

Diagnosing and Managing Ocular Emergencies and Urgencies

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Disclosures:

- Maculogix: advisory board
- Sun Pharmaceuticals: speakers bureau,
- Avellino: advisory board,
- Dompe: advisory board,
- RVL Pharmaceuticals: advisory board

Expected Learning Outcomes

- Needs Assessment:
 - To enable optometrists to provide care for patients who present with an ocular emergency
- At the end of the presentation, attendees should be able to:
 - triage a patient as a potential emergency utilizing specific case history questions and understanding of what constitutes an emergency.
 - recognize the signs and symptoms of a patient experiencing an acute onset red eye (e.g. viral conjunctivitis), diagnostic criteria and be able to prescribe appropriate treatment and management.
 - the signs and symptoms of a patient experiencing a sudden decrease in vision from a retinal disease (e.g. central serous retinopathy), diagnostic criteria and appropriate treatment and management.
 - the signs and symptoms of a patient experiencing an acute changes in vision secondary to a retinal tumor, diagnostic criteria and be able to prescribe appropriate treatment and management

Eye Care and the Emergency Department

- Non-injury related ocular ER visits comprised 51% of ocular-related visits⁷
- Only 3% of ocular-related ER visits required hospitalization⁷
- **75% of the time**, there was a **clinically significant change in the diagnosis** when care was first delivered at the ED or PCP and then followed up by a visit to an eye care specialist

Distribution of clinical conditions associated with ocular related non-urgent ER visits

Condition	Percent of Non-Urgent Visits (%)
Conjunctivitis, unspecified	46.7
Blepharoconjunctivitis	6.3
Severe Allergic Conjunctivitis	6.2
Atopic Conjunctivitis	4.7
Hemorrhagic Conjunctivitis	4.1
Hordeolum	3
Blepharitis, unspecified	2.7
Disorders of the Optic Nerve	1.8
Myrdiasis	1.5
Chalazion	1.5

7. Erin A.Nash and Curtis E. Margo, "Patterns of Emergency Department Visits for Disorders of the Eye and Ocular Adnexa," Archives of Ophthalmology, volume 116, September 1998, pp. 1222 - 1226.
8. Hau S, Ioannidis A, Masaoutis P, Verma S. Patterns of ophthalmological complaints presenting to a dedicated ophthalmic Accident & Emergency department: inappropriate use and patients' perspective. Emerg Med J. 2008 Nov;25(11):740-4.

What Classifies an Emergency?

- Any condition in which the patient has the potential for:
 - vision loss,
 - currently experiencing vision loss,
 - permanent structural damage,
 - pain or discomfort,
 - or is an “emergency” for the patient.
- It is important to be able to triage a walk-in patient and, more importantly, a call-in patient.

What questions to ask?

Onset	suddenly noticed or sudden onset?
Visual Loss	any loss of vision? loss vs. blurry vision one eye or both part of visual field or all transient vs. permanent
Pain	is there pain? constant? scale (1-10)
Redness	is there any redness? location?
Associated Factors	contact lens wear? trauma? discharge? photophobia? medical history (eg. DM)

Visual Loss

- Visual loss varies greatly in meaning from patient to patient
 - ranging from blur to complete blindness and may affect one or both eyes
- Components include:
 - acuity,
 - visual field,
 - color and brightness may be affected jointly or separately
- Detailed history and extent of vision loss crucial

Profound Loss of Vision

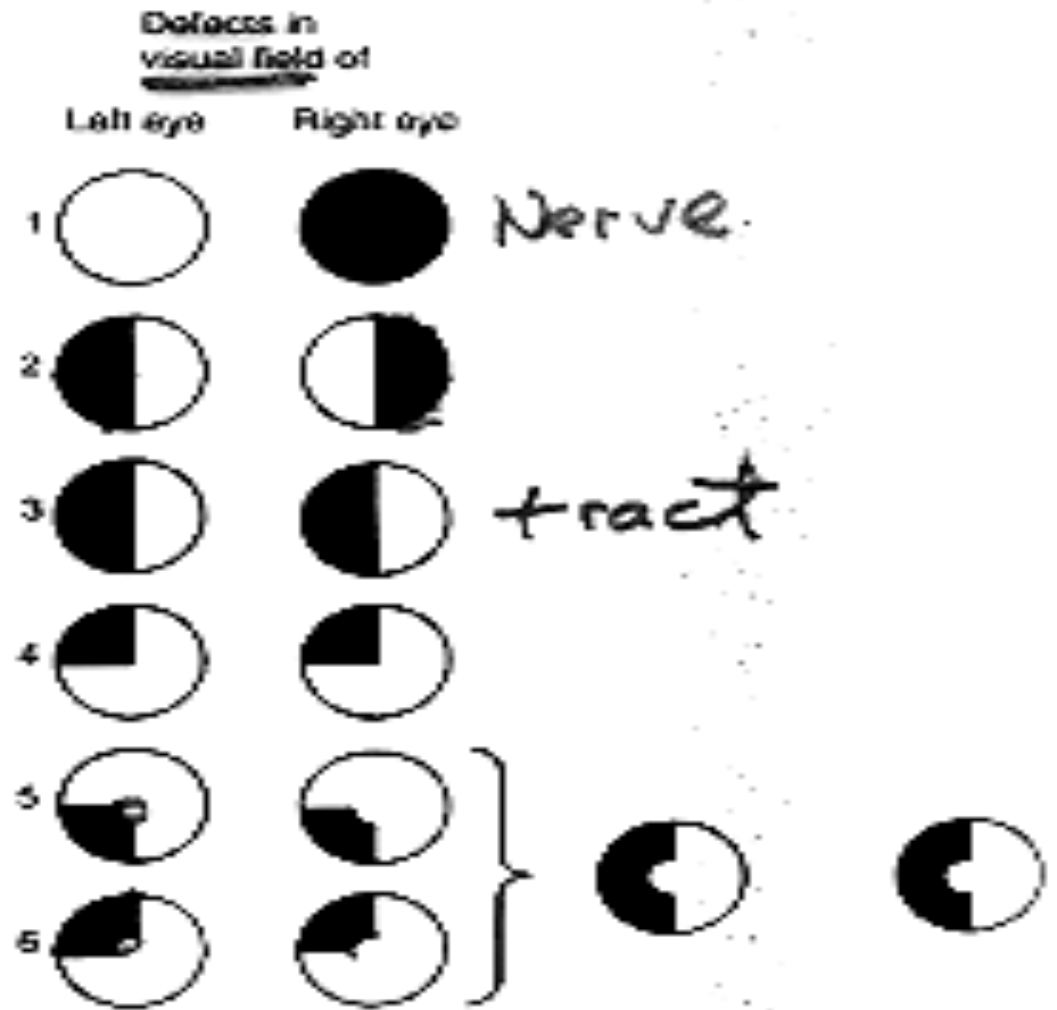
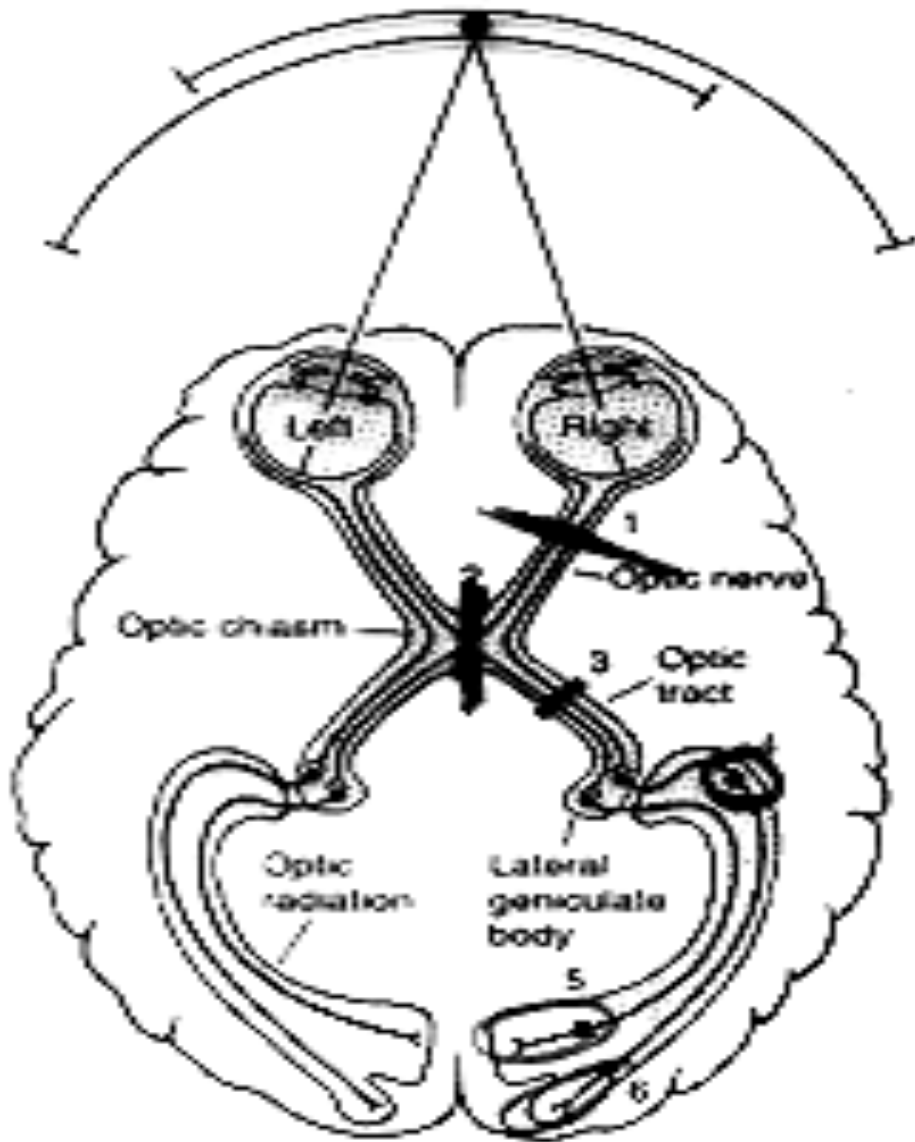
- Referring to a complete or greatly diminished vision affecting the whole field
- Common causes of severe vision loss:

Vascular	central retinal vein occlusion, central retinal artery occlusion, vitreous heme
Inflammatory	optic neuritis
Infiltrative	optic neuropathy
Mechanical	retinal detachment

Monocular vs. Binocular

- Ocular or optic nerve pathology causes monocular vision loss
- lesion at or posterior to chiasm causes binocular vision loss
 - VF defects become more congruous the further back in the visual pathway
 - Homonymous VF defects noted posterior to chiasm
- Difference between mono vs. bino usually straightforward, keeping the following in mind:
 - Patients occasionally mistake homonymous hemianopsia (similar loss of visual field in both eyes) for a monocular loss

Visual Defects



Monocular

- Differentiate between eyes that have lost all useful vision and those that have blurred vision
- Blurring of vision is not localized and may be caused by pathology anywhere from cornea to optic nerve
- Need to get anatomical diagnosis first before considering the cause

General Appearance

- Level of consciousness
 - When introducing yourself be aware of the patient's gross level of consciousness?
 - Is the patient awake, alert and responsive?
- Personal Hygiene and Dress
 - Is it appropriate for the environment, temperature, age and social status of the patient?
 - Is the patient malodorous or disheveled?

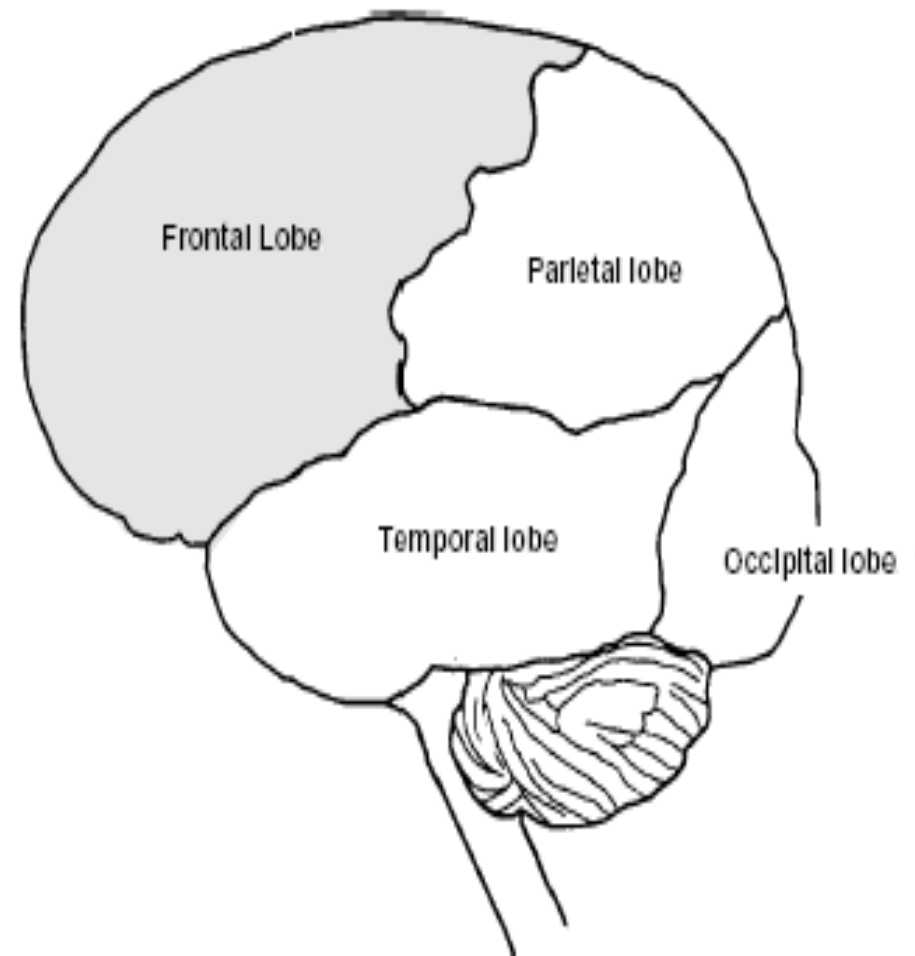


General Appearance

- Posture and Motor control
 - What posture does patient assume while sitting in the exam chair
 - Are there any signs of involuntary motor activity such as tremors
 - E.g. damage to the cerebellum may produce a tremor that usually worsens with movement of the affected limb

Neurological Screening: Cerebrum

- Frontal lobe
 - Emotions, drive, affect, self-awareness, and responses related to emotional states
 - Motor cortex associated with voluntary skeletal movement and speech formation (Broca)



Right vs Left Brain Injury

- So what happens if one side of the brain is injured?
 - People who have an injury to the right side of the brain "don't put things together" and fail to process important information.
 - As a result, they often develop a "denial syndrome" and say "there's nothing wrong with me."

Right vs Left Brain Injury

- The left side of the brain deals more with language and helps to analyze information given to the brain.
 - If you injure the left side of the brain, you're aware that things aren't working (the right hemisphere is doing its job) but are unable to solve complex problems or do a complex activity.
 - People with left hemisphere injuries tend to be more depressed, have more organizational problems, and have problems using language.

