CORNEAL DYSTROPHIES AND DEGENERATIONS: DIAGNOSIS AND TREATMENT

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GOALS

- Differentiate dystrophy vs. degeneration
- Normal vs. abnormal
- Classify the disease by location
  - Layers of the cornea
  - Central vs. peripheral
- Determine appropriate treatment

Review the Layers of the Cornea

- Tear film 7-11 um
- Epithelium 50 um
- Epithelial BM <128 nm
- Bowman 8-14 um
- Stroma 500 um
- “Dua Layer” 15 um
- Descemet 5-10 um
- Endothelium 5 um

CORNEAL DYSTROPHY

- Rare conditions
- Slowly progressive, bilateral, central location
- Primary involvement of single corneal layer *
- Variable penetration and severity
- No associated systemic or ocular disease
- No sex predilection.
- Onset by age 20, stabilize by age 40 (except Fuchs)
- Autosomal dominant (50%)

CORNEAL DEGENERATION

- Non-familial, late onset
- Asymmetric, unilateral, central or peripheral
- Changes to the tissue caused by inflammation, age, or systemic disease.
- Characterized by a deposition of material, a thinning of tissue, or vascularization
Corneal Degenerations

From Periphery to Center (arbitrary division)

- arcus senilis
- lipid keratopathy
- white limbal girdle of Vogt
- senile furrow
- Terrien’s marginal degeneration
- Hassall-Henle bodies
- calcific band keratopathy
- calcareous degeneration
- spheroidal degeneration
- iron deposition
- Coats’ white ring
- crocodile shagreen
- corneal farinata
- Salzmann’s corneal degeneration
- corneal keloids
- corneal amyloid degeneration

GENETICS

- Most corneal dystrophies are autosomal dominant:
  - Heterozygous only one of the DNA strands effected
  - Homozygous more severe disease and recurrence in transplanted corneas is more prevalent.
- Autosomal recessive: 25% get it
- X linked: only men
- Not much has changed in the diagnosis of corneal disease however our increased understanding of the genetics has allowed us to classify better.

GENETICS

- Chromosomes 1,5,9,10,12,16,17,20,21
- Long arm of chromosome 5, 5q31
  - Gene codes for protein keratoepithelin which is involved in Bowman’s and stroma attached to Descemet’s layer
  - Gene codes for 683 amino acids
  - Lattice, Granular, Avellino, Reis-Bückler Dystrophies
- Discovering the pathways may aid in the use of drugs to interfere with the deposition of substances
- Many chromosomes explain the phenotypic variations. Any change of sequence in the amino acid chain can cause variations of the disease

EPITHELIUM

- 50 um non-keratinized stratified squamous epithelium
- 5-10 layers central 8-10 peripheral
- Superficial layers have microvilli that attach tears.
- Exfoliation q 5-7 days
- Deeper layers (Basilar Columnar cells) have hemidesmosomes
  - connect the epithelium to basement membrane which connects to Bowman’s Layer.

EPITHELIAL BASEMENT MEMBRANE “DISORDER” EBMD

- Most common anterior corneal disorder
- DYS: inherited, single layer, bilateral
- DEG: Prevalent 43%, 25% unilat,> 29y, trauma
- Abnormality of epithelial turnover, maturation, and production of BM and adhesion complexes
- Thickened BM ultimately weakens the epithelium and causes recurrent corneal erosion (RCE).

