Corneal endothelium

- Single layer of mitochondria rich cells on inner surface of cornea
- Embryologically derived from neural crest
- Attached to the other layers of cornea through an acellular layer of collagen, Descemet’s membrane
- Governs fluid and solute transport across posterior surface of cornea
  - Maintains cornea in the dehydrated state required for optical transparency
Endothelial dystrophies

- Fuchs’ endothelial dystrophy
- Posterior polymorphous dystrophy
- Congenital hereditary endothelial dystrophy

Endothelial dysfunction

- Corneal edema
- Visual compromise and pain
Fuchs’ Endothelial Dystrophy
Fuchs’ dystrophy

- Bilateral, noninflammatory, degenerative disease of the endothelium with reduced Na/K pump activity leading to accumulation of focal outgrowths called guttae, corneal edema, and loss of vision

- Autosomal dominant

- Onset in 5-6th decade of life

- F >> M

- Corneal findings: central guttae, stromal thickening, Descemet’s folds

- Associated with narrow angles and glaucoma
Central guttae
Low endothelial cell count
Surgical considerations with Fuchs’

• Visually significant cataract and borderline corneal function
  – CCT < 620 and no evidence of stromal edema ➔ cataract surgery alone recommended
  – CCT > 620, frank stromal edema, or 10% difference in corneal thickness in morning compared to evening ➔ combined cataract surgery with PK/DSEK
Surgical considerations with Fuchs’

- Corneal edema requiring transplantation with mild-moderate cataract
  - Retrospective studies indicate that most patient undergoing corneal transplant eventually require cataract surgery
  - Combined transplantation and cataract surgery recommended to avoid increased costs and delay in visual rehabilitation
Posterior Polymorphous Dystrophy
PPMD

• Bilateral, nonprogressive disease thought to be 2/2 focal metaplasia of endothelial cells into a population of aberrant keratinized epithelial-like cells

• Autosomal dominant

• Onset in 2-3\textsuperscript{rd} decade of life

• Corneal findings: vesicular changes, endothelial band lesions, irregular placoid opacities of the posterior corneal surface and multilayered endothelium (irregular scalloped edges)

• Associated with PAS, iris atrophy, corectopia, and glaucoma (overlap with ICE syndrome)

• Most patients asymptomatic
Congenital Hereditary Endothelial Dystrophy
CHED1

- Autosomal dominant
- Diffuse thickening and lamination of Descemet’s membrane with sparse, atrophic endothelial cells
- Occurs congenitally or during 1-2\textsuperscript{nd} year of life
- Diffuse corneal clouding and thickening that slowly progresses over 1-10 years
CHED2

- Autosomal recessive
- Diffuse thickening and lamination of Descemet’s membrane with sparse, atrophic endothelial cells
- Congenital nonprogressive disease, but more severe than CHED1
- Diffuse corneal clouding and thickening often associated with nystagmus
Treatment

- **Fuchs’**
  - Early: conservative, hypertonic ointment, IOP lowering
  - Advanced: transplantation (fairly good prognosis)

- **PPMD:** Most patients asymptomatic.
  - Early symptomatic: conservative
  - Advanced: transplantation (prognosis related to presence of PAS and glaucoma, higher chance of recurrence)

- **CHED**
  - CHED1: PK in advanced cases
  - CHED2: PK usually required given severity
References

• Yanoff, et al.
• BCSC