Legends of the Posterior Segment

Blair Lonsberry, MS, OD, MEd., FAAO
Professor of Optometry
Pacific University College of Optometry
blonsberry@pacificu.edu
Disclosures

**Paid consultant for:**
Alcon: Honoraria, Speakers Bureau
Carl Zeiss Meditec: Honoraria, Speakers Bureau
Eyevance: Honoraria, Speakers Bureau
Optovue: Honoraria, Speakers Bureau
Shire: Honoraria, Advisory Board, Speakers Bureau
Sun Pharmaceuticals: Honoraria, Advisory Board
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)
Which of the following OCT’s goes with this patient?
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
CSR versus RD
Central Serous Retinopathy

- 80% to 90% of cases resolve spontaneously within 3 months
- Treatment options:
  - include laser photocoagulation,
  - "safety-enhanced" PDT,
  - 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.
Arturo: 50 y/o Russian Male

- RK 1991 -> 20/20 with hyperopic correction:
  - +5.50 -1.50X090
- TA: 32/18
- Pach
  - 544 μ
  - 558 μ
- Gonio –CBB
- - PMHx
- - meds
1 Mo Later

- TA: 24,25 RE; 18 LE
  - (Initial IOP 32/18)

- How do you account for the difference?
- Illustrates the importance of establishing a baseline
Case Courtesy of Dr. Mark Dunbar
Fixation Monitor: Gaze/Blind Spot
Fixation Target: Central
Fixation Losses: 0/14
False POS Errors: 3%
False NEG Errors: 0%
Test Duration: 05:38
Fovea: 35 dB

Case Courtesy of Dr. Mark Dunbar
The Future of Glaucoma Diagnosis and Management???
OCT En Face

RPC

RPC Vessel density

RNFL Thickness

Images and data courtesy of Robert Weinreb, MD and Linda Zangwill, PhD, UC San Diego
Moderate Glaucoma

**OCT En Face**  
**RPC**  
**RPC Vessel density**  
**RNFL Thickness**

Images and data courtesy of Robert Weinreb, MD and Linda Zangwill, PhD, UC San Diego
Advanced Glaucoma

OCT En Face  RPC  RPC Vessel density  RNFL Thickness

Images and data courtesy of Robert Weinreb, MD and Linda Zangwill, PhD, UC San Diego
QUICKIE
CHRPE vs Nevus
CHRPE vs Hamartomas

SURE 4. Retinal pigment epithelial hamartomas (pigmented ocular fundus lesions) associated with familial adenomatous polyposis Gardner 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
From: Enhanced Depth Imaging Optical Coherence Tomography of Small Choroidal Melanoma: Comparison With Choroidal Nevus
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.

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 underway. To date, 22 patients have been treated, and there have been no serious adverse events. All patients who have been followed for 6 months or longer have had a final vision at last follow-up within one letter of their baseline
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

- Microvascular damage
- Ischemia
- Increased permeability
- Leakage
- Macular edema
- Neovascularization
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
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)

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

Month 6 Primary Endpoint
Mean Change from Baseline BCVA

BRVO

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 approx 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)

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

PRN Lucentis available for all patients: 6M tx period

- 0.5 mg
- Ranibizumab 0.3 mg
- Ranibizumab 0.5 mg

12M trial

CRVO

Month 6 Primary Endpoint
Mean Change from Baseline BCVA

CRVO

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

Sham/0.5 mg (n=130)

0.3 mg Ranibizumab (n=132)

0.5 mg Ranibizumab (n=130)
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 acetozolamide (500 mg IV or po),
  – topical ocular hypertensive drops (lupidine, B-blocker),
  – anterior chamber paracentesis,
  – consider admission to hospital for carbogen Tx (high carbon dioxide)
13 YR Female

CC: noticed that her left eye became blurry and objects were “wavy” a couple of days ago. Sudden onset and she had experienced a headache over the left eye just prior to the vision going blurry.

