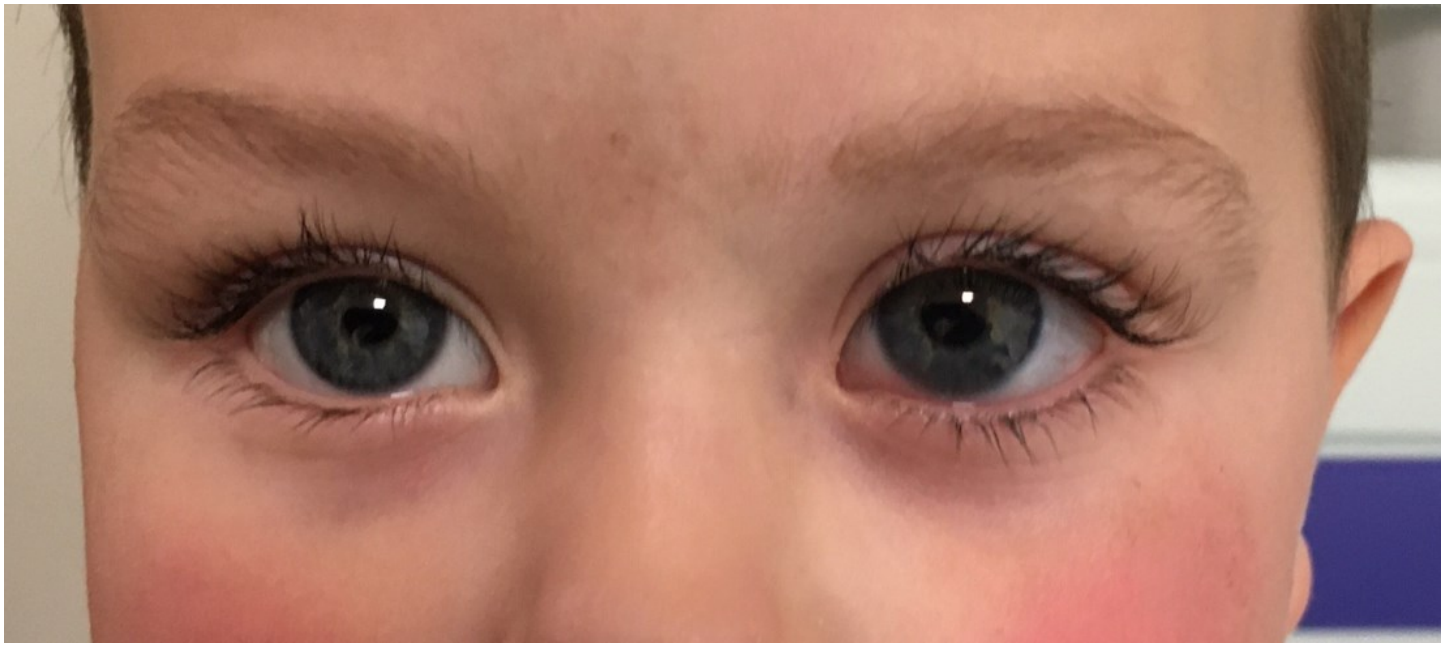


Childhood Glaucoma

How is childhood glaucoma different to adult glaucoma?

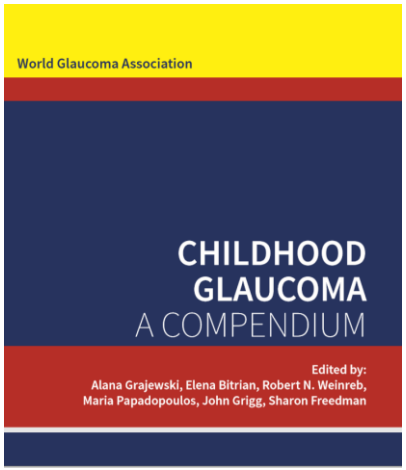
How is childhood glaucoma different to adult glaucoma?

- Aetiology
- Clinical signs
- Difficulty of assessment
- Treatment strategies
- Surgical procedures

CGRN Definition Childhood Glaucoma

2 or more:

- IOP≥ 21 mmHg
- Optic disc cupping with progression
- Increase in cup/disc ratio or asymmetry >0.2 or focal rim thinning
- Haab striae or enlarged corneal diameter
- >11 mm in newborn
  - » >12 in child less 1 year old
  - » >13 any age
- Progressive myopia with increase in ocular dimensions
- Visual field defect consistent with glaucomatous optic neuropathy



CGRN Definition Childhood Glaucoma Suspect

At least one:

- IOP> 21 mmHg on two separate occasions
- Suspicious optic disc appearance for glaucoma (*i.e.* increased cup to disc ratio)
- Suspicious visual field for glaucoma
- Increased corneal diameter or axial length in setting of normal IOP

