Corneal Dystrophy/Degeneration:

What Every Optometrist Should Know

Disclosure

- Presenter is on speakers panel of Alcon, Allergan, Abbott, Bausch + Lomb, Merck, STAAR Surgical, TearLab, HPS and Odyssey
- Past-President of the Optometric Council on Refractive Technology (OCRT)
- OSSO Board Member
- Presenter has NO financial interest in any products mentioned
- Except he does have stock in a certain coffee company...

Corneal Dystrophies

- Group of corneal diseases that are genetically determined and have been traditionally classified with respect to the corneal layer affected
- Defined as a corneal opacity or alteration which is most often bilateral and progressive and centrally located
- Tend to be avascular and involve all the areas of the cornea
- New classification system describes: old name, new name, defective gene, inheritance pattern, phenotype of disorder and typical complications.
Anterior Dystrophies

Meesman's Dystrophy
- Autosomal dominantly
- Symptoms:
  - Foreign body sensation due to epithelial erosion
  - Decreased visual acuity is usually minimal
- Signs:
  - Micropapillae/irregular epithelial cysts that are most prominently seen in the inferior lateral zone
  - Slowly progressive
  - Bilateral, symmetric
  - Develops in the first or second years of life
- Treatment:
  - Superficial cornal debridement
  - PTK

Epithelial Basement Dystrophy (EBMD)
- Abnormal corneal epithelial regeneration and maturation
- Abnormal basement membrane
- Very common dystrophy
- Considered age related
  - Prevalence increases with age
  - Often around 50-70 yo
- Late onset supports degeneration vs. dystrophy
EBMD

- 10-65% of patients are symptomatic
- Symptoms
  - Foreign body sensation
  - Watery, itchy, dry eyes
  - Stinging
  - Air conditioning
  - Blurred vision
  - Dry eye
  - Inverted IOT
  - Discomfort
  - Other symptoms

EBMD

- Appears as a map, dot or fingerprint
- Chalky patches
- Intraepithelial microcysts
- Fissures within central 2/3 of cornea
- Bilateral and asymmetric
- Females > Males
- Negative staining is a good indicator

EBMD

- Treatment of EBMD
  - Monitor cornea for any RCT
  - Avoid long-term use of topical DE
  - Externals
  - Copious artificial tears
  - Bimatoprost
  - Recheck
  - Bimatoprost may cause astigmatism
  - Topical plugs
  - Consider eyelids
  - Surgery may be needed
  - PTK
  - Ectasia
  - Monitor for changes in visual acuity or comfort
STROMAL DYSTROPHIES

Granular Dystrophy
(Greenouw Type I)

- Discrete white granular opacities in central anterior corneal stroma
- Increasing number, density, size and depth in age
- RCE's are commonly associated with pain
- Sub-epithelial scarring/dense stromal deposits reduce visual acuity
- PKP if disease progresses

GRANULAR DYSTROPHY

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Granular Dystrophy

GRANULAR DYSTROPHY

GRANULAR DYSTROPHY

Macular Dystrophy
(Groenouw Type II)

- Grayish opacities with indistinct edges in superficial stroma
- Over time
  - Extends into deeper stromal layers
  - Intervening stroma becomes hazy
  - Visual acuity is decreased
  - Light sensitivity and pain
- Surgery is expected by 20-50 years old
Lattice Dystrophy (Type I)
- Clinically appears
- Linear or focal branching deposits within the anterior stroma
- Central cornea becomes opaque and scars decreasing the visual acuity
- Autosomal Dominant
- < 20/40 Decade
- > 60/60 decade decrease VA
- RCE scars associated with lattice
- Surgical intervention recommended with decreased acuity
Lattice Dystrophy

Central Crystalline Dystrophy of Schnyder
- Central discoid opacification posterior to Bowman's membrane in anterior stroma
- Opacities consist of:
  - Small needle-shaped refractive crystals
  - White
  - Polymorphous
  - May extend into deeper stroma avoiding epithelium
- Vision is relatively unaffected
- Associated with cholesterolemia

Schnyder's Crystalline Dystrophy

Other Stromal Dystrophies
- Avellino
- Gelatinous Drop Like
- Fleck
- Central Cloudy
- Posterior Amorphous
POSTERIOR MEMBRANE DYSTROPHIES

