

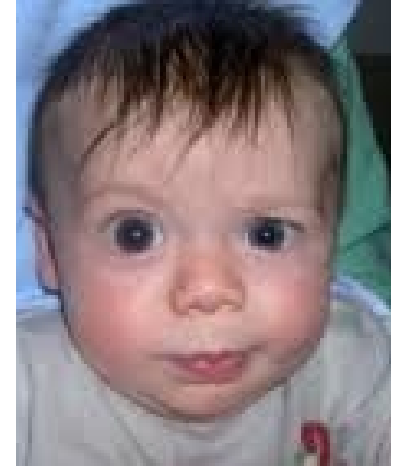


Clinical Subtypes of Primary Congenital Glaucoma (PCG)

By,

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The aim of this presentation is to show different clinical subtypes of PCG and how PCG subtypes are clues for surgical approach



1. Clinical subtypes according to IOP, corneal diameter and corneal integrity

- **El Hazmi et al 2005** published a larger study showing classification of PCG into mild, moderate, or severe forms, based on IOP, corneal diameter, and corneal integrity can be helpful for surgical decision making.

EXTENDED REPORT

Correlation between surgical success rate and severity of congenital glaucoma

A Al-Hazmi, A Awad, J Zwaan, S A Al-Mesfer, I Al-Jadaan, A Al-Mohammed

Br J Ophthalmol 2005;89:449-453. doi: 10.1136/bjo.2004.047761

Aim: To study the correlation between severity of primary congenital glaucoma (PCG) and success of three types of surgery.

Methods: This was a retrospective review of all records of patients diagnosed with PCG up to age 1 year who underwent goniotomy, trabeculotomy, or combined trabeculotomy-trabeculectomy with mitomycin C as initial procedure between 1982 and 2002 at the King Khalid Eye Specialist Hospital, Riyadh, Saudi Arabia. 532 paediatric glaucoma patients below age 1 year (820 eyes) with a minimum 1 year follow up were identified. The main outcome measures used for the surgeries were postoperative intraocular pressure, stability of the corneal diameter, and maintenance of corneal clarity. Surgical success was defined as a postoperative intraocular pressure of ≤ 21 mm Hg without additional medical or surgical therapy, and with decreased corneal oedema, stabilised corneal diameter, and no additional optic nerve damage for at least 1 year after surgery. Complications, time of surgical failure, and follow up were recorded.

Results: The eyes were grouped into mild (249), moderate (342), and severe (229) PCG, based on intraocular pressure, corneal diameter, and clarity. All three surgical procedures resulted in high success rates of 81-100% for the mild form of PCG. Eyes classified with moderate glaucoma had a 13%, 40%, and 80% success rate respectively for goniotomy, trabeculotomy, and combined trabeculotomy-trabeculectomy with mitomycin C. The success rate for severe PCG was 10% and 70% for trabeculotomy and combined surgery respectively. Goniotomy was never done for eyes with this condition.

Conclusion: Clinical classification of PCG is helpful for surgical decision making. The mild form has a high surgical success regardless of the procedure chosen. Combined trabeculotomy-trabeculectomy with mitomycin C gave the best results for moderate and severe cases of PCG.

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El Hazmi et al
2005

- The 820 eyes with PCG were classified into mild, moderate, and severe cases based on IOP, corneal diameter, and corneal integrity or clarity.

Table 1 Classification used for primary congenital glaucoma

Severity	IOP (mm Hg)	Corneal diameter (mm)	Corneal clarity
Mild	<25	<13	Good
Moderate	25-35	13-14.5	Fair
Severe	>35	>14.5	Poor

- The mild form of congenital glaucoma generally responds successfully to surgery, no matter which procedure is chosen.
- Combined trabeculotomy-trabeculectomy with adjunctive mitomycin C results in better IOP control in the moderate and severe forms.

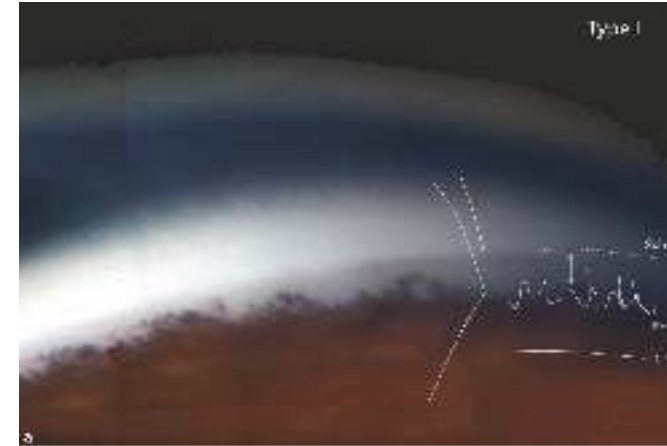
Table 4 Outcome of three surgical procedures for primary congenital glaucoma in 820 eyes

