

FAVORITE CASES GRAND ROUNDS

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FINANCIAL DISCLOSURES

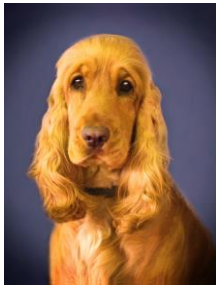
- None
- brsutton@indiana.edu



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CASE # 1

- Well I'll be dog gone!



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CASE # 1

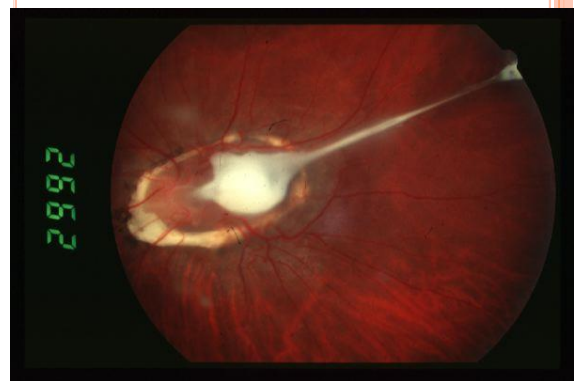
- 41 year old white male
- In for yearly exam, "always had poor vision in left eye"
- Diagnosed with coloboma OS many years ago
- No medical history
- BVA 20/15 OD, hand motion @ five feet OS

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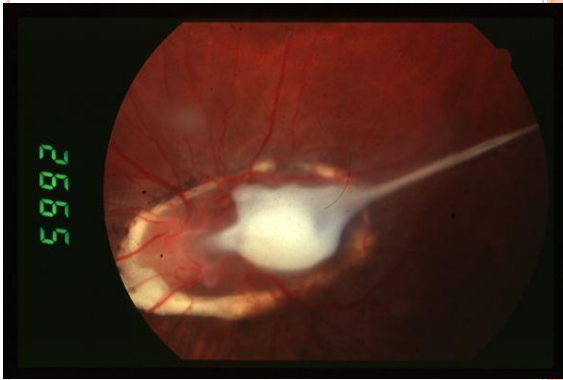
CASE # 1

- 45 prism diopter constant LXT
- Other entrance testing and slit lamp findings unremarkable
- Fundus as shown OS, unremarkable OD

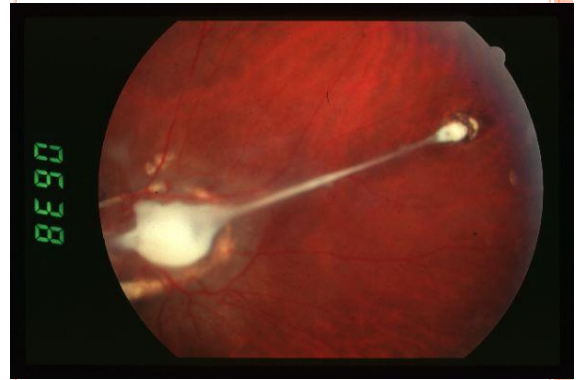
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TOXOCARA CANIS

- Nematode carried by dogs. Usually southeastern US. Major problem worldwide
- Ingested by humans as the result of eating tainted soil (dog feces) or occasionally undercooked meat
- Eggs can remain viable in humans for years then activate into mobile larvae. Entire life cycle can be carried out in dogs, larvae only in humans
- Larvae enter the eye via the blood stream, result in formation of a granuloma

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TOXOCARA CANIS

- Two forms: VLM and OLM
- Do not typically co-exist together
- VLM occurs between ages one and four, OLM later in childhood into adolescence
- VLM symptoms of fever, weight loss, vomiting, etc. Vague nature of symptoms often prevents accurate diagnosis

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TOXOCARA CANIS

- Typical ocular finding is a large granulomatous scar with fibrous bands radiating from the lesion and RPE hyperplasia
- Associated vitritis and even iritis are possible
- Treatment options include Albendazole 400mg BID in children (800 adults), Thiabendazole, cryotherapy, and photocoagulation.