Ocular Hx: she currently wear glasses for distance

Medical Hx: she is currently not diagnosed with any health problems and is not taking any medications
Entrance Skills

VA with current Rx: 20/30 OD and 20/30 OS
Entrance skills unremarkable
Amsler: metamorphopsia OS
BCVA: 20/20 OD with increased minus, no improvement possible in the left eye
IOP’s: 13 mm Hg OD and OS
Fundus Photos
OCT
Retina Consult

• Referred patient to retina and they confirmed the diagnosis of VKH.
• She was begun on oral prednisone 60 mg per day and she was re-evaluated in 1 week.
• At the follow up, there was reduction in her serous retinopathy and vision was improved.
From the Experts

• Vogt-Koyanagi-Harada (VKH) disease is a multisystemic disorder characterized by granulomatous panuveitis with exudative retinal detachments that is often associated with neurologic and cutaneous manifestations.

• VKH disease occurs more commonly in patients with a genetic predisposition to the disease, including those from Asian, Middle Eastern, Hispanic, and Native American populations.
From the Experts

• VKH:
  – Patients have no prior history of ocular trauma or surgery
  – Patients have no evidence of another ocular disease based on clinical or laboratory evidence
  – Patients have bilateral ocular involvement.
From the Experts

• VKH:
  – The neurologic and auditory signs include the following:
    • Malaise, fever, headache, nausea, abdominal pain, stiffness of the neck and back, or a combination of these factors; headache alone is not sufficient to meet the definition of meningitis
    • Tinnitus
    • Cerebrospinal fluid pleocytosis
  – Integumentary signs include the following:
    • Alopecia: loss of body hair
    • Poliosis: loss of pigment in hair
    • Vitiligo: loss of skin pigmentation in blotchy pattern
VKH Treatment

• For most patients with bilateral serous detachments and severe visual loss, begin therapy with systemic prednisone (1-2 mg/kg/day).

• The length of treatment and subsequent taper must be individualized for each patient.
  – Most patients require therapy for 6 months and occasionally up to 1 year before successful tapering of systemic corticosteroids.
  – Systemic therapy should not be discontinued during the 3 months following the onset of the disease because of the risk for recurrence.
What does this look like???
Patient 1 year later: resolved with PVD
OS 1 year after the OD resolution
Macular hole

- Unilateral, decreased vision
  - Often in 60-80 year old women
  - Anyone w/ a history of trauma
- Symptoms:
  - Decreased vision, metamorphopsia
    - 20/200 for full thickness holes
- Signs:
  - Red hole in the macula
  - (+) Watzke-Allen sign
Macular hole

• Stages
  – Stage 1a -> impending hole. Normal foveal depression with yellow spot/dot in fovea.
  – Stage 1b -> Abnormal foveal depression with yellow ring.
Macular hole

- Stages
  - Stage 2 -> Small full-thickness hole. 20/80 - 20/400.
  - Stage 3 -> Full-thickness hole w/ cuff of SRF. No PVD
  - Stage 4 -> Full-thickness hole with cuff of SRF, with complete PVD.
Macular hole

• Stages
  – Stage 2 -> Small full-thickness hole. 20/80 - 20/400.
  – Stage 3 -> Full-thickness hole w/ cuff of SRF. No PVD
  – Stage 4 -> Full-thickness hole with cuff of SRF, with complete PVD.
# New Macular Hole Staging

Table 2. Correlation between Commonly Used Clinical Macular Hole Stages and the International Vitreomacular Traction Study Classification System for Vitreomacular Adhesion, Traction, and Macular Hole

<table>
<thead>
<tr>
<th>Full-Thickness Macular Hole Stages in Common Use</th>
<th>International Vitreomacular Traction Study Classification System</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 0</td>
<td>VMA</td>
</tr>
<tr>
<td>Stage 1: impending macular hole</td>
<td>VMT</td>
</tr>
<tr>
<td>Stage 2: small hole</td>
<td>Small or medium FTMH with VMT</td>
</tr>
<tr>
<td>Stage 3: large hole</td>
<td>Medium or large FTMH with VMT</td>
</tr>
<tr>
<td>Stage 4: FTMH with PVD</td>
<td>Small, medium, or large FTMH without VMT</td>
</tr>
</tbody>
</table>
# New Macular Hole Staging

