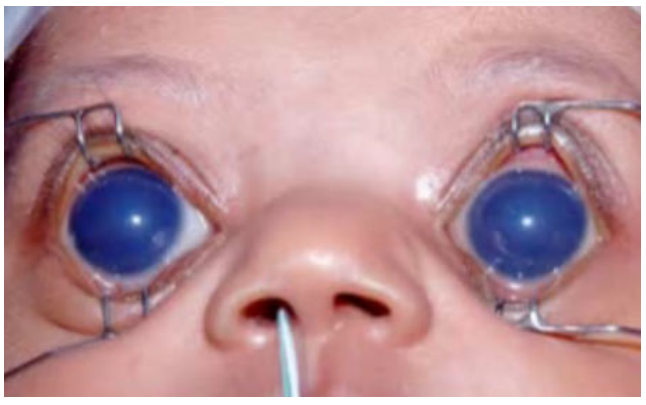


Congenital glaucoma/ Buphthalmos

CONGENITAL GLAUCOMA

Learning Objectives:

1. Definition
2. Pathogenesis
3. Clinical Presentation
4. Differential diagnosis
5. Management

Congenital Glaucoma

- A condition of raised intraocular pressure in early childhood with developmental anomaly of the anterior chamber angle region without any associated abnormality of eye or adnexa, which obstructs the drainage of aqueous humour.
- It appears between birth and 4 yrs of age.
- Both eyes are usually involved, but to varying severity
- Boys are more affected than girls.
- Affects 1 in 10,000 births

- A hereditary factor is occasionally present (Autosomal recessive).

Types

1. Primary developmental/congenital glaucoma.

2. Developmental glaucoma with associated ocular anomalies

3. Secondary congenital/Juvenile glaucoma

Pathogenesis

- Failure/abnormal development of the anterior chamber angle and trabecular meshwork during intrauterine development.
- Maldevelopment of trabeculum including the iridotrabecular junction *(trabeculodysgenesis)*

- Angle develops from mesodermal tissue

either in the form of failure of resorption of mesodermal sheet

Or

abnormal cleavage of AC angle

This is responsible for impaired aqueous outflow resulting in raised IOP.

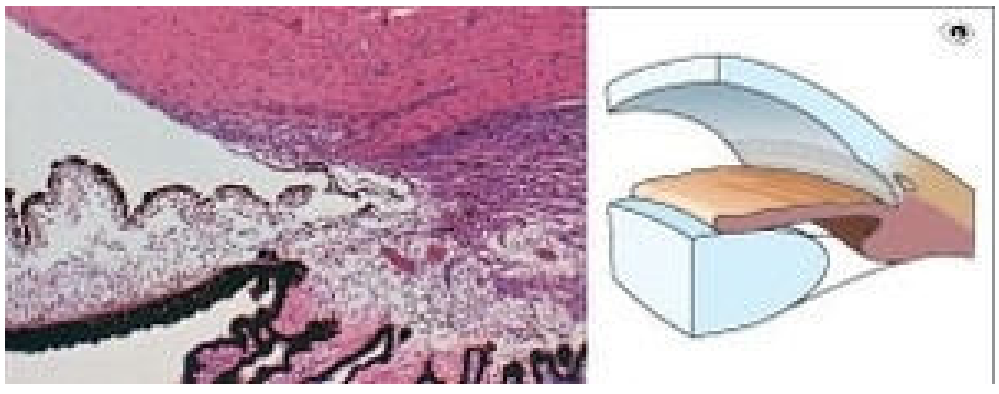


Figure1: The normal chamber angle: on the left is a histological cross-section; on the right is a drawing of the same