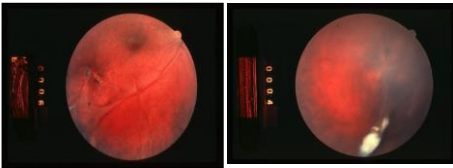
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TOXOCARA CANIS

- Steroids can be utilized to curb inflammation
- Success of treatment is very limited. Death of the organism leads to a greater inflammatory reaction so treatment is sometimes not indicated

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TOXOCARIASIS



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TOXOCARIASIS



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TRUE COLOBOMA



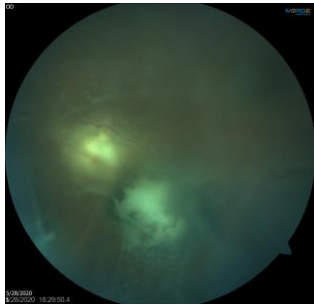
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TOXOPLASMOSIS



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ACTIVE TOXOPLASMOSIS



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HISTOPLASMOSIS: FLORIDA STRAIN!

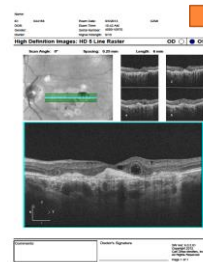


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WHAT AM I?



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CASE # 2

- We've got it in the fold!



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CASE # 2

- 87 year old white female
- Chief complaint " blurry vision in right eye"
- History of cataract extraction and yag capsulotomy OS
- HTN
- Entrance testing non-contributory

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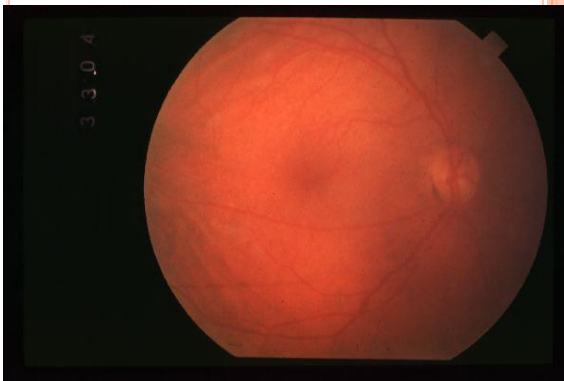
CASE # 2

- BVA 20/70 OD, 20/20 OS
- NS cataract OD, centered IOL OS with open capsule
- Fundi as shown

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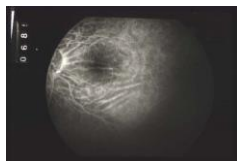
CHOROIDAL FOLDS

- Shrinkage or compression of the scleral tissue leading to folding of the attached choroid/Bruch's membrane/RPE complex
- Bilateral folds usually the result of age related scleral shrinkage in hyperopic females (as in this case). Benign
- Unilateral folds are more ominous and can be the result of orbital tumors, hypotony, CNVM's, or posterior scleritis

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CHOROIDAL FOLDS

- Alternating light and dark bands, show up clearly on IVFA as light and dark streaks



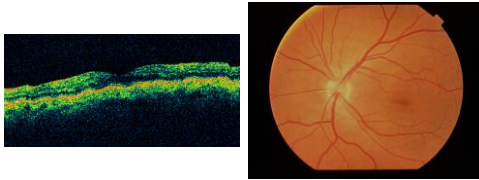
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CHOROIDAL VS. RETINAL FOLDS

- | | |
|---|---|
| <ul style="list-style-type: none"> ○ Choroidal... ○ Usually roughly horizontal ○ Usually posterior pole ○ Light and dark streaks ○ Visible on IVFA | <ul style="list-style-type: none"> ○ Retinal... ○ Often stellate alignment ○ Can be anywhere ○ Similar but finer ○ Visible on IVFA only if vascular traction |
|---|---|

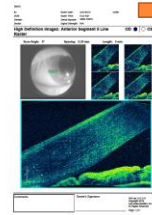
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CHOROIDAL FOLDS DUE TO HYPOTONY



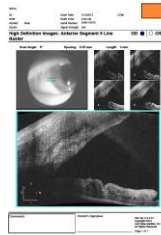
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WOUND LEAK WITH CHOROIDALS

