UNVEILING THE WINDOW TO VISION:
UNDERSTANDING THE CORNEA, ITS DISEASES, AND TRANSPLANTATION

Dr. Kristina V. Thomas, MD
Cataract, Cornea, and Refractive Surgery

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Goals

1. Gain a comprehensive understanding of the cornea
2. Explore common corneal diseases and treatment options
3. Understand the different types of cornea transplantation

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Disclosures

- NONE

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Introduction

- Transparent, avascular tissue
  - Glucose from aqueous
  - Oxygen from tear film and limbal vessels
- 11-12mm horizontally and 10-11mm vertically
- One of body's highest densities of nerve endings
  - 100x as sensitive as conjunctiva
- Refractive index: 1.376
  - *1.3375 is ued in calibrating keratometers to account for combined power of the anterior and posterior corneas
- Average radius of curvature: 7.8mm (6.7 – 9.4mm)/ 43.25D
- Major source of astigmatism in the optical system

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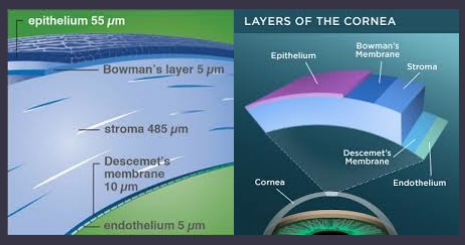

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Epithelium

- Stratified (5 or 6 layers) of squamous non-keratinized epithelium
- Superficial cells → flattened
- 50-60 μm
- Tight junctions in superficial cells prevent tear fluid from getting into stroma
- Continuous proliferation of basal cells at the limbus
  - Subsequently differentiate into superficial cells in 7-14 days

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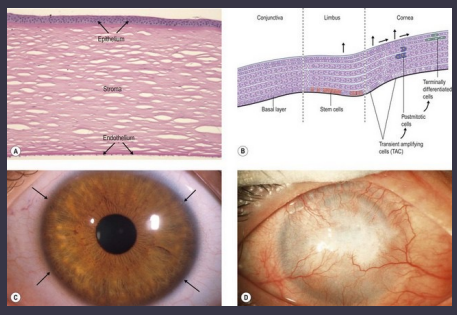
Tissue Engineering for Reconstruction of the Corneal Epithelium
Ursula Schlötzer-Schrehardt, Naresh Polisetti, Johannes Menzel-Severing and Friedrich E. Kruse



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Anatomy and Vision

- Crisp Vision
  - Requires a smooth corneal surface
  - Healthy tear film and epithelium
  - Tight packing of epithelial cells
    - Produces uniform refractive index
    - Minimal light scattering

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Bowmans Layer and Stroma

- Bowman's Layer
  - Acellular
  - Does not regenerate
  - located between the epithelial basement membrane and anterior cornea stromal
  - Precise function remains unknown
  - Removed in PRK patients
- Stroma
  - Thickest layer
  - Matrix of collagen and proteoglycans type I, V, VI
    - Concentrations vary from anterior to posterior stroma
  - Completely transparent
    - Lattice arrangement of collagen fibrils in extracellular matrix
    - Acts as a diffraction grating to reduce light scattering by destructive interference
    - Size of lattice elements < the wavelength of visible light

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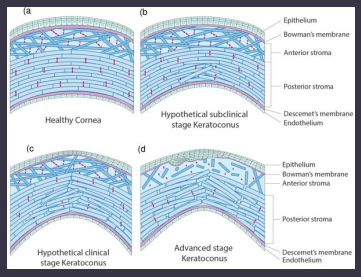
Biomechanics of keratoconus: Two numerical studies
Nicolas Falgayrettes,Etienne Patoor,Franck Cleymand,Yinka Zevering,Jean-Marc Perone. https://doi.org/10.1371/journal.pone.0278455



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Endothelium and Descemet's

- Single layer of flat hexagonal cells
- Forms boundary between Stroma and AC
- Cell loss results in cells enlarging/ and spread of neighboring cells to cover the defective area
  - Does not regenerate
- Responsible for pumping fluid out of the stroma to prevent edema
- Descemet's Membrane
  - Basement membrane of endothelium
  - Increases in thickness from birth
  - Allows entry of nutrients and macromolecules into the stroma
  - Regenerates

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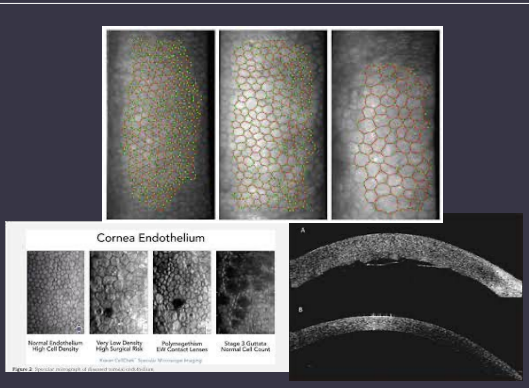

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Examination of the Cornea

- Question:

What is the most important test for cornea patients?

