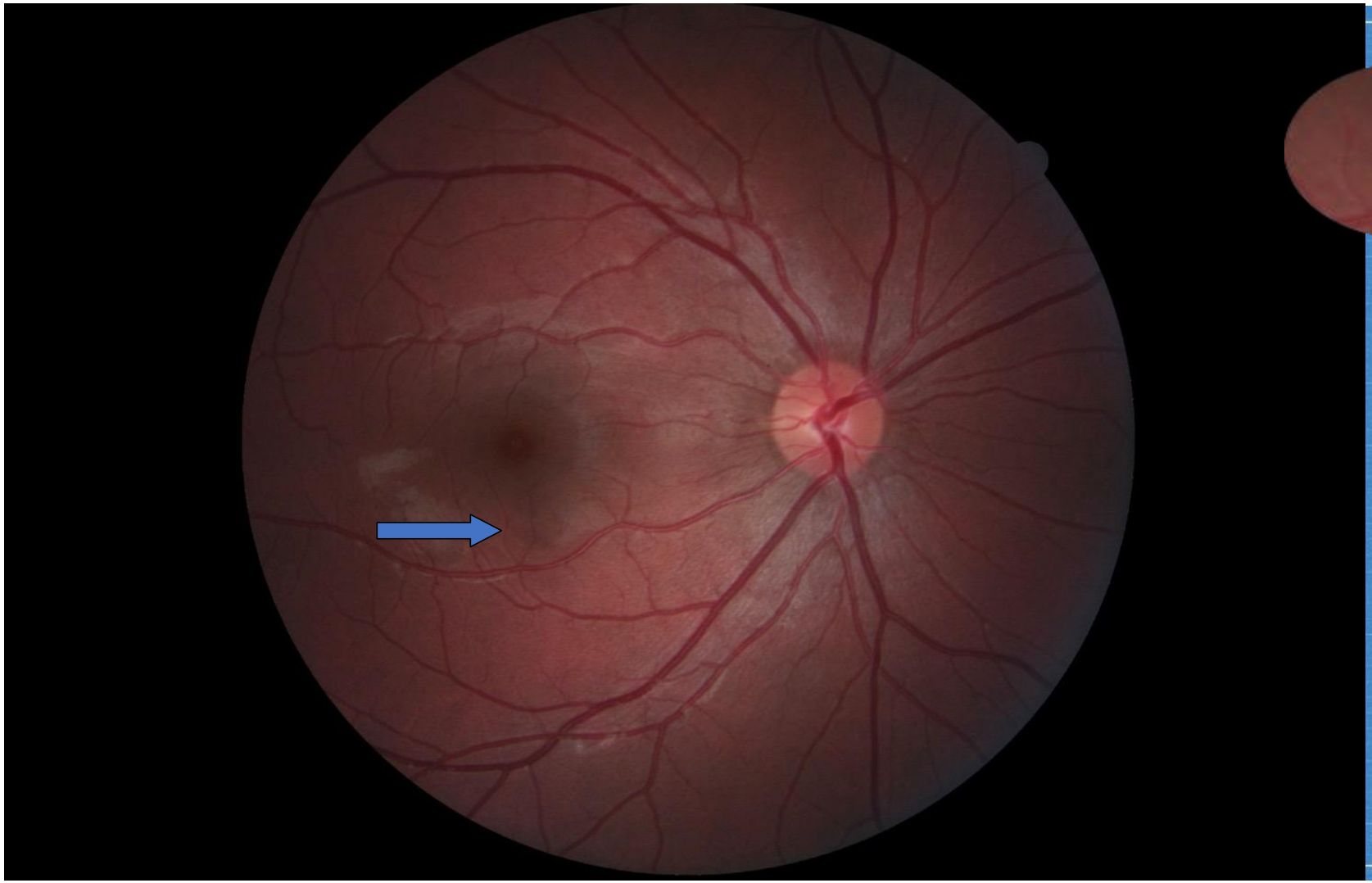
Case Report #1

- Visual acuity:
  SC OD: 20/20-3, OS: 20/20, OU: 20/15
- Entrance testing:
  PERRL (-)APD
  Confrontation fields: FTFC (peripheral) OU
  EOM: Full OU
  Color vision: normal OU using HRR test

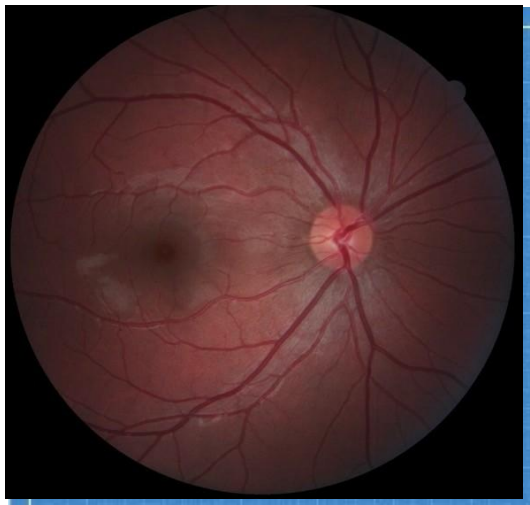
Case Report #1

- Amsler Grid:
  - OD: Patient described a superior central scotoma just above the center of vision.
  - OS: no abnormalities.