EBMD

- The basal cells produce abnormal finger-like projections that bend intra-epithelially and trap cells/debris that form cysts.
- MAPS : multi-lamination of BM and collagen
- DOTS: grey opacities, cysts
- FINGERPRINT:
  - reduplication of BM
### SLX of EBMD
- Negative NaFL pattern and instantaneous TBUT
- No Rose Bengal Stain
- When Microcysts surface and erupt, + NaFL
- Asymptomatic vs. Variable degrees of Blur, diplopia, photophobia, dryness, FBS, or pain.
  TX: Lubricants, hypertonics

### TREATMENT FOR EBMD
- Indicated if vision or comfort are compromised.
- Manage co-existing ocular surface disease
- Environment/diet
- Lubricants
- Punctal occlusion
- Bandage Contact Lens (BCL)
- Surgical: PTK

### LUBRICANTS
- Avoid preservatives or surfactants
- Electrolytes nourish eye
- Avoid bland ointments: hypo-osmotic and retain fluid
- Hyperosmotic agents
  - Muro 128: Solution (2-5%) vs. ointment (5%)
  - Ung: comfort, > concentration
  - Treat 6 weeks Soln/3-6mo ung
- Warm Packs: QID 2-3 weeks

### What’s New in Juice FreshKote
- RX only Artificial Tear
  - Patents, concentration, monitor
- Treats all 3 layers
  - Amisol Clear restores lipid
  - 2% polyvinyl pyrrolidone and polyvinyl alcohol improve both aqueous and mucin layer
- Polixetonium preservative with low toxicity, anti-microbial, anti-fungal, & facilitates wetting
- High oncotic pressure
  - Reduces edema, establishes epithelial integrity and may prevent damage
- 3-6 x per day, OK with CLS
- Focus Laboratories

### ALTERNATIVE DROPS
**N-ACETYL CARnosine**
- “inactive ingredient”
- Visual Ocuity®: A Professional Product from Longevity Science®
- Can-C, International Anti-ageing Systems, UK
- HPMC 0.3% and Glycerin 1.0%
- Anti-oxidant compound combined with CMC
- Carnosine penetrates the lipid membrane of the lens to reduce opacification
- Improves VA/glare

### Autologous Serum Drops
- Utilizes patient’s own blood serum
- Blood is drawn and the serum is spun down and mixed with artificial tears. Devoid of cells and clot factors
- Replaces individualized growth factors
- Replaces individualized antibodies
- Serum contains growth factors, fibronectin, Vit. A and anti-proteases
- Requires blood donation 2-3 times year $150-$300 Hospital/Lieters
- Consider 5-10% serum albumin drops qid instead
PUNCTAL OCCLUSION: THE IDEAL PLUG

- Easy and comfortable to insert
- Negligible corneal contact, no sensation
- Visible upon inspection only
- Reversible: easy to remove by a professional
- Inert material with no allergic response
- Effective in the treatment of dry eye
- Responsibility = Consent
- Increase contact time of natural or artificial tears on the eye.

Superficial Punctate Keratitis of Thygeson (SPKT)

- Chronic, usually bilateral disorder characterized by central focal epithelial lesions and no stromal involvement.
- Fine or dense/ Single or Multi Avg of 15-20 lesions (1 to 50)
- Corneal sensation not effected although occasional hypoesthesia
- Conjunctiva is not inflamed*

SPKT

- NaFl/RB staining and elevated during active disease process
- Each lesion comprised of multiple lesions
- Change position over time
- Conjunctiva: usually not inflamed unless during the developmental stage: 1-2 wks

Etiology of SPKT Unknown

- Possibly Viral due to latency, recurrence, lesion appearance, duration
- PCR testing proved that it is NOT HSV 1 or 2, VZV or adenovirus
- Still investigating HPV since both have minimal inflammation
- Prolonged SPKT Associated with Salzmanns Nodular Deg.
- Suggested association with eczema, urticaria, asthma
- HLA-DW3 and DR3 association:
  gluten sensitive, DM2, Lupus, Graves

Etiology of SPKT Unknown

- R/O etiology of
  - Punctate epithelial erosions PEE vs.
  - Sub-epithelial infiltrates SEI vs.
  - Superficial Punctate keratitis SPK

“Pink EYE”

