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…

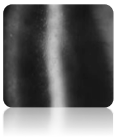
Corneal Dystrophies

- 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

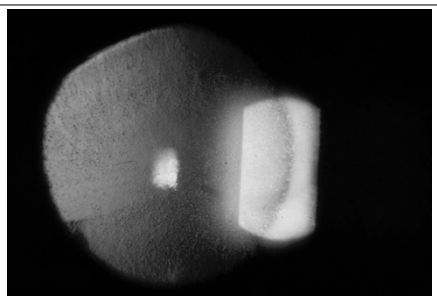
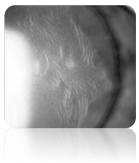


Photo Courtesy Tracy Swartz OD, FAAO

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
    -

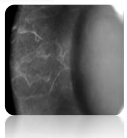
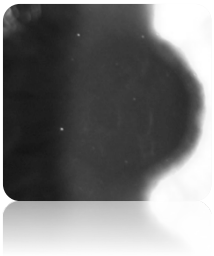


Photo Courtesy Tracy Swartz OD, FAAO

EBMD

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



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EBMD

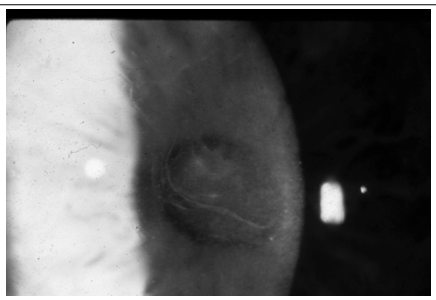


Photo Courtesy Tracy Swartz OD, FAAO

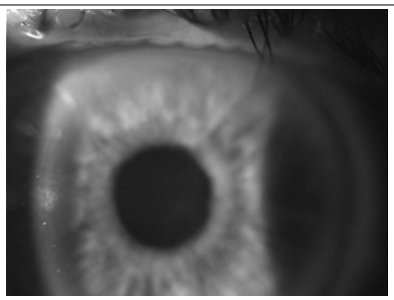
EBMD

-
  - 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

Melton and Thomas

Photo Courtesy Tracy Swartz OD, FAAO

EBMD POST-PTK



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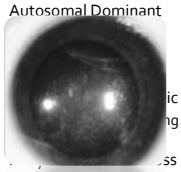
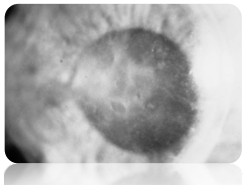
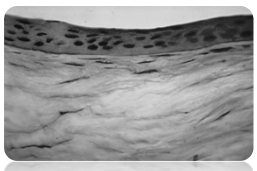
Bowman's Layer Dystrophies

- Reis-Buckler
  - Autosomal Dominant
  - ic
  - ng
  - ss
  - Corneal Surface is smooth
- Thiel-Behnke
  - Autosomal Dominant
  - Later moderate visual loss
  - Corneal sensation normal
  - May present with RCE's



Reis-Buckler





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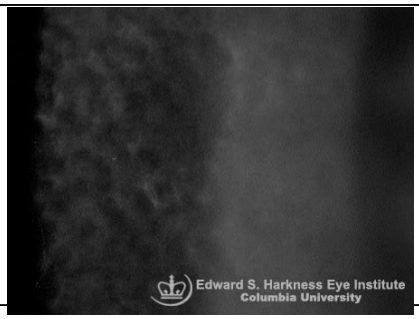
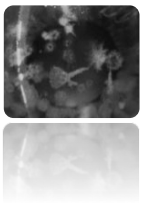
Thiel-Behnke Dystrophy



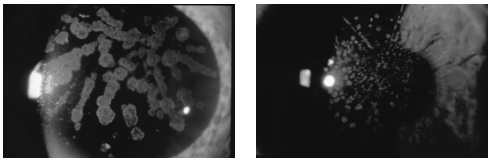
STROMAL DYSTROPHIES

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



GRANULAR DYSTROPHY



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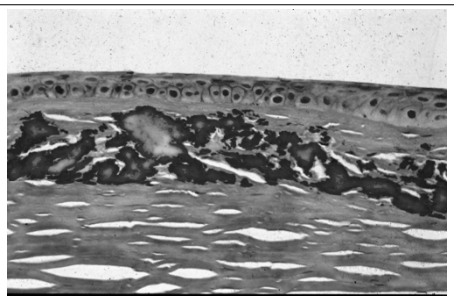
GRANULAR DYSTROPHY



