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Original Article

Surgical Management of Congenital Glaucoma - A Long-term Clinical Study and its Outcome

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Abstract

Introduction: Congenital glaucoma (CG) is more common, aggressive, and difficult to control. It results in raised intraocular pressure and diminished visual acuity in children. Many surgical procedures are described in the literature in its surgical management. Trabeculectomy in chronic simple, narrow-angle, aphakic, and secondary glaucoma is good. Its response in CG is scanty ion literature. The present study evaluates results of different procedures adopted in surgical treatment of CG.

Aim: The aim of this study is to evaluate the long-term results of surgery for CG.

Study Design: This is a retrospective comparative study.

Materials and Methods: Children aged below 14 years with CG surgically treated by methods such as goniotomy, trabeculotomy, trabeculectomy, combined trabeculotomy - trabeculectomy, cyclocryotherapy and Nd-Yag cyclophotocoagulation from January 2010 to December 2016.

Observations and Results: A total of 36.42 (85.71%) of the children were diagnosed as CG at birth. A total of 66 eyes (89.18%) were diagnosed as primary infantile glaucoma, 6 eyes (8.10%) had developmental glaucoma, and 2 eyes (2.70%) had secondary glaucoma. The mean corneal diameter 11.8±0.35 (range 9-18) mm and mean cupping 0.8 (range 0.3-1.1). The mean intraocular pressure was reduced from 27.8 ± 3.45 to 15.66 ± 1.50 mmHg. Trabeculectomy proved to be the most useful in achieving reduction in the mean intraocular pressure to 14 ± 1.0 mmHg.

Conclusions: The safe surgical procedure in treating CG was primary trabeculectomy. The success rate of all surgical procedures decreased on long-term follow-up. The visual outcome was satisfactory in 78.37% of the total eyes operated with a follow-up of 5 years.

Key words: Congenital and primary glaucoma, Glaucoma, Goniotomy, Trabeculectomy, Trabeculotomy

INTRODUCTION

Congenital glaucoma (CG) refers to glaucoma associated with developmental anomalies in the eye since birth. They may be associated with anomalies of the anterior chamber angle and trabecular meshwork alone or associated with other ocular or systemic/developmental anomalies. Pediatric glaucoma is a potentially blinding disease accounting for about 4.2% of childhood blindness.[1] Hoskin[2] based on anatomy classified developmental glaucoma into (1) isolated trabeculodysgenesis: A - flat iris insertion: Anterior insertion, posterior insertion mixed insertion, B - concave iris configuration, (2) iridotrabeculodysgenesis: A - anterior stromal defects, hypoplasia, and hyperplasia, B - anomalous iris vessels: Persistence of tunica vasculosus lentis, anomalous superficial vessels, C - structural anomalies: Holes, colobomata, aniridia, and (3) corneotrabeculodysgenesis, A - peripheral (axenfeld anomaly), B - mid peripheral (Reiger's anomaly), and C - central (Peter's anomaly). Pediatric glaucoma is a potentially blinding disease accounting for about 4.2% of childhood blindness.[3] In India, according to a population-based study in South India, the prevalence of Primary CG (PCG) is 1 in 3,300.[1] The prevalence has varied from 1 in 10,000-20,000 in the West[4] to 1 in 1250 in the gypsy population of Slovakia.[4] Males constitute 65% of the cases. The most cases are sporadic. About 10% show a hereditary pattern, usually autosomal

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recessive with variable inheritance.[5,6] Trabeculectomy is one of the most commonly performed operations for glaucoma. Fairly good results were obtained with trabeculectomy in chronic simple glaucoma,[7-9] narrow angle glaucoma,[10] aphakic glaucoma,[8] and secondary glaucoma.[10] Aim of the present communication is to report the follow-up of trabeculectomy in 50 cases of CG. In this context, the present study was conducted to evaluate the long-term results of surgery for CG in terms of reduced intraocular pressure (IOP) control and residual visual acuity in children under 14 years who underwent different surgical procedures for primary and secondary CG.