Childhood Glaucoma Classification Overview

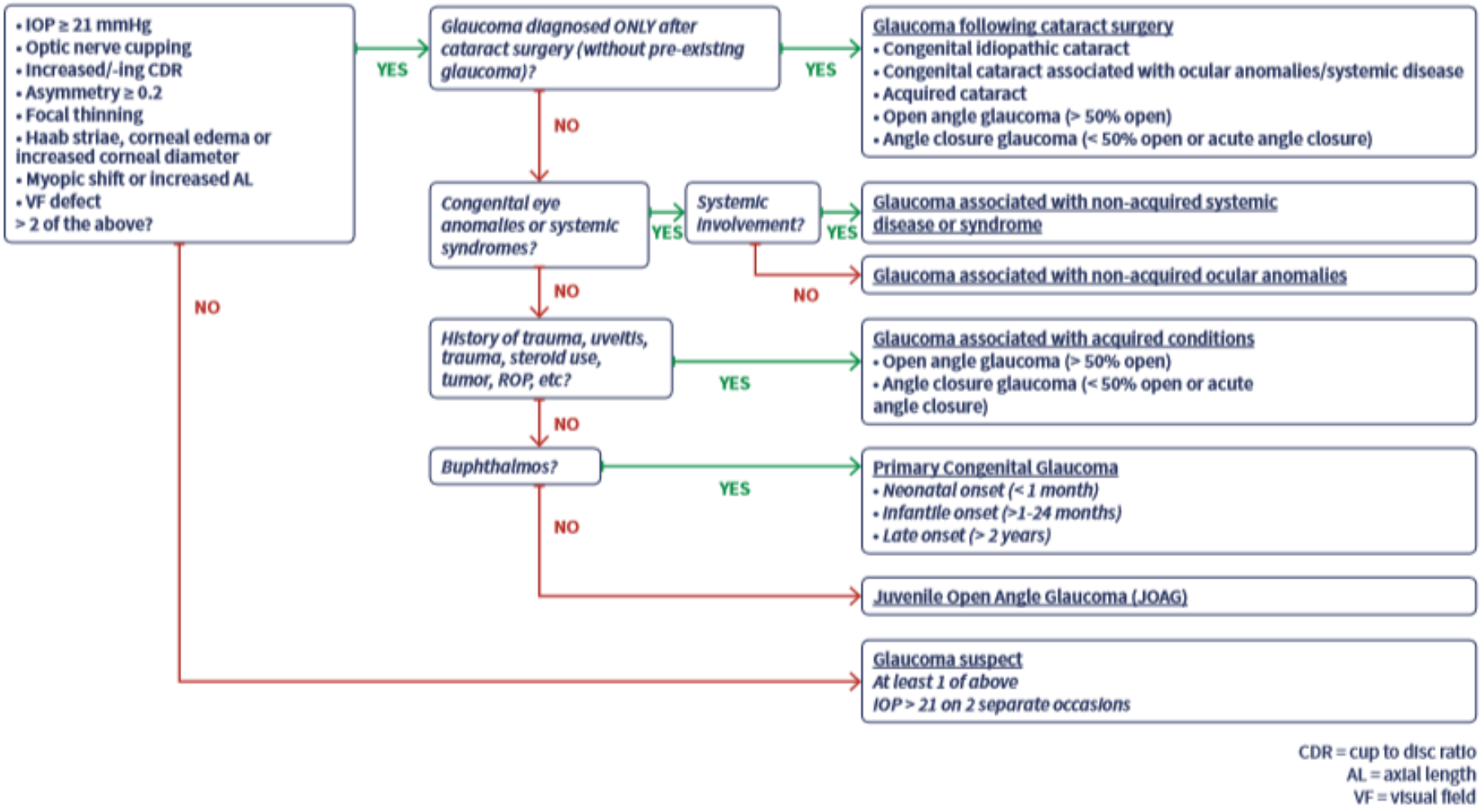
Primary

- Primary congenital glaucoma (PCG)
  - i. Neonatal or newborn onset (0-1 month)
  - ii. Infantile onset (>1-24 months)
  - iii. Late onset or late-recognized (>2 years)
- Juvenile open angle glaucoma (JOAG)

Secondary

- Non-acquired ocular anomalies
  (*i.e.* Axenfeld Riegers Syndrome, Peters anomaly, aniridia)
- Non-acquired systemic disease or syndrome
  (*i.e.* Sturge-Weber syndrome, Marfan syndrome, Congenital rubella)
- Acquired condition
  (*i.e.* uveitis, trauma, steroid induced, tumor, retinopathy of prematurity)
- Following cataract surgery

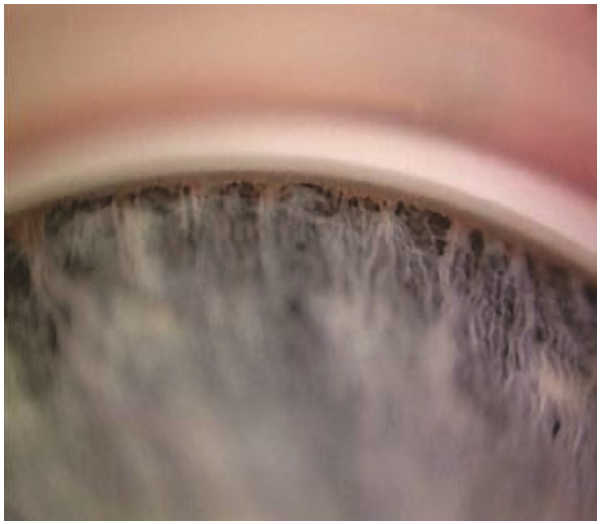
Childhood Glaucoma Classification Algorithm