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Examination of the Cornea

- Answer: ***Visual Acuity***

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Examination of the Cornea

- Must **push** the patient to get accurate acuity
- Unlike Retina and Glaucoma specialists, Cornea specialists do not have FA or OCT to diagnose and rule-out disease
- Just a small (less than 1mm) change in cornea can cause no visual symptoms at all or decrease VA to 20/400.
  - Location
  - Etiology
  - Severity

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Examination of the Cornea

Example:

- Pt with dry ARMD comes in for dry eye f/u and VA is decreased from 20/20 OU (last visit) to 20/40 OD and 20/60 OS
- What is the etiology of the decreased Vision?
  - Change in refraction? What is the PH VA?
  - Dry eye worsening?
  - Retina etiology? Is dry AMD now wet?
  - Visual acuity inaccurate?

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Examination of the Cornea

- Exam: dryness IMPROVED
- PH VA was not done
  - PH very important as helps Cornea specialist to r/o retinal etiology
  - If pt PH to 20/20, either refractive error or cornea issue
- VA was repeated and pt was "pushed" by having them blink repeatedly and a drop of AT
  - VA 20/20 OU
  - Therefore no need to dilate patient
- Had VA not been rechecked, pt would have been dilated unnecessarily to look for retina etiology.

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Visual Acuity for Cornea

- Need to push Cornea patients when testing VA
  - Remind them to blink
  - Place Artificial Tears
  - Compare VA to last visit VA to decide next step

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Examination of Cornea

- Fluorescein Stain
  - Non-toxic, water-soluble, orange dye
  - Assesses the viability of corneal epithelium using cobalt blue filter
  - Prefer placing with paper strip to control amount placed into the eye
  - Positive Staining
    - Stains punctate and macro-ulcerative epithelial defects
    - Diffuses into the corneal stroma and causes a green flare in AC
  - Negative Staining:
    - Highlights non-staining lesions that project through the tear film
  - Tear Break Up Time (TBUT)
    - Number of seconds that elapse between the last blink and the appearance of the first dry spot in the tear film

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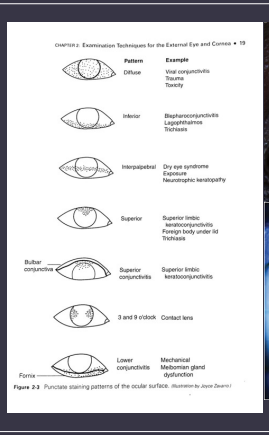

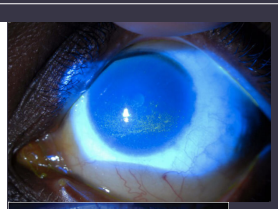
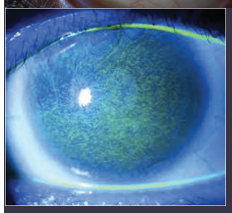

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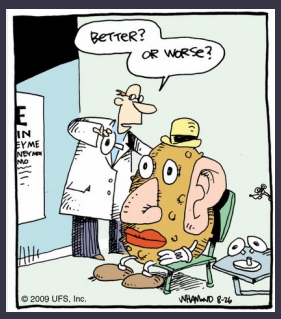
Examination of the Cornea: Astigmatism



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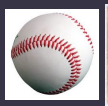
Regular Astigmatism

- Cornea shaped more like a football
  - with one meridian being significantly more curved than the meridian perpendicular to it.
  - shape is symmetric
- Corrected with cylindrical lenses

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Irregular Astigmatism

- Irregular astigmatism
  - Surface of the cornea is marred by peaks, ridges, valleys, and other abnormal shapes.
  - The shape is NOT symmetrical
    - Since the cornea is not uniformly smooth, light cannot be collected and focused onto the lens/retina properly.
  - Mild
    - Slightly blurry or distorted vision
  - Severe
    - Multiple images to appear in each eye that are disorienting and sometimes debilitating.

