

# **IDIOPATHIC CENTRAL SEROUS CHORIORETINOPATHY**

Revised 2002

## **INCIDENCE**

- 30- 50 YEAR OLD (SELDOM IN PATIENTS < 30 OR GREATER THAN 55 YEARS)
- MEN 90%
- RACE - NO PARTICULAR PREDILECTION, RARE IN BLACKS

## **PAST HISTORY**

- NO SYSTEMIC ABNORMALITIES ( POSSIBLY HIGHER INCIDENCE OF HYPERTENSION)
- NO HEREDITARY ASSOCIATION
- TYPE A PERSONALITY

## **SYMPTOMS**

- METAMORPHOPSIA MICROPSIA SCOTOMA
- DECREASED VISION
- DECREASED COLOR
- HYPEROPIC SHIFT
- ABNORMALPHOTOSTRESSTEST

## **SIGNS**

- SEROUS DETACHMENT OF MACULA
- SMALL RPE DETACHMENT
- SUBRETINITIC DEPOSITS- WHITE OR GRAY
- PIGMENT EPITHELIAL CHANGE IN BOTH EYES

## **FLUORESCEIN ANGIOGRAPHY**

- PINPOINT LEAK - SINGLE OR MULTIPLE DOT OF HYPERFLUORESCENCE 80%
  - SMOKESTACK 20%
- RPE WINDOW DEFECTS
- GUTTERS

## **PATHOGENESIS**

UNKNOWN

## DIFFERENTIAL DIAGNOSIS

- RETINAL EPITHELIAL DETACHMENT
- VKH SYNDROME
- MALIGNANT HYPERTENSION
- TOXEMIA OF PREGNANCY OCCULTSRN
- CHOROIDAL NEVI, TUMORS
- OPTIC PITS
- RESOLVING CSR NO LEAK SEEN ON FA

## NATURAL COURSE

- 95% SPONTANEOUS RESOLUTION 2-4 MONTHS
- 30- 35 % RECURRENCE
- 5% DEVELOP SRN

## TREATMENT

- OBSERVATION AND REASSURANCE
- STEROIDS OF NO VALUE
- LASER
  - SPEEDS RESORPTION OF SRF
  - IF LEAK PRESENT > 4 MONTHS
  - VISUALLY IMPAIRED
  - NO EVIDENCE THAT LASER IMPROVES LONG- TERM VISUAL ACUITY
  - DOES NOT PREVENT RECCURRENCES

## AGE-RELATED MACULAR DEGENERATION

- PROGRESSIVE, DEGENERATIVE DISEASE OF THE RPE, BRUCH'S MEMBRANE, AND CHORIOCAPILLARIS
- 165,000 NEW CASES EACH YEAR IN USA
- LEADING CAUSE OF LEGAL BLINDNESS OVER AGE 65
- 10 MILLION PEOPLE HAVE SOME FORM OF THE DISEASE

## AGE-RELATED MACULAR DEGENERATION

- DRUSEN
- DIRTY GRAY SUBRETINAL MEMBRANE
  - NEUROSENSORY RETINAL DETACHMENT
- SUBRETINAL HEMORRHAGE
- HARD LIPID EXUDATE
- CYSTOID EDEMA OF RETINA
- RPE ATROPHY AND HYPERPLASIA
- RPE RIPS
- FIBROVASCULAR DISCIFORM SCARRING

- VITREOUS HEMORRHAGE

## DRUSEN

HARD, CONFLUENT, SOFT, CALCIFIED, CUTICULAR, BASAL LAMINAR

## FLUORESCEIN ANGIOGRAPHY IN ARMD

- DRUSEN-HYPER OR HYPOFLUORESCENCE
- EARLY FA- LACY, IRREGULAR, NODULAR, HYPERFLUORESCENCE
- LATE FA- DIFFUSE LEAKAGE OF DYE
- HYPOFLUORESCENT BORDER SURROUNDING SRN