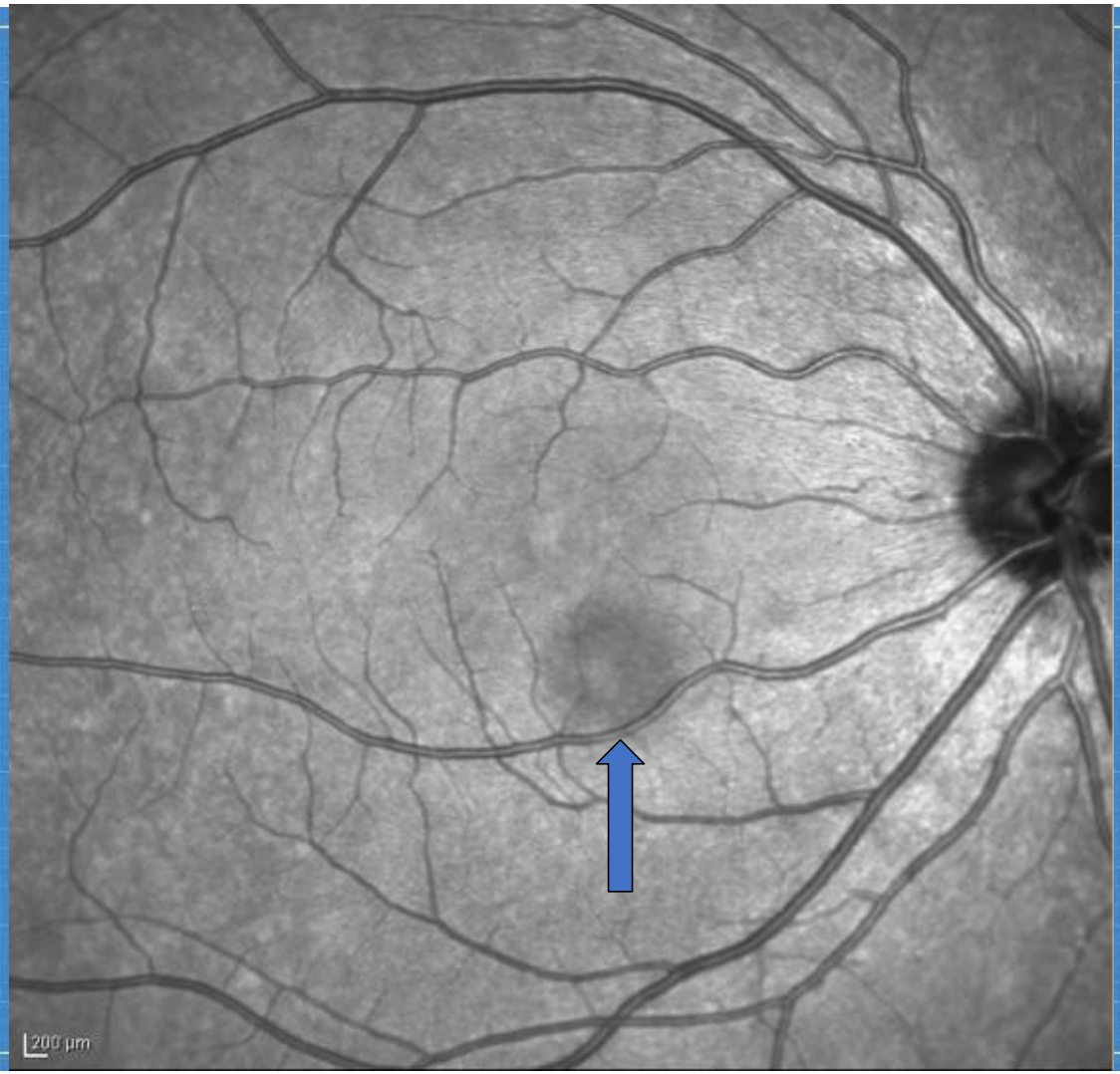


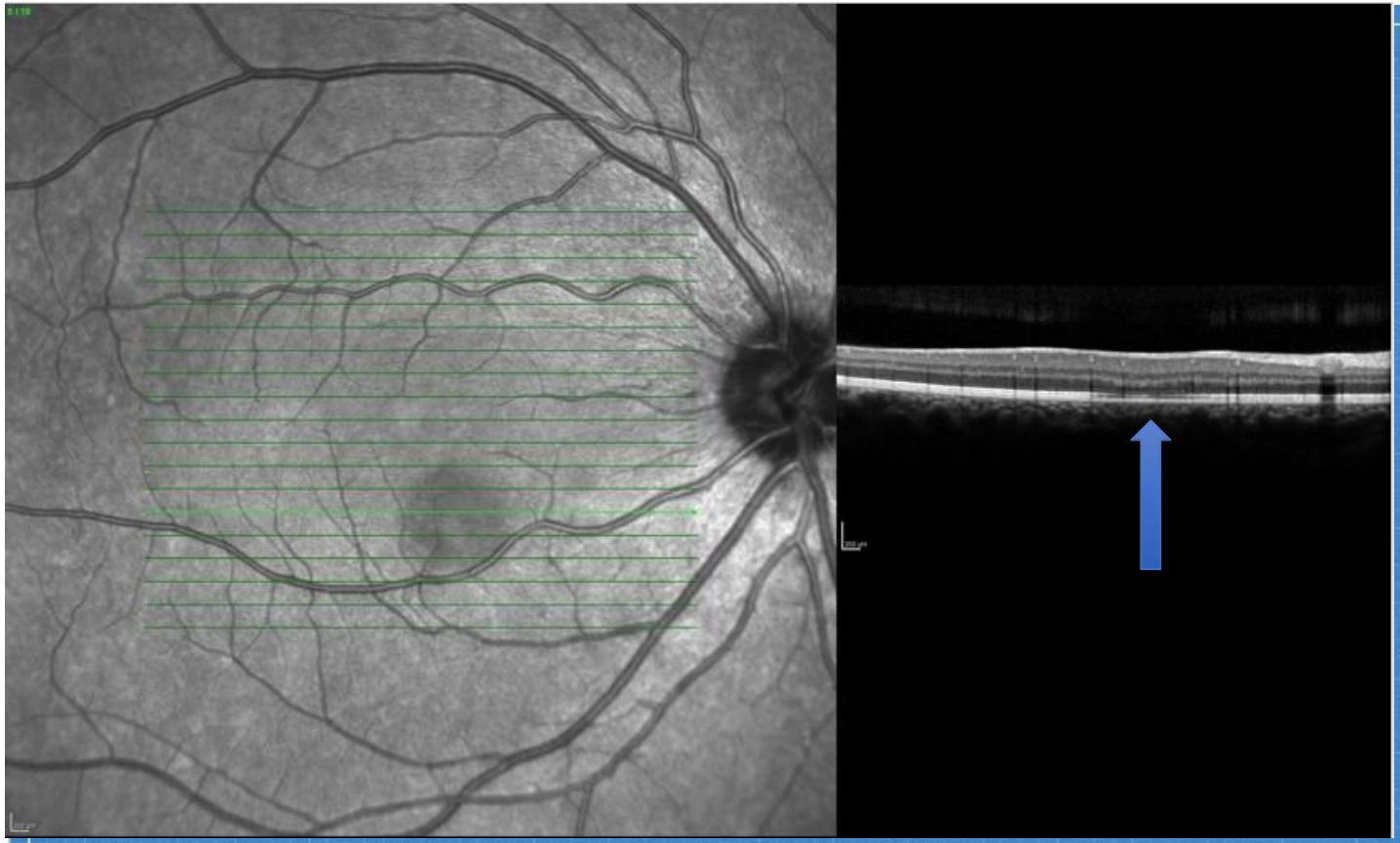


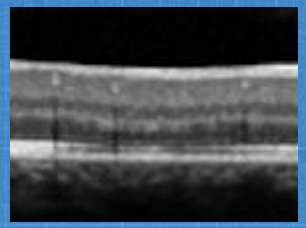
Fundus photo OD



Best visualized with red-free

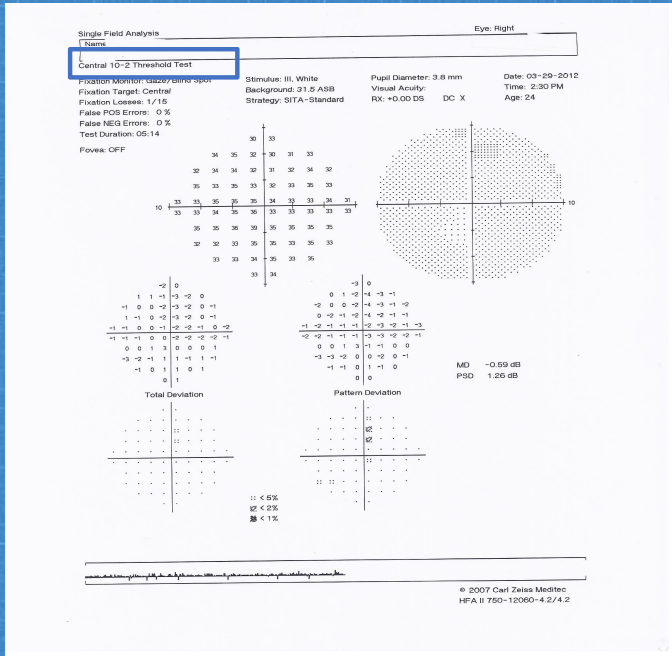


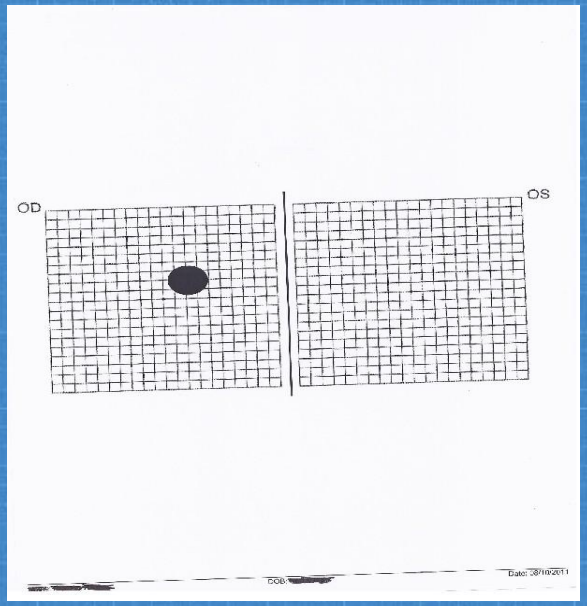


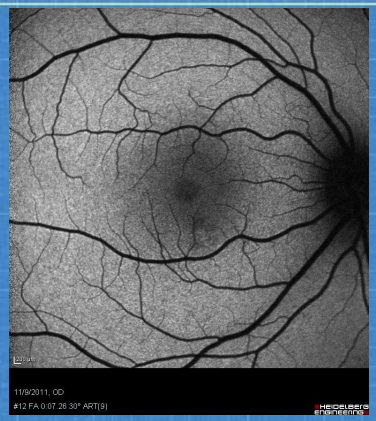


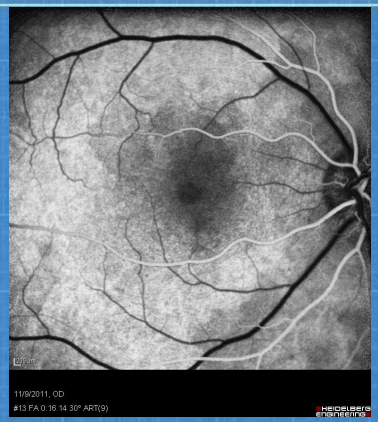
- OCT through lesion: Note disruption in PIL corresponding with adjacent lesion on fundus photo

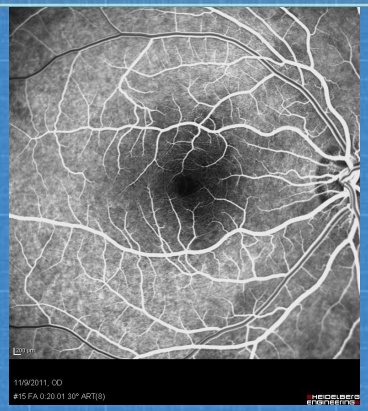
Visual field

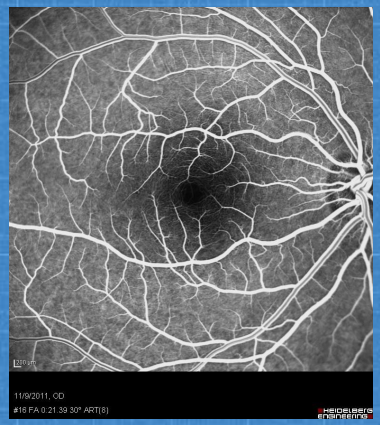


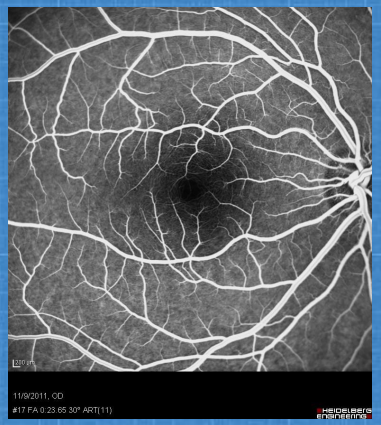


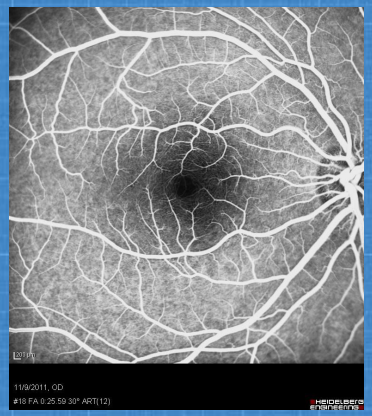












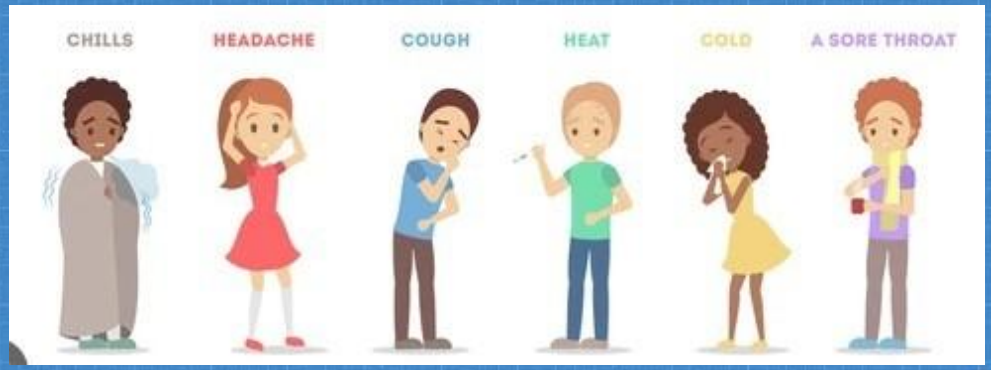
Normal fluorescein angiogram of the right eye

Case Report #2

43-year-old Caucasian female

"I have been noticing a small blind spot in the vision in my right eye" (unknown duration)

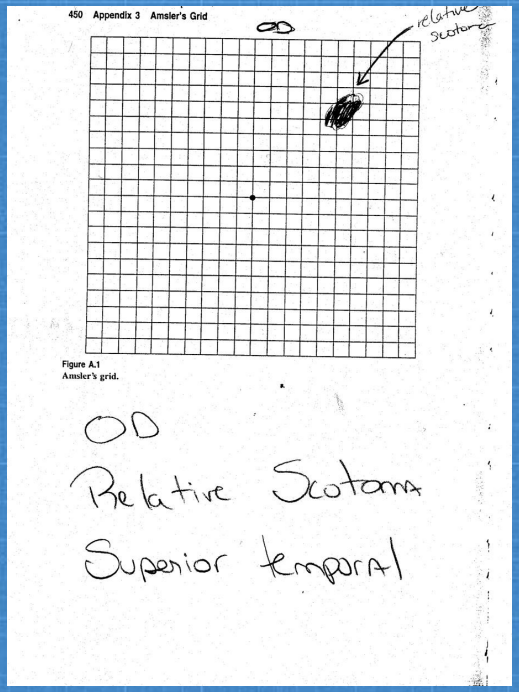
- Flu like symptoms recently
- Remainder of history unremarkable



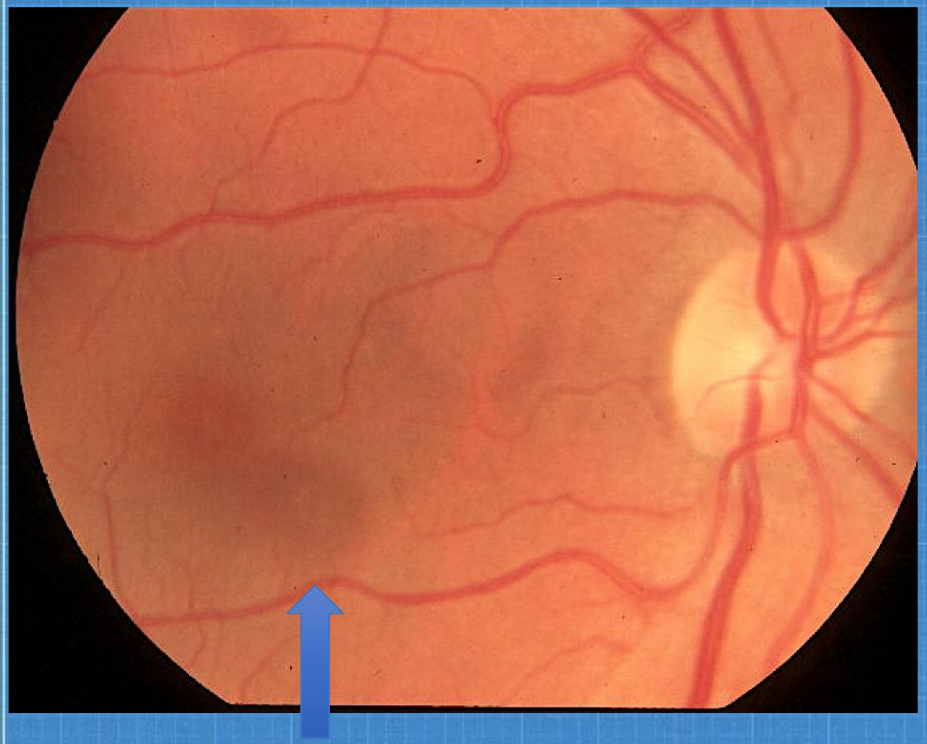
Case Report #2

- Visual acuity:
  - SC OD: 20/20, OS: 20/20
- Entrance testing:
  - PERRL (-)APD
  - Confrontation fields: FTFC (small scotoma superior to fixation)
  - EOM: Full OU
  - Color vision: normal OU using HRR test

