

## A STUDY ON DEVELOPMENTAL GLAUCOMA

Meenakshi Babu<sup>1</sup>, Vimala Thangaraj<sup>2</sup>, Sharmila Devi Vadivelu<sup>3</sup>, Sudarvizhi Arumugam<sup>4</sup>, Manupriya Murali<sup>5</sup>

<sup>1</sup>Assistant Professor, Department of Ophthalmology, Regional Institute of Ophthalmology, Government Ophthalmic Hospital, Madras Medical College, Chennai.

<sup>2</sup>Assistant Professor, Department of Ophthalmology, Regional Institute of Ophthalmology, Government Ophthalmic Hospital, Madras Medical College, Chennai.

<sup>3</sup>Assistant Professor, Department of Ophthalmology, Regional Institute of Ophthalmology, Government Ophthalmic Hospital, Madras Medical College, Chennai.

<sup>4</sup>Postgraduate, Department of Ophthalmology, Regional Institute of Ophthalmology, Government Ophthalmic Hospital, Madras Medical College, Chennai.

<sup>5</sup>Postgraduate, Department of Ophthalmology, Regional Institute of Ophthalmology, Government Ophthalmic Hospital, Madras Medical College, Chennai.

### ABSTRACT

#### BACKGROUND

Developmental glaucoma includes congenital glaucoma and glaucoma associated with other developmental anomalies, either systemic or ocular. In all forms it occurs in about 1 in 10,000 live births. The aim of the study is to analyse aetiology of developmental glaucoma and its mode of treatment and outcome.

#### MATERIALS AND METHODS

This is a retrospective study conducted in Regional Institute of Ophthalmology, Government Ophthalmic Hospital, Chennai during the period of June 2005 to September 2007. 66 eyes of 37 patients were included in the study

#### RESULTS

Most common age group affected was less than 3 years with predilection for female children. Most common systemic anomaly was Sturge Weber syndrome seen in 4 cases, followed by congenital heart disease which accounted for 3 cases. Most common complication associated with developmental glaucoma in this study was corneal decompensation which occurred in 17 eyes followed by cataract in 6 eyes. Myopia was the most common refractive error. The success rate was 81%, 67% and 67% at the end of 3 months, 6 months and 1 year respectively for trabeculectomy. The success rate for trabeculotomy with trabeculectomy was 88%.

#### CONCLUSION

Developmental glaucoma is a global problem. A proper diagnostic evaluation under general anaesthesia is required. Surgery remains the principle therapeutic modality in the management of developmental glaucoma. It is important that appropriate surgery should be chosen and performed with technical perfection in a tertiary care centre where there is adequate facility to ensure safe anaesthesia and skilful surgery with a lifetime follow up.

#### KEYWORDS

Developmental Glaucoma, Trabeculectomy, Trabeculotomy with Trabeculectomy, Trabeculotomy with Trabeculectomy with Mitomycin C.

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#### BACKGROUND

Developmental glaucoma refers to glaucoma associated with developmental anomalies of eye present at birth. It includes congenital glaucoma and glaucoma associated with other developmental anomalies, either systemic or ocular. In all forms it occurs in about 1 in 10,000 live births. Isolated

congenital glaucoma refers to a special form of glaucoma. These eyes have an isolated maldevelopment of trabecular meshwork not associated with other developmental ocular anomalies or ocular disease that can raise intra ocular pressure. Isolated congenital glaucoma is the most common glaucoma of infancy, occurring in 1 in 30,000 live births.<sup>1</sup>

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Corresponding Author:

Dr. Vimala Thangaraj,

Assistant Professor, Department of Ophthalmology,

Regional Institute of Ophthalmology,

Government Ophthalmic Hospital,

Madras Medical College, Chennai.

E-mail: dr\_vimala\_t@yahoo.com

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#### Classification of Developmental Glaucoma

##### • Hoskins Anatomic Classification<sup>2</sup>

1. Isolated trabeculodysgenesis-maldevelopment of trabecular meshwork.
2. Flat iris insertion
3. Anterior insertion
4. Posterior insertion
5. Mixed insertion
6. Concave iris insertion



7. Iridotrabeculodysgenesis
8. Anterior stromal defects-hyperplasia/hypoplasia
9. Anomalous iris vessels –persistent tunica vasculosa, anomalous superior vessels,
10. Structural anomalies –holes, coloboma, aniridia
11. Corneotrabeculodysgenesis
12. Peripheral
13. Mid peripheral
14. Central
15. Microcornea/megalo cornea

## MATERIALS AND METHODS

This is a retrospective study conducted in Regional Institute of Ophthalmology, Government Ophthalmic Hospital, Chennai during the period of June 2005 to September 2007. 66 eyes of 37 patients were included in the study. Children who were not cooperative for office examination were examined under general anaesthesia. Intraocular pressure measurement was done by handheld Perkins tonometer. Parameters that were recorded included age and sex of the patient, eye involved and its association with any systemic and/or ocular anomalies, visual acuity if possible, IOP measured by handheld Perkins tonometer, corneal diameter measured by vernier calipers, fundus examination by direct ophthalmoscopy, A-Scan and B Scan biometry for axial length and posterior segment evaluation.