30 YR WM

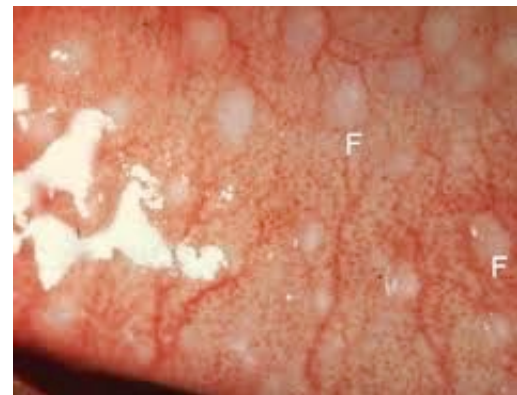
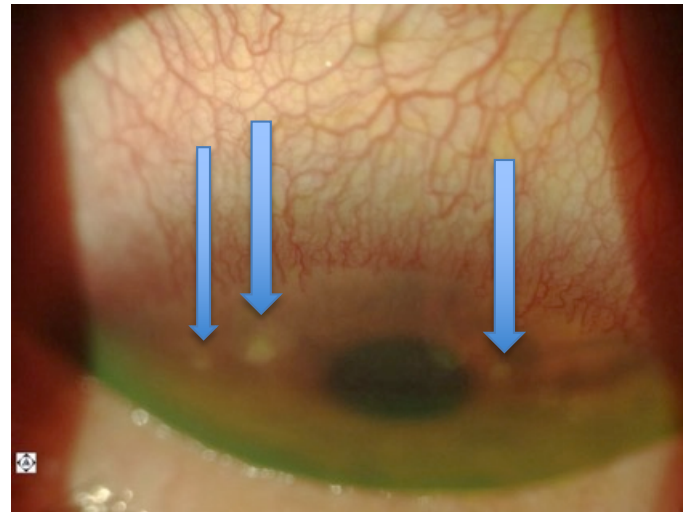
- Patient calls from his PCP office asking if we can see him today because he has had red/painful eyes for over a week and has not resolved
- Medical history:
 - Past week has been experiencing painful urination and discharge
 - New sexual partner apprx 10 days ago, who also had developed a red eye
 - Chlamydia and gonorrhea testing were negative
 - Has tested positive for HSV2 but no current flare up

30 YO WM

- Medications:
 - In the past week patient:
 - 2 courses of azithromycin (1 gram each)
 - Injection of rocephin
 - Injection of penicillin G
 - Currently taking doxycycline 100 mg bid
 - Valtrex 1 gram 3 times per day for 7 days (d/c 1 day ago)
 - Was on Vigamox qid for 7 days (d/c 1 day ago)
- VA: 6/7.5 (20/25) OD, OS
- Entrance skills unremarkable though some pain on eye movement

30 YO WM

- SLE:
 - 2+ injection conjunctival both eyes
 - 1-2+ lid edema
 - Mixed papillary and follicular response
 - 1-2+ diffuse SPK (no staining noted above infiltrates)
 - No cells or flare noted



30 YO WM

- AdenoPlus:
 - Performed on the right eye (patient felt that was the worst eye)
 - Negative

30 YO WM

- Started patient on the miracle drop
 - Tobradex 4 times per day and scheduled patient to come back the next day
- 1 day f/u
 - Patient was feeling better
 - Less redness and much reduced photophobia and discomfort
 - No improvement on painful urination or discharge and is now seeing blood in his urine
 - Continue tobradex 4 times per day and RTC in 4 days for f/u with dilation and told to contact PCP to update on the blood in the urine

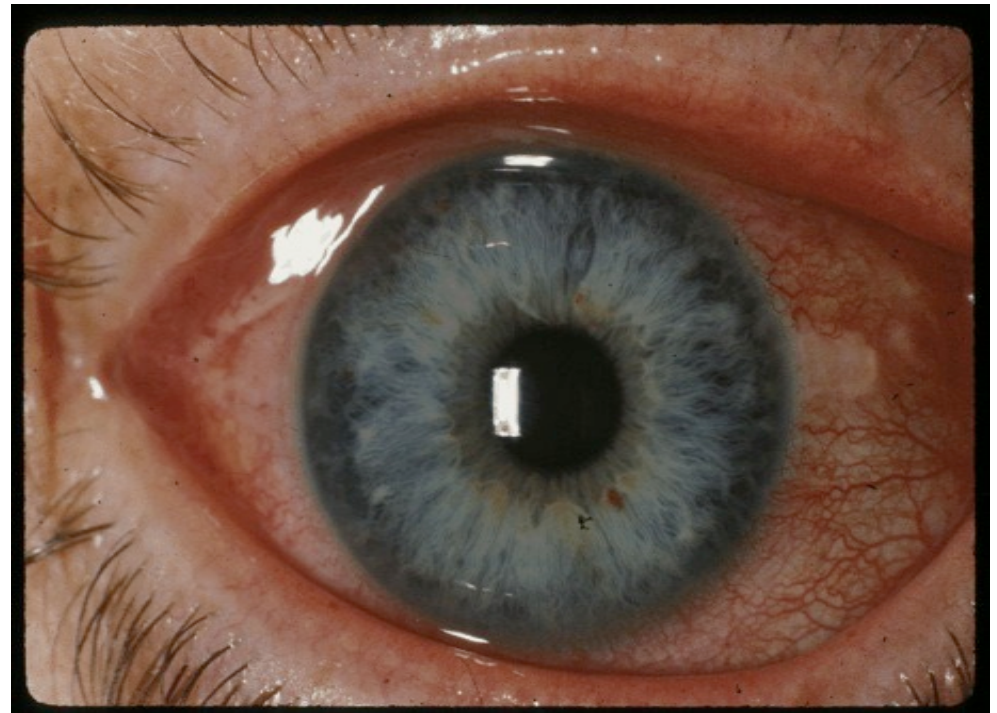
30 YO WM

- 4 day f/u:
 - Patient says his eyes are doing great and that all of his urogenital problems abruptly stopped on Saturday
 - Discussion with PCP: Kidney stone
 - What was going on with the eye?
 - Viral conjunctivitis likely EKC

What did we learn from this?

Viral Conjunctivitis

- Most common infectious keratitis presenting on emergent basis
- 62% caused by adenovirus
- Two major types:
 - Pharyngoconjunctival fever (PCF)
 - Epidemic keratoconjunctivitis (EKC)



Viral Conjunctivitis



- PCF: history of recent/current upper respiratory infection
 - classic triad of fever, pharyngitis, and acute follicular conjunctivitis.
 - occurs more commonly in children, is caused by serotypes 3 and 7, and is spread by respiratory secretions.
 - tearing and foreign body sensation that is initially unilateral.

Viral Conjunctivitis



- PCF:
 - corneal involvement is not a key feature, there is occasionally a punctate keratitis;
 - SEIs are rare.
 - self-limiting condition that varies in severity and may last from 4 days to 2 weeks
 - Treatment if symptomatic though topical steroids are rarely needed.

Viral Conjunctivitis: EKC (Epidemic Keratoconjunctivitis)

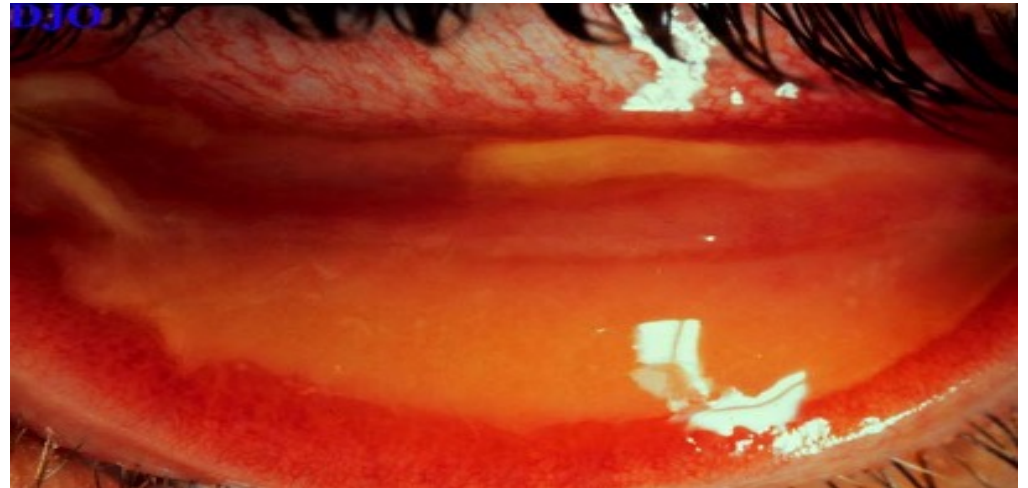
- EKC initially manifests as a flu-like syndrome consisting of fever, malaise, and myalgias followed by the appearance of ocular signs and symptoms, including a red eye, eyelid edema, excessive tearing, irritation, foreign body sensation, and photophobia.
- EKC frequently begins as a unilateral condition but, in 70% of cases, will become bilateral within the first week of symptoms as a result of hand-to-eye transmission
 - Adenovirus 8 common variant leading to “rule of 8’ s”
 - First 8 days red eye with fine SPK
 - Next 8 days deeper focal epithelial lesions
 - Following 8 potential development of infiltrates
 - Resolution

<https://www.aao.org/eyenet/article/epidemic-keratoconjunctivitis-prevention-strategie>

Viral Conjunctivitis: Signs and Symptoms

- Gritty sensation
- Watery discharge
- Sticky in mornings
- *Follicular response*
- Chemosis
- Injection
- SPK
- Infiltrates possible
- Positive lymph nodes

- *Pseudomembranes in severe cases*
- Subconjunctival hemes



Management

- Considering the use of anti-inflammatory treatment to relieve patient symptoms and improve comfort??
 - E.g. Lotemax^R QID OU
- EKC patients are typically very uncomfortable and would benefit from anti-inflammatory treatment
 - especially if infiltrates or pseudomembrane present
- studies have shown that steroids are effective in reducing inflammation during the acute phase of EKC and decreasing the likelihood of development of corneal subepithelial infiltrates.
- However, the studies also showed that their use increased viral replication and titers and prolonged the mean duration of viral shedding
- routine corticosteroid use is generally not indicated for EKC
 - when managing a severe EKC inflammation, you should carefully weigh the risks and benefits of steroids

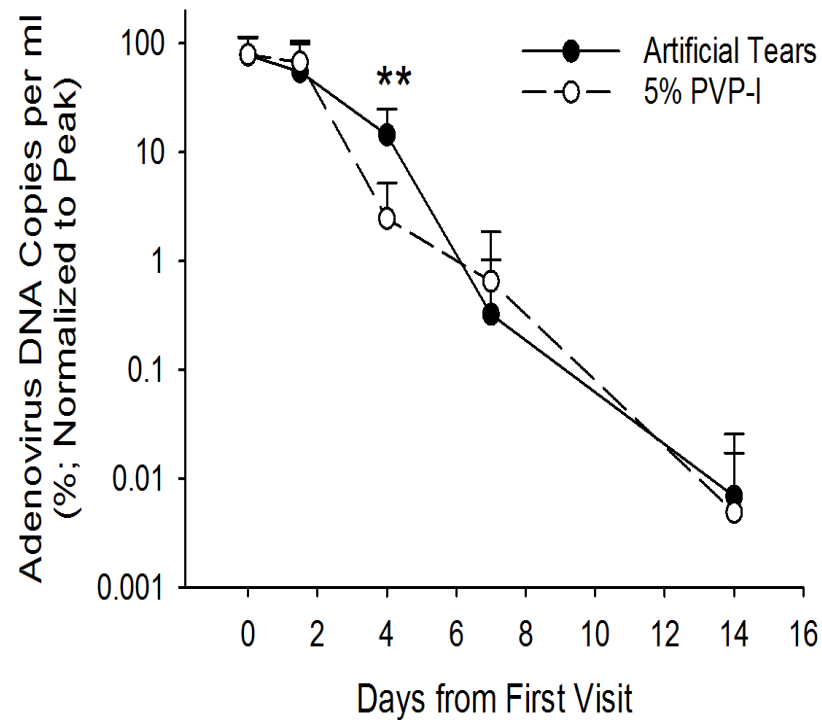
<https://www.aao.org/eyenet/article/epidemic-keratoconjunctivitis-prevention-strategie>

Management

- Betadine (Melton-Thomas Protocol):
 - Proparacaine
 - 4-5 drops of Betadine 5%
 - Get patient to close eye and gently roll them around
 - After one minute, lavage the eye
 - Lotemax 4 times a day for 4 days
- Alternative: Betadine swabsticks.
 - 5% Betadine solution only comes in 30 ml bottles cost \$14.00.
 - Case of 200 Betadine swabsticks apprx. 45 dollars.

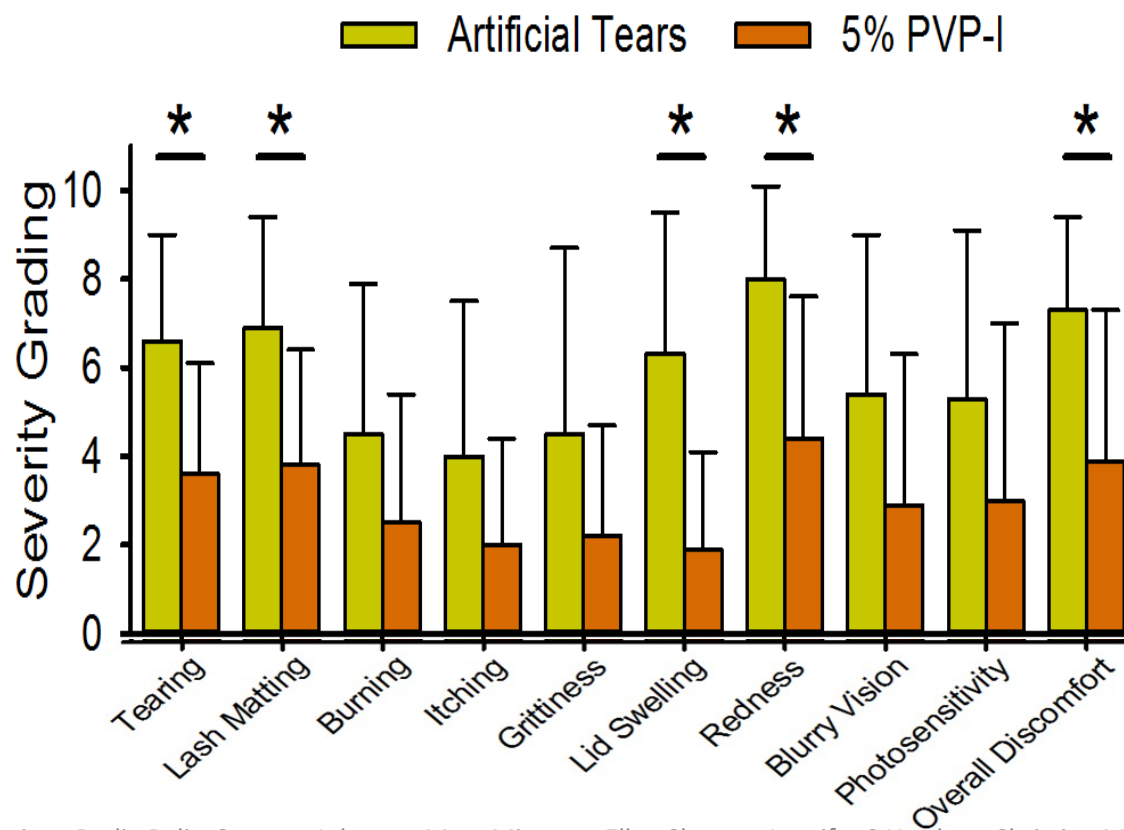
Reducing Adenoviral Patient-Infected Days (RAPID) Study

Effect of 5% PVP-I on qPCR-Derived Viral Titers

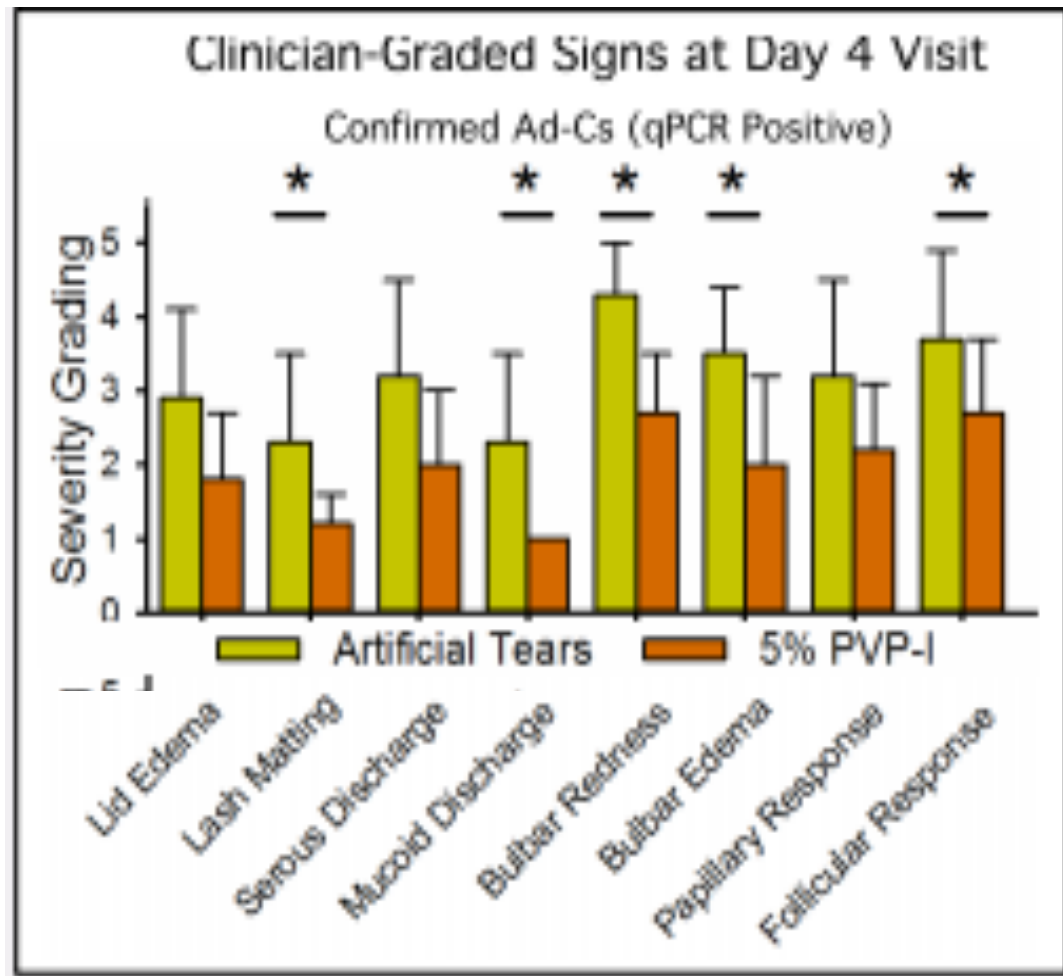


Andrew Hartwick, Tammy Than, Bojana Rodic-Polic, Spencer Johnson, Mary Migneco, Ellen Shorter, Jennifer S Harthan, Christina Morettin, Meredith Whiteside, Christian K. Olson, Mathew Margolis, Julia Huecker, Gregory Storch, Mae O Gordon; Reducing Adenoviral Patient-Infected Days (RAPID) Study: A Randomized Trial Assessing Efficacy of One Time, In-Office Application of 5% Povidone-Iodine in Treatment of Adenoviral Conjunctivitis. *Invest. Ophthalmol. Vis. Sci.* 2019;60(9):6257. doi: <https://doi.org/>.