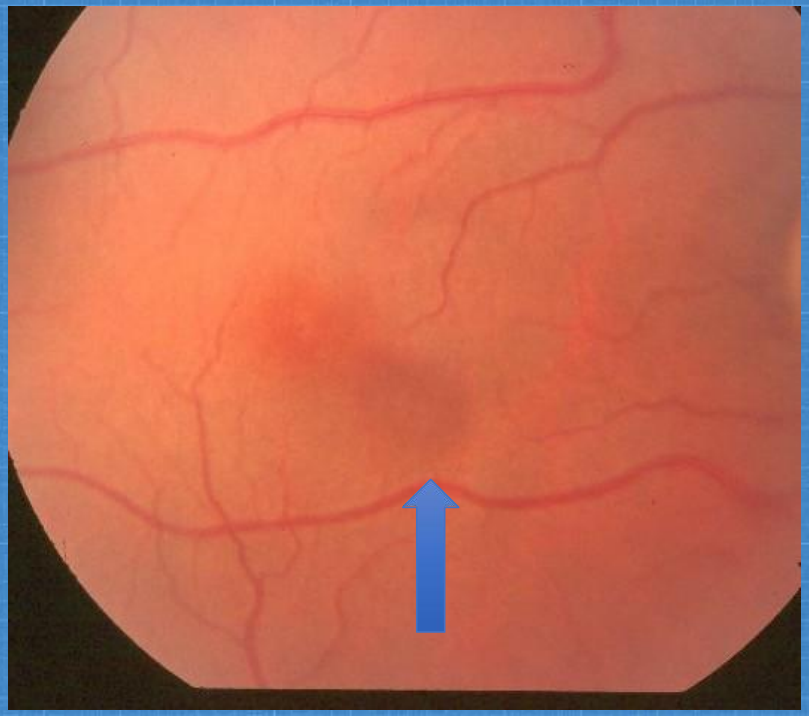
Amsler grid


450 Appendix 3 Amsler's Grid
OD
relative scotoma
Figure A.1
Amsler's grid.
OD
Relative Scotoma
Superior temporal

Photos

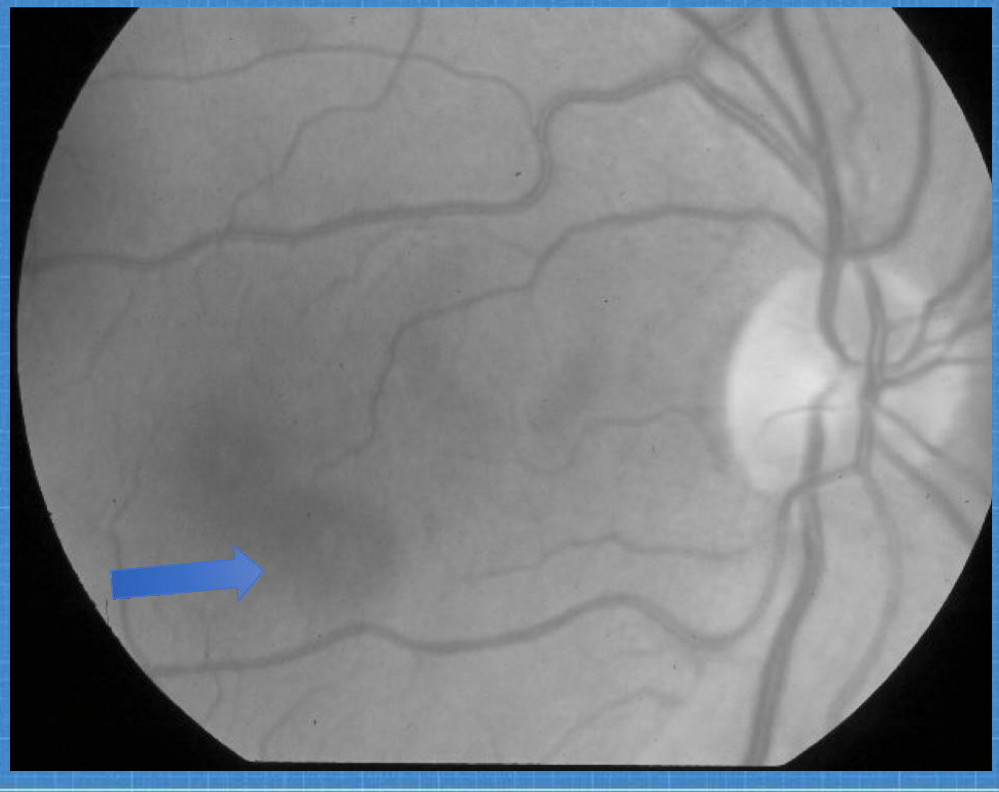


Fundus appearance at initial presentation

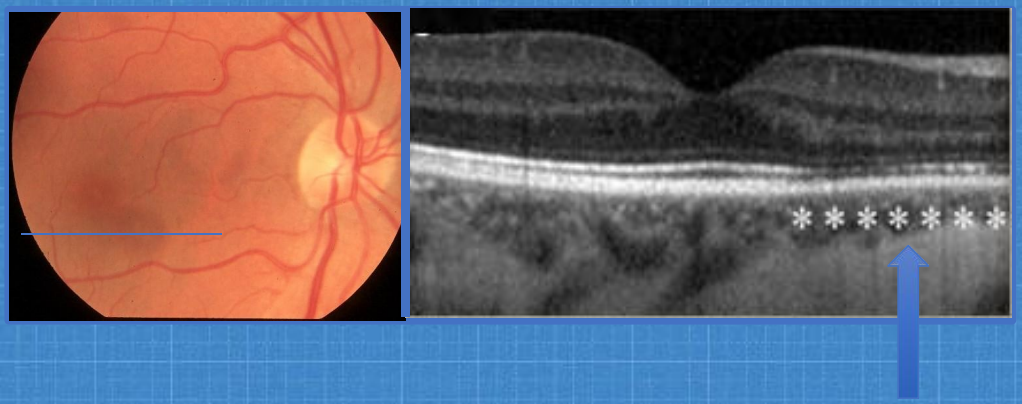


2 months later

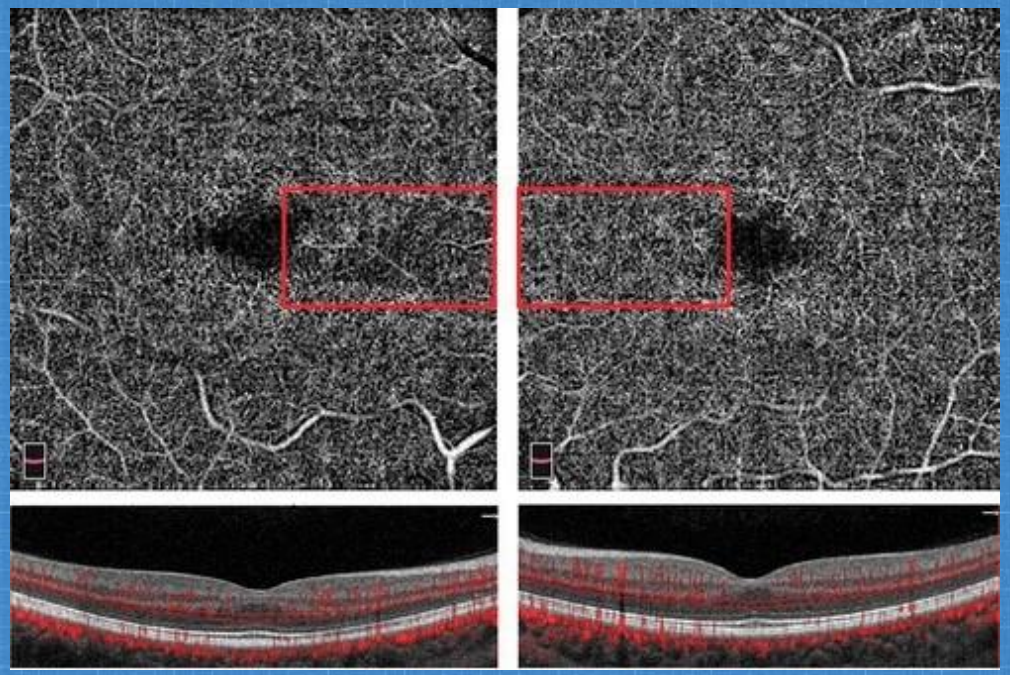
Red-free Image



OCT



OCT Angiography



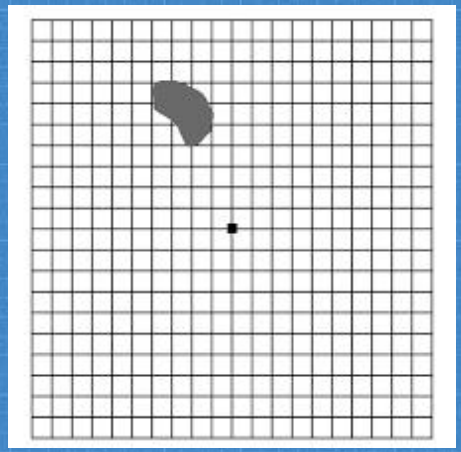
Flow deficit of the deep capillary plexus of the right eye (right image) with normal capillary density in the corresponding area of the left eye (left image)

Case Report #3

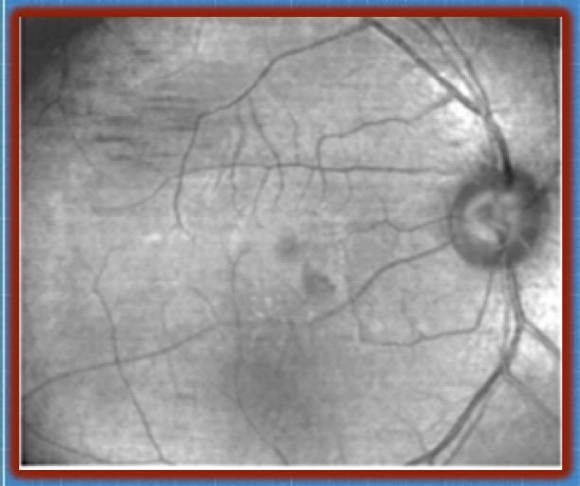
31-year-old female

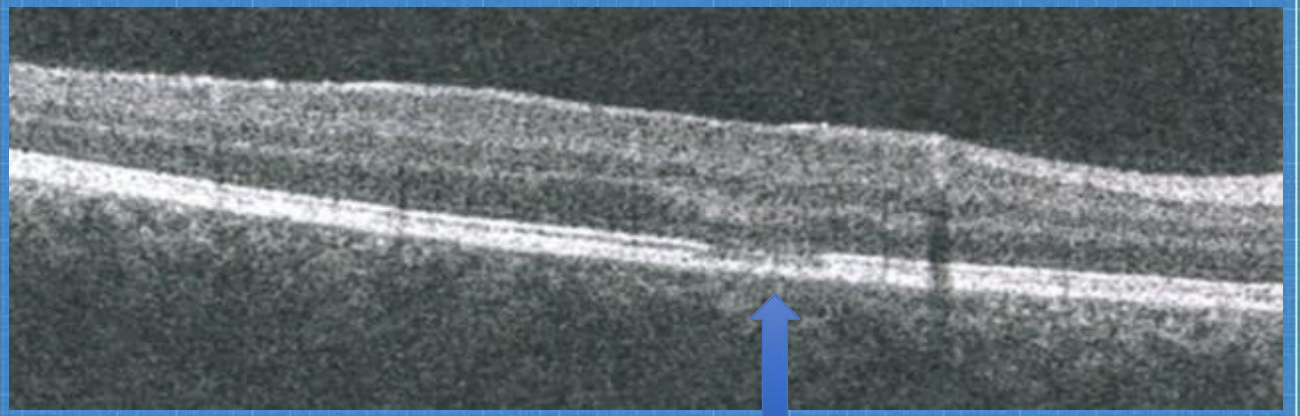
"Gray spot/shadow in my right eye x 2-3 days"

- Negative pertinent history
- BCVA 20/20 OD and OS
- Amsler: Small paracentral scotoma OD

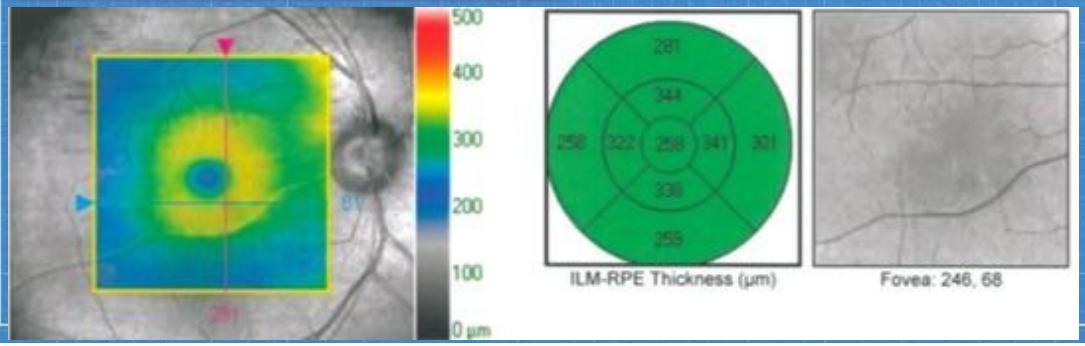


OCT

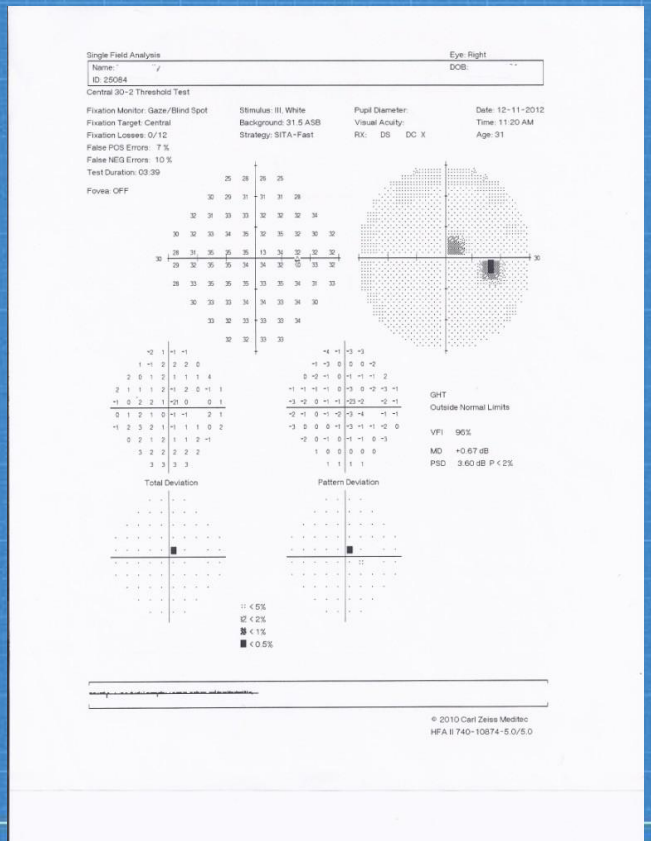


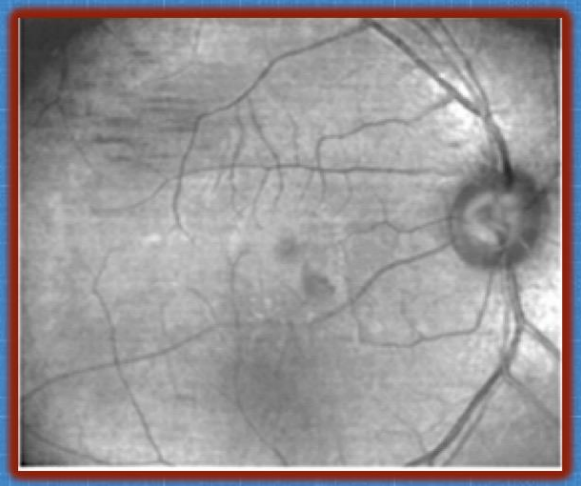


Disruption of PIL



Visual field/FAF

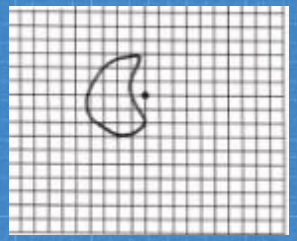




Diagnosis?