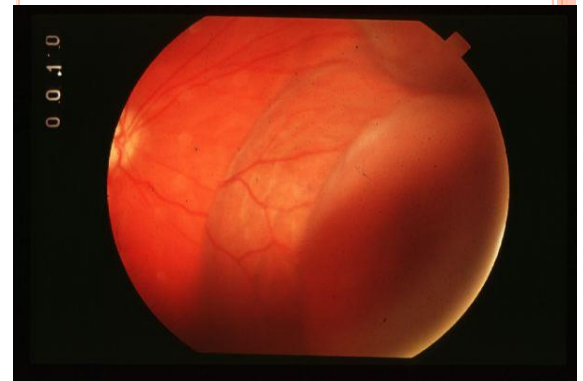


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WOUND LEAK POST REPAIR



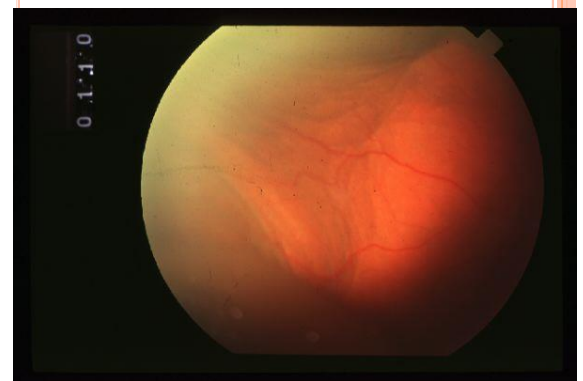
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CHOROIDAL FOLDS



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PATTON'S FOLDS



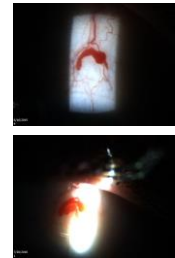
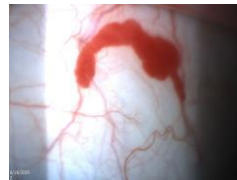
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WHAT AM I?



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HOW ABOUT ME?



June 2015, July 2015, July 2016

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CASE # 3

- Vasculitis? I don't think so.....



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CASE # 3

- 23 year old AA female presenting for low vision consultation
- BCVA of 20 / 40 -2 OD and 20/800 OS
- Large, constant LXT, significant VF constriction OS > OD
- Other entrance tests and slit lamp findings unremarkable. Fundus appearance.....

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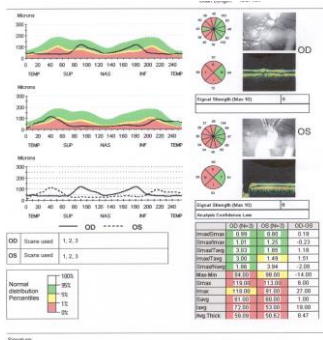
CASE # 3



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LEBER'S HEREDITARY OPTIC NEUROPATHY

- Hereditary mitochondrial disease process affecting the retinal ganglion cells
- Maternal inheritance pattern (mitochondrial DNA in embryo comes only from the egg)
- Genetic mitochondrial point mutations that have been fully identified / mapped. 95% of cases due to three genetic mutations: 11778,3469,14484
- Only 20-50% of male carriers and 10% of female carriers get optic atrophy and vision loss
- Environmental triggers include smoke, excessive alcohol, poor nutrition / vitamin deficiency, trauma
- Because of inheritance pattern, males can not pass to offspring

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LHON

- Males more commonly afflicted but females can be
- Typically strikes in early adulthood, but can strike later. 95% prior to age 50, most typically under age 30
- Late onset cases after age 50 have a 5:1 male prevalence
- Most commonly affects one eye followed by the fellow eye within several weeks (75%), but can be simultaneous (25%)
- Clinical trials ongoing with gene therapy.....early results promising

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GENE THERAPY TRIALS

- Gensight Biologics GS010
- Single intravitreal injection using an adenoviral vector to carry information to the mitochondria
- 15 patients in early trial
- 48 weeks after the injection.....
- < 2 years from disease onset, 30 letter average gain in VA
- > 2 years from disease onset, zero letter average gain in VA
- Phase 3 trials Rescue and Reverse: no better than sham at resulting in a 15 letter gain in vision: But.....may have had "cross-over" effect and helped the other eye

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LHON

Reported associations

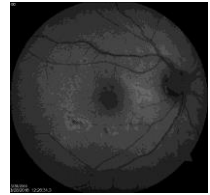
- Can get pseudopapilledema secondary to peripapillary NFL swelling
- Reports of vasculitis and pseudovasculitis
- Often don't get decreased pupillary responses; sparing of the melanopsin-expressing RGC's that mediate pupil response

Our patient.....