Day 4: qPCR + Participant-Reported Symptoms



Andrew Hartwick, Tammy Than, Bojana Rodic-Polic, Spencer Johnson, Mary Migneco, Ellen Shorter, Jennifer S Harthan, Christina Morettin, Meredith Whiteside, Christian K. Olson, Mathew Margolis, Julia Huecker, Gregory Storch, Mae O Gordon; Reducing Adenoviral Patient-Infected Days (RAPID) Study: A Randomized Trial Assessing Efficacy of One Time, In-Office Application of 5% Povidone-Iodine in Treatment of Adenoviral Conjunctivitis. *Invest. Ophthalmol. Vis. Sci.* 2019;60(9):6257. doi: <https://doi.org/>.



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Management

- Antivirals used in HSV keratitis have traditionally thought to be ineffective in treatment of viral conjunctivitis
 - Ganciclovir: In a double-masked, controlled, and randomized study it was found to shorten the mean time of recovery from 18.5 days to 7.7 days in patients who were treated vs. those who just received artificial tears.
 - Tabbara K, Jarade E. Ganciclovir effects in adenoviral keratoconjunctivitis. 2001; ARVO abstract 3111 (suppl); S579
 - In clinical trial Avenova^R: proposed end date November 2020
 - The investigators propose a study to evaluate the role of Avenova[®] (0.01% hypochlorous acid) in the treatment of common ocular viral infections.
- Important to stress limited contact with others, frequent hand washing, not sharing of towels, etc.

Efficacy of Hospital Germicides against Adenovirus 8, a
Common Cause of Epidemic Keratoconjunctivitis in Health Care

Facilities. ANTIMICROBIAL AGENTS AND CHEMOTHERAPY, Apr. 2006, p. 1419–1424

An important finding from our study was that of the four disinfectants recommended by the CDC and Association for Professionals in Infection Control and Epidemiology for elimination of adenovirus type 8 from ophthalmic instruments, two **(70% isopropyl alcohol and 3% hydrogen peroxide) were found to be ineffective**. Based on these data, 3% hydrogen peroxide and 70% isopropyl alcohol are not effective against adenovirus that is capable of causing epidemic keratoconjunctivitis and similar viruses and should no longer be used for disinfecting applanation tonometers.

EKC Disinfection

- Commercial grade disinfectants that include compounds such as:
 - peracetic acid,
 - aldehydes [glutaraldehyde and ortho-phthalaldehyde],
 - chlorine-based products [1,900 to 6,000 ppm available free chlorine],
 - ethanol mixed with quaternary ammonium compounds)
- E.g. Cidex, DisCide

Case History

- 38 black male, complaining that the vision in his right eye is blurry.
 - Got the current Rx 3 weeks previously, and started out good but in last couple of days OD vision has become blurry
- Medical Hx: no current health concerns and no medications

Entrance Skills

- Va' s: OD: 20/25, OS: 20/20
- Pupils: PERRL
- CVF: full to finger count
- EOM' s: FROM
- Amsler: central metamorphopsia OD
- HVF: 10-2 (see VF)

SINGLE FIELD ANALYSIS

NAME: ██████████ ID: 110724 EYE: RIGHT DOB: 03-08-1969

CENTRAL 10-2 THRESHOLD TEST

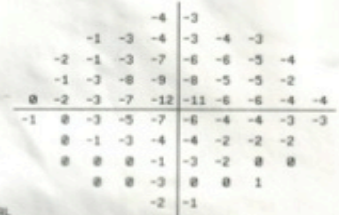
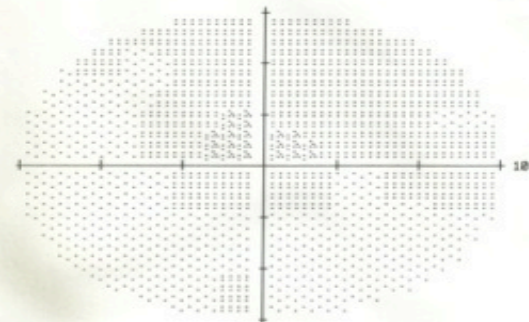
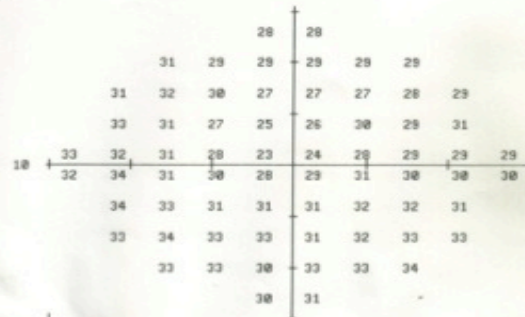
FIXATION MONITOR: GAZE/BLINDSPOT
 FIXATION TARGET: CENTRAL
 FIXATION LOSSES: 2/15
 FALSE POS ERRORS: 0 X
 FALSE NEG ERRORS: 0 X
 TEST DURATION: 06:32

STIMULUS: III, WHITE
 BACKGROUND: 31.3 ASB
 STRATEGY: SITR-STANDARD

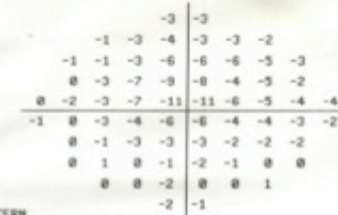
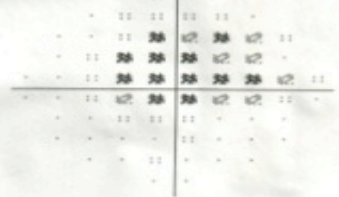
PUPIL DIAMETER: 5.5 MM
 VISUAL ACUITY:
 RX: +4.00 DS -2.00 DC X 115

DATE: 09-19-2007
 TIME: 10:07 AM
 AGE: 38

FOVER: OFF



TOTAL DEVIATION



PATTERN DEVIATION



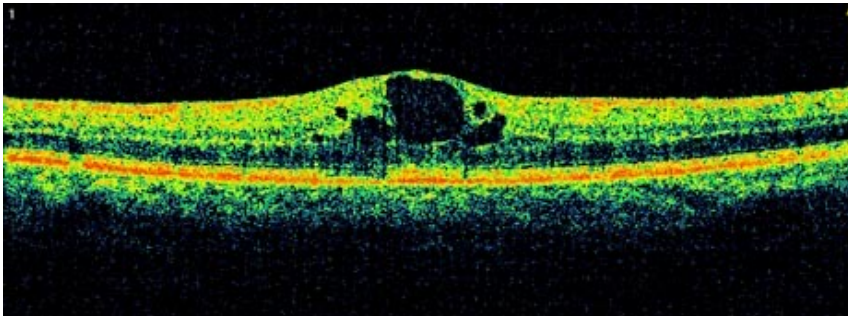
MD -3.12 DB P < 2%
 PSD 2.60 DB P < 1%

11 < 5X
 10 < 2X
 9 < 1X

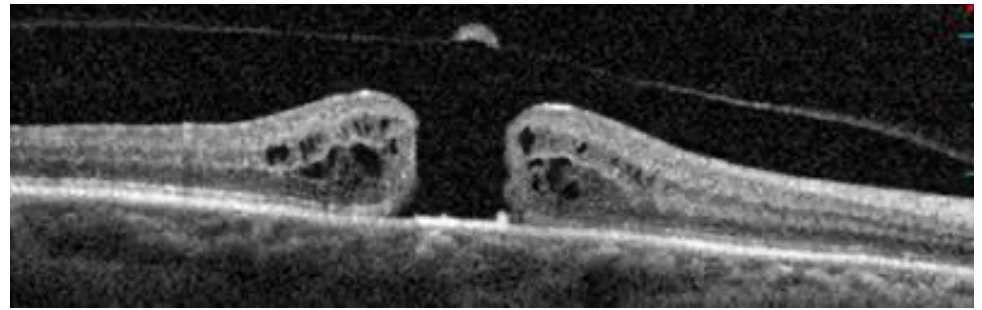
PACIFIC UNIVERSITY FAMILY EYE CENTER
 511 SW 10TH AVE
 PORTLAND, OR

Which of the following OCT's goes with this patient?

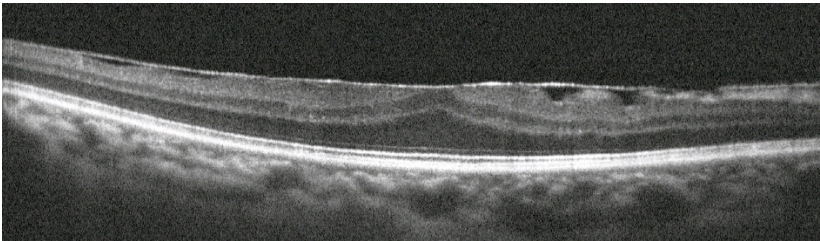
1



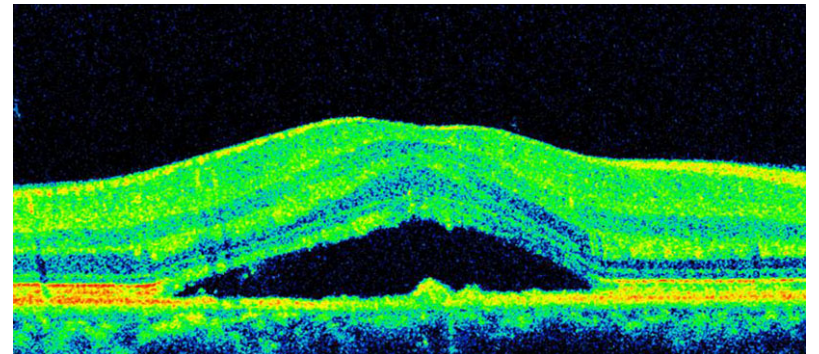
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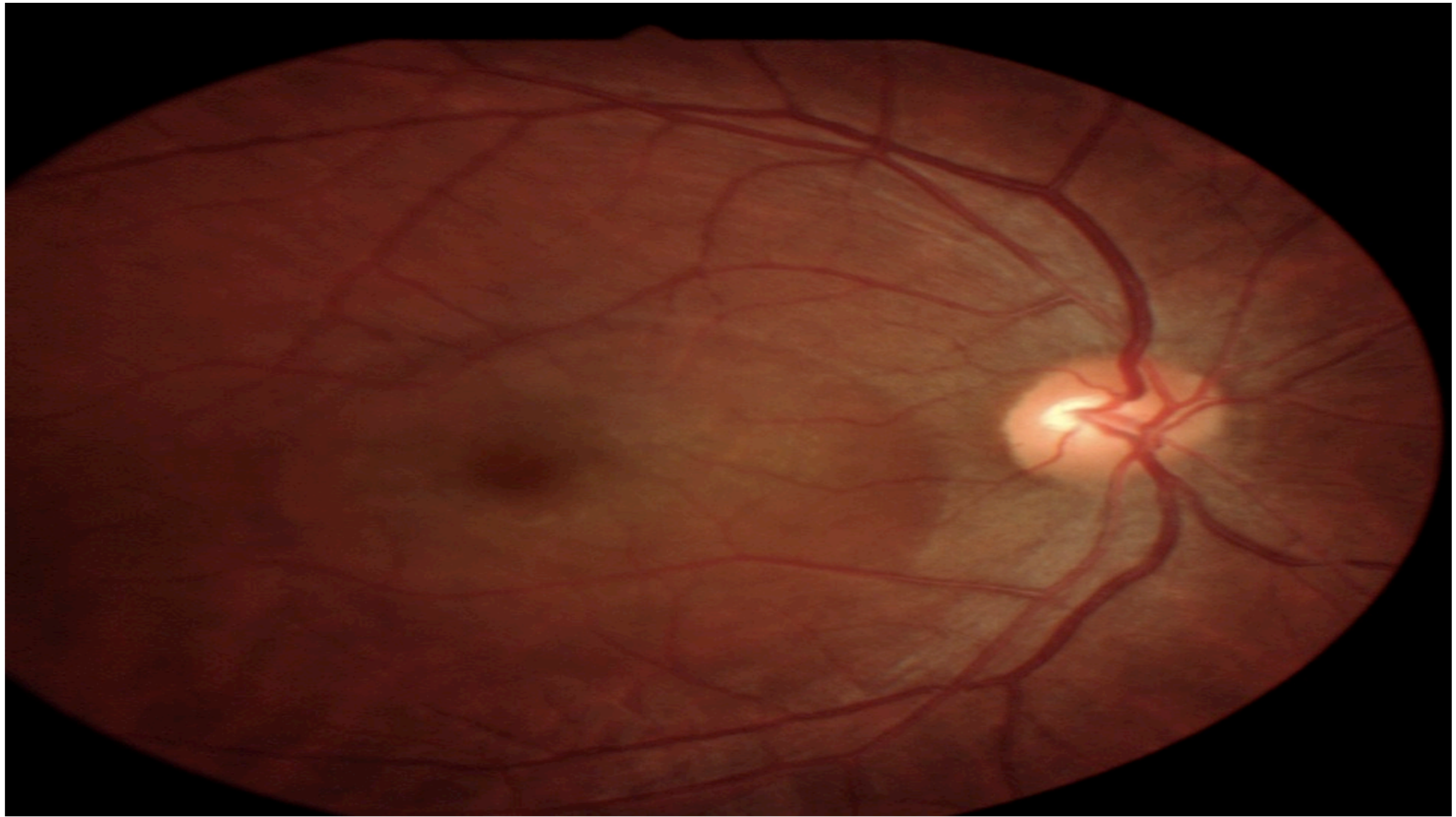


3



4





Central Serous Retinopathy

- an exudative chorioretinopathy characterized by an exudative neurosensory retinal detachment with or without an associated detachment of the retinal pigment epithelium (RPE)
- Patients experience blurry vision, metamorphopsia and micropsia
- individuals between 20 and 50 years of age

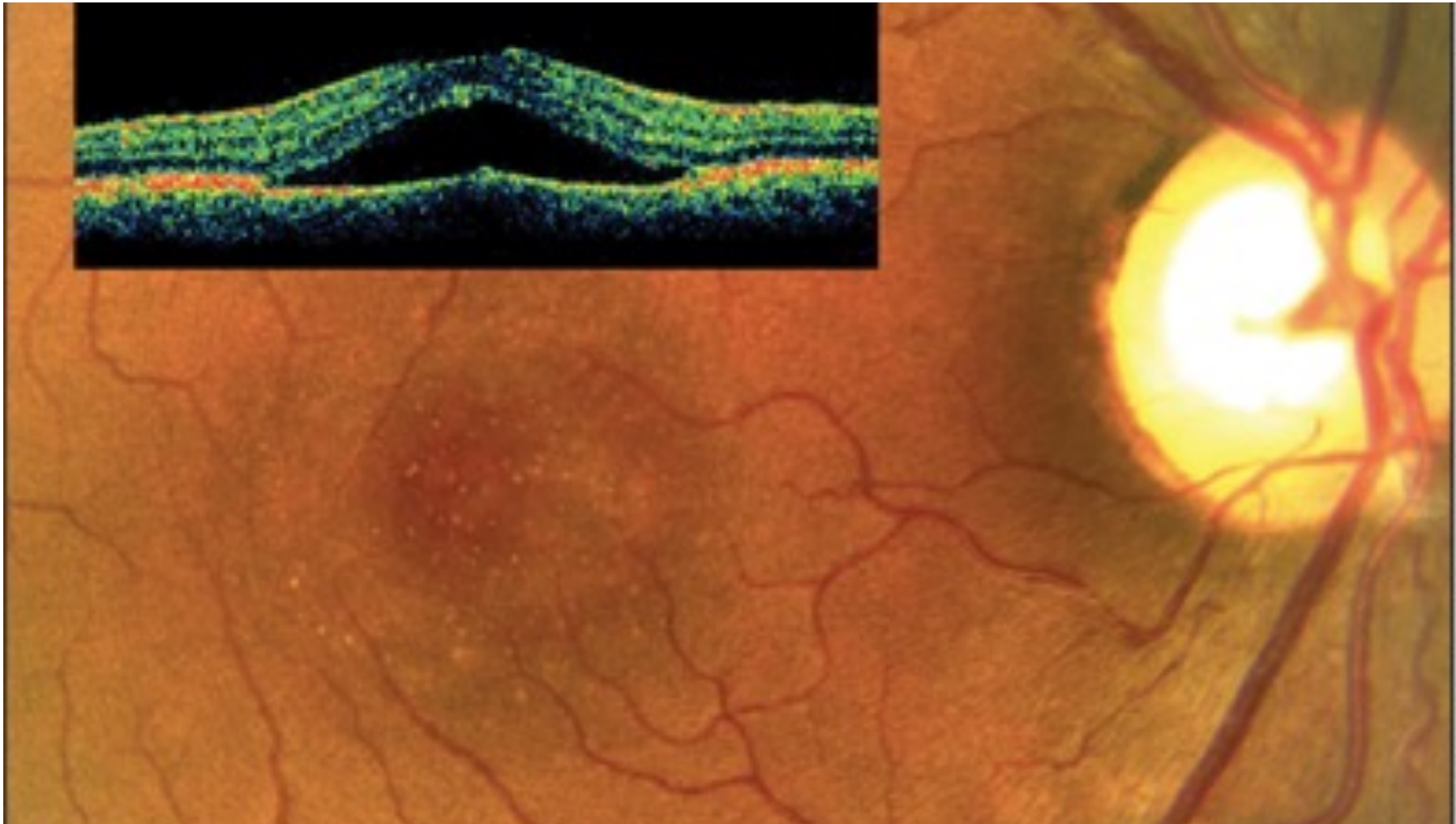
Central Serous Retinopathy

- incidence in men vs women is approximately 6:1
- associated with stress and stress hormones (ie, corticosteroids and epinephrine);
- individuals with a "type A personality" who are under stress
- recurrence in the ipsilateral eye is approximately 30% and CSR in the fellow eye was 32%