MATERIALS AND METHODS

The present study was a prospective study conducted in a tertiary teaching Hospital in Northern Kerala attached to Kannur Medical College, Kannur. A total of 42 children attending the OPD of Ophthalmology with diagnosis of CG were included in the study. An Institutional Ethical Committee clearance was obtained and committee approved consent letter was used for the inclusion of children in the study. The study period was between January 2010 and December 2016 (6 years). A thorough clinical history including family history of childhood glaucoma and consanguinity was elicited. Demographic data were collected using standard collection forms; pre- and post-operative ocular examination status, IOP, visual acuity, type of ocular surgery, age at the diagnosis of the disease and operation, intra- and post-operative notes, and complications were recorded. The clinical diagnosis was established based on measurements of IOP and by the presence of accompanying signs and symptoms of sustained, elevated IOP such as buphthalmos and corneal enlargement and edema as well as tearing and photophobia. All children were examined under chloral hydrate (50-100 mg/kg) sedation in the OPD. Ocular assessment includes microscopic evaluation of the anterior segment of the eye with slit lamp. IOP measurement was done under topical anesthesia by using handheld tonometer, Goldmann applanation tonometer, and pneumotonometer. The horizontal corneal diameter assessed by a ruler or caliper. Gonioscopy and fundoscopy performed by Koeppe lens if the cornea was clear. The selected eyes were operated using the following surgical procedures to lower IOP. The different procedures undertaken were goniotomy, trabeculotomy, trabeculectomy, combined trabeculotomy - trabeculectomy (CTT), cyclocryotherapy (CCT), and contact Nd-Yag cyclophotocoagulation (CYC). All the procedures were performed under general anesthesia, using standard techniques that are described in text books. The minimum follow-up required to be included in the study was 6 months from the date of glaucoma surgery. Post-operative drug therapy included topical dexamethasone, neomycin, and polymyxin B; 3 h and ointment at bedtime. Antiglaucoma drugs systemically used wherever necessary. During the stay at hospital, all children were examined on days 1-3 after surgery. After discharge, the follow-up was at weeks 1, 4, and 8 and then every 3 months thereafter for 5 years. At every examination, the anterior chamber depth, corneal appearance, bleb appearance, IOP, fundus, and any complications were looked for and recorded. All the data were analyzed using standard statistical methods like mean ± standard deviation, and the student's *t*-test was used. Because of the variability in length of follow-up among patients, life-table (survival) analysis (Kaplan-Meier method) was used to estimate the success rate at various post-operative intervals.

OBSERVATIONS AND RESULTS

Totally, 74 eyes of 42 children were operated on including 29 male and 13 females. The mean age at the diagnosis was 10.25 ± 2.20. Nearly 64% of the children were diagnosed as CG at birth. There were 29 male and 13 female children with M:F ratio of 2.23:1. The mean corneal diameter 11.8 (range 9-18) mm and mean cupping 0.8 (range 0.3-1.1). The mean cup to disc ratio was 0.6 among the 37 eyes. The mean intraocular pressure reduction was from 27.8 ± 2.10 to 15.45 ± 0.90 mmHg in 45 eyes (60.81%), following a mean number of operation/eye of 2.2, and a mean follow-up of 74.6 months. The mean age of the patients at surgery was 38.6 ± 3.40 months (Table 1). Among the 42 children, 30 children had bilateral CG (71.42%) and the remaining 12 (28.57%) were presenting with unilateral disease (right eye - 08 and left eye - 04), (Table 1). Family history of CG was positive in 11.42 (26.19%), (Table 1). A total of 27.42 children showed visual acuity <6.60 (64.28%) and above 6.60 visual acuity was in 15.42 (35.71%) children (Table 1).

Among the 74 eyes, 66 eyes (89.18%) were diagnosed as primary infantile glaucoma, 6 eyes (8.10%) had developmental glaucoma, and 2 eyes (2.70%) had secondary glaucoma (Table 2).

