WORLD VIEW

The Armenian Eye Care Project: surgical outcomes of complicated paediatric glaucoma

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Aim: To review surgical results of the Armenian Eye Care Project's (AECPP) efforts in paediatric glaucoma and to identify additional morbidity associated with its itinerant nature.

Methods: A non-randomised, retrospective historical cohort study of complicated paediatric glaucoma surgeries was performed by the AECPP at the Republican Eye Hospital. 38 eyes of 34 paediatric patients underwent mitomycin C trabeculectomies (MMCT) ($n = 20$) or Ahmed glaucoma valve (AGV) placements ($n = 18$) for complicated paediatric glaucomas. The median age was 12.5 years with a range of 6–17 years. Three clinical groups were represented: congenital glaucoma, Frank Kamenetsky syndrome (a glaucoma associated with uveitis), and Rieger's syndrome.

Results: Preoperative and postoperative intraocular pressures were similar for the two groups (MMCT v AGV = 30 v 32 mm Hg and 13.5 v 14.8 mm Hg, respectively; $p > 0.05$, t test). The AGV group was twice as likely to use medications postoperatively (44% v 23%; $p < 0.05$, t test). The MMCT group was more likely to have a three line drop in visual acuity than the AGV group (28% v 12%; $p < 0.05$, t test). The decreased visual acuity found postoperatively was most commonly secondary to cataract formation.

Conclusion: Total success rates were similar in the two surgical treatment groups. These compared favourably with success rates reported for non-itinerant surgery. Postoperative complications worsened by the itinerant nature of the project were few and represented 5% of the total surgical interventions. Ultimately, the installation of fellowship trained specialists is the most effective approach to minimising this problem and is recommended by the AECPP as a strategy for developing and recovering countries.

The initial goals of the Armenian Eye Care Project (AECPP) were to treat difficult cases, provide hospital infrastructure, develop western style educational programmes within Armenia, and develop a fellowship training programme. One of our major efforts was directed towards paediatric glaucomas resistant to or not appropriate for goniotomy and trabeculectomy. Historically, this group of patients is very difficult to treat. In addition to intraocular pressure control to prevent loss of vision, other important considerations include the production of cataracts and related amblyopia. This can be further complicated by a setting where a surgeon visits a hospital, performs paediatric glaucoma surgery, and then leaves after a short period of time. The follow up care in glaucoma surgery is an important integral part of surgical care for the patient. However, many cases found by visiting surgeons are serious and vision will be lost if surgical intervention does not occur. We, therefore, sought to examine our surgical missionary experience at the Malayan Eye Institute (formerly Republican Eye Hospital) in Yerevan, Armenia, in terms of effectiveness and difficulties that arose both perioperatively and secondary to its itinerant nature.

DESIGN AND METHODS

The study is a non-randomised, retrospective historical cohort study of complicated paediatric glaucoma surgeries performed by the AECPP at the Malayan Eye Institute. The AECPP is staffed by volunteer ophthalmologists who make biannual site visits. All paediatric glaucoma surgeries were performed by one surgeon (RH). Patients typically presented when a disease process produced a visible change in the appearance of a patient's eye or when the patient became visually symptomatic. Armenian patients also are referred from former Soviet republics and Russia. Patient treatment records are maintained by the

patients/parents and carried by them to the hospital for visits. Postoperatively, patients are seen by the operating physician at 6 month intervals, with the majority of follow up care performed by Armenian ophthalmologists. Goldmann applanation was initially unavailable. Maklikov indentation tonometry was used and converted to mm Hg by subtracting a conversion factor of 6 in higher ranges (>25) and a conversion factor of 5 in lower ranges (>10). This correlation, when compared against Tonopen pressures, seemed to correlate well (SD 1–2 mm Hg). Surgical success was defined as follows: complete success = IOP < 21 mm Hg without drugs; qualified success = IOP < 21 mm Hg with medications; failure = IOP > 21 mm Hg with/without drugs or additional glaucoma surgery needed or visually devastating complications. A total of 17 of the eyes had previously failed surgeries. Thirty eight eyes of 34 patients underwent surgery. The median age for the entire group of patients was 12.5 years with a range of 6–17 years. In this group of patients, a randomised trial was not possible as ocular morphology dictates the possible methods of intervention and posterior vitrectomy was initially unavailable.