After diagnosing developmental glaucoma, patients who were high risk for surgery under general anaesthesia were started with medical management.

Surgery was done once patients obtained GA fitness. All the surgeries were done by a single experienced surgeon. 25 eyes were started with medical therapy. 40 eyes underwent surgery, of which 21 eyes underwent Trabeculectomy, 15 had trabeculotomy with trabeculectomy, 1 eye had trabeculectomy with derroofing of Schlemm's canal, and 1 eye associated with Weil Marchesani syndrome underwent extra capsular cataract extraction (ECCE) with Anterior vitrectomy. 2 painful blind eyes were enucleated. Post operatively corneal clarity was assessed. Repeat intra ocular measurements were done on follow up. Patients with failure of surgery underwent repeat surgery. Repeat surgeries included trabeculotomy with trabeculectomy, trabeculotomy with trabeculectomy with Mitomycin C. Patients with failure of second surgery underwent third surgery. Patients were followed up and refractive errors were corrected, and spectacles were prescribed if possible. Cases with amblyopia were started on occlusion therapy.

## Inclusion Criteria

Developmental glaucoma in the paediatric age group (from birth to 12 years of age)

## Exclusion Criteria

Patients with traumatic glaucoma, post inflammatory glaucoma, those with other causes of epiphora, corneal haziness without raise in intra ocular pressure and other

causes of megalophthalmos without rise in intraocular pressure were excluded from the study.

## RESULTS

### Age Distribution

Age	<1 yr	13-36 months	37-60 months	5-7 yrs
Number	14	12	7	4
Percentage	38	32	19	11

**Table 1. Age Distribution**

The most common age of presentation in our study was below 1 year of age followed by 1-3 years of age.

### Sex Distribution

Sex	Male	Female
Number	13	24
Percentage	35	65

**Table 2. Sex Distribution**

In our study female children were more commonly affected which constituted about 65% of cases.

### Laterality

Laterality	Both	RE	LE
Number	29	6	2
Percentage	78	16	6

**Table 3. Laterality**

Most of the cases had bilateral involvement. Cases which had unilateral involvement were mostly due to phakomatoses.

### Associated Systemic Anomalies

Systemic involvement	Number
Sturge Weber	4
Mucopolysaccharidoses	2
Congenital heart disease	3
Microcephaly with seizure disorder	2
Oculomelanocytosis of OTA	1
Cleft lip and cleft palate	1
Down's syndrome	1

**Table 4. Associated Systemic Anomalies**

14 cases in this study had associated systemic anomalies. Of these, 4 cases had Sturge Weber syndrome.

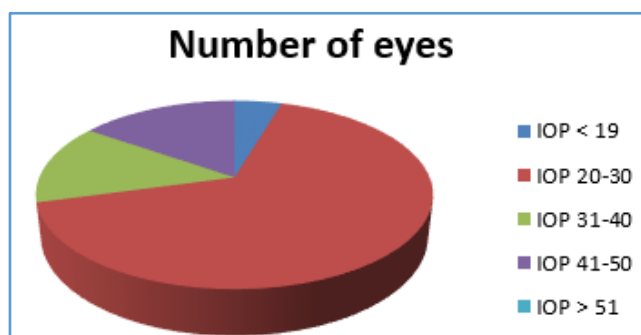
### Associated Ocular Anomalies

2 cases had associated ocular anomalies of which 1 had Weill-Marchesani and the other had Rieger's anomaly.

### Intraocular Pressure

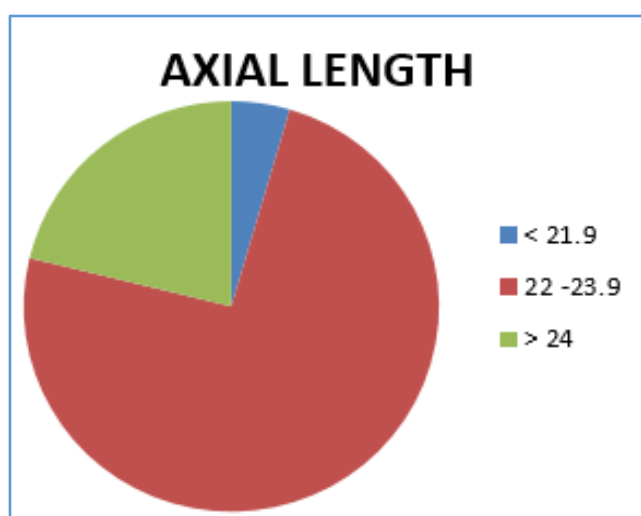
Of the 66 eyes that were studied 43 eyes had IOP recording between 20 and 30 mm of Hg. Only 1 eye had IOP more than 51 mm of Hg. IOP below 20 mm of Hg was recorded in cases with Retinal Detachment (RD). One case with RD had

associated choroidal detachment and had gone for phthisis bulbi. The second case of RD had corneal diameter of 16 mm and an axial length of 30 mm.