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Irregular Astigmatism

- Can have defined pattern or undefined pattern
- Defined
  - Keratoconus, Pellucid Marginal dystrophy, pterygium, CL warpage, s/p refractive surgery (PRK/LASIK)
- Undefined
  - DES, Trauma, corneal scarring, limbal/corneal dystrophies, Salzmann Nodular Degeneration, MDFD

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When to get a Topography

- Any disorder where there may be corneal irregularities
  - Irregular astigmatism
  - Corneal Dystrophy
    - Keratoconus, Pellucid Marginal Degeneration, Salzmann Nodular degeneration, MDFD
  - Pterygium
  - Corneal scar or Corneal trauma
  - Unknown decrease in VA
  - Increasing astigmatism/ Cylinder in MRx
  - s/p LASIK and c/o decreased VA
  - Prior to LASIK

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Topography vs Tomography

- Topography
  - Non invasive imaging for mapping the surface curvature and shape of ANTERIOR corneal surface
  - Placido disc
    - Evaluates the cornea based on reflection of concentric rings (mires)
    - Widely spaced rings → flatter
    - Closely spaced rings → steeper
  - Examples: Orbscan, Atlas, Nidek OPD
- Tomography
  - Computes a 3D image of the cornea and assesses ANTERIOR and POSTERIOR cornea
  - Scheimpflug
    - Evaluates the cornea using a camera that captures cross sections of cornea as it rotates
  - Examples: Pentacam, Sirius, Galilei,

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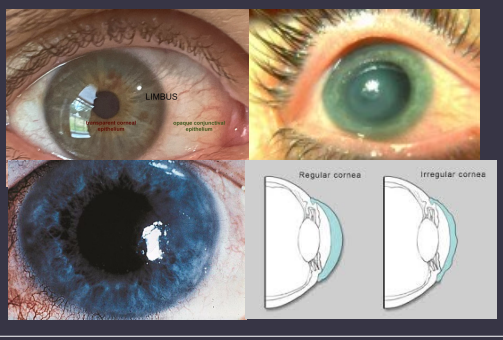
Dry Eye Syndrome



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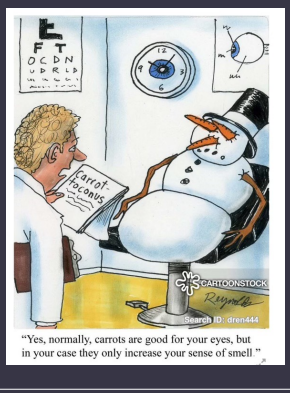
Keratoconus



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Keratoconus

- Disorder in which the central or paracentral cornea undergoes progressive thinning and bulging
- Takes on shape of a cone
- Prevalence: 50 per 100,000.
- No predictable hereditary pattern
  - +FH in 6-8% of cases
  - DNA believed to be a gene for KCN on chromosome 21
- In general, progresses during adolescent years and then stabilizes
  - Although can progress at any time
- Signs
  - Rizzutti's sign: conical reflection on the nasal cornea with light from temporal side
  - Munsons's sign: bulging of lower lid on downgaze
  - Scissoring on red reflex on retinoscopy

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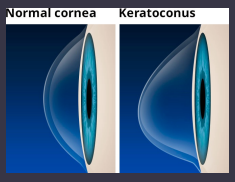
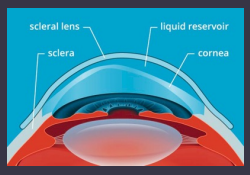
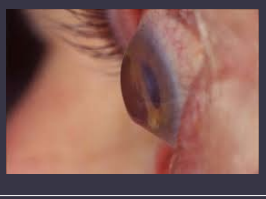
Keratoconus







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Keratoconus

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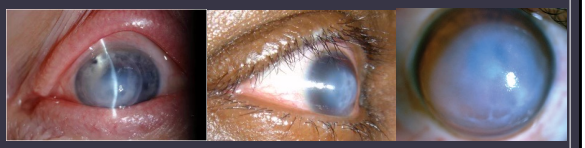
Acute Hydrops

- Break in Descemet's membrane allows aqeuous enter into stroma
- Severe Corneal edema suddenly
- Decreased Vision, light sensitivity, tearing, pain.