Posterior Polymorphous
- Autosomal dominant
- Teens to 20's
- Vesicles at Descemets Endothelium
- Signs
  - Vesicle bands
  - Diffuse opacities
  - Edema
  - Corneal steepening
  - Increase IOP

Posterior Polymorphous (PPMD)
- Vesicles are hallmark of PPMD
- Bilateral
- Trabecular meshwork can become covered with epithelial cells and basement membrane
- Synechiae can be present

Fuch's Dystrophy
- Autosomal dominant inheritance
- Bilateral / Asymmetry
- Late onset > 50 y.o.
- Females affected 3 times more than males
  - 5-7% develop edema
- Characterized
  - Corneal guttata
  - Extracellular accumulation of abnormal endothelial secretion
  - Appears in 30-40 year old
Fuch's Dystrophy

- Characterized
- Ciliary Goutta
  - Small refractive "drops" or corneal endothelium
  - Affects the "pump" action of the endothelium
- Edema
  - Greater in the AV
  - Declines as day goes on
  - Longstanding edema may lead to central scarring
  - KCS common

Fuch's Dystrophy

- Symptoms vary with degree of goutta and compromise of the endothelial tissue
- Moderate goutta
  - May affect visual function
  - May induce mild-moderate edema
  - Halo around lights
  - May vision in A.M.
- Severe goutta
  - Vision decreases
  - Possible bullous develops

Fuch's Dystrophy

- Treatment
  - Early stages of disease
  - Increase topical drops
  - Hypromellose 0.1%
  - BCL used if bullous present
  - EDUCATION
  - Visual function is significantly compromised
  - Penetrating keratoplasty
  - DALK or EK (endothelial keratoplasty)
  - DSAEK
Puch's Dystrophy

- DLEK
  - Recipient cornea is stripped of Descemet's membrane and endothelium
  - Transplantation of donor cornea through small incision
  - Results in
    - Improved endothelial function, corneal clarity and intraocular pressure
    - Minimally affects refraction
    - Can provide rapid visual recovery
    - Maintains structural integrity of the cornea

Congenital Hereditary Endothelial Dystrophy (CHED)

- Rare congenital dystrophy
- First weeks 6 months old
- Bilateral-symmetric
- Noninflammatory clouding
- Signs
  - Opacification extending to limbus with clear zones
  - Thickening
  - No neovascularization
  - No increase in IOP

- Nystagmus present
- VA can be as low as 20/200
- No neovascularization
- No increase in IOP
- Diagnosis of exclusion
Congenital Hereditary Endothelium Dystrophy

Iridocorneal Endothelial Syndrome (ICE)

ICE
- F6M
- Diagnosed 3rd to 5th Decade
- 3 Main features
  - Iris changes
  - Cornea swelling
  - Glaucoma
- Unknown etiology

ICE
- Abnormal endothelium
- Irido-corneal adhesions
- 80-100% develop glaucoma
- Increase IOP
- Edema
- Iris
  - Mild to severe atrophy
  - Nodules may be present
  - Glowy membranes on iris
- Condition can be relentless and difficult to treat
Corneal Degenerations

- Defined as a deterioration or change from a higher to a lower form, especially change of tissue to a lower or less functionally active.
- Non-inherited
- Unilateral or bilateral
- Asymmetric
- Develop in later years
- Variable progression
- Systemic disease can be associated

Degenerations

- Arcus
- Spheroïdal degeneration
- Amyloid
- Limbal gridle of Wegi
- Band keratopathy
- Salzman’s nodular degeneration

Degenerations

- Coats white ring
- Hassall-Henic bodies
- Crocodile leash green
- Senile furrow
- Dellen
- Pterygial
- Pterygium
Ectatic Disorders

Keratoconus

- Ectatic corneal dystrophy
- Bilateral with asymmetry
- Manifests in 20-30s
- Most likely a multigenic disease
  - Complex mode of inheritance
  - Environmental factors influence manifestation
- Etiology
  - Decreased enzyme activities
  - Decreased levels of enzymes inhibitors
- Destruction of normal corneal matrix results in thinning and scarring
**Keratoconus**