## Table 2. Correlation between Commonly Used Clinical Macular Hole Stages and the International Vitreomacular Traction Study Classification System for Vitreomacular Adhesion, Traction, and Macular Hole

<table>
<thead>
<tr>
<th>Full-Thickness Macular Hole Stages in Common Use</th>
<th>International Vitreomacular Traction Study Classification System</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 0</td>
<td>VMA</td>
</tr>
<tr>
<td>Stage 1: impending macular hole</td>
<td>VMT</td>
</tr>
<tr>
<td>Stage 2: small hole</td>
<td>Small or medium FTMH with VMT</td>
</tr>
<tr>
<td>Stage 3: large hole</td>
<td>Medium or large FTMH with VMT</td>
</tr>
<tr>
<td>Stage 4: FTMH with PVD</td>
<td>Small, medium, or large FTMH without VMT</td>
</tr>
</tbody>
</table>
New Macular Hole Staging

Table 2. Correlation between Commonly Used Clinical Macular Hole Stages and the International Vitreomacular Traction Study Classification System for Vitreomacular Adhesion, Traction, and Macular Hole

<table>
<thead>
<tr>
<th>Full-Thickness Macular Hole Stages in Common Use</th>
<th>International Vitreomacular Traction Study Classification System</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 0</td>
<td>VMA</td>
</tr>
<tr>
<td>Stage 1: impending macular hole</td>
<td>VMT</td>
</tr>
<tr>
<td>Stage 2: small hole</td>
<td>Small or medium FTMH with VMT</td>
</tr>
<tr>
<td>Stage 3: large hole</td>
<td>Medium or large FTMH with VMT</td>
</tr>
<tr>
<td>Stage 4: FTMH with PVD</td>
<td>Small, medium, or large FTMH without VMT</td>
</tr>
</tbody>
</table>

Small FTMH w/o traction

154 microns

237 microns
# New Macular Hole Staging

**Table 2. Correlation between Commonly Used Clinical Macular Hole Stages and the International Vitreomacular Traction Study Classification System for Vitreomacular Adhesion, Traction, and Macular Hole**

<table>
<thead>
<tr>
<th>Full-Thickness Macular Hole Stages in Common Use</th>
<th>International Vitreomacular Traction Study Classification System</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 0</td>
<td>VMA</td>
</tr>
<tr>
<td>Stage 1: impending macular hole</td>
<td>VMT</td>
</tr>
<tr>
<td>Stage 2: small hole</td>
<td>Small or medium FTMH with VMT</td>
</tr>
<tr>
<td>Stage 3: large hole</td>
<td>Medium or large FTMH with VMT</td>
</tr>
<tr>
<td>Stage 4: FTMH with PVD</td>
<td>Small, medium, or large FTMH without VMT</td>
</tr>
</tbody>
</table>

**Medium FTMH w/o traction**

250-400 microns
New Macular Hole Staging

Table 2. Correlation between Commonly Used Clinical Macular Hole Stages and the International Vitreomacular Traction Study Classification System for Vitreomacular Adhesion, Traction, and Macular Hole

<table>
<thead>
<tr>
<th>Full-Thickness Macular Hole Stages in Common Use</th>
<th>International Vitreomacular Traction Study Classification System</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 0</td>
<td>VMA</td>
</tr>
<tr>
<td>Stage 1: impending macular hole</td>
<td>VMT</td>
</tr>
<tr>
<td>Stage 2: small hole</td>
<td>Small or medium FTMH with VMT</td>
</tr>
<tr>
<td>Stage 3: large hole</td>
<td>Medium or large FTMH with VMT</td>
</tr>
<tr>
<td>Stage 4: FTMH with PVD</td>
<td>Small, medium, or large FTMH without VMT</td>
</tr>
</tbody>
</table>

Large FTMH with traction

> 400 microns