**Tales from the Trenches: Posterior Segment**
Mark T. Dunbar, OD, FAAO
Bascom Palmer Eye Institute
University of Miami, Miller School of Medicine
Miami, FL 33136
mdunbar@med.miami.edu

---

**Mark Dunbar: Disclosure**
- Consultant for Allergan Pharmn
- Optometry Advisory Board for:
  - Allergan
  - Carl Zeiss Meditec
  - Artic Dx
  - Alcon Nutrition

Mark Dunbar does not own stock in any of the above companies

---

**Leonardo**
57 y/o Hispanic Male
- “Routine” exam
- Has had poor vision for ~ 25 yrs or so
- VA: 20/70 RE; 20/60 LE
- CVF: FTFC OU
- Pupils: ERLL – No APD
- SLE – Tr NS

---

**Thoughts…?**
Differential Diagnosis?
28 y/o Hispanic Female
33 Wks Pregnant: 3 Wk Hx of ↓ VA

4/27/2009

28 y/o Hispanic Female
33 Wks Pregnant: 3 Wk Hx of ↓ VA

Seen 2 Wks Later with resolution of her serous detachments

35 y/o HIV (+) Male
CD4 ~ 400

Patient first presented to BPEI in 3/99
Normal findings noted

28 y/o Hispanic Female
33 Wks Pregnant: 3 Wk Hx of ↓ VA

8/29/2005
Idiopathic Central Serous Chorioretinopathy (ICSC)
- Condition of unknown etiology
- Localized detachment of sensory retina
- Anxious males: 3:1 to 10:1
  - Incidence doubles in women 30-40 vs 20-30 y/o
- History of emotional stress
- “Type A personality”
- Common Caucasians, Hispanics, Asians

Central Serous Chorioretinopathy (CSC)
- Condition 1st described by Albrecht von Graefe 1866
- Relapsing central leutic retinitis
- Several terms commonly used today
  - Idiopathic central serous chorioretinopathy
  - Central serous chorioretinopathy (CSC)
  - Central serous retinopathy (CSR)
**Age of Onset**
- Ages 30-55 y/o
- Europe 40-60 y/o

**Corticosteroids and CSC**
- Strong relationship with increased cortisol levels
- Steroid users
- Organ transplant
- Medical conditions requiring steroid: SLE, RA
- Pregnant women
- Increased levels of free circulating endogenous cortisol

**46 y/o White Male**
- Presented with blurred vision LE X 1 mo
- Also notes a floater in the same eye
- Va: 20/20 RE; 20/25 LE

Fig 4
ICSC

- Detachment of sensory retina
  Due to leakage from small underlying PED
- Absence of foveal reflex
- Yellow dot in center of fovea: Xanthophil
- Vision rarely less than 20/25
- Patients often report micropsia
Fibrin with Atypical ICSC
- Some patients with ICSC can exude more of a fibrin response
- Recent studies suggest the fibrin may actually be fragments of the photoreceptor outer segments
  - They accumulate when the normal process of phagocytosis of the photoreceptor outer segments become disrupted due to the serous detachment of the retina

Central Serous Chorioretinopathy (CSC)
2 Main Types
- Common classic CSC
- More widespread alteration of the RPE with chronic shallow SRF
  - Chronic CSC
  - May be associated with chronic corticosteroid use

Pathophysiology of CSC
- Normal physiology
  - There is a balance in the tissue osmotic and hydrostatic pressures which results in fluid flow from the retina toward the choroid
- Abnormal choroidal vascular hyperpermeability
  - Excessive tissue hydrostatic pressure in choroid leads to mechanical disruption of the RPE barrier -> damage to the RPE Cells -> egress of fluid under the retina

CSC: Treatment
- Argon laser directly to site of leakage (PED) if detachment persists > 6 months
- Do NOT use oral steroids
- PDT for chronic CSC

Who can tell me what “Sclopetaria” is?
- Closed globe injury that results from high velocity object bumping, but not perforating the sclera
- Full-thickness defect in Choroid, Bruch’s membrane, and Retina, but Intact Sclera.
- Tissue replaced with dense fibrous connective tissue.

Chorioretinitis Sclopetaria

- Closed globe injury that results from high velocity “missile energy” bumping, but not perforating the sclera.
- Full-thickness defect in Choroid, Bruch’s membrane, and Retina, but **Intact Sclera**.
- Tissue replaced with dense fibrous connective tissue. 


BB Gun Related Injuries

- Nearly 30,000 Americans present to ERs with BB- and pellet gun-related injuries each year.
- Most incidents are unintentional and typically occur in young males.
- CDC Surveillance report of 47,000 BB gun-related injuries between 1992-1995:
  - 50% of injuries occurred in children between 10 and 14 years of age
  - 2,839 (6%) of patients suffered direct eye trauma.