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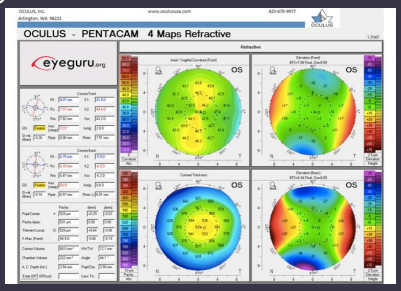
Tomography: Regular Astigmatism



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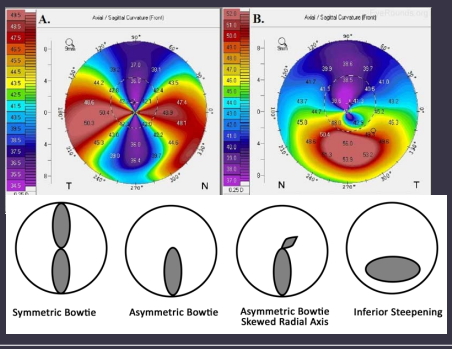
Keratoconus



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Anterior Basement Membrane Dystrophy

- Other names: Epithelial Basement Membrane Dystrophy, Map-Dot-Fingerprint Dystrophy, Cogan's microcystic dystrophy
- Abnormality of epithelial turnover, maturation, and production of basement membrane
- Histology
  - Thickened basement membrane
  - Abnormal epithelial cells with microcysts
  - Fibrillar material between epithelial basement membrane and Bowman's layer
- AD inheritance but more often sporadic
- Clinical findings:
  - Fingerprint lines
  - Map lines
  - Dots or microcysts

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Anterior Basement Membrane Dystrophy

- Occurs in 6-18% of population
  - More common in women
  - More common over age 50
- Symptoms
  - Decreased vision, fluctuating
  - Recurrent epithelial erosions

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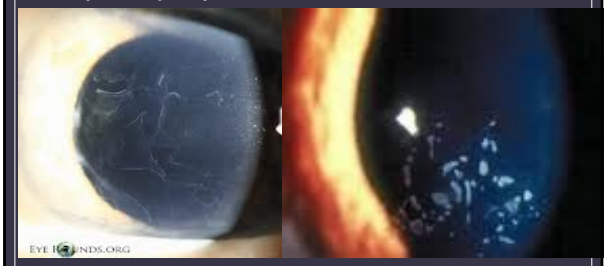
Anterior Basement Membrane Dystrophy



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Salzmann Nodular Degeneration

- Progressive disorder with gray-white or blue-white elevated nodules developing on the cornea
- More common in middle-aged and older women
  - More common bilaterally
- Associated with ocular surface disease
  - Chronic DES, Blepharitis, MGD
  - Phyctenulosis, vernal keratoconjunctivits, viral diseases
- Associated with CL wear

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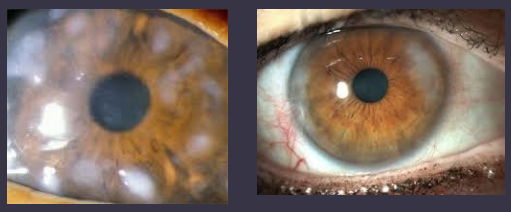
Salzmann Nodular Degeneration



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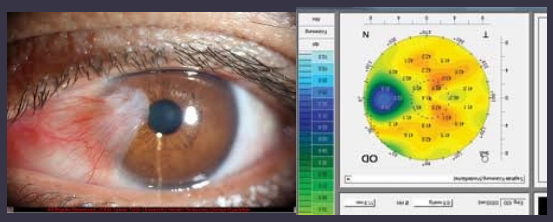
Pterygium



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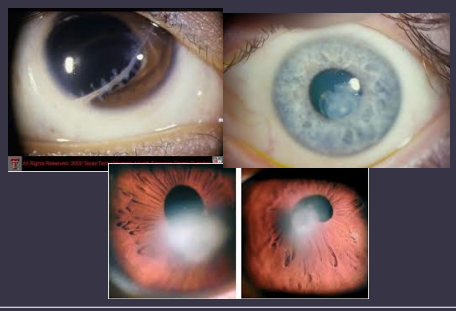
Corneal Scars



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When is Pachymetry important?

