



Retinal Imaging Grand Rounds

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Financial disclosures

Nothing to disclose



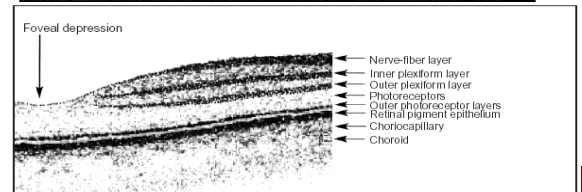
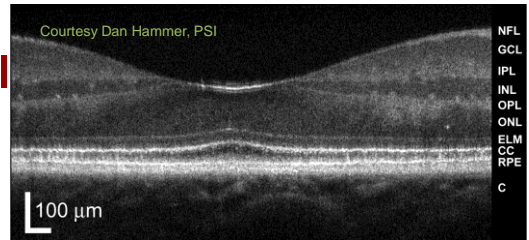
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OCT

- *in vivo* histology
- Working mechanism: similar to B-scan but uses infrared light instead of sound to assess reflectivity
- Resolution: 3-5 microns with SD/SS technology
- Different optical reflectivity in various tissue structures



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OCT evolution

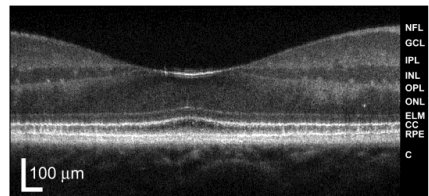
- Time Domain.....then.....
- Spectral Domain (most current commercial instruments).....now....
- Swept Source and OCTA. Swept Source.....
- Twice as fast (twice as many A-scans / second) as SD OCT
- Allows for wide field imaging (12mm vs. 6-9 mm). Easily gets ONH and macula in the same scan
- Longer wavelength of light, so can image much more effectively through media opacities, and penetrates much better into the choroid (2.6 mm depth vs. 2.3mm)



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PIL (Photoreceptor integrity line) / Ellipsoid zone / ISOS junction

- Line seen at junction of inner and outer segments of the photoreceptors
- Extremely useful for evaluating disease state and visual potential
- Very important!



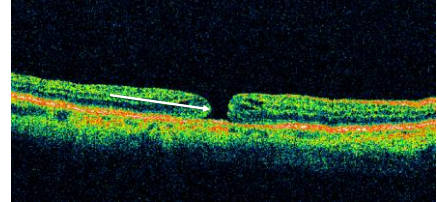
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Vitreoretinal Interface Disorders

- Epiretinal Membrane
- Vitreomacular adhesion / Vitreomacular Trtraction
- Partial Thickness Macular Hole
- Full thickness Macular Hole
- Macular pseudohole

Macular hole sizes (full thickness)

- Small \leq 250 microns
- Medium 250-400 microns
- Large $>$ 400 microns
- Horizontal diameter at narrowest point



Grading system for VMA / VMT

- VMA with no change in foveal contour: Stage 0
- VMT with disruption of foveal contour: Stage 1
- VMT with small or medium FT hole: Stage 2
- VMT large full thickness hole: Stage 3
- Any full thickness hole without VMT: stage 4
- Lamellar hole
- Pseudohole from ERM

VAST study : How common is VMA / VMT?

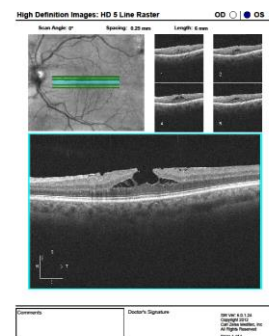
- 1950 eyes
- Age 40-89 years
- Phakic
- No pre-existing maculopathy
- No history of vitrectomy or Jetrea
- VMA prevalence of 39%
- VMT prevalence of 1%
- Most common in 40's and 50's, then decreases with age (25% VMA & 2% VMT over age 63)
- AA 55% less than Caucasians
- Not significantly associated with sex, refractive error, or visual acuity status

ERM

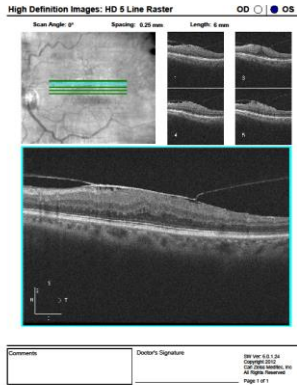
- Membranous growth of glial cells on retina surface
- Can be asymptomatic or very bothersome
- Metamorphopsia is common
- More common after PVD
- Tractional macular holes, cysts, CME, neurosensory RD's; retinal and choroidal folds, etc.

20/25!

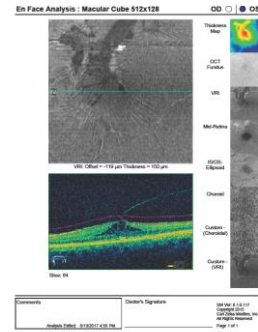
- ERM
- Foveal schisis



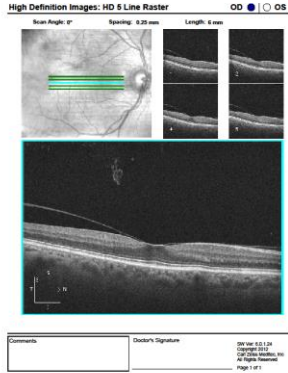
- VMT
- ERM



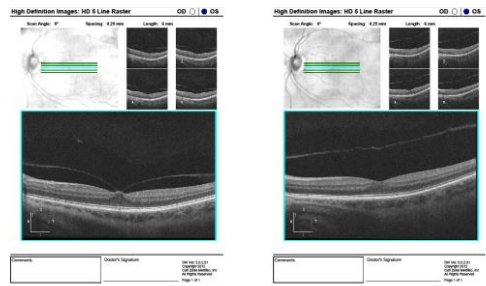
Enface



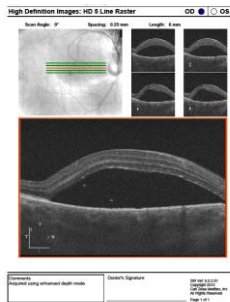
- VMA
- "Floater"



VMT spontaneous resolution after 3 months



ICSC



Pachychoroid diseases

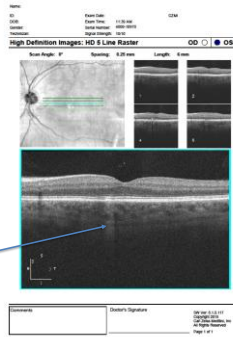
All share attenuated choriocapillaris, dilated choroidal veins, thickened choroid (definitely thick at 390 microns or greater), RPE dysfunction, and potential for CNV

Pachychoroid Pigment epitheliopathy, CSR, Polypoidal choroidal vasculopathy, Peripapillary pachychoroid, FCE

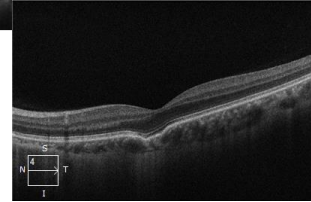
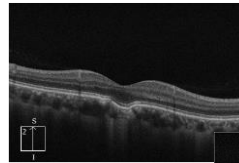
Use EDI on OCT