## AGE-RELATED MACULAR DEGENERATION

- NON EXUDATIVE (DRY) 90%
- EXUDATIVE (WET) 10%

## AGE RELATED MACULAR DEGENERATION SEVERE VISUAL LOSS (20/200 OR WORSE)

- NON EXUDATIVE (DRY) 12%
- EXUDATIVE (WET) 88%

## GEOGRAPHIC ATROPHY

CENTRAL AREOLAR PIGMENT EPITHELIAL ATROPHY LOSS OF RPE, OUTER  
RETINA, CHORIOCAPILLARIS

## ARMD RISK FACTORS

- INCREASING AGE
- HYPEROPIA
- LIGHT IRIS COLOR
- POSITIVE FAMILY HISTORY
- CIGARETTE SMOKING

## CONDITIONS MASQUERADING AS SRN

- CSR
- TRAUMA
- AMPPPE
- BEST
- CUTICULAR DRUSEN
- INFLAMMATION

- RPEDETACHMENT
- DRUSEN
- ARTERIAL MACROANEURYSM
- SERPIGINOUS CHOROIDITIS

#### MACULAR PHOTOCOAGULATION STUDY ARMD

- EFFICACY OF ARGON BLUE GREEN LASER IN TREATMENT OF SRN
- ANGIOGRAPHIC EVIDENCE OF SRN AT LEAST 200 MICRONS FROM CENTER OF FAZ
- POST TREATMENT RECURRENCE RATE 59%

#### MACULAR PHOTOCOAGULATION STUDY ARMD

SEVERE VISION LOSS ( LOSS OF > 6 LINES OF VISION)

- NO TREATMENT 60%
- TREATMENT 25%

#### AGE-RELATED MACULAR DEGENERATION

- RISK OF SRN IN FELLOW OF PATIENT WITH SRN IN ONE EYE IS 10-12% A YEAR

#### MPS KRYPTON RED LASER STUDY

- SRN WITHIN FAZ
- JUXTAFOVEAL 62% VISION REMAINS STABLE OR IMPROVES
- SUBFOVEAL 28% VISION REMAINS STABLE OR IMPROVES

#### LASER WAVELENGTH

- ARGON BLUE-GREEN 488 & 514
- ARGON GREEN 514
- KRYPTON 647
- DYE YELLOW 577
- DYE RED 630

#### ADVANTAGES OF KRYPTON

- NOT ABSORBED BY LENS AND CORNEA
- NOT SCATTERED OR ABSORBED BY LENS OR VITREOUS OPACITIES. E.G. CATARACT, HEME NOT ABSORBED BY XANTHOPHYLL
- DEEPER BURN, OUTER RETINAL LAYERS, CHOROID, RPE

PROBLEMS WITH KRYPTON

- NOT ABSORBED BY FRESH BLOOD
- NOT ABSORBED WELL BY HYPOPIGMENTED EYES

PRESUMED OCULAR HISTOPLASMOSIS SYNDROME (POHS)

1. PRESUMED OHS BECAUSE MUCH OF THE EVIDENCE THAT HISTOPLASMA CAUSES THE SYNDROME IS CIRCUMSTANTIAL. SEVERAL CASES WITH PROBABLE HISTOPLASMA CAPSULATUM IN EYES WITH SRN HAVE BEEN REPORTED
2. FUNGUS THAT IS ENDEMIC TO OHIO/MISSISSIPPI RIVER VALLEYS, EASTERN AND MIDWESTERN USA
3. 90% OF PATIENTS HAVE POSITIVE SKIN REACTION TO HISTOPLASMIN
4. OCULAR TRIAD
  - PERIPAPILLARY CHORIORETINAL SCARRING
  - PERIPHERAL ATROPHIC SCARS- HISTO SPOTS MACULAR SRN (NO VITIRITIS)
5. DISSEMINATED CHORIODITIS SIMILAR TO POHS ASSOCIATED WITH EPSTEIN- BARR VIRUS
6. VISUAL LOSS DUE TO SRN
7. MACULAR PHOTOCOAGULATION STUDY
  - EFFICACY OF LASER TX EVALUATED
  - ARGON LASER EFFECTIVE IN PREVENTING SEVERE VISION LOSS WHEN SRN 200- 2500
  - MICRONS FROM FAZ

**MPS POH SYNDROME**

	<b>Loss &gt; 6 lines</b>		<b>&gt; 20 / 200</b>
	<i>18 mos</i>	<i>36 mos</i>	<i>36 mos</i>
Treatment	9%	10%	95%
Control	34%	45%	61%