Commotio Retinae

- Whitening of outer retinal layers
- Shock waves traversing the eye
- Cherry red spot and decreased vision in Berlin’s edema
- Good prognosis

Traumatic Macular Hole

- Knapp first described in 1869
- Now known to be less than 10% of full thickness macular holes
- 6% of pts suffering contusive injuries develop full thickness macular hole
- Develops from concussive forces in a countercoup manner

Traumatic Macular Hole

- Intact hyaloid, mechanism likely traction / ILM rigidity
  - Outward extension of the equator causes a flattening of the retina and tangential traction
- Hydration theory: dehiscence of the ILM disrupts hemostasis and causes intraretinal swelling -> leads to macular hole formation
Idiopathic Macular Holes

- VA 20/400 to 20/60
- 1/3 DD full thickness round hole
- Surrounding cuff of fluid
- Yellow deposits in the base of the hole
- Translucent operculum (anterior) 50%
- May have associated ERM (10-20%)

Idiopathic Macular Holes

Pathogenesis

- Anterior-posterior vitreous traction
- 1989 Gass/Johnson:
  Tangential traction due to shrinkage and contraction of the prefoveal vitreous cortex

Stages of Macular Holes

- IA: Yellow spot or ring in macula
- IB: Loss of foveal depression
- II: Partial tear in the sensory retina
- III: Fully developed full thick mac hole
- IV: Macular hole with posterior vitreous separation

Vitreous Surgery for Macular Holes

  - 52 patients
  - PPV/Removal vitreous cort, Fld/Gass exchange
  - 58% anatomic success, 73% visual success
  - Overall 42% success rate
- Kelly, Wendel: Ophth Nov 1993
  - 170 patients
Macular Hole Surgery

- Patel/Wendel Sem Ophthalmol 1994, 152 pts
  - Macular hole < 3m duration 80% success
  - Macular hole > 3 mo – 2 yrs 74% success
  - Macular hole > 2 yrs duration 61% success

Macular Holes: Loss of Vision

- Loss of neurosensory retinal tissue
- Rim of subretinal fluid around the hole (microdetachment)

Macular Hole Surgery

Postoperative Period
- Face down for 2 weeks
- Has evolved to face down for 1 wk
- Silicone Oil sometimes for patients who need to travel on planes or over mountains

Intravitreal Gas Tamponade

Why Face Down Positioning (FDP)

- The mechanism by which the tamponade agent facilitates macular hole sealing is uncertain
- Two possible effects are:
  - Mechanically tamponade the macula
  - Isolate the healing macula from vitreous fluid
- Theory: provide a template over which the nascent bridging preretinal membrane can form

57 y/o Hispanic Female
Decreased VA LE X 2 Mo

Retina, April 2009
Vitreomacular Traction in the Era of OCT
- Not rare!
- A group of disorders caused by incomplete PVD
- Leads to persistent traction on the macula
- Produces in most cases CME and decreased visual acuity
- Can be idiopathic
- Can occur with ERM and macular hole

Differential diagnosis of Full Thickness Macular Hole
- ERM with pseudo hole
- Lamellar hole
- Solar maculopathy

Next Case
25 y/o AA Female
Type I DM

28 yo Jeweler

- Referred by another jeweler who is friends with my wife
- Blurred vision RE > LE X 1 mo
  - Also red eyes OU
- PHx: RK done 10 yrs ago
  - Saw the RK Dr 1-2 mo ago – told “dry eyes”, quite smoking!
- Reports to be in good health

28 yo Jeweler

- VA: 20/40 RE; 20/20 LE
- CVF: FTFC OU
- Pupils – Equally reactive, NO APD
- No preauricular adenopathy
- Diffuse injection OU

28 yo Jeweler

- Anterior Segment
  - RK Scars OU
  - AC: 1 + C/F RE; 3 + C/F LE
  - Iris:
    - RE: organized fibrin membrane around the pupil – no synechia
      - Nodule inferior
    - LE: No fibrin, No nodule
  - Lens: fibrin, debris, pig ant cap R>L

28 yo Jeweler

As he is dilating -> More history

- 3 Vices
  - Alcohol – 10 scotches/night
  - Very promiscuous – loves women
  - Smokes
- 20 lb weight loss over the holidays
- Attributes this to work and not eating
28 yo Jeweler

- Panuveitis with Retinal Vasculitis
  - Periphlebitis
  - Vascular occlusions
- Iridocyclitis with iris nodules
- Moderate vision loss RE

What is the etiology?