Measure with calipers at the fovea
100X HD single line is excellent

Choroid-sclera junction

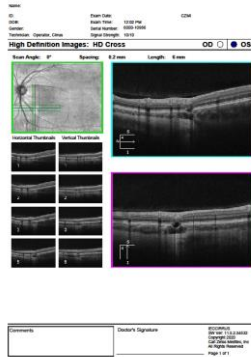


Focal choroidal excavation

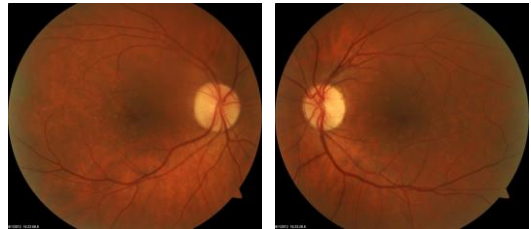


Conforming (here) or nonconforming

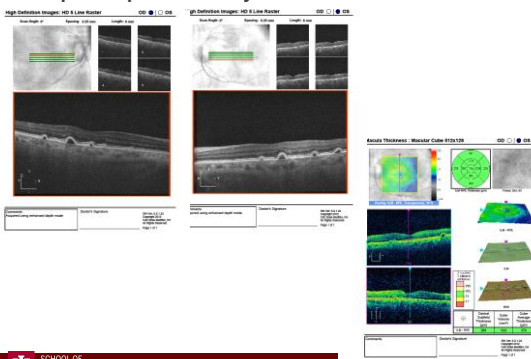
Non-conforming FCE



"Drops of water on a windshield"



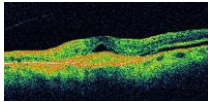
Multiple idiopathic PED syndrome



Macular Degeneration

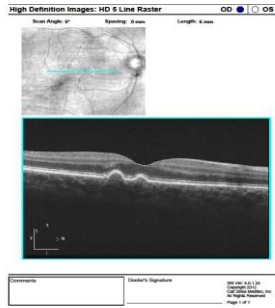


AMD

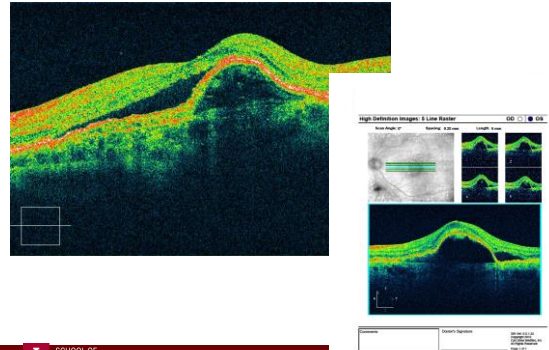


CNV

Drusen

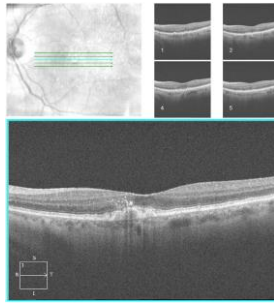


Fluid.....or something else?

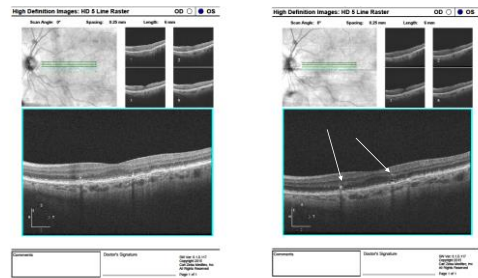


Hyper-reflective foci

- Dot shaped intraretinal lesions at the apex of drusen
- Often correspond to focal hyperpigmentation
- Start in the outer retina and migrate inward
- Likely represent pigment granules
- Ancillary AREDS II OCT study showed them to be associated with a 5X risk of geographic AMD in two years. No extra risk of CNV

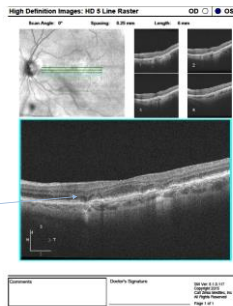


Hyper-reflective foci



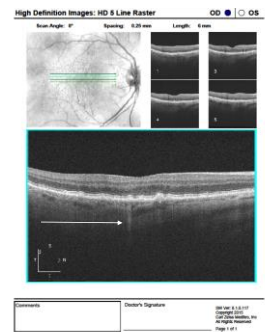
Nascent geographic atrophy / iRORA (incomplete RPE and outer retinal atrophy)

- Thinning of the OPL and INL with a hyporeflective wedge and choroidal transmission defect
- No photoreceptor or RPE loss
- Strongly associated with impending GA
- No extra risk of CNV
- cRORA: (Complete RPE and outer retinal atrophy) on OCT: may not yet show up on photos, but will on FAF



Sub-RPE hyper-reflective columns/ hypertransmission defects

- Increased transmission of signal columns beneath the RPE (hyper-reflective)
- Overlying RPE appears intact
- May represent fine cracks in the RPE
- Opposite appearance of shadows cast by retinal blood vessels
- Extra risk of geographic disease and CNV

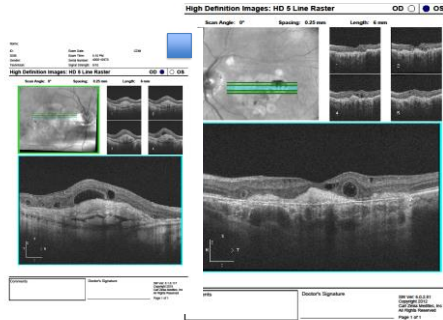


Miscellaneous Retinal Conditions



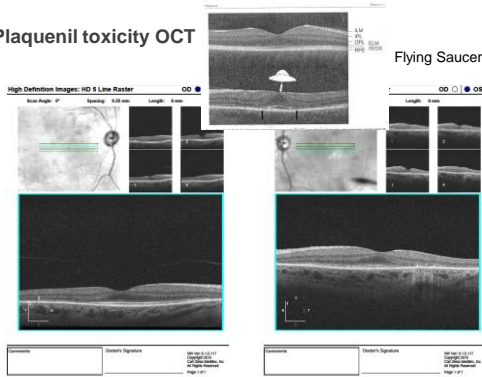
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ORT's



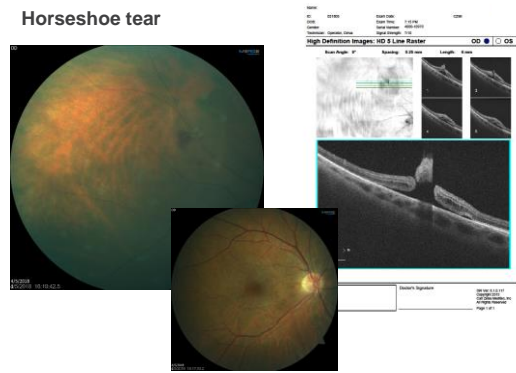
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Plaque toxicity OCT



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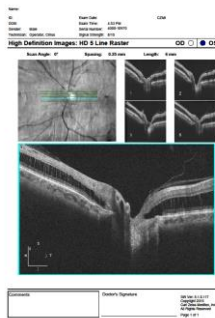
Horseshoe tear



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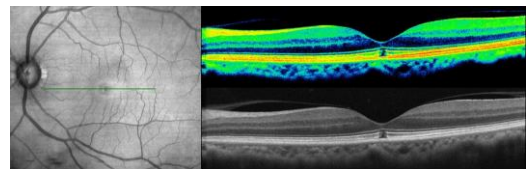
Peripapillary retinoschisis

- Underappreciated
- 3-5 % of glaucoma patients
- Macula rarely involved
- Can have a visible laminar pit or not
- Can be affected by IOP
- Can affect RNFL readings
- Can resolve
- Also seen in .5% or so of normals



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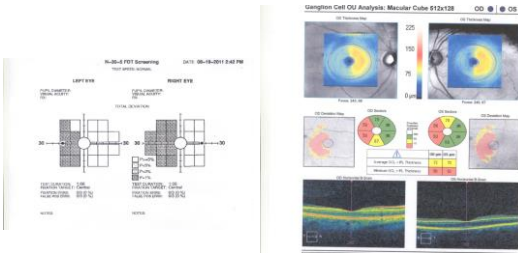
Solar maculopathy (images courtesy Dr. Jerome Sherman)