- **Diagnoses**
  - Slit lamp findings
  - Munson's sign
  - Central corneal thinning
  - Fleck-chewing
  - Scarring at Bowman's layer or anterior stroma
  - Age is same (vertical striated
  - Irregular astigmatism
  - Keratogingival disjunction with both contact and glasses
  - Topographically
  - Inferior steepening

**Keratoconus**

- Gestates for approximately 10-20 years and then stabilizes
- Severity is variable between patients
- Often asymmetric appearance
- Thinning can be extensive:
  - Resulting in rupture in Descemet's membrane
  - This results in a posterior corneal decompensation
  - Indoprop

**Keratoconus**

- **Hydrops**
  - Symptoms
    - Sudden decrease in best corrected vision
    - Foreign body sensation
    - Pain
  - Signs
    - Conjunctival hyperemia
    - Prominent central or inferior corneal edema
    - Goblet
  - Self-limited in 3-5 weeks; endothelial cells regenerate at ruptured Descemet's membrane
Keratoconus

- Treatment
- Hydros
- Hypoammonetics
- Antibiotics to avoid secondary infection
- IOP
- RGP's
- 9:20 sphere, Kori Design Valley Center
- Hydros
- Synechiae after Hybrid

Keratoconus Treatment Flow
The New Paradigm

- Disease Identification & Management
  - Spectacle, Contacts, Custom Lenses
- Identification of Surgical Need
  - Coronal Lente intolerance or Risk of Blowing
- Work-Up, INTAC/Surgery, 1-Day & 3-Month Post-Op
  - 2 to 3 Day Patient Recovery
- Ongoing Follow-Up
  - Include Initial CI, Fit
- Post-Op Management & Outcome Analysis
  - Referral if Consultations or Aesthetic Outcome
- Long-Term Follow-Up
  - Include CI Filing, Periodic Monitoring (Order/YPF)

Riboflavin (CXL)

- Increase in cross-links
- Strenthenes Cornea
- Riboflavin eye drops are applied to the cornea
- The riboflavin is activated by a UV-light

Corneal Crosslinking
Riboflavin & UV

- Increase in corneal rigidity
- Reduced Corneal thickness
- Young's modulus
- Increased 4x in Human corneas
**Corneal Crosslinking with Riboflavin (CXL)**

**Corneal Crosslinking Clinical Applications**
- Keratoconus/juvenile keratoconus
- Corneal stabilization
  - PTK
  - PKP
  - MMC
  - Toric PK
  - Referred PK/RK/DS
  - CRT/Rehatomology
- Corneal ulcers
- Myopia control

**Corneal Crosslinking Clinical Applications**
- Intracorneal ring segments
  - FDA approved for nearsightedness 1998
  - FDA approved under HDE 2004
- Provide structural support to thinned peripheral cornea
- Flattens cone
- Pulls cone toward center of cornea
- Decreases irregular astigmatism
Pellucid Marginal Degeneration (PMD)

- Bilateral thinning of the inferior peripheral cornea
- Thinning occurs 1-2 mm above inferior limbus
- Separated by an area of uninvolved cornea between limbus and thin zone
- Hydrops may present in the thinner area
- Commonly seen in and to 3rd decade
- Non-hereditary
- M-F

PMD

- Subjective symptoms
  - Increase in astigmatism
  - Unexplained decrease in visual acuity
- Affected area is clear of lipid or vascularization
- Corneal topography has distinct inferior steepening
  - Crab claw
  - Kissing flaps
  - Beard and mustache

PMD

- Treatment
  - glasses
  - Traditionally may be sufficient with PMD
  - Multifocal lens
  - Contact lens
  - Challenges fix with increase astigmatism (ATH)
  - Asymmetrical astigmatism
- Surgical intervention
  - PK
  - Inferior lamellar patch graft
Terrien's Marginal Degeneration

- Rare bilateral asymmetric disease
- Unknown etiology
- Superior peripheral cornea thins eccentrically
- Lateral deposition
- Decubitus
- Organization
- Can perforate
- No changes in epithelium

Terrien's Marginal Degeneration

- Occurs at any age or sex
- Although more typical in middle-aged males
- No signs of inflammation
- No injection of conjunctiva
- No A/C chamber reaction
- Increase in regular and irregular astigmatism
- Asymptomatic
- Change in vision may be a prompt