Sarcoidosis

- Multisystem granulomatous disorder of unknown etiology characterized by intrathoracic involvement
- World wide distribution - more common in developing countries
- Multiple theories considered including infectious agents, allergies, hypersensitivity's: none conclusive

Sarcoidosis

- All races affected, blacks more in US
- Females more common 60/40
- 75% < 40, Children uncommon
- Area of active disease is Lung

Joanne: 50 y/o White Female

- Grew up in the Wisconsin
- Always “pretty highly myopic”
- Began having eye problems in early 20’s that ultimately required laser treatment
- Lost her central acuity in RE, but did “well” in the LE

Joanne: October 2008
Ocular Histoplasmosis

- Condition caused by mild or subclinical systemic infection with *Histoplasma capsulatum*
- Predominantly found in eastern half of the U.S. especially the Ohio River Valley
- 2,000,000 people who live (have lived) in endemic areas have “histo spots”
  - 100,000 will lose vision in 1 or both eyes

Ocular Histoplasmosis

- Multiple "punched out" chorioretinal scars
- Peripapillary atrophy
- Lesion involving the macula -> NVM

Ocular Histoplasmosis

- Most frequent finding assoc with CNVM:
  - Localized serous or hemorrhagic detachment of retina
  - Poorly defined, round/oval, light gray, subretinal lesion
  - Subretinal blood, or exudate
  - Bleeding/exudate occurs beneath the retina not beneath the RPE

Ocular Histoplasmosis

- Laser photocoag proven beneficial by MPS
  - Extrafoveal
    - 5 yrs: SVL 12% Tx vs 42% NonTx
    - 60-70% had VA > 20/40
  - Juxtafoveal
    - 5 yrs: SVL 12% Tx vs 28%
    - Adequately Tx eyes averaged 20/40
  - Natural Hx: 14-23% of NonTx ≥ 20/40

Macular Telangiectasis… What is it?
Retinal Telangiectasis

Term proposed by Reese to describe retinopathies characterized by dilated and incompetent vessels

Coat’s Syndrome/Disease

- Condition 1st described by George Coats in 1908
  - Massive retinal exudation with or without retinal vascular disease
  - 3 disease processes identified in his group of patients
    - AMD
    - von Hippel Lindau
    - Congenital retinal telangiectasis

Coat’s Syndrome/Disease

- 1912 Leber described condition characterized by multiple retinal aneurysms w/ little or no leakage -> “Leber’s miliary aneurysms”
- 1956 Reese linked the 2 diseases as a spectrums of the same disease process
  - Begins as telangiectasis of the retinal vessels
  - Followed by progressive exudation
  - Can lead to retinal detachment

Coat’s Syndrome/Disease

- Coats syndrome is now recognized to be a form of congenital retinal telangiectasis
- Unilaterally
- Young males
  - Can the disease occur in older patients?
  - What is the spectrum of the disease

Idiopathic Juxtafoveal Retinal Telangiectasis (JRT)

Gass JD, Blodi BA. Ophthalmology 1993

- Unknown etiology
- Telangiectatic retinal vessels, temporal to the fovea
- Associated findings:
  - Dilated capillaries
  - Minimal exudation
  - Retinal crystals
  - Right angle venules
  - Retinal pigment hyperplasia

Idiopathic Juxtafoveal Retinal Telangiectasis (JRT)

Gass JD, Blodi BA. Ophthalmology 1993

- Unilateral or bilateral
- Males or females
- Classification:
  - Type I: (A&B) Form of Coats -> unilateral
  - Type II: (A&B) Bilateral, M=F, most common, present in mid-50’s, 20/40-20/60
  - Type III: (A&B) Rare
Newer imaging technologies have helped identify some interesting differences between the two groups:

**Group 1**
- Still considered to fall within the spectrum of Coats’ disease
- More likely to have profound vascular changes, with more obvious aneurysmal dilations and prominent cystic changes within the macula

**Group 2 patients:**
- Central lamellar cyst, which the retina “drapes” over the cyst
  - Visible on OCT
  - Hallmark diagnostic sign for a group 2 patient.
- Demonstrate a loss of retinal transparency
- Smaller, subtler telangiectatic changes within the capillaries
- RPE changes not seen in group 1 patients
  - RPE changes explain why group 2 patients can develop CNV

**Clues to the Diagnosis**
- Intraretinal vascular ‘changes’
  - No subretinal or deep hemorrhage
- No other obvious retinal vascular disease
  - Crossing changes
  - No scattered retinal hemorrhages, Ma, microvascular changes elsewhere
- Fluorescein angiogram is diagnostic

**JRT = Macular Telangiectasis**

**What’s in a Name?**
- Macular telangiectasia: 2 groups
  - Group 1 - macular aneurysmal telangiectasia (MAT)
  - Group 2 - macular perifoveal telangiectasia (MPT)

**Macular Telangiectasis**

**Macular Telangiectasis**

https://web.emmes.com/study/mactel/
- Established in 2005
- Prospective 4 year survey of at least 200 patients drawn from centers in Europe, North America and Australia
- Genetic basis of disease
- Mechanisms of vision