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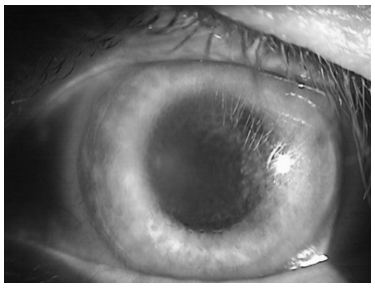
Granular Dystrophy



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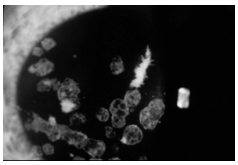
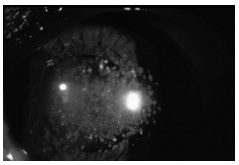
GRANULAR DYSTOPHY

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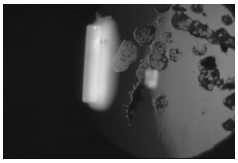
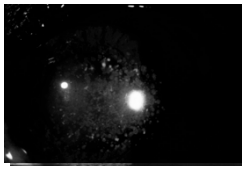
GRANULAR DYSTROPHY

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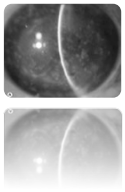
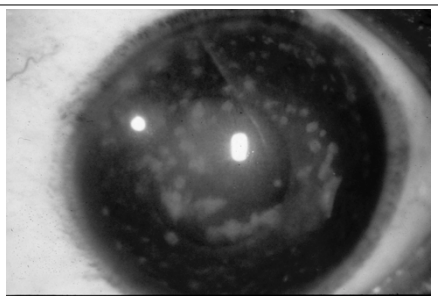
Macular Dystrophy (Groenouw Type II)

- 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



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Macular Dystrophy

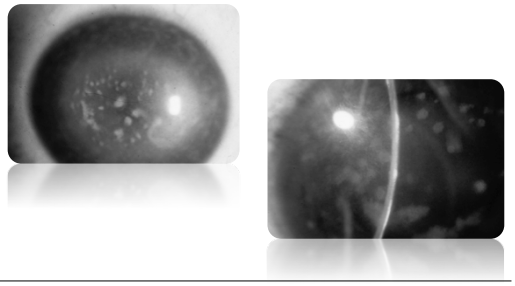
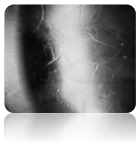


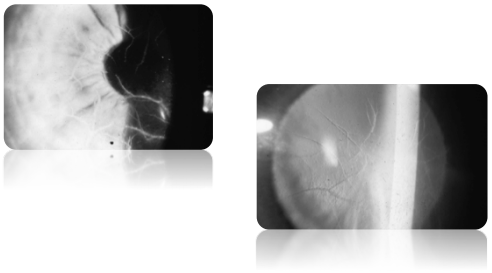
Photo Courtesy Tracy Swartz OD, FAAO

Lattice Dystrophy (Type I)

-
  - 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
-
-



Lattice Dystrophy



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Lattice Dystrophy

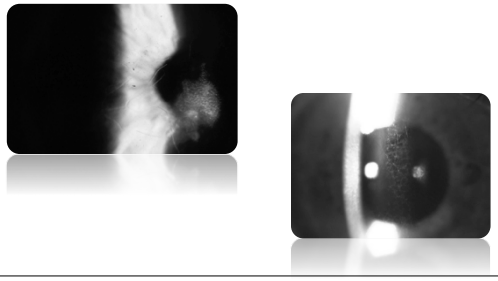
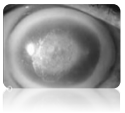