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WHAT AM I? OD DAY ONE



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OS DAY ONE



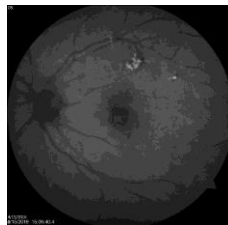
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3 WEEK FU ON ORAL STEROIDS OD



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3 WEEK FU OS ON ORAL STEROIDS



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CASE # 4



- "Looks like drops of water on a windshield"



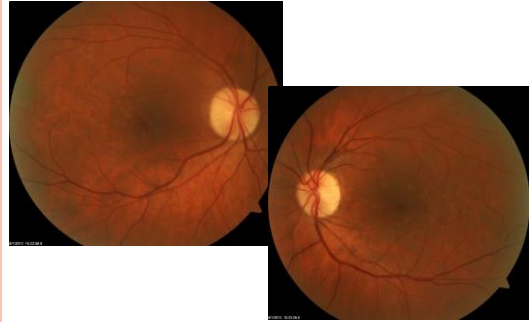
54

CASE # 4

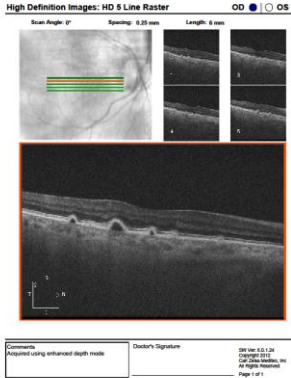
- 60 YO Caucasian female
- In for exam to check on cataracts
- Arthritis, Asthma, OSA
- BCVA 20/25- OD, 20/20 OS
- IOP 13,12
- All preliminary testing normal
- 1+ NS, trace PSC OU
- ONH Drusen OU
- Multiple, small PED's OU within the arcades

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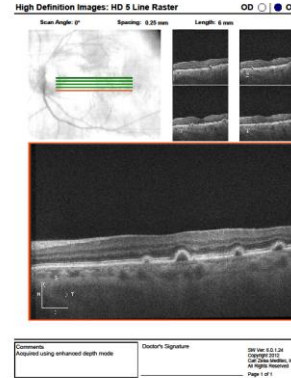
CASE # 4



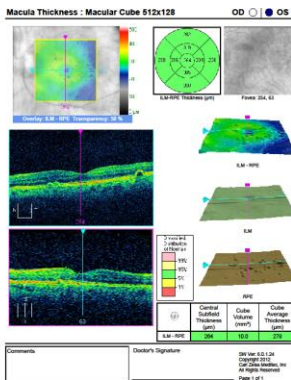
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MULTIPLE IDIOPATHIC PED SYNDROME

- Very rare, with few cases in the literature
- Multiple PED's with little or no neurosensory retinal detachment
- Usually females, often related to pregnancy
- One theory is a variant of ICSC involving only the RPE
- May be related to sleep apnea, which has been linked to ICSC
- What about Balversa?

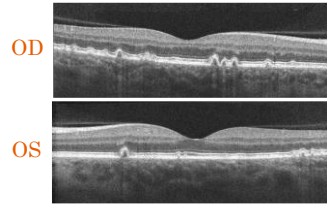
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Another pt. with Multiple Idiopathic PED Syndrome

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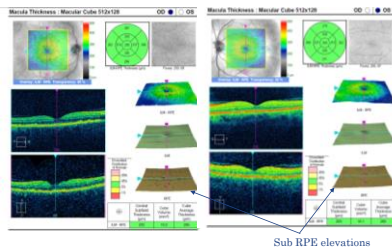
CIRRUS SD-OCT



Findings: multiple serous pigment epithelial detachments

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MACULAR CUBE



Sub RPE elevations

63

WHAT AM I?



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CASE # 5

- “We all have it doc....”