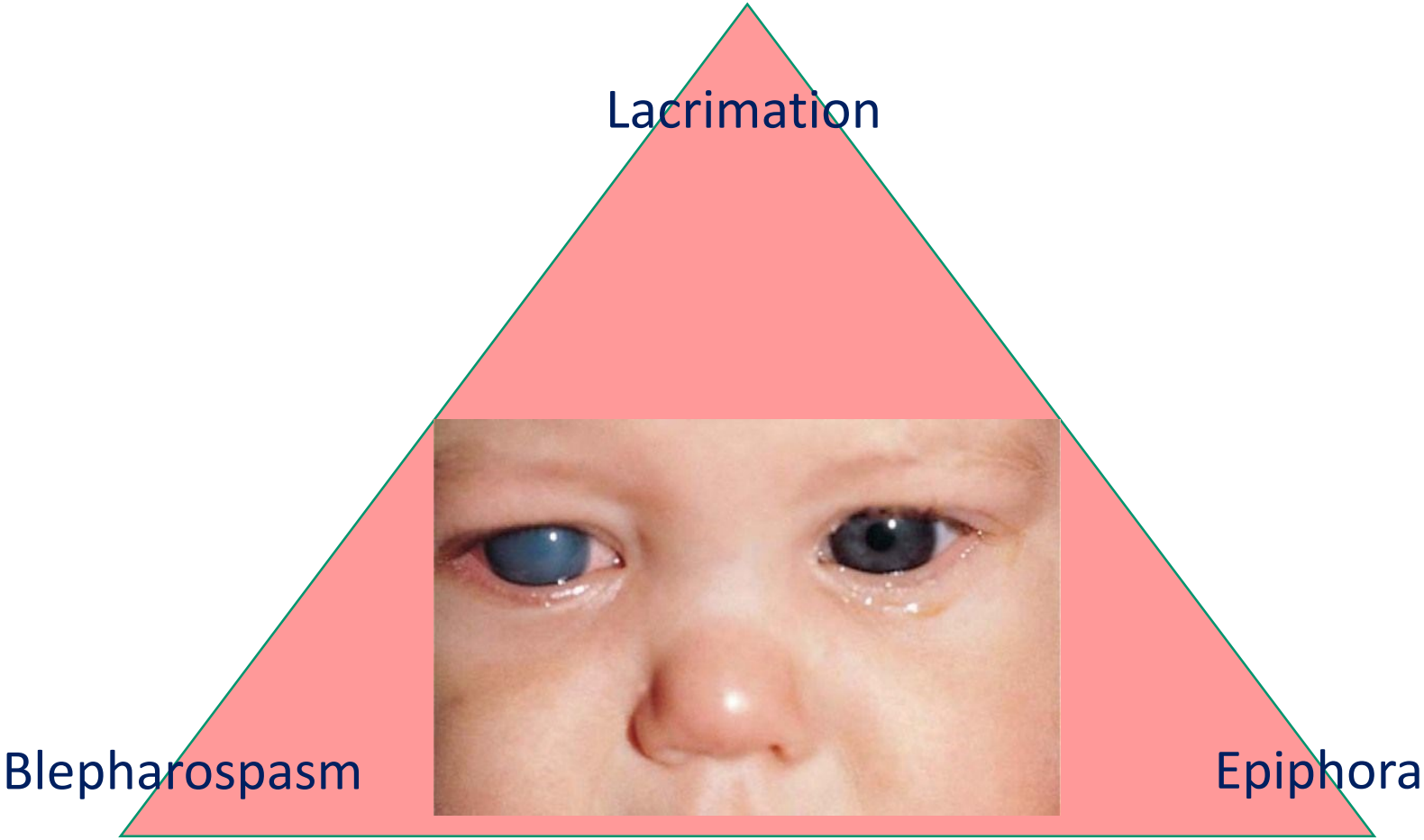
CDR = cup to disc ratio
AL = axial length
VF = visual field

Primary Congenital Glaucoma

- Usually sporadic, FH in 20% AR with variable penetrance
  - *CYP1B1,* variable expressivity
  - Sibling risk <5% but examine all (esp if consanguinous)
- Bilateral in 80%, often asymmetrical
- 1/10,000 births, higher prevalence with consanguinity
- 2M: 1F
- Isolated trabeculodysgenesis
- “Barkan membrane”
  - thickened trabecular sheets
  - obstruct outflow
  - prevent posterior migration of the iris / ciliary body
  - Iris appears hypoplastic

