Childhood Glaucoma Surgery in Developing Countries

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Introduction

The surgical management of childhood glaucoma is one of the biggest challenges in the field of glaucoma and an even greater challenge in the developing world. Despite its rarity, childhood glaucoma is one of the leading causes of admission to schools for the blind in both developed and developing countries. Glaucoma accounts for 3–5% of blindness in children worldwide [1, 2], but in certain regions such as Latin America and the Caribbean, it is the cause of blindness in 7% of cases. Overall, the number of blind children in the world is estimated to be 1.4 million, with a disproportionate number, approximately three-quarters, living in the poorest regions of Africa and Asia [3].

Surgery is the main modality of treatment for childhood glaucoma, and a proportion of patients need multiple surgical interventions for long-term control of intraocular pressure (IOP). While early diagnosis and appropriate surgical technique are important in the management of this condition, the treatment does not end there. It is almost a lifelong commitment for both the parents and doctors caring for these children. Childhood glaucoma blindness in the developing world can be minimized by earlier detection and treatment and by strategies to reduce cases that are lost to follow-up.

Although therapies have improved the prognosis of many childhood glaucomas, the number of complex glaucoma cases being seen has increased, needing expert care and multiple interventions. Although there is paucity of data on the surgical management of childhood glaucoma from the developing world, the aim of this chapter is to highlight the challenges of managing childhood glaucoma in developing countries, the different surgical options available, and their outcomes.

Challenges of Managing Childhood Glaucoma in Developing Countries

Several problems and issues are associated with the effective management of childhood glaucoma in developing countries [4] and include:

1. The burden of disease is greater in the developing world. Primary congenital glaucoma (PCG) is the most common form of childhood glaucoma. A recent published report from an urban center in Miami, Florida, USA, reported that PCG constituted the largest group (32%) among

Electronic Supplementary Material The online version of this chapter (https://doi.org/10.1007/978-3-319-54003-0_11) contains supplementary material, which is available to authorized users.

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A. L. Grajewski et al. (eds.), *Surgical Management of Childhood Glaucoma*,
https://doi.org/10.1007/978-3-319-54003-0_11

all types of childhood glaucoma [5]. However, its incidence varies widely, occurring in approximately 1 in 20,000 in Western populations [6, 7] but more frequently in the developing world. PCG is often transmitted in an autosomal recessive pattern and may occur up to ten times more frequently in certain ethnic and religious populations where consanguineous marriage is socially and culturally acceptable. The highest reported prevalence is found in Slovakian gypsies at 1 in 1250 [8], and among Saudi Arabians, it is 1 in 2500 [9]. In southern India, prevalence of PCG is estimated at 1 in 3300 live births, accounting for 4.2% of overall childhood blindness [10].

2. More severe disease phenotype and late presentation of the disease in an advanced stage when it is less amenable to successful surgical treatment.
3. Limited number of trained personnel with the appropriate skill sets (surgeon, anesthesiologist, nursing staff, and other professionals) for microsurgical treatment and postoperative care.
4. Cultural fear of surgical treatment, which must be overcome.
5. Incurable disease that needs ongoing follow-up, which is often inadequate in treated cases leading to poor prognosis. So the initial benefits of glaucoma surgery may be lost if there is no continuity of care for these children.
6. Limited access to health care and long travel distances to health-care centers.
7. Poor socioeconomic status of the family and lack of affordability of health care.
8. Barriers to affordability and availability in rural areas of topical medications along with variable nature and efficacy (quality) of available generic medication.

Surgical Therapy of Childhood Glaucoma in Developing Countries

Goniotomy

The classic surgery for PCG is goniotomy as described by Barkan [11]. PCG, which occurs in India, the Middle East, South Africa, and several other parts of the developing world, is mostly familial compared to the West and responds less favorably to goniotomy [4]. Furthermore, in developing countries, the disease is of a more severe phenotype and presents late with severe corneal edema precluding goniotomy [12, 13]. Compared to Western and European populations, a large number of Indian children (>80%) present with a severely cloudy cornea at birth, thus in whom goniotomy is technically impossible [14]. In such cases trabeculotomy ab externo is advocated as the initial surgical procedure of choice by several authors [15–19]. However, if corneal clarity and diameter of the cornea permit, goniotomy may be attempted by surgeons who are trained and competent in performing the surgery (see Chap. 4).

Trabeculotomy Ab Externo

Trabeculotomy ab externo was simultaneously and independently described by Burian [20, 21] and by Smith [22] in 1960 and is our preferred angle surgical technique due to a number of advantages [19, 23] over goniotomy in the management of childhood glaucomas in our part of the world. The popularity of trabeculotomy ab externo as an initial procedure in the surgical management of PCG has been championed by a number of authorities [23, 24] (see Chap. 4).