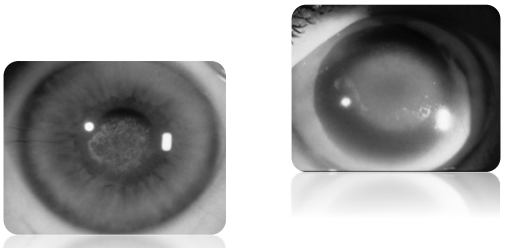
Photo Courtesy Tracy Swartz OD, FAAO

Central Crystalline Dystrophy of Schnyder

- Central discoid opacification posterior to Bowman's membrane in anterior stroma
- Opacities consist of:
  - Small needle shaped refractileals crystals
    - White
    - Polychromatic
  - May extend into deeper stroma-avoiding epithelium
-
- Associated with cholesterolimia



Schnyder's Crystalline Dystrophy



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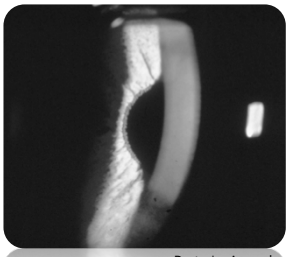
Other Stromal Dystrophies

- Avellino
- Gelatinous Drop Like
- Fleck
- Central Cloudy
- Posterior Amorphous



Posterior Amorphous

Photo Courtesy Tracy Swartz OD, FAAO

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

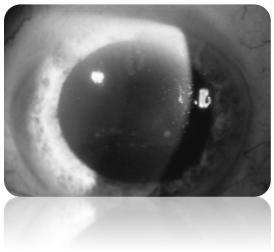
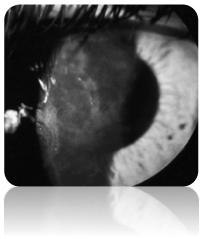


Photo Courtesy Tracy Swartz OD, FAAO

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 / Asymetry
- 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
    - Small refractile "drops" 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

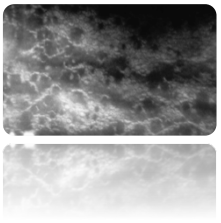
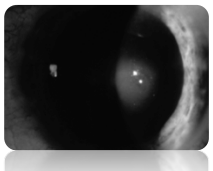


Photo Courtesy Tracy Swartz OD, FAAO

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.
-
  - Vision decreases
  - Possible bullous develops

FUCH'S DYSTROPHY



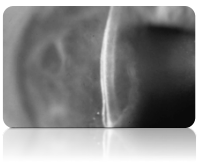


Photo Courtesy Tracy Swartz OD, FAAO

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)

Fuch's Dystrophy

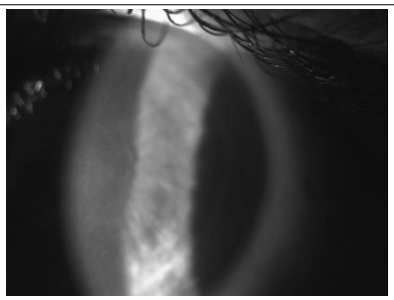


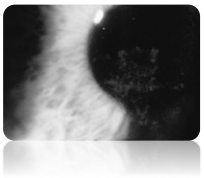
Photo Courtesy Tracy Swartz OD, FAAO

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

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



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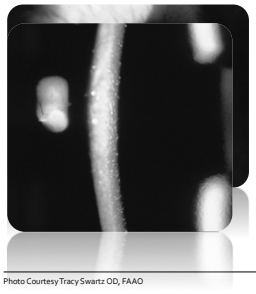
Congenital Hereditary Endothelium Dystrophy



- 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

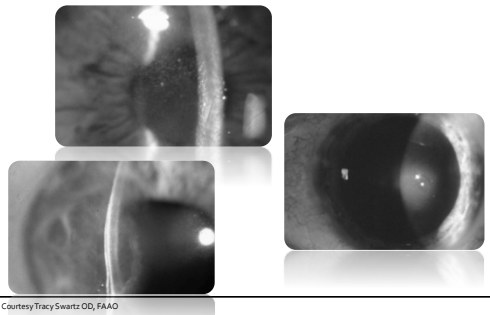


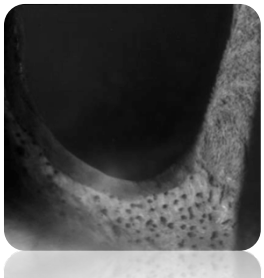
Photo Courtesy Tracy Swartz OD, FAAO

Iridocorneal Endothelial Syndrome

ICE

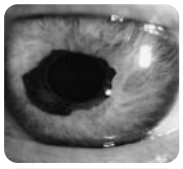
ICE

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



ICE

-
-
-
  - Increase IOP
  - Edema
-
  - Mild to severe atrophy
  - Nodules may be present
  - Glassy membrane on iris
- to treat