Central Serous Retinopathy

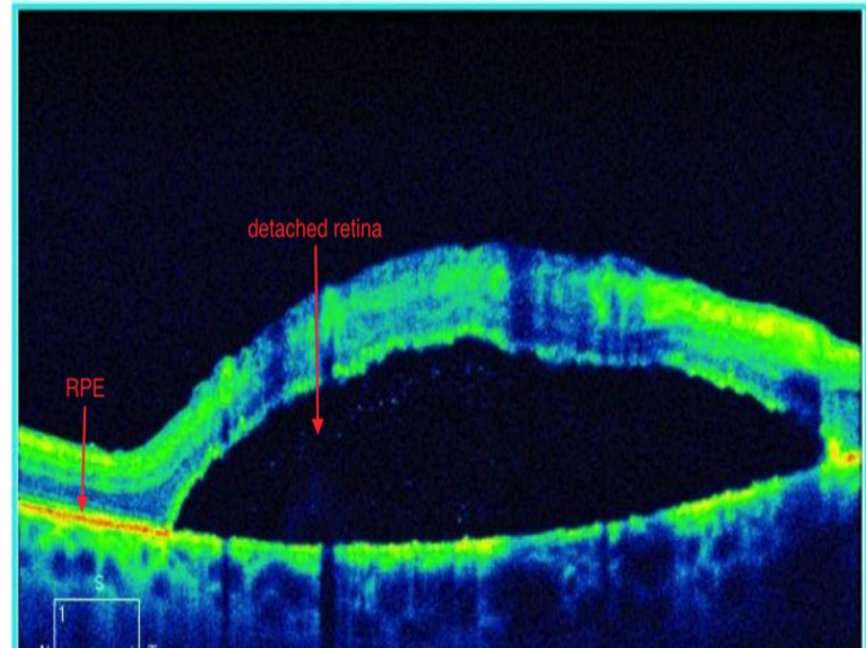
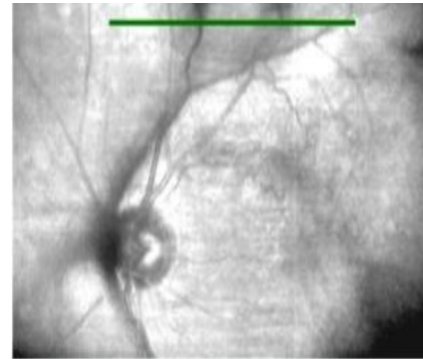
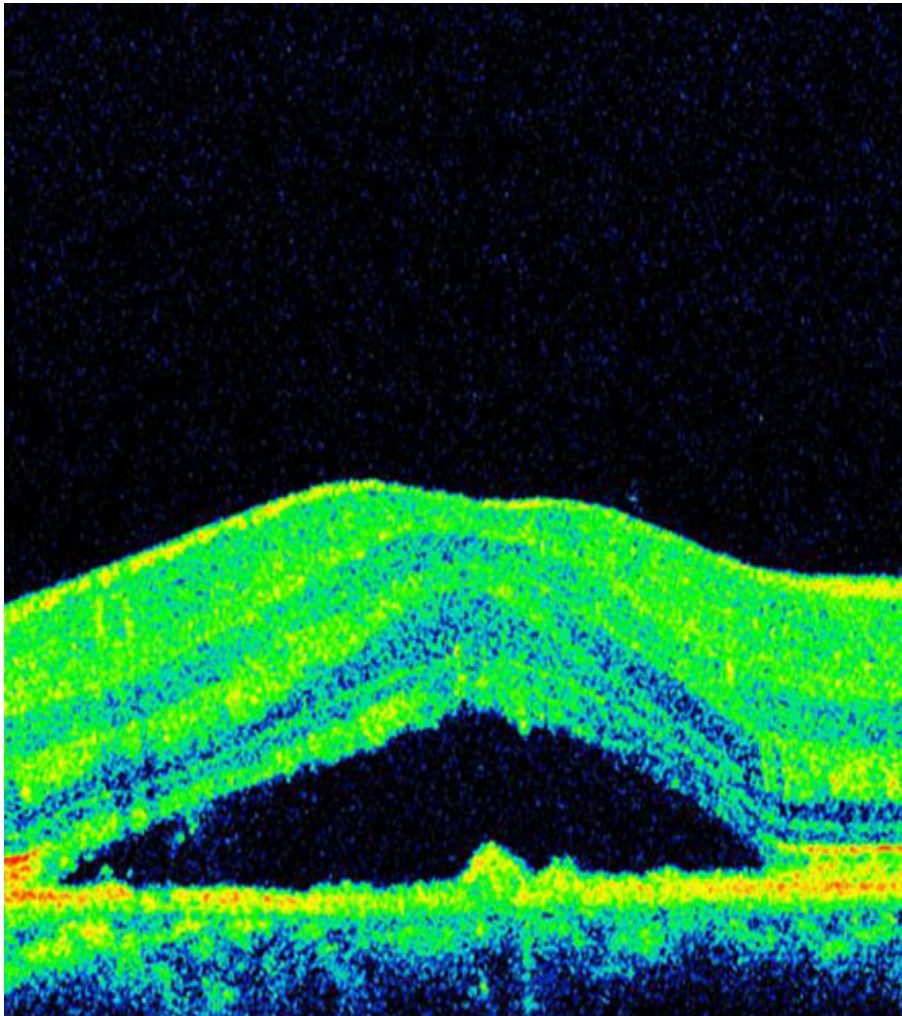
- systemic associations of CSCR include:
 - Sleep apnea syndrome
 - Systemic hypertension
 - Psychopharmacologic medications
 - Systemic lupus erythematosus
 - Gastroesophageal reflux disease
- Association between *H. pylori* infection, peptic ulcer disease and CSCR has been reported in some studies

Central Serous Retinopathy



FIGURES APPEAR COURTESY OF THE AUTHC

CSR versus RD



Central Serous Retinopathy

- 80% to 90% of cases resolve spontaneously within 3 months
- Treatment options:
 - include laser photocoagulation,
 - Anti-VEGF
 - Results remain inconclusive, and long term benefits warrant more studies.
 - "safety-enhanced" PDT (current "preferred" treatment option)
 - PDT causes vascular remodeling of the choroid and choroidal hypoperfusion.,
 - Acetazolamide reduced the time for subjective and objective CSR resolution, but it had no effect on final VA or recurrence rate. Most patients in the experimental group in that study had side effects from the acetazolamide, including paresthesias, nervousness, and gastric upset

Central Serous Retinopathy

- Treatment options:
 - Topical NSAIDs:
 - Conflicting reports
 - Michael Singer, MD, from Medical Center Ophthalmology in San Antonio reported an increase in resolution time by 50%
 - PRADEEP VENKATESH, MD reports that NSAIDS treatment could possibly slow down or cause a rebound CSR

Latest Treatment Under Investigation

- Eplerenone is a mineralocorticoid antagonist receptor currently used in the treatment of hypertension and congestive heart failure.
- Literature has demonstrated improved resolution of CSR with no serious adverse effects.
- Several randomized clinical trials are currently underway.
 - Currently, its use in CSCR remains investigational and is not considered standard of care

Case

- 65 year old Caucasian patient presents with sudden onset loss/blurring of vision in the right eye
- PMHx: HTN for 15 years, takes “water pill”
- VA’ s: 20/60 OD, 20/25 OS
- Pupils: PERRL -APD
- CVF: Inferior defect right eye, no defects noted in the left eye

Vision Loss Without Pain: Diabetes/Diabetic Retinopathy

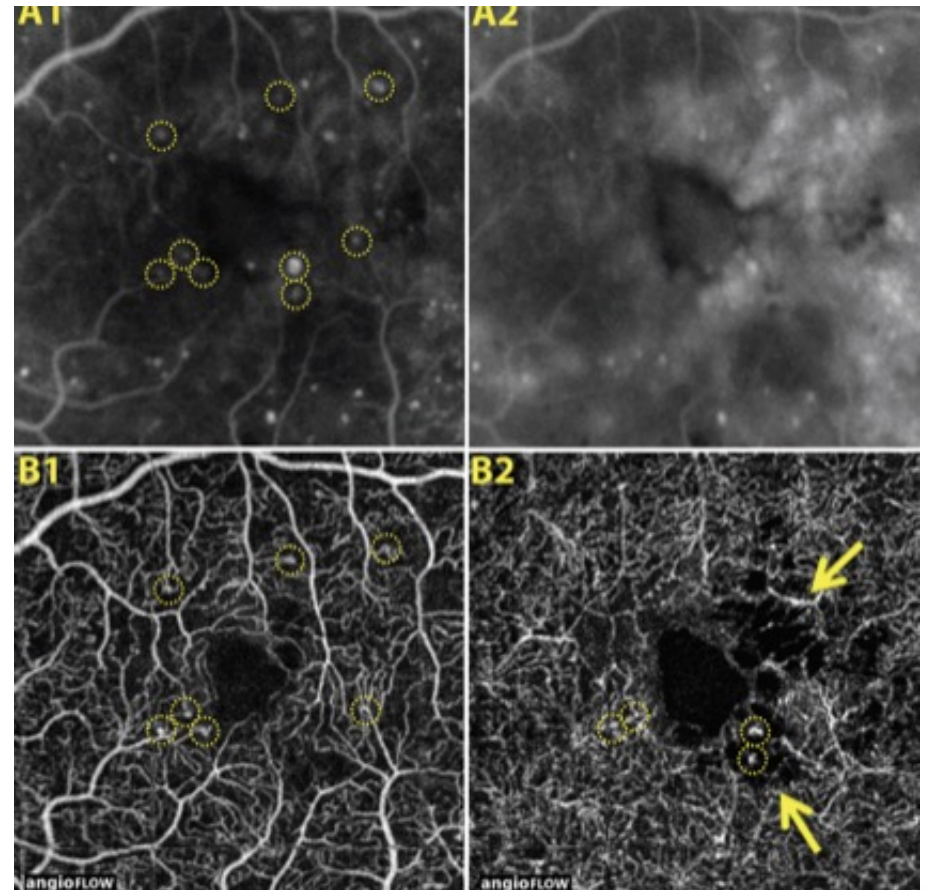
- Microvascular complications resulting in capillary closure & abnormal permeability
- S&S include;
 - blurring of vision (maculopathy and refractive error shifts),
 - sudden drop in vision (vitreous heme),
 - dot and blot hemes,
 - exudate,
 - cotton wool spots,
 - neovascularization (iris, retina and disc)

Diabetic Retinopathy

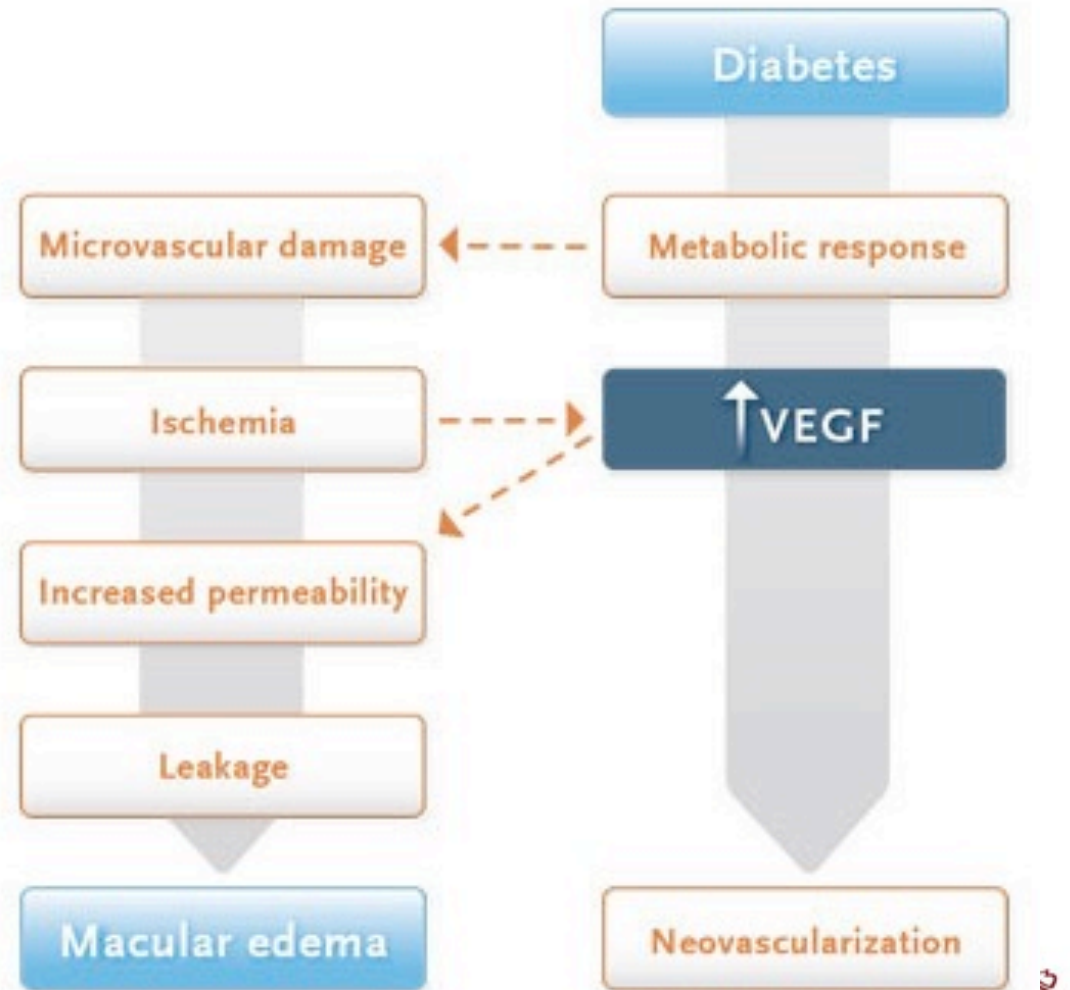
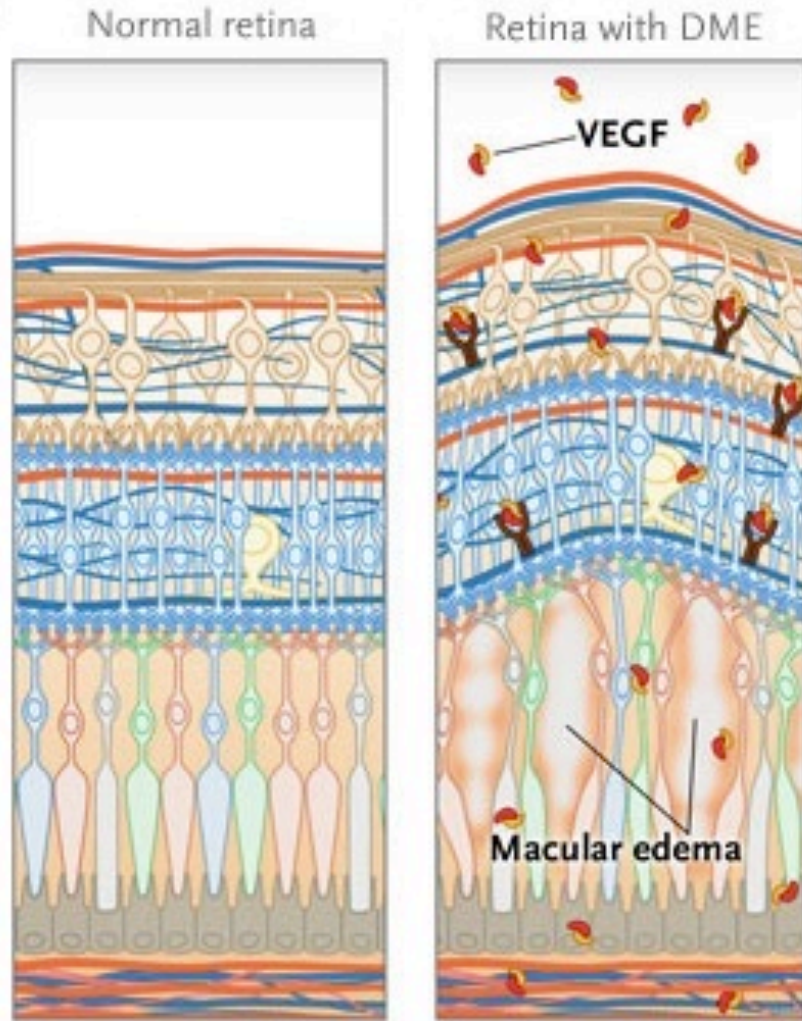
CSME (DME)



CSME (DME) OCTA



VEGF and DME

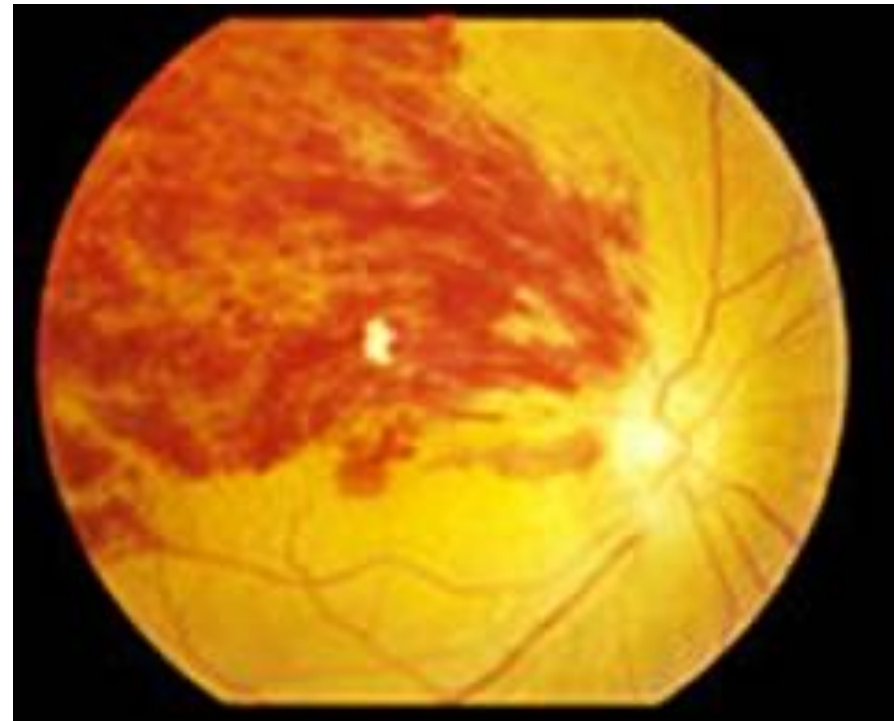


Vision Loss Without Pain: Vein Occlusion

- Associated with:
 - hypertension,
 - coronary artery disease,
 - DM and
 - peripheral vascular disease.
- Usually seen in elderly patients (60-70), slight male and hyperopic predilection.
- Second most common vascular disease after diabetic retinopathy.

