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Bilateral Congenital Glaucoma – A Rare Case Report

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Abstract

Background: Glaucoma in childhood is a potentially blinding condition, yet, there is a lack of epidemiologic and clinical data regarding this pathology. Unlike glaucoma in adults, which is notoriously difficult to detect since there are no symptoms, glaucoma in children does present with signs and symptoms which can be detected by paediatricians and even parents, who are usually the first contacts of these children. **Methodology:** A detailed evaluation under general or local anaesthesia is advisable to establish the diagnosis and plan for management. Medical therapy has a limited role and surgery remains the primary therapeutic modality. While goniotomy or trabeculotomy ab externo is valuable in the management of congenital glaucoma, primary combined trabeculotomy–trabeculectomy offers the best hope of success in advanced cases. **Result:** The childhood glaucoma research network (CGRN) has provided a new classification system of paediatric glaucoma's. Careful diagnosis and timely intervention leads to the most promising outcome and gives these children a long morbidity free life. Early diagnosis, prompt therapeutic intervention and proper refractive correction are keys to success. **Conclusion:** Management of residual vision and visual rehabilitation should be an integral part of the management of children with low vision and lifelong follow-up is a must.

Key Words: Congenital glaucoma, trabeculotomy, trabeculectomy and combined trabeculotomy–trabeculectomy

Case Report

The baby was a one-day old male baby, delivered via emergency LSCS. It was a full term delivery with birth weight of 2.330kg in Sree Balaji medical college and hospital on 24th September 2019. The delivery was uneventful and the general condition of both mother and baby were stable. Ophthalmologist were called to the NICU for an opinion regarding the bilateral bluish discoloration of the baby's eyes. On examination of the baby there was bilateral bluish discoloration of the cornea, along with increased corneal diameter. Due to the diffused edema, details of structures posterior to cornea could not be perceived. Under local anesthesia corneal diameter and IOP were measured.



Figure 1 and 2: bilateral bluish discoloration of the cornea



Figure 3 and 4: measurement of IOP and corneal diameter under LA

		Right eye	Left eye
Corneal diameter	Horizontal	12mm	12mm
	Vertical	11mm	11mm
IOP	5.5gm	40.0 mmhg	41.5 mmhg
	7.5gm	36.0 mmhg	35.8 mmhg

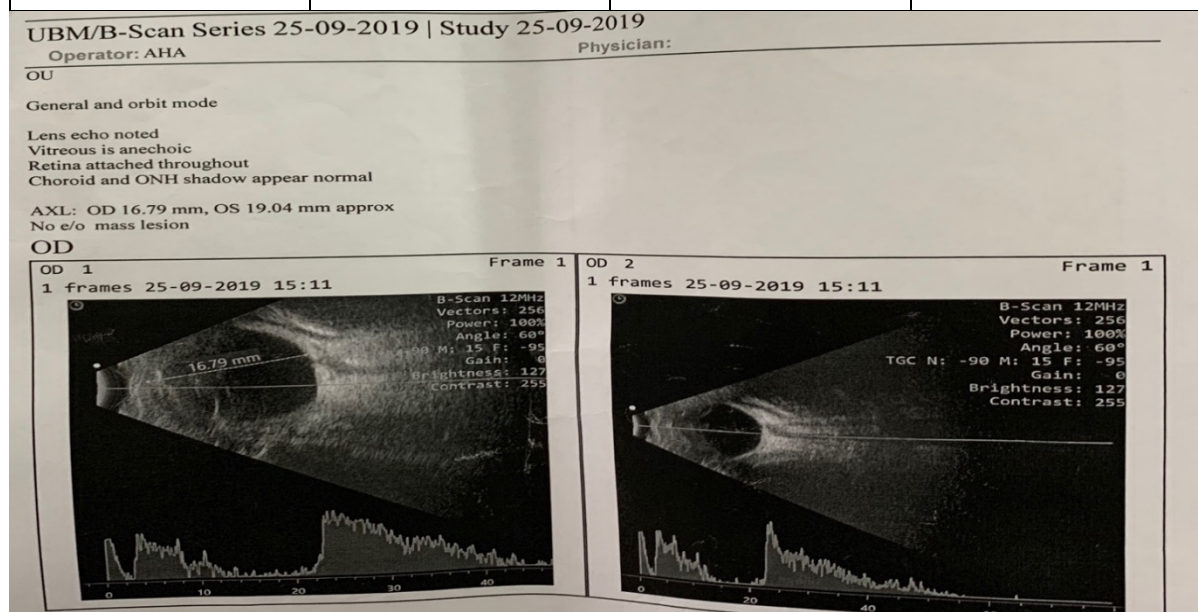


Figure 5: (USG) – Bscan was done to rule out posterior segment pathology

Management of The Baby

The baby was started on a topical beta blocker eye drops and antibiotics. The baby was then referred to a glaucoma specialist with experience in congenital glaucoma treatment. After a thorough examination under anaesthesia, trabeculotomy was performed in the right eye and at a later point in time in the left eye. The baby is on regular follow up with the ophthalmologist and the paediatrician.