Among the 6 eyes of 3 patients who had goniotomy, 3 eyes failed within 3 months, repeat goniotomy had to be done in 2 eyes. The mean post-operative follow-up in this group was 4.60 ± 0.85 years. A total of 2 eyes had good visual acuity at the end of 5 years follow-up. Totally, 3 eyes developed scar in the cornea which affected the visualization of the angle and subsequently the result of surgery. The mean reduction of IOP was 17 ± 1.25 mmHg. The success rate was 2/74 (2.70%), (Table 3). In 5 eyes in this study trabeculotomy was done. Scar developed in 2/05 eyes. Corneal haze decreased in 3/05 eyes. The success rate in

terms of gain in visual acuity was 3/74 (4.05%), (Table 3). A total of 37 eyes in the study underwent trabeculectomy. The mean reduction in IOP was 14 ± 1.0 mmHg. The success rate was 31/74 (41.89%) in terms of gain in visual acuity. Scar development was in 4/74 eyes (5.40%) and decrease in corneal haze was observed in 33/74 (44.59%) eyes (Table 3). CTT was done in 16 eyes. Mean IOP was found reduced to 15 ± 1.45 mmHg. Visual acuity was good in 9/74 (12.16%), average in 4/16 (5.40%) of the eyes operated. Scar was found in 2/16 and decreased corneal haze in 12/16 eyes (75%), (Table 3). A CCT procedure was under taken in 05 eyes. IOP was reduced to 16 ± 1.0 mmHg at the end of follow-up for 5 years and the success rate was 4/05 (80%) and there was no corneal haze or scar formation (Table 3). Nd-Yag cyclophotocoagulation was done in 5 eyes. The success rate was 100% and there were eyes with scar and decreased corneal haze in all the eyes (Table 3).

At the end of the study and follow-up of 5 years, the overall end results in terms of lowered IOP, improved visual acuity and reduced corneal haziness, optic disc diameter, and incidence of scar were analyzed in all the 74 eyes operated. The mean age at the diagnosis was 10.25 ± 2.20. 36/42 (85.71%) of the children were diagnosed as CG at birth. A total of 66 eyes (89.18%) were diagnosed as primary infantile glaucoma, 6 eyes (8.10%) had developmental glaucoma, and 2 eyes (2.70%) had secondary glaucoma. The mean corneal diameter 11.8 ± 0.35 (range 9-18) mm, and mean cupping 0.8 (range 0.3-1.1). The mean intraocular pressure reduction was from 27.8 ± 3.45 to 15.66 ± 1.50 mmHg; following a mean number of operation/eye of 2.2, and a mean follow-up of 94.6 ± 4.20 months. The mean age of the patients at surgery was 38.6 ± 3.40 months. Trabeculectomy proved to be the most useful in achieving reduction in the intraocular pressure to 14 ± 1.0 mmHg. There were no severe complications recorded in the study. Good recovery of visual acuity was found in 42/74 eyes (56.75%) operated and average recovery in 16/74 eyes (21.62%) and poor recovery in 16/42 eyes (21.62%), (Table 3). During the post-operative period hyphema was noted in 16/74 eyes and total resolved in 10 days. Eccentric pull of pupil was noted in 5/74 eyes operated. Choroidal and exudative retinal detachment occurred in 1 child following trabeculectomy. The detachment resolved in 2 weeks.

Table 1: The demographic data in the study (*n*-42)

Observation	Result
Mean age at diagnosis in months	10.25±2.20
Mean age at time of surgery in months	34.4±3.40
Gender	
Male	29
Female	13
Number of affected eyes	
Right	30+08
Left	30+04
Consanguinity	11.42 (26.19%)
Mean pre-operative IOP (mmHg)	27.8±2.10
Mean corneal diameter (mm)	11.8±0.35
Mean cup to disc ratio	0.8 (37 eyes)
Clear cornea	16 eyes
Corneal edema	22 eyes
Corneal scar	04 eyes
Mean number of pre-op medications	2.6
Visual acuity	
Poor visual acuity (NLP=No light perception, LP=Light perception, HM=Hand motion CF=Count fingers, CSM=Central, steady, maintain)	27
Average (6/60, 6/36, 6/24)	10
Good (6/12, 6/9 and 6/6	05

Table 2: The final clinical diagnosis (*n*-42)

Type of glaucoma	*n* (%)
Primary infantile glaucoma	66 (89.18)
Developmental glaucoma	06 (08.10)
Secondary glaucoma	02 (02.70)

DISCUSSION

CG remains a responsible cause for blindness in children accounting for 4-18% of cases.[11,12] In Indian literature few studies showed incidence of Congenital Glaucoma accounted for 4.2-7% of all causes causing blindness in children.[12,13] CG cases are usually sporadic but 10-40%