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CASE # 5

- 58 year old Caucasian male
- Complaining of flashes OD for four days. Has floaters, but longstanding with no increase
- HIV+, hyperlipidemia, asthma
- Atripla, Levoquin, Pilocarpine
- Told by a retinal specialist 17 years prior that he has a progressive retinal disease but would not go “completely blind”
- BCVA of 20 / 20 in each eye
- IOP 18 OD, 19 OS
- Entrance testing unremarkable
- Anterior segment unremarkable OU
- Posterior segment reveals significant drusenoid changes OU with pigment mottling OU

66

CASE # 5

- Posterior segment also reveals a fresh PVD OD with no holes, tears, or breaks
- Posterior segment appearance.....

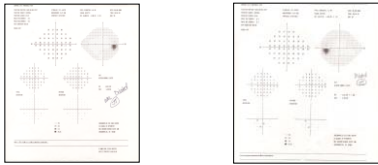
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CASE # 5



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CASE # 5



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CASE # 5

- So what have we here.....?
- Doyme's Honeycomb Dystrophy!
- AKA: Mallattia-Leventinese Dystrophy

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CASE # 5

- Doyme's Honeycomb Dystrophy
- Described by Doyme in England in 1898
- Malattia-Leventinese Dystrophy described by Alfred Vogt in Switzerland in 1925
- Now believed to be phenotypic variants of the same condition
- Both caused by genetic mutation in EFEMP1 gene

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CASE # 5

- The affected gene encodes a protein that is expressed in the retina and the RPE
- Leads to drusen formation early in life
- Located all throughout the posterior pole, including nasal to the disc
- The drusen coalesce over time leading to radial (MLD), honeycomb (DHD), or mosaic patterns (MLD and DHD)
- Symptoms are rare until early middle age
- Macula is affected with drusen, so can get vision loss and SRNVM

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CASE # 5

- Because the condition is genetic, the role of nutritional supplements is unclear
- Manage with regular follow-up, amsler at home, possibly supplements



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CASE # 5

- Differential diagnoses:
 - 1) AMD : affects macular area only. More likely to have RPE loss, more likely to have an SRNVM
 - 2) Familial or basil laminar drusen: cluster in groups throughout the posterior pole, especially in arcades, but do not coalesce. No effect on vision.

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CASE # 5

- This patient monitored over time with regular follow-up and amsler
- Remains symptom free with good vision



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ANOTHER DOYNE'S EXAMPLE



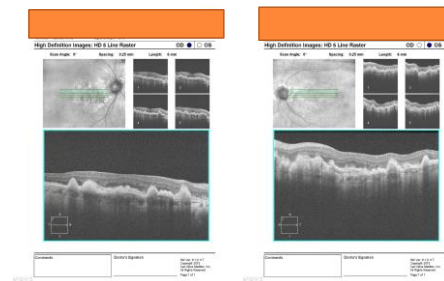
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DOYNE'S 5 YEARS LATER



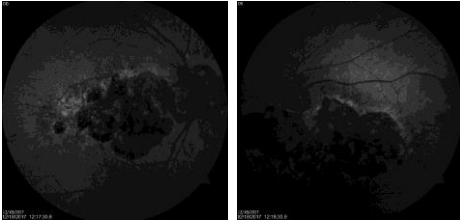
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DOYNE'S OCT



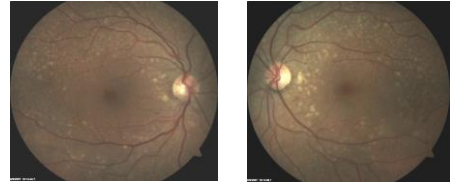
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DOYNE'S FAF



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CASE # 5
ANOTHER DOYNE'S EXAMPLE



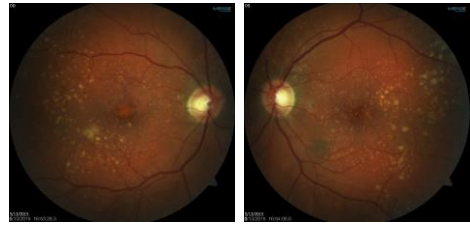
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CASE # 5
ANOTHER DOYNE'S EXAMPLE



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CASE # 5: ANOTHER DOYNE'S



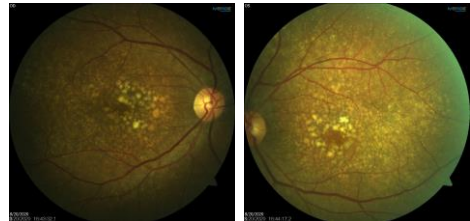
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CASE # 5: AND ONE MORE.....