Family History

On taking detailed Family history, the grandmother revealed she had a twin delivery and both her daughters (which include the baby's mother) had similar complaints at birth. They underwent a certain surgical procedure back then which was around 40 days post birth, that on ocular examination appears to be peripheral iridotomy. Most probably by Scheie's procedure. On examination of the mother, there was a very apparent alternating divergent squint

(exotropia), in right eye there is an updrawn pupil, whereas in left eye, a peripheral iridotomy scar is seen in the 7'o clock position. Bedside visual acuity both eyes were $>5/60$.



Figure 8: post trabeculotomy of the right eye
Figure 9: post trabeculotomy of both eyes

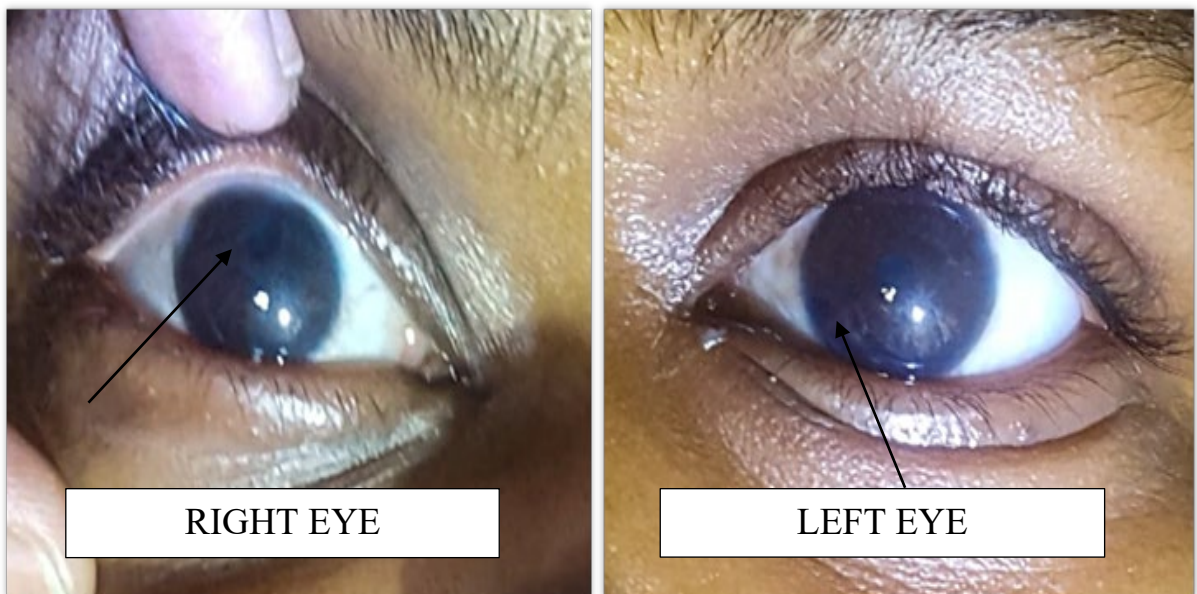


Figure 6: right eye of the mother showing updrawn pupil
Figure 7: left eye of the mother showing a peripheral iridotomy scar

Results And Discussion

Primary Congenital Glaucoma

Congenital glaucoma was the most common form of paediatric glaucoma accounting for nearly 38% of all childhood glaucoma's (Taylor *et al.*, 1999). In India a large scale study calculated the incidence to be 1 in 3300 in Andhra Pradesh.³ Primary congenital glaucoma is rare, with an incidence of 1: 10000 population. It can be classified as

- True Congenital Glaucoma(40%) in which IOP is elevated during intrauterine life;
- Infantile Glaucoma (55%) which manifest prior to age 3 and
- Juvenile Glaucoma which is the least common, in which the IOP rises between 3 and 16 years of age.