\*\*\*\*\*RECURRENCE RATE 30%

## **DIABETIC RETINOPATHY STATISTICS**

1. Second leading cause of new blindness in USA
2. Leading cause in 20-64 year age group
3. Diabetic population 25% some form of retinopathy
4. Diabetic population 5% PDR
5. 7 years DM 50% diabetic retinopathy
6. 17-25 years 90% some degree of retinopathy
7. PDR 26% after 26-50 years of DM

## **PHYSIOLOGIC ABNORMALITIES**

- Impaired autoregulation of retinal vessels
- Abnormal retinal blood flow
- Breakdown blood-retinal barrier

## **MECHANISMS OF DAMAGE**

- Thickened basement membranes
- Sorbitol toxicity
- Erythrocyte abnormalities
  1. Hemoglobin A1C reflects glucose control over 2-3 month period
  2. Rigidity of RBC, sticky RBC
  3. Increased platelet adhesion, increased factor VIII

## **EARLIEST SEQUELAE OF DR**

- Breakdown of blood-retinal barrier vitreous fluorophotometry
- Thickened basement membrane
- Pericyte dropout

## **CLASSIFICATION OF DIABETIC RETINOPATHY**

- BACKGROUND (BDR OR NPDR)
- PREPROLIFERATIVE DR (PPDR)
- PROLIFERATIVE DR (PDR)
- DIABETIC MACULOPATHY

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## **BACKGROUND DIABETIC RETINOPATHY**

- Intraretinal changes
- Microaneurysms
- Dot and blot hemorrhages
- Hard exudates
- Macular edema

## **PREPROLIFERATIVE DIABETIC RETINOPATHY**

- Cotton wool spots
- Venous beading and Loops
- IRMA
- Extensive retinal hemorrhages/microaneurysms
- Capillary occlusions ( Capillary nonperfusion)

\*\* 50% progress to PDR 12-24 months

\*\* Severe PPDR presence of at 3 of these lesions in at least two quadrants

## **PROLIFERATIVE DIABETIC RETINOPATHY**

- New vessel formation- NVD, NVE
- Fibrovascular proliferation
- Vitreous hemorrhage
- Traction retinal detachment
- Rubeosis irides

## **DIABETIC MACULOPATHY**

- FOCAL
1. DIFFUSE
  2. ISCHEMIC
  3. COMBINATION

## **EARLY TREATMENT DIABETIC RETINOPATHY STUDY (ETDRS)**

1. Is photocoagulation effective in the treatment of diabetic macular edema?
2. When in the course of diabetic retinopathy is it most effective to initiate PRP?
3. Is aspirin treatment effective in altering the course of diabetic retinopathy?

## **ETDRS TREATMENT CRITERIA MACULAR EDEMA**

### **CLINICALLY SIGNIFICANT EDEMA**

1. Thickening of the retina at or within 500 microns of the center of the fovea
2. Hard exudates at or within 500 microns of the center of the fovea with retinal edema
3. Zone of retinal thickening 1 disc diameter or larger, any part of which is within 1 disc diameter of the center of the fovea

## **ETDRS TREATMENT FOR MACULAR EDEMA**

Focal and/or Grid treatment

- **Focal** - applied to microaneurysms
  - 50-200 micron
  - 0.05-0.1 sec. duration
  - whiten or darken aneurysm
- **Grid**- thickened retina showing diffuse leakage or CNP on fluorescein
- 50-200 micron
- 0.1 exposure

## RESULTS ETDRS

	TREATMENT	NON-TREATMENT
<b>VISUAL LOSS</b>	<b>12%</b>	<b>24%</b>
Doubling visual angle e.g. 20/40-20/80		

## ETDRS INDICATIONS FOR TREATMENT

1. Treatment stabilizes visual acuity
2. Treatment in patients with clinically significant macular and good vision appears to be an appropriate choice

## DIABETIC MACULOPATHY POOR PROGNOSIS

- PERIFOVEAL CAPILLARY NONPERFUSION
- CYSTOID MACULAR EDEMA
- HARD EXUDATES IN FOVEOLA
- VISUAL ACUITY 20/200 OR LESS

## DIABETIC RETINOPATHY STUDY

1. Laser reduced risk of severe visual loss by 50%
2. Risk factors assoc with SVL
  - vitreous or preretinal hemorrhage
  - neovascularization
  - NV on or 1 DD from optic nerve
  - severity of new vessels
3. Incidence of SVL directly related to number of risk factors
4. Eyes with 3 or more risk factors are at high risk for developing SVL