Terrien's Degeneration

- Circumferential yellow demarcation
- Lipid and fine pannus
- Often resembles a pterygium
- Perforation is rare, without trauma
- Hydrops may occur
- Topography
  - Corneal flattening at juncture of fornix
  - Steepening 90 degrees from flat area
  - Spherical and regular central area
**Terrien's Marginal Degeneration**

- Management
  - Asymptomatic thus education and supportive
  - Initiated red eyes on occasion
  - Lesions/cyst
  - Early refractive treatments
    - Spectacles
    - Contact lenses
    - IOP
    - Explant lenses
  - Surgical intervention includes PK

**Mooren's Ulcer**

- Painful relentless chronic ulcerative keratitis
- Initially starts peripherally and progresses circumferentially and centrally
- Idiopathic

**Mooren's Ulcer**

- Divided into 3 distinct variations
  - Unilateral Mooren's
  - Progressive keratitis in elderly
  - Bilateral Aggressive Mooren's
    - Younger patients
    - Circumferentially progresses towards central ulceration
  - Bilateral Indolent Mooren's
    - Middle-aged patients
    - Progressive peripapillary swelling
    - Iritis
    - Little inflammation
Mooren's Ulcer

- Pathophysiological mechanism unknown
  - Possibly autoimmune
- Presents
  - Redness
  - Irritation
  - Photophobia
  - Pain
  - Often worse than inflammation indicates
  - Visual disruption - irregular astigmatism
  - Acanthamoeba

Mooren's Ulcer

- Treatment
  - Steroids:
    - Pred Forte qid
  - Cycloplegia
  - Topical antibiotics:
    - 4th generation for Acanthamoeba
  - Oral steroids
  - Conjunctival resection
  - Immunosuppressive therapy

Let's Put It All Together

Case 3

- 83 y.o. male
- Presents to office for general eye exam
- Hyperopic
- Equatorial macular
- Last eye exam was NEVER
- "I hate Dr's you are all crazy-I am here because I need my drivers license!"
Case 1
- Ocular Marshmellowitis
- Arcus Senilis
- Limbal girdle of Voigt
- Terrien's Marginal Degeneration

Case 2
- 53 y.o. nursery school teacher
- "I noticed a white spot in my eye"
- The last 6 months, becoming a lot
- We play with glue a lot
- +/- NIDDM; HTN;
- UCVA 20/25-OU

Case 2
- Reiss-Bucklers
- Keratoconjunctivitis Sicca
- Schnyder's Crystalline Dystrophy
- Astellino Degeneration
- Snot
- Hardening and gelled over time

Case 3
- 53 y.o. Wal-Mart greeter
- "I want LASIK surgery"
- UCVA
- OD 20/20
- OS 20/25
- +/- NA
- 20/20, 20/25
- No corneal staining
- Hx of pain eye from time to time in AM
Case 3
- Keratoconus
- EBMD
- Pluritis
- Who cares! Can't we just be done with this lecture already-seriously enough of these ridiculous questions: "Bored with this!"

Case 4
- 37 y.o. professional roller-blader
- "My eye is irritated, red, and I don't see as well as I used to"
- "My girlfriend is a pre-school teacher"
- "I use Visine!"

Case 4
- Crocodile Shagreen
- Pterygium
- Macular Dystrophy
- Phlegm
- More snot!

Case 5
- 39 y.o. male
- "Need help, abnormal firm nodules on top of my eye"
- "In even the slightest light"
- "Make me wear glasses while applying my hair gel at the male houseplit"
- "Must want for new shades. They were great and is REBA!"
- "My eyeballs!"
Case 5

- Arcus Inversus
- Lattice Degeneration
- Hair Net Dystrophy
- Macular Dystrophy
- Punishment for bringing ABBA back!

Case 6

- 58 y.o. feline exerciser
- "I have not had an exam in a few years"
- Hx of taking drop with "yellow" top
- Wants a new Rx
  - VA: 20/40
  - IOP: 23 mmHg

- PPMO
- WB
- SUY
- CHEE
- LOST
- OAT
- GDx
- ICE
- Too cold, Too cold
Thank you