Ahmed glaucoma valve (AGV) group

Patients who entered this group generally had sclera too thin for the creation of scleral flaps and/or conjunctiva too scarred for filtration surgery. A total of 18 eyes underwent implantation of an AGV. The technique has been described previously.^{1–4} The AGV was chosen because of Third World availability, ease of insertion, and decreased postoperative care demands. Important considerations are to place the drainage tube through a smaller fistula needle tract (25 gauge) in cases with thinner/buphtalmic sclera. This minimises peritube filtration and perioperative hypotony. In addition, implants are placed in inferior positions only in monocular patients to minimise morbidity associated with postoperative strabismus. The AGV should not

Figure 1 Frank Kamenetsky syndrome (a severe glaucoma associated with uveitis). There are some overlapping features with Rieger's syndrome but there is a history of enteritis, uveitis, and the systemic findings of Rieger's syndrome are absent.

be held in forceps over the valve mechanism housing as this may damage the plastic rivets and allow fibrovascular ingrowth and device failure.⁵ Finally, in cases where the sclera is too thin to allow fixation, the implant may be sutured to the tendinous insertion of the rectus muscle.

Mitomycin C trabeculectomy (MMCT) group

Patients who entered this group generally had shallow anterior chambers from synechial closure of the filtering angle, less conjunctival scarring, and better visual quality. MMCT for primary and secondary surgical treatment of paediatric glaucomas has been previously described.^{6–8} A total of 21 MMC trabeculectomies were performed. Intraoperative

Figure 2 Rieger's syndrome presentation with marked iris abnormalities typical of those patients undergoing surgery.

mitomycin C was used at a concentration of 0.25 mg/ml. A volume of 0.5 ml of the solution was applied with a truncated Weck cell sponge for 2 minutes with the conjunctiva held elevated and then for an additional 2 minutes with the conjunctiva draped over the sponge for a total of 4 minutes. Trabeculectomy flaps are usually closed with 4–6 sutures and released postoperatively by selective laser suture lysis. Manually released sutures were not used. In adapting to the situation in Armenia, a number of modifications were made to the trabeculectomy operation. Scleral flaps for trabeculectomies were cut thicker to allow effective tamponade postoperatively. In addition, the sclerostomies were cut in a conservative manner allowing 0.5–0.75 mm shoulders adjacent to the sclerostomy. These modifications allowed the use of 2–4 sutures to achieve a physiological outflow and decreased the need for laser suture lysis. Laser suture lysis was available during the surgical missions but not at other times.

RESULTS

The mean age of the two groups was virtually identical and three major diagnosis groups were represented: congenital glaucoma, Frank Kamenetsky syndrome (a severe glaucoma associated with uveitis; Fig 1), and Rieger's syndrome (Fig 2; Table 1).

Preoperative and postoperative intraocular pressures were similar for the two groups (MMCT *v* AGV = 30 *v* 32 mm Hg and 13.5 *v* 14.8 mm Hg, respectively; *p* > 0.05, *t* test). The data are also presented in Tables 2, 3, and 4.

Table 1 Patient demographics

Race	Armenians	White race
Number of patients	34	
Number of eyes	38	
Age (years)	AGV (n=18)	MMCT (n=20)
Mean	12.5	12.7
SD (years)	2.8	2.4
Range (years)	6–16	10–17
Congenital glaucoma	12 (66%)	6 (30%)
Frank Kamenetsky syndrome	3 (17%)	11 (55%)
Rieger's syndrome	3 (17%)	3 (15%)