## DIABETIC RETINOPATHY STUDY

Demonstrated overall reduction of SVL (< 5/200) from 15.9% in untreated eyes to 6.4% in treated eyes, a reduction of 60%

## DIABETIC RETINOPATHY STUDY

	SEVERE VISUAL LOSS	
	TREATMENT	NO TREATMENT
NVD < 1/2 DD WITH VIT HEME	4.3%	25.6%
NVD > 1/2 DD WITHOUT VIT HEME	8.5%	26.2%
NVD > 1/2 DD WITH VIT HEME	20.1%	36.9%
NVE < 1/2 DD WITH VIT HEME	7.2%	29.7%

## **PAN RETINAL PHOTOCOAGULATION**

- Decreases quantity of vasoproliferative factor
- Improves the diffusion of oxygen from the choroids
- Destroying sick retina allows available oxygen to be used more efficiently

## **COMPLICATIONS OF LASER TREATMENT**

- Decreased peripheral vision
- Decreased night vision
- Color vision
- Loss of 1-2 lines visual acuity
- Macular edema
- Temporary loss of accommodation
- Narrow angle glaucoma
- Choroidals
- Foveal burn
- Retinal hole

## **INDICATION FOR VITRECTOMY**

- Vitreous hemorrhage
- Traction retinal detachment
- Combined TRD and Rhegmatogenous RD
- Progressive fibrovascular proliferation
- Rubeosis irides and Vitreous hemorrhage

## **VITREOUS HEMORRHAGE EARLY INTERVENTION**

Wait 6 months unless:

- Bilateral vitreous hemorrhages
- Rubeosis irides
- TRD threatening or involving macula

\*\* Must follow with serial B scans to evaluate for retinal detachment

## **CYSTOID MACULAR EDEMA**

Definition

Accumulation of extracellular (+/-) intracellular fluid in the macular region

History

Voght - 1918 - Retinitis Pigmentosa

Irvine, Nichols - 1953 – Aphakia

Gass - Norton - 1966 - Fluorescein

Causes

- Retinal vascular disease
  - Diabetic maculopathy

- Venous occlusive disease
  - Central
  - Branch
- Anomalous vascular disease
  - Leber's vascular disease
  - Localized Leber's (macular telangiectasia)
  - Coat's disease
  - von Hippel-Lindau syndrome (retinal angiomatosis)
  - Juxtapapillary hemangioma

#### Surgery

Cataract	Retinal detachment
Pseudophakia	Vitrectomy
Keratoplasty	Cryopexy, laser
Glaucoma	Keratorefractive surgery

#### Inflammation

- Intraocular surgery
- Intermediate uveitis (pars planitis)
- Uveitis (iritis)

#### Drug toxicity

- Epinephrine
- Nicotinic acid

#### Hereditary

- Dominantly inherited CME
- Retinitis pigmentosa

#### Degeneration

- Age-related macular degeneration
- Idiopathic pre-retinal macular gliosis

#### Choroid/RPE disease

- Choroidal subretinal neovascularization

#### Tumors

Choroidal hemangioma	Choroidal nevus
Choroidal malignant melanoma	Choroidal osteoma
Ocular hypotony	

#### Pathology

- Light microscopy (aphakic)
  - Accumulation of extracellular fluid in the outer plexiform layer
  - Cystoid spaces
  - Neuronal degeneration

- Retinal periphlebitis
- Electron microscopy
  - Swelling and degeneration of Mueller cells (intracellular edema)
  - Degeneration of ganglion, bipolar, and photoreceptor cells
- Experimental (post cataract extraction or - trabeculectomy Rhesus)
  - Disruption of outer blood-retinal-barrier (RPE)
  - Disruption of inner blood-retinal-barrier (ret. vessels) associated with vitreous loss
  - Serous detachment of sensory retina
  - Intra and extracellular fluid spread in sensory retina

**Pathophysiology**

- Etiology
  - Mechanical - vitreous traction
  - Inflammatory - prostaglandins leukotrienes
  - Systemic factors
  - Age
  - Light - UV release free radicals stimulate production of prostaglandins
  - Mueller cell dysfunction
- Bruch's Membrane
  - Permeability
    - Age
    - Lipid
  - RPE pump

**Incidence of aphakic/pseudophakic CME**

- Angiographic CME
- Clinical CME
- Chronic CME

Clinically significant: 3 to 5%

80% with CME have 20/40 or better vision

Fluorescein angiography: 40 to 60% at 6 wk.