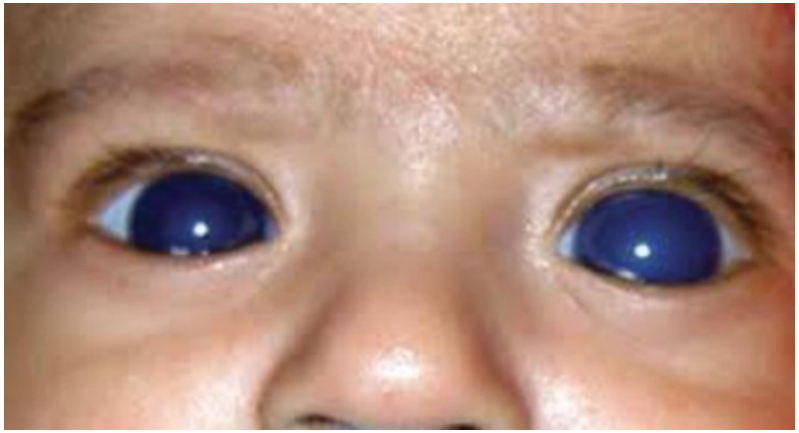


PCG - presentation

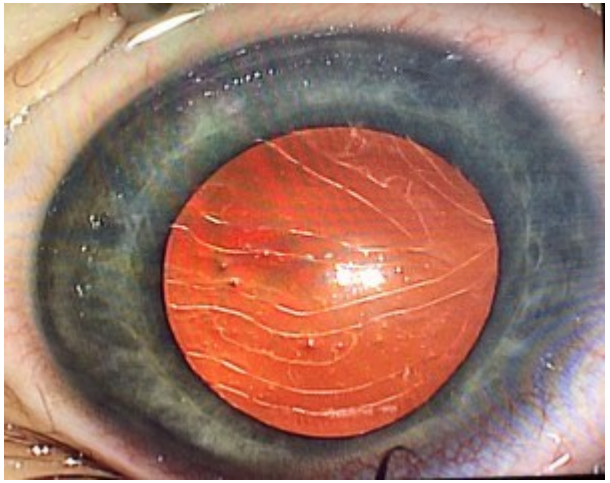


PCG signs

PCG signs

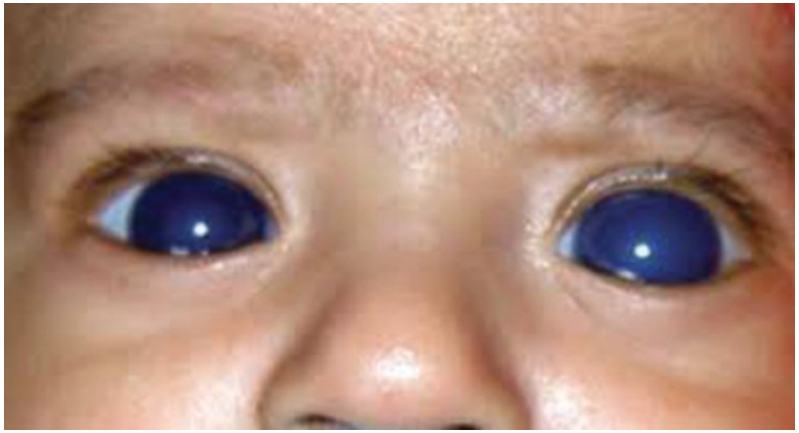


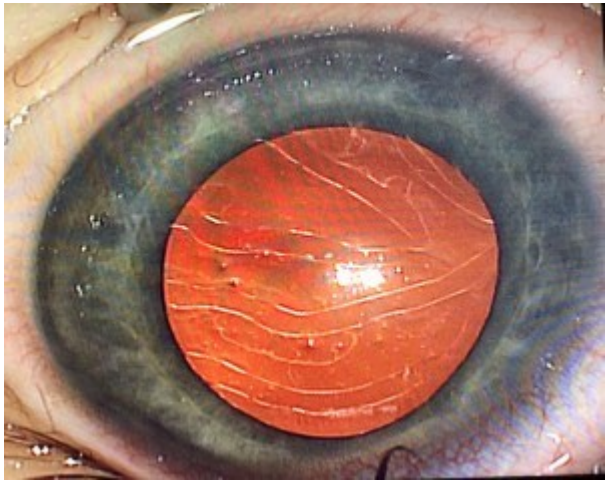
- Globe enlargement / buphthalmos
- Increased HCD
- Corneal oedema
- Raised IOP (Icare overestimates IOP)
- Haabs striae
- Myopia
- Disc cupping
- EUA
  - Abnormal gonioscopy
  - Baseline CCT
  - Increased axial length
  - All anaesthetic agents (exc ketamine) lower IOP
  - Blood for genetic testing



PCG differential diagnosis

- Increased HCD
  - XL megalocornea
  - *LTBP2* mutations
  - Congenital high myopia
  - Connective tissues disorders: OI, marfan
- Corneal striae
  - Trauma
- Corneal oedema
  - Trauma
  - Corneal dystrophies eg CHED
  - Metabolic disorders eg MPS
  - Anterior segment dysgenesis
- Lacrimation / epiphora
  - CNLDO
  - keratitis
- Disc cupping
  - Large optic nerves – physiological
  - Congenital pits or coloboma
  - Late optic nerve hypoplasia with CVI