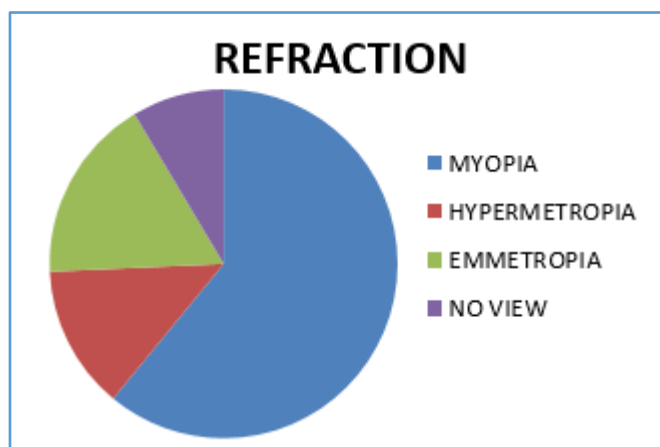


### Axial length

Majority of cases had axial length of 22 – 24 mm. 14 cases had axial length of more than 24 of which the highest recorded was 30 mm and this case had an associated Retinal detachment.



### Refraction



Most common refractive error was myopia which was present in 32 eyes, followed by emmetropia in 9 eyes. Hypermetropia occurred in 7 eyes. In this study myopia was

the most common refractive error presenting in approximately half of the eyes.

### Complications

Complications of developmental glaucoma that were encountered in this study were as follows.

Complications	Number of Eyes
Corneal decompensation	17
Cataract	4
Retinal detachment	1
RD with choroidal detachment	1
Phthisis	2

**Table 5. Showing Complications of Developmental Glaucoma**

### Medical Treatment

Of the 37 patients, 13 patients (25 eyes) received betaxolol eye drops one drop twice daily. Among the 13 patients, 2 patients were not willing for surgery, 1 patient had cleft lip with cleft palate, 2 patients had seizure disorder, 3 patients had congenital heart disease, 4 patients were preterm with low birth weight. 1 eye had become phthisical.

### Surgical Treatment

40 eyes had surgical treatment. Of these trabeculectomy was done as primary procedure in 21 eyes. Of the 21 eyes, 13 eyes had successful control of intra ocular pressure and 8 failed. Of the 8 eyes, 4 eyes underwent trabeculectomy with trabeculectomy with Mitomycin C, 2 cases were taken up for trabeculectomy with trabeculectomy, 1 case underwent lensectomy with cyclodialysis. 1 eye became phthisical. Of the 4 eyes which underwent Trabeculectomy with Trabeculectomy with Mitomycin C, 3 were successful and 1 case which had oculomelanocytosis of OTA underwent a third surgical procedure - Thermosclerostomy. Trabeculectomy with trabeculectomy was done as primary procedure in 15 eyes. All cases which underwent Trabeculectomy with trabeculectomy as the first procedure were successful until the follow up period. A case of Weil Marchesani syndrome underwent Extra Capsular Cataract extraction with anterior vitrectomy as the primary procedure and this procedure was successful in this case. 2 cases with painful blind eye were enucleated. Post-operative clearing of cornea and reduction of intra ocular pressure were taken as parameters to decide whether the procedure was successful.

### Primary Procedure

Type of surgery Done	No. of Eyes	Success	Failure
Trabeculectomy	21	13	8
Trabeculectomy with Trabeculectomy	15	15	
Trabeculectomy with Deroofing of Schlemm's canal	1		1
ECCE with Anterior Vitrectomy	1	1	
Enucleation	2		

**Table 6. Type of Surgery Done and Success Rate**

**Repeat surgery was done in failed cases**

Type of Surgery	No. of Eyes	Success	Failure
Trabeculotomy with trabeculectomy with Mitomycin C	4	3	1
Trabeculotomy with Trabeculectomy	2		2
Lensectomy with Cyclodialysis	1	1	
No Intervention (phthisis bulbi)	1		

**Table 7. Second Surgery**

Type of Surgery	No. of Eyes	Success	Failure
Thermosclerostomy	1	1	–
Trabeculotomy with trabeculectomy with mitomycin C	2	2	–

**Table 8. A Third Surgery was Done for Three Eyes**

**Visual Outcome**

- 17 eyes of 11 patients had visual acuity greater than 6/60, of which 5 eyes had visual acuity greater or equal to 6/12 with correction.
- 5 eyes of 4 children had vision of No PL and 2 eyes had been enucleated.
- 12 infants (22 eyes) fixed and followed light. Vision recording was not possible in 3 infants (6 eyes) as they were less than 1 month of age.
- 14 eyes of 7 patients had vision of less than 6/60 of which 3 children had only perception of light.