STANDARD TESTS

- No testing done – expensive, time consuming
- Diagnosis based on hx/exam
- Misdiagnosis ~ 50% of cases
- Most often, treated empirically
- Antibiotics – between 95%-99% of all cases
- Steroids – may pose risk in misdiagnosis
- “If antibiotics are not effective, it must be viral.”
- Other bacterial infections, such as Strep, use a confirmation test.
  - FDA Cleared
  - CLIA Waived
  - 10 minute results
  - Detects all 51 serotypes of adenovirus
  - 33%-40% of all acute
  - 80%-90% of viral
  - CPT Code 87809
  - Cost $13 Reimburse $17
SPKT
- Mean age 29 (2 to 70)
- Usually Bilateral
- Favor the central visual axis
- Long duration with remissions and exacerbations
- Asymptomatic (esp. later) vs. FBS, epiphora, photophobia
- Treat the symptoms

Treatment for SPKT
- Lubricants for comfort
- BCL to smooth surface
- Lack of response to systemic or topical AB, debridement/ cautery of tissue
- Good response to steroids however long taper and can prolong the course or worse
- Antivirals?
- Cyclosporine
  - Reinhard showed 70% suppression with 2%

MEESMAN’S DYSTROPHY
- Diagnosed within first year of life
- A "peculiar" substance is produced which thickens BM
- Numerous epithelial vesicles that extend to limbus*
  - Contain debris, cells, GAG
- No scarring. Photophobia. Irritation
- May have slight decrease in VA.
- CLS are not contraindicated and may be therapeutic when rupturing
- LISCH: whorl-shaped clusters

RECURRENT CORNEAL EROSION
- Traumatic erosions due to thickened BM with poor hemi-desmosomal attachments.
- May result from incomplete healing following trauma
- Associated with EBMD (50%) or Lattice Dystrophy
- Onset in the am due to edema or shearing effects
- Symptoms may be more severe than it appears
- Epithelial loss surrounded by pooling and loose ends
- Prophylaxis
  - Treatment: lubricants/ plugs/BCL

Treatment for RCE
- Prophylaxis with lubricants/hyperosmotic agents/BCL
- Treat like a corneal abrasion: heals slower
- Debridement
- Anterior Stromal Puncture
- PTK with PRK

BANDAGE CONTACT LENSES
- To aid in healing by offering protection
- To provide comfort for decompensating corneas with erupting microcysts
- To aid in dehydration
- To produce a more regular refracting surface
- To aid in drug delivery
- To reduce inflammation
INDICATIONS FOR BCLS

- **ACUTE**
  - Traumatic abrasion
  - Following FB removal
  - RCE
  - Chemical burns
  - Thermal Burns
  - Shield Ulcer

- **CHRONIC**
  - Severe dry eye
  - Bells Palsy exposure
  - Cicatrical disease
  - Nocturnal lag
  - Conjunctival elevations that reduce wetting
  - Whorl Keratopathy

INDICATIONS FOR BCLS

- **SURGICAL RESULT**
  - Retinal surgery causes epithelial defects
  - PRK
  - PTK

- **DISEASE**
  - Thygeson’s
  - Salzmann’s
  - Granular Dystrophy
  - Lattice Dystrophy
  - EBD
  - Bullous
  - Band Keratopathy
  - Piggyback

- **DISEASE**
  - RGP induce abrasions for ectasias

CONTRAINDICATIONS FOR BCL

- Non-compliant patient
- Poor Hygiene
- Socio-economic
- High risk for infection
- Non-consent

Faster Recovery with BCL

- Donnenfeld reported in A.Ophthalmology 1997
- Compared patients treated with:
  - Pressure patch /AB vs. BCL vs. BCL/Topical NSAID
- No difference in re-epithelialization time
- Psychometric Analysis: patients prefer BCL/NSAID
- Return to normal activities in 1.37 days
- Soak lens in ANTIBIOTIC
  - Caution with preservative toxicity, especially BZK
- Other options: Collagen Shields

GOALS IN FITTING BCL

- CL should have smooth surface
- Minimal ET
- Wettability
- High dK
- High modulus when lid edema is present
- Full coverage, minimal movement

