Corneal Dystrophy/Degeneration:
What Every Optometrist Should Know
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Disclosure
- Presenter is on speakers panel of Alcon, Allergan, Abbott, Bausch + Lomb, Merck, STAAR Surgical, TearLab, WFS and Odyssey
- Past-President of the Optometric Council on Refractive Technology (OCR)
- OMSO 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:
  - Meissner's disease: epithelial cysts that are most prominently seen in the central pellucid zone
  - Slowly progressive
  - Bilateral, symmetric
  - Develops in the first 5 or 6 years of life
- Treatment:
  - Superficial corneal 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
  - Occurs around age 50
- Late onset supports degeneration vs. dystrophy
EBMD

- 10-65% of patients are symptomatic
- Symptoms
  - Foreign body sensation
  - Watery, itchy eyes
  - Irritability
  - Allergies
  - Blurred vision
  - Dry eye
  - Intermittent blurry vision
  - Discomfort
  - Other common RKS

EBMD

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

EBMD

- Treatment of EBMD
  - Monitoring cornea for any RKS
  - Azithromycin oral drops every 6 hours for 6 weeks
  - Subconjunctival injection
  - Topical antibiotics
  - Topical steroids
  - Reducing contact lenses while active
  - Punctal plugs
  - Corneal bandage
  - Surgery may be needed
  - PTK
  - FDT
  - Monitor for changes in visual acuity or comfort
### EBMD POST-PTK

![Image of EBMD POST-PTK](image1)

### Bowman's Layer Dystrophies

- **Reis-Buckler**
  - Autosomal Dominant
  - Corneal surface is smooth

- **Thiel-Behnke**
  - Autosomal Dominant
  - Corneal sensation normal
  - May present with RCE's

### Reis-Buckler

![Image of Reis-Buckler](image2)

### Thiel-Behnke Dystrophy

![Image of Thiel-Behnke Dystrophy](image3)

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Edward S. Harkness Eye Institute
Columbia University
Granular Dystrophy
(Greenough 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
- Subepithelial scarring/dense stromal deposits reduce visual acuity
- PKP if disease progresses
Granular Dystrophy

GRANULAR DYSTOPHY

GRANULAR DYSTOPHY

Macular Dystrophy (Grbaczowa 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 30-50 years old
Macular Dystrophy

Lattice Dystrophy (Type I)
- Clinically appears
  - Linear reticular branching deposits within the anterior stroma
  - Central cornea becomes opaque and tears decreasing the visual acuity
- Autosomal Dominant
- < 1 decade
- > 1 decade decrease VA
- RPE rare associated with lattice
- Surgical intervention recommended with decreased acuity
Lattice Dystrophy

- Central discoid opacification posterior to Bowman’s membrane in anterior stroma
- Opacities consist of:
  - Small needle-shaped refractive crystals
  - White
  - Polychromatic
  - May extend into deeper stroma avoiding epithelium
- Vision is relatively unaffected
- Associated with cholesteroloma

Central Crystalline Dystrophy of Schnyder

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
  - Iridal
  - 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 guttae
  - Excessive accumulation of abnormal endothelial cells
  - Appears in 30-50 years of life

10/22/13
Fuch's Dystrophy

- Characterized
  - Corneal Guttata
    - Small, visible "droplets" on corneal endothelium
  - Edema
    - Hypertrophy of AK
    - Impaired blood flow to endothelium

- Symptoms
  - Moderate guttata
    - May affect visual function
    - May induce mild to moderate edema
    - Halo around lights
    - Hazy vision
  - Severe guttata
    - Vision decreases
    - Possible bullous develops

Fuch's Dystrophy

- Treatment
  - Early stage of disease
  - Increased fluid flow
  - Hypotonic drops
  - BCL (if bullous present)
  - Education
  - Visual function is significantly compromised
  - Penetrating keratoplasty
  - DCSK or endothelial keratoplasty (DSK)
  - DMEK donor-lens endothelial keratoplasty (DMEK)
**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, correct clarity and intraocular
    - Minimally affects refraction
    - Can provide rapid visual recovery
    - Maintains structural integrity of the cornea

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**Congenital Hereditary Endothelial Dystrophy (CHED)**

- Rare congenital dystrophy
- First weeks to 6 months old
- Bilateral symmetric
- Non-inflammatory clouding
- Signs:
  - Opacification extending to limbus with clear zones
  - Thinning
  - No new/no extra tissue
  - No increase in IOP

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**Congenital Hereditary Endothelial Dystrophy**

- Nystagmus is present
- VA can be as low as 20/200
- No new/no extra tissue
- No increase in IOP
- Diagnosis of exclusion
Congenital Hereditary Endothelium Dystrophy

Iridocorneal Endothelial Syndrome
ICE

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

ICE

- Abnormal endothelium
- Iris corneal adhesions
- 90-100% develop glaucoma
- Increased IOP
- Edema
- Iris
  - Mild to severe atrophy
  - Nodules may be present
  - Glaucome membroane 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
- Sphairoidal degeneration
- Amyloid
- Limbal gridle of Vegi
- Band keratopathy
- Salzman’s nodular degeneration

Degenerations

- Coats white Ring
- Hassall-Henle bodies
- Crocodile lash green
- Senile furrow
- Delen
- Pterygeac
- Pterygium
Ectatic Disorders

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

Keratoconus

- Etiology
  - Increased enzyme activities (decreased levels of enzyme inhibitors)
  - Toxicity
  - Destruction of normal corneal matrix; results in thinning and scarring
Keratoconus