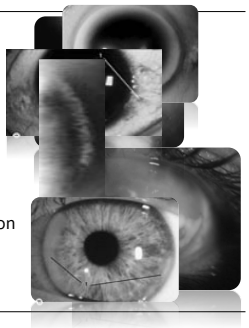
Corneal Degenerations

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

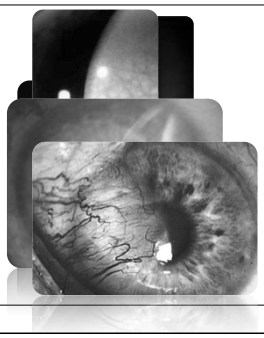
Degenerations

- Arcus
- Spheroidal degeneration
- Amyloid
- Limbal girdle of Vogt
- Band keratopathy
- Salzman's nodular degeneration



Degenerations

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



Ectatic Disorders

Keratoconus

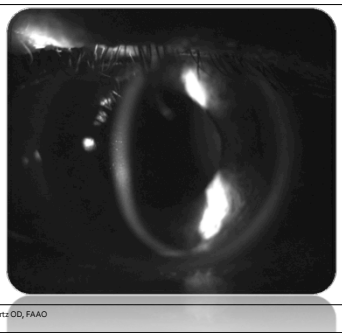


Photo Courtesy Tracy Swartz OD, FAAO

Keratoconus

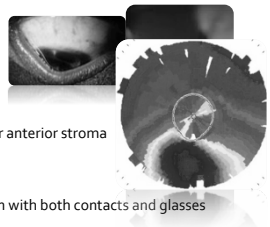
- Ectatic corneal dystrophy
- Bilateral with asymmetry
- Manifests in 20-30'ss
- Most likely a multigenic disease
  - Complex mode of inheritance
  - Environmental factors influence manifestation
-
  - Increased enzyme activities /decreased levels of enzyme inhibitors= toxicity
  - Destruction of normal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

Keratoconus

Photo Courtesy Tracy Swartz OD, FAAO

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

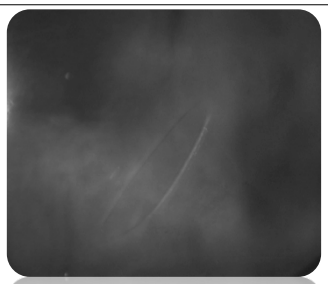
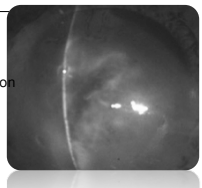


Photo Courtesy Tracy Swartz OD, FAAO

Keratoconus

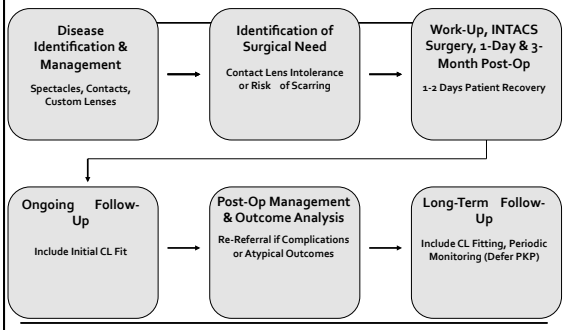
- Hydrops
  - Symptoms
    - Sudden decrease in best corrected vision
    - Foreign body sensation
    - Pain
  - Signs
    - Conjunctival hyperemia/redness
    - Prominent central or inferior corneal edema
    - Clouding
  - Self-limited in 8-10 weeks as endothelial cells regenerate at ruptured Descemet's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