Table 2 MMCT group

Patient	Age	IOP		Vision		Medications		Follow up
		Pre	Post	Pre	Post	Pre	Post	
1	16	45	11	20/200	20/200	5	0	6
2	16	45	14	20/200	20/200	5	0	6
3	15	35	35	20/30	20/300	2	2	12
4	16	29	11	4/200	20/100	2	0	12
5	15	26	14	20/40	20/200	2	0	18
6	17	26	10	HM	HM	2	0	18
7	11	27	10	4/200	20/600	2	0	12
8	12	26	19	5/200	2/200	1	1	12
9	10	26	24	5/200	NLP	1	2	36
10	10	30	10	20/60	20/60	2	0	18
11	10	29	16	20/200	20/200	2	0	24
12	10	40	8	20/500	20/250	2	0	18
13	12	34	10	20/200	20/200	2	0	24
14	11	44	10	20/60	NLP	3	0	24
15	11	24	12	20/40	20/40	3	0	6
16	11	24	7	20/600	20/600	2	0	60
17	14	26	11	2/200	4/200	2	1	6
18	11	26	9	20/40	20/50	3	0	6
19	11	22	6	20/50	20/40	3	0	6
20	15	24	24	LP	NLP	2	2	18

Table 3 AGV group (18 eyes)

Patient	Age (years)	IOP		Vision		Medications		Follow up (months)
		Preop	Postop	Pre	Post	Pre	Post	
1	16	62	13	20/300	20/300	2	0	60
2	6	26	17	LP	LP	2	1	12
3	14	32	16	20/400	20/400	2	1	12
4	12	28	10	LP	LP	2	0	18
5	10	29	24	HM	2/200	2	2	18
6	12	24	8	5/200	5/200	2	0	12
7	13	29	20	CF	CF	2	1	30
8	9	25	16	20/300	20/300	2	0	18
9	10	44	29	4/200	LP	2	2	30
10	14	25	10	LP	LP	1	0	16
11	11	24	17	CF	LP	1	2	24
12	14	29	11	HM	HM	0	0	18
13	10	22	17	2/200	2/200	1	1	24
14	15	30	12	LP	LP	2	0	18
15	16	30	13	20/300	20/300	3	0	18
16	16	29	11	LP	LP	2	0	12
17	14	45	10	HM	HM	2	0	18
18	14	42	13	CF	CF	4	1	6

Table 4 Summary of surgical results

	AGV (n=18)	MMCT (n=21)
Follow up (months)		
Range	6–60	6–60
Mean	18.8	16
SD	9.6	12.5
Vision		
W/n 2 lines	15 (83%)	12 (57%)
Improved	1 (5%)	3 (14%)
Worse	2 (12%)	6 (28%)
Intraocular pressure		
Preop	32.0 (10.1) mm Hg	30.1 (7.4) mm Hg
Postop	14.8 (5.4) mm Hg	13.5 (7.0) mm Hg
Preop medications		
Mean	1.9	2.2
SD	0.8	1.1
Postop medications		
Mean	0.7	0.4
SD	0.8	0.75
% Eyes postop medications	44%	23%
Success		
Total	16 (88%)	18 (86%)
Complete	10 (55%)	16 (76%)
Qualified	6 (33%)	2 (10%)
Failure	2 (12%)	3 (14%)

The AGV group was twice as likely to use drugs postoperatively (44% v 23%; $p < 0.05$, t test) as the MMCT group.

The MMCT group was more likely to have a three line drop in visual acuity than the AGV group (28% v 12%; $p < 0.05$, t test). The decreased visual acuity found postoperatively was, most commonly, secondary to cataract formation (Tables 2, 3, 4, and 5).

The success rates for the two groups were similar: MMCT v AGV = 88 v 86%. A slightly higher number of complete successes were found in the MMCT group (Tables 2, 3, 4, and 5). Hypotony in the MMCT group with shallow anterior chambers did create significant morbidity in terms of corneal and lenticular problems (Table 5). There was one choroidal haemorrhage in each group of patients. The AGV group had one patient with a limited choroidal haemorrhage not affecting visual acuity. The MMCT group had one patient who developed a massive choroidal haemorrhage after suture lysis with loss of all vision. This haemorrhage occurred after the surgeon had left and was not drained by on-site personnel. Finally, follow up periods of both groups were comparable

Table 5 Complications

	Number	(%)
AGV (18 eyes)		
Tube extrusion	1	5
Reposition of tube	1	5
Transient shallowing of AC	2	10
Limited choroidal haemorrhage	2	10
Corneal blood staining	1	5
Scleral graft thinning	1	5
MMCT (20 eyes)		
Corneal blood staining	2	10
Massive choroidal haemorrhage	1	5
Choroidal detachment requiring drainage	1	5
Shallow AC requiring reformation	3	14
Corneal decompensation	2	10
Aqueous misdirection	1	5
Lenticular opacification	6	29

with a median of at least 1.5 years (Tables 2, 3, 4). There was one case of fibrovascular downgrowth secondary to a long term extruded drainage tube. This was successfully treated with inversion of the resulting fibrous cylinder under viscoelastic and cauterisation of the entry fistula tract with a micropoint intraocular cautery.