**Surgical technique**

**Postop period**

	<u>4 mo.</u>	<u>8 mo.</u>	<u>16-24 mo.</u>
ICCE with IOL	17%	12%	15%
ECCE with IOL	8%	6%	4%
ICCE without IOL	14%	15%	9%

## Treatment

- Prevention
  - Meticulous surgical techniques
  - Limitation of UV and blue (440 nm) light exposure
    - Intraoperative: microscope
    - Postoperative: daylight
  - ECCE with capsulotomy delayed 6 months
  - Indomethacin: 48 hours preop plus 2 weeks postop
- Medical therapy
  - Uncertain benefit
    - Indomethacin (anti-prostaglandins)
    - Corticosteroids (anti-inflammatory)
    - Carbonic anhydrase inhibitors
- Surgical therapy
  - Laser photocoagulation - not effective
  - Anterior vitrectomy
    - Vitreous in wound
    - Chronic inflammation
    - Persistent CME

## IDIOPATHIC EPIRETINAL MEMBRANE

### History

- 1972 Wise - complete description; etiology; venous impedance
- 1975 Bellhoren, Wise, Friedman, Henkind - gliosis on electron microscopy

### Definition

- Preretinal membrane growing on the surface of the retina in an eye otherwise free of obvious predisposing factors, e.g. surgery, venous obstruction
- Historical terms epiretinal membrane
  - Idiopathic preretinal macular fibrosis
  - Surface wrinkling
  - Cellophane Silk screen
  - Macular pucker
  - Primary retinal folds
  - Silent central retinal vein occlusion
  - Shrinkage of the internal limiting membrane - produces surface wrinkling
  - Vitreoretinal interface maculopathy
- Secondary retinal fibrosis/gliosis
  - Venous obstruction
  - Diabetic retinopathy
  - Post-trauma
  - Post-surgery (esp. retinal detachment)
  - Inflammation
  - Post-photocoagulation
  - Senile macular degeneration

- Heredoretinal degeneration
- "Congenital" preretinal macular gliosis
- Etiology
  - ILM breaks (resulting from posterior vitreous detachment) - glial proliferation
  - ? Inflammation
  - ? Vascular insufficiency
  - Fibrosis – neovascularization
  - RPE – surgery

#### Incidence

- Clinically - 10% > 50 years of age
- Pathologically 2-10%
- Bilateral - 20%
- Sex - M:F = 2:3

#### Classification

- Mild
  - Glinting reflex
  - No vascular changes
- Moderate
  - Obvious membrane
  - Traction lines (striae)
  - Vascular tortuosity
  - No obscuration of retinal vessels
- Severe
  - Obscuration of retinal vessels
  - Marked vascular tortuosity
  - Heterotropia of macula

#### Associated findings

- Macular edema - disruption of normal perifoveal capillaries
  - Fluorescein leakage
    - 60% of cases with clinical edema
    - 34% of all cases with IPRMG
  - Lamellar macular holes/cysts
  - Pseudo hole
    - Posterior vitreous detachment - 66%

#### Symptomatology

- None - 40% - diagnosis incidental
- Metamorphopsia
- Decreased vision - Vision loss result of: Retinal distortion CME
- Flashes/Floaters

## Prognosis

- Initial
  - 20/30 or better = 66%
  - 20/50 or better = 84%
  - 20/100 or less = 11 %
  - 20/200 or less = 4%
  - Amsler grid distortion = 40%
- Final
  - Stable - 85%
  - Decreased
    - 14% - lamellar macular hole/cyst
    - 1 % - gliosis alone
    - Improved - spontaneous peeling
- Treatment
  - Observation
  - Photocoagulation
  - Vitrectomy and membrane peeling - 20/100 or less
  - Membranes - glial cells, fibrocytes, myofibroblasts, and RPE cells
  -