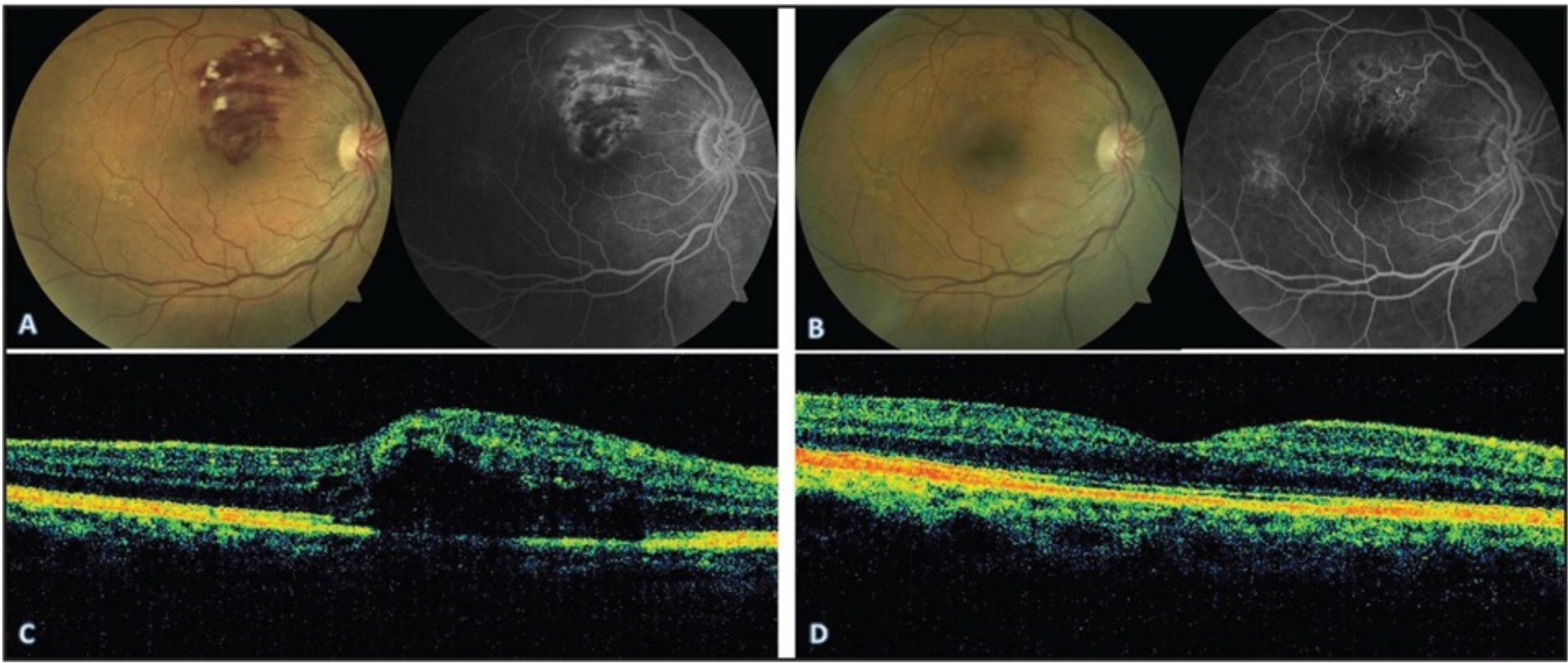
Branch Retinal Vein Occlusion: Signs/Symptoms

- BRVO: sudden, painless, visual field defect.
 - patients may have normal vision.
 - quadrantic VF defect,
 - dilated tortuous retinal veins with superficial hemes and CWS
 - typically occurs at A/V crossing (sup/temp)

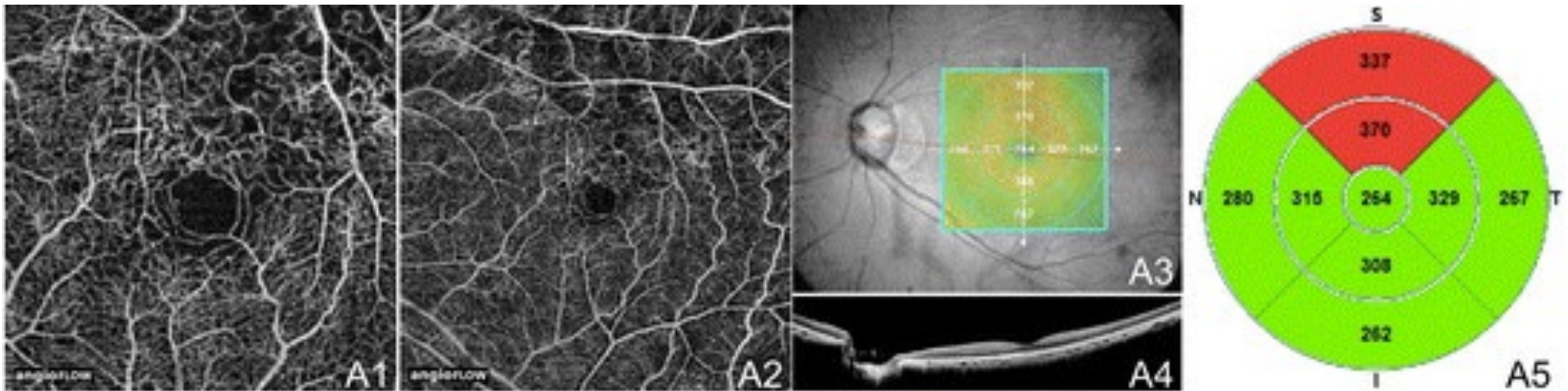


BRVO

- BRVO more common than CRVO and has more favorable prognosis
 - Overall 50-60% of BRVO patients will maintain VA of 20/40 or better
- Visual loss results from:
 - Macular edema
 - Foveal hemorrhage
 - Vitreous heme
 - Epiretinal membrane
 - RD
 - Macular ischemia
 - Neovascularization complications



<http://www.healio.com/ophthalmology/journals/osli/>





Study Design (n=397) BRVO

BRANCH retinal Vein Occlusion study safety/efficacy

Macular Edema Secondary to BRVO

1:1:1 Randomization

Sham
(n=132)

Ranibizumab
0.3 mg
(n=134)

Ranibizumab
0.5 mg
(n=131)

Monthly Injections (last at 5M)
Rescue Laser (if eligible beginning at Month 3)

12M

Month 6
Primary
Endpoint

PRN ranibizumab for all patients
Rescue Laser (if eligible beginning at Month 9)

Ranibizumab
0.5 mg

Ranibizumab
0.3 mg

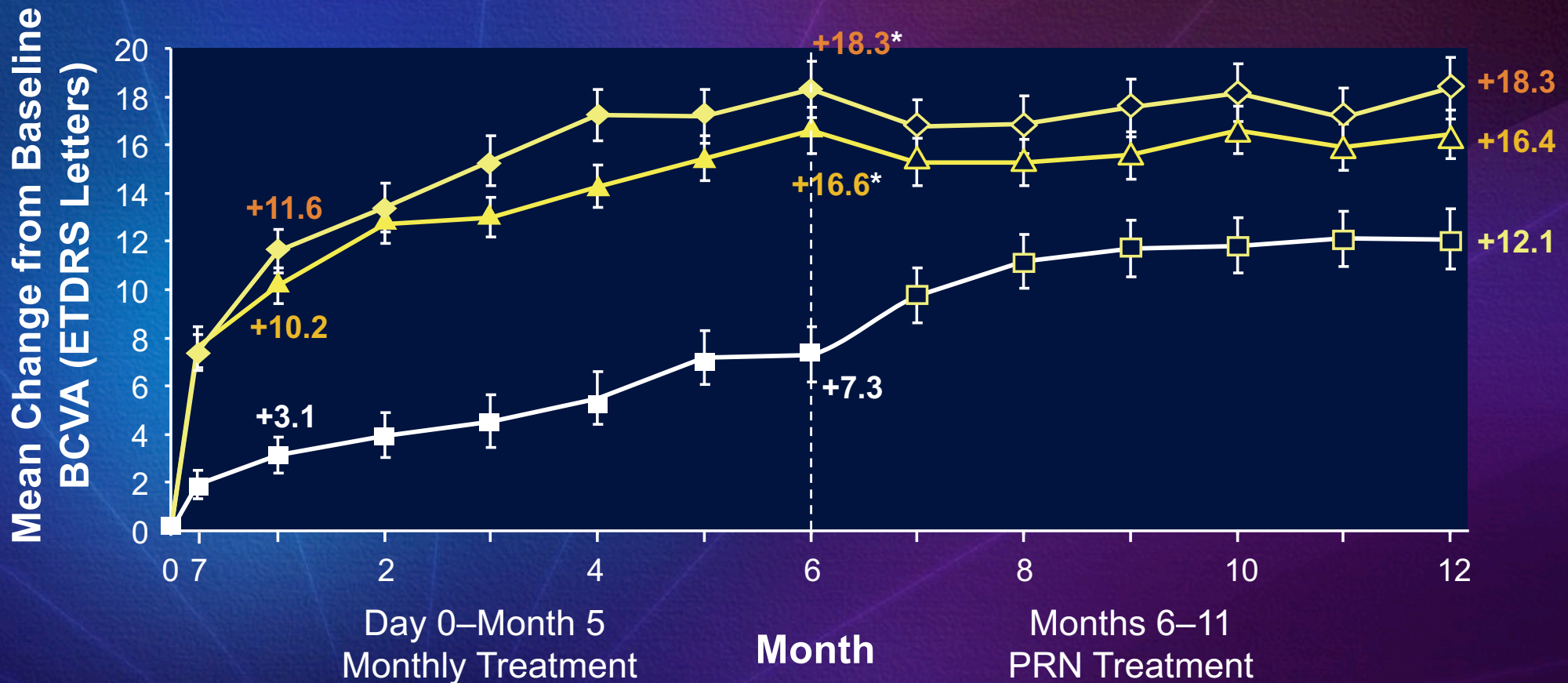
Ranibizumab
0.5 mg



Mean Change from Baseline BCVA

BRVO

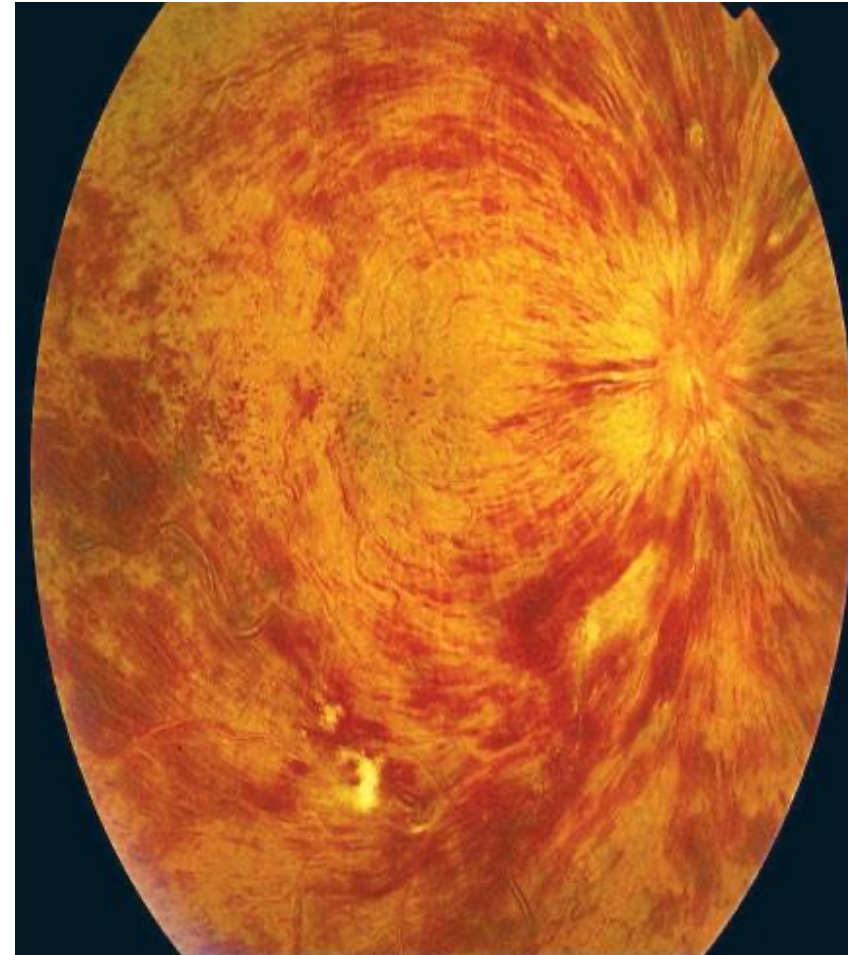
■ Sham/0.5 mg (n=132) ▲ 0.3 mg Ranibizumab (n=134) ◆ 0.5 mg Ranibizumab (n=131)



The gain of additional 3 lines occurred at a rate of 61% of 0.5 AVT grp, 55% for 0.3 AVT & 29% placebo

Central Retinal Vein Occlusion: Signs/Symptoms

- CRVO: thrombus occurring at lamina is classical theory but new evidence indicates that the occlusion is typically in the optic nerve posterior to the lamina cribrosa
 - decreased VA ranging from near normal to hand motion with majority 20/200 range
 - dilated tortuous vessels, with numerous retinal hemes and CWS



Central Retinal Vein Occlusion

- Visual morbidity and blindness are primarily from:
 - persistent macular edema,
 - macular ischemia and
 - neovascular glaucoma

Central Retinal Vein Occlusion

- CRVO's can be ischemic or non.
 - Classical definition of ischemic is 10-disc area of non-perfusion found on angiography
 - RAPD and ERG maybe better predictor
 - VA's typically worse in ischemic
 - Increased number of cotton wool spots with decreased VA maybe predictive

Central Retinal Vein Occlusion

- Ischemic CRVO may lead to iris neovascularization and neovascular glaucoma
 - Estimated apprx 20% of CRVO's are ischemic with 45% of those developing neo
- Regular examinations (1-2 wks) to monitor for ischemia or neo development
 - should include gonio as angle neo can precede iris rubeosis



Study Design CRUISE (n=392)

CRVO

Central Retinal vein occlusion Study: Efficacy & safety

Macular Edema Secondary to CRVO

1:1:1 Randomization

Sham
(n=130)

Ranibizumab
0.3 mg
(n=132)

Ranibizumab
0.5 mg
(n=130)