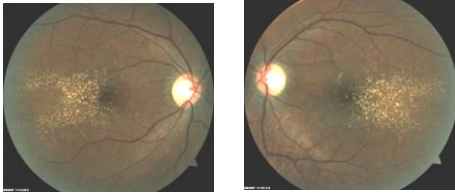
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CASE # 5: YET ONE MORE DOYNE'S.....



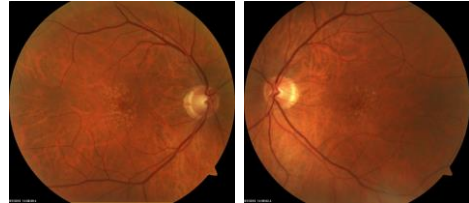
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CASE # 5
FAMILIAL / BASIL LAMINAR DRUSEN EXAMPLE



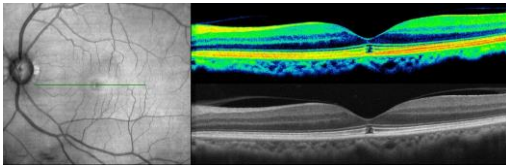
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CASE # 5
AMD EXAMPLE



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WHAT AM I? SCANS COURTESY OF DR.
JEROME SHERMAN



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CASE # 6

- “Are you sure this picture is not upside down?”



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CASE # 6

- 43 year old AA male complaining of poor vision, pain, itching and watering OD
- History of RD OD due to trauma and subsequent repair 10 years prior
- Taking Naphcon-A and Hydrocodone
- BCVA of LP OD, 20 / 30 + OS
- IOP 50 OD, 15 OS
- EOM's normal
- VF normal OS
- Pupil unreactive OD, but “reverse APD” OS
- White appearance to upper half of iris with the naked eye

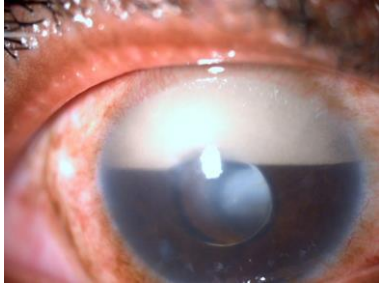
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CASE # 6

- Anterior segment evaluation.....
- Eyelid edema OD
- Aphakic OD
- Solid, clear bubble in central AC OD with top half of chamber filled with a white substance
- 2+ Conj. injection OU
- “reverse pseudohypopyon”
- Anterior segment OS unremarkable
- Posterior segment OS unremarkable. No view of fundus OD
- Anterior segment appearance.....

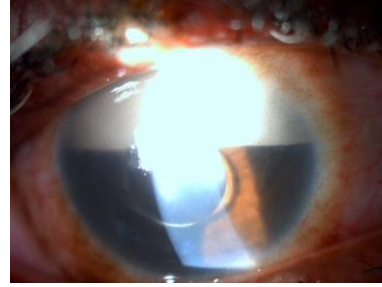
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CASE # 6



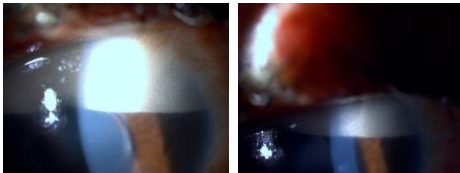
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CASE # 6



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CASE # 6: LOOKS LIKE STYROFOAM!



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CASE # 6

- o So what have we here.....?
- o Silicone oil emulsification!

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CASE # 6

- o Silicone oil tamponade is routinely used in complicated retinal detachment repair
- o It must be removed later to avoid potential complications
- o One complication is emulsification



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CASE # 6

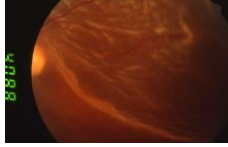
- o Emulsification basically means turning to soap
- o Silicone oil turns in to smaller droplets of soap like material when surface tension decreases significantly
- o This occurs when the oil contacts various biological products



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CASE # 6

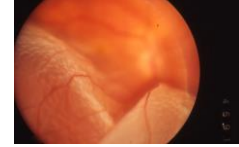
- Proteins, lipids, and phospholipids
- Particularly HDL
- Occurs up to 50% of the time if oil is not removed
- Happens in the vitreal cavity, then droplets travel to the AC, especially in an aphakic patient