- **HIGH Water** = provides mechanism for dehydration and slower drug release.
- **LOW Water** = when evaporation is not desired.
- Minimal movement to avoid rupture of hemi-desmosomal bonds. Complete coverage.
- Disposables / EW/low ET

FDA Approved vs. STD of CARE

- Cooper Vision PermaLens Therapeutic
  - 71% H2O dK 34
- Bausch & Lomb Plano-T
  - 38% H2O, dK 9.2
- CIBA Focus Night & Day
  - 24% H2O, dK 140
- CIBA CSI-FW
  - 38% H2O, dK 13
- B & L Purevision
  - 36% H2O dK 101
- Acuvue
  - 58% H2O dK 28
- Acuvue Advance/Oasys
  - 47% H2O dK 60/103
- B&L Soflens
  - 66% H2O dK 32
- Cooper ProCLear Compatibles
  - 62% H2O dK 34
INFECTION PROPHYLAXIS

- Erthromycin ung or Bacitracin ung q 2-4h or
- Polytrim gtt +/- qid
- Tobramycin +/- qid
- Ciloxan/ Ocuflox qid
- Zymar/ Vigamox qid
- Submerge BCL
- DOSAGE & TOXICITY

FOLLOW-UP CARE FOR BCL

- 24 Hours
  - May note 25-50% improved
- If improvement q 2-5 days
- Monitor high risk patients daily
  - CL wearers
  - HSV, Immuno-compr, DM
  - Monocular, children
  - Central or Large abrasion
- Do not remove BCL too early- wait 5-7 days until after it appears to be resolved- late phase healing
- If condition worsens or no improvement, consider referral for tarsorrhaphy or conjunctival flap

ALTERNATIVES TO BCL

- TARSORRHAPHY
  - Surgically close the palpebral fissure by suturing the superior and inferior lids at the lateral aspect
- STAMLER LID SPLINT
  - Adhesive on one side with enough rigidity on the other to hold the eyelid in the closed position
  - Allow for use of meds and examination

AMNIOTIC MEMBRANE

What does PROKERA® do?
- Helps the eye heal: The amniotic membrane tissue in PROKERA® has natural therapeutic actions that help damaged or inflamed eye surfaces heal faster. Eyes treated with PROKERA® have quicker healing, less pain, less scarring, and less inflammation.
- Protects the eye surface: The amniotic membrane in PROKERA® is thin and clear like the tissue on the surface of your eye and protects your eye’s damaged tissue while inserted.

USES OF PROKERA

- What does PROKERA® treat?
- PROKERA® is used by physicians all over the world to treat eye diseases such as recurrent corneal erosion, infectious/inflammatory keratitis, Herpes, superficial epithelial defects, severe dry eye syndrome, and other corneal diseases.

PREVENTION OF RCE

- Patients with RCE show a chronic increased level of metallo-proteinase enzymes (MMP 2&9) which may dissolve the basement membrane and fibrils due to inadequate neutralization.
- Treatment is to inhibit metallo-proteinases
  - Doxycycline: oral, 50mg BID
    - 2 months treatment time. Reduced MMP 70%.
- Topical Steroids
  - Pred Forte, FML, Lotemax, TID, 2-3 weeks
- No recurrences in 21 months.
Doxycyline Hyclate
- 20 mg Doxycyline Hyclate
- Sub-antimicrobial dosage
  - (<50mg)
- Enzyme modulation of inflammation
- By OCuSOFT
- Kit comes with lid scrub foam
- Claims to be a more potent collagenase inhibitor than minocycline and therefore less SE
- Long term use

ANTERIOR STROMA MICROPUNCTURE
- Disturb Bowman’s Layer to promote tighter adhesion and stimulate cornea to produce functional BM complexes
- Topical anesthetic and a 27g cannula: use forceps to bend needle to avoid puncture
- Closely spaced (.5mm) punctures damaged/adjacent
  - Anterior Stroma :100-150 u
  - Apply tangential force
  - Avoid Visual axis since minimal scarring can occur
  - RR 40%