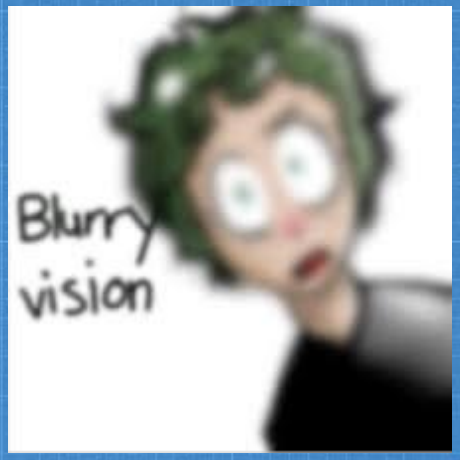
Acute Macular Neuroretinopathy (AMNR)

- Rare condition that causes sudden paracentral scotoma in young women
- Pathophysiology is unclear and there is no specific treatment



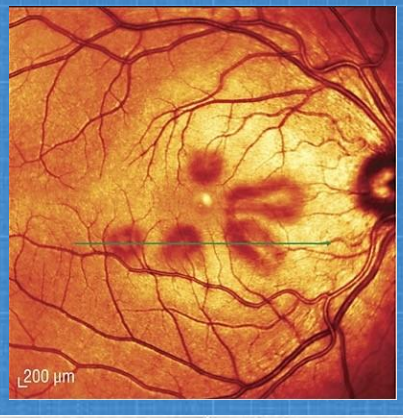
Natural History

- Sudden onset of mild visual impairment
- Unilateral or bilateral with normal to slightly abnormal visual acuity
- Patients usually complain of a visual disturbance that may change colors paracentral to fovea in the affected eye
- Visual field defects are directly associated with the area seen on the retina



Clinical Diagnosis

- Lesions are distinct and appear as dark reddish-brown, wedge-shaped areas that point toward the fovea
- Best viewed with red-free light
- PIL- IS/OS affected, neurosensory retina
- The lesions may develop rapidly or over days to weeks



Lesion began to fade with complete resolution by 4 months

Ancillary Testing

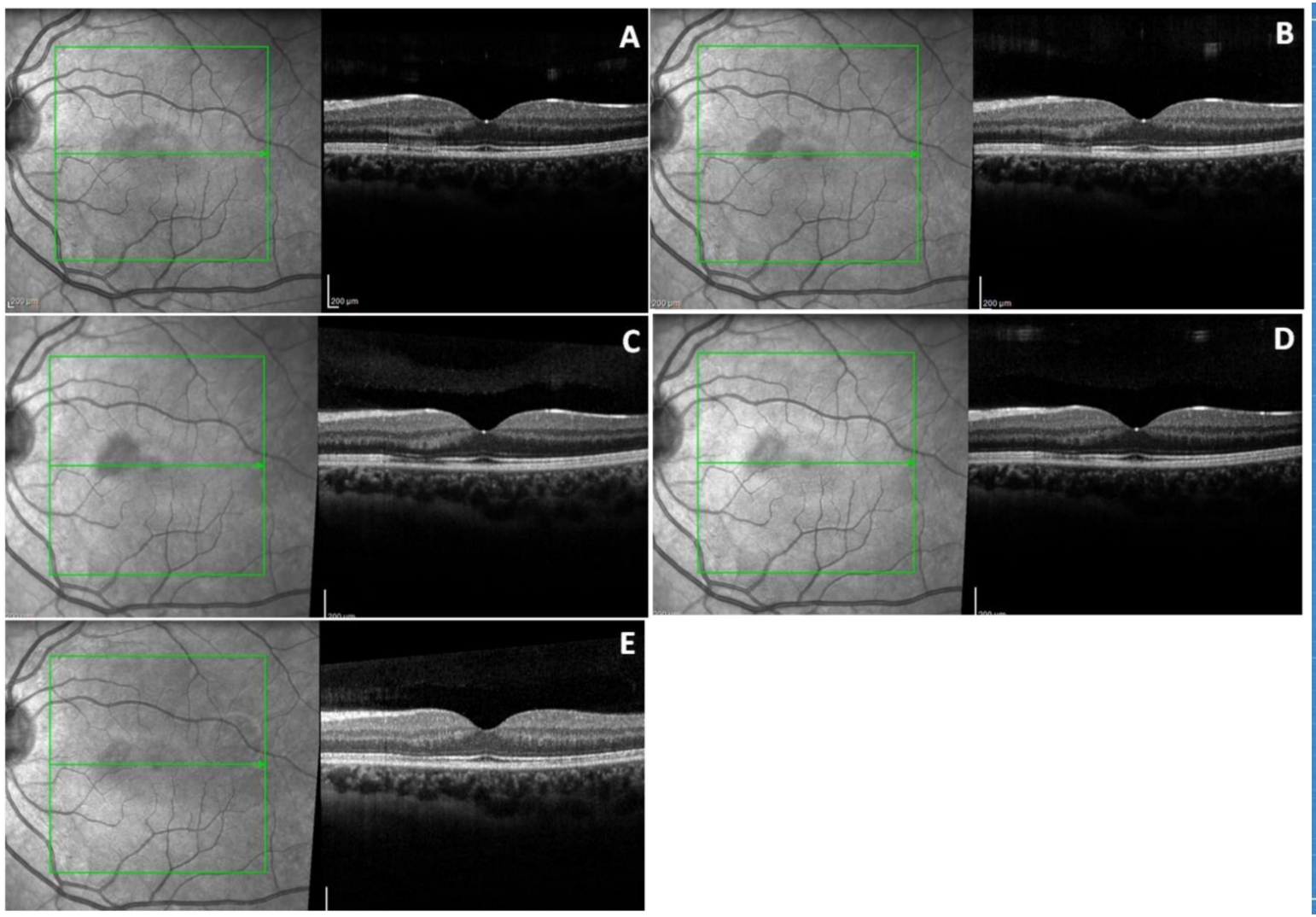
- Fluorescein angiography of acute macular neuroretinopathy is typically normal
- There may be slight hypofluorescence of the lesions
- Optical coherence tomography has proven to be an excellent diagnostic tool for this condition

OCT

WHAT STAGE OF THE DISEASE ARE YOU SEEING???

- Superficial involvement at the level of the outer plexiform layer early in the course of the disease
- Chronic- segmental absence of tissue involving the inner sensory/outer sensory junction of the retinal layers generally not disturbing the RPE layer

OCT



Incidence of AMNR

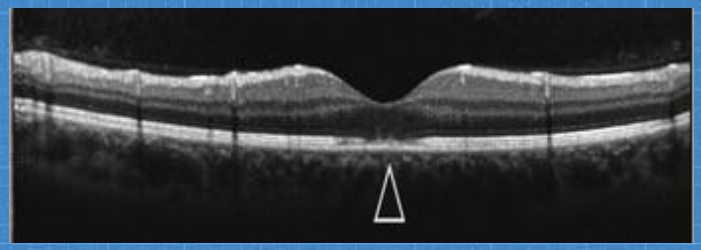
- The incidence of AMN significantly increased from 0.66/100,000 visits in 2019 to **8.97/100,000 visits in 2020**
- Acute macular neuroretinopathy is seen more frequently in women than men
- Women were in their reproductive years, with a mean age of 27 years and typically taking oral contraceptives

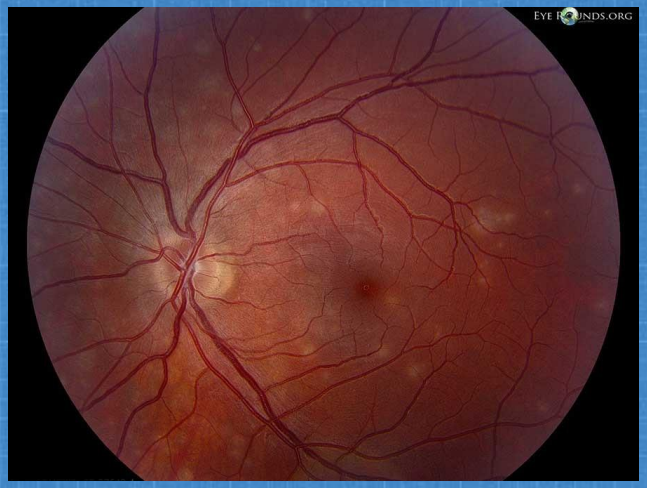
Causes of AMINR

- Unknown… UNDERLYING ISCHEMIC ETIOLOGY???
- Oral contraceptives
- Hypotension
- Vasoconstrictive agents (caffeine, epinep adrenaline)

Differential diagnosis

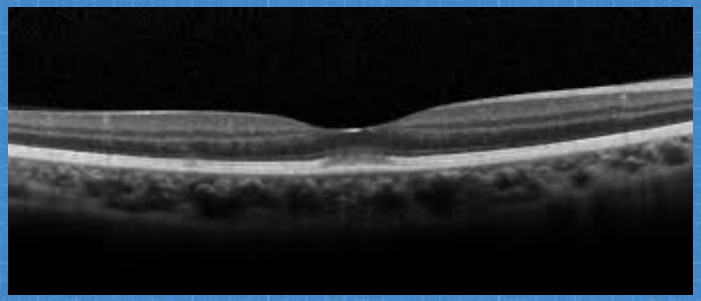
MEWDS (Multiple Evanescent White Dot Syndrome)

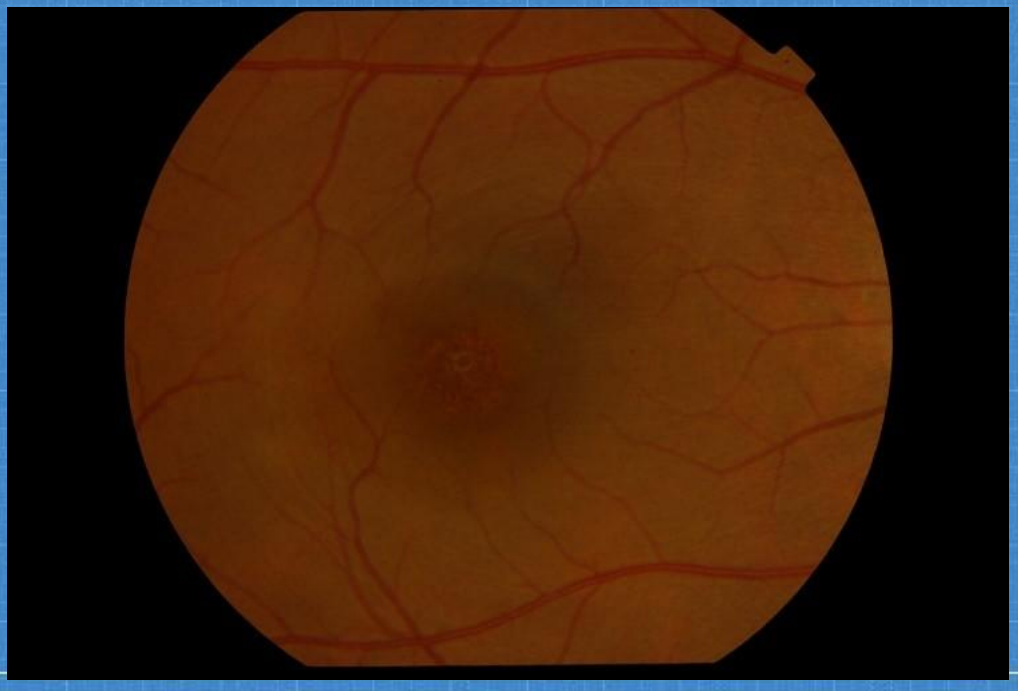




Differential diagnosis

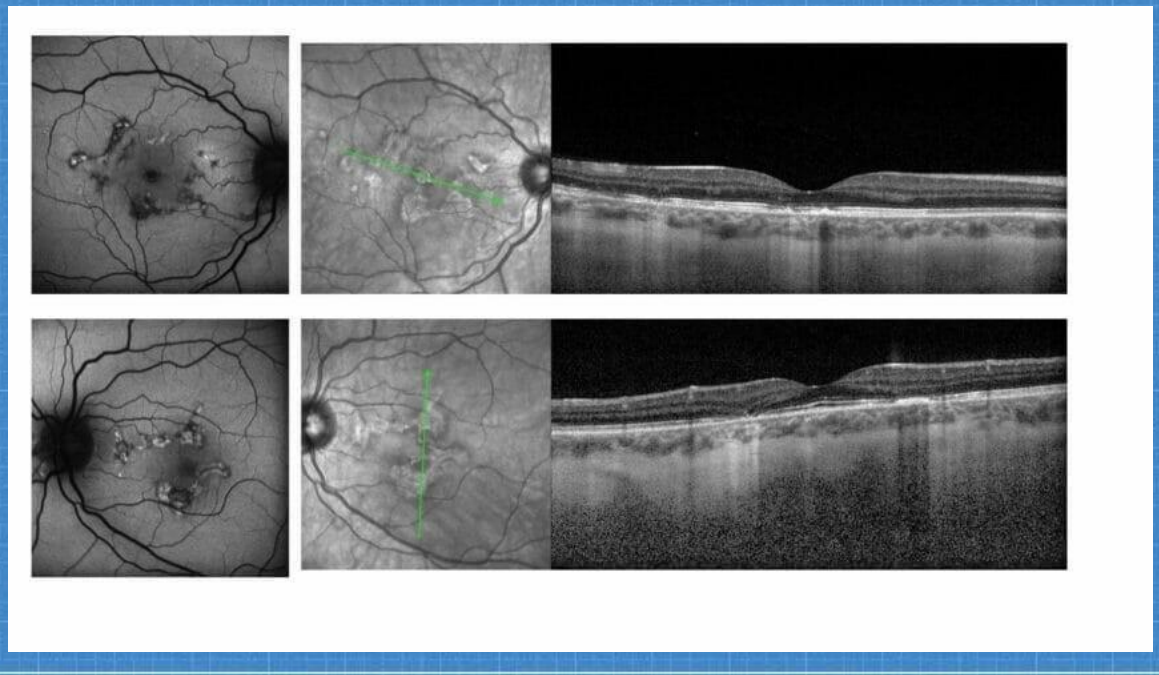
Acute retinal pigment epitheliitis (Krill's Disease)