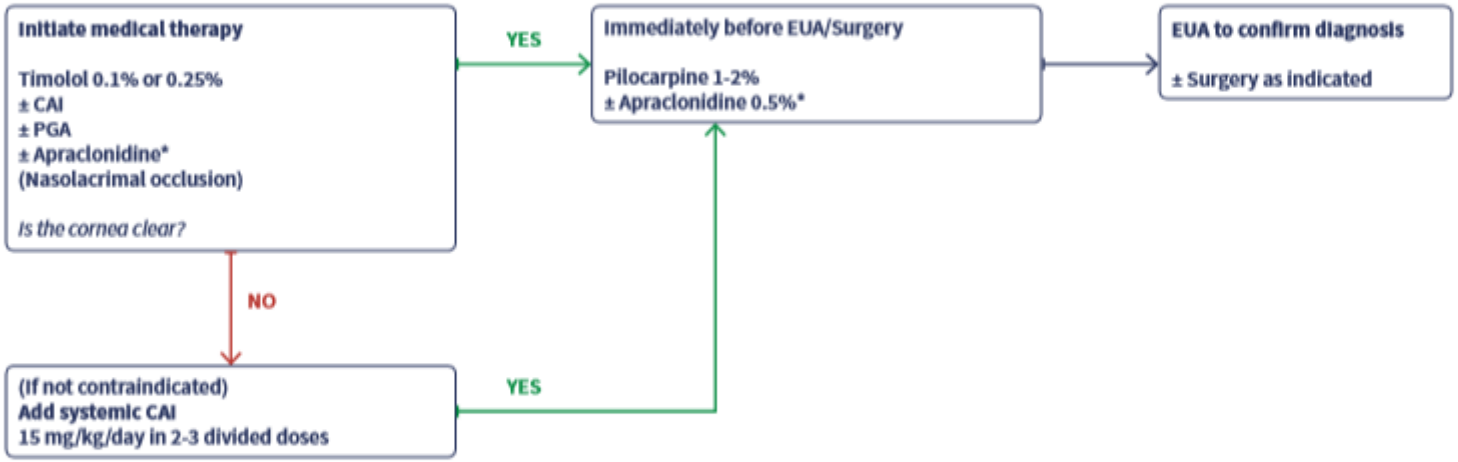
Glaucoma drops in children

Medication type	Indications	Contraindications / side effects
β-blockers Non-selective (timolol, levobunolol, carteolol) Selective (betaxolol)	1st or 2nd line therapy Non-selective drugs more effective Selective drugs safer with asthma	Bronchospasm, bradycardia Avoid in premature or small infants or reactive airways Start 0.1% or 0.25% in small children
Carbonic anhydrase inhibitors Topical (dorzolamide, brinzolamide; 2-3 times/day) Oral (5mg/kg 3 times a day 2-4 ti	1st or 2nd line therapy Topical better tolerated that oral but not as effective	Later option in corneal transplant patients Metabolic acidosis may occur with oral therapy
Miotics Echothiophate Pilocarpine	Echothiophate rarely used Pilocarpine after angle surgery	Headache, myopic shift, pro-inflammatory effect, diarrhea (echothiophate)
Adrenergic agonists Epinephrine	Rarely used	Hypertension, tachycardia
α_2-agonists Apraclonidine 0.5%		Use with caution only if>6/12
Brimonidine use lowest concentration 0.10 % Do not use	During/after angle surgery, short term 2nd or 3rd line therapy	**Do not use in children <40 lbs** Bradycardia, hypotension, hypothermia, hypotonia, apnea (specially with β-blocker)
Prostaglandins (latanoprost, travoprost, bimatoprost, tafluprost)	1st, 2nd or 3rd line with JOAG, 2nd or 3rd line with others	Systemically safe Long eyelashes, redness Trial in uveitic glaucoma as last resort

PCG management options

Case scenario 1

Scenario 1 – 3 month old baby, full-term and otherwise well, with bilaterally large and cloudy corneas, suspected of having primary congenital glaucoma. Pressures are in the 30s both eyes, and angle surgery is planned in the next few days.



*Apraclonidine should be used with great caution, with warning to parents of potential side effects.

CAI = carbonic anhydrase inhibitor
PGA = prostaglandin analogue
EUA = examination under anesthesia

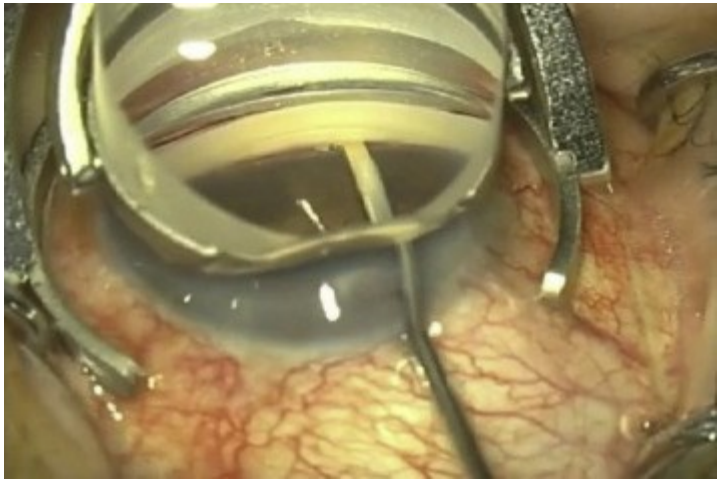
Table 1 Comparison of Angle Surgery techniques

From: Childhood glaucoma surgery in the 21st Century

	Goniotomy	Trabeculotomy	Trabeculotomy (360°)		Trabeculotomy-Trabeculectomy
		(probe)	Suture	Illuminated microcatheter	
Conjunctival incision	–	+	+	+	+
Possible with opaque cornea (angle structures not clearly visible)	–	+	+	+	+
Angle visualisation	+	–	– (only with gonio lens)	+	–
Specialised instruments/skills	++	+	+	++	++
Extent of surgical trauma	+	++	++	++	+++
Safety	++	+	+	++	+
Duration of surgery	+	++	++ to +++	++ to +++	++
Can be repeated	++	++	–	–	+
Bleb-related complications	–	–/+	–/+	–/+	+

Papadopoulos, M., Edmunds, B., Fenerty, C. *et al.* Childhood glaucoma surgery in the 21st Century. *Eye* **28,** 931–943 (2014). https://doi-org.ezp.lib.cam.ac.uk/10.1038/eye.2014.140

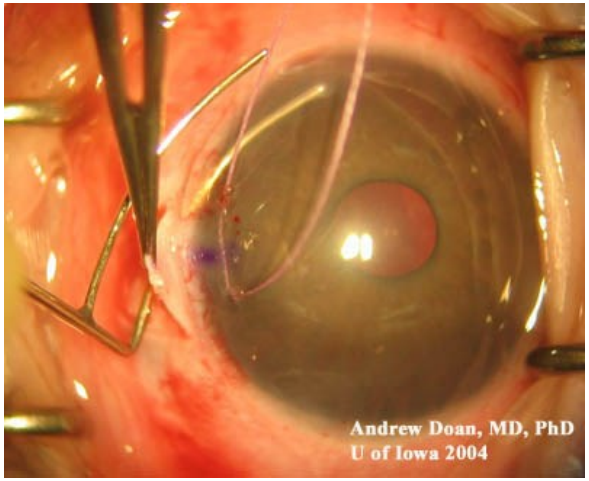
PCG surgical planning – angle surgery



Goniotomy

- Needs a clear cornea
- Requires repeating
- Quoted high success rate
- Works less well if HCD >14, AL >24mm, early onset