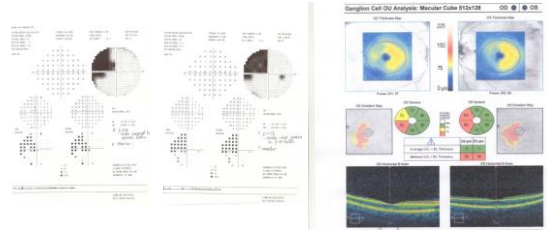
Damage to outer photoreceptors/ inner RPE

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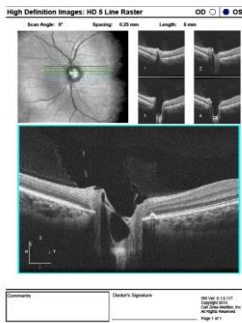
Stroke and GCC loss



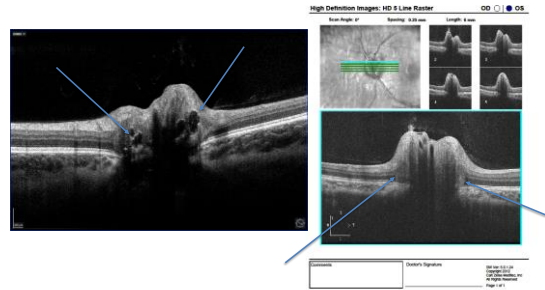
Stroke and GCC loss



PARTIAL PVD



ONH DRUSEN

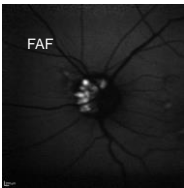


• Improved with EDI

PHOMS

ONH drusen detection with OCT

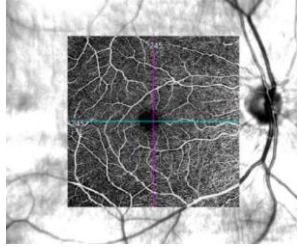
- Optic Disc Drusen Consortium Consensus.....
- Always use EDI
- Blood vessels are more solid, cast a shadow, and can show as figure 8
- Drusen always prelaminar
- Drusen always hyporeflective, but can conglomerate, causing some internal reflectivity in that case
- Drusen often have a hyperreflective border, especially superiorly
- The old concept of a hyporeflective fluid wedge at the edge of the nerve in true papilledema DOES NOT APPLY with SD-OCT. Was a time domain OCT artifact.



OCTA

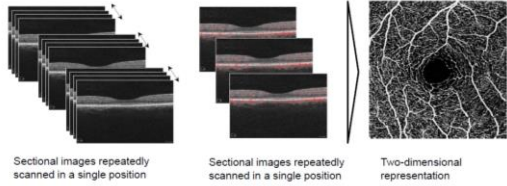
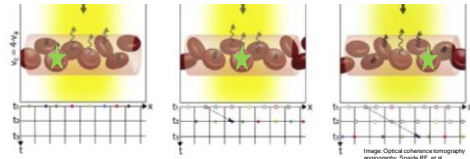
What is it?

- Additional application of the spectral domain OCT
- Noninvasive, imaging modality that provides volumetric images of retinal and choroidal vasculature



How does it work?

- Uses motion contrast to detect flow
- Based on properties of light reflected from blood cells



Sectional images repeatedly scanned in a single position

Sectional images repeatedly scanned in a single position

Two-dimensional representation

Advantages

- Noninvasive
- Dye-less
- Fast
- Higher resolution
- Can view both structure and vascular information
- Imaging of retinal and choroidal vasculature
- Volumetric data with segmentation allows for precise identification of lesion
- No leakage

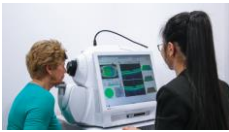


Photo: Zeiss

- Comprehensive information about structure and vascular perfusion at multiple levels of the retina and choroid

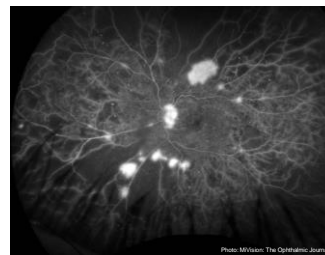
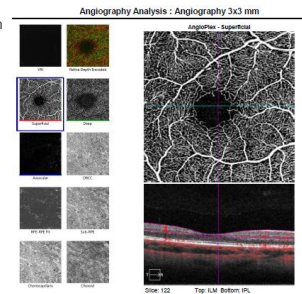


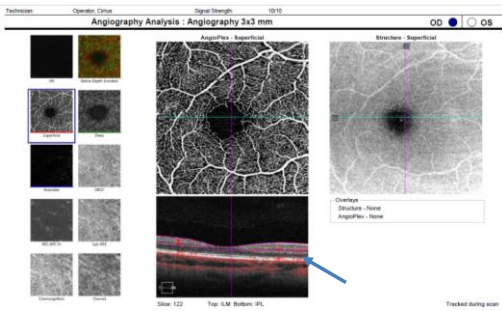
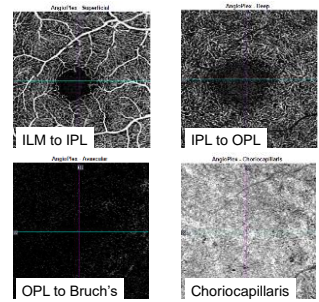
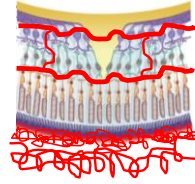
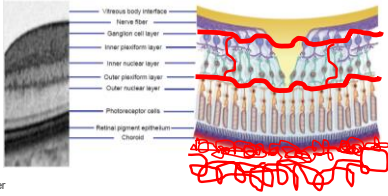
Photo: Mivison: The Ophthalmic Journal

Disadvantages

- Limited field of view
- Artifacts
- No dye leakage
 - Cannot detect leakage, pooling, or staining
- Slow moving blood may result in poor detection of pathology

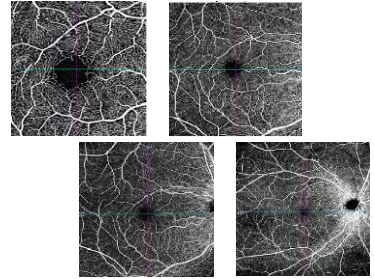
Retinal Circulation

- Central Retinal Artery
 - Superficial capillary plexus
 - Within ganglion cell layer
 - Deep capillary plexus
 - Below inner nuclear layer
- Choroid
 - Choriocapillaris
 - Fine capillaries
 - Sattler's and Haller's layer
 - Medium and larger diameter vessels



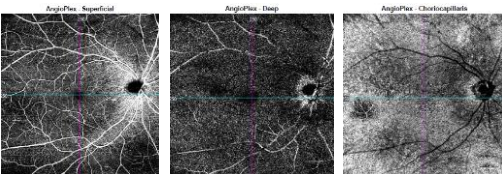
Scans

- 3mm x 3mm
 - Smaller scan size, with high mag
 - Best resolution
- 6mm x 6mm
 - Larger scan size, with less mag
 - Optimum coverage and resolution
- 8mm x 8mm
 - Larger scan size
 - Weaker image quality
- 12mm x 12mm
 - Largest scan size
 - Weakest image quality

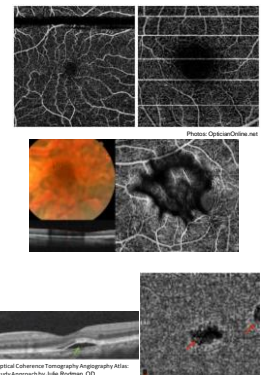


Artifacts

- Projection
 - Fluctuation in shadows from superficial vessels in deeper structures that instrument identifies as real



- Motion
 - Image displacement due to blink or saccade
- Media opacities
 - Cornea, cataract, vitreous
- Shadowing
 - Region of attenuated signal due to overlying lesions
 - Dense: Hemorrhage, fibrosis, RPE hyperplasia
 - Not Dense: PED, macular edema

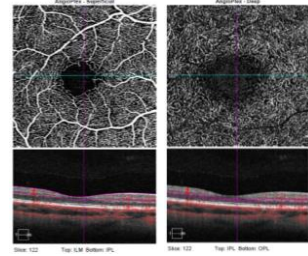


How is this useful?