CORNEAL DEBRIDEMENT
- Soften epithelium
  - 1-2 gtt topical anesthetic q 15-30 seconds for 2-3 minutes
- Use cotton swab, spatula, spud or jewelers forceps
- Remove flaps by pulling edges toward center
- Don’t pull directly up or out
- Remove flaps down to tight, firm edges.
- Tx abrasion (>50-100%)
  - Recurrence Rate 18%

Bowman’s Layer
- Acellular modified layer of anterior stroma
- Type 1 collagen fibers randomly arranged
- Pores for corneal nerves to pass
- Fxn? Not found in many species with good vision and normal epithelial-stroma junctions.
- Not replaced and when damaged, causes significant opacification which effects VA

REIS-BUCKLER DYSTROPHY
- Bilateral, symmetric, AD, by age 5
- Bowman’s layer is obliterated and replaced with randomly arranged regular collagen that thickens.
- Linear, ring-like or “Honey comb”
- Marked VA loss due to superficial stromal haze or topographical changes from elevation of tissue
- Painful if recurrent erosions occur.
- TX: PKP or LK around age 50 but may recur in grafts

ANTEORIER MOSAIC
- Dystrophy or Degeneration
- AKA: Anterior Crocodile Shagreen
- Breaks in Bowman’s that appear like central grey polygonal opacities with clear spaces.
- Blanches with limbal pressure.
- Asymptomatic
**BAND KERATOPATHY DEGENERATION**

- Deposition of Calcium salts in Bowman’s layer
- Located interpalpebral region
- History of uveitis, renal failure, prolonged use of miotics, syphilis, interstitial keratitis, hyperparathyroidism
- TX: Chelation with EDTA 1%
- TX: Therapeutic CL

**Treatment: Cosmetic Contact Lens**

- Black Underprint: color is applied to a dark background to mask and make a scar more uniform. This darkens and mutes the overlaid color.
- Store in glass vials
- 53% H2O to maintain dye

**SALZMANN’ S NODULAR DEGENERATION**

- Bluish, superficial nodular elevations
- Inflammatory/Non-inflammatory event that exposes the cornea and results in excess COLLAGEN plaques that replace Bowmans
- Post-chronic-keratitis
- Asymptomatic to very painful and sight threatening depending on location and severity
- TX: BCL/AB/NSAID, SCLERAL PTK,PKP

**STROMAL DYSTROPHYS**

- 90% of corneal thickness
- 22%: Comprised of collagenous lamellae (type 1) interspersed with keratocytes and ground substance (proteoglycans, glycoproteins, serum)
- GAGS: affect hydration, thickness, transparency
- 78%: rest is water.
- Abnormal Substance found within the cells or fibrils that have distinct histological-stains

**Name of Dystrophy**
**Name of Deposition**
**Pathology Stain**

- **Marilyn Monroe Always**
- **Gets Her Man**
- **Los Angeles County**
- **Southern California Ocean**

**MACULAR DYSTROPHY**

- Clouding due to build-up of mucopolysaccharides
- Begins centrally & superficially then extends limbus to limbus thru all layers
- Thinning without clear spaces
- Primary involvement of the endothelium: guttata*
- Begins in 1st decade of life: aggressive causing early & severe VA loss
- Predominant in Virginia area
- Autosomal recessive*
- TX: PKP
- Macular / Mucopolysaccharide / Alcian Blue stain
**GRANULAR DYSTROPHY**

- Central, anterior to mid-stromal deposits of Hyaline
- AD
- Discreet white spots (translucent) to powdery rings
- Clear areas between lesions in early years
- Erosions can break thru BM.
- Autosomal dominant w/ complete penetrance*
- Granular / Hyaline/ Masson Trichrome

**GRANULAR DYSTROPHY TREATMENT**

- Pinhole effect may maintain VA (20/20) until the lesions coalesce and reduce VA=20/200.
- PKP was only treatment and recurrences were common
- Present treatment includes PTK and BCL:
  - Smooth epithelial surface to treat monocular diplopia
  - Pain management following PTK or erosions
  - Induced anisometropia
  - Spectacle distortions of high plus lens

