Evaluation of primary congenital glaucoma at Assiut University Hospital

Omar A. Hasan, Hasan L. Fahmy, Ali N. Riyadh, Mahmoud F. Rateb

Department of Ophthalmology, Faculty of Medicine, Assiut University, Assiut, Egypt

Correspondence to Omar A. Hasan, MBChB, MSC., Assistant Lecturer of Ophthalmology at Assiut University Hospital, 71515, Assiut, Egypt.
Fax: 01000525341
Tel: 00201000525341;
e-mail: oabdelkarem@gmail.com

Received 30 June 2022
Revised 04 September 2022
Accepted 02 October 2022
Published 21 June 2023

Journal of Current Medical Research and Practice
2023, 8:106–110

Purpose
This study aims to evaluate cases of primary congenital glaucoma at Assiut University Hospital.

Patients and methods
A total of 32 eyes of 19 infants with primary congenital glaucoma were enrolled in this study. All eyes underwent combined trabeculotomy-trabeculectomy with mitomycin C under general anesthesia. Follow-up of the surgical outcome was achieved for each patient by measuring the postoperative ocular tension monthly for 3 months and then every 3 months till the end of a year.

Results
A total of 20 (62.5%) eyes showed successful intraocular pressure (IOP) lowering after the surgery, with mean IOP of 15.3 mmHg, with range between 11 and 19 mmHg measured under general anesthesia using halothane. A total of nine (28.1%) eyes required postoperative antiglaucoma medications to normalize the IOP and three (9.3%) eyes required glaucoma drainage device surgery to normalize the IOP.

Conclusion
Eyes that required antiglaucoma medications or eyes that underwent glaucoma drainage device surgery had common features between them as positive consanguinity between parents, early age of presentation, high initial IOP at time of presentation, and corneal haziness.

Keywords:
congenital, glaucoma, trabeculectomy

J Curr Med Res Pract 8:106–110

2357-0121

Introduction

Congenital glaucoma is a developmental glaucoma occurring before the age of 3 years owing to an obstruction that prevents adequate drainage of aqueous humor caused by abnormal development of the trabecular meshwork and anterior chamber angle. This arbitrary age has been estimated as it corresponds to the age at which the eye grows in response to high intraocular pressure (IOP) [1].

Children with congenital glaucoma typically present with globe enlargement (buphthalmos), edema and opacification of the cornea with rupture of Descemet's membrane (Haab's striae). Additional clinical features include thinning of the anterior sclera and iris atrophy, anomalously deep anterior chamber, and structurally normal posterior segment except for progressive glaucomatous optic atrophy [2].

Moreover, visual acuity may be reduced and/or visual fields may be restricted. In untreated or treated late cases, blindness occurs [1,2].

Aim

This study aims to evaluate cases of primary congenital glaucoma at Assiut University Hospital regarding the epidemiological pattern, clinical presentation, surgical outcome of the conventional combined trabeculotomy and trabeculectomy with mitomycin C, and the risk factors for poor prognosis.

Patients and methods

IRB number: 17100944.

Type of the study: a prospective interventional study was conducted.

Patients

A total of 32 eyes of 19 infants (10 males and nine females) (13 bilateral and six unilateral) with primary congenital glaucoma were enrolled in this study in the period between October 2019 and January 2021.

The age of these infants ranged from 1 week to 6 months.

The diagnosis was established by presence of the following characteristic signs of primary congenital

glaucoma such as corneal haziness, increased corneal diameter (megalocornea), and buphthalmos.

Exclusion criteria

Infants with history of previous surgery, secondary glaucoma, and glaucoma associated with other ocular congenital anomalies were excluded.

Methods

Examination under sedation

Examination under sedation using halothane revealed corneal diameter more than 13 mm, increased ocular tension more than 25 mmHg using a Schiotz tonometer, and glaucomatous optic nerve head cupping.

Investigations

Ultrasound A scan was used to measure the axial length of the eye, which was typically more than 22 mm, and B scan was used to detect the presence of pathological excavation of the optic disc and to exclude presence of intraocular tumors.

Surgical procedure

Combined trabeculotomy-trabeculectomy with mitomycin C under general anesthesia was done in all eyes.

Surgical steps

Superior quadrant fornix-based flap was created. A 3 mm × 3 mm half-thickness scleral flap was made. Mitomycin C (0.3 mg/ml)-soaked pieces of microsponge were applied under the scleral flap and the conjunctiva for 2–3 min. The area was washed thoroughly with 30 ml of balanced salt solution. A 2-mm radial incision was made starting from the gray zone up to the white zone, followed by entering Schlemm's canal externally. The incision was slowly deepened until seeping aqueous humor was observed. The Schlemm's canal was dissected by 120° in both directions using a trabeculotome probe. Trabeculectomy was done by cutting a 1 mm × 2 mm deep scleral flap. Peripheral iridectomy was done. The scleral flap and conjunctiva were sutured with 8–0 absorbable sutures.