Differential diagnosis

Acute posterior multifocal placoid pigment epitheliopathy

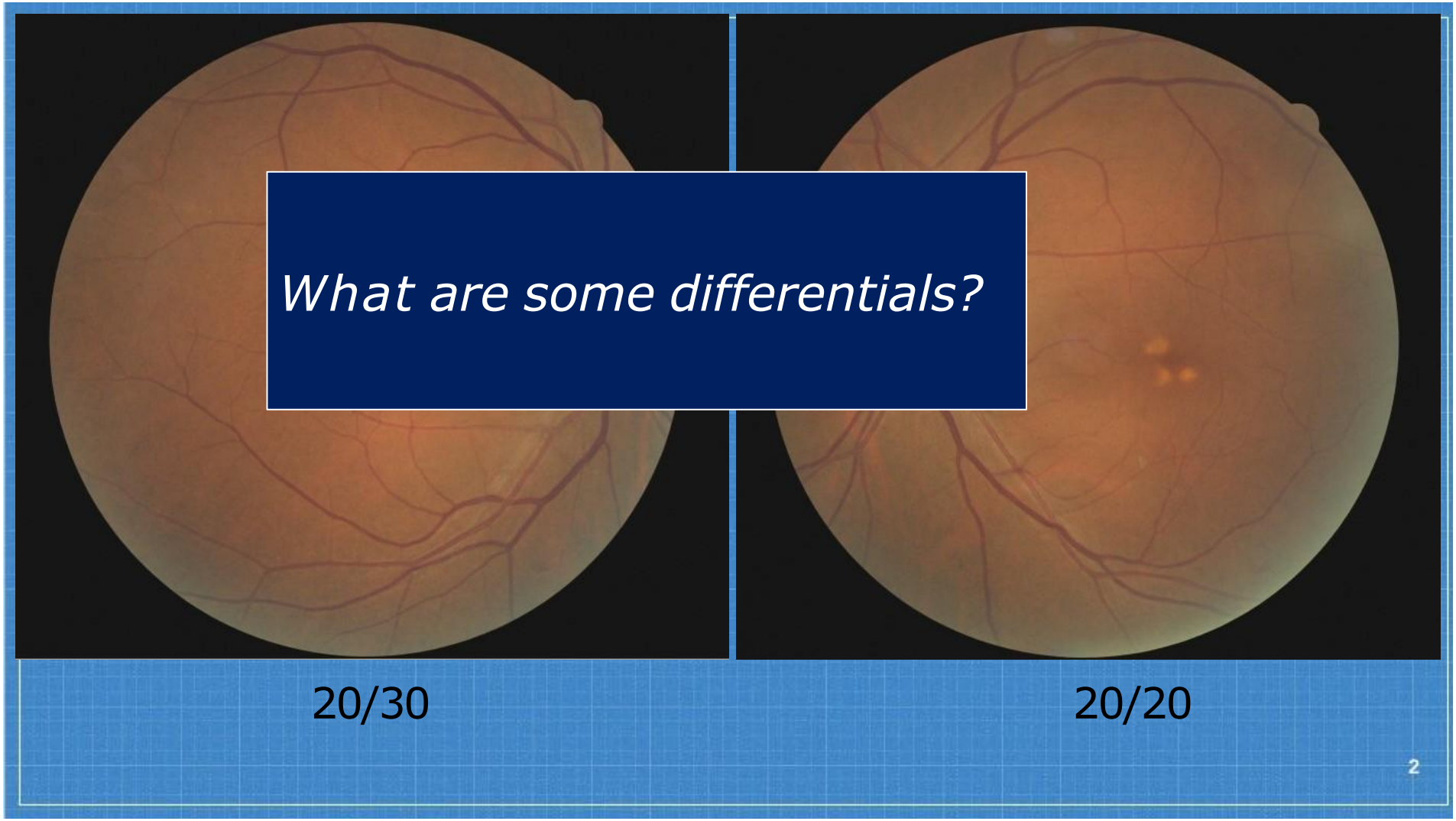


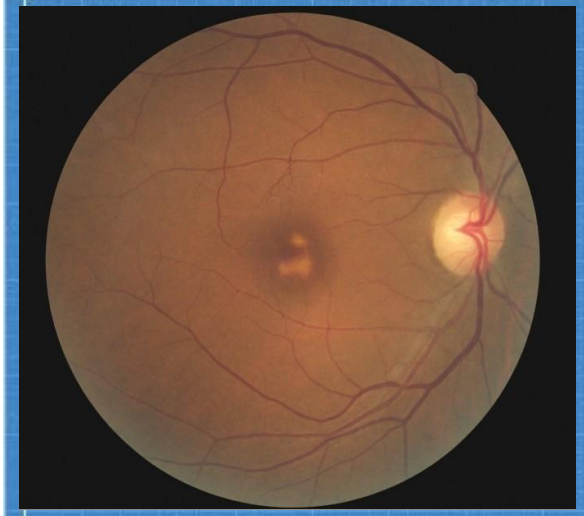
67-year-old Black female

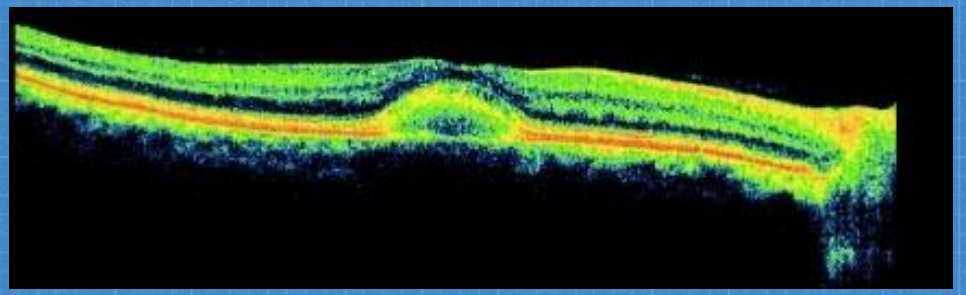
Decreased vision OD>OS; has stopped reading completely

PMH:
(+)Migraines
(+)Hypothyroidism

POH:
(?ARMD): given vitamins in past

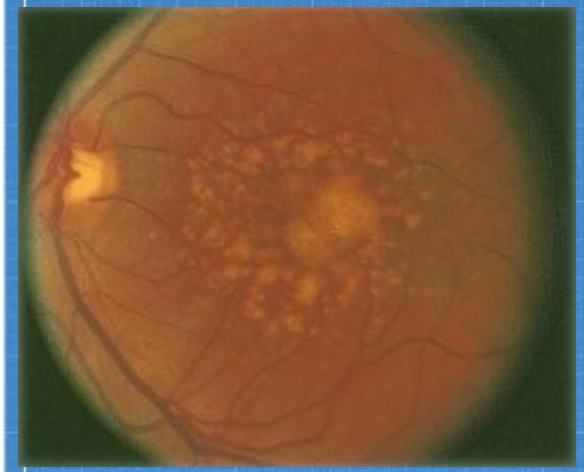

What are some differentials?
20/30
20/20
2

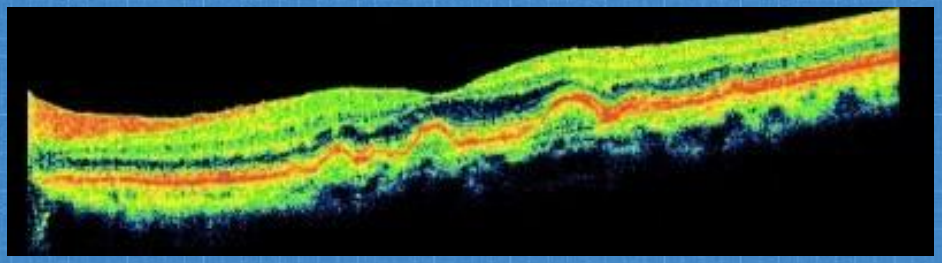




Sunny Side Up: Vitelliform Dystrophy

This condition is often misdiagnosed as AMD. Find out how to differentiate





What is AOFVD?
(Adult onset foveomacular vitelliform dystrophy)

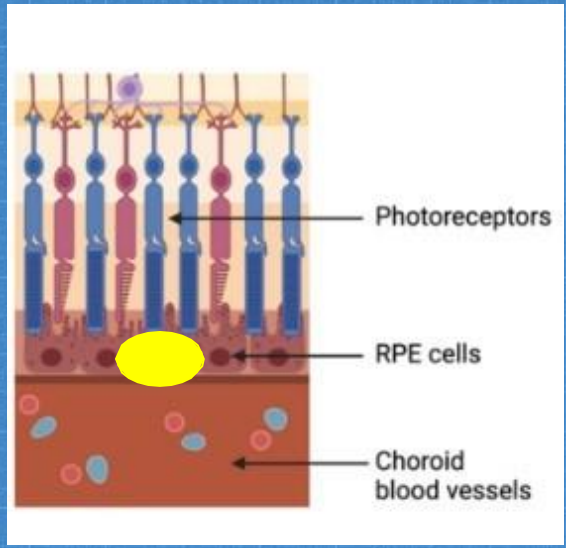
- Patients present in the early stages with minimal to no symptoms
- Progressive disease with no treatment

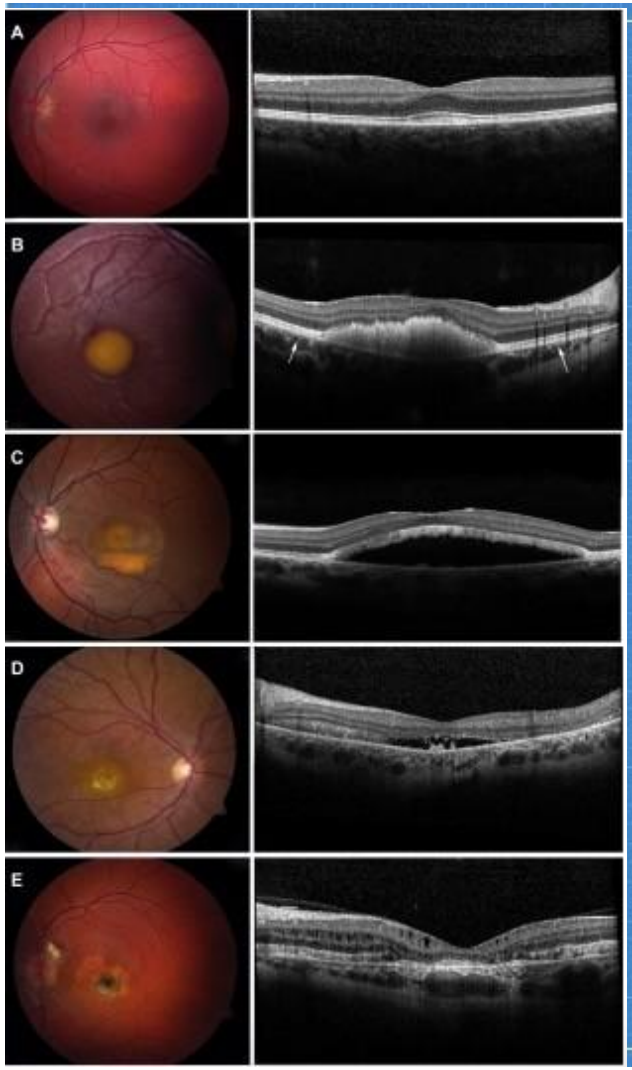
Because of relatively mild vision symptoms during most stages and its resemblance to AMD, this condition is often misdiagnosed.

How do we differentiate from AMD?