Keratoconus

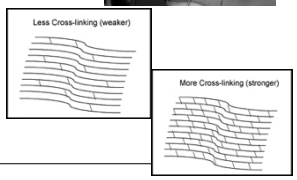
- Treatment
  - Hydrops
    - Hyperosmotics
    - Antibiotics to avoid secondary infection
    - PKP
  - RGP's
    - Bi-Aspheric I-Kone Design Valley Contax
    - Hyrokone
    - SynergEyes High Dk Hybrid

Keratoconus Treatment Flow
The New Paradigm



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

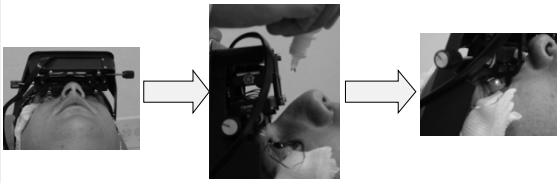
- rigidity
  - 329% human corneas
- Young's modulus
  - Increased 4.5X in human corneas

laboratory science

Stress-strain measurements of human and porcine corneas after riboflavin–ultraviolet-A-induced cross-linking

Wollensack G, Spoerl E, Seiler T. Stress-strain measurements of human and porcine corneas after riboflavin-ultraviolet-A-induced cross-linking J Cataract Refract Surg 2003; 29:1780-1785

Corneal Crosslinking with Riboflavin: CXL



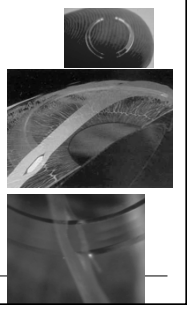
Corneal Crosslinking Clinical Applications

- keratectasia
- 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

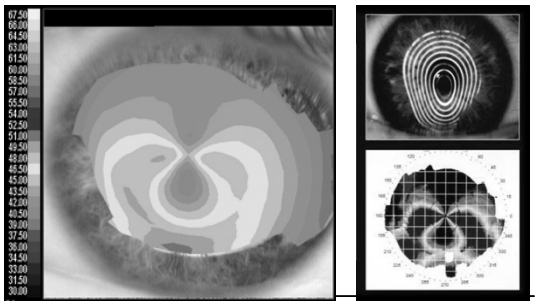
- Intracorneal ring segments
  - FDA approved for nearsightedness 1998
  - FDA approved under HDE 2004
- Provide structural support to thinned peripheral cornea
- Flattens cone
- Pulls cone toward center of cornea
  - Decreases irregular astigmatism



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

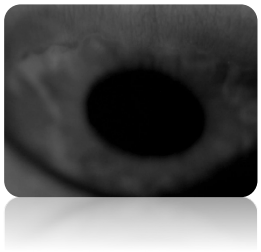
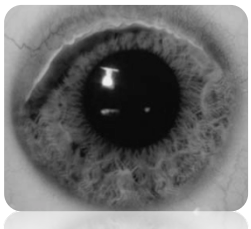


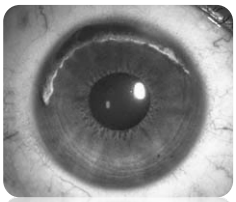
Photo Courtesy Tracy Swartz OD, FAAO

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

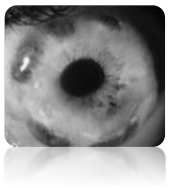
Terrien's Degeneration



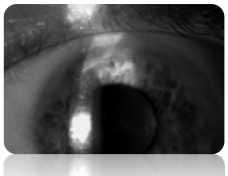


Terrien's Marginal Degeneration

- 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



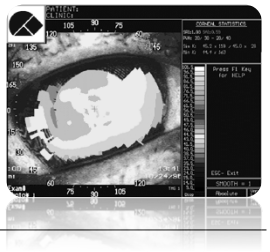


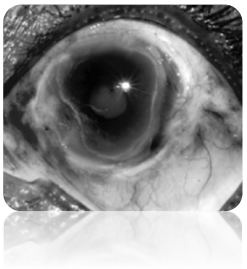
Photo Courtesy Tracy Swartz OD, FAAO

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

- Pathophysilogical 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 floroquinolone
  - Oral steroids
  - Conjunctival resection
  - Immunosuppressive therapy

Thank you