Monthly Injections (last at 5M): 6M tx period

12M trial

PRN Lucentis available for for **all patients**: 6M tx period

Month 6
Primary
Endpoint

0.5 mg

Ranibizumab
0.3 mg

Ranibizumab
0.5 mg



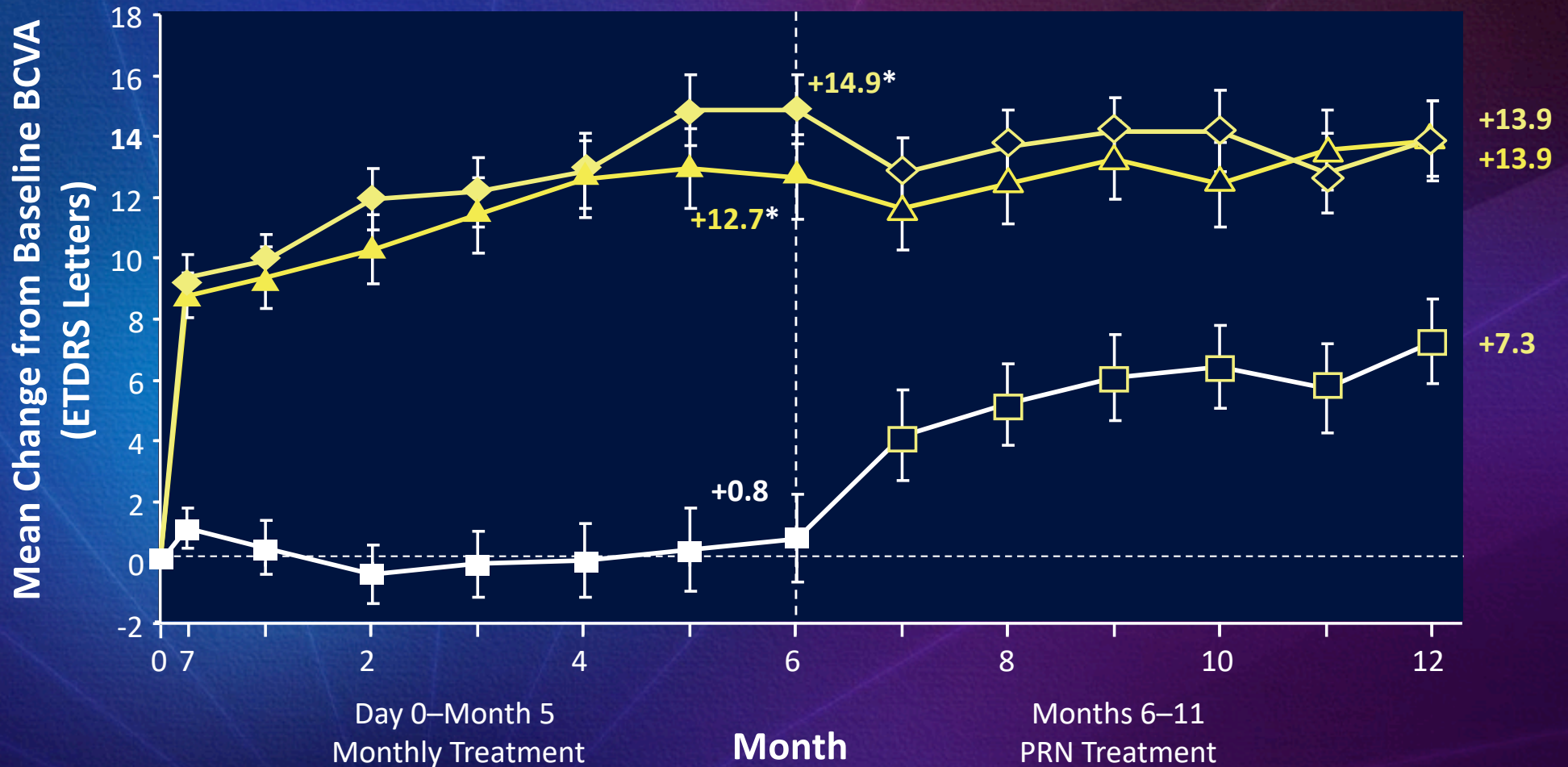
Mean Change from Baseline BCVA

CRVO

Sham/0.5 mg (n=130)

0.3 mg Ranibizumab (n=132)

0.5 mg Ranibizumab (n=130)



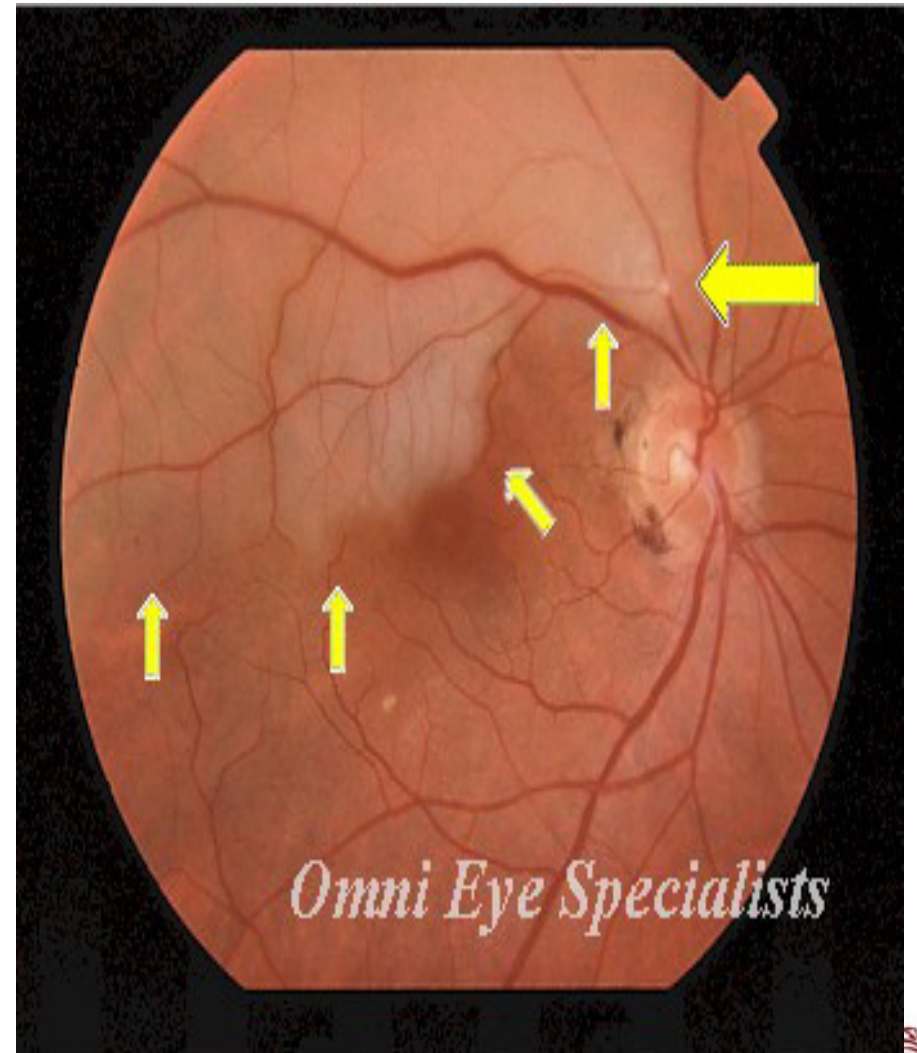
Pts with ≥ 3 line improvement was noted in 48% of .5 AVT, 26 of .3 AVT & 17% of sham

Vision Loss Without Pain: Artery Occlusion

- Primarily embolic in nature from cholesterol, calcifications, plaques.
- Usually occurs in elderly associated with:
 - hypertension (67%),
 - carotid occlusive disease (25%),
 - DM (33%) and
 - cardiac valvular disease.
- Sudden loss of unilateral, painless vision
 - defect dependent upon location of occlusion

Vision Loss Without Pain: Artery Occlusion

- BRAO typically located in temporal retinal bifurcations.



CRAO

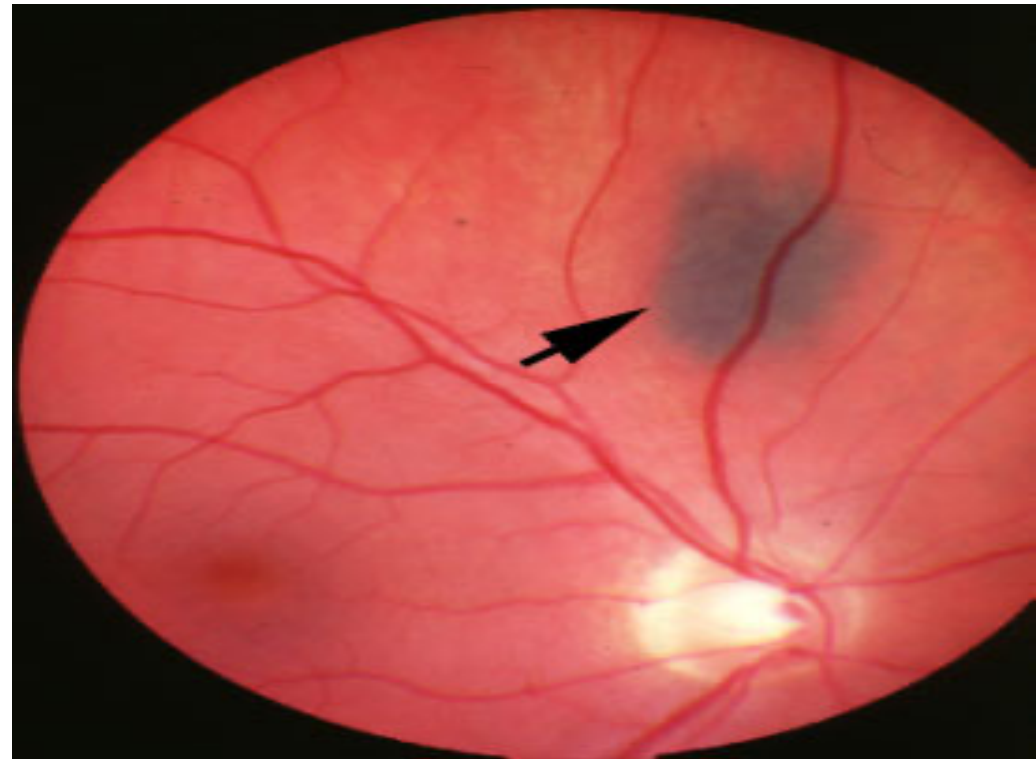
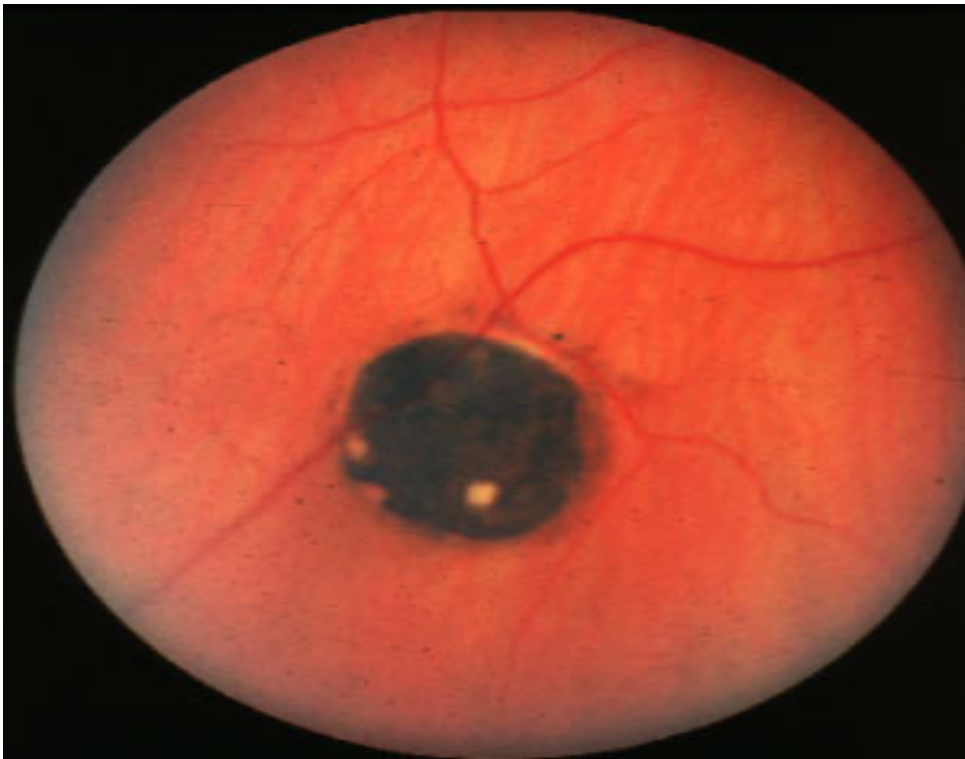
- CRAO has profound vision loss with history of amaurosis fugax.
 - Vision is usually CF (count fingers) to LP (light perception) with positive APD.
 - Diffuse retinal whitening with arteriole constriction, cherry red macula.



Ophthalmic Emergency

- Treatment is controversial due to poor prognosis and questionable benefit.
- Treat immediately before workup, if patient presents within 24 hours of visual loss:
 - Digital ocular massage,
 - systemic acetazolamide (500 mg IV or po),
 - topical ocular hypertensive drops (Iopidine, B-blocker),
 - anterior chamber paracentesis,
 - consider admission to hospital for carbogen Tx (high carbon dioxide)

CHRPE vs Nevus



CHRPE vs Hamartomas

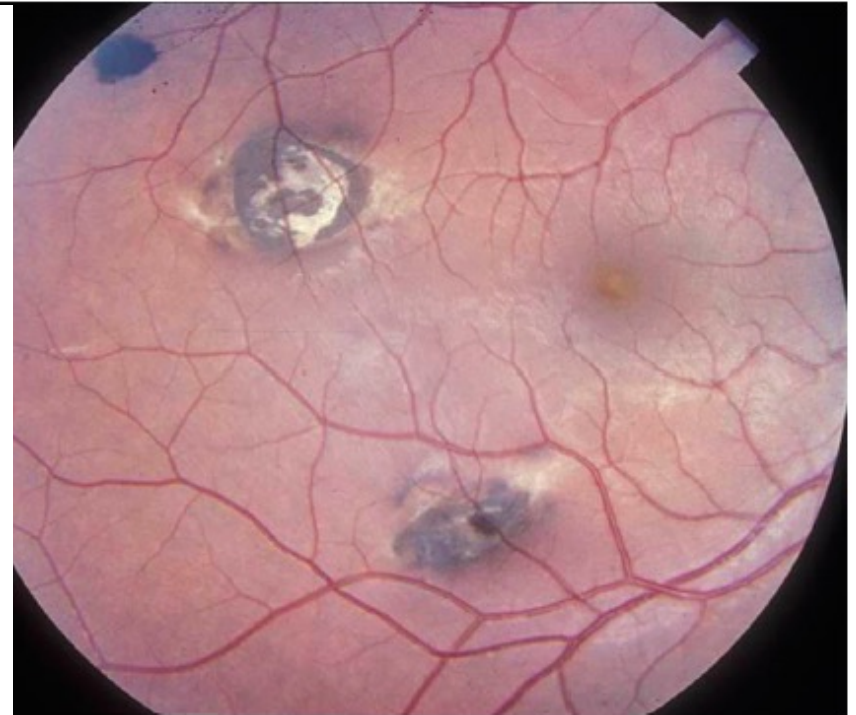
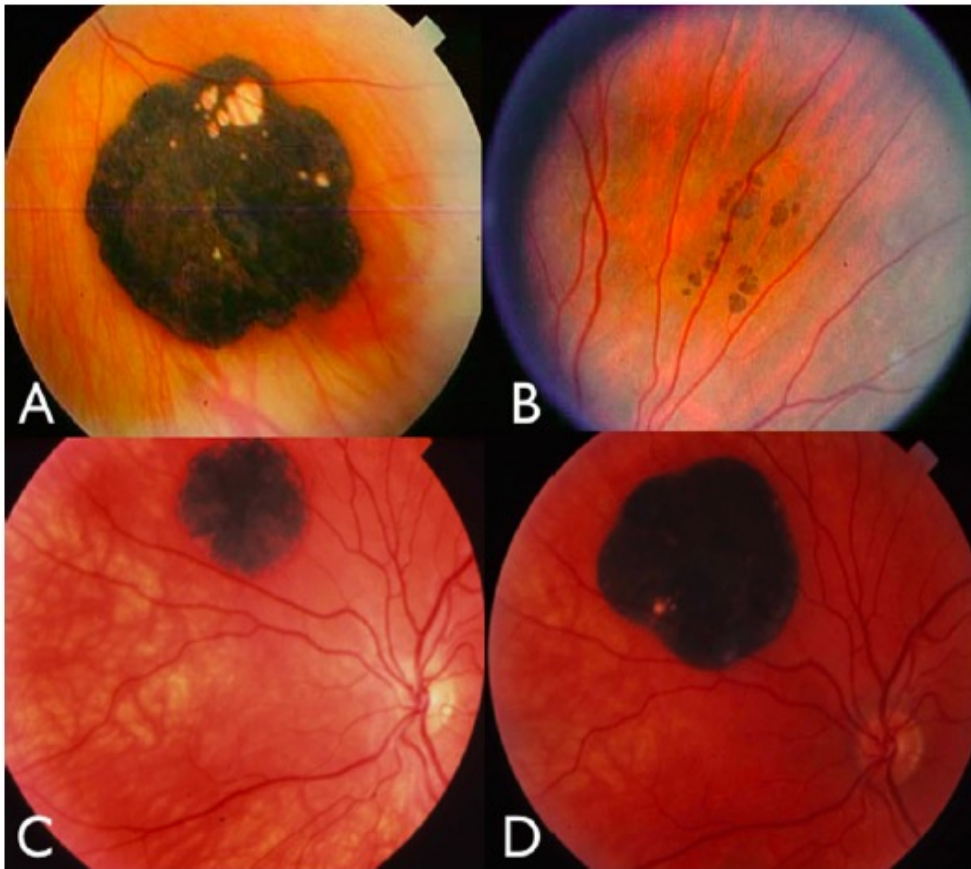


FIGURE 4. Retinal pigment epithelial hamartomas (pigmented ocular lesions) associated with familial adenomatous polyposis (FAP) syndrome).