Severity	Goniotomy			Trabeculotomy			CTM ^a		
	No	Success (%)	Failure (%)	No	Success (%)	Failure (%)	No	Success (%)	Failure (%)
Mild	145	117 (81)	28 (19)	102	93 (90)	10 (10)	2	2 (100)	0 (0)
Moderate	109	14 (13)	95 (87)	163	65 (40)	98 (60)	70	56 (80)	14 (20)
Severe	0	0	0	153	15 (10)	138 (85)	76	53 (70)	23 (30)
Total	254	131 (52)	123 (48)	418	173 (41)	246 (59)	148	111 (75)	37 (25)

^aCTM=combined trabeculotomy-trabeculectomy with mitomycin C.

2. Clinical subtypes according to angle pathology

- According to **Sampaolesi et al 2009** there are two types of pathological chamber angle in children with PCG
- **Type I** has thin pathological mesodermal remnants that cover the ciliary band of the chamber angles and sometimes slender extensions reach the Schwalbe line.
- **Type II** chamber angles are identified by the presence of thick pathological mesodermal remnants with apparent high iris insertion with black triangles and pillars. In both types, it is impossible to distinguish the ciliary body band.



Clinical application of angle pathology

Type I	Type II
Pathological mesodermal remnants	Apparent high insertion of iris
Axial length: up to 23 mm	Axial length: >23 mm
Corneal diameter: up to 13 mm	Corneal diameter: >13 mm
Trabeculotomy	Combined surgery: trabeculotomy + trabeculectomy
<i>Sampaolesi et al 2009</i>	



3. PCG Subtypes according to Age of Onset

David S. Walton 2012 showed that There are 4 variants based on the age of presentation as follows:

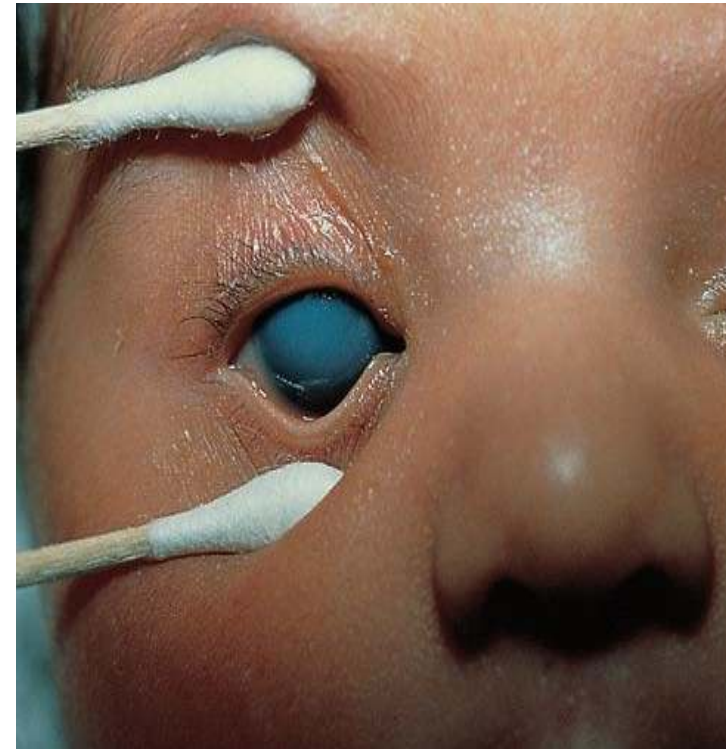
1. **Newborn** onset (0-1 month)
2. **Infantile** onset (between 1-12 months)
3. **Late onset** or late-recognized (12 months)
4. **Spontaneously arrested** cases (very rare)

•*David S. Walton. MD, Pediatric Ophthalmology 2012 at the annual meeting of the American Academy of Ophthalmology.*



Newborn PCG (0-1 month)

- **Newborn PCG** represents about 10% of all cases of PCG. It is usually recognized within the first few days of life based on striking corneal opacification that is the result of elevated IOP and corneal anomalies.
- These children also have a very thin iris and hypoplasia of the filtration angle with minimally exposed trabecular meshwork.
- *Marchini et al. Pediatric glaucoma: current perspectives Pediatric Health, Medicine and Therapeutics 2014;5 15–27*



Newborn PCG (0-1 month)

