Secondary Glaucomas

• Elevated IOP related to a specific cause
• Some types may be prevented or treatable
• May be unilateral
• Elevated IOP leads to typical glaucomatous changes
  – optic nerve and visual fields

Secondary Glaucomas

• Pigmentary
• Pseudoexfoliation
• Traumatic
• Neovascular
• Steroid
• Uveitic

Pigment Dispersion Syndrome (PDS) and Pigmentary Glaucoma (PG)

• Inherited disorder
• Abnormal irido zonular contact
  – exaggerated by physiologic pupillary movement and accommodation
• Disruption of iris pigment epithelium (IPE)
• Deposition of dispersed pigment granules throughout the anterior segment
• Pigment deposited in angle
• Transient > Then Permanent IOP Elevation

PDS and Pigmentary Glaucoma

• Trabecular meshwork clogs with pigment
  – Pigment Dispersion Syndrome- no elevation in IOP
  – Pigmentary Glaucoma- IOP elevated
• Pigment liberation may be triggered by exercise or pupillary dilation
  – Brief spike in IOP after exercise such as basketball
  – See brown snowstorm of pigment in anterior chamber w brief IOP spike
Pigmentary Glaucoma

Pigmentary Glaucoma

PDS and Pigmentary Glaucoma

• Diagnostic Triad
  – Corneal pigmentation (Krukenberg spindle)
  – Slit-like, radial, mid-peripheral iris transillumination defects
  – Dense trabecular pigmentation

• Iris insertion typically posterior
• Peripheral iris tends to be concave

PDS and PG-Clinical Correlations

• Autosomal dominant genetic origin
  – Phenotypical expression begins in early 20s

• Caucasians

• Gender
  – men and women equally affected with PDS
  – men develop glaucoma 3X as often
    • Men develop at younger age also

• Refractive error
  – 60-80% myopes
  – 20% emmetropes
  • -1.00 to +1.00 D

PDS and Pigmentary Glaucoma - Natural History

• Active phase - pigment liberation
  – probably begins in mid-20s
  – continues to mid-40s

• Regression phase
  – pigment liberation ceases in middle age
    • lens enlarges pushing iris away
    • loss of accommodation (presbyopia)
  – transillumination defects may disappear
    • migration of pigment epithelial cells adjacent to defects

PDS and Pigmentary Glaucoma - Therapy

• Begin therapy early to prevent development of glaucoma
  – Prevent progression of disease
    • therapy is not just to lower IOP

• Difficult decision when to initiate therapy
  – IOP elevated at any time
  – Monitor transillumination defects
PDS and Pigmentary Glaucoma- Therapy

- Miotics: reduce iris concavity and iridozonular contact
  - immobilize pupil
  - creates convex iris
  - bring iris forward away from lens zonules
    - inhibits iris pigment liberation
  - young can not tolerate miotics

- Laser Iridotomy
  - allows communication between anterior and posterior chambers
  - may not completely inhibit pigment liberation
  - may not completely prevent pigment release with exercise
- Argon/Selective Laser Trabeculoplasty (ALT)
  - success rate greater in younger patients

Pseudoexfoliation Syndrome (PEX)

- Ocular and systemic condition
  - unknown etiology
- Excessive presence of extracellular material
- Abnormal production and/or turnover of extracellular matrix material
- Intraocular signs most obvious
- Involves all structures in anterior segment of eye

Pseudoexfoliation or Exfoliation What is the Best Name?