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**Granular Dystrophy**

70 YO AAF
- 1992 VA 20/50
- 1997: 20/80 & RCE
- PKP vs. PTK
- SRX pre: +1.00
- K: 42.00/41.00
- SP 2 mo: +8.75
- K: 36.75/ 37.75
- SP 6 mo: +6.50
- TX: Acuvue +7.00

25 YO WF
- C/O anisometropia/haze
- RE +7.50 i 2.00 x 010 20/30
- LE +.25 i 2.25 x 170 20/30
- CL FIT
  - DIL +3.50 8.00/11.2 20/25
  - PV pl -1.75 x 180
- Refit OD at 4 months pg
  - Hydrasoft Options
  - +8.75 -2.00 x 010 20/25 !!!

**LATTICE DYSTROPHY**

- Branching refractile filaments of AMYLOID
- Symptoms occur early in life, age 20-30, AD
- RCE are common
- Resultant scars and late intervening haze can blur VA
- Lines thicken with age & penetrate deeper layers
- Autosomal Dom/ Recessive
- TX: PKP
- Lattice / Amyloid / Congo Red

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**TYPES OF LATTICE DYSTROPHY**

- TYPE 1
  - Poor VA by age 40-60
- TYPE 2
  - Merotoja syndrome
  - Bilateral Facial palsy, skin thickens,
  - Depressed brows, prominent
  - VA loss >65y
- TYPE 3
  - Floppy ears, protruding lips, Auto-R
  - Lger deposits, mid stroma, no RCE
  - VA loss > 60 y

**AVELLINO DYSTROPHY**

- Avellino, Italy
- Typical granular dystrophy with axial anterior stromal haze and mid-stroma discreet linear opacities.
- Congo red
CENTRAL CRYSTALLINE DYSTROPHY OF SNYDER
- Deposits of cholesterol crystals in anterior stroma
- Premature peripheral arcus
- Vision is generally good
- Usually normal serum lipid profile: +/- hyperlipidemia
- Expressivity is variable
- B120 gene on chromosome 1 is responsible for lipid metabolism and transport

WHITE LIMBAL GIRDLE OF VOGT
- Effects >50% population between 40-60
- With/Without clear zone
- Represents subepithelial degeneration and sometimes calcium deposition
- Does not affect visual acuity
- Located in the horizontal meridian

POSTERIOR EMBRYOTOXIN
- Extremely prominent ring of Schwalbe
- Eye is normal but may be associated with correctopia, aniridia, or corneal conditions that are part of systemic syndromes

ARCUS SENILIS
- Effects >60% population between 40-60 years
- Peripheral lipid deposition
- Located anterior to Descemet’s layer and in Bowman’s layer
- Juvenile form usually represents hyper-lipidemia
- Be suspicious of carotid disease if this is present to a greater degree in one eye.

FURROW DEGENERATION
- Peripheral thinning in the elderly
- Lucid interval of Arcus
- No inflammation
- Vision unaffected

DELLEN
- Peripheral 50% thinning of one or more layers
- Runs along the limbus parallel to area of swelling
- Limbal elevation causes dryness which leads to further excavation
- CL, lid disease, OSD
### TERRENIUS MARGINAL DEGENERATION
- Intact epithelium with progressive thinning
- Non-inflammatory
- Supero-nasal location
- Attacks young men (3:1)
- Produces AR or oblique astigmatism seen on topography
- Treat irregular astigmatism with RGP

### ANTI-INFLAMMATORY THERAPY
- **CYCLOSPORINE**
  - .05% and .1%
  - Reduction in artificial tear use
  - Increase in goblet cell density
  - Decrease in corneal staining
  - Improved Schirmer test scores
  - Improved visual