Nevi Trivia

- 31% of choroidal nevi show slight enlargement over time without the transformation to a melanoma (Ophthalmology 2011)
- The prevalence of choroidal nevi in the white U.S. population ranges from 4.6% to 7.9%
 - If it is assumed that all choroidal melanomas arise from preexisting nevi, then the published data suggest a low rate (1/8845) of malignant transformation of a choroidal nevus in the U.S. white population. (Ophthalmology 2005)
- Choroidal melanoma risk for metastasis, ranging from 16% to 53% (at 5 years of follow-up) depending on the size of the tumor at the time of diagnosis. (Arch Ophthalmol 1992)

TFSOM—“To Find Small Ocular Melanoma”

Thickness: lesions >2mm

Fluid: any subretinal fluid (suggestive of serous retinal detachment)

Symptoms: photopsia, vision loss

Orange pigment overlying the lesion

Margin touching optic nerve head (<3mm)

- None of these factors = 3% risk of a nevus converting to melanoma in five years.
One of these factors = 8% risk of conversion in five years. Two or more factors = 50% risk of conversion in five years. For any changes noted during the course of follow-up, refer the patient to a retinal practice or an ocular oncology service.

TFSOM-UHHD:

“To Find Small Ocular Melanoma Using Helpful Hints Daily”

Thickness: lesions >2mm

Fluid: subretinal fluid

Symptoms: photopsia, vision loss

Orange pigment overlying the lesion

Margin touching optic nerve head (<3mm)

Ultrasound Hollowness

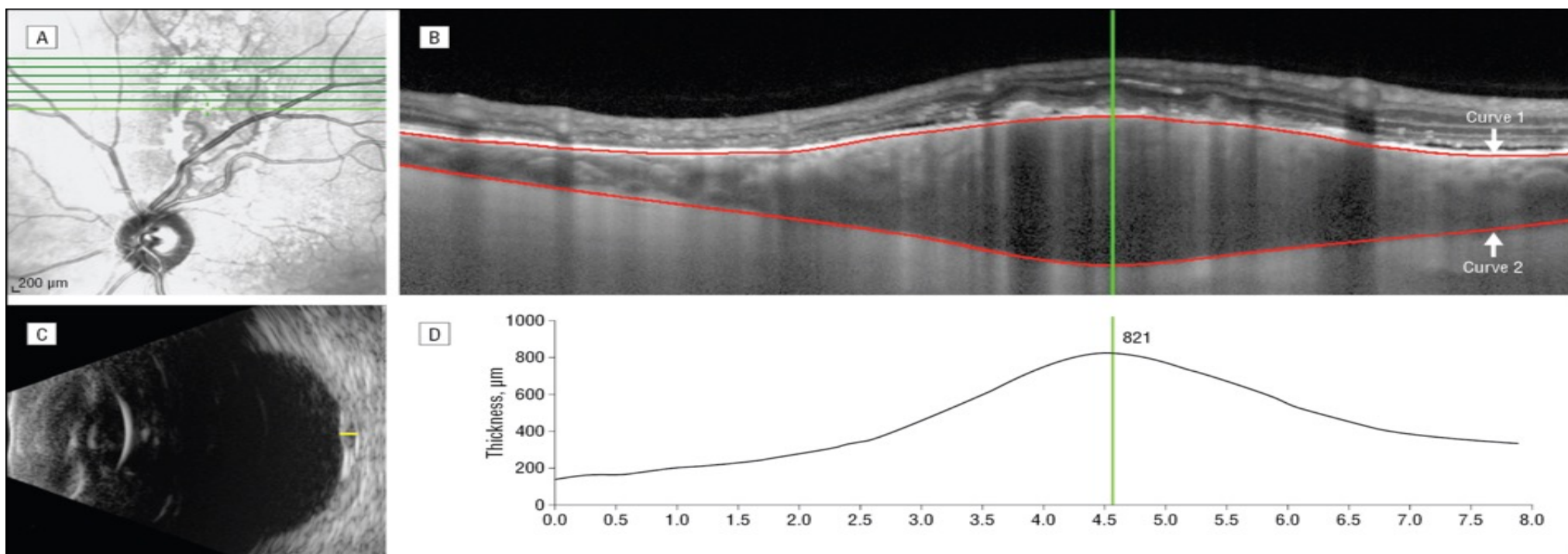
Halo absence

Drusen absence

- Choroidal nevi showing no features should be initially monitored twice yearly and followed up annually
- 1 or 2 features should be monitored every 4 to 6 months.
- Nevi with 3 or more features should be evaluated at an experienced center for management alternatives and possible treatment owing to the high risk of ultimate growth

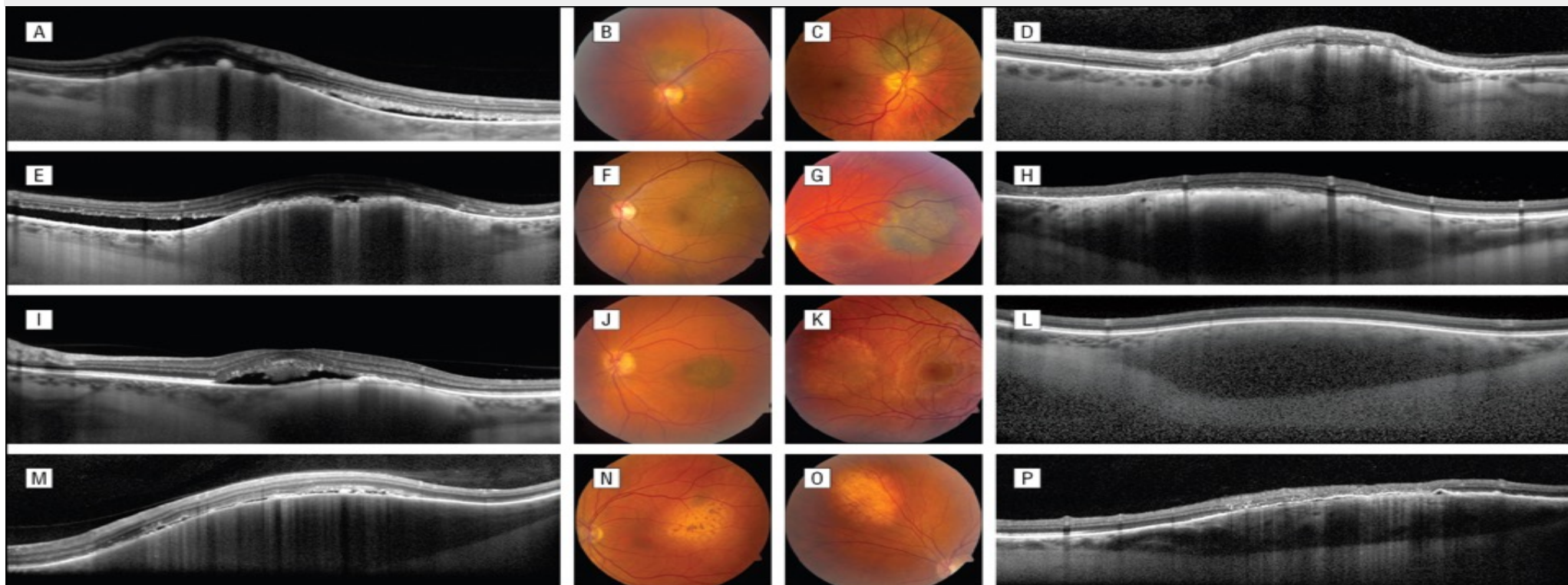
From: **Enhanced Depth Imaging Optical Coherence Tomography of Small Choroidal Melanoma: Comparison With Choroidal Nevus**

Arch Ophthalmol. 2012;130(7):850-856. doi:10.1001/archophthalmol.2012.1135



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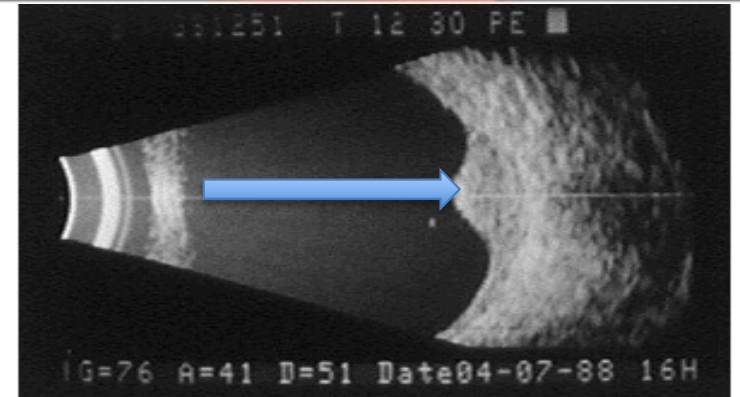
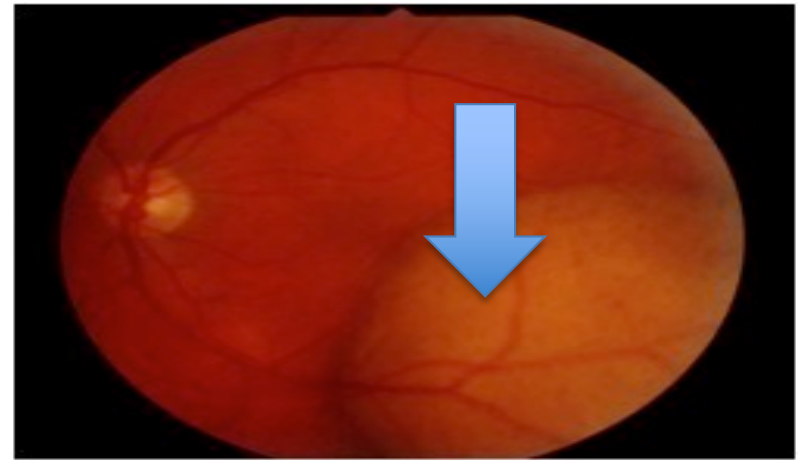


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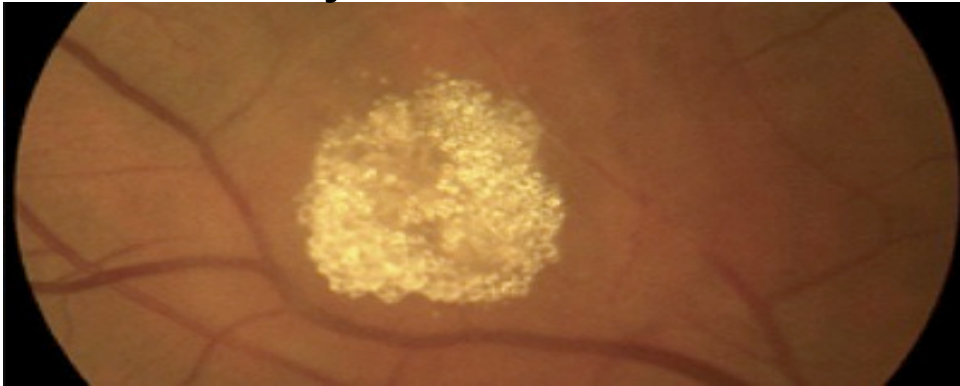
Case

- 65 yr old white male
 - Notices spot in vision in his left eye
 - Diabetes for 15 years
- Vision: 20/20 (6/6) and 20/40 (6/12)
- Dilated exam:
 - Large lesion noted in left eye (not noted in exam 6 months previously)
 - See photo and B-scan

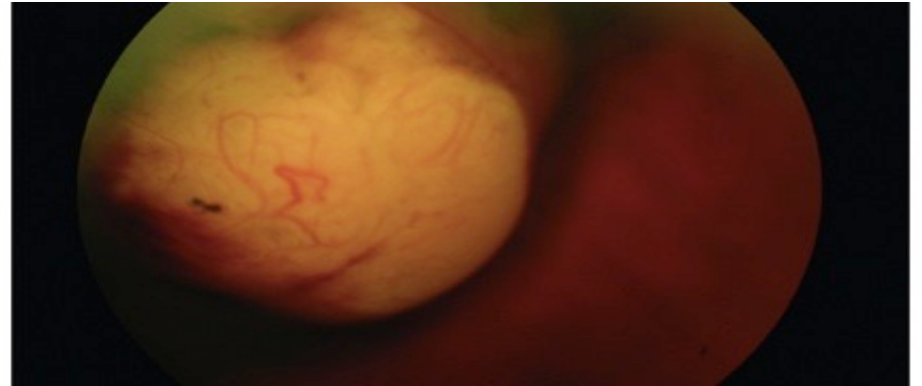


Ocular Tumors

Astrocytic Hamartoma



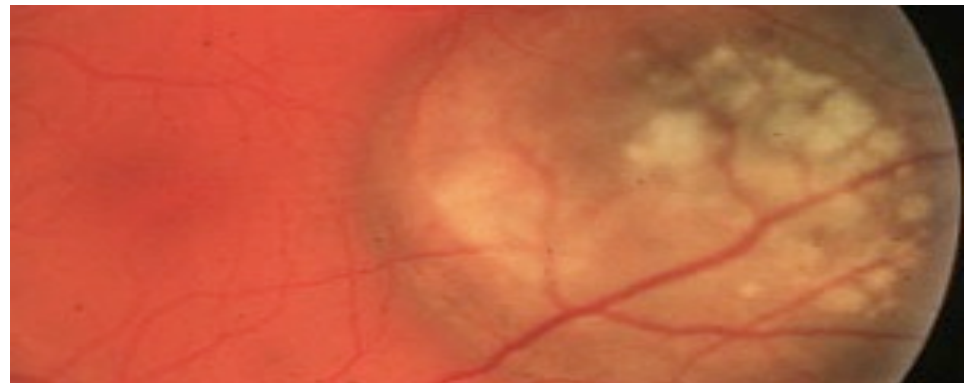
Amelanotic Melanoma



Retinoblastoma



Metastatic Choroidal Tumor



Choroidal Melanoma Metastases

- 80 to 90% of metastases from uveal melanoma occurred in the liver, less common sites being the skin and lung.
 - Gragoudas ES, Seddon JM, Egan KM, et al. Long-term results of proton beam irradiated uveal melanomas. *Ophthalmology*. 1987;94:349–53.

Melanoma and Mortality

- Tumor Size:
 - 5-year mortality after enucleation:
 - 16% for small melanoma,
 - 32% for medium melanoma, and
 - 53% for large melanoma.
 - the prognostic importance of tumor size:
 - each 1-mm increase in melanoma thickness adds approximately 5% increased risk for metastatic disease at 10 years
- Tumor genetics:
 - Chromosome monosomy 3 (apprx 50% of patients)
 - 50% of them develop metastasis within 5 years of diagnosis
 - 70% mortality within 4 years of ocular treatment
 - one of the most important independent risk factors of poor survival

New Treatment for Choroidal Melanoma

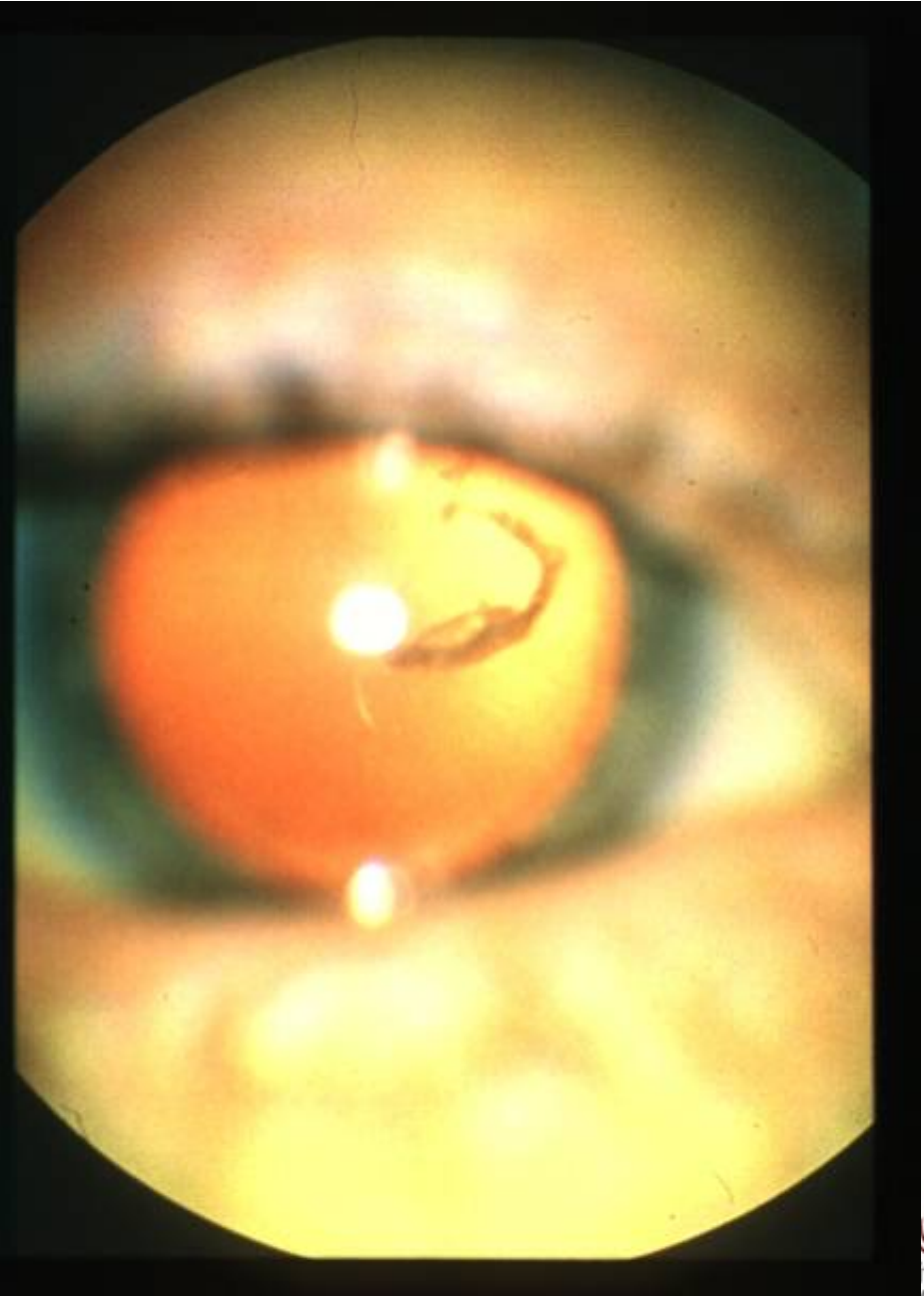
- light-activated AU-011 agent represents the first potential new therapy for choroidal melanoma
- AU-011 is a viral nanoparticle conjugate delivered by intravitreal injection, which targets tumor cells in the choroid and then is activated by ophthalmic laser to disrupt the tumor cell membrane, leading to necrosis.
- Two year prospective study complete