PCG surgical planning – angle surgery



Ab externo trabeculotomy

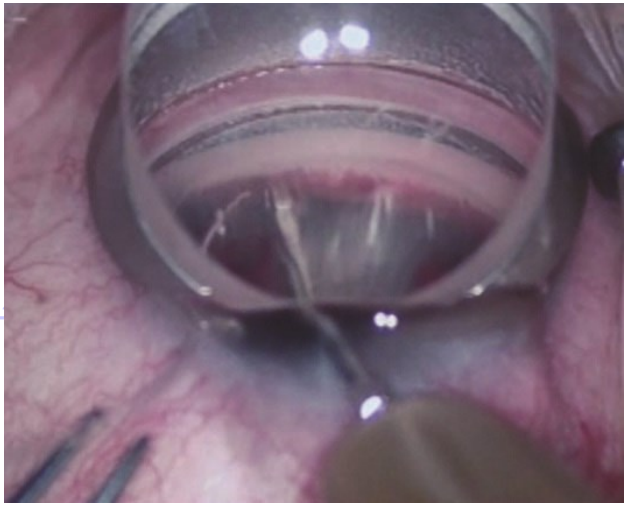
- OK with cloudy cornea
- Difficult to find SC
- Conj scarring
- Similar success rate

Ab interno trabeculotomy

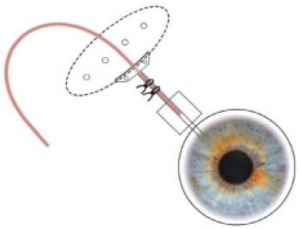
Gonioscopy assisted trabeculotomy

itrack

https://www.youtube.com/watch?v=UDZS7S-



PCG surgical planning – glaucoma drainage device



- Molteno tube / Ahmed valve
- Best for glaucoma following cataract surgery, uveitis
- Advantages
  - Effective in lowering IOP long term 50%
  - Most likely to survive future surgery
- Disadvantages
  - Long surgical time
  - Often need topical therapy too
  - High surgical revision rate – plate encapsulation / erosion
  - Higher risk of corneal decompensation

PCG surgical planning – MMC trabeculectomy

- First line in most secondary glaucoma types except those with poor visual prognosis eg aphakia / pseudophakia
- Second line to Angle surgery in PCG
- Moorfields safer surgery (MMC, posteriorly directed drainage
- MMC 0.2mg/ml, AC maintainer
- Risk of endophthalmitis with cystic bleb

PCG surgical planning

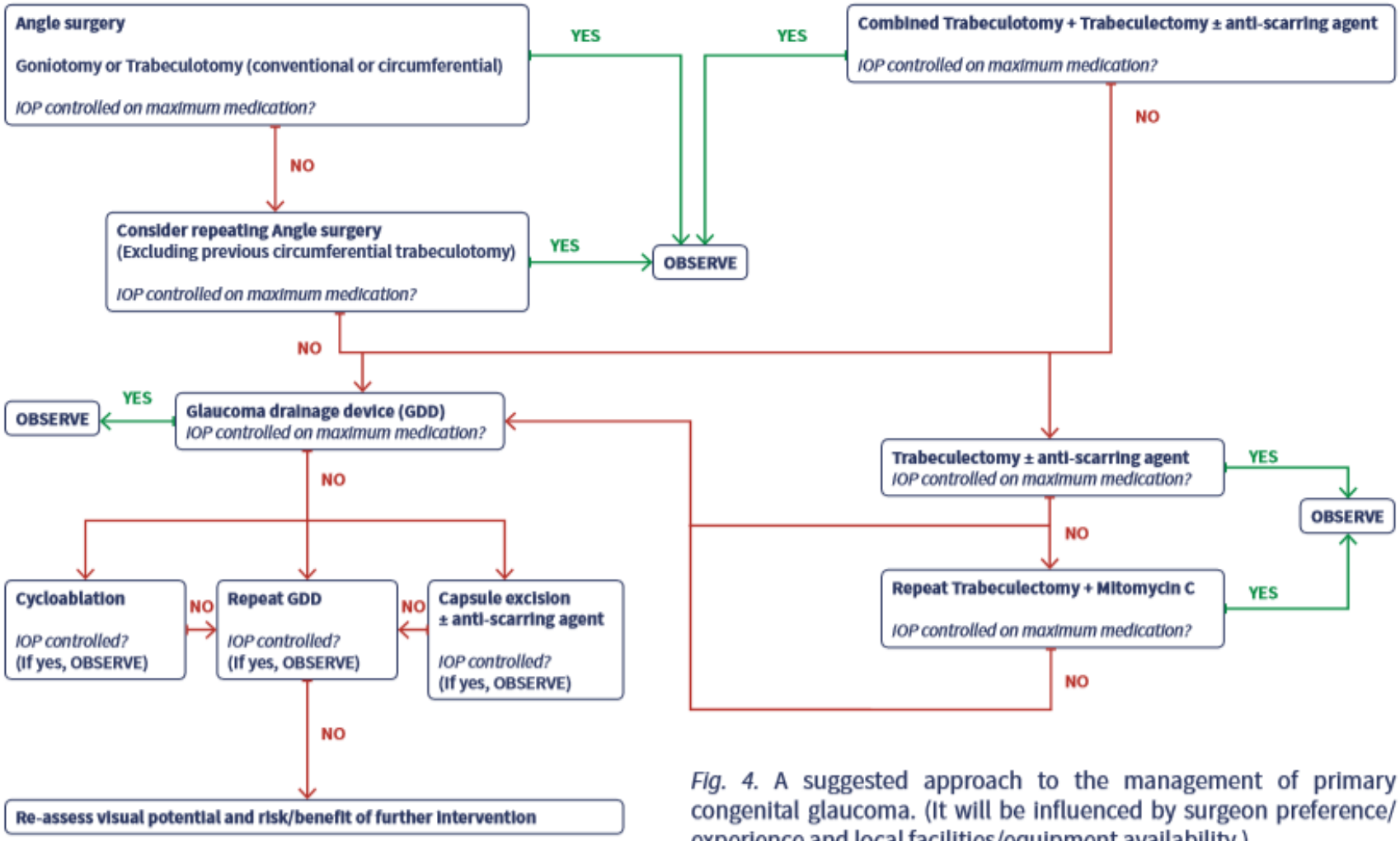
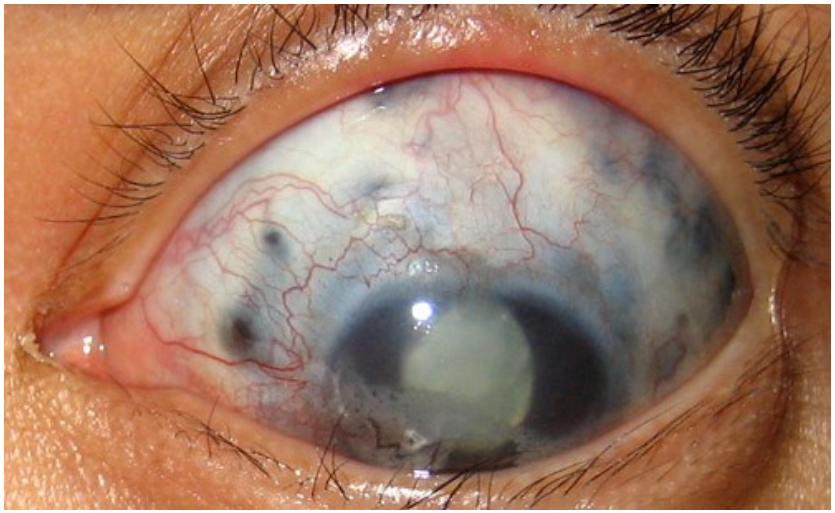