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CASE # 6

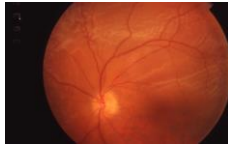
- “inverse pseudohypopyon”
- Leads to corneal endothelial toxicity and edema, band keratopathy, and increased IOP / glaucoma
- In a seeing eye, prompt removal is indicated



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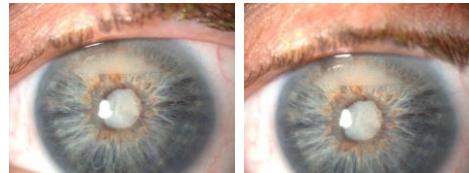
CASE # 6

- This patient was given cycloplegia for ciliary spasm
- Also offered pressure lowering agents for comfort, but deferred
- Sent for consideration of surgical removal of emulsified oil
- Lost to follow up



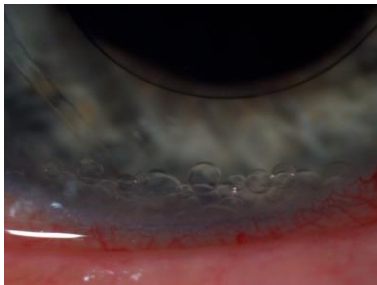
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ANOTHER EXAMPLE OF EMULSIFICATION



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MORE AC OIL WITH AC IOL

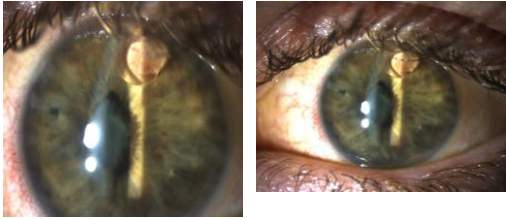


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SILICONE OIL BUBBLE



Can also get silicone oil bubbles after intravitreal injections. How???

103

WHAT AM I ?



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CASE # 7 "HIDING IN PLAIN SIGHT"

- o 49 YO Caucasian male
Presents for an exam with a complaint of broken safety glasses. No other issues
- o History of follicular lymphoma 3 years prior, treated with chemotherapy. In complete remission
- o No current medical conditions
- o No medications or OTC meds/supplements
- o BVCA 20/20 OD, 20/20 OS
- o Low CHA in both eyes
- o All preliminary testing normal, full confrontation VF
- o Amsler grid normal OU
- o IOP 12 mmHg OD, 12 mmHg OS
- o Anterior segment unremarkable OU
- o Posterior pole as shown, periphery attached and free of pathology OU

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POSTERIOR POLE OU

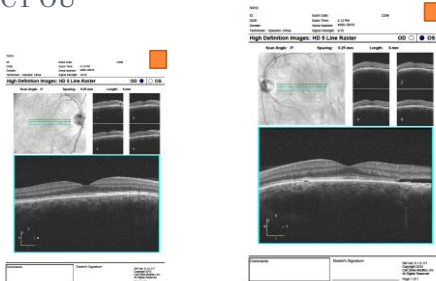


Very mild pigment mottling, inf. macula

Pigment mottling inf. & temp. to macula, flat

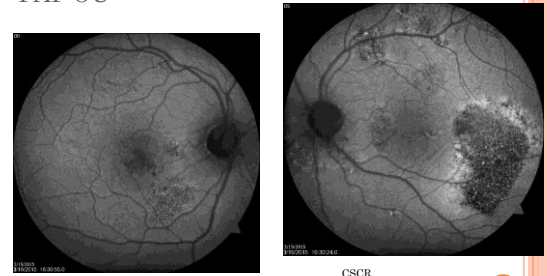
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OCT OU



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FAF OU



CSCR

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FUNDUS AUTOFLUORESCENCE (FAF)

- 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 autofluoresces in the 300nm-600nm wavelength range, which is very close to visible light (400nm-700nm), so visible light can excite an emission
- Valuable diagnostic and monitoring tool in an ever increasing list of ocular conditions
- Can show damage well before it is visible to examination or in regular photos

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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



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CENTRAL SEROUS CHORIORETINOPATHY