### RESTASIS® ALLERGAN
(Cyclosporine Ophthalmic Emulsion 0.05%)
- Indicated for patients who do not get relief with compresses and lubricants
- Restores tear production
- Increases goblet cells
- Excellent safety profile
- BID dosing
- Mild stinging

### STEM CELL DEFICIENCY
- Defects in renewal and repair causes invasion of conjunctival epithelial cells onto the cornea
- **SIGNs:**
  - Dull corneal reflex
  - Ingrowth of thickened fibrovascular pannus
  - Keratitis
  - Scarring
  - Calcification

### FUCH’S SUPERFICIAL MARGINAL KERATITIS
- Affects middle aged adults
- It is characterized by periods of relapses and remissions of irritation and redness.
- Begins as superficial marginal keratitis that advances non-uniformly sparing the central cornea.
- Advancing keratitis is demarcated from the central cornea by a gray line.
- Active keratitis is accompanied by stromal infiltrates

### FUCH’S SUPERFICIAL MARGINAL KERATITIS
- Chronic disease leads to progressive circumferential peripheral corneal thinning with vascularized pseudopterygia growing over these areas.
- Histopathologic studies shown corneas to be thinned 20-25% in the periphery.
- Inflammatory cells in the cornea consisted of mostly lymphocytes and PMN but also mast cells & basophils.
- BV leaking lipids
- These studies suggest no clear cut etiology of the disease.
TREATMENT FUCH’S SUPERFICIAL MARGINAL KERATITIS
- A.T., topical steroids, and topical antibiotics during acute exacerbations
- Topical Cyclosporine 1% BID
- Fit RGP contact lenses (Improve Vision)
- Lamellar Keratoplasty (reports of recurrence in the graft)
- Combined superficial keratectomy with a conjunctival autograft. (Kotecha and Raber)
  Method used to retard recurrent pseudopterygium formation

Normal Changes to the Endothelium
- Descemet’s layer thickens from 3-17 μ
- There is a decrease in the # of endothelial cells
  - from 3500 cells/mm² to 1200
  - this single layer spreads out: lacks mitosis
- High density mitochondria : 90% pump
- Lenses produce reversible polymegathism

Abnormal Changes to the Endothelium
- Endothelial cells become more irregular
- Cells secrete collagen towards Descemet’s causing multilamination = guttata
- This breaks down the barrier function and results in stromal and epithelial edema

POSTERIOR POLYMORPHOUS DYSTROPHY- PPMD
- Can be present at birth
- Wide variety of expression
  - Non-symptomatic
  - Grouped vesicles cause blur
  - Stromal edema
  - Correctopia and irido-corneal adhesions resulting in glaucoma if they enter TM

POSTERIOR POLYMORPHOUS DYSTROPHY- PPMD
- Isolated to coalescent vesicles that intervene between normal endothelial cells.
- Areas of normal or thickened Descemet’s membrane representing a collagenase material
- These vesicles can lead to stromal edema.
- Association with keratoconus

FUCH’S DYSTROPHY
- Bilateral, asymmetric, begins in 5th or 6th decade
- More predominant in women (3x)
- Initially pigment dusting
- Non-symptomatic

Guttata represent clear, vesicular endothelial secretions that project into the potential space between the endothelium and Descemet’s I
**FUCH’S DYSTROPHY STAGE 2**
- Guttata interrupt the normal pumping mechanism = edema
- Edema begins around Descemet’s and Bowman’s layers and then spreads the entire thickness.
- Pts experience glare/hazy VA
- Bullae appear: they reduce vision and cause pain when they rupture, especially in am

**FUCH’S DYSTROPHY STAGE 3**
- Edema is reduced but subepithelial connective tissue grows and causes reduced vision.
- Patient is comfortable due to reduced corneal sensitivity.
- Elevated IOP, peripheral neovascularization, and corneal erosions.