- **Inner Retinal Diseases**
 - Diabetes
 - Vein Occlusions
 - Mac Tel
 - Paracentral Acute Middle Maculopathy
- **Outer Retinal Diseases**
 - CNVM
 - AMD
 - CSCR
 - Others
- **Optic Nerve Head Diseases**
 - Glaucoma
 - Neovascularization of the Disc

Evaluating Inner Retinal Disorders

- Evaluate OCTA and B-scans
 - SCP
 - DCP
- Evaluate FAZ



Diabetic Retinopathy

- Hallmark of diabetes is changes in vasculature
- OCTA may reveal diabetic retinopathy before it is clinically detectable
- Non-proliferative Diabetic Retinopathy
 - Enlargement of Foveal Avascular Zone
 - Areas of capillary nonperfusion and dropout
 - MAs

Enlargement of FAZ

- Represents macular ischemia
- Enlarged in both SCP and DCP in diabetes compared to healthy patients

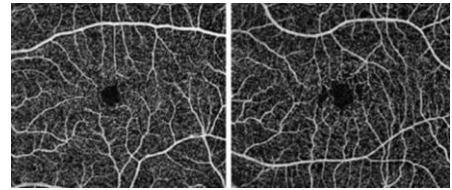
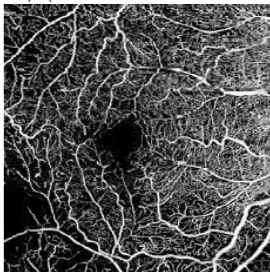


Photo: Quantitative optical coherence tomography angiography features for objective classification and staging of diabetic retinopathy. Adam ML, et al.

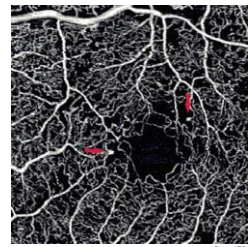
Capillary Nonperfusion

- Represents ischemia and areas of capillary dropout
- Vessel density is defined as the proportion of blood vessel area over the total measured area



Microaneurysms

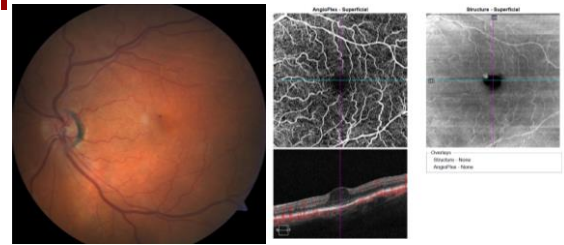
- May or may not be visible
 - Due to slow moving blood in MAs





- Visible MAs showing flow

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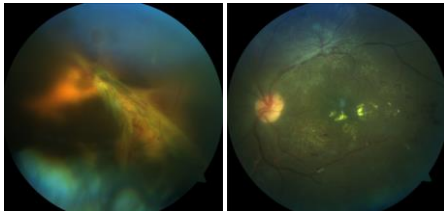


- No-flow MA

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Proliferative Diabetic Retinopathy

- Differentiating IRMA from neovascularization
 - Based on location
 - Using flow overlay



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IRMA

- Shunt vessels that act to supply areas of non-perfusion
- Located intraretinally
- Flow overlay shows no ILM breach

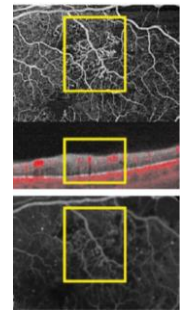


Photo: Differentiating intraretinal microvascular abnormalities from retinal neovascularization using optical coherence tomography angiography. Arora M, et al.

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Proliferative Neovascularization

- Breached the ILM and extends into vitreous
- Flow overlay shows positive flow signal within the neovascular complex

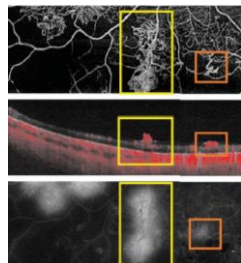
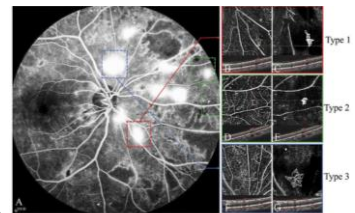


Photo: Differentiating intraretinal microvascular abnormalities from retinal neovascularization using optical coherence tomography angiography. Arora M, et al.

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NVE detailed analysis with OCTA (on IVFA, all types leak and look the same)

- **Three Types**
 - **Type 1:** Originates from venous side with tree-like appearance
 - May have higher risk of vitreous hemorrhage or RD
 - **Type 2:** Originates from capillary networks in an octopus-like shape
 - May respond better to PRP
 - **Type 3:** Originates from IRMAs in sea fan shape



Classification of NVE with a combination of FA and OCTA. A. NVEs demonstrated as FA with marked dye leakage. B and C: A Type 1 NVE originating from the venous side with a tree-like appearance. B and E: A Type 2 NVE originating from capillary networks, with an octopus-like shape. F and G: A Type 3 NVE originating from IRMAs, with a sea fan shape. Photo: Novel three types of neovascularization elsewhere determine the differential clinical features of proliferative diabetic retinopathy. Pan J, et al.

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Classification of VRI in NVE

- **Arch bridge**
 - Extends along partially detached vitreous, attached to retina by vascular "roots"
- **Flat**
 - Extends along attached vitreous surface with short attachments to retina
- **Forward**
 - Transverses vitreous surface and extends into vitreous cavity
- **Flat-forward**
 - Combination

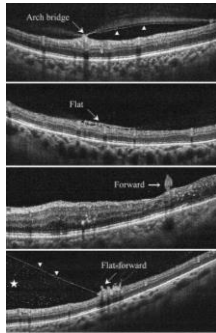
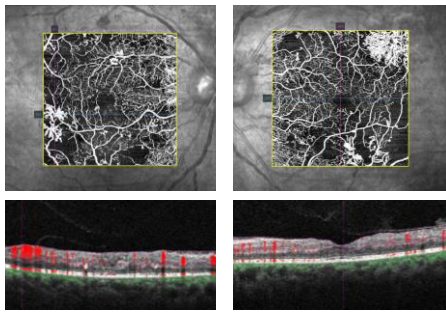
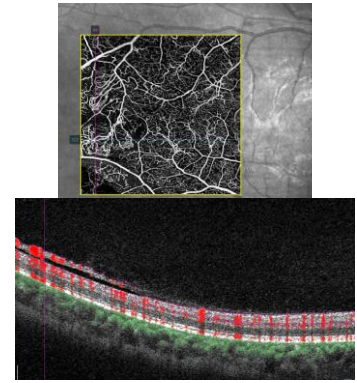


Photo: Novel three types of neovascularization elsewhere determine the differential clinical features of proliferative diabetic retinopathy. Park J, et al.

