Corneal Dystrophy/Degeneration: What Every Optometrist Should Know
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Disclosure
- Presenter is on speakers panel of Alcon, Allergan, Abbott, Bausch + Lomb, Tear Lab, RPS and BVI
- 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:
  - Myriads of tiny intraepithelial cysts that are most prominently seen in the interpalpebral zone
  - Slowly progressive
  - Bilateral, symmetric
  - Develops in the first 1 or 2 years of life
- Treatment:
  - Superficial corneal debridement
  - PTK
Meesman's Dystrophy

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

**EBMD**
- 10-69% of patients are symptomatic
- Symptoms
  - Foreign body sensation
  - Blurred vision
  - Dry eye
  - Discomfort
EBMD

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

Photo Courtesy: Tracy Swartz, OD, FAAO

EBMD

Photo Courtesy: Tracy Swartz, OD, FAAO

EBMD

- Monitoring cornea for any RCE
- Kote
- Restasis bid
- Bandage contact lens while active
- Punctal plugs
- Consider humidifier
- Surgery may be needed
- PTK
- Puncture
- Monitor for changes in visual acuity or comfort

Photo Courtesy: Tracy Swartz, OD, FAAO
**EBMD POST-PTK**

**Bowman’s Layer Dystrophies**

- Reis-Buckler
  - Autosomal Dominant
  - Corneal Surface is smooth
- Thiel-Behnke
  - Autosomal Dominant
  - Recent vision loss
  - Corneal sensation normal
  - May present with RCE's

**Reis-Buckler**
Thiel-Behnke Dystrophy

STROMAL DYSTROPHIES

Granular Dystrophy (Groenouw Type I)
- Discrete white granular opacities in central anterior corneal stroma
- Increasing number, density, size and depth as age
- RCE's are commonly associated with pain
- Sub-epithelial scarring/dense stomal deposits reduce visual acuity
- PKP if disease progresses
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 20-30 years old
Macular Dystrophy

- Linear, refractive branching deposits within the anterior stroma
- Central cornea becomes opaque and scars decreasing the visual acuity
- Autosomal Dominant
- 1st Decade
- > 4th decade decrease VA
- 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 refractile crystals
    - White
    - Polychromatic
    - May extend into deeper stroma-avoiding epithelium
- Associated with cholesterolimia
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 Descemet's/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
- Corneal guttata
  - Excessive accumulation of abnormal endothelial secretions
  - Appears in 30-40th year of life

Photo Courtesy Tracy Swartz OD, FAAO
Fuch's Dystrophy

- Corneal Guttata
  - Visual reflects “drips” on corneal endothelium
  - Affects the “pump” action of the endothelium

- Edema
  - Greater in the AM
  - Desiccates as day goes on
  - Long standing edema may lead to corneal scarring
  - RCE's common

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Fuch's Dystrophy

- Symptoms vary with degree of guttata and compromise of the endothelial tissue

- Moderate guttata
  - May affect visual function
  - May induce mild-moderate edema
    - Halos around lights
    - Hazy vision > a.m.

- Severe guttata
  - Vision decreases
  - Possible bullae develops

FUCH'S DYSTROPHY

PhotoCourtesy: Tracy Swartz, OD, FAAO
Fuch's Dystrophy

- Treatment
  - Early stages of disease
  - Increase artificial tears
  - Hyperosmotics qhs
  - BCL used if Bullous is present
  - EDUCATION!
- Visual function is significantly compromised
  - Penetrating keratoplasty
  - Deep Lamellar endothelial keratoplasty (DLEK)
  - Descemet stripping automated endothelial keratoplasty (DSAEK)

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Fuch’s Dystrophy

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

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Photo Courtesy Tracy Swartz OD, FAAO
Congenital Hereditary Endothelium Dystrophy (CHED)

- Rare congenital dystrophy
- First weeks-6 months old
  - Opacification extending to limbus with clear zones
  - Thickening
  - No neo/No extra tissue
  - No increase in IOP

Photo Courtesy: Tracy Swartz OD, FAAO

Congenital Hereditary Endothelium Dystrophy

- Nystagmus is present
- VA can be as low as 20/100
- No neo/No extra tissue
- No increase in IOP
- Diagnosis of exclusion

Photo Courtesy: Tracy Swartz OD, FAAO

Congenital Hereditary Endothelium Dystrophy

[Images of ocular anatomy]
Iridocorneal Endothelial Syndrome

ICE

- F>M
- Diagnosed 3rd to 5th Decade
- 3 Main features
  - Iris changes
  - Corneal swelling
  - Glaucoma
- Unknown etiology

- Increase IOP
- Edema
- Mild to severe atrophy
- Nodules may be present
- Glassy membrane on iris
- 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
- Spheroidal degeneration
- Amyloid
- Limbal girdle of Vogt
- Band keratopathy
- Salzmann's nodular degeneration
Degenerations

- Coats white Ring
- Hassal-Henle bodies
- Crocodile shagreen
- Senile furrow
- Dellen
- Pingueculae
- Pterygium