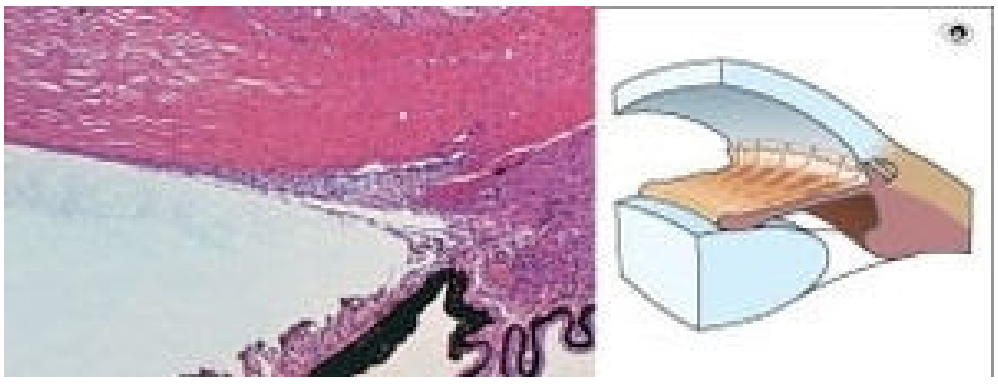


Figure 2: An underdeveloped chamber angle

Clinical Presentation

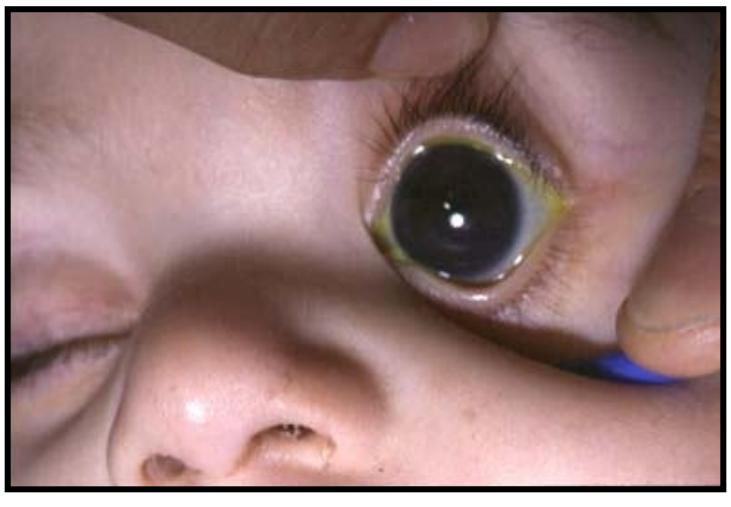
Symptoms:

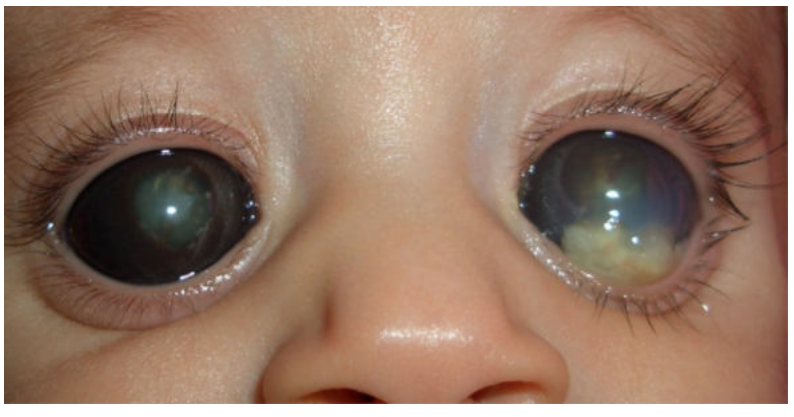
- Photophobia
- Blepharospasm
- Lacrimation/watering

Clinical Presentation

Signs:

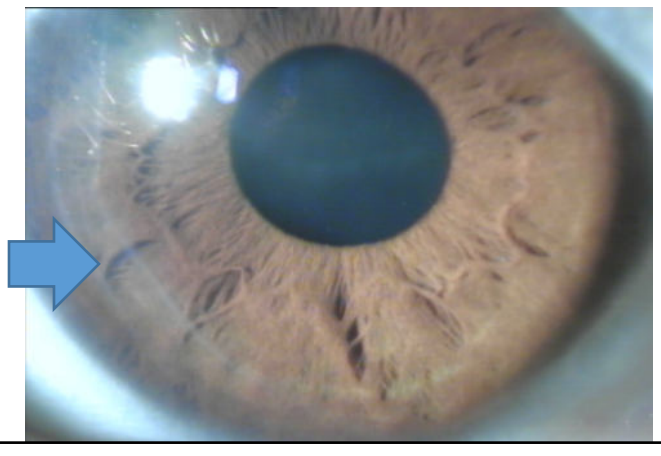
- Enlargement of the globe (buphthalmos) is a result of elevated intraocular pressure.
- Bluish discoloration patches over sclera
  [due to thinning of sclera underlying uveal tissue becomes visible]