- PXF: material on lens wo elevated IOP
- PXS: syndrome including elevated IOP but no glaucoma
- PXG: syndrome w glaucoma

Pseudoexfoliation Syndrome

Epidemiology:
- Most common identifiable cause of glaucoma
- Increases with age and varies by race
- Elderly
- Caucasians more common
- Often Become Bilateral over time
- Labile IOP
- Aggressive form of glaucoma

Pseudoexfoliation Syndrome

- 15-40% probability of developing glaucoma
- 66% present unilaterally
- 17% odds of Gl. In contralateral eye in 10Y
- Compares unfavorably to POAG
- Cataracts more likely, CE more complicated
Pseudoexfoliation Syndrome

Clinical Features:
- **Lens:** central deposition of white material, a clear zone, and a granular, layered, peripheral zone
- **Iris:** loss of ruff, sphincter transillumination
- **Cornea:** clumps of white material, pigment
- **Angle:** splotchy pigmentation and Sampaolesi line
- **Zonules:** loose with white deposits

Pseudoexfoliation Syndrome (PEX)

- **Clinical Aspects**
  - Pigment and PXE deposits on anterior segment surfaces
  - Ocular hypertension
  - Iris stromal atrophy and hemorrhage
  - Poor mydriasis
  - Melanin dispersion with mydriasis
  - Defect in blood-aqueous barrier
  - Pseudouveitis
  - Posterior synechiae

Pseudoexfoliation Glaucoma

- Both age and elevated IOP increase risk of conversion of OHTN to glaucoma
- There are individuals w XFS who never develop glaucoma
- Electron microscopy has revealed exfoliation fibers in conjunctiva of clinically uninvolved fellow eye
  - Bilateral presentation with asymmetry common

Traumatic Glaucoma

- Any form of trauma may disrupt aqueous humor outflow pathways and elevate IOP
  - Iridodialysis
    - separation of iris from ciliary body
  - Cyclodialysis
    - separation of iris and ciliary body from sclera
  - Angle recession
    - attachment b/w ciliary body and chamber angle only partially disrupted

Traumatic Glaucoma

- **Angle recession**
  - Injury to anterior chamber angle with secondary damage to trabecular meshwork
  - Appears as wide open angle
    - As compared to other eye
  - Associated with hyphema
    - 60% of hyphemas have form of angle recession
    - 6% develop glaucoma
    - Development f glaucoma depends upon number of quadrants involved
      - > 180 degrees- increased likelihood of ^ IOP
Angle Recession

Traumatic Glaucoma

- Angle recession glaucoma
  - manage as other open angle glaucomas
  - damage to TM affects outflow
  - prostaglandins, beta-blockers, alpha agonists, CAI
    - miotics and ALT not typically useful

Inflammatory Glaucoma

- What is the cause of the inflammation?
  - Work with uveitis specialist, internist or rheumatologist to determine appropriate diagnostic workup
  - Specific entity such as Syphilis, Sarcoid, etc.
- How/Why is it IOP elevated?
  - Clogging of meshwork, posterior synechia, etc.

Inflammatory Glaucoma

- Elevated IOP due to three categories
  - Secondary open angle
    - Inflammatory cells and protein clog trabecular meshwork
    - Trabeculitis
    - Steroid induced
  - Secondary closed angle
    - Peripheral anterior synechiae and/or
    - Poster synechiae
      - Pupillary block and Iris Bombe
    - Combination of above

Raised IOP in Uveitis

- Reviewed 257 pts specialty uveitis clinic
- Mean age 45 years old
- 51% White/49% Black
- 53% Male/47% Female
- Acute Uveitis- mean follow up 7.97 years
  - Increased IOP 26.0%
  - Required Therapy 15.1%
Uveitis and Glaucoma Mechanisms

- Mild Inflammation leads to reduced aqueous production
  - Low IOP
- Severe inflammation leads to breakdown of blood-aqueous barrier
  - Accumulation of inflammatory cells, protein, fibrin in trabecular meshwork
  - May also cause direct damage
  - Elevate IOP

Uveitis and Open Angle Glaucoma Mechanisms

- Direct inflammation of Trabecular Meshwork
  - Trabeculitis
  - Associated w Herpetic Disease
- Steroid response w elevated IOP