Ectatic Disorders

Keratoconus

Photo Courtesy Tracy Swartz, FAAO
Keratoconus

- Ectatic corneal dystrophy
- Bilateral with asymmetry
- Manifests in 20-30's
- Most likely a multigenic disease
  - Complex mode of inheritance
  - Environmental factors influence manifestation
  - Increased enzyme activities / decreased levels of enzyme inhibitors = toxicity
  - Destruction of normal corneal matrix results in thinning and scarring

Keratoconus

Diagnosis

- Slit lamp findings
- Munson's Sign
- Central corneal thinning
- Fleischer's ring
- Scarring at Bowman's layer or anterior stroma
- Vogt's striae (vertical striae)
- Irregular astigmatism
  - Resulting in difficult refraction with both contacts 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 aqueous infusion into stroma
  - Hydrops

**KERATOCONUS**
**DESCEMET’S BREAK**

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Keratoconus

- Hydrops
- Symptoms
  - Sudden decrease in best corrected vision
  - Foreign body sensation
  - Pain
- Signs
  - Conjunctival hyperemia/redness
  - Prominent central or inferior corneal edema
  - Clouding
- Self-limited in 8-10 weeks as endothelial cells regenerate at ruptured Descemet's membrane
Keratoconus

- Treatment
  - Hydrops
  - Hyperosmotics
  - Antibiotics to avoid secondary infection
  - PKP
  - RGP's
    - Bi-Aspheric I-Kone Design Valley Contax
    - Hyrokone
    - SynergEyes High Dk Hybrid

Keratoconus Treatment Flow
The New Paradigm

- Disease Identification & Management
  - Spectacles, Contacts, Custom Lenses

- Identification of Surgical Need
  - Contact Lens Tolerance or Risk of Scarring

- Post-Op Management & Outcome Analysis
  - Re-referral if Complications or Atypical Outcomes

- Work-Up, INTACS Surgery, 1-Day & 3-Month Post-Op
  - 1-2 Days Patient Recovery

- Ongoing Follow-Up
  - Include Initial CL Fit

- Post-Op Management & Outcome Analysis
  - Re-referral if Complications or Atypical Outcomes

- Long-Term Follow-Up
  - Include CL Fitting, Periodic Monitoring (Slate PKP)

Riboflavin
(CXL)

- Increase in cross links
- Strengthens Cornea
- Riboflavin eye drops are applied to the cornea
- The riboflavin is activated by a UV-light
Corneal Crosslinking
Riboflavin & UVA

- **rigidity**
  - 343% human corneas

- **Young’s modulus**
  - Increased 4.5X in human corneas


Corneal Crosslinking with Riboflavin:
CXL

Corneal crosslinking procedure.
Corneal Crosslinking Clinical Applications

- Keratoclastic
- Corneal stabilization
- Intacs
- CK, LTK
- RK, HK
- Extended PRK/LASEK
- CRT/Orthokeratology
- Corneal ulcers
- Myopia control

Corneal Crosslinking Clinical Applications

**Treatment:** Corneal Ectasia/iatrogenic KC

- 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 2nd to 3rd decade
- Non-hereditary
- M=F
Pellucid Marginal Degeneration

PMD
- Subjective symptoms
  - Increase in against-the-rule astigmatism
  - Unexplained decrease in visual acuity
- Crab claw
- Kissing doves
- Beard and mustache

PMD
- Treatment
  - Glasses
  - Traditionally may be sufficient with PMD
  - Matching astigmatism
  - Contact lens
  - Challenging fits with increase astigmatism (ATR)
  - Asymmetrical astigmatism
  - Surgical intervention
    - PK
    - Inferior lamellar patch graft
Terrien’s Marginal Degeneration

- Rare bilateral asymmetric disease
- Unknown etiology
- Superior peripheral cornea thins/Ectatic
  - Lipid deposition
  - Vascularization
  - Opacification
  - Can perforate
  - No changes to 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 Marginal 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 furrow
  - Steepening 90 degrees from flat area
  - Spherical and regular central area

Terrien's Marginal Degeneration

- Management
  - Asymptomatic thus education and supportive
  - Irritated red eyes on occasion
    - Lotemax qid
  - Early refractive treatments
    - Spectacles
    - Contact lenses
    - RGP
    - Piggyback 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 ulceration in elderly
  - Bilateral Aggressive Mooren’s
    - Younger patients
    - Circumferentially progresses towards central ulceration
  - Bilateral indolent Mooren’s
    - Middle aged patients
    - Progressive peripheral guttering
      - Bilaterally
      - Little inflammation

Mooren’s Ulcer

- Pathophysiological mechanism unknown
  - Possibly autoimmune
- Presents
  - Redness
  - Tearing
  - Photophobia
  - PAIN
    - Often worse than inflammation indicates
  - Visual disruption-irregular astigmatism
  - Iritis
Mooren’s Ulcer

- Treatment
- Steroids
  - Pred Forte q1h
- Cycloplegia
- Topical antibiotic
  - 4th generation fluoroquinolone
- Oral steroids
- Conjunctival resection
- Immunosuppressive therapy

Thank you