Trabeculectomy with or Without Antiscarring Therapy

Trabeculectomy is a procedure that most ophthalmologists are familiar with and is technically easier than goniotomy or trabeculotomy. Hence some surgeons in developing countries perform only trabeculectomy. However, many others do not consider it as a first-line procedure for PCG in view of the higher incidence of complications and lower success rates reported in the early literature [25–27]. Nevertheless, several subsequent reports documented successful results following primary trabeculectomy for PCG that are comparable to goniotomy or external trabeculotomy [28–32]. In refractory PCG following

failed angle surgery or combined trabeculotomy-trabeculectomy (CTT), trabeculectomy with mitomycin C (MMC) [33, 34] may be an option (see Chap. 5).

Combined Trabeculotomy-Trabeculectomy (Ab Externo)

The prevalence of children presenting with glaucoma and opaque corneas precluding goniotomy, coupled with encouraging reports of primary trabeculectomy in PCG, prompted Indian surgeons to combine trabeculotomy ab externo with trabeculectomy as the initial surgery for PCG in the Indian patient population [35]. In mild forms of PCG, i.e., mild angle anomaly and corneal haze, we perform trabeculotomy ab externo in isolation. However, in most cases of PCG with megalocornea and associated significant corneal edema, we prefer CTT. In secondary childhood glaucomas, if Schlemm canal is technically possible to explore, we perform CTT. However, if Schlemm canal is difficult to dissect or is anatomically absent, we perform trabeculectomy alone.

Surgical Technique:

1. A limbus-based conjunctival flap is raised 7 mm from the superior limbus with blunt-tipped Westcott scissors and plain forceps. The dissection is normally done in the episcleral plane. Hemostasis is meticulously maintained throughout the dissection of the conjunctival flap with bipolar wet-field cautery.
2. Retracting the conjunctival flap gently toward the pupil, *light cautery is applied on the sclera to outline the sides of a 4 mm equilateral triangle* with its base at the limbus. *The authors prefer triangular flap* as it allows adequate exposure of Schlemm canal and involves less scleral dissection than a rectangular flap.
3. *A one-half thickness scleral incision* is then made with a no. 11 blade. Here we must bear in mind that the sclera in a buphthalmic eye is usually much thinner than in the adult eye.
4. *The partial thickness scleral flap is then dissected* toward the limbus using a no. 15 blade. The flap is held with Pierse-Hoskin forceps during the dissection. Care should be taken to maintain the same plane while dissecting the scleral flap, especially near the limbus.
5. *Surgical landmarks and anatomy of the limbal region should be carefully identified* before one can proceed to the next step. Closest to the limbus is a transparent band of deep corneal lamellae, behind which is a narrow grayish-blue band, which is an external landmark of the trabecular meshwork. The grayish-blue band is followed by white, opaque sclera. The junction of the posterior border of the grayish-blue band and the opaque sclera is the external landmark for finding Schlemm canal. In most eyes, this is situated between 2 and 2.5 mm behind the surgical limbus. The second landmark is one or more perforator vessels entering the sclera, indicating the area of Schlemm canal. Another landmark is a depression or dip at the area of Schlemm canal—the continuity of the two zones is not smooth; there is a dip between blue and white zone, which marks the junction.
6. *A 2 × 2-mm-deep block is outlined* without penetrating the anterior chamber (AC), which marks the area of the sclerostomy.
7. *A central radial incision is then made across the scleral spur.* The objective of this radial incision is to cut the external wall of Schlemm canal and to avoid entering the AC. It is important to bear in mind that Schlemm canal is separated from the AC only by the trabecular meshwork.

   This is the most delicate step in the surgery and demands utmost microsurgical skill. Under high magnification the radial incision is gradually deepened with a no. 11 blade until it is carried through the external wall of Schlemm canal, at which point there is a gush of aqueous, occasionally mixed with blood. *In our experience, a drop of aqueous is more common than a drop of blood.* The dissection is carefully continued through the external

wall until the inner wall is characteristically slightly pigmented and is composed of criss-crossing fibers. Vannas scissors are used to enlarge the lumen of the canal. Some surgeons confirm passage into the canal by passing a 6-0 nylon/prolene suture into the canal, as described by Smith [22].