Fig. 4. A suggested approach to the management of primary congenital glaucoma. (It will be influenced by surgeon preference/ experience and local facilities/equipment availability.)

Outcomes

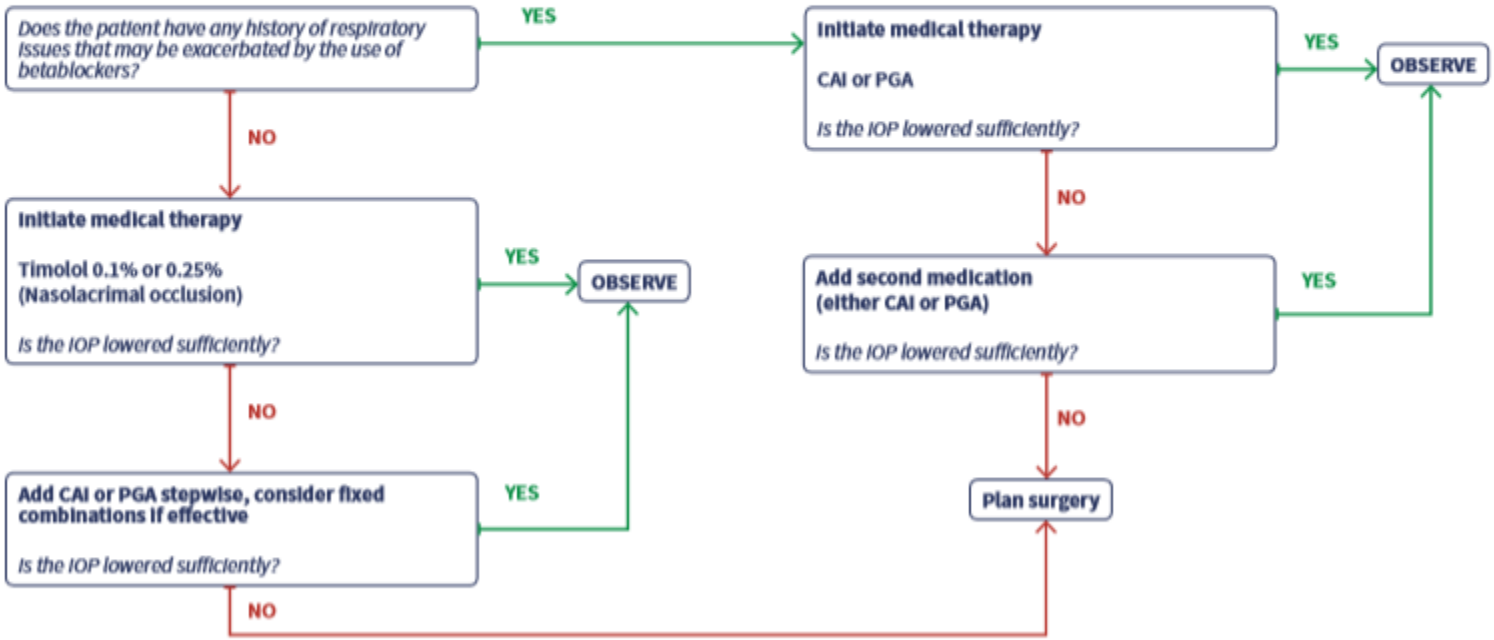
- 50% achieve vision of 6/15 or better
- Unilateral: 40% achieve this (amblyopia)



