

A CASE REPORT ON CONGENITAL GLAUCOMA**Dr. Seema¹, Dr. Ramesh Kaundal^{2*} and Dr. Deepti Parmar³**¹M.B.B.S. M.S.Ophthalmology, Medical Officer DDUZH Shimla.²M.B.B.S. M.S.Surgery, Medical Officer C.H. Theog.³M.B.B.S. M.S.Ophthalmology, Senior Resident, IGMC Shimla.***Corresponding Author: Dr. Ramesh Kaundal**

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INTRODUCTION

PCG is a rare eye disorder which accounts for 0.01–0.04% of total blindness. The disease is usually manifested at birth or early childhood (before 3 years of age). The incidence of PCG is different in different populations.^[1] The majority of patients (about 60%) are diagnosed by the age of 6 months, and 80% are diagnosed within the first year of life. A slight predominance of males is common (about 65%), and involvement is usually bilateral (about 70%). Most cases of PCG occur sporadically. Patients with a familial pattern usually show a recessive pattern with incomplete or variable penetrance and possibly multifactorial inheritance. An autosomal dominant and pseudo-dominant mode of inheritance has also been reported.^[2,3] PCG refers to a specific form of developmental glaucoma characterized by an isolated maldevelopment of the trabecular meshwork (isolated trabeculodysgenesis) not associated with other developmental ocular anomalies or ocular disease that can raise the IOP. Also called primary infantile glaucoma, it is the most common form of developmental glaucoma. Most western textbooks describe a classic triad of symptoms comprising epiphora, photophobia and blepharospasm (attributable to IOP-induced corneal epithelial edema). However, one study conducted in a tertiary institution shows that large eyeball size and hazy eyes (from corneal edema) may be the more common presenting features in the Indian subcontinent.^[4] Medical therapy usually provides a supportive role to reduce the IOP temporarily, to clear the cornea, and to facilitate surgical intervention. Laser therapy has a limited role in developmental glaucomas. The most effective and definitive form of treatment of most developmental glaucomas is surgical. Primary surgical treatment is usually with goniotomy or trabeculotomy, although combined trabeculotomy with trabeculectomy may be useful in certain populations with a high risk of failure of goniotomy or trabeculotomy. Refractory pediatric glaucomas may be managed by trabeculectomy with anti-fibrosis drugs, glaucoma drainage implants and cyclodestructive procedures.^[5]

A 4.5 year-old male child was brought by her mother for routine eye evaluation. Increased size of eyeball was observed which were perceived by mother as beautiful big eyes. The kid was not seen by an ophthalmologist until PRESENT age.

No history of eye surgery or eye trauma could be elicited. Medical History was noncontributory. Family and Social History was nonsignificant Visual Acuity was documented as OD-- Fix/follow; OS--Fix/follow. Intra-ocular pressure (NCT) was recorded as 14mmHg, OD; and 18 mmHg, OS. External and anterior segment examination revealed OD, Normal; OS: normal.

Since the child was fairly cooperative all the necessary investigations could be carried out without sedation or GA. Gonioscopy revealed high iris insetion OS, high iris insertion OD. Dilated fundus exam (DFE) revealed OD—increased cupping; OS--Increased cupping. Horizontal corneal diameter was OD, 14 mm; OS, 14.5

mm. Axial eye length (echography) was calculated as OD, 24.3 mm; OS 24.5 mm.

Increased intraocular pressure, increased horizontal corneal diameter, and increased axial eye length point helped us reach a diagnosis of buphthalmos due to primary congenital glaucoma. The history of approximately 2 years of increased eye size OS would place the onset of disease around 1 year of age; typical onset of primary congenital glaucoma is between 3-6 months of age. The absence of any angle recession, posterior embryotoxon/ iris strands/iris hypoplasia/corectopia, Peter's anomaly, or aniridia help solidify this diagnosis of congenital glaucoma presenting only as buphthalmos.

MANAGEMENT OF RESIDUAL VISION IN PEDIATRIC GLAUCOMA

Unfortunately, even with the best treatment in the best centers, many children with congenital glaucomas end up with "low vision". Visual rehabilitation and low vision

aids can help these children lead a normal or near-normal life. After proper assessment, telescopes (hand-held or spectacle-mounted) may be prescribed to improve distant vision while hand or pocket magnifiers (2× to 3×) may be prescribed to improve near vision. Structured training programs in the use of these devices should be planned and discussed with the child and parents.

CONCLUSION

Once considered virtually untreatable, pediatric glaucomas have now become reasonably manageable in most cases, thanks to scientific advances. Let us pray that ongoing research in this field will assist to further help the preservation or restoration of vision of children suffering from glaucomas.

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