Axenfeld’s Anomaly and Syndrome

- Iris strands extend across the angle to insert into a prominent Schwalbe’s ring
- About 50% of patients will develop juvenile glaucoma (in which case is called Axenfeld’s syndrome)
- Glaucoma most often occurs in childhood but may occur at anytime

- Associated systemic findings: hypertelorism, congenital glaucoma, Marfan’s syndrome, Lowe’s syndrome, Pierre Robin syndrome, Hallerman-Strieff syndrome

Figure 5.4
Posterior embryotoxon in an otherwise normal eye.

Figure 5.3
Posterior embryotoxon is thick, centrally displaced anterior border ring of Schwalbe (arrow).
Rieger’s Anomaly and Syndrome

- An autosomal dominant disorder, in which one will see a prominent Schwalbe’s ring or detached SL, iris strand attachments, hypoplasia of iris, pseudo-polycoria and corectopia, often bilateral but asymmetrical.
- Gonioscopy reveal broad attachments anterior to SLine.

- Associated ocular defects: **glaucoma** (50 to 60% of patients between 5 and 30 years of age), corectopia, pseudo-polycoria, hyaline corneal opacities in Descemet’s membrane.
- Associated systemic findings: **dental** (small teeth & absence of the teeth), skull, facial, skeletal anomalies, chromosomal anomalies, and Down’s syndrome.

- Rieger’s syndrome: the condition that combines Rieger’s anomaly plus the skeletal anomalies and/or other nonocular developmental anomalies.
- Sometimes referred to Axenfeld-Rieger syndrome.
Figure 5.8
Bieger's anomaly.
Circumscribed Posterior Keratoconus

- One or more localized crater defect on the posterior corneal surface with a concavity facing toward the anterior chamber, usually unilateral
- A rare disorder exhibiting a noninflammatory thinning of the cornea

Variable stroma haze usually centrally and unilateral with a normal anterior surface

- Unrelated to usual form of keratoconus and visual acuity not usually interfered

Associated ocular abnormalities:
- Anterior lenticonus (a globular protuberance of anterior surface of lens), aniridia, Fleischer’s ring, other signs of ASD, and glaucoma

- Anterior lenticonus, aniridia, Fleischer’s ring, anterior synechia
- Hypoplastic lens, the budge of corneal clouded, shall only central clouded, normal growth

Figure 5.9 Posterior keratoconus

Peter’s Anomaly- very rare, often bilateral

- May show a posterior corneal defect and leukoma as only sign or may be seen with iris adhesions to the leukoma and also lens apposition to the leukoma
- Anteriorly displaced lens results in a shallow anterior chamber along with the peripheral anterior synechia and can cause secondary glaucoma (keratolenticular adhesions)

- Associated ocular defects: glaucoma and microphthalmia
- Systemic associations: cleft palate, congenital heart defects, skeletal anomalies, craniofacial dysplasia
- **Sclerocornea**
  - A congenital anomaly characterized by a nonprogressive, noninflammatory, usually bilateral opacities of the peripheral, central, or entire cornea with deep or superficial vascularization

- Tissue destined to become stroma becomes sclera instead and may be circumferential (peripheral sclerocornea) or involve the entire cornea
- Associated ocular abnormalities, cornea plana, various cleavage anomalies, aniridia, microphthalmos, nystagmus, strabismus, decreased corneal sensation, cataract, and glaucoma

- Associated systemic findings: chromosomal abnormalities, mental retardation, deafness, craniofacial, digital, and skin abnormalities