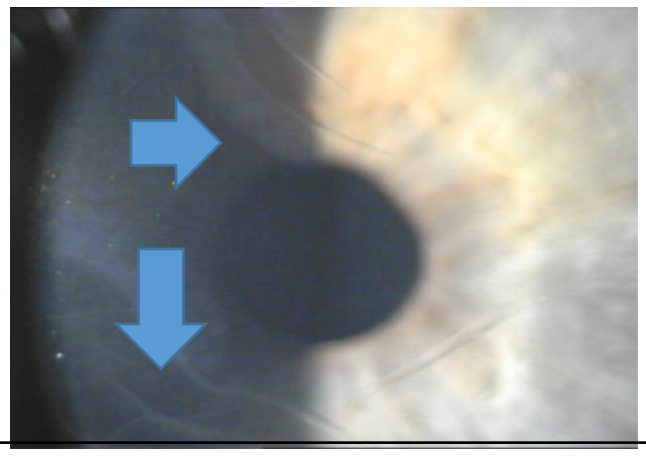


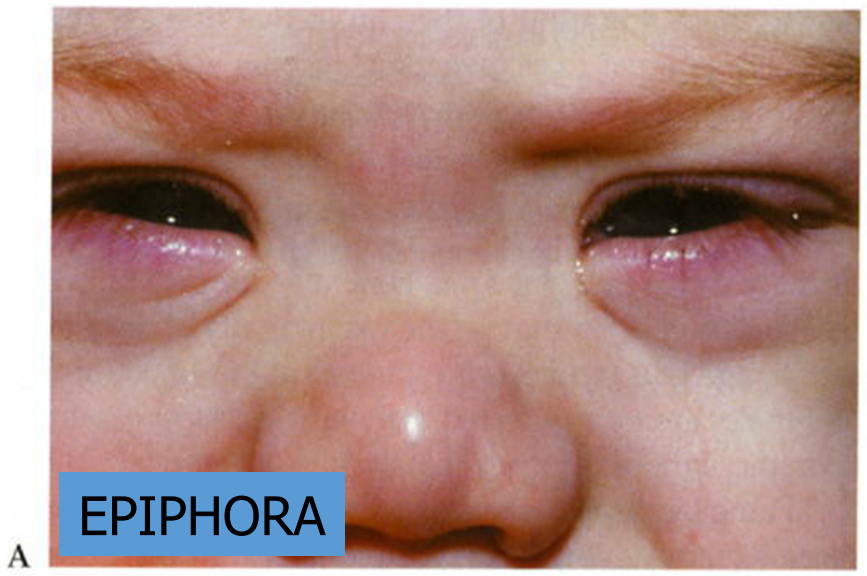


Clinical signs

- ***Corneal signs.***
  i. *Corneal oedema.*
  ii. *Corneal enlargement. (corneal diameter > 13mm)*
  iii. *Tears and breaks in Descemet's membrane*
  (Haab's striae).







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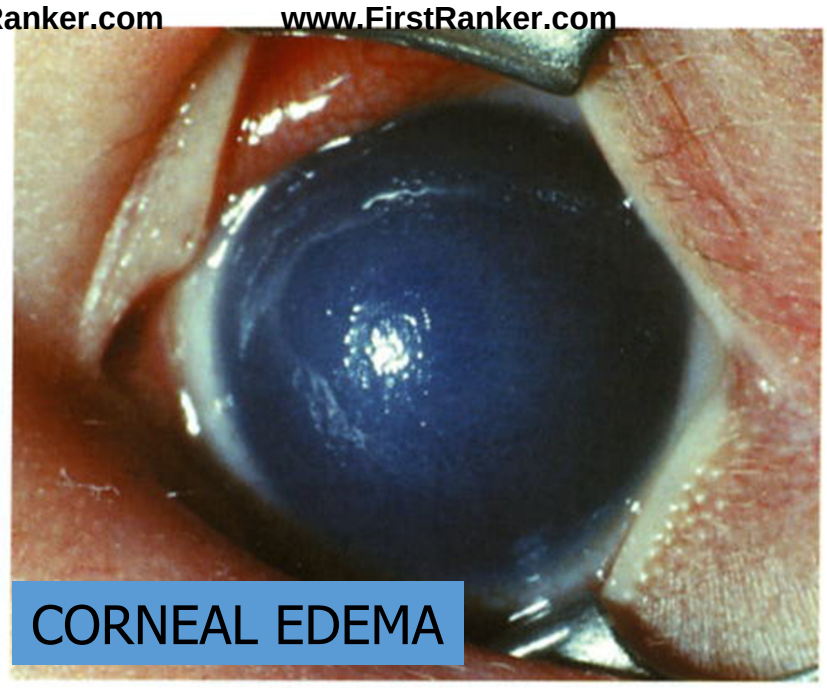