Follow-up

Follow-up of the surgical outcome was achieved for each patient by measuring the postoperative IOP, evaluation of corneal transparency, measuring the corneal diameter and measuring the ocular axial length monthly for 3 months and then every 3 months for 12 months.

Success of surgery was determined mainly by stable IOP under 21 mmHg without antiglaucoma medications, which was considered the primary surgical outcome. Other signs were considered as secondary surgical outcomes, such as stable axial length, stable corneal diameter, and improvement of corneal transparency.

Statistical analysis

Categorical variables were described by number and percent, where continuous variables were described by mean and SD. χ^2 test was used to compare between categorical variables, whereas comparison between continuous variables was done by Mann–Whitney test. A two-tailed *P* value less than 0.05 was considered statistically significant. All analyses were performed with the IBM SPSS 28.0 software (IBM, SPSS Inc., Chicago, IL, USA).

Results

Baseline data of infants in the current study

A total of 19 infants were enrolled in this study. Of those infants, 10 (52.6%) were males and nine (47.3%) were females. The mean age of presentation of the infants was 2.8 months, with range between 15 days and 6 months. Of the 19 infants in this study, 13 (68.4%) had bilateral primary congenital glaucoma, 15 (78.9%) of the 19 infants had positive consanguinity between their parent, and 12 (63.1%) of the 19 infants had congenital heart disease diagnosed by echocardiography during the preoperative assessment, where 10 of them had atrial septum defect and two of them had patent ductus arteriosus.

Examination data of infants in the current study

Mean baseline IOP measured using the Shiotz tonometer by examination under sedation using halothane was 30.7 ± 10.2 mmHg, with range from 23.1 to 50.6 mmHg. The mean corneal diameter measured horizontally and vertically was 13.1 ± 1.2 mm, with range from 11 to 15 mm, and the mean axial length measured with ultrasound A scan was 20.9 ± 1.4 mm, with range from 19 to 23 mm.

Surgical outcome data in the current study

A total of 20 (62.5%) eyes showed successful IOP lowering after the surgery, with mean IOP of 15.3 mmHg, with range between 11 and 19 mmHg measured under general anesthesia using halothane, nine (28.1%) eyes required postoperative antiglaucoma medications to normalize the IOP, and three (9.3%) eyes required glaucoma drainage device surgery to normalize the IOP.

Distribution data according to surgical outcome

Table 1.

Relationship between patient demographic outcomes

There was no significant statistical difference between eyes with successful IOP lowering after the first surgery and eyes required antiglaucoma medications or glaucoma drainage device surgery regarding sex of the infant ($P = 0.784$), laterality of the disease ($P = 0.815$), and parental consanguinity ($P = 0.098$) (Table 2).

Relationship between clinical presentation and surgical outcome

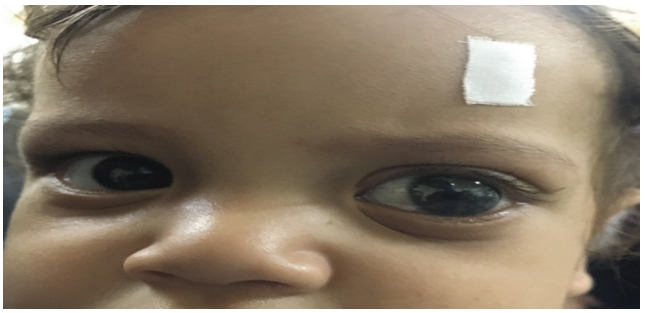
The need for postoperative antiglaucoma medications or glaucoma drainage device surgery was highly significantly correlated with the younger age of presentation ($P < 0.001$) and the higher presenting IOP ($P < 0.001$) (Table 3).

There was no significant statistical difference between eyes with successful IOP lowering after the first surgery and eyes that required antiglaucoma medications or glaucoma drainage device surgery regarding corneal diameter at presentation ($P = 0.785$) (Figs. 1–3).

Discussion

In our study, 32 eyes of 19 infants with primary congenital glaucoma were included. Of those infants,

Figure 1



Left congenital glaucoma.