- There is a **debate** in the literature about **angle surgery** for newborn PCG where the filtration angle is not adequately developed to support good results.
- However, recent studies show 60% success rate of first **angle surgery** is for newborn PCG.
- *Sushmita Kaushik, et al. Indian J Ophthalmol 2021;69:2088-94.*

initial and re-surgery for childhood glaucoma in Northern India

Gupta, Faisal Thattaruthody, Deepika Dhillon, Kira Sagarika Suehi, Srishti Raj, Surinder S Pandav

describe the effectiveness of goniotomy for childhood glaucoma patients with pediatric glaucoma who underwent goniotomy between 2010 and 2019 in Northwest India were prospectively analyzed. Goniotomy as a re-do surgery of the untreated angle in failed filtering surgery (IOF) ±18 mm Hg with or without 2 topical medications underwent goniotomy during this period (37.9% of all pediatric glaucoma surgeries). 132 of 211 (62.5%) primary pediatric glaucoma surgeries, 112, and 54 eyes had a six months, 1-year and 2-year follow-up. In Primary Congenital Glaucoma (PCG) were 79.7% for primary PCG eyes, the success rate was 62% at 1 year. Among non-PCG eyes, the success rate was 87.5% and 57.1% success rates, respectively. On longer follow-up, eyes with shorter axial length at presentation were significantly predictive of success ($P = 0.02$, respectively). At 1 year, in the primary surgery group, 28.9% had moderate (better than LOGMAR 1.0) and were no significant intraoperative or post-operative complications. Goniotomy is an effective surgery for childhood glaucoma in Indian eyes. It is safe for conjunctival and scleral dissection and antifibrotic agents.

Infantile PCG (between 1-12 months)

- **Infantile PCG**, which is the largest clinical subtype of PCG, is usually diagnosed between 1 and 12 months of age. These children present with large eyes and also have corneal findings that are secondary to the increased IOP.

- **Gonioscopy** will reveal an open angle and a normal trabecular meshwork, but with variable narrowing and opacification of the ciliary body band and a minimally visible scleral spur.

- **Angle surgery** for infantile PCG has a reported success rate that exceeds 90%.



Late-recognized PCG (12 months)

- Late-recognized PCG includes children whose disease is diagnosed past 1 year of age.
- The main presenting features of late-recognized PCG is corneal enlargement is the main finding in children ages 1 to 4 years old, and they may also have some photophobia. Late recognized PCG is associated with moderate angle anomalies.
- Angle surgery for Late-recognized PCG also has a high success rate.

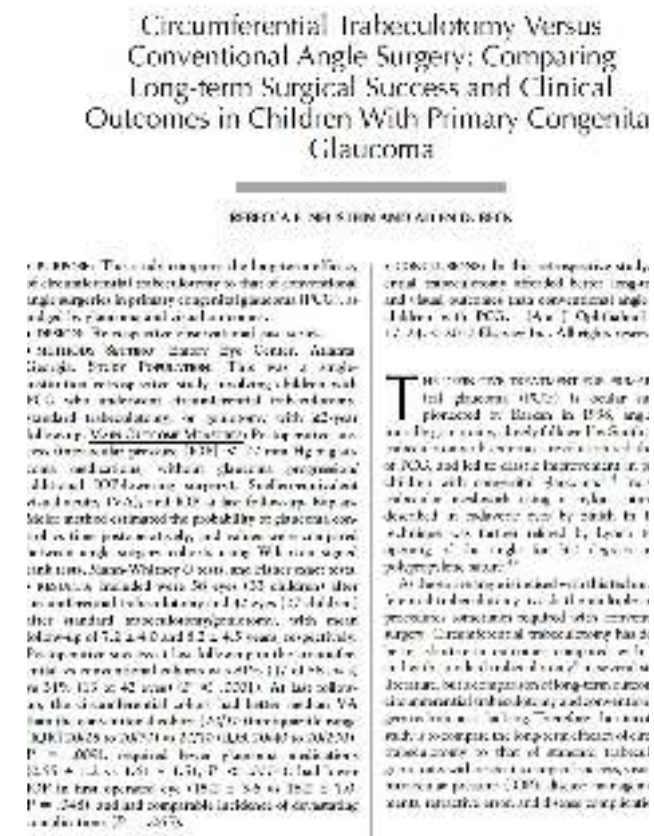


•David S. Walton. MD, *Pediatric Ophthalmology 2012 at the annual meeting of the American Academy of Ophthalmology.*

Circumferential versus conventional angle surgery

- Currently, angle surgery is considered in terms of 360° degree angle surgery (360° degree trabeculotomy by either ab-interno (GATT) or ab-externo approaches) **increased** the success rate of PCG.

- *Am J Ophthalmol* 2017;183:17–24.





Best Wishes