7.10.1. Congenital Eyelid Anomalies (I)

Epiblepharon

- rarely causes corneal staining
- cilia often do not touch the cornea except in downgaze
- Figure 10-6 (© 2020 American Academy of Ophthalmology)
- form a horizontal fold of tissue that causes the cilia to assume a vertical position
- lower eyelid pretarsal muscle and skin ride above the lid margin
- eyelid margin is in normal position
- most common in Asian children
- tends to diminish with maturation of facial bones
  - may not require treatment
- chronic corneal epithelial irritation
  - indication for surgical treatment
  - surgical treatment
- excess skin and muscle fold are excised just inferior to the eyelid margin

Epicanthus

- usually bilateral
- medial canthal fold
- pathogenesis
  - immature midfacial bones
  - fold of skin and subcutaneous tissue
- pseudostrabismus
  - child may appear esotropic
- 4 types
  - most prominent in the upper eyelid
    - can be a normal variation of the Asian eyelid
    - epicanthus tarsalis (1)
  - most prominent in the lower eyelid
    - frequently associated with BPES
    - epicanthus inversus (2)
  - involves the upper and lower eyelids equally
    - epicanthus palpebralis (3)
  - arises from the eyebrow region and runs to the lacrimal sac
    - epicanthus supraciliaris (3)
- management
  - observation is recommended until the face achieves maturity
  - epicanthus inversus rarely resolves with facial growth
    - most forms become less apparent with normal growth of facial bones
  - isolated epicanthus
    - linear revisions (Z-plasty or Y–V-plasty)
  - epicanthus tarsalis in the Asian patient
    - Y–V-plasty
    - ± construction of an upper eyelid crease
- Figure 10-5 (© 2020 American Academy of Ophthalmology)

Ankyloblepharon

- partial or complete fusion of eyelids by webs of skin
- can usually be opened with scissors after being clamped for a few seconds with a hemostat

Euryblepharon

- Figure 10-4 (© 2020 American Academy of Ophthalmology)
- Figure 10-3 (© 2020 American Academy of Ophthalmology)
- ± associated with BPES
- clinical presentation
  - lateral portion of the eyelids is typically more involved
    - vertical shortening + horizontal lengthening of the involved eyelids
  - palpebral fissure often has a downward slant
    - inferiorly displaced lateral canthal tendon
  - impaired blinking
  - poor closure
  - lagophthalmos
  - exposure keratitis
- surgial reconstruction
  - if symptomatic
    - lateral canthal repositioning
    - suspension of suborbicularis oculi fat to lateral orbital rim to support lower eyelid
  - for excess horizontal length
    - lateral tarsal strip or eyelid margin resection
  - ± skin grafts

Blepharophimosis–Ptosis–Epicanthus Inversus Syndrome (BPES)

- blepharophimosis syndrome
- autosomal dominant
  - chromosome 3
  - FOXL2 gene
- clinical presentation
  - blepharophimosis
  - severe ptosis
  - epicanthus inversus (fold of skin extending from the lower to upper eyelid)
  - lateral lower eyelid ectropion
  - hypoplasia of the superior orbital rims
  - telecanthus
  - hypertelorism
  - poorly developed nasal bridge
  - ear deformities
- Figure 10-1 (© 2020 American Academy of Ophthalmology)
- 2 types
  - type I
    - early loss of ovarian function
  - type II
- management
  - multiple surgeries required
  - indications for surgical correction
    - visually disruptive ptosis
      - usually requires frontalis suspension
    - telecanthus
      - medial canthal repositioning places traction on the upper eyelid and may exacerbate the ptosis
      - multiple Z-plasties or Y–V-plasties + transnasal wiring of the medial canthal tendons
    - ectropion
    - orbital rim hypoplasia

Congenital Ectropion

- etiology
  - isolated finding
    - rare
  - often associated with
    - BPES
    - Down syndrome
    - ichthyosis
- clinical presentation
  - vertical insufficiency of the anterior lamella of eyelid
  - chronic epiphora
  - exposure keratitis
- treatment
  - mild cases
    - observation
  - severe/symptomatic cases
    - treated like a cicatricial ectropion
      - horizontal tightening of the lateral canthal tendon
      - vertical lengthening of anterior lamella with full-thickness skin grafting
- complete eversion of upper eyelids
  - occasionally occurs in newborns
  - etiology
    - inclusion conjunctivitis
    - anterior lamellar inflammation or shortage
    - Down syndrome
  - management
    - topical lubrication
    - short-term patching of both eyes
    - full-thickness sutures
    - temporary tarsorrhaphy
    - definitive repair
- Figure 10-2 (© 2020 American Academy of Ophthalmology)

Introduction

- types
  - isolated
  - associated with other eyelid, facial, or systemic anomalies
- most congenital eyelid anomalies occur during the second month of gestation