Table 1: Comparison between results regarding demographic and clinical data

	Eyes with normal IOP after the surgery (*n*=20)	Eyes that required medications to normalize the IOP (*n*=9)	Eyes that required another surgery to normalize the IOP (*n*=3)
Age (months)			
Range	3-6	0.5-0.92	0.58-0.58
Mean±SD	4.01±0.77	0.75±0.17	0.58±0.0
Sex [*n* (%)]			
Male	11 (55.0)	5 (55.6)	1 (33.3)
Female	9 (45.0)	4 (44.4)	2 (66.7)
Bilateral PCG [*n* (%)]			
Bilateral	16 (80.0)	8 (88.9)	2 (66.7)
Unilateral	4 (20.0)	1 (11.1)	1 (33.3)
Parental consanguinity [*n* (%)]			
Yes	16 (80.0)	9 (100.0)	3 (100.0)
No	4 (20.0)	0	0
IOP (mm Hg)			
Range	23.1-30.12	33.53-36.49	36.17-50.60
Mean±SD	27.2±1.50	34.82±0.99	42.10±7.55
Corneal diameter (mm)			
Range	11.91-15.52	11.05-14.35	12.09-14.09
Mean±SD	13.19±0.99	12.95±1.00	13.1±1.00

IOP, intraocular pressure; PCG, primary congenital glaucoma.

Table 2: Statistical significance of results regarding demographic data

	Eyes with normal IOP after the surgery (*n*=20) [*n* (%)]	Eyes that required medications or another surgery to normalize the IOP (*n*=12) [*n* (%)]	*P*
Sex			
Male	11 (55.0)	6 (50.0)	0.784
Female	9 (45.0)	6 (50.0)	
Bilateral PCG			
Bilateral	16 (80.0)	10 (83.3)	0.815
Unilateral	4 (20.0)	2 (16.7)	
Parental consanguinity			
Yes	16 (80.0)	12 (100.0)	0.098
No	4 (20.0)	0	

IOP, intraocular pressure; PCG, primary congenital glaucoma.

Table 3: Statistical significance of results regarding clinical data

	Eyes with normal IOP after the surgery (*n*=20)	Eyes that required medications or another surgery to normalize the IOP (*n*=12)	*P*
Age (months)			
Range	3-6	0.5-0.92	<0.001**
Mean±SD	4.01±0.77	0.7±0.15	
IOP (mmHg)			
Range	23.1-30.12	33.53-50.60	<0.001**
Mean±SD	27.22±1.50	36.64±4.68	
Corneal diameter (mm)			
Range	11.91-15.52	11.05-14.35	0.785
Mean±SD	13.19±0.99	12.99±0.96	

**Highly statistically significant. IOP, intraocular pressure.

Figure 2

Bilateral congenital glaucoma with corneal edema.

Figure 3

Bilateral congenital glaucoma with clear cornea.

10 (52.6%) were males and nine (47.3%) were females. Of the 19 infants in this study, 13 (68.4%) had bilateral primary congenital glaucoma, six (31.5%) of the 19 infants in this study had unilateral primary congenital glaucoma, and 15 (78.9%) of the 19 infants had positive consanguinity between their parents. Mokbel *et al.* [3] studied the childhood glaucoma profile in Dakahlia in 2018 and found that males represented 63.8% of primary congenital glaucoma cases and 80% of cases were bilateral. Wagdy *et al.* [4] studied the epidemiological pattern of primary congenital glaucoma among children attending Menoufia University Hospital in 2021 and found that males represented 68% of primary congenital glaucoma cases and 84% of cases were bilateral, Malik *et al.* [5] mentioned that 83% of cases of primary congenital glaucoma in Saudi Arabia were bilateral and 90% of cases had positive consanguinity between their parents. Most of the results of these studies are comparable to the results of our study regarding the epidemiological characteristics of primary congenital glaucoma.

In our study, 12 (63.1%) of the 19 infants had congenital heart disease diagnosed by echocardiography during the preoperative fitness, where 10 of them had atrial septum defect and two of them had patent ductus arteriosus. Mansour *et al.* [6] studied ocular pathology in congenital heart disease in 240 patients presented to the medical center of the American University of Beirut in 2005 and found that 55% of cases had positive eye findings. Lewis *et al.* [7] discussed the role of genes in primary congenital glaucoma in 2017 and mentioned that various congenital heart defects are associated with developmental glaucoma. The results of these study agreed with the results our study regarding the probability of presence of congenital heart disease in infants with primary congenital glaucoma, which requires meticulous preoperative assessment by a pediatric cardiologist to evaluate the severity of condition before proceeding to the operation theater under general anesthesia.