## **IDIOPATHIC MACULAR CYSTS AND HOLES**

### History

- 1871 Noyes - Ophthalmoscopic description in a 13 year old girl – trauma
- 1924 Lister - non-trauma origin ? vitreous
  - Age - 60 to 80
  - Sex - Male:Female = 1:3
  - Bilateral - 25% to 30%

### Description

- Cyst
  - Thinning of retinal structures
  - Thickening of retina
  - Amsler grid - central distortion
- Hole - size = 0.9 mm
  - No anterior edge
  - Klein's retinal tags (yellow dots in base)
  - Cuff of edema - retinal detachment
  - Amsler - central scotoma
- Fluorescein angiography
  - Normal retinal vasculature
  - Window defect – central
  - loss of xanthophylls
  - loss of pigment of RPE
  - No leakage

## Pathophysiology

- Cystoid macular edema (aphakia, diabetes, CRVO, BRVO)
- Vitreous traction
- Trauma
- Atrophy - macular degeneration
- Surgery
- Inflammation

## Risk Factors

- Systemic hypertension
- Hormonal
  - Estrogen
  - Hysterectomy
- Vitreoretinal traction from PVD

## Visual Prognosis

- Cysts 20/20 - 20/50
- Holes 20/80 - 20/200
- Cysts holes 50%
- Normal macula to cyst/hole "rare"
- Retinal detachment (non-localized) rare - pathologic myopes Trauma

## Treatment

- Control retinal edema
- ? Laser hole detachment
- Vitrectomy
- Transient macular cyst/hole
  - PVD trauma
    - Retina elevated
    - RPE window defect
    - Disappears
      - Release traction
      - RPE - pigment regenerates/reconstitutes

## **PHOTIC (SOLAR) RETINOPATHY**

### Light - Damage

- Light absorbed short period (10<sup>-9</sup> - 10<sup>-12</sup> seconds)
- Mechanical disruption cell organelles
  
- Thermal – Photocoagulative
- RPE absorbs light - 0.05 - 0.5 seconds
- Temperature  $\Delta$  10-20 degrees C > body
- Coagulation of retinal proteins
  
- Photochemical - visible wave length

- Light absorbed - minutes – hours
- Damage  $\lambda < 500$  nm
  - esp. 320-400 nm. (near ultraviolet)
- Non-temperature change
- Light induces super-oxide radicals attack cell walls

#### Clinical picture

- Symptoms
  - Central scotoma
  - Dyschromatopsia
  - Metamorphopsia
  - Va reduced to 20/25 - 20/200
- Most patients recover 3-6 mo. Some have permanent reduction in visual acuity
- Foveolar lesion - small yellow-white spot which fades resulting in cyst or small lamellar hole or mottling of RPE
- Fluorescein angiogram - usually normal or RPE disturbance

#### Pathological – Photochemical

- Acute
- Retinal Edema
- Photoreceptor and RPE damage
  
- Subacute
- Subretinal macropage influx
  
- Chronic
- Regeneration of photoreceptors
- RPE – depigmented
- Proliferative
  
- Severe damage
  - Chronic breakdown outer blood retinal barrier
  - Subretinal neovascularization
- ? CME

#### Etiology

- Believed to be caused by visible light, blue and/or near ultraviolet. Near UV (325-400 nm.) 6 times more toxic than blue light (441 nm.)
- Thermal
- Sun - ozone, eclipse, drugs
- Laser
- ARC welding
- High tension wire
- Photochemical

- Sun
- Operating room microscope - 5 to 7 minutes
- Slit lamp
- Indirect ophthalmoscope - 15 minutes
- Endo illuminators
- Fiberoptic - e.g. urology

#### Factors

- Damage caused is proportional to the area of the pupil
- Pupil size - area =
 

Oxygen (anesthesia)	Threshold for damage
Temperature	Threshold for damage
Vit A	Threshold
Vit C	Threshold (antioxident)
Duration exposure	
Diabetes	
Photosensitizes	
Hydrochlorothiazides	
Phenothiazines	
Focus - aphakes vs. pseudoaphakes	
Intense light exposure may release lysosomal enzymes which can damage cell membranes	

