

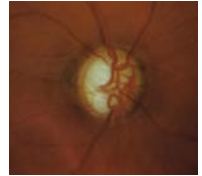
Infantile Glaucomas

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What is Infantile Glaucoma?

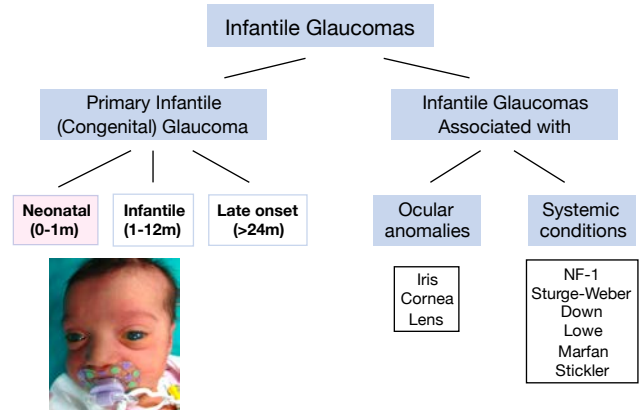
- Progressive optic neuropathy
- **Elevated intraocular pressure**
- Onset within the first 3 years of life
- Genetic basis
- Uniformly blinding disease if left untreated
- Significant visual morbidity and legal blindness



Epidemiology

- Frequency: 1:10,000 to 1:20,000
- Bilateral 70%
- More common in males (65%)
- Most (80%) will have onset within the first year of life
- More common in certain ethnicities
 - Middle eastern
 - Slovakian Roms
- Consanguinity increases the risk of PCG

Classification



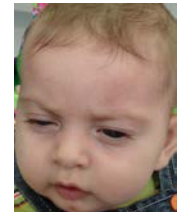
Pathogenetic Mechanisms

- Aqueous humor cannot leave the eye
- Developmental anomaly of the drainage angle
 - Trabecular meshwork and Schlemm's canal
- Neural crest cell migration
- Arrest of differentiation of the angle structures
- Histopathologic appearance resembles that of a 28w gestational age
- 3 distinct genetic loci



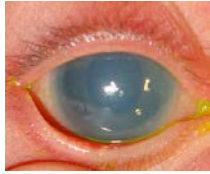
Presentation

- **Classical triad:**
 1. Photophobia
 2. Blepharospasm
 3. Epiphora
- Within the first 3 years of age:
 1. Enlarged globe
 2. Corneal stretching
 3. **Abnormal red reflex**
- Beyond 3 years of age:
 - Scleral stretching
 - **Myopia**



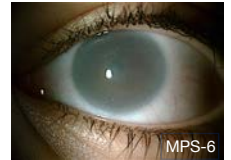
Corneal Edema

- Reason for abnormal red reflex
- Elevated intraocular pressure
- Presenting symptom in newborns
- → **Photophobia**
- → **Epiphora**
- May lead to persistent corneal opacification



Differential Diagnosis of Corneal Edema

1. Birth trauma
2. Keratitis (Rubella, HSV)
3. Corneal dystrophies
4. Corneal dysgenesis
5. Metabolic disorders
 - MPS
 - Cystinosis



Corneal Enlargement

- Pressure related corneal stretching
- May go unnoticed if bilateral
- Nomograms per age
- Only within the first 4 years
- Irreversible
- DDx: X-linked megalocornea



Epiphora

- Not an isolated finding in glaucoma
 - Light aversion, corneal enlargement/edema
- Congenital NLD obstruction
- Epiblepharon
- Corneal epithelial defect
- Conjunctivitis
- Keratitis
- Intraocular inflammation



Management and Prognosis

- Need EUA to confirm IOP and diagnosis
- Multiple procedures frequently required
- Systemic toxicity of topical hypotensive medications
 - Beta blockers
- Require life-long monitoring
- **Prognosis**
 - IOP control in 80% of patients
 - VA <20/50 in 50% of patients