**FUCH’S DYSTROPHY TREATMENT**
- Hypertonic solutions to draw fluid out
  - Sodium Chloride
  - Muro 128 (2% or 5%) solution, 5% ointment-PF
  - Fresh Kote
- BCL to aid in comfort for ruptured bullae
- Lubricants for comfort
- Lower IOP
- Conjunctival flap
- Corneal transplant to restore vision/ DSEK

**DSEK: Descemet Stripping Endothelial Keratoplasty**
- Faster visual recovery
- Less astigmatism created since there are no sutures
- Eye is much stronger and more resistant to injury since only the diseased tissue rather than the entire cornea is replaced
- Surgery time is quicker
- Chance of rejection is reduced
- Procedure can be combined with cataract surgery
  - VA 20/30-20/40
  - 1-2 D Hyperopic Shift, thicker

**Visante Applications**
- Anterior Segment Imaging and Surgery
  - Corneal Imaging and Measurement
    - imaging and evaluation of corneal pathologies
    - penetrating keratoplasty
    - lamellar keratoplasty
    - endothelial keratoplasty
    - keratoconus imaging and assessment
    - anterior segment imaging through opaque corneas

**Refractive Surgery**
- Corneal laser refractive surgery: pre-op, enhancement options
- Phakic IOLs
- Corneal refractive implants: Intacs

**Anterior Segment Imaging and Surgery**
- Corneal Imaging and Measurement
- Iris Imaging and Evaluation
- Trauma Assessment
Keratoconus is a clinical term to describe a condition in which the cornea assumes a conical shape because of thinning and protrusion.

Keratoconus - Keratometry
- Initially, mires get small and then there is a lack of parallelism
- Expand perimeters by use of +1.25 SPH and add 7 D to your reading
- Steepening begins infero-temporally and progresses clockwise
- TOPOGRAPHY - more sensitive
- PLACIDO RINGS - get closer

Retinoscopy
- Scissors Reflex
- Against motion that breaks apart
- Represents multiple refractive powers within the optic zone

Pseudokeratoconus
- Corneal warpage topography can mimic KC
- Repeat topography must be performed and a measurable change would indicate pseudo-KC
- Evaluation of elevation maps at steep zone:
  - Predicts the elevation or depression of the cornea if the best fit sphere was on cornea

Keratoconus - Slit Lamp Findings
- Fleischer Ring
- Vogt’s Striae
- Stromal Thinning
- Stromal Scars
- Swirl-like Pattern
- Enlarged Corneal Nerves
- Acute Hydrops

Pellucid Marginal Degeneration
- 20-40yo, no gender preference, slow progression
- Thinning occurs below the steep curvature
- Stromal thinning is concentric to the lower limbus and runs from 4-8:00, 1-2mm wide
- Clear, epithelialized, and non-vascularized.
- Absence of lipid: ddx from Moorens or Terriens
- Vertical stress lines and hydrops can occur
- Beer-belly cornea-
PROGNOSIS FOR PELLUCID

- Lens fitting is difficult due to inferior apex
- Central rings show AR/Inferior rings show WR
- Fitting flat causes bearing and on K (steep) causes too much seal off
- Larger lenses needed due to low positioning/glare
- CAREFUL MONITORING-
- Poor SX Candidate

KERATOLOBUS

- A diffuse thinning of the cornea to 1/3-1/5 the normal thickness
- It is noted early in life and progression is minimal
- Associated with Ehlers-Danlos Syndrome and Leber’s Congenital Amaurosis
- Acute hydrops

POSTERIOR KERATOCONUS

- Rare developmental defect
- Focal indentations of the posterior cornea with overlying stromal scarring
- Anterior curve not effected
- Descemets’ membrane and endothelium are always present but may be abnormal in the area of thinning

associated Ocular Disease
- Lens abnormalities, choroidal or retinal sclerosis, PPMD, retinal coloboma, optic nerve hypoplasia, ptosis, iron rings, and posterior synechia,
- Systemic Associations
  - Mental retardation, webbed neck, hypertelorism, short stature, superior placed lateral canthi, genitourinary abnormalities

POSTERIOR KERATOCONUS

Thank you
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