NVE example

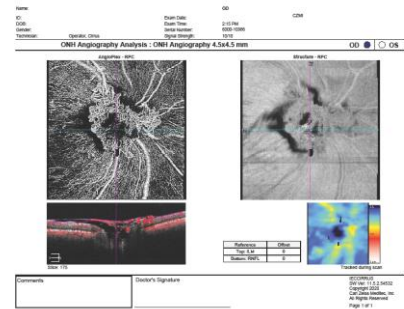
- Type 3, "flat"



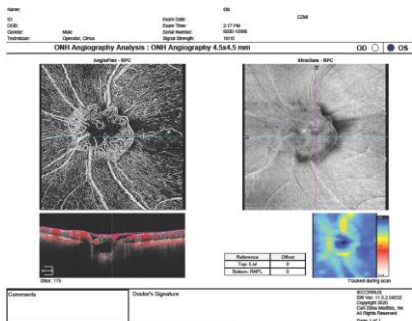
IRMA

NVE

NVD in PDR

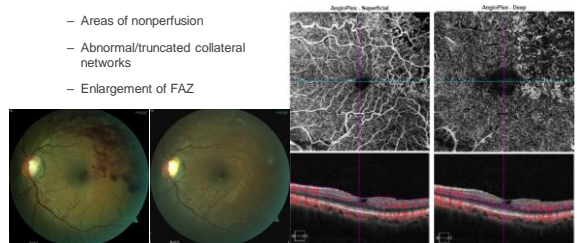


NVD in PDR

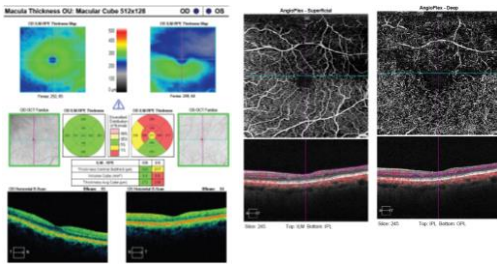


Vascular Occlusions

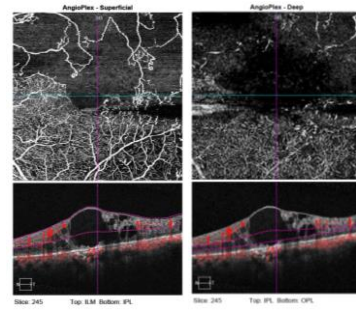
- BRVO/CRVO
 - Areas of nonperfusion
 - Abnormal/truncated collateral networks
 - Enlargement of FAZ



Hemi-CRVO

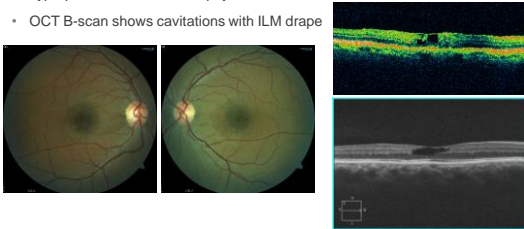


Hemi-CRVO



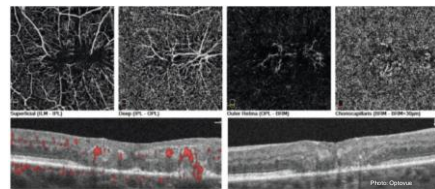
Macular Telangiectasia Type 2

- Most Common
- Ectatic capillaries of macula that dive into deeper retinal tissues
- Fundoscopy shows hemorrhages, right-angled venules, RPE plaques and hyperplasia, and retinal atrophy
- OCT B-scan shows cavitations with ILM drape

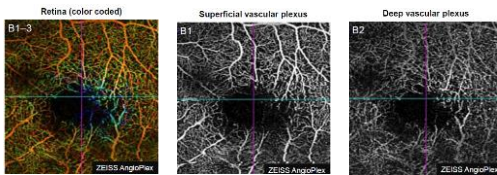


Mac Tel 2

- OCTA is perfectly suited for Mac Tel 2, due to its distinct vascular changes
- Right-angled draining venules
- Ectatic capillaries at SCP



Mac Tel 2



Paracentral Acute Middle Maculopathy (PAMM)

- Ischemia of the retinal intermediate/deep capillary plexus
- Presents with acute scotoma, may have decreased VA (depending on foveal involvement) or asymptomatic
- May see gray or white lesion on fundus exam in acute stage
 - Resolves over a few weeks, then cannot appreciate funduscopically
 - Diagnosis relies heavily on OCT and OCTA appearance



PAMM

- Isolated presentation
 - Idiopathic
 - Other reported associations: flu-like illness, post-vaccination, medications, high caffeine intake, migraines, pregnancy, cosmetic fillers and more
- Finding in retinal vasculature disease
 - Diabetic retinopathy, HTN retinopathy, sickle cell, purtscher's retinopathy
 - Arterial and venous occlusions

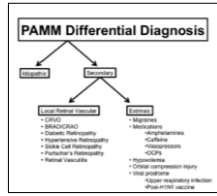
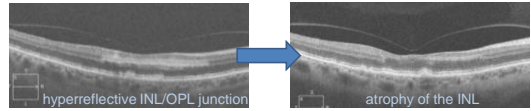


Photo: Rahmy E. Retina, 2015

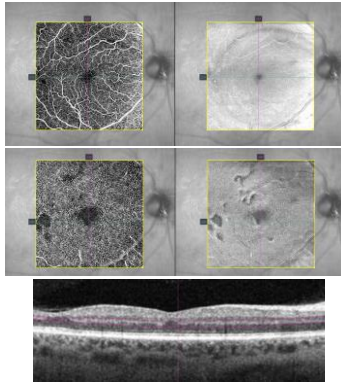
PAMM

OCT findings: Acute stage → hyper-reflective band at junction of the outer plexiform layer (OPL) and inner nuclear layer (INL)
Late stage → atrophy and thinning of the INL

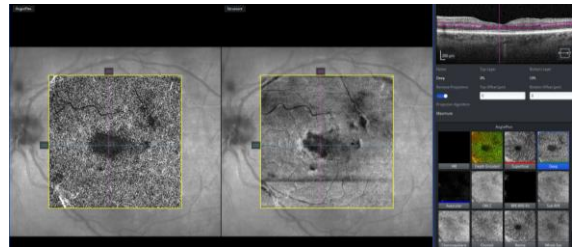


PAMM

- Late stage PAMM
- OCTA - shows areas of dropout, mainly evident when evaluating the deep capillary plexus slab
- En Face - localizes lesion to mid-retina

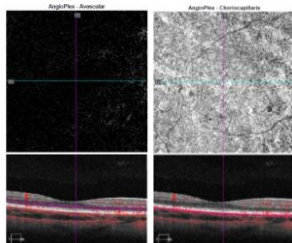


PAMM



Evaluating Outer Retinal Disorders

- Evaluate OCTA and B-scans
 - Avascular
 - Choriocapillaris



Age-related Macular Degeneration

- Devastating vision loss caused by geographic atrophy or CNVM
- CNVM consists of abnormal growth of blood vessels from the choroid
- OCTA can help detect presence of CNVM



CNVM 1

- Occult
- Located below RPE
 - May only show in choriocapillaris scan

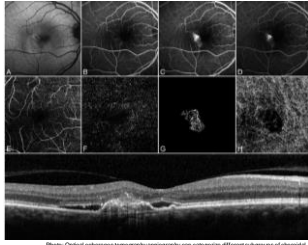
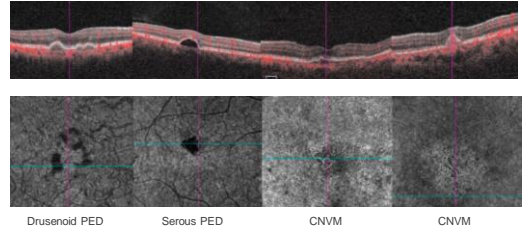


Photo: Optical coherence tomography angiography can categorize different subgroups of choroidal neovascularization secondary to age-related macular degeneration. Antigo, A, et al.