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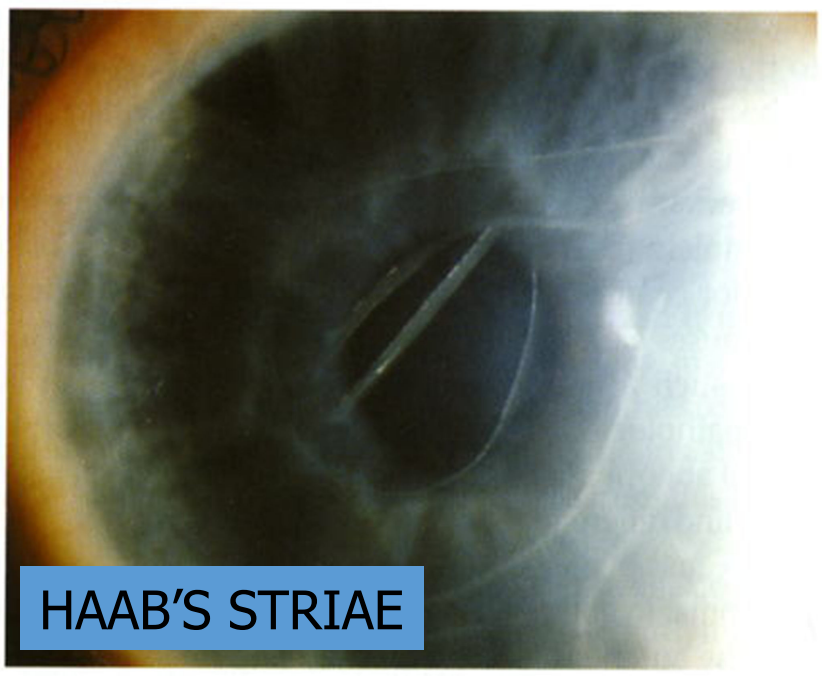


C

D

Clinical signs

- ***Anterior chamber becomes deep.***
- ***Iris may show iridodonesis and atrophic patches***

  in late stage.
- ***Lens becomes flat due to stretching of zonules and*** may

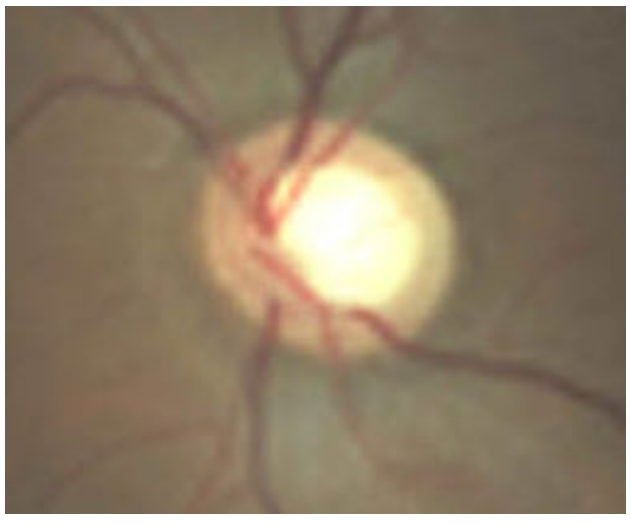
  even subluxate.

Clinical signs

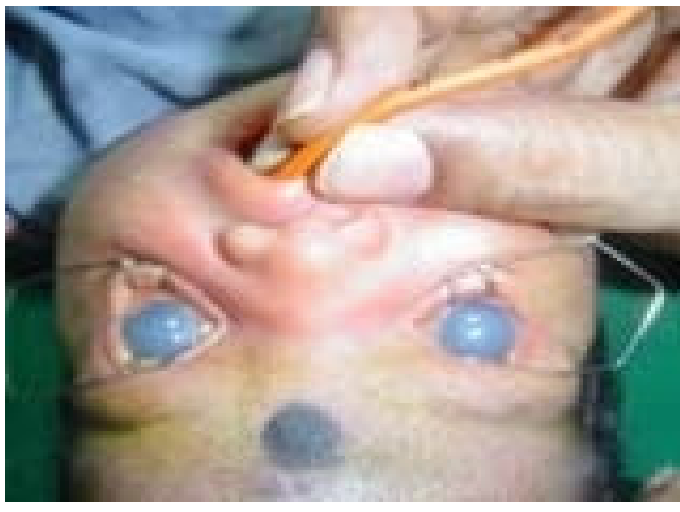
- ***IOP is raised which is neither marked nor acute.***