In our study, 32 eyes of 19 infants with primary congenital glaucoma were included and underwent combined trabeculotomy and trabeculectomy with mitomycin C with follow-up for 1 year. A total of 20 (62.5%) eyes showed successful IOP lowering after the surgery, with mean IOP of 15.3 mmHg, with range between 11 and 19 mmHg, measured under general anesthesia using halothane; nine (28.1%) eyes required postoperative antiglaucoma medications to normalize the IOP; and three (9.3%) eyes required glaucoma drainage device surgery to normalize the IOP. The eyes that required antiglaucoma medications or glaucoma drainage device surgery had common features between them, such as positive consanguinity between parents, early age of presentation, high initial IOP at the time of presentation, and corneal haziness. Farid and colleagues studied the surgical outcome of primary congenital

glaucoma at Benha University Hospital in 2021 and mentioned that surgical success was achieved in 72.3% of cases without antiglaucoma medications and in 18% of cases with antiglaucoma medications. This study also mentioned that age at the time of surgery was positively correlated with success; the older the age, the better the outcomes. However, preoperative IOP was negatively correlated; the lower the preoperative IOP, the better the outcomes [8]. Bayoumi studied the surgical management of primary congenital glaucoma at Alexandria University Hospital from 2005 to 2012, with 5-year follow-up, and mentioned that 60% of cases were controlled by combined trabeculotomy and trabeculectomy with mitomycin C, 20% of cases required another surgery, and 20% were controlled by antiglaucoma medications. They mentioned that there is an agreement in cases that needed more than one surgical procedure; the most common presentation was corneal haze, indicating that this presentation was one of the most severe and with the worst prognosis among the presentations of congenital glaucoma [9]. Al-Hazmi and colleagues studied the correlation between severity of primary congenital glaucoma and success of surgery in 820 eyes from 1992 to 2002 in Saudi Arabia. This study considered IOP above 35 mmHg and poor corneal clarity as severe form of primary congenital glaucoma. The success rate of combined trabeculotomy and trabeculectomy with mitomycin C was 100, 80, and 70% for mild, moderate, and severe cases of primary congenital glaucoma, respectively [10].

Most of the results of these studies are comparable to the results of our study regarding the surgical outcome of combined trabeculotomy and trabeculectomy with mitomycin C and the risk factors for poor results such as positive consanguinity between parents, early age of presentation, high initial IOP at time of presentation, and corneal haziness.

In conclusion, this study is one of few studies to address primary congenital glaucoma in Egypt and more specifically, its most populous part, the region of Upper Egypt, where there is a high prevalence of primary congenital glaucoma owing to high consanguineous marital relations. In general, the outcome of combined trabeculotomy and trabeculectomy with mitomycin C in our study was comparable to outcome in other studies either inside Egypt or worldwide. Eyes that required antiglaucoma medications or glaucoma drainage device surgery had common features between them such as positive consanguinity between parents, early age of presentation, high initial IOP at time of presentation, and corneal haziness.

Financial support and sponsorship
Nil.

Conflicts of interest
None declared.

References

1 Allingham RR, Damji K, Freeman S, Moroi S, Shafranov G. Congenital glaucomas and developmental glaucomas with associated anomalies. In: Allingham RR, Damji KF, Freedman S, Moroi AE, Rhee DJ, editors. *Shields textbook of glaucoma.* 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2005. 235–271.

2 Francois J. Congenital glaucoma and its inheritance. Ophthalmologica 1972; 181:61–73.

3 Mokbel TH, El Hefney EM, Hagras SM, ALNagdy AA, Badawi AE, Kasem MA, *et al.* Childhood glaucoma profile in Dakahelia, Egypt: a retrospective study. Int J Ophthalmol 2018; 11:674–680.

4 Wagdy F, Helmy H, Kasemy Z. Epidemiological pattern of Primary Congenital Glaucoma among Egyptian Children aged 0-4 years Attending Minoufia University hospital. Egypt J Ophthalmol (Mansoura Ophthalmic Center) 2021; 1:138–150.

5 Malik R, Khandekar R, Boodhna T, Rahbeeni Z, Al Towerki AE, Edward DP, *et al.* Eradicating primary congenital glaucoma from Saudi Arabia: The case for a national screening program. Saudi J Ophthalmol 2017; 31:247–249.

6 Mansour A, Bitar F, Traboulsi E, Kassak KM, Obeid MY, Megarbane A, Salti HI. Ocular pathology in congenital heart disease. Eye 2005; 19:29–34.

7 Lewis CJ, Hedberg-Buenz A, DeLuca AP, Stone EM, Alward WLM, Fingert JH. Primary congenital and developmental glaucomas. Hum Mol Genet 2017; 26(R1):R28–R36.

8 Farid MF, Anany M, Awwad MA. Surgical outcomes of primary congenital glaucoma in children under one year from the Nile Delta. Clin Ophthalmol 2021; 15:1145–1151.

9 Bayoumi N. Surgical management of primary congenital glaucoma in egypt. J Egypt Ophthalmol Soc 2016; 109:85.

10 Al-Hazmi A, Awad A, Zwaan J, Al-Mesfer SA, Al-Jadaan I, Al-Mohammed A. Correlation between surgical success rate and severity of congenital glaucoma. Br J Ophthalmol 2005; 89:449–453.