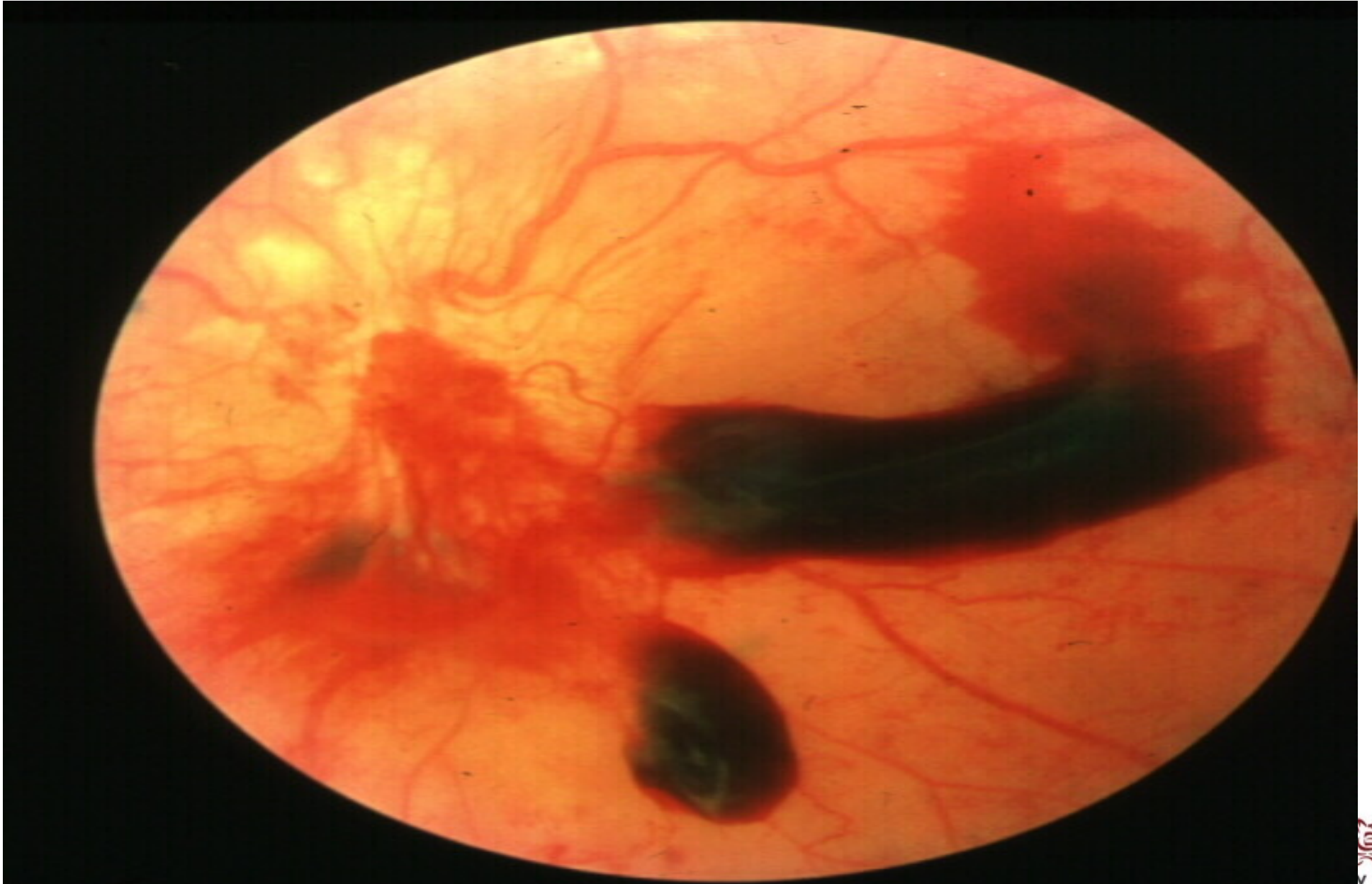
New Treatment for Choroidal Melanoma

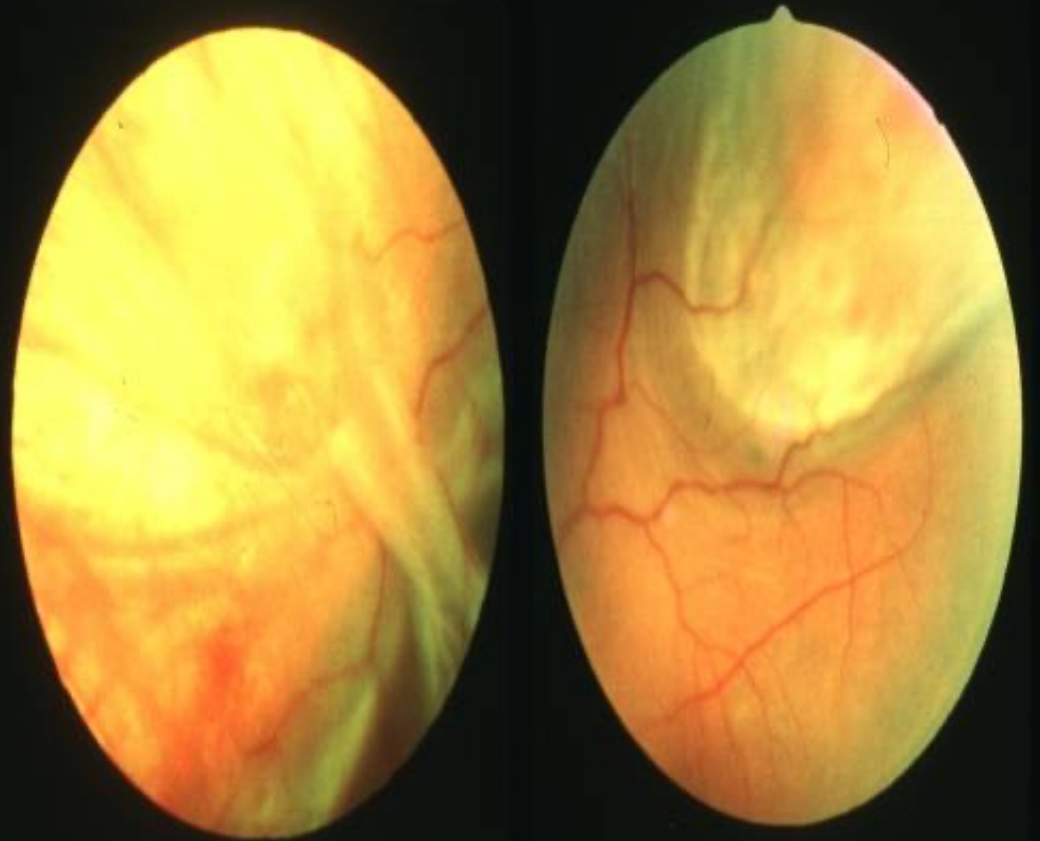
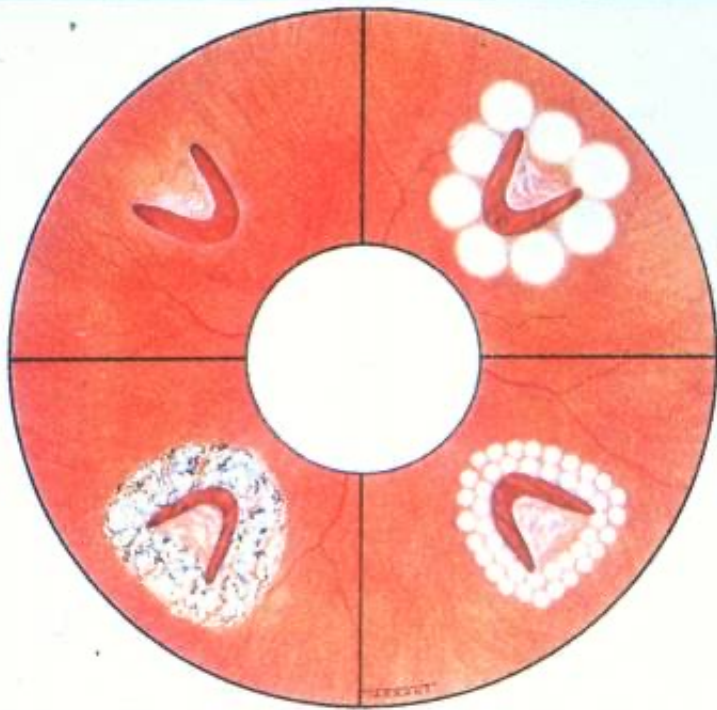
- Total cohort of 36
 - 12 patients in the single-dose cohort demonstrated a modest tumor control rate of 67% with a follow-up period of 9 to 24 months, and
 - 22 patients in the multiple-dose cohort (2 patients lost to follow-up) demonstrated a modest tumor control rate of 77% with a follow-up period of 0.5 to 18 months.
 - Subjects treated with the maximum safe and tolerated dose (80 μ g with 2 lasers) with 0.5 months to 6 months follow-up have a tumor control rate of 92% (13 of 14 subjects).
 - Vision was preserved in all patients at 3 months or longer up to 24 months.

Flashes and Floaters

- Patients often present complaining of “spots” or “cobwebs” in front of their eyes
- Causes of floaters include: posterior vitreous detachment (PVD), retinal tear, vitreous heme, uveitis.
- Since PVD and retinal tears present the same way, a RT has to be eliminated
- Ask the patient whether spots move with eye and continue to move after the eye has stopped
- Large spots could be blood clots

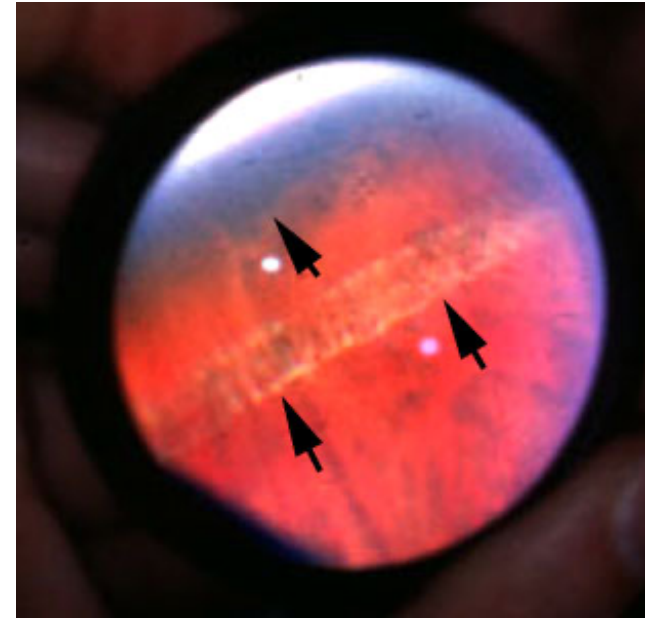






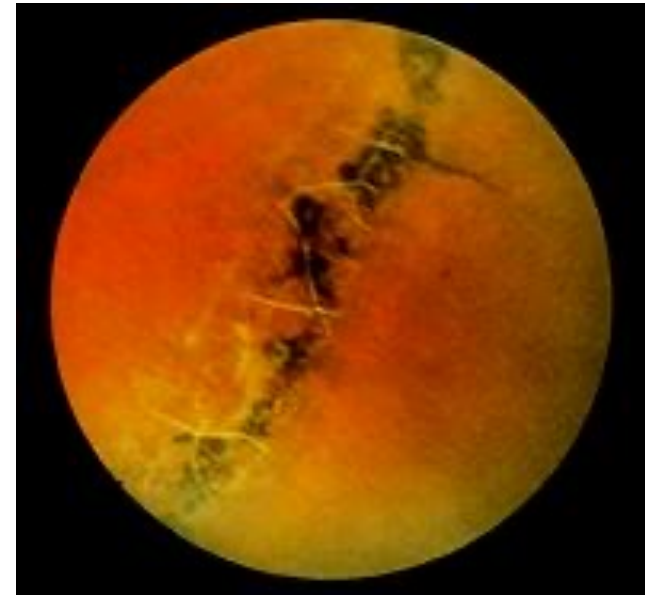
PDS Clinical Features

- *Posterior segment*
 - Lattice degeneration occurs in 8-11% of the general population
 - The incidence of atrophic holes in lattice degeneration ranges from 18-42%
 - Lattice retinal degeneration has been reported to be evident in 20-33% of cases of PDS and PDG
 - greater than would be expected for the associated myopia



PDS Clinical Features

- *Posterior segment*
 - retinal breaks occur more frequently than in normal eyes, affecting 12% of eyes with PDS and PDG
 - risk of retinal detachment is only 0.1-0.7% in the “normal” phakic eye
 - retinal detachments have been reported to occur in 5.5–6.6% of PDS cases
 - higher than expected for the degree of myopia and is independent of miotic use



Flashes and Floaters: PVD

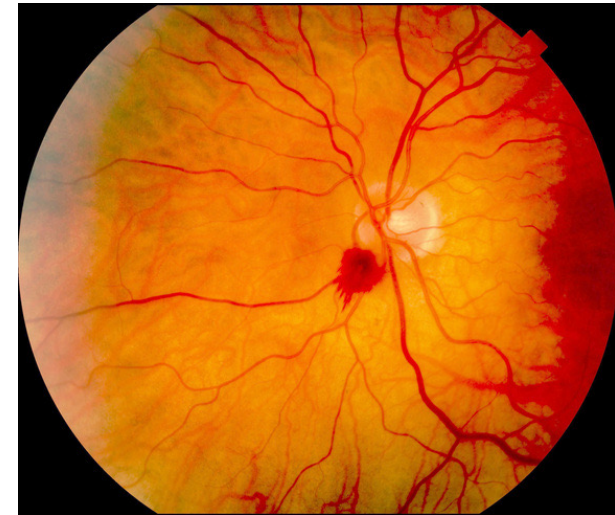
- Sudden onset typically means a PVD, retinal tear or heme
- If the spots appear after flashing light, then retinal tear must be eliminated
 - 10-15% of patients with acute symptomatic PVD are found to have a retinal tear
- The prevalence of PVD increases with age, with axial length, and following cataract surgery and trauma.
- PVD is rare in persons under 30 years of age, and it increases from 10 percent between ages 30 and 59, to 27 percent between ages 60 and 69, and to 63 percent in persons age 70 years and older.

Flashes and Floaters: PVD

- Patients with myopia experience PVD approximately 10 years earlier than those with emmetropia or hyperopia.
- Key is to rule out potentially sight threatening condition for the floaters, ie retinal tear.
- Patients with retinal condition such as lattice degeneration and myopes need to be educated about S&S of RD (flashes and floaters)
 - 8-11% population has lattice
 - Risk of RD with lattice is <1%
 - 30-50% of patients with a RD have lattice

Flashes and Floaters: PVD

- A hemorrhagic PVD (i.e. vitreous hemorrhage secondary to a PVD) can occur in about 7.5% of PVDs.
- This occurs when a retinal blood vessel is torn during vitreous separation.
- The risk of having an underlying retinal tear increases to nearly 70% in the case of a hemorrhagic PVD.
- Symptoms of a hemorrhagic PVD may include a more significant decrease in vision secondary to the blood dispersed throughout the vitreous cavity.



Flashes and Floaters: Management

- A patient who presents with a sudden onset PVD without retinal breaks or hemorrhage requires repeat peripheral examination in six weeks, as the risk of retinal complications is highest within the six weeks following vitreous detachment.
- If no retinal breaks are seen at that point, routine yearly examination is all that is needed