- ***Axial myopia may occur because of increase in*** axial length which may give rise to anisometropic amblyopia

- ***Optic disc may show variable cupping and thus damage to otic nerve in the form of atrophy***



Diagnosis



- Clinical clues
  - Enlarged eyes; tearing, and photophobia (avoidance of light).
  - Often, babies also rub their eyes.

- If CG is suggested, a thorough examination under general anesthesia is necessary.
  - to avoid blepharospasm (spasmodic closure of the eyes). to prevent a transient rise in the IOP.
  - Besides measuring the IOP, anesthesia allows a thorough investigation of all segments of the eye and, in particular, the optic disc

Diagnosis

1. *Measurement of IOP with Schiotz or preferably* hand held Perkin's applanation tonometer.
2. *Measurement of corneal diameter by callipers.*
3. *Ophthalmoscopy to evalute optic disc.*
4. *Gonioscopic examination of angle of anterior*

chamber reveals trabeculodysgenesis with either flat or concave iris insertion as described in pathogenesis.

Differential Diagnosis

- Cloudy Cornea: In unilateral cases the commonest cause is trauma with rupture of Descemet's membrane (forceps injury).

  In bilateral cases causes may be trauma, mucopolysaccharidosis, interstitial keratitis and corneal endothelial dystrophy.

Differential Diagnosis

- *Large cornea due to buphthalmos should be* differentiated from megalocornea.
- *Lacrimation in an infant is usually considered to* be due to congenital nasolacrimal duct blockage and thus early diagnosis of congenital glaucoma may be missed.
- *Photophobia may be due to keratitis or uveitis.*

Differential Diagnosis

- *Raised IOP in infants may also be associated*

  with

- retinoblastoma,
- retinopathy of prematurity,
- persistent primary hyperplastic vitreous,
- traumatic glaucoma and
- secondary congenital glaucoma seen in rubella, aniridia and Sturge-Weber syndrome.

