

Primary congenital glaucoma

- 1: 10 000 births
- 65% male
- Sporadic in 90%
- AR with incomplete penetrance in 10%
- Isolated trabeculodysgenesis:
- Maldevelopment of the trabeculum, including the iridotrabeular junction, which is not associated with any other ocular abnormalities

Symptoms of Glaucoma

- Classic Triad:
 - Photophobia
 - Epiphora/tearing
 - Blepharospasm

EUA

EUA Essentials
(remember anaesthetic agents - JOP, except ketamine)
IOP - use 2 methods of IOP measurement ie Perkins and Tonopen
HCD
Refraction
Anterior segment exam
retroillumination and direct for Haab's striae/ MCO
gonioscopy
Disk assessment with photography if available
Consider A-scan for axial length

What's normal?

- Corneal diameter
 - Newborn 9.5-10.5mm
 - 1 year 11.0mm
 - 2-3 years 12.00mm
- Cup:Disk Ratio > 0.3
 - 70% Infantile glaucoma
 - 3% Normal Newborns

What's Normal

- Normally refraction up to +4.00 in 4 month old
- Axial Length
 - Mean AL of term infant = 16.5mm
 - Rapid growth of the eye in first 18 months - average growth = 3.75mm
 - By 13 years age AL = 23mm

The Normal Infant Angle

- iris usually inserts posterior to scleral spur, but spur is hard to identify
- anterior extension of ciliary body is seen as a distinct band anterior to iris insertion
- iris insertion into the angle is flat, because angle recess has not yet formed
- trabecular meshwork appears thicker & more translucent than that of adult
- absence of acquired pigmentation of trabecular meshwork is normal

Congenital Glaucoma

- anterior insertion of iris directly into trabecular meshwork
- most commonly flat or concave insertion may also be seen
- surface of the trabecular meshwork may have a stippled appearance & meshwork may appear thicker than normal
- no pigmented band is present, but a thin section of ciliary body may be visible through thickened trabeculum
- peripheral iris may show a thinning of the anterior stroma

Ambroise Pare (1517-1590)

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- "Oeil de boeuf est une maladie d'oeil quand il est gros et eminent sortant hors la teste comme on voit les boeufs les avoir"

- Sant-Yves 1722 - abundant aqueous listed as a cause of globe enlargement
- Beger 1744, Mackenzie 1830: association of increased ocular tension and buphthalmos
- Weber 1856; described glaucomatous disk cupping; Von Graefe correlated high IOP and signs of glaucoma
- Von Muralt 1869: congenital hydrophthalmos recognized as glaucoma
- Grahamer 1884, Haab 1899: described ruptures in Descemet's membrane

J. Ringland Anderson 1939

- *"There is no division of opinion regarding the severity of the visual loss and its progressive nature in spite of treatment ... Little hope of preserving sufficient sight to permit the earning of a livelihood can be held out to them."*

Anderson JR. *Hydrophthalmia or Congenital Glaucoma*. 1939. London, Cambridge University Press.

The Goniotomy

- Carlo de Vincentis¹ 1895
 - L'incisione dell'angolo
- Otto Barkan² 1936
 - Goniotomy



¹ De Vincentis, C. "Incisione dell'angolo irideo nel glaucoma". *Ann Ottalmol*. 1893; 22: 540
² Barkan, O. "A new operation for chronic glaucoma". *Am J Ophthalmol*. 1936; 19:951. Barkan, O. "Operation for congenital glaucoma". *Am J Ophthalmol*. 1942; 25: 852.

Differential Diagnosis

- Raised IOP
- Enlarged Cornea / Globe
- Descemet's Membrane Tears / Bands
- Corneal Haze / Scarring
 - Iris Atrophy / Malformation
- Optic Nerve Head Cupping

Khan AQ Conditions that can be mistaken as early childhood glaucoma. *Ophthalmic Genet.* 2011 Sep;32(3):129-37. Epub 2011 Feb 22.

DDx: (i) raised IOP

- Co-operation (or lack of it)
- General Anaesthesia
 - increased intrathoracic pressure & ketamine can increase IOP, inhalational agents can reduce it
- Corneal Thickness
 - CCT is usually reduced in kids with childhood glaucoma, but may be increased in other conditions eg aphakia, aniridia
 - importance is unknown in kids - see Freedman SF. Central corneal thickness in children—does it help or hinder our evaluation of eyes at risk for glaucoma? *J AAPOS.* 2008;12(1):1-2.