Uveitis + Angle Closure Glaucoma Mechanisms

- Chronic inflammation leads to formation of peripheral anterior synechiae
  - Direct angle closure
- Chronic inflammation leads to posterior synechiae
  - Pupillary block

Inflammatory Glaucoma

- Treat Inflammation and follow IOP
- If glaucomatous damage is occurring or very high IOP, reduce IOP
- Start with aqueous suppressant agents
  - Timolol bid
- Reserve prostaglandin use as may increase inflammation

Inflammatory Glaucoma

- IOP increases once inflammation is controlled
  - Patient has pre-existing damage to outflow channels
  - Inflammation w reduced IOP masked problem
  - Treat with glaucoma agents
- Steroid responders
  - If unsure if steroid responder, change to weaker steroid and follow IOP
  - If IOP falls confirmation of steroid response
  - Use steroid to control inflammation and if needed, reduce IOP with glaucoma agents temporarily
Inflammatory Glaucoma

• Therapy
  – Topical steroids
    • Mainstay of therapy unless infectious etiology
    • Prednisilone Acetate 1% every 1-2 hours
    • Taper as inflammation resolves
    • If inflammation rebounds, increase dosage to previous levels
    • Vary dosage depending upon severity
    • Mistake often made is not to use often enough

Uveitic Glaucoma

Management

• Periocular Injection/Oral Steroids
  – Greater risks of side effects
  – Use w severe inflammation

• Nonsteroidal Anti-inflammatory drugs
  – Mild inflammation
  – Topical NSAIDs
    • May be helpful, especially as adjunctive therapy

Steroid Glaucoma

• Increase in IOP after the use of steroids
  – topical most common form
    • dexamethasone, prednisolone
    • less common with newer steroids
    – loteprednol, rimexolone
  – often starts within 10 days of use
  – 6-20 mm Hg rise
  – need to suspect condition developing

• Inhaled steroids for asthma/emphysema commonly used and also may rise IOP

Steroid Glaucoma

• Becoming a more common and significant problem with the increase in use of intravitreal dosing
  – IVTA (Kenalog)
  – Sustained-release fluocinolone acetonide inserts (Retisert)
    • 40-50% risk of developing elevated IOP

• Use of IVTA more common for treatment of AMD, Cystoid Macula Edema, Diabetic Macula Edema, Uveitic edema, and as adjunct to panretinal photocoagulation and PDT.

Steroid Glaucoma

• Time to onset of elevated IOP variable
  – From days to months
  – Average of 75 days

• Magnitude of IOP elevation also variable
  – Mild to severe IOP increase
  – Transient to permanent

Steroid Glaucoma

• Elevated IOP develops 2-6 weeks after begin use
  – Rare cases onset abrupt or delayed

• Steroid potency correlates w degree of IOP rise

• Management
  – Discontinue steroid if possible
  – Switch to NSAID or steroid-sparing agent
  – Reduce IOP with glaucoma agents such as beta blockers
Management of Steroid Glaucoma

- Always take baseline IOP
- Patient needs to return within 2-3 weeks of use
- Examine monthly thereafter
- May occur at any time
- If rise noted, try to switch or d/c steroid
- Cover with glaucoma medication
- Follow after episode as glaucoma suspect

Neovascular Glaucoma

- Secondary Closed Angle Glaucoma
- Acutely painful congested eye
- Elevated IOP of 40-80 mmHg
- Corneal edema
- Neovascularization of iris (rubeosis irides)
- Neovascularization of angle

Neovascular Glaucoma

- "Zippering" of angle by synechia
  - History of ischemic retinopathy
- Diabetic retinopathy #1 cause
- CRVO #2 cause
- Typical onset 3-5 months after occlusion
  - "90 day glaucoma"

Management of Neovascular Glaucoma

- Difficult to treat
- PRP
- Filtration surgery
- Cyclocryotherapy
- Decrease IOP if useful vision remains
- Pain control if no useful vision remains
  - Retrobulbar alcohol injection/ Enucleation