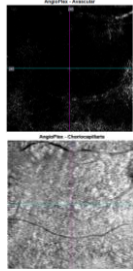
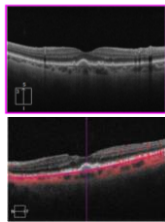
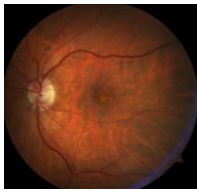
CNVM 1

- Use flow overlay to help determine if blood flow is present in a suspicious area



Drusenoid PED Serous PED CNVM CNVM

Drusenoid PED



CNVM 2

- Classic
- Located above the RPE

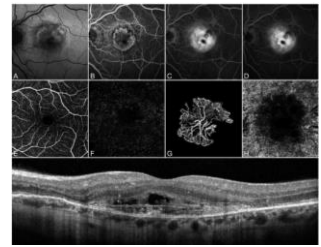


Photo: Optical coherence tomography angiography can categorize different subgroups of choroidal neovascularization secondary to age-related macular degeneration. Antigo, A, et al.

CNVM 3

1. Retinal Angiomatous Proliferation (RAP) Lesion
2. "Hot Spot" on ICGA
3. Develops within outer retina and progresses posteriorly into subretinal space

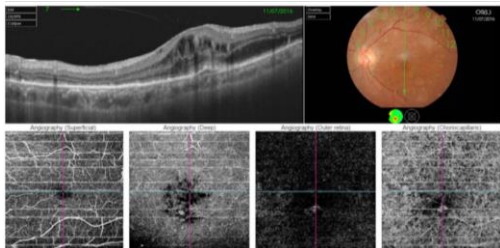


Photo: Diagnosis and Follow-Up of Nonexudative Choroidal Neovascularization With Multiple Optical Coherence Tomography Angiography Devices: A Case Report. Lina M, et al.

New Type: Nonexudative CNVM

- CNVM visible on OCTA but does not leak on dye angiography

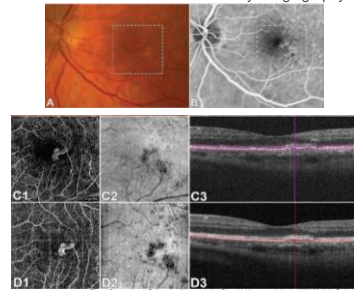
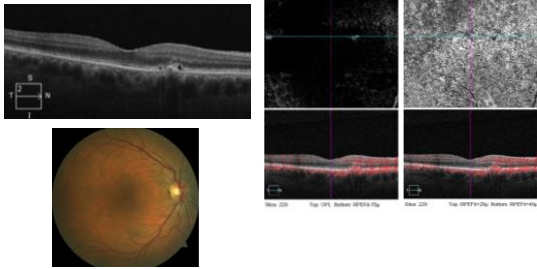
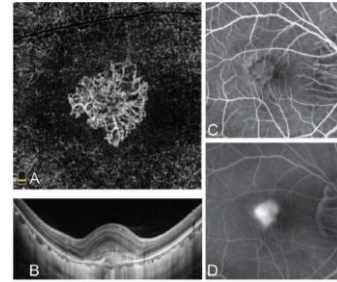


Photo: Diagnosis and Follow-Up of Nonexudative Choroidal Neovascularization With Multiple Optical Coherence Tomography Angiography Devices: A Case Report. Lina M, et al.

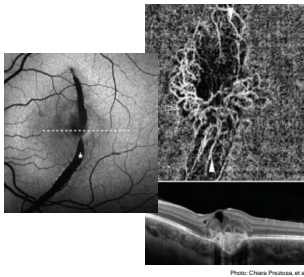
Nonexudative CNVM



Myopic CNVM



Choroidal Rupture CNVM



IVFA

What is fluorescein dye?

- Sodium fluorescein is an inert dye that is yellow-orange in color. 10% and 25% strengths
- Absorbs wavelengths in the blue range and fluoresces at 520-530 nm
- When injected intravenously, the dye leaks from all vessels in the body except those in the central nervous system including the retina (the retina is neural tissue and respects the blood-brain barrier)
- IVFA "discovered" by two Indiana University medical students

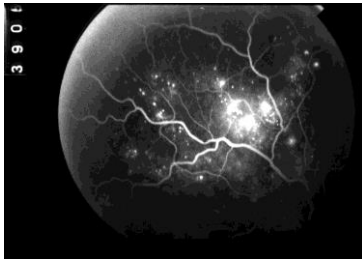
What's normal?

- The choriocapillaris has fenestrations which allow the dye to leak into the extravascular space, leading to "choroidal flush"
- The RPE serves as a barrier to prevent this dye from leaking forward and also limits its visibility
- Normal retinal vessels do not leak dye



What's not normal?

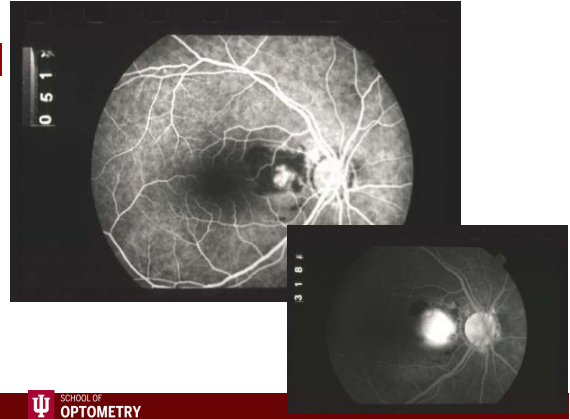
- Abnormalities can be broken down into two main categories: hypofluorescence and hyperfluorescence
- Hypofluorescence** is the result of either a **filling defect** or **blockage**
 - A filling defect is the result of capillary non-perfusion (diabetes, post vein occlusion) or a blockage such as an arterial embolus or sickle cell induced clot. Arteriosclerosis can also result in a lack of filling
 - Hypofluorescence due to blockage is the result of some entity blocking the background fluorescence of the choroidal flush or obscuring the retinal vasculature
 - Examples include pre-retinal, intra-retinal, and sub-retinal hemorrhages as well as exudates, pigment, and masses.



Fundus Autofluorescence (FAF)

What's not normal?

- Hyperfluorescence** is the result of either **loss of normal blockage** of the background flush or **leakage from abnormal vessels**
- Window defects, atrophy and chorioretinal scars can lead to loss of the RPE and hyperfluorescence due to lack of blocking
- Hyperfluorescence from leakage occurs with neovascularization, microaneurysms, edema, and compromised vessels
- IVFA is mostly useful today in situations when widefield evaluation of the peripheral retina is needed, and situations where activity (leakage vs. no leakage) is in question.



Fundus Autofluorescence: FAF

- Images ocular fluorophores
 - lipofuscin distribution in the RPE
- Lipofuscin autofluoresces in the 300nm-600nm wavelength range, which is very close to visible light (400nm-700nm), so visible light can excite an emission
- Gaining traction over about the last 15 years or so
- Recently becoming more integrated into clinical practice, as diagnostic and monitoring tool with applications in multiple disease states

Imaging Modalities

Confocal Scanning Laser Ophthalmoscope (cSLO)

- Example: Heidelberg Spectralis
- Uses a low energy laser and averages up to 30 scans
- Pros: high contrast, high resolution

Ultrawide field

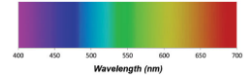
- Example: Optos
- SLO + ellipsoid mirror combines for 200° field of view

FAF Camera

- Example: Zeiss, Topcon, Canon...
- Uses a single, VERY bright flash (300 watt-seconds) yielding a single image
- Pros: quick, less motion artifact, less expensive SLO