Khan AQ Conditions that can be mistaken as early childhood glaucoma. *Ophthalmic Genet.* 2011 Sep;32(3):129-37. Epub 2011 Feb 22.

DDx: (ii) enlarged cornea / globe

- megalocornea without glaucoma
- high myopia
- proptosis
- isolated corneal ectasia

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DDx: (iii) Descemet's tears

- birth injury
 - can occur due to compression by forceps
 - splits in Descemet's are usually vertical
 - PPD
- Infantile Corneal Opacity:*
"STUMPED"
Sclerocornea
Tears in DM (birth trauma)
Ulcer
MPS
Pressure (glaucoma)
Endothelial (CHED, CHSD,
PPD) Dermoid/ Rubella
keratitis/ Cystinuria

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DDx: (iv) corneal haze

- CHED
- PPD
- Congenital Stromal Dystrophy
- Posterior Amorphous Corneal Dystrophy
- MPSs
- Peter's anomaly / Sclerocornea (ASDAs)

Infantile Corneal Opacity: "STUMPED"
Sclerocornea
Tears in DM (birth trauma)
Ulcer
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Pressure (glaucoma)
Endothelial (CHED, CHSD, PPD) Dermoid/
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DDx: (v) optic nerve head cupping

- physiological cupping
- PVL-related optic nerve hypoplasia
- papillorenal syndrome (MIM#120330)
- progressive cupping in preterm kids*

Khan AO. Conditions that can be mistaken as early childhood glaucoma. *Ophthalmic Genet.* 2011 Sep;32(3):129-37.

* Park HJ, Hampo C, Demer JI. Longitudinal study of optic cup progression in children. *J Pediatr Ophthalmol Strabismus.* 2011 May-Jun;48(3):151-6.

Classifying paediatric glaucoma

- Primary / Developmental Glaucoma
- Secondary / Acquired Glaucoma
- Trabecular Meshwork Endothelialization

Yeung HJ, Walton DS. Clinical classification of childhood glaucomas. *Arch Ophthalmol.* 2010 Jun;128(6):680-4.

1. Primary / Developmental Glaucoma

- PCG
 - Newborn PCG, Infantile PCG, Late-recognised PCG
- Juvenile OAG
- Primary ACG
- Primary Glaucoma Associated with Systemic Disease
 - eg SWS, Lowe Syndrome, Rubinstein Taybi
- Primary Glaucoma Associated with Profound Ocular Anomalies
 - eg Aniridia, PPD, Axenfeld-Rieger Anomaly

Yeung HJ, Walton DS. Clinical classification of childhood glaucomas. *Arch Ophthalmol.* 2010 Jun;128(6):680-4.

Medical Management

- temporising or short-term adjunctive use only
- plasma levels higher than adults
- relatively ineffective in general
- CAIs quite good (vs adults)
- prostaglandins in older kids
- alpha-2 agonists have a poor side effect profile

Surgical Management

- Angle Surgery
 - Goniotomy
 - Trabeculotomy
- Trabeculectomy
- Seton
- Cyclodestruction

Goniotomy

- microscope, goniolens eg Barkan lens
- one hand holds lens, the other holds gonionknife or 25G needle on Healon syringe
- can incise 90-120 degrees of angle with needle/ fine blade
- transient hyphaema common
- do temporal approach first, if needs to be repeated do nasal approach (positioning difficult)
- Advantages: short procedure, avoidance of conjunctiva
- Disadvantages: requires clear cornea, difficult to learn

Trabeculotomy

- with trabeculotome or 6/0 Prolene suture
- cut-down onto Schlemm's canal
- easier than goniotomy and doesn't require clear cornea but damages conjunctiva

Refractory Glaucoma

- Trabeculectomy
 - with or without MMC
 - 2-4mg/ml for 2-3 minutes
- Setons
 - Ahmed Valve
 - Molteno
- Cyclodestruction
 - can be used as a temporising measure eg in aphakic glaucoma
 - can be UBM guided