Treatment

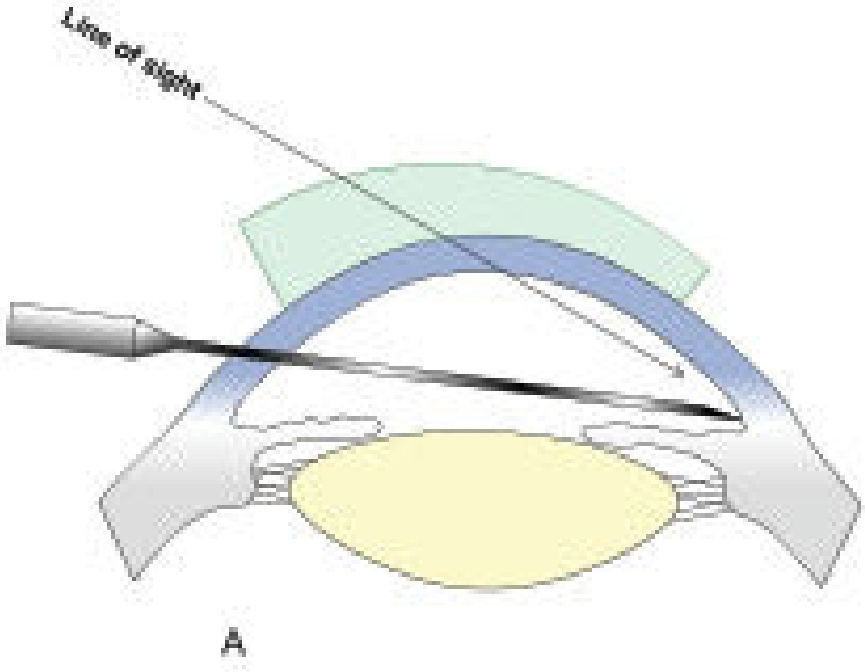
- The treatment is primarily surgical

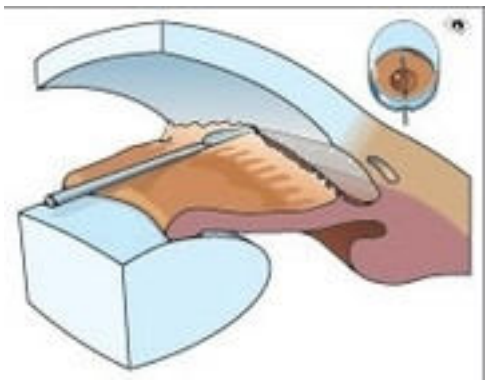
(medical treatment is given temporarily- beta blockers,hyper osmotics, acetazolamide) .

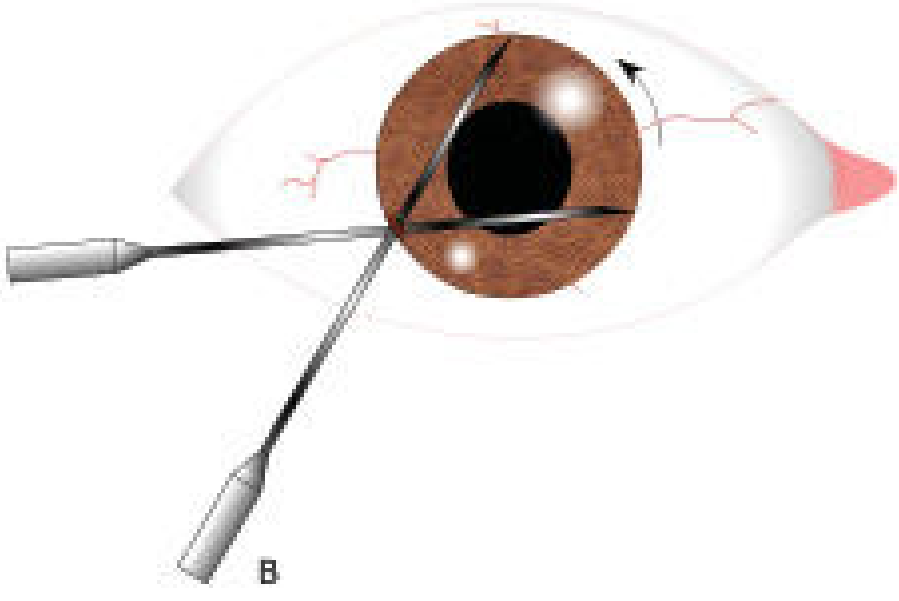
- Different surgical procedures

  (according to the degree of the maldevelopment and the clarity of the cornea)
  - **Goniotomy (85% success)**
  - **Trabeculotomy**
  - **Trabeculotomy + trabeculectomy (best results)**
- Supplemental treatment options are
  - Medical therapy
  - Implant surgery
  - Cyclodestructive procedures