DISCUSSION

Three major diagnosis groups were represented: congenital glaucoma, Frank Kamenetsky syndrome (Fig 1), and Rieger's syndrome (Fig 2). In all cases of congenital glaucoma, patients had either failed goniotomy/trabeculotomy or were so grossly buphthalmic that goniotomy/trabeculotomy was not a reasonable surgical option secondary to derangement of outflow structures. Frank Kamenetsky syndrome has been described in the Soviet era ophthalmic literature as a disease with iris hypoplasia, focal stromal defects, and secondary glaucoma that is transmitted in a sex linked recessive fashion. Armenian ophthalmologists include a disease under this diagnosis that starts with a prodromal enteritis followed by a severe iridocyclitis leading to iris atrophy, focal iris stromal defects, and secondary angle closure.⁶ It is possible that these represent two different types of secondary glaucomas (with uveitis separating the two) but we are not able to separate them further in this report. The pathology, epidemiology, and treatment of

Frank Kamenetsky syndrome will be studied further with USAID funding. Rieger's syndrome was a surprisingly common aetiology and its genetics allowed earlier identification of some patients with glaucoma.

Ocular morphology frequently dictated the type of surgical intervention. In grossly buphthalmic eyes, thin sclera precluded the creation of scleral flaps and AGVs were placed. Conversely, phakic eyes with shallow anterior chambers secondary to peripheral anterior synechiae were selected for MMCT. Posterior vitrectomy was unavailable during the time of the study.

Laser suture lysis was performed only during the time the surgeon visited. Had laser suture lysis been available for the entire postoperative period, final intraocular pressures may have been lower in the MMCT group of patients.

Trabeculectomy without antimetabolites in the treatment of congenital glaucoma has dismal success rates (30% >3 years). These early results have prompted use of mitomycin C in complicated paediatric glaucomas. With mitomycin C as antifibrosis therapy, success rates between 56% and 95% have been reported.⁷⁻⁹ However, enthusiasm for this increased success rate should be tempered by cataract formation and the risk of infection that these children will face in the future.

Reported success rates for aqueous drainage implants in congenital glaucoma are widely variable—44–90%.^{1-4 10-17} Our success rates in both groups were comparable with those in the literature without delegated postoperative care. However, between the two groups of patients, there are some characteristic differences. Lenticular opacifications and lower intraocular pressures were more common in the MMCT group, whereas strabismus, higher intraocular pressures, and a greater need for postoperative antiglaucoma medications and revisions were found in aqueous drainage implant groups.

Although our surgical complication rate is reassuring for itinerant missionary ophthalmic surgeons, some complications could have had much lower morbidity with the presence of an on-site fellowship trained glaucoma specialist. Specifically, one choroidal haemorrhage in a patient's remaining eye was not drained and light perception was lost. One case of prolonged flat anterior chamber secondary to aqueous misdirection led to corneal decompensation and cataract formation in an 11 year old child. Finally, postoperative bleeding at the site of a scleral flap resulted in a hyphaema with transient closure of a trabeculectomy flap. The resultant high intraocular pressure produced corneal blood staining. The complications of the choroidal haemorrhage and the corneal decompensation may have been moderated by an on-site fellowship trained glaucoma specialist. This represents a 5% level of complications worsened by the itinerant nature of the care provided. This is sobering but needs to be weighed against the certainty of visual loss in 100% of these patients without intervention.

Itinerant missionary ophthalmic surgical care is a start towards elevation of ophthalmic care in recovering or developing countries. We believe that any truly effective strategy to raise the level of ophthalmic care in selected countries involves, at its core, fellowship training. This provides the candidate with advanced clinical skills and the necessary

paradigm shift to continuously deliver an elevated standard of care. We applaud the courage of those individuals who have returned to their country after fellowship training to help their people.

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