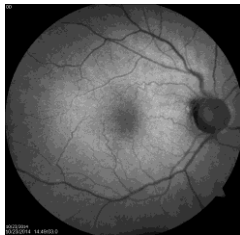
FAF Basics

- Images are entirely based upon the presence of lipofuscin in the RPE
 - In the eye, a byproduct of photoreceptor outer segment phagocytosis
 - Accumulates in the RPE with age and certain diseases
 - Also accumulates in other tissues and organs with age or disease (brain, liver, heart)
- Lipofuscin is excited with blue light, then emissions peak at green-yellow



FAF interpretation

- Sick or stressed RPE hyper fluoresces
- Dead, absent, or hypertrophic RPE hypo fluoresces
- The optic nerve head, blood vessels, and fovea are always hypo fluorescent (dark)
- Normal fundi are diffusely, mildly hyper fluorescent and grainy



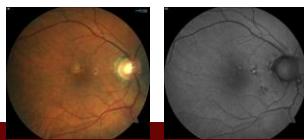
Conditions in which FAF is useful

- AMD
- ICSC (central serous retinopathy)
- Plaque toxicity
- Nevi / melanomas, choroidal lesions
- Macular / retinal dystrophies
- White dot syndromes
- MacTel
- ONH Drusen



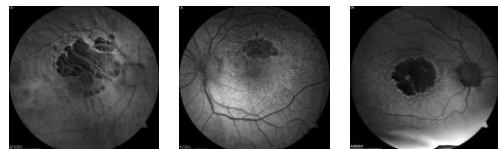
FAF: AMD

- Useful in detecting subretinal drusenoid deposits (SDDs) and monitoring geographic RPE loss
- Very common for FAF to reveal RPE loss / stress that is not as visible or even not visible at all to examination or color photography.
- Drusen can be normal, hyper or hypo fluorescent
 - Small drusen may not be visualized on FAF
 - Large, soft drusen tend to hyper fluoresce



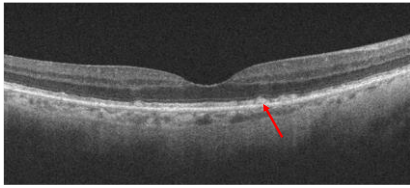
Geographic atrophy FAF

- Rate of progression of geographic atrophy is most dependent upon the FAF pattern at the junctional zone.
- Hyper fluorescence at the junctional zone is a bad sign
- Hypo fluorescence / normal fluorescence portends slow progression



Subretinal Drusenoid Deposits (SDD)

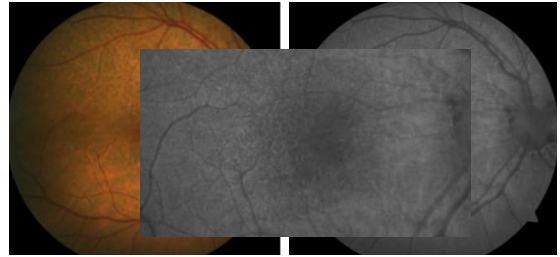
- Formerly known as reticular pseudodrusen
- Higher risk of progression to advanced AMD (wet and GA)
- Can not distinguish SDDs from drusen on color photos/funduscopy – best seen on OCT/FAF



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SDD FAF

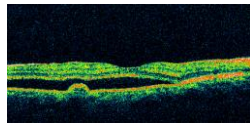


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Idiopathic Central Serous Chorioretinopathy

- FAF paints a completely different picture than funduscopy / color photography
- Recurrent and often multifocal - best appreciated using FAF
- Guttering/Troughing - best highlighted on FAF
 - Characteristic of chronic disease



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111

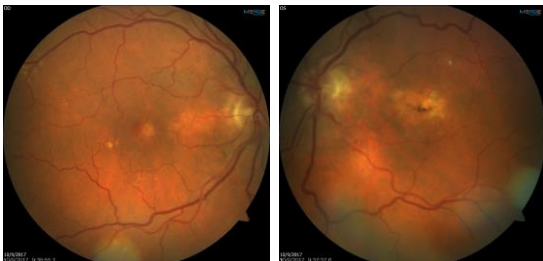
Acute ICSC



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112

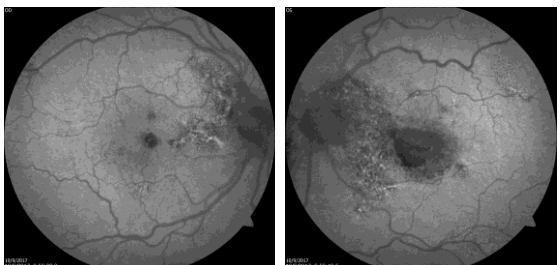
Chronic ICSC



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113

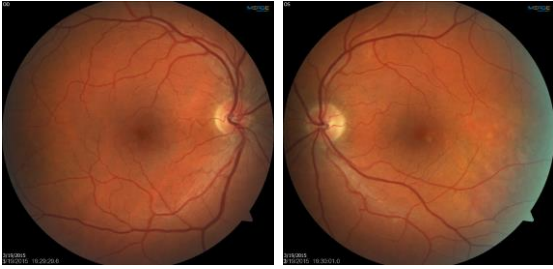
Chronic ICSC - FAF



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114

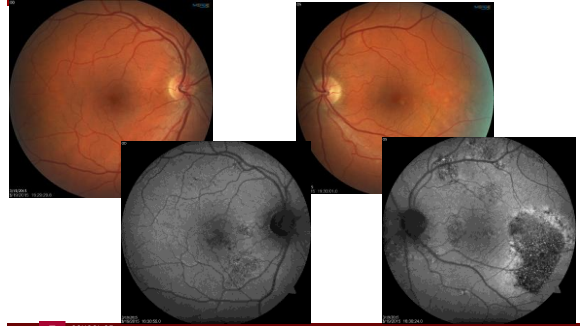
ICSC



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ICSC – FAF reveals multifocality



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116

ICSC “Troughing”



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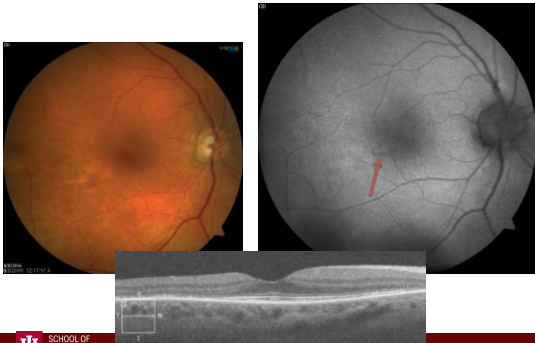
Troughing



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118

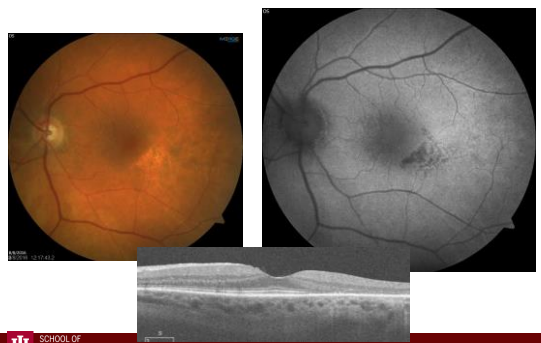
Plaquenil toxicity



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119

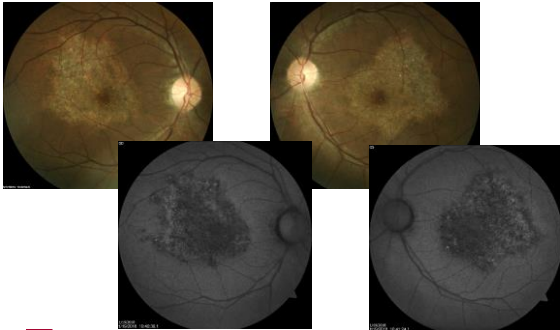
Plaquenil toxicity



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120

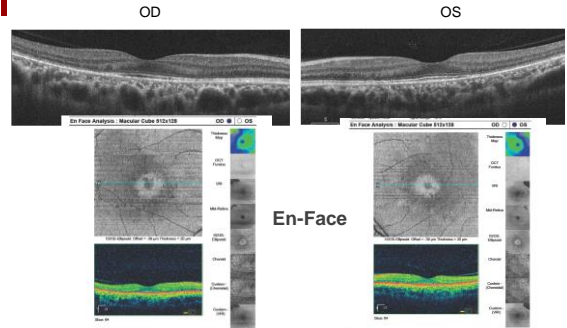
Chloroquine retinopathy



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Chloroquine OCT



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Risk factors for Choroidal Melanoma (updated in 2019)