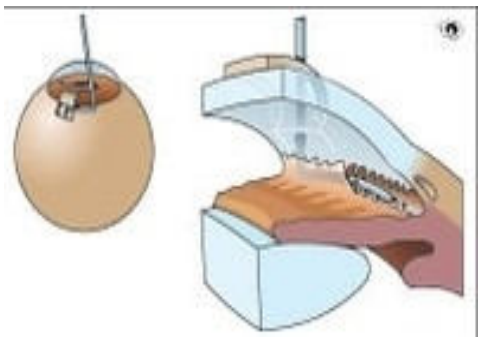
Goniotomy

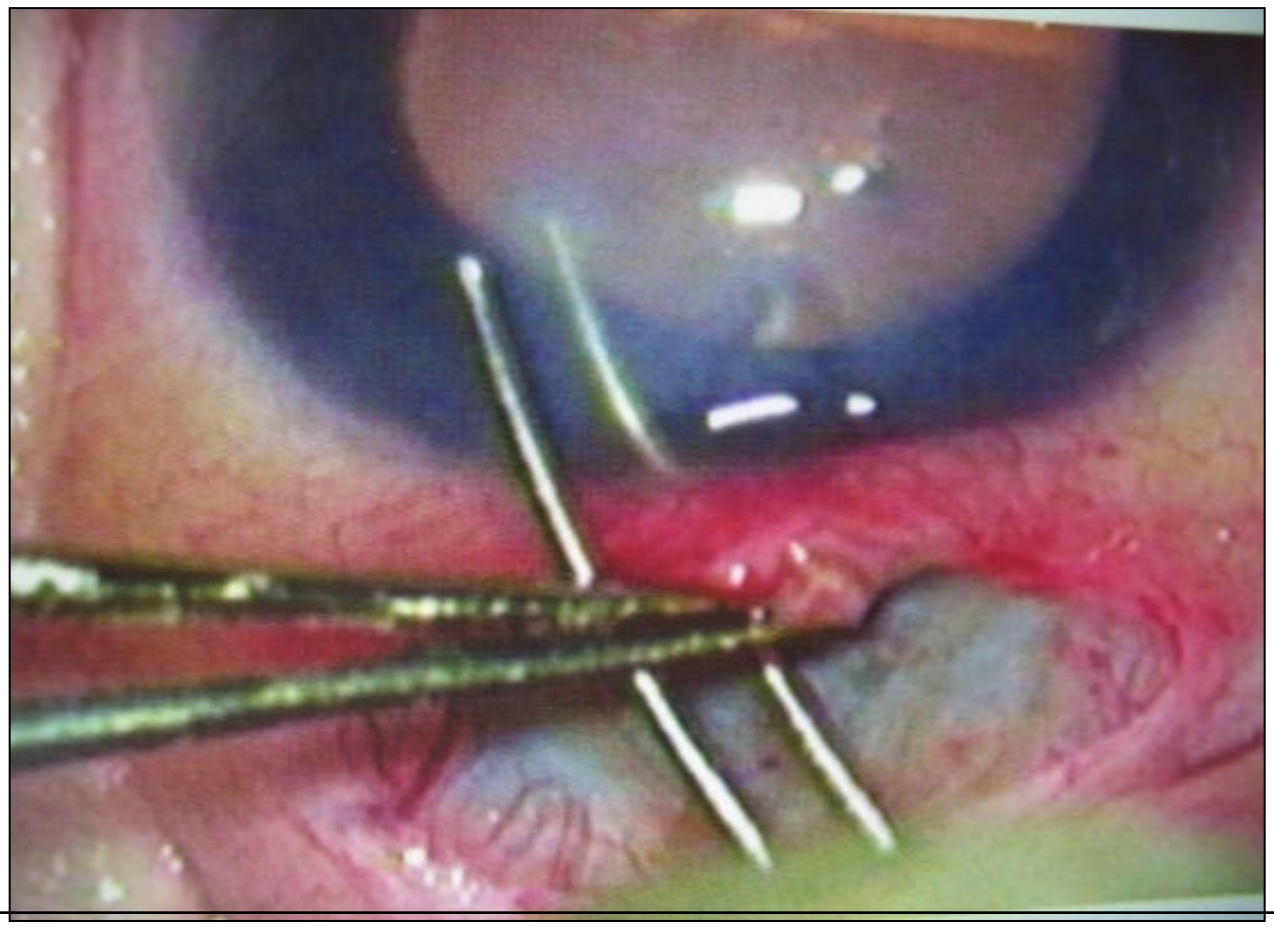






Trabeculotomy





Trabeculectomy

Infantile Glaucoma

- **Infantile glaucoma** is also **congenital glaucoma**
- However, **intraocular** pressure starts to rise at some time during the first years of life.
- The cause for this IOP increase is basically the same as in **congenital glaucoma**, but it occurs later since the **anterior chamber** angle is more mature than when glaucoma is present at birth.
- The IOP may be normal during the first years of childhood and then gradually increase.

Juvenile Glaucoma

- Juvenile glaucoma is an IOP increase that occurs in an older child or young adult (10- 35 yrs)and is often inherited.
- During a thorough examination, the ophthalmologist may find discreet evidence of an incomplete maturation of the chamber angle,
- The clinical features as well as treatment of juvenile glaucoma are quite similar to adult Primary Chronic Open-Angle Glaucoma (POAG)

Developmental glaucoma with associated anomalies

- Glaucoma with iridocorneal dysgenesis
- Glaucoma with aniridia
- Glaucoma with ectopia lentis
- With phakomatosis- Sturge weber syndrome, neuro fibromatosis
- Lowe's, naevus of ota, nanophthalmos, congenital ectopian uveae, microcornea & rubella syndrome