

# Primary Congenital Glaucoma

## Case Description

2-month-old boy with right eye tearing, frequent blinking, and forceful closure of his eyelids

## Image Description



enlarged cornea of the right eye

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## Differential Diagnosis

- primary congenital glaucoma**
  - 80% bilateral
  - 10-40% familial
  - epiphora
  - photophobia
  - blepharospasm
  - corneal edema
- forceps injury**
  - vertical/oblique descemet tears
  - unilateral
- NLD obstruction**
- CHED**
  - bilateral
  - AD
    - presents during 1st-2nd year of life
    - progresses over 1-10 years
  - CHED1
  - AR
  - CHED2
    - congenital
    - stationary
- CHSD**
  - congenital stromal corneal dystrophy (CSCD)
  - AD
- congenital megalocornea**
  - bilateral
  - XLR → boys
  - >14 mm
- anterior uveitis**
- corneal abrasion/foreign body**
- congenital rubella**
  - microphthalmia
  - corneal clouding
  - cataract
  - glaucoma
  - salt & pepper retinopathy
    - most common ocular manifestation
    - 25%-50%
  - hearing loss
  - cardiac defects
- Sturge-Weber syndrome**
  - sclerocornea
  - trauma/tear
  - ulcer
  - metabolic
    - bilateral
    - mucopolysaccharidoses
    - cystinosis
  - Peters
    - at birth
  - PPMD
    - bilateral
    - grouped vesicles, scalloped bands
  - dermoid (limbal)
- STUMPPeD**

## Data acquisition

### History

- onset/course
- epiphora
- photophobia
- blepharospasm
- discharge
- forceps delivery
- maternal infections during pregnancy
- congenital glaucoma
- CHED/CHSD
- congenital megalocornea
- metabolic diseases
- family hx

### Physical Exam

may need to be done under general anesthesia

- vision**
- APD**
- external exam**
  - Sturge-Weber syndrome
    - port wine stains
  - forceps mark
  - dacryocystocele, discharge from lacrimal puncta
- IOP**
- anterior segment**
  - corneal diameter >12 mm
  - increased tear lake
  - corneal edema
  - horizontal Descemet tears
  - corneal thickness (pachymetry)
  - anterior uveitis
    - AC reaction
  - abnormal angle anatomy
  - gonioscopy
  - optical disc cupping
  - diffuse choroidal hemangioma
  - Sturge-Weber syndrome
  - congenital rubella
    - salt & pepper retinopathy
  - myopic shift
  - cycloplegic refraction
- dilated funduscopy**



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## Additional Testing

A-scan for axial length

congenital glaucoma

## Assessment

### Medical

- to lower IOP before surgery
- adjunct treatment after surgery
- avoid brimonidine
- relative contraindication <2 years of age
- treatment of choice for primary congenital glaucoma

### Surgical

- if clear cornea
  - goniotomy
- cloudy cornea
  - 2 failed goniotomies
    - trabeculotomy
  - age > 1.5 yr
    - trabeculectomy with mitomycin C
    - tube shunt
    - cyclodestructive procedures
- + genetic counseling

### Referrals

## Treatment

### Pathogenesis & natural history

### treatment options

### prognosis

### Inform patient's family about

### amblyopia

### myopia

### corneal scarring

### glaucomatous optic atrophy

### life-long treatment (medical/surgical) may be necessary

### follow-up

### but useful vision can be maintained with timely treatment

### prognosis best for patients whose glaucoma is diagnosed between 3 and 12 months

### vast majority respond to angle surgery