



FAMILIAL BILATERAL CONGENITAL GLAUCOMA – A RARE CASE REPORT

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ABSTRACT Congenital glaucoma is a rare disease affecting children early in life. Primary congenital glaucoma was considered untreated with inevitable blindness, however recent advances in biochemical & genetic studies, the introduction of new diagnostic tools, intraocular pressure lowering medications & improvement of surgical techniques have led to a better understanding of this devastating disease & preserving the vision of affected children.

KEYWORDS :

INTRODUCTION:

Congenital glaucoma is a heterogeneous group of diseases with following classification.

Congenital - is existent or becomes evident at birth
infantile – becomes evident during early childhood (<3 yrs)
juvenile – becomes apparent in later childhood (>3 yrs)

incidence of congenital glaucoma is about 1 in 10,000 – 1 in 18,000 live births & depends heavily on ethnic groups.

Congenital glaucoma is highly prevalent in inbred populations & consanguinity is strongly associated. In 75% of cases both the eyes are affected, males are more affected than females.

90% of cases occur sporadically & only 10% of cases have an increased frequency in their family, mostly with an autosomal recessive pattern of inheritance.

CASE REPORT:

A 29 yrs old lady, 2nd gravid, para 1 live 1, with previous LSCS, married for 6 yrs, second degree consanguinity, booked & immunised patient on regular antenatal visits, with a history of she herself a twin baby with congenital glaucoma has undergone surgical correction at 40 days of life, (medical reports – showed peripheral iridotomy, with squint, a known case of gestational diabetes on oral hypoglycemic agents (sugars under control), came to us in early labour at 38 wks of gestation, emergency LSCS was done in view of previous LSCS in early in labour, an alive term male baby of 2.5 kg, on 25/9/19 at 4:02 am, APGAR of 8/10 & 9/10 with bilateral corneal clouding, ophthalmologist opinion obtained on examination baby had bilateral corneal opacities, with increased corneal diameter. baby was started on a topical beta blocker & antibiotic, referred to glaucoma specialist with experience in congenital glaucoma, trabeculotomy was done in right eye on day 5 of life, followed by in left eye after 2 weeks. now the baby is on regular follow up with paediatrician & ophthalmologist.



Post surgery image

DISCUSSION:

Congenital glaucoma is caused by impaired aqueous outflow due to abnormalities in development of anterior chamber, there are several pathogenesis:

Anomalous trabecular meshwork development. (neural crestopathy)
Barkan membrane covering the trabecular mesh work.

Anterior insertion of the ciliary body (due to arrest in the normal migration uvea) contributes to narrowing / collapsing of schlemm

canal.

Absent schlemm canal.

Resistance to the flow of aqueous causes rise in intraocular, that induces a decline in viable retinal ganglion cells, just as in adult glaucoma which leads to damage of visual field & eventually a decline of visual acuity. visual loss results from descemet breaks, corneal edema, optic neuropathy and eventually buphthalmos & amblyopia.

Classical triad of symptoms are:

Epiphora
Photophobia
Blepharospasm & corneal opacities.

Signs include enlarged/ asymmetric optic cupping, IOP > 10-12 mmHg, iris heterochromia, ectopia lentis, aniridia. The treatment of congenital glaucoma needs a team approach to normalise IOP, correct existing errors of refraction & anterior segment architecture & to enable physiologic maturation of visual pathway. the preferred mode of treatment for congenital glaucoma is surgical.

Goniotomy & trabeculotomy collectively known as “ANGLE SURGERY” is the preferred treatment with 90% success rate. Goniotomy – when cornea is clear & trabeculotomy – when cornea is cloudy. Early & accurate diagnosis of congenital glaucoma is vital, so that appropriate management can be initiated before irreversible damage to the optic nerve takes place.

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