Congenital Glaucoma

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- Congenital glaucoma is a major cause of blindness in children, despite its low incidence (1:10,000 births) legal blindness 50%

- This category includes isolated congenital glaucoma (also called primary congenital glaucoma) and glaucomas associated with other developmental anomalies, either systemic or ocular.

- boys 65%

- both eyes 75%

Inheritence

- Most cases sporadic
- 10% inheritence as AR with incomplete penetrance

Pathogenesis

- Trabeculodysgenesis
- other major ocular pathology

- A hereditary factor is occasionally present.
- The IOP elevation is caused by the failure of the **anterior chamber** angle and the **trabecular meshwork** to develop appropriately during intrauterine development.
- In these infants, the **aqueous** humor does not properly drain, but since the production of **aqueous** humor is nevertheless normal,

The intraocular pressure is high

Consequences of an Increased IOP during Infancy

- Depending on the IOP level, **glaucomatous damage** is inevitable after weeks, months or even years.
- This basically occurs via the same mechanisms as in the adult.
- In addition to **optic nerve** damage, the globe (eyeball) enlarges because the **sclera** in the eye of a baby is distensible.

Classification

- True congenital 40 %
- Infantile 55 % prior 3rd birthday
- Juvenile 5 % between 3-16 years

- Juvenile glaucoma is the term used to designate cases in which the pressure rise develops after the third birthday but before the age of 16 years.

- The enlargement of the eye (buphthalmos) is least common, despite the elevated intraocular pressure. Gonioscopy is normal or reveals trabeculodysgenesis. This condition may simulate the primary open-angle glaucoma.

Diagnosis

- 1-Corneal haze (lacrimation photophobia blepharospasm)
- 2-Bophthalmos (before 3 years) (myopia, deep AC, lens subluxattion)
- 3-Breaks in descemets membrane(Haab striae)
- 4-Optic disc cupping(?enlargement of scleral canal)

Surgery

- Goniotomy clear cornea diameter <14 mml 85% success rate
- Trabeculotomy
- Trabeculectomy
- Combined

Follow up

Differential Diagnosis

- Cloudy cornea (Birth Trauma- Rubella-Metabolic DisorderEndothelial dystrophy)
- Large cornea(megalocornea -high myopia
- Lacrimation
- Secondary glaucomas(Tumour PHPPV-ROP-Trauma-inflammation-Ectopia lentis)

Primary Angle Closure Glaucoma

Angle closure
Iridotrabecular apposition /adhesion
Obstruction to aqueous outflow
Raised Intraocular pressure. •

Anatomical features : Shallow AC
Thicker lens
Shorter Axial length
Smaller corneal diameter
Occludable Angle:

Stages in Natural history •

1 Primary angle-closure suspect (PACS) • Gonioscopy shows posterior meshwork ITC in three or more quadrants but no PAS. • Normal IOP, optic disc and visual field.

2 Primary angle-closure (PAC) • Gonioscopy shows three or more quadrants of ITC with raised IOP and Normal optic disc and field.

3 Primary angle-closure glaucoma (PACG) • Gonioscopy shows ITC in three or more quadrants. • Optic neuropathy. • ITC –iridotrabecular contact , PAS – Peripheral anterior synechiae

Risk Factors

Positive family history for angle closure
Age over 40-50 yrs
Women
Hyperopia Pseudoexfoliation

Pathogenesis

pupillary block-plateau iris

Relative Pupillary block • Failure of aqueous flow through the mid dilated pupil leads to a pressure differential between the anterior and posterior chambers, with resultant anterior bowing of the lax iris [Iris bombe] blocks trabecular meshwork and iridolenticular contact

Non-pupillary block relating with plateau iris • Plateau iris configuration is characterized by a flat central iris plane in association with normal central anterior chamber depth. The angle recess is very narrow, with a sharp iris angulation over anteriorly positioned and/or orientated ciliary processes• Plateau iris syndrome describes the occurrence of angle-closure despite a patent iridotomy in a patient with morphological plateau iris.

Ocular manifestations

- Symptoms: Decreased vision

Halos around lights

Frontal headache

Ocular pain

Nausea and vomiting

1. Acute congestive glaucoma

Elevated IOP risen rapidly

Conjunctival congestion Corneal epithelial / stromal edema

Shallow or flat peripheral AC

mid dilated [vertical oval] pupil absent / sluggish pupil reaction

Fellow eye generally shows an occludable angle

2. Chronic presentation

Episodic (intermittent) ITC is associated with the formation of discrete PAS, • Disc cupping /nerve fibre defects with or without visual field defect

3 Resolved acute (post-congestive) angle closure

• Folds in Descemet membrane (if IOP has been reduced rapidly), optic nerve head congestion and choroidal folds.

• Later iris atrophy [spiral-like configuration], irregular pupil, posterior synechiae and glaukomflecken

Sequence of events:

- Acute angle closure:

sudden ,circumferential ,iridotrabecular apposition-rapid severe rise in IOP

- Intermittent angle closure : Self limiting episodes of ITC ,milder signs & symptoms of former
- Creeping angle closure: slowly progressive ITC –Elevated IOP
- Chronic angle closure :

irreversible ,iridotrabecular adhesion , asymptomatic unless significant raised IOP.

Provocative tests

• Pharmacological test: pupillary block mechanism in mid dilated state ,increased tension of iris . -Performed with short acting mydriatic [phenylephrine eye drops] -if test proves positive –acute attack may be triggered

• **Paraphysiological test :** Dark room prone test – pupil dilates in dark,lens moves forwards in prone. - Patient sits for 30 minutes in dark with head prone ,no sleeping - IOP checked rapidly ,positive if increases by 8 mm Hg

Management of PACG ATTACK

A-Medical treatment

B-Minor mechanical manibulation

C-Peripheral laser iridotomy :

D-Surgical peripheral iridectomy

E-Argon laser iridoplasty

F-Lens extraction

G-Trabeculectomy ·

Management of PACG ATTACK

A-Medical treatment

Osmotic agent and acetazolamide
Pilocarpine
B-blocker
alpha agonists
Topical corticosteroid
systemic pain medication If patient is vomiting
Intramuscular metoclopramide

B-Minor mechanical manibulation

Corneal indentation using 4 mirror lens Leave patient supine to allow lens move back in position If patient in pain Attack broken Pupil constricted IOP lowered Corneal clearing

C-Peripheral laser iridotomy :

• A procedure ,where hole is made in iris periphery allowing aqueous to drain from PC into TM Helps eliminate high aqueous pressure behind iris and iris falls back. Done using Nd:YAG laser ,150-200 microns size 3-6 mj of power based on thickness Topical pilocarpine 30 mins before laser therapy,. Post op steroids and antiglaucoma meds Examine patency and size of iridotomy with gonioscopy

D-Surgical peripheral iridectomy

• Removal of iris tissue by knife or scissors • 2-3 mm peripheral corneal incision in superotemporal site. • Alternatively ,conjunctival peritomy and scleral limbus incision ,nylon sutures wound closure • Externalised iris piece held with toothed forceps , incised with fine scissors.

E-Argon laser iridoplasty

• Aim to shrink and flatten iris tissue without damage • Energy is defocussed so can be given in cloudy cornea also

F-Lens extraction

Removal of lens with or without opacity due to lens size / malposition. Best outcomes with small incision phaco.

G-Trabeculectomy •

Trabeculectomy lowers IOP - creating a fistula, to allow aqueous outflow from the anterior chamber to the sub-Tenon space. The fistula is protected or ‘guarded’ by a superficial scleral flap • When medical therapy has failed to achieve adequate control of IOP.