Vitelliform and AMD share a malfunction at the level of the RPE and choroid

- Vitelliform lesions have high levels of lipofuscin
- Believed to be the result of accumulation of photoreceptor outer segments that failed to be digested by the RPE leading to buildup in the subretinal space





Previtelliform

Vitelliform- Egg yolk lesion under the fovea

Pseudohypopyon- Layering of heavy proteinaceous material inferiorly, due to gravity; superior part of lesion looks clear or fluid filled

Vitelliruptive- Resembles scrambled eggs; atrophy starting due to resportion of vitelliform material

Atrophic- material completely resolved; loss of RPE and outer retina

https://www.aaojournal.org/article/S0161-6420%2816%2932058-9/fulltext

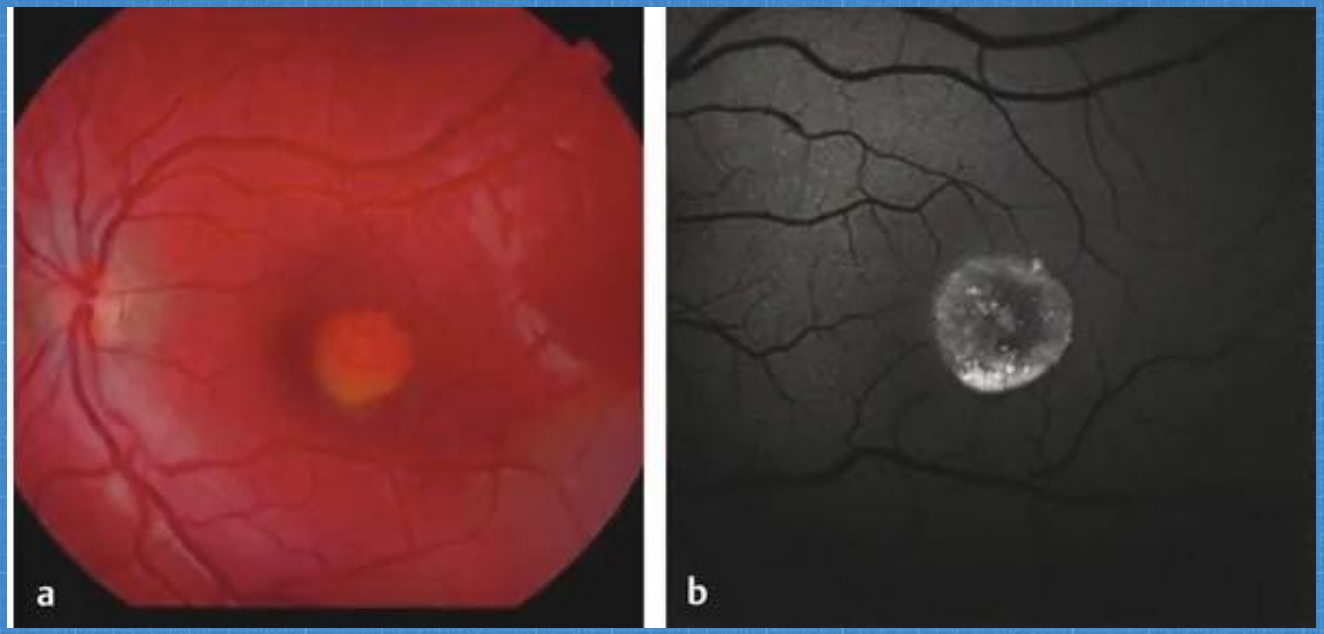
So…. How do we differentiate?

AOFVD is usually:

- Bilateral; not associated with surrounding atrophy; absence of drusen
- Earlier age of onset; 30-40s
- Subretinal lesion, uniform hyper-reflective lesion

- Ancillary testing
  - OCTA, FAF

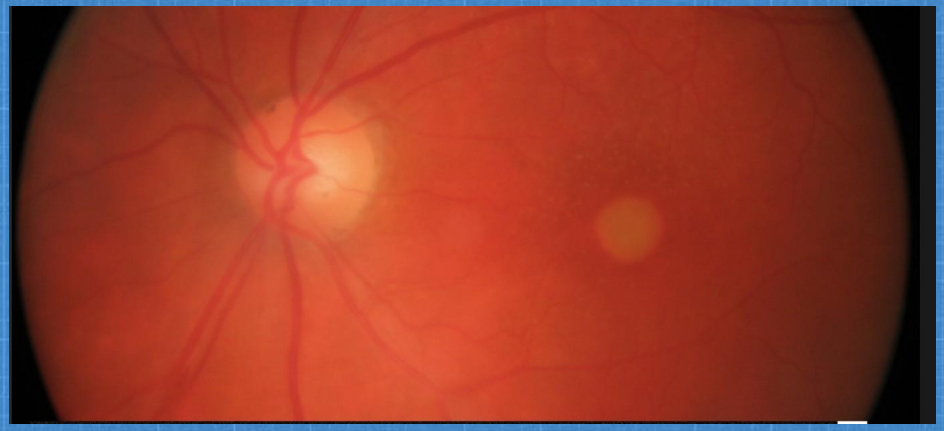
Fundus autofluorescence

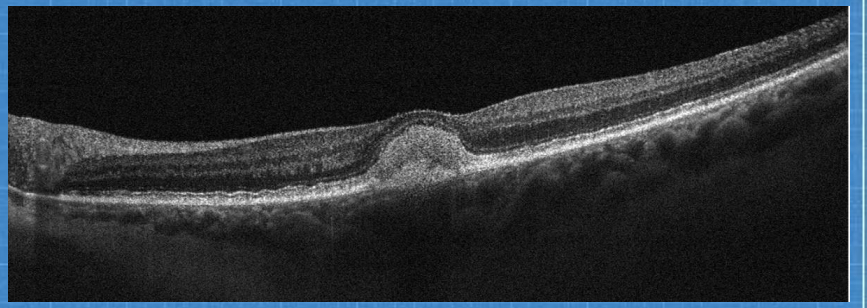


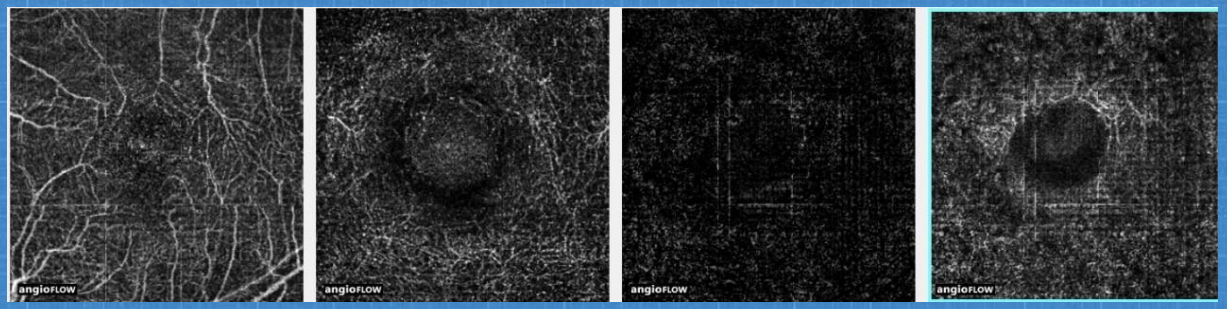
Hyperautofluorescent lesion corresponding to the yellow spot

https://entokey.com/best-vitelliform-macular-dystrophy-best-disease/

OCTA

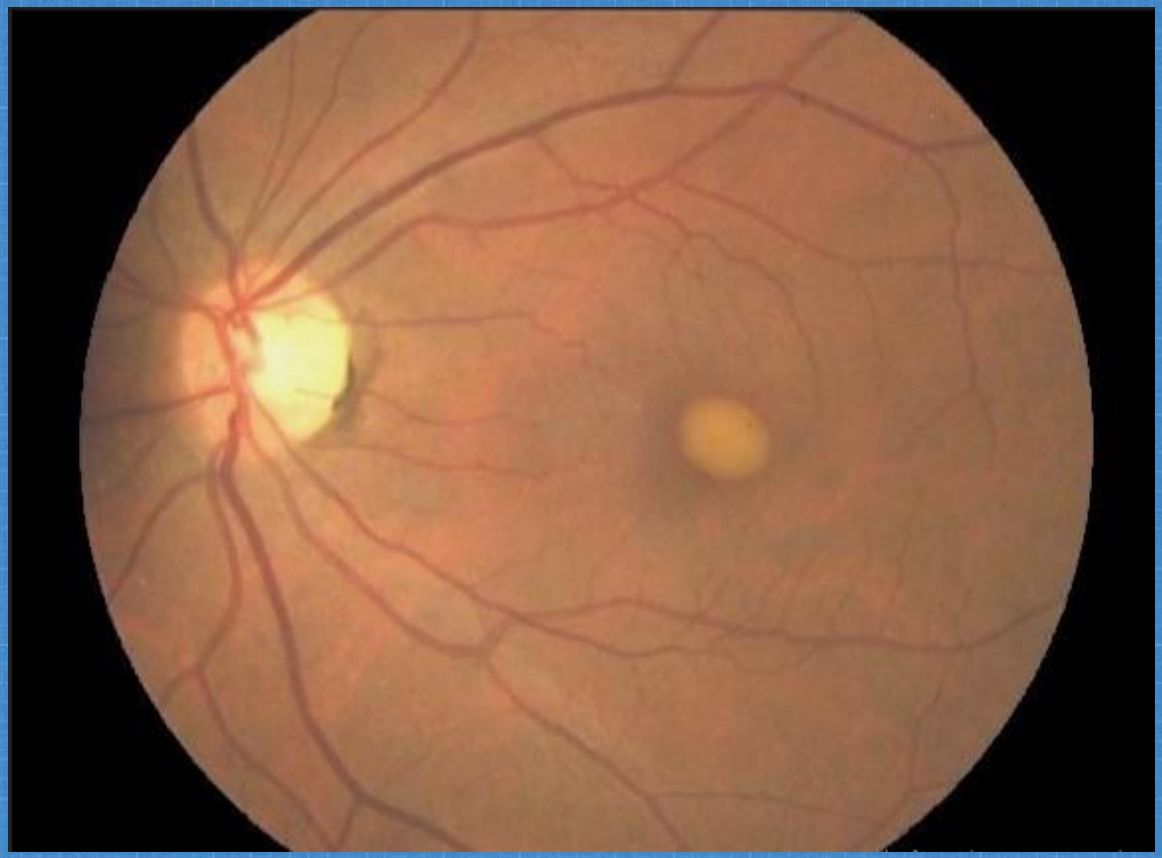


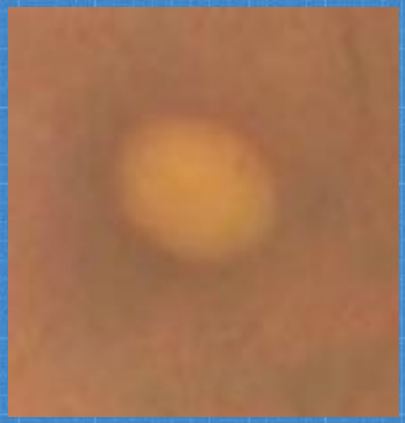


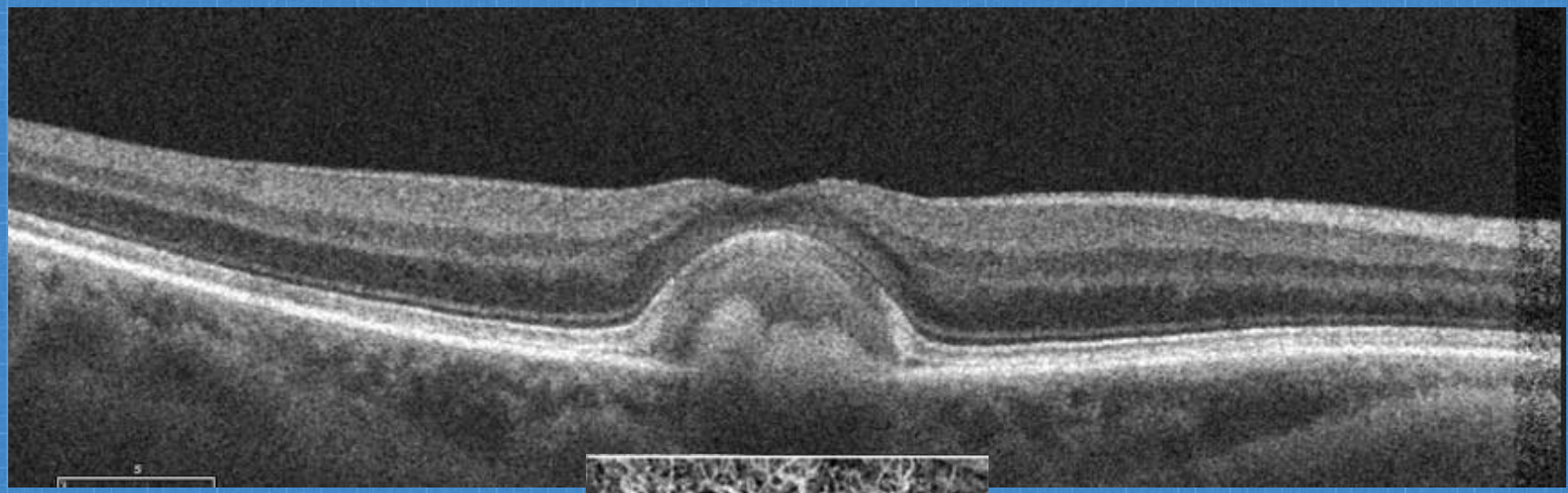


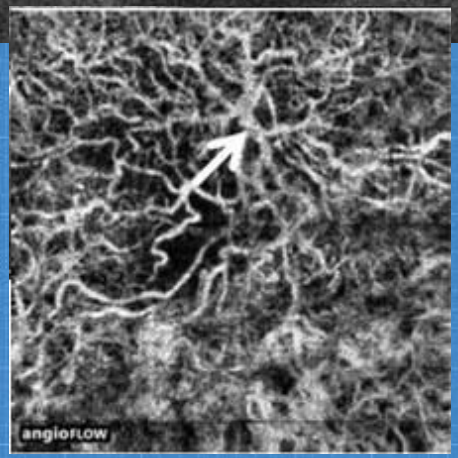
Vitelliform lesions blocks signal; can pick up leakage

…Another









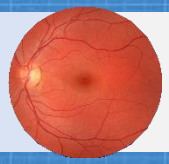
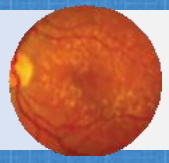
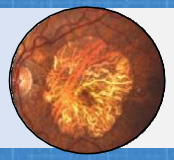
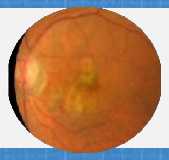
What do you see?