- Any disease process where there may be cornea edema (thickening of the cornea)
- S/P Cornea Transplant (DSAEK, PKP)
- Fuch's Dystrophy
- Corneal endothelial dystrophy
- Failed PK or DSAEK
- Cornea Edema
- Bullous Keratopathy
- Prior to Refractive procedure (to look for corneal thinning)

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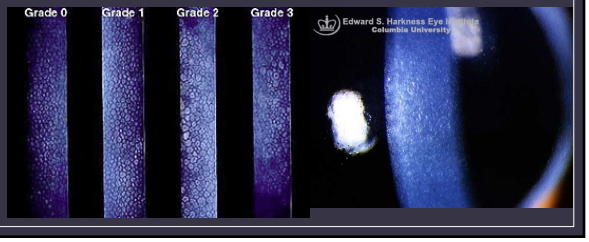
Fuch's Dystrophy

- Genetic disorder which causes the cornea to swell
  - AD or sporadic
- Endothelial cells are lost and damaged
- Symptoms: glare, halos, decreased vision (initially upon wakening)



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Corneal Transplation

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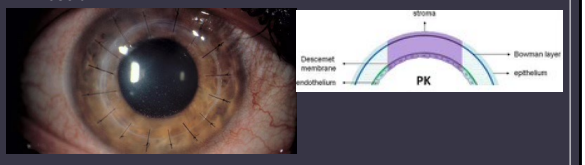
Penetrating Keratoplasty (PK)

- Full thickness corneal replacement
- Replaces all 5 layers of the cornea
- Keratoconus patients, Bullous Keratopathy, Severe Fuchs, Corneal Scars



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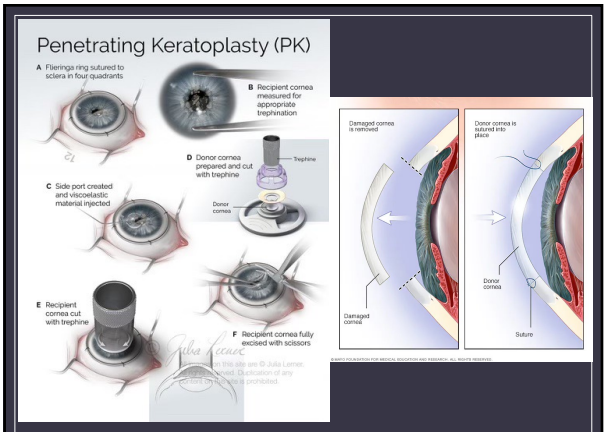

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Descemet's Stripping Automated Keratoplasty (DSAEK):

- Partial thickness cornea transplant
- Descemet's membrane and endothelium are excised and replaced with DM, endothelium, and posterior stroma from donor
- Fuch's Dystrophy, Pseudophakic Bullous Keratopathy, Endothelial dysfunction

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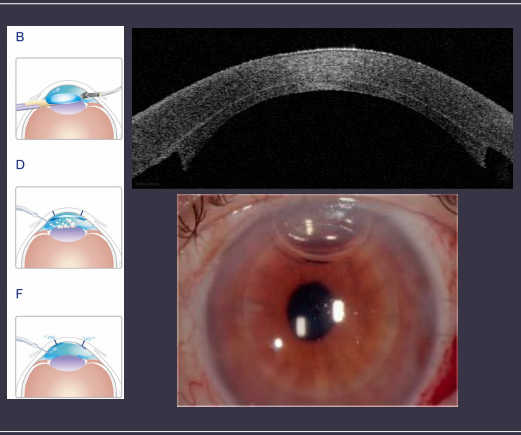

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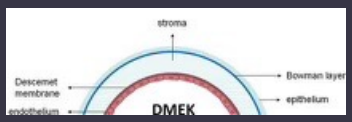
Descemet's Membrane Endothelial Keratoplasty (DMEK)

- Partial thickness cornea transplant
- Descemet's membrane and endothelium are excised and replaced with DM and endothelium only
- Fuch's Dystrophy, Pseudophakic Bullous Keratopathy, Endothelial dysfunction



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DMEK or DSEK?

- DMEK provides faster and slightly better visual recover
- DMEK reduced rejection rates

- DSEK lower rate of rebubbling
- DSEK preferred for eyes with complicated anterior chamber
  - Aphakea, tube shunts, iris defects, hypotony, etc

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Descemet's Stripping Only (DSO)

- Alternative to DMEK
- Smaller area of central unhealthy endothelium and DM is removed but no tissue is implanted
  - Wait to see if the surrounding endothelial cells migrate from periphery to recover the central area
- Advantages
  - Zero risk of rejection
  - No need for steroid drops long term to prevent rejection
- Disadvantages
  - Can only remove 4mm central area
  - Recovery takes at least 3-4 weeks
  - Option for patient's in early stages of Fuch's

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Thank You

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