Table 3: The mean values of parameters after follow-up of 5 years (*n*-74)

Type of surgery- number of eyes - 74	IOP mmHg	Visual acuity			Optic cup	Corneal diameter	Scar Present	Corneal haze	
		Poor- 16	Average- 16	Good- 42				↑	↓
Goniotomy-06	17±1.25	04	01	01	0.8±0.06	12.8±0.35	3	03	03
Trabeculotomy - 05	16±1.60	02	01	02	0.9±0.05	11.90±0.5	2	02	03
Trabeculectomy - 37	14±1.0	06	08	23	0.8±0.07	10.70±0.4	4	04	33
CTT 16	15±1.45	03	04	09	0.9±0.04	11.35±0.5	2	04	12
CCT - 05	16±1.0	01	01	03	0.9±0.05	11.80±0.4	-	00	05
Nd-Yag-cyclo-photocoagulation -05	16±1.2	00	01	04	0.9±0.04	12.30±0.3	-	00	05

Poor visual acuity, NLP: No light perception, LP: Light perception, HM: Hand motion, CF: Count fingers, CSM: Central, steady, maintain, Average visual acuity: Vision 6/60, 6/36, 6/24, Good visual acuity: 6/12, 6/9 and 6/6

are familial with frequent association with consanguinity. In majority of familial cases the transmission is autosomal recessive with variable expression and penetrance of 40-100%.[14] Al-Hazmi *et al*.[5] reported family history in (21%) of his patients and (61%) were the products of consanguineous marriages in Saudi's population. In the present study, the consanguinity was observed in 11/42 (26.19%). Among all the surgical methods described in the surgical management of CG, trabeculectomy was found to give 48% absolute success and another partial success in 28% of their patients by Alex Joseph.[15] However, this method was found to be less effective in CG.[9] According to Alex[15], the trabeculectomy has the advantage over the other drainage operations by drainage of aqueous into the Schlemm's canal beside, subconjunctival drainage which does not produce a large conjunctival bleb. Trabeculectomy can be easily repeated without much disfigurement which may be essential in CG. The complications observed after trabeculectomy are limited both in number and clinical importance. PCG in the present study was 66/74 eyes (89.18%). This could be explained to little extent due to consanguinity of 11.42 children (26.19%). Trabeculectomy was undertaken in 37 eyes and 23/74 showed recovery of good visual acuity (31.08%) and average in 8/74 (10.81-21.62%); total 41.89%. This success rate is comparatively higher than other modalities adopted in 37 eyes in the present study showing 27/74 (36.48%). There is no statistical difference between the two groups in the study (*P* - 0.842, *P*- significant at <0.05, using student *t*-test). Al-Hazmi *et al*.[14] reported low success rate of goniotomy (42%) and trabeculotomy (29%) in the early cases treated at King Khalid Eye Specialist Hospital between 1982 and 1990. In the present study the corresponding results were 2.70% and 4.05% respectively. This study observed an overall success rate in terms of gain in visual acuity was 78.37% of the total eyes operated with a follow-up of 5 years. Al-Hazmi *et al*.[14] reported that success rate of trabeculectomy increase with age, 32% for patients younger than 6 months and 85% for patients older than 4 years of age. In the present study, the overall mean age at surgery was 38.6 ± 3.40 months. Mullaney *et al*.[3] reported that the success rate of CTT was 78% for PCG and 45% for developmental and secondary CG. The present study showed success rate in regards with gain in visual acuity was 17.56%. CCT showed 100% success rate among the 5 eyes operated on and overall success rate was 5/74 6.75%. A total of 27/42 children showed visual acuity less than 6.60 (64.28%); legally blind and above 6.60 visual acuity was in 15/42 (35.71%) children.

CONCLUSION

The safe surgical procedure in treating CG was primary trabeculectomy. No significant different between trabeculectomy and other modalities of surgical treatments. The success rate of all surgical procedures decreased on long term follow-up. The visual outcome was satisfactory in 78.37% of the total eyes operated with a follow-up of 5 years. The main cause of visual loss is corneal opacity.

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