- CSCR: related to steroids / cortisol, either by taking steroids or endogenously elevating steroids through stress
- Type A personalities often if stress related
- Also linked to sleep apnea and testosterone injections / supplementation. Some reports of link to excessive energy drink consumption and link to high doses of ADHD medications
- Focal RPE damage / PED with secondary neurosensory retinal detachment
- Metamorphopsia, decreased vision
- Recurrent and often multifocal
- Abnormally thick choroid: One of the "Pachychoroid" diseases
- Observation often at first
- Topical NSAIDs ?, PDT laser, spironolactone or similar agent, possibly Diamox

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PACHYCHOROID DISEASES

- All share attenuated choriocapillaris, dilated choroidal veins, thickened choroid (definitively thick at 390 microns or greater), RPE dysfunction, and potential for CNV
- Pachychoroid Pigment Epitheliopathy
- CSCR
- Pachychoroid Neovascularopathy
- Polypoidal Choroidal Vasculopathy



Image courtesy of Dr. Sherrol Reynolds

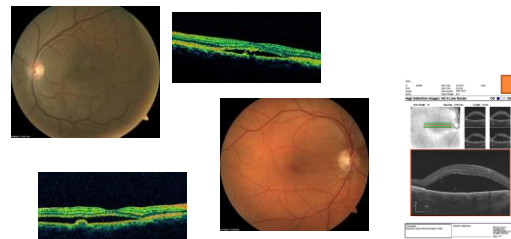
112

CSCR

- Fundus Autofluorescence paints a completely different picture than fundoscopy / color photography
- RPE damage and death, "troughing"
- Much greater multifocality

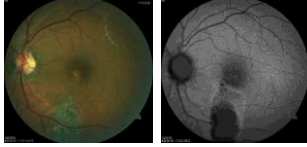
113

CSCR OCT



114

CSCR FAF "TROUGHING"



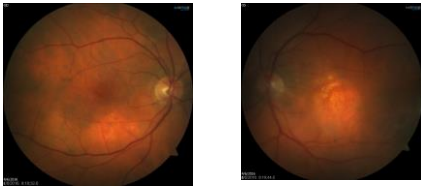
115

CSCR FAF



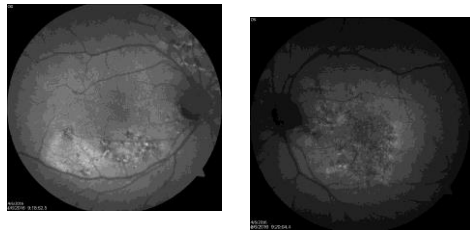
116

WET AMD?? HISTORY OF MULTIPLE ANTI-VEGF SHOTS. IN FOR A SECOND OPINION



117

NOPE.....CSCR



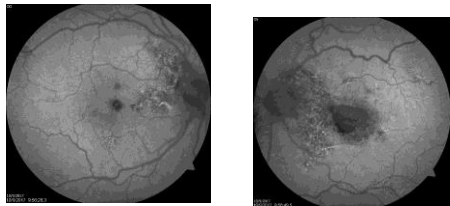
118

CSCR



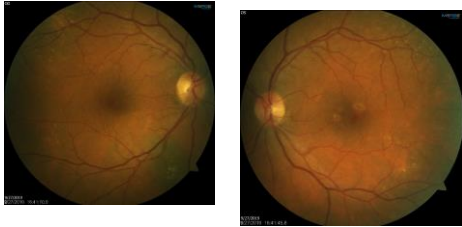
119

CSCR FAF



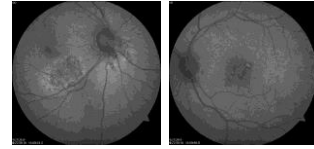
120

CSCR



121

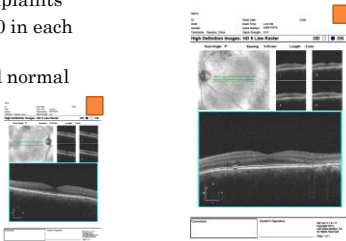
CSCR FAF



122

PATIENT FROM THE CASE 6 MONTHS LATER

- Still no complaints
- BCVA 20/20 in each eye
- Amsler grid normal OU



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THE END!

- Any Questions?



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