- Diagnoses
- Slit lamp findings
- Munzoff’s sign
- Central corneal thinning
- Fleckling
- Scarring at Bowman’s layer or anterior stroma
- Age is stable (vertical strand)
- Irregular astigmatism
- Keratoglobus (intrastromal with both contact glasses)
- Topographically
- Inferior stroming

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 of Descemet’s membrane
  - This results in posterior stromal edema
  - Hydrops

Keratoconus

- Signs
  - Conjunctival hyperemia
  - Prominent central and inferior corneal edema
  - Gobbling
  - Self-limited in 6-8 weeks; endothelial cells regenerate at ruptured Descemet’s membrane

Keratoconus

- Hydrops
- Symptoms
  - Sudden decrease in best corrected vision
  - Foreign body sensation
  - Pain
- Signs
  - Conjunctival hyperemia
  - Prominent central and inferior corneal edema
  - Gobbling
  - Self-limited in 6-8 weeks; endothelial cells regenerate at ruptured Descemet’s membrane
Keratoconus

- Treatment
  - Hydration
  - Topical medications
  - Intraocular gas injection
  - Hyperopic LASIK
  - RGP lenses
  - Skeletal laser osteoplasty
  - Hyaluronic acid injection
  - Artificial corneal tissue replacement

Keratoconus Treatment Flow
The New Paradigm

- Disease Identification & Management
- Spectacle, Contacts, Custom Lenses
- Identification of Surgical Need
- Contact Lens intolerance or Risk of Scarring
- Work-Up, INTACS Surgery, 1-Day Post-Op
- 3 Month Post-Op
- Post-Op Management & Outcome Analysis
- Follow-Up
- Ongoing Follow-Up
- Intra-Corneal Implant (ICL)
- Long-Term Follow-Up
- Ongoing Management
- Intraocular lens implantation
- Cornal crosslinking
- Riboflavin 2% (CXL)
- Increase in corneal thickness
- Strengthen corneal
difference in corneal
- Riboflavin eye drops are applied to the cornea
- The riboflavin is activated by a UV-light

Corneal Crosslinking
Riboflavin & UV-A

- Increase in corneal rigidity
- G21 penetration rate
- Young’s modulus
- Increased G, G21 in human corneas

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**Corneal Crosslinking with Riboflavin: CXL**

**Corneal Crosslinking Clinical Applications**
- Keratoconus/juvenile keratoconus
- Corneal stabilization
- LASIK
- PRK
- RLE
- Advanced Hybrid LASIK
- CRT/multikeraology
- Corneal ulcers
- Myopia control

**Corneal Crosslinking Clinical Applications**
- Intracorneal ring segments
- FDA approved for nearsightedness 1998
- FDA approved under IDE 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 unaffected cornea between limbus and thinning
- Hydrops may present in the thinner area
- Commonly seen in 2nd to 3rd decade
- Non-inherited
- M:F

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

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

- Rare bilateral asymmetric disease
- Unknown etiology
- Superior peripheral cornea thinning
- 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 conjunctive
- No A/C chamber reaction
- Increase in regular and irregular astigmatism
- Asymptomatic
- Change in vision may be a prompt

**Terrien's Degeneration**

**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 fornix
  - Strengthening 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
  - Lesional
  - Early reductive treatments
    - Spectacles
    - Contact lenses
    - IOP
    - PTK
  - 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
    - Presents slowly in elderly
  - Bilateral Aggressive Mooren's
    - Younger patients
    - Circumferentially progresses towards central ulceration
  - Bilateral Incontinent Mooren's
    - Middle-aged patients
    - Progressive peripheral guttation
    - Blindly
    - Little inflammation
Mooren's Ulcer

- Pathophysiological mechanism unknown
  - Possibly autoimmune
- Presents
  - Redness
  - Irritation
  - Photophobia
  - Pain
  - Often worse than inflammation indicates
  - Visual disturbance or irregular astigmatism
  - Acute

Mooren's Ulcer

- Treatment
  - Steroids:
    - Prednisolone
  - Cycloplegia
  - Topical antibiotics:
    - 4th generation fluoroquinolones
  - Oral steroids
  - Conjunctival resection
  - Immunosuppressive therapy

Let's Put It All Together

Case 3

- 63 y.o. male
- Presents to office for general eye exam
  - Irritated
  - Repeatedly denied
  - 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 Marshmallowitis
- 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 left eye is worse, redness & tearing
- No diabetes with glucose
- NIDDM & HTN
- UCVA 20/25 OU

**Case 3**
- 57 y.o. Wall-Mart greeter
- "I want LASIK surgery"
- UCVA
  - OD 20/30
  - OS 20/50
  - OD 0.30 x 60
  - OS 0.25 x 60
- No corneal staining
- Hx of pain eye from time to time in AM
Case 3
- Keratoconus
- EBMD
- Hornitis

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 eyes are 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
- "Constant dryness of eyes in my nose and in my mouth"
- "I have really dry eyes and when I apply Vaseline to this make them better"
- "I must want to reiterate this are increased tear production and is ABAE"
- "My eye hurts!"
**Case 5**

- RCVA
- 26 years
- 26 Aug
- Pachymetry
- 50 um
- 70 cells
- Mild staining noted on the cornea
- Deep striae more centrally located
- Strong aerosol smell

**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 mm Hg

**Case 6**

- PPMD
- WB
- SUY
- CHED
- LOST
- OAT
- GDx
- ICE
  - Too cold, Too cold
Thank you