#### Therapy

- Steroids
- Prevent
  - Filters - < 500 nm.
    - Scope
    - IOL
    - Glasses
  - Antioxidants
    - Selenium, zinc, Vitamin E
- Avoid
  - Phenothiazines
  - Psoralen compounds
  - Hematoporphyrin
  - Retinoic acid
  - Allopurinol

### ANGIOID STREAKS

#### History

- 1889 - Doyne – funduscopy
- 1891 - Plange - reddish brown streak
- 1892 - Knapp - angioid streak
- 1917 - Kofler - cracks in Bruch's

- 1929 - Grondblad and Strandberg - angioid and PXE

#### Incidence

1 :70,000 - 1 :160,000

#### Pathology

- Peau d'orange - alteration of Bruch's membrane leading to undulations in the retinal pigment epithelium/traction of extraocular muscles.
- Salmon patches - local changes - Bruch's, RPE and choriocapillaris break in Bruch's
  - decrease pigment of RPE
  - increase pigment of RPE
  - atrophy of choriocapillaris
  - fibrovascular proliferation

#### Associated diseases

- Pseudoxanthoma elasticum
- Paget's disease (osteitis deformans)
- Ehlers - Danlos
- Sickle hemoglobinopathy
- Optic nerve drusen

#### PSEUDOXANTHOMA ELASTICUM

##### Incidence

1 :40,000 - 1 :160,000

##### Hereditary

- Connective tissue? inborn error of metabolism
- Types
  - Autosomal dominant 1
  - Autosomal dominant 2
  - Autosomal recessive 1
  - Autosomal recessive 2

##### Organ involvement

- Skin
- Eye
- Gastrointestinal
- Cardiovascular

##### 23 cases angioid streaks - Montefiore Hospital

- Ages - 9-77 years Mean – 56
- Male:Female - 2:1
- Black:White - 1:2
- Macular Involvement - 14 (61 %) (6 bilateral)

## Vascular anomaly

Opticociliary artery - 54 - F – PXE

## Associated systemic disease

- PXE 9 (40%)
- Paget's 1 (14%)
- Sickle cell 1 (4%)
- No associated disease (52%)
- Disc drusen 3 (13%)
- Retinitis pigmentosa 1 (4%)

## **RPE/CHOROIDAL INFLAMMATORY DISEASE**

### ACUTE MACULAR NEURORETINOPATHY

- (Bos and Deutman) Cloveleaf, wedge-shaped, flower petal, grayish-to-pink lesions, . bilateral, young patients.
- Rapid central vision loss (20/30) - 20/40); often follows flu-like symptoms.
- Resolution and improved vision in weeks to months.
- May occur after systemic allergic reaction to iodine contrast. One case occurred following treatment for uterine bleeding after C- section.
- Fluorescein angiography: petals may hypofluoresce, or be normal.

### RECURRENT MULTIFOCAL CHOROIDITIS

#### Terminology

- Multifocal choroiditis and pan uveitis (Dreyer -and Gass, Nozik and Palestine).
- Punctate inner choroidopathy (Watzke)

#### Clinical findings: simulates POHS:

- Variable vitreous inflammation (RMC, no; MCP, yes)
- Anterior uveitis occurs 50%
- Yellow-gray choroidal lesions; sensory detachment that resolves spontaneously
- Block early, stain late
- Most patients negative for histo
- Female predilection, unilateral (25%)
- Punched out lesions
- SRNV (40%) subretinal fibrosis. Steroids for subfoveal SRNV. Amsler
- Moderate myopia
- Macular lesions arranged in branching pattern

### ACUTE RETINAL PIGMENT EPITHELIITIS

- (KRILL AND DEUTMAN) acute onset of visual disturbances in one or both eyes in young adults. May follow viral syndrome.
- Gradual and almost complete recovery in two months
- Findings
  - Discrete, multiple, round, 1/4 DD dark spots in macular RPE
  - More vision lost than macular changes

- Halo-like depigmented zones around RPE spots (hyperfluoresce)
- F.A.: may be normal
- No treatment