8. *The internal arm of the trabeculotome is introduced into the canal* with external parallel arm as a guide. Once 90% of the trabeculotome is within the canal, *it is rotated into the* AC until 75% of the probe arm length has entered, and the instrument is withdrawn. About 2–2½ clock hours of the internal wall of Schlemm canal and trabecular meshwork are disrupted by rotation of the trabeculotome into the AC. The same procedure is repeated on the other side. In total, about 100–120° of trabecular meshwork is opened by this technique.
9. *Excess force should not be used while introducing the probe into the canal*, to avoid creating a false passage. If the probe does not slip easily down the canal, it should be withdrawn and dissection of the outer wall continued until the surgeon is satisfied that all fibers of the outer wall are severed. The probe is then reintroduced into the canal to complete trabeculotomy.

   As the probe passes into the AC, minimal resistance is felt while disrupting the inner wall of the canal. *There may be minimal intracameral bleeding from the inner wall,* leaving a small hyphema that often resolves in a few days.
10. *The probe is swept in a plane parallel to the iris.* If done incorrectly, this may cause iridodialysis. Anterior rotation can cause trauma to the Descemet membrane.
11. *The trabeculotomy has been completed and now trabeculectomy has to be performed.* The deep block is excised using Vannas scissors.
12. *An iridectomy is then completed.* It is imperative that the base of the iridectomy opening is wider than the trabeculectomy opening to prevent ostium block and iris pillar attachment to the ostium causing pupillary peaking.
13. *The scleral flap is then closed* with one to three 10-0 nylon sutures, one at the apex and one on each lateral side of the triangular flap. The knots should be buried to avoid later exposure through the conjunctival tissue.
14. *Conjunctiva and Tenon capsule are then closed* with a running suture of an absorbable material (e.g., 8-0 vicryl).
15. In highly buphthalmic eyes, Schlemm canal may not be located with certainty. In such cases it is possible to convert the procedure to a trabeculectomy despite the lack of successful trabeculotomy.
16. *Some surgeons prefer to perform a paracentesis opening* with a beveled corneal incision at the beginning of the surgery. In such a situation, the anterior chamber is reformed with balanced salt solution, and patency of the trabeculectomy can be tested at the conclusion of the surgery. The authors, however, do not prefer making a paracentesis opening.
17. If bilateral surgery is needed, both procedures are performed at the same operating sessions, although an entirely different set of draping, gloves, drops, instruments, and irrigating solutions for each eye is necessary.
18. Subconjunctival dexamethasone injection (0.2 ml) is given, and a drop of cycloplegic and antibiotic is instilled into the conjunctival sac before a patch and shield are applied to the eye.

Refer to Video 11.1 for the surgical technique of primary CTT on a 3-month-old child with primary congenital glaucoma.

When primary surgery fails, medical treatment may be initiated with topical carbonic anhydrase inhibitors, beta-blockers, and prostaglandin analogues (in that order) after ruling out contraindications. If the medical treatment is ineffective, either trabeculectomy with antiscarring agents or a GDD is our next choice based on the health of the conjunctiva and severity of the disease. We consider trabeculectomy with MMC (0.4 mg/ml for 2 min) in the superotemporal or superonasal quadrant with unscarred conjunctiva. When trabeculectomy fails, bleb needling with

adjunctive antimetabolite use is an option, especially if the sclerostomy is patent and the flap edge is visible [36].

Glaucoma Drainage Device (GDD) Surgery

GDDs are indicated when primary surgery fails or in certain secondary glaucomas even as a primary procedure. However, in certain parts of the developing world, the cost of these devices prohibits their regular use. Although the Ahmed glaucoma valve (AGV) (New World Medical, Rancho Cucamonga, CA, USA) has been available in India for the past 1–2 decades, the Baerveldt implant has never been made available. Since 2014, we have been using a prototype similar to the Baerveldt, the Aurolab aqueous drainage implant (AADI), developed by the Aravind Group (Madurai, Tamil Nadu, India). We use both flow-restricted, e.g., AGV, and non-flow-restricted, e.g., AADI, implants. The choice of device is dependent on the surgeon's comfort and the availability of the GDD. Both types of GDD are equally effective in controlling IOP. However, studies in adults suggest that the need for medications is higher with flow-restricted GDDs, and hypotony-related problems are higher with non-flow-restricted GDDs [37]. One has to weigh the benefits and risks on an individual patient basis and choose the procedure that is safe and effective. Our indications for GDD surgery are failed CTT surgery in PCG as a primary procedure or after failed trabeculectomy surgery in eyes with aniridia, Sturge–Weber syndrome, and other secondary childhood glaucomas and as primary surgery for secondary glaucomas following congenital cataract surgery, uveitis, trauma, steroid use, post-keratoplasty, and post-vitreoretinal surgery or in eyes with severely scarred conjunctiva.