Glaucoma following cataract surgery

Case scenario 2

Scenario 2 – 6 month old baby with bilateral aphakia, just noted to have elevated IOP to the 30s and mild corneal haze and slight enlargement of one eye. Optic nerve looks normal without cupping. The other eye is unremarkable with
IOP of 15mmHg. Anterior chamber is deep and iris plane is flat in both eyes.
a) Baby is healthy otherwise and full-term.
b) Baby has had respiratory syncytial virus (RSV) infection and requires use of a nebulizer periodically



CAI = carbonic anhydrase inhibitor
PGA = prostaglandin analogue

Glaucoma following cataract surgery

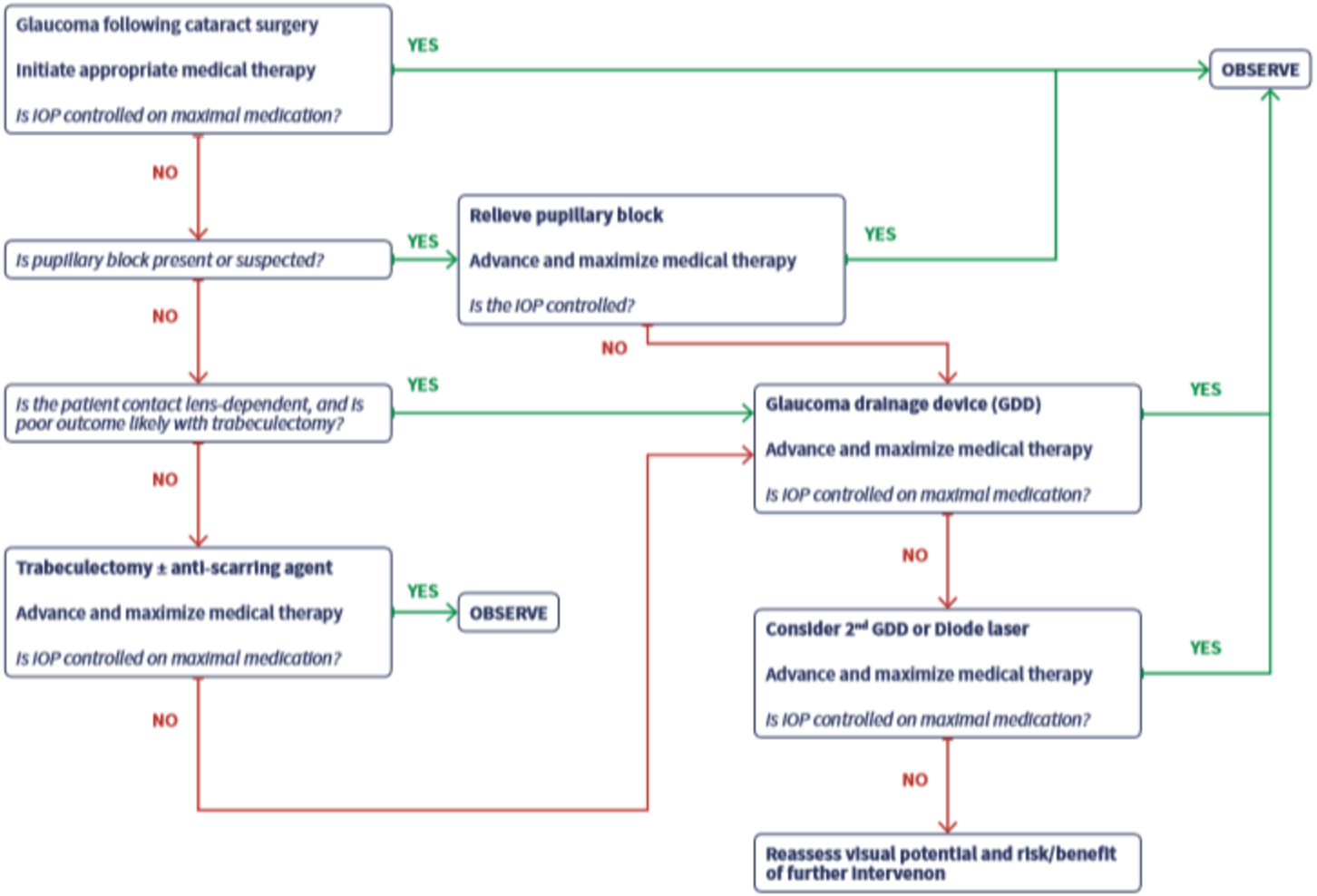


Fig. 8. A suggested approach to the management of glaucoma following cataract surgery. (It will be influenced by surgeon preference/ experience and local facilities/equipment availability.)

Steroid induced glaucoma

- Due to increased outflow resistance
  - upregulation of glucocorticoid receptors in trabecular meshwork cells
  - Increased expression of GAGS, elastin and fibronectin
  - Suppression of phagocytic activity, increased debris in meshwork
- Risk factors
  - POAG of 1st degree FH
  - Previous IOP rise on steroids
  - Age <6 years
  - Type 1 DM, connective tissue disease,
  - High myopia
  - The higher the steroid potency and penetration, the greater the hypertensive response
- Topical and peri-ocular steroid, 50% get IOP rise following i/v vitreal-S
- Lesser risk from oral or inhalational
- Discontinue or change steroid therapy and treat glaucoma
- Where long term steroid is needed, treat and face the consequences!