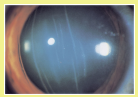
8.9.4. Secondary Abnormalities Affecting the Fetal Cornea

Arcus Juvenilis
- sector
  - deposition of lipid in peripheral corneal stroma
- not associated with abnormalities of serum lipid

Birth Trauma
- first few postnatal days
- vertical or oblique
- 
  Figure 9-10 (© 2020 American Academy of Ophthalmology)
- ruptures in Descemet membrane and the endothelium
- progressive corneal edema
  - can again become edematous later in life
  - edema may or may not clear
- healing
  - hypertrophic ridge of descemet membrane
- high astigmatism
- amblyopia
- differential diagnosis
  - congenital glaucoma

Primary Congenital Glaucoma
- dysplasia of the anterior chamber angle
- at birth or within the first few years of life
- epiphora
- photophobia
- blepharospasm
- buphthalmos
- corneal enlargement >12 mm
  - megalocornea=13.0-16.5 mm
- corneal edema
  - 25% at birth
  - 60% at 6 month
- tears in Descemet membrane
  - Haab striae
  - oriented horizontally or concentric to the limbus

Intrauterine Keratitis: Bacterial and Syphilitic
- mechanism of corneal damage
  - direct action of the infecting agent
  - teratogenic effect resulting in malformation
  - delayed reactivation of the agent after birth
- compare with circumscribed posterior keratoconus (non-inflammatory)
  - posterior corneal defect
- von Hippel internal corneal ulcer
  - signs of inflammation may be present after birth
    - corneal infiltrates and vascularization
    - keratic precipitates
    - uveitis
  - iris adhesions
  - lens is usually involved
- congenital syphilis
  - fetal death or premature delivery
  - first decade of life
    - children with untreated congenital syphilis
  - rapidly progressive corneal edema
  - interstitial keratitis
    - deep stroma adjacent to descemet membrane
      - cornea assumes a salmon pink color
      - salmon patch
  - abnormal vascularization
    - over several weeks to months blood flow ceases
      - "ghost" vessels

Congenital Corneal Keloid
- bilateral
- failure of normal differentiation of corneal tissue
- associated systemic conditions
  - Lowe disease (oculocerebrorenal syndrome)
  - ACL syndrome (acromegaly, cutis gyrata, cornea leukoma syndrome)
    - autosomal dominant inheritance
- associated ocular findings
  - cataracts
  - aniridia
  - glaucoma
- histology
  - thick collagen bundles haphazardly arranged
  - focal areas of myofibroblastic proliferation

Congenital Corneal Anesthesia
- epidemiology
  - genetics
    - autosomal recessive
    - chromosome 9q31-q33
  - rare
- clinical presentation
  - infancy and childhood
  - bilateral
  - sterile epithelial ulcerations
  - painless corneal opacities
  - classification
    - group I
      - isolated trigeminal anesthesia
    - group II
      - mesenchymal anomalies
        - Goldenhar syndrome
        - Möbius syndrome
        - familial dysautonomia
    - group III
      - focal brainstem signs
- differential diagnosis
  - herpes simplex virus keratitis, recurrent corneal erosion, and dry eye
- management
  - thorough systemic examination
  - neuroradiologic studies
  - frequent topical lubrication
  - punctal occlusion
  - nighttime eyelid splinting
  - lateral tarsorrhaphy
  - amniotic membrane transplantation
  - scleral contact lenses
  - conjunctival flap

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