Shields mnemonic: To Find Small Ocular Melanoma Using Helpful Hints Daily



To Find Small Ocular Melanoma Doing IMaging (TFSOM-DIM)

Thickness >2 mm (on ultrasound)

Fluid subretinal (on OCT)

Symptoms of vision loss (by VA testing)

Orange pigment (on FAF)

Melanoma hollow (on ultrasound)

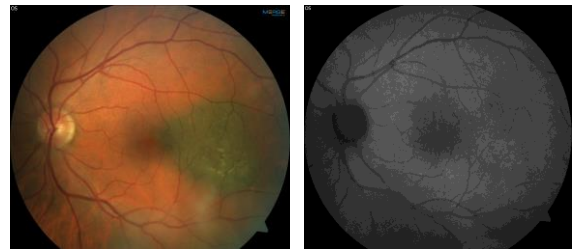
Diameter >5mm (by photography)

Risk factors	5 year risk
0	1.1%
1	11%
2	22%
3	34%
4	51%
5	55%

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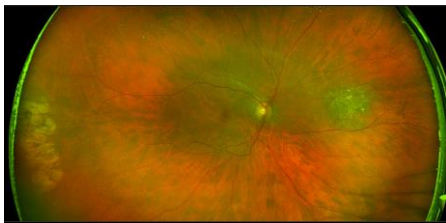
Choroidal nevus FAF



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Choroidal Nevus



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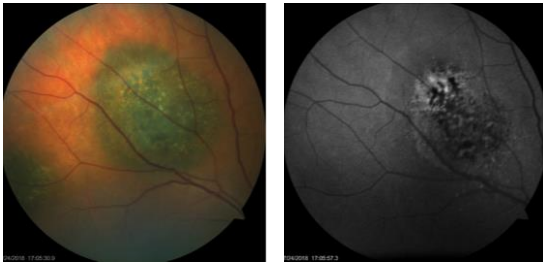
Choroidal Nevus FAF



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126

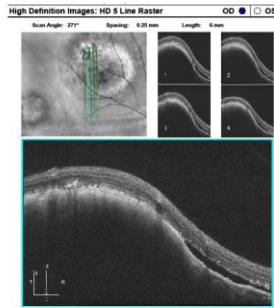
Choroidal Melanoma



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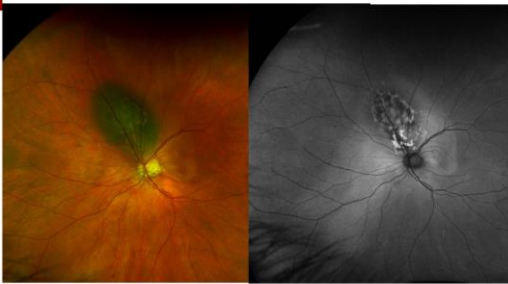
OCT – Shows subretinal fluid



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De Novo Choroidal Melanoma



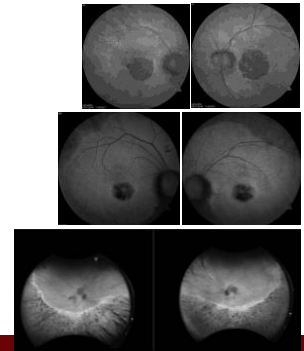
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Macular/Retinal Dystrophies

Utility of FAF:

- Aid in proper diagnosis
 - Earlier diagnosis
 - Examine family members at risk of diagnosis before clinically visible changes
- Track progression over time



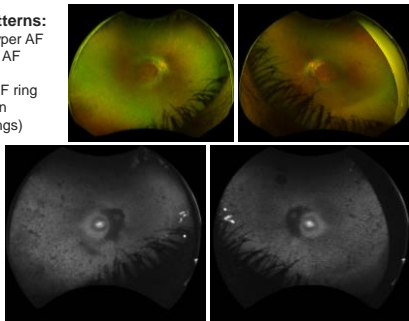
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Retinitis Pigmentosa

Macular FAF patterns:

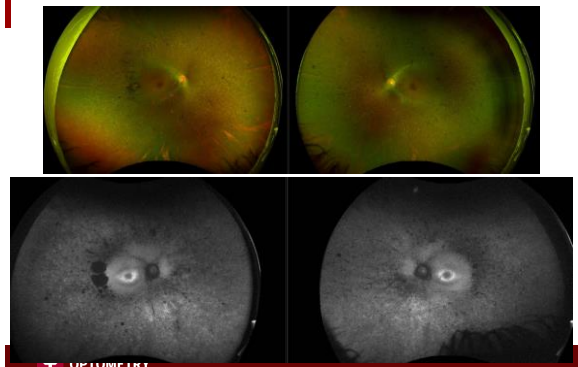
- central foveal hyper AF
- perifoveal hyper AF ring
- macular hyper AF ring
- bull's-eye pattern (double hyper rings)



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Retinitis pigmentosa: Mother



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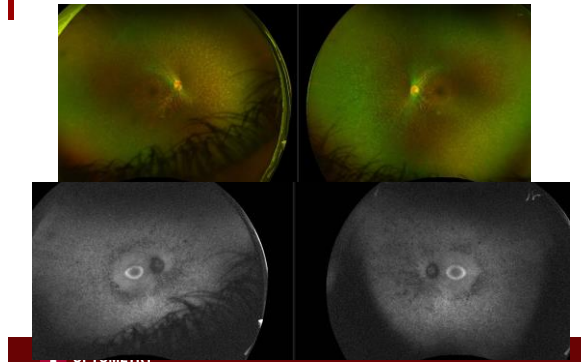
132

RP: Twin daughter #1



133

RP: Twin daughter # 2



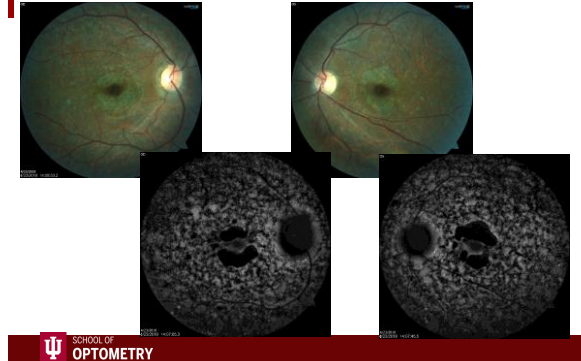
134

RP: sister / aunt



135

Stargardt disease

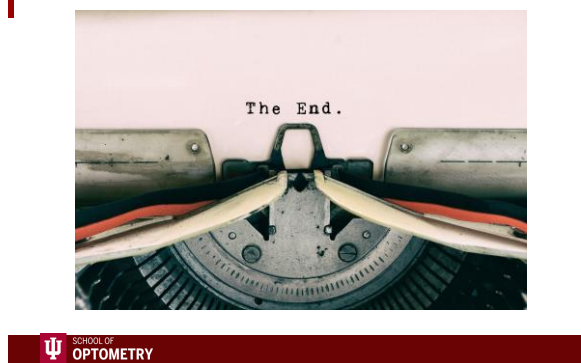


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Fundus Flavimaculatus



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138