PCG is thought to be caused by impaired aqueous outflow due to maldevelopment of the AC angle. The prognosis is dependent on severity and age of onset/ diagnosis; in true congenital glaucoma. The Classical triad of congenital glaucoma includes epiphora, photophobia, and blepharospasm. Characteristically, the patients are male (65%) with bilateral involvement (70%) and diagnosed within the first year of life (80%). Visual loss results from Descemet breaks, corneal oedema, and optic neuropathy, and eventually buphthalmos, and amblyopia. Buphthalmos is a large eye, that is a result of stretching due to elevated IOP prior to the age of 3 years. The thinned sclera often appears blue due to increased visualization of the underlying uvea. Haab striae are curvilinear healed breaks in descemet membrane, complications include myopia and lens subluxation. We have a very small interval or a window period where these children can be successfully treated. If not, the disease can be potentially blinding (Fung *et al.*, 2013).

The current standard of care involves Medical Therapy (prior to angle surgery) as first line therapy usually in the form of Beta-blocker + pilocarpine. Medical therapy after incisional surgery includes prostaglandin analogue/ beta-blocker as first line and Carbonic anhydrase inhibitor as second. Alpha- agonists are contraindicated in infants due to high risk of central depression. Systemic agents are usually needed only temporarily prior to surgery. Medical therapy provides a temporizing measure to clear the cornea to facilitate examination and surgical intervention. Surgical intervention is the definitive treatment. The first two and the

most relevant two surgical procedures devised for PCG are goniotomy and trabeculotomy (Dandona *et al.*, 1998).

Goniotomy creates a route for aqueous drainage through Schlemm's canal by incision of the trabecular meshwork under direct visualization. The success of goniotomy is thought to be approximately 80% with a single procedure. In India, nearly all patients present with clouding and goniotomy is technically impossible (Sood *et al.*, 2018)

Trabeculotomy on the other hand involves disrupting the tissue between Schlemm's canal and the anterior chamber using an ab externo approach to create direct communication and in contrast to goniotomy can be done in an eye with corneal opalescence. High success rates ranging from 87% to 92% in cases done before one year of age have been reported (Gupta *et al.*, 2016). After angle surgery, trabeculectomy may be useful. However primary trabeculectomy is not a first-line procedure in congenital glaucoma in view of a higher incidence of complications and lower success rate in normalizing intraocular pressure approximately 50% fail in the initial 5–10 years (Mukkamala *et al.*, 2015).

Surgeons have advocated added trabeculectomy whenever there is incomplete canalization in cases of trabeculotomy with equivocal results. Usually amenable to surgical treatment, good results are possible if the disease is diagnosed and appropriately treated in time. Ninety-eight percent of the PCG require surgical treatment. Addition of antifibrotics further increases the success rates as proven in many Indian studies (Møller, 1977).

Netland and Walton first introduced glaucoma drainage devices GDDs to be used in paediatric glaucoma. Over the years various GDDs have been utilized for intraocular pressure control in children. Refractory paediatric glaucoma remains a challenge. Glaucoma drainage devices appear to be the most predictable and possibly safest procedure to consider after failed conventional angle surgery. Regular follow up and monitoring of IOP, corneal diameter and other parameters is required long term, Amblyopia and refractive error should be managed aggressively.

Genetics of Childhood Glaucoma

The majority of primary congenital glaucoma cases are sporadic but 10% - 40% are familial with frequent association with consanguinity. In the most familial cases, transmission is autosomal recessive with variable expression and penetrance of 40% - 100%.¹⁶ Three loci for PCG have been found GLC3A (2p21), GLC3B (1p36), and GLC3C (14q24.3). Two candidate genes: CYPB1 and LTBP2 have also been implicated. The majority of congenital glaucoma

map to GLC3A locus on chromosome 2 (2p21). Several CYP1B1 mutations in various ethnic backgrounds have been implicated in the pathogenesis. Various distinct mutations were identified in the coding region of CYP1B1 in patients of PCG-affected families, of which many mutations are novel in the Indian population. Families linked to these loci display autosomal recessive inheritance pattern. Genetic counselling can be provided to at-risk families that will aid in the prevention of PCG-related blindness.

Conclusion

The paediatric eye has its own inherent difficulties. This combined with the failure to report the symptoms, makes these children the most challenging scenarios that the ophthalmologist may encounter. Careful diagnosis and timely intervention leads to the most promising outcome and gives these children a long morbidity free life.

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Study significance: This combined with the failure to report the symptoms, makes these children the most challenging scenarios that the ophthalmologist may encounter. Careful diagnosis and timely intervention leads to the most promising outcome and gives these children a long morbidity free life.

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