…And a review of AMD using OCT!

The Beckman Classification

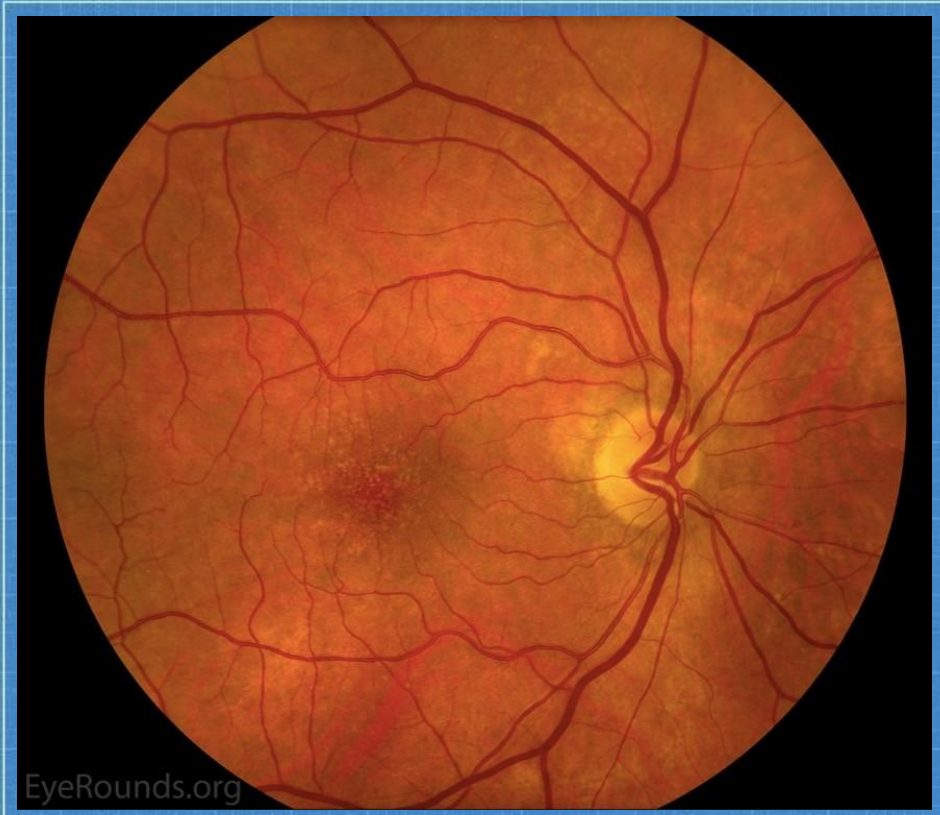
4 Stages of AMD

PROGRESSION

No AMD		No drusen or small drusen ≤ 63 µm No AMD pigmentary abnormalities
Early AMD		Medium drusen > 63 µm and ≤ 125 µm No AMD pigmentary abnormalities
Intermediate AMD		1 large druse > 125 µm and/or Any AMD pigmentary abnormalities
Advanced AMD	 	**2 forms:** Geographic Atrophy and Neovascular AMD

Geographic Atrophy Neovascular AMD

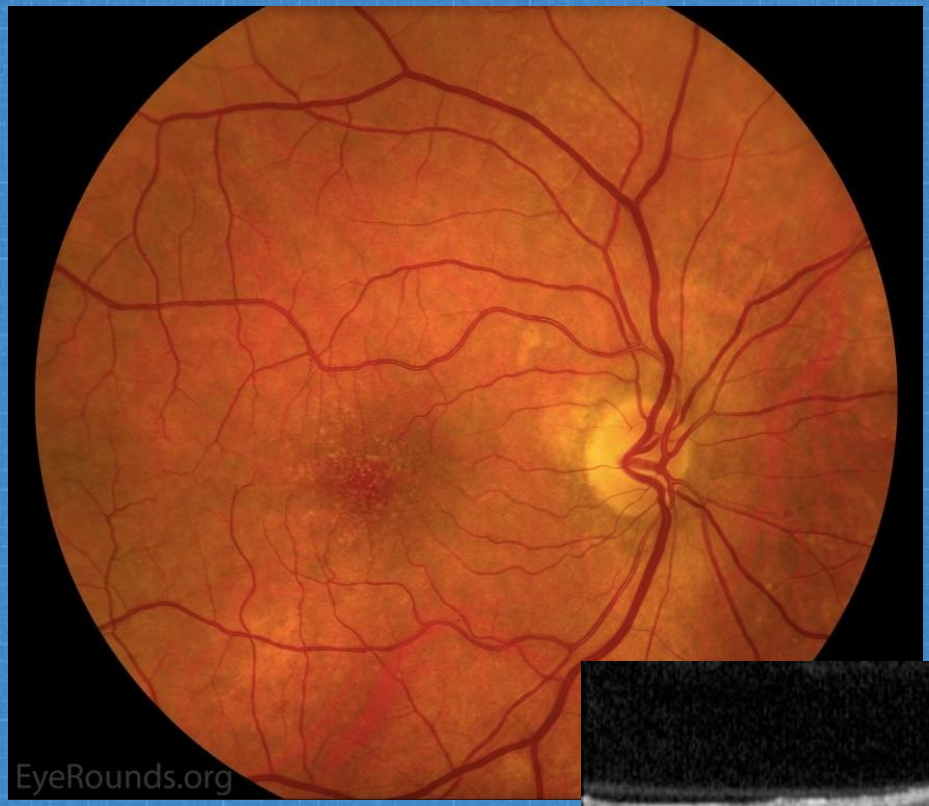
- sources: AMD, age-related macular degeneration. Ferris FL, et al. Ophthalmology. 2013;120(4):844-851.

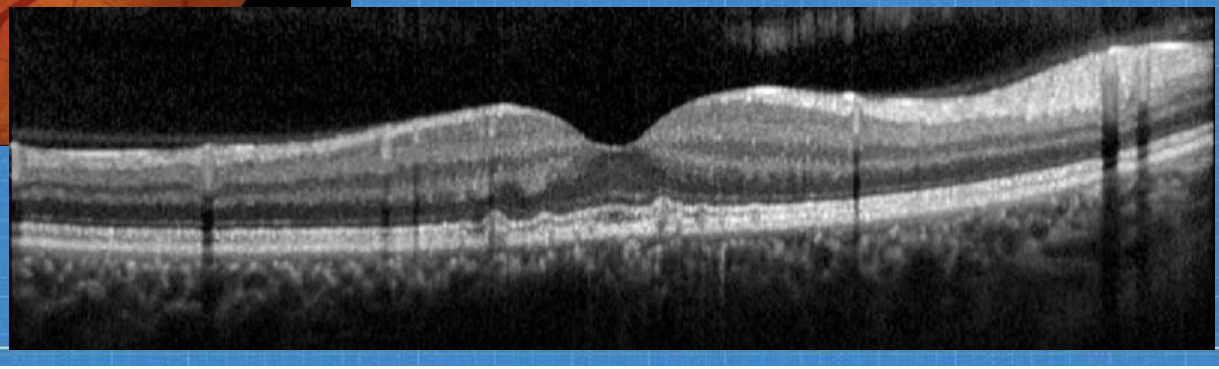


What might this look like clinically?

Early AMD

Medium drusen > 63 µm and ≤ 125 µm
No AMD pigmentary abnormalities


EyeRounds.org



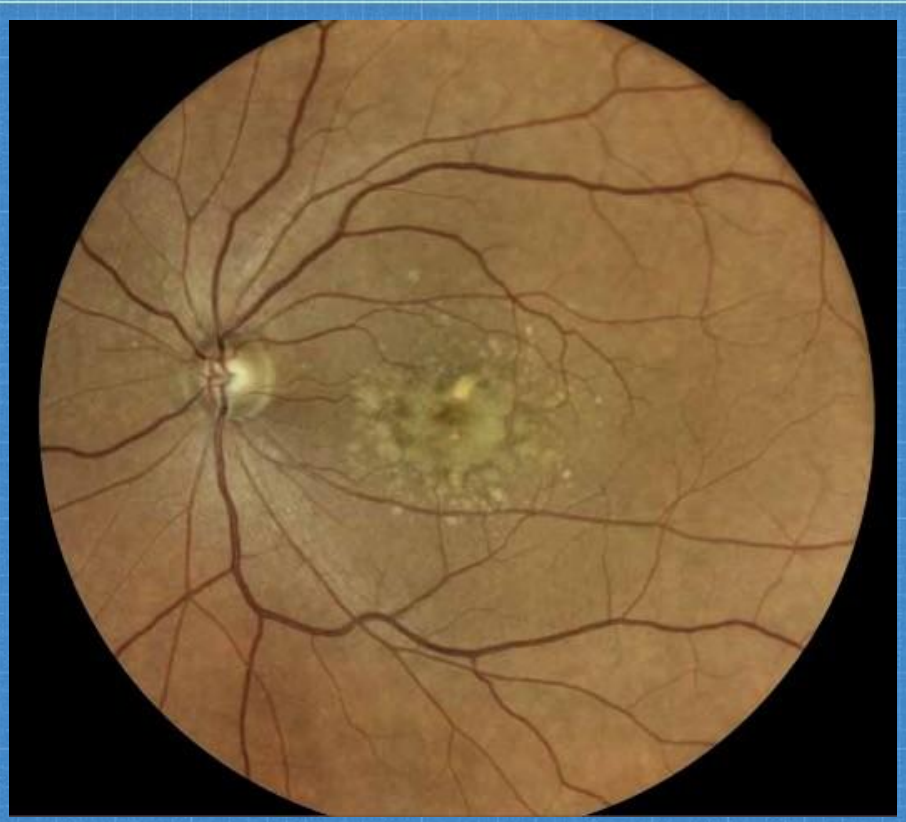
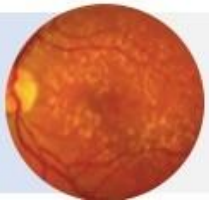


Image courtesy of iCare

What might this look like clinically?