## ACUTE MULTIFOCAL POSTERIOR PLACOID PIGMENT EPITHELIOPATHY

### Findings

- Young, healthy, both sexes
- Rapid visual loss, one or both eyes
- Posterior, flat, placoid, gray-white lesions
- Rare sensory R.D.
- Vitreous cells: 50%
- Preceding flu symptoms (33%), virus?
- Occasional associations:
  - Thyroiditis, cerebrovasculitis, adenovirus 5, lymphadenopathy, hepatomegaly, erythema nodosum, regional enteritis, CSR pleo-cytosis.
  - Perivenous exudation, venous dilatation and tortuosity, papilledema, papillitis, episcleritis, irido-cyclitis
- Occasionally unilateral. Second eye involved a few days to weeks later
- May be recurrent
- Fundus lesions resolve rapidly, 2-4 weeks
- Vision improved, usually 20/30; improved 1-3 months
- Gray-white lesions fade within one week, and replaced by depigmented RPE, clumping occurs.
- Possible choriocapillaris occlusion, probably RPE involvement primarily
- Urinary casts during active phase (Priluck)

### Testing

- Fluorescein angiography
  - Blocks early, stains late
  - ERG, EOG normal
- Chronic
  - RPE window effect
  - RPE mottling
  - SRN rare

### Differential

- Serpiginous (GHPC): lesions resolve more slowly, marked choriocapillaris and large choroidal vessel occlusion, chronic, recurrent; leaves patient severely disabled, 30% SRNV
- Rubella DUSN
- Toxemia (multifocal occlusion of choriocapillaris)
- Neoplastic infiltrates of sub-APE space
- Birdshot choroidopathy
- Tapetoretinal degeneration

## Treatment

- None
- ? steroids

## GEOGRAPHIC HELICOID PERIPAPILLARY CHOROIDOPATHY

### Findings

- Young or middle-aged individual, bilateral (one eye may follow)
- inflammation of RPE and choroids
- Recurrent attacks (weeks to years later) new areas occur adjacent to old areas ("skip area" can occur)
- Usually starts near disc and grows toward macula and periphery (may start in macula)
- No systemic abnormalities
- Occasional anterior segment and vitreous inflammation (33%)
- Hot disc, occasional retinal vasculitis overlying RPE lesions, BVO, NVD, NVE
- SRNV may develop (25%) at edge of chorioretinal atrophy
- Irregular, gray-white, geographic edematous RPE. May appear identical to acute AMPPPE
- Serous detachment infrequent
- Hypofluorescence early (nonperfusion of choroid or obstruction of choroidal fluorescence) and stain late (spreads in from edge)
- Older lesions are flat, pigmented and atrophic large choroidal vessels may drop out. Fibrous metaplasia of RPE (gray-white tissue) may occur in area of chorioretinal atrophy (50%)
- Cellular inflammation at level of choriocapillaris and RPE causing necrosis and scar formation
- Oral steroids, retrobulbar steroids may halt progression of acute disease
- EAG, EOG usually normal

## HARADA'S DISEASE

### Findings

- Pigmented individuals (Asian, Black, American Indian ancestry)
- Rapid visual loss due to sensory retinal detachment, granulomatous anterior/posterior uveitis
- Initial yellowish color in RPE beneath sensory R.D., multiple pigment epithelial detachments, hot disc
- VKH syndrome: headache, vomiting, malaise, CSF pleocytosis, vitiligo, poliosis, alopecia, dysacusis, focal neurologic signs
- Bullous sensory detachments may have shifting fluid
- Uveal effusion
- Treatment systemic, retrobulbar steroid
- Late pigment epithelial changes after resolution of sensory detachment
- Diffuse, salt and pepper RPE change
- Ultrasonography shows posterior uveal tract thickening
- Fluorescein: patchy choroidal nonperfusion, pinpoint areas of hyperfluorescence

that increase to large placoid areas of staining

- Occasional late SRNV, NVD
- Serous detachment is caused by diffuse granulomatous choroiditis of unknown etiology. Subpigment epithelial plaques of inflammatory cellular exudate may be the cause of the yellowish subretinal lesions and the placoid areas of late staining
- Differential diagnosis: (Bilateral detachments) CSR, acute leukemia, metastatic carcinoma, uveal melanocytic proliferation associated systemic carcinoma, uveal effusion syndrome, benign reactive lymphoid hyperplasia of the uveal tract, toxemia, hypertension.