Who can tell me what “Sclopetaria” is?

Chorioretinitis Sclopetaria
- 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

_JAMA. 1995 Jun 14;273(22):1749-54
MMWR BB- and Pellet Gun-Related Injuries

Fig 1
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 hemostatis 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

- Originally described as a “syndrome”
- Incomplete or partial PVD at the ON
- Results in traction at the macula
  - Often in a “dumbbell” shaped configuration
- Produces macular edema – CME
- Necessitates pars plana vitrectomy
- Rare

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

Differential diagnosis of Full Thickness Macular Hole

- ERM with pseudo hole
- Lamellar hole
- Solar maculopathy

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

39 y/o White Female

- Was seen 2 mo ago for routine exam
- History of high myopia
- Refracted and examined
  - Rx is ~ -10 D myope (with minimal astigmatism) each eye
- Noted to be in good ocular health
  - Fundus LE a small, depigmented area of the RPE inferior nasal to the macula
Diagnosis

- CNV – related to pathologic myopia
- Tx: Avastin injection
Not Everybody Has Been This “Lucky”

38 y/o White Female – change in vision RE (only seeing eye)
- History of high myopia
- Previous laser LE 5 yrs prior – no improvement in VA
- Noted blur and metamorphopsia RE X 1 wk

High Myopia
- 3th most common disease assoc w CNV
- Myopic degenerative changes seen in 25-33% of all high myopes
  - CNV 5-10% -> 60-75% subfoveal
- Laser limited success due to thinned, fragile nature of Bruch's membrane
  - Laser Tx, scar spreads >> usually larger than what the natural course of the disease
CNV & Degenerative Myopia

- Typically Type II – grow under the retina
- Up to 74% subfoveal
- < Incidence w/ ↑ post staphyloma
  - Suggests may need preserved choriocapillaris for CNV to develop

Myopic Degeneration

- Peripapillary scleral crescent
- Tilted optic nerve
- Thinning of the sensory retina/RPE
- Staphyloma
- Fuch’s spot
- Lacquer cracks
- CNV
- Peripheral retinal degeneration
- Retinal Detachment

33 yo Hispanic Female
Blurred VA LE 20/40

- Do you see any gray/green changes that could be consistent with CNV?
- Is there fluid in the macula?
  - Subretinal hemorrhage or exudate?
- Is the macula flat or is there any elevation?

4 mo later

Next Case
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

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

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?

28 yo Jeweler

Differential Diagnosis

- HIV related
  - CMV
- Syphilis
- Sarcoid
- Behcets
- Eal’s
- SLE

27 yo Jeweler

Work up

- HIV
- CBC with Diff
- ANA
- Chest X-ray
- ACE
- CRP
- FTA-ABS
- RPR
- PPD
- Anti-cytoplasmic antibodies
  - pANCA
  - cANCA
- Cardiolipin
- Hepatitis B panel
- Hepatitis C
- Comprehensive metabolic panel

27 yo Jeweler

Working Diagnosis

- Sarcoid

Treatment

- PF q 1h
- Hyosine hs
- RTC 10 days

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
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

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
OCT

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

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

1912 Leber described condition characterized by multiple retinal aneurysms w/ little or no leakage -> “Leber’s miliary aneurysms”

Leber T. Albrecht von Graefe’s Arch Klin Ophthalmol 1912

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

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
**Macular Telangiectasis**

- 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

---

**The Relationship between Coats and Type I JRT**

- 142 (99%) of 150 consecutive eyes with Coat’s Disease had massive retinal exudation involved midperiphery and or peripheral fundus
- 2 (1%) eyes had telangiectasis restricted to the macula area
- Typically in younger males

---

**JRT = Macular Telangiectasis**

**What’s in a Name?**

- Macular telangiectasia: 2 groups
- Group 1 - macular aneurysmal telangiectasia (MAT)
- Group 2 - macular perifoveal telangiectasia (MPT)

---

**The Relationship between Coats and Type I JRT**

- 53 eyes in 50 patients
- 12 eyes had massive retinal exudation and telangiectasis involving the peripheral fundus
- 22 eyes had smaller, focal areas of peripheral telangiectasis
- 12 eyes had juxtafoveal involvement
The Relationship between Coats and Type I JRT


“Idiopathic retinal telangiectasis with exudation is a spectrum of disease, which is synonymous with Coats’ disease”