**Follow Up**

Cases were followed up for 3 months, 6 months, 1 year depending on the time of surgery.

**Trabeculectomy**

At the end of 1 year, out of 21 eyes which underwent trabeculectomy 13 eyes had good control of intra ocular pressure. 8 eyes which underwent Trabeculectomy had failed. Of the 8 eyes, 7 were taken up for repeat surgery and 1 eye became phthisical.

**Trabeculotomy with Trabeculectomy**

At the end of 1 year of follow up 15 eyes out of 17 eyes which underwent trabeculotomy with trabeculectomy had successful control of intra ocular pressure. 2 eyes had failed.

**Trabeculotomy with Trabeculectomy with Mitomycin C-** Out of 4 eyes followed up for 1 year 3 had good control of IOP. 1 eye had failed at end of 1 year.

The success rate was 81%, 67% and 67% at the end of 3 months, 6 months and 1 year respectively for trabeculectomy. The success rate for trabeculotomy with trabeculectomy at the end of 1 year was 88%. In this study all 4 cases of Sturge Weber syndrome which underwent Trabeculotomy with Trabeculectomy were successful and were managed without additional medical therapy. All 4 cases had good visual acuity.

Post op IOP mm Hg	No. of Patients	Percentage
Upto 20	31	82
21-25	7	18

**Table 9. Postop Intraocular Pressure**

Of the 40 eyes operated, 31 eyes had IOP less than 20 mm of Hg and 7 eyes had IOP between 21 and 25 mm of Hg.

**DISCUSSION**

66 eyes of 37 patients were included in the study. Most common age group affected was less than 3 years which accounted for 70% of cases. Female children were more commonly affected than male children in this study. 78% of cases had bilateral involvement.<sup>3</sup> In another study it was found that the overall frequency of bilateral disease was 92.3%. Most common systemic anomaly was Sturge Weber syndrome seen in 4 cases, followed by congenital heart disease which accounted for 3 cases. Among the 66 eyes, 43 eyes had preoperative IOP recording between 20 and 30 mm of Hg. Only 1 eye had IOP more than 51 mm of Hg. IOP below 20 mm of Hg was recorded in cases with Retinal Detachment (RD).<sup>4</sup> It is comparable with another study in which the mean preoperative IOP was 26.9±5.2 mmHg (range, 14–42 mmHg). Majority of our cases had axial length of 22–24 mm.<sup>5</sup> In another study it was found that axial lengths of children under 2 years were 21.31\_+0.97 mm. Most common complication associated with developmental glaucoma in this study was corneal decompensation in 17 eyes followed by cataract in 6 eyes, Retinal Detachment in 2 eyes and phthisis bulbi in 2 eyes. Myopia was the most common refractive error. 45% of cases had myopia.<sup>6</sup> In another study among 41 patients, 33 (80.5 %) were myopic. The success rate was 81%, 67% and 67% at the end of 3 months, 6 months and 1 year respectively for trabeculectomy. The success rate for trabeculotomy with trabeculectomy at the end of 1 year was 88%.<sup>7</sup> It is comparable with another study in which the cumulative chance of success of the primary trabeculectomy was 72% and the success of trabeculotomy with trabeculectomy was 93.5%.

**Postoperative IOP**

Of the 40 eyes operated, 31 eyes had IOP less than 20 mm of Hg and 7 eyes had IOP between 21 and 25 mm of Hg.<sup>8</sup> It is comparable with another study in which the mean postoperative IOP was 13.1 ± 5.8 mmHg; 13.7 ± 4.4 mmHg; and 13.3 ± 6.0 mmHg in the congenital, infantile, and juvenile types of developmental glaucomas, respectively.

**CONCLUSION**

Developmental glaucoma is a global problem. A proper diagnostic evaluation under general anaesthesia is required. Surgery remains the mainstay of management of developmental glaucoma. Trabeculectomy can be done as a primary procedure in the management of primary developmental glaucoma, while combined Trabeculotomy with Trabeculectomy offers the best hope of success.

Trabeculotomy with trabeculectomy with Mitomycin C proves successful in cases with failure of primary procedure. It is important that the appropriate surgery should be performed with technical perfection in a tertiary care centre where there is adequate facility to ensure safe anaesthesia and a lifetime follow up.

Visual rehabilitation is as important in the management of developmental glaucoma as is the control of IOP. Visual rehabilitation involves correction of refractive errors, opacities in the media such as corneal scarring and cataract, and amblyopia management.

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