Intermediate AMD



1 large druse > 125 µm and/or

Any AMD pigmentary abnormalities

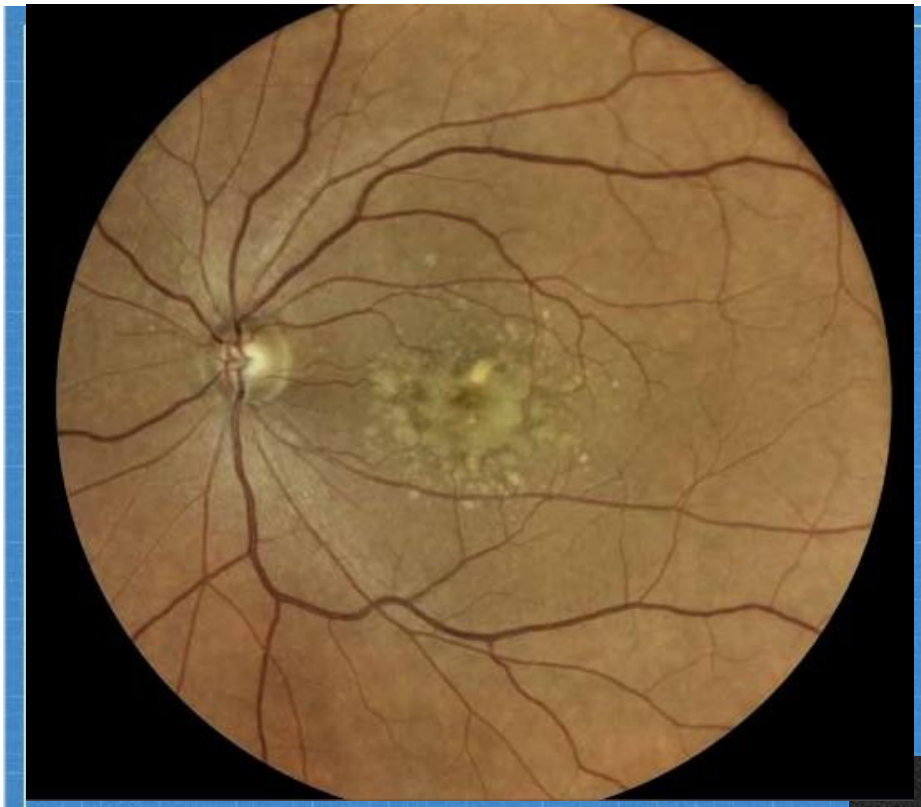
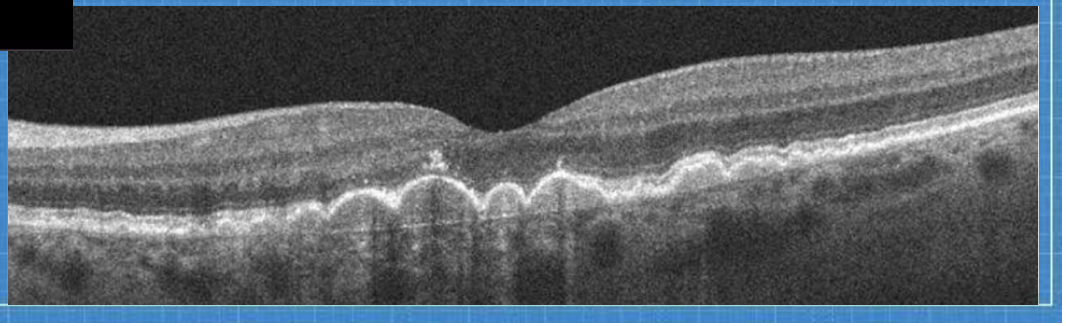
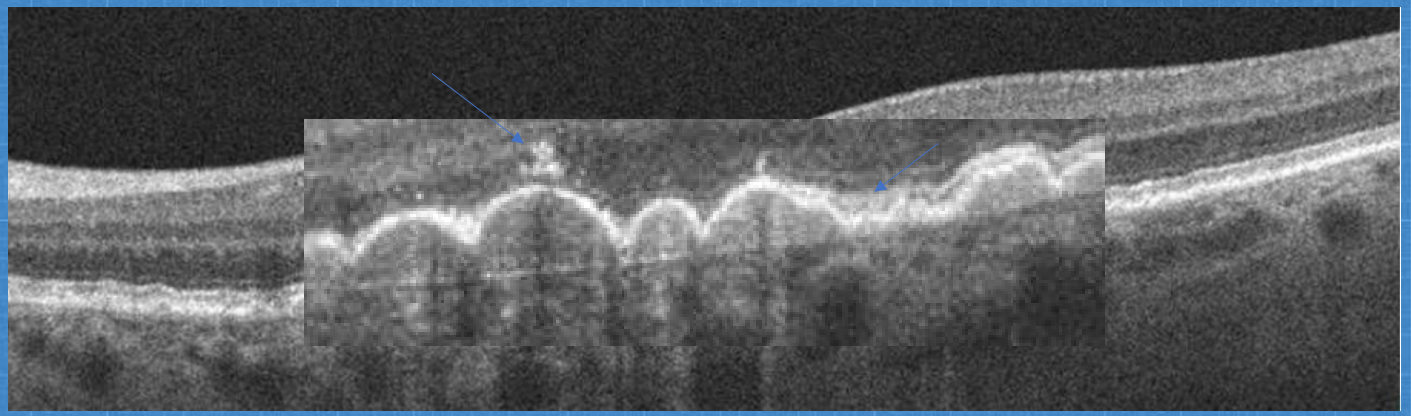
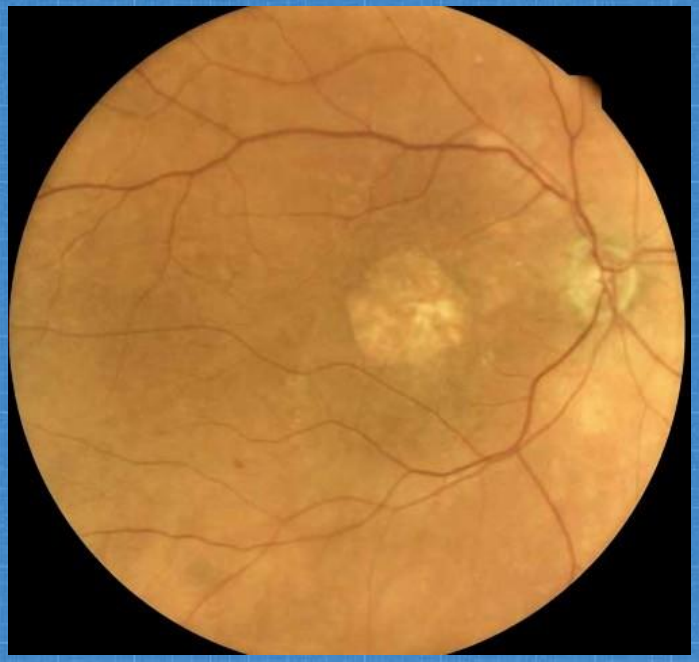


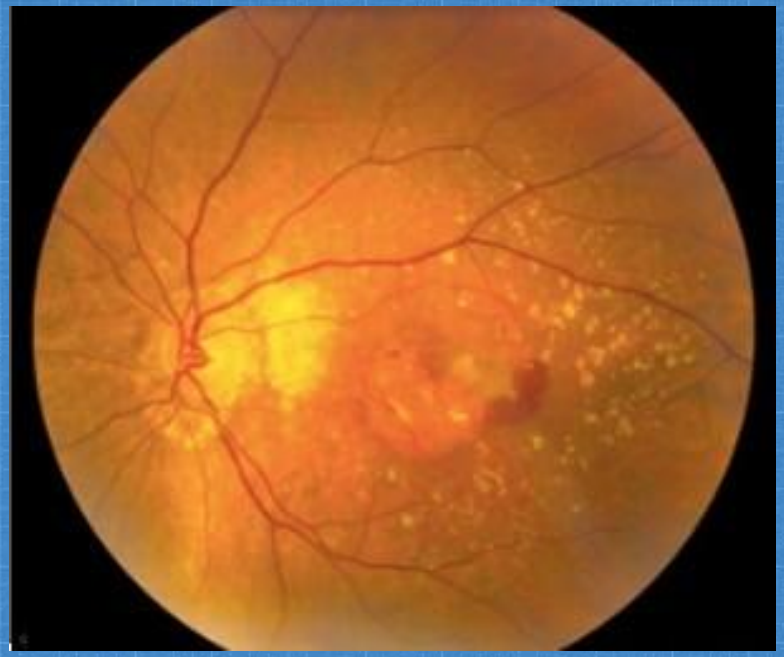
Image courtesy of iCare





What might this look like clinically?





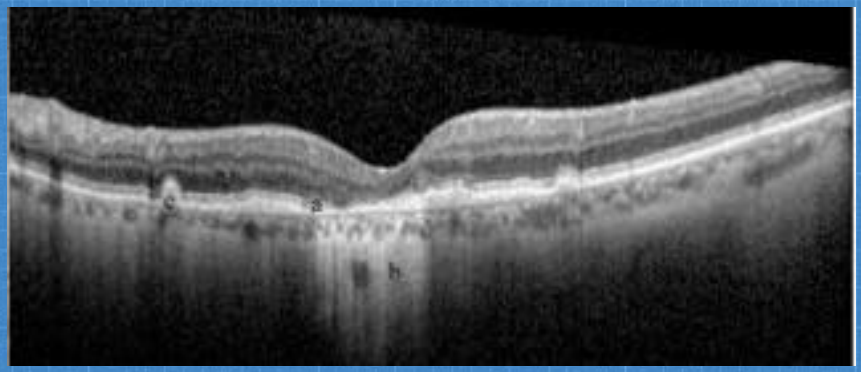
Advanced AMD

Geographic Atrophy

Neovascular AMD

2 forms: Geographic Atrophy and Neovascular AMD

Geographic Atrophy outside of fovea



Advanced AMD

Geographic Atrophy

Neovascular AMD

2 forms: Geographic Atrophy and Neovascular AMD

Advanced AMD

Geographic Atrophy

Neovascular AMD

2 forms: Geographic Atrophy and Neovascular AMD

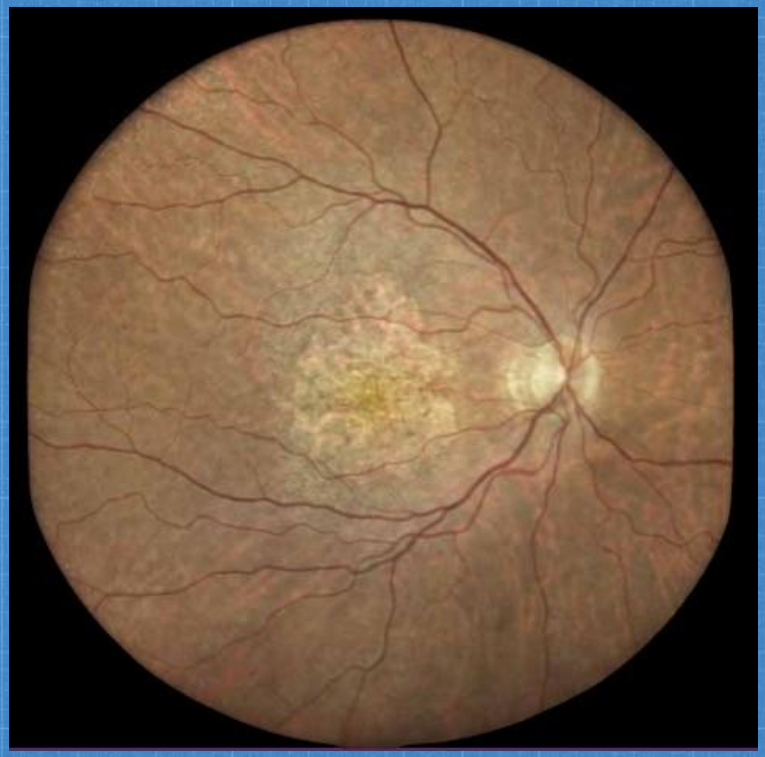
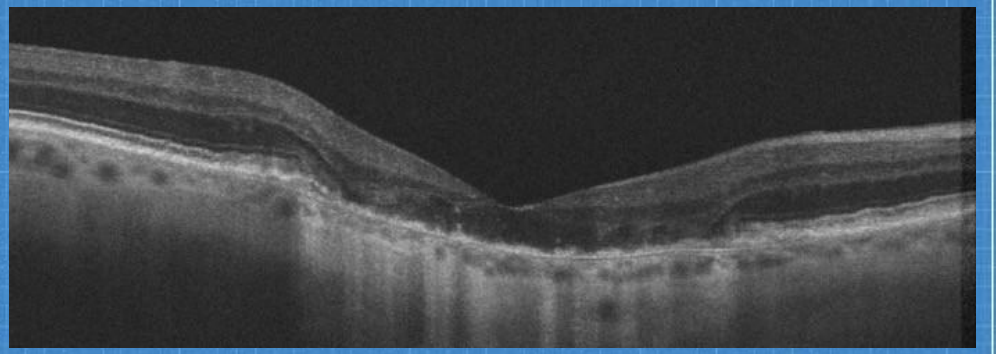


Image courtesy of iCare

Geographic Atrophy involving fovea



Advanced AMD

Geographic Atrophy

Neovascular AMD

2 forms: Geographic Atrophy and Neovascular AMD

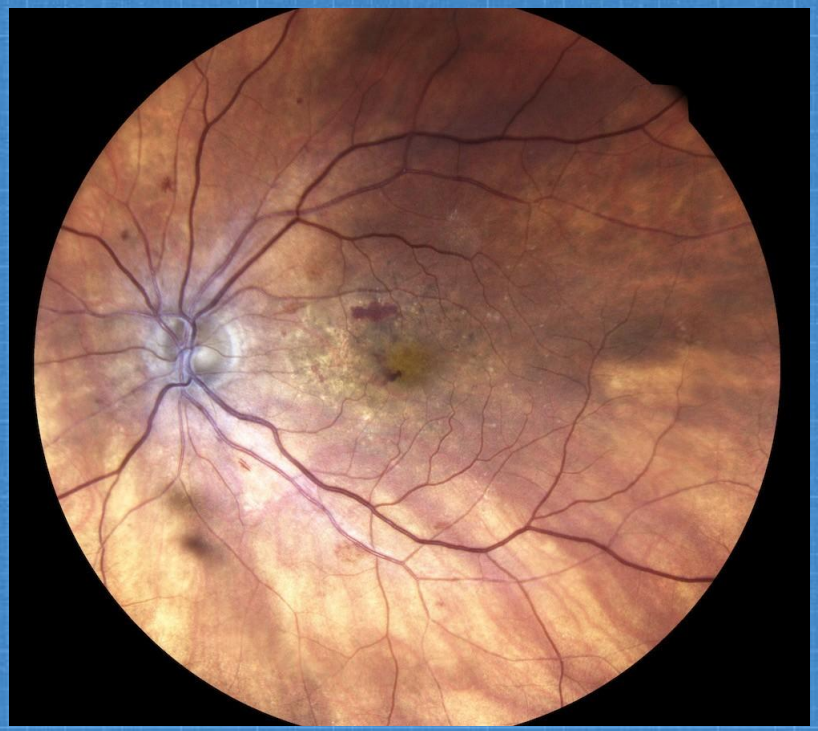
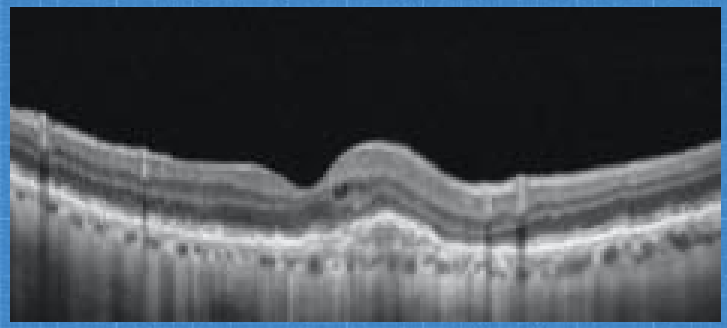


Image courtesy of iCare

Neovascular AMD




THANK
YOU