Proper planning, patient selection, and meticulous surgery are important for safe and successful surgery. With regard to technique, we prefer a superotemporal location and most often or always a limbal-based conjunctival flap (6 mm limbal-based conjunctival opening, 6 mm posterior to the limbus to reduce the size of the conjunctival opening). We prefer double ligature of the tube using 6/0 Vicryl. We use a 24G needle to perform 3–4 mm needle track, and the AC is entered parallel to the limbus. For eyes having a pars plana GDD, a 25G needle entry is preferred. We prefer to place the tube in the ciliary sulcus in pseudophakic eyes, leaving a long tube to allow visualization of the tube tip through the pupil and to avoid the tube blocking. In eyes with aniridia, a short tube with oblique entry into the AC oriented away from the crystalline lens can help prevent lenticular touch. We leave longer tubes in uveitic eyes to decrease the risk of occlusion from peripheral anterior synechiae. Pars plana tube insertion is preferred in vitrectomized eyes or eyes with shallow anterior chamber or those with penetrating keratoplasty. The conjunctiva is closed with 8/0 Vicryl in a continuous fashion with a mattress suture, ensuring traction-free closure. Continuous mattress conjunctival closure provides a posterior suture line, watertight closure, and with minimal discomfort. Refer to Videos 11.2 and 11.3 for the surgical technique of an AADI and AGV implant.

We find the use of ICare tonometer (Icare Finland Oy, Vantaa, Finland) for IOP estimation and ultrasound biomicroscopy to evaluate the bleb dimensions and the fluid pocket very useful in decreasing the number of examinations under anesthesia in these children. Ultrasound also helps to identify posterior segment complications that may occur in the immediate or late postoperative period.

A hypertensive phase occurs at 3–4 weeks after AGV implantation and the 9–10th week following an AADI implant. Hypertensive phase is defined as elevated IOP >21 mmHg after initial low or normal IOP in the presence of cystic and tense bleb. Starting prophylactic glaucoma medications post AGV implantation can decrease the rate of hypertensive phase. The high IOP during the hypertensive phase is managed with topical and oral glaucoma medications. In our experience the number of eyes needing medications for the hypertensive phase was close to 46% with flow-restricted and 26% with non-flow-restricted implants (unpublished data). With IOP control, the majority of the corneas are clear (Fig. 11.1); however, aftercare includes frequent follow-up,

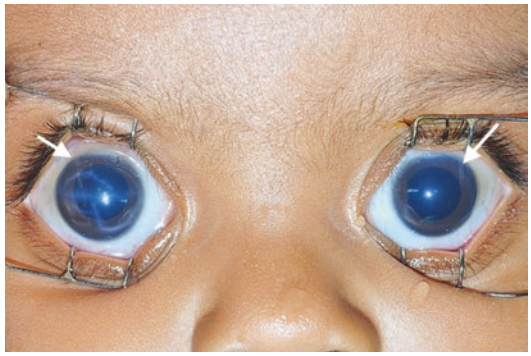

Fig. 11.1 Postoperative appearance after Ahmed glaucoma valve implantation in a child with primary congenital glaucoma. Both eyes show clear corneas and Haab striae in the right eye and well-placed tubes (*arrows*) in a child with failed combined trabeculotomy and trabeculectomy

not just for IOP evaluation but for tube- and plate-related complications, which are much higher in children compared to adults [38, 39].

Laser Surgery

Cyclodestructive procedures are preferred in patients with limited visual potential, with high risk of intraocular complications with incisional surgery, and with severely scarred conjunctiva precluding GDD surgery or as an adjunct when the GDD surgery is failing. The commonly used cycloablative procedure is contact transscleral cyclophotocoagulation (TSCPC) using Nd:YAG and 810 nm diode laser with a G-probe [40]. The power used is 1500–2000 mW with a soft pop. In children we limit the treatment to 180° or to 20–25 shots to prevent hypotony and phthisis. The laser can be repeated if needed, and repeat laser is not done earlier than 3–4 months after first laser. The placement of the G-probe and laser delivery is based on the limbal anatomy and the globe enlargement. Transillumination to identify the area of the ciliary body helps to deliver the laser appropriately. We avoid the areas of thinning and staphyloma, as well as pigmented areas, to help prevent inadvertent perforations. We also avoid delivering laser at the 3 and 9 o'clock position to avoid ciliary nerve damage increasing risk of subsequent corneal anesthesia and corneal complications. We restrict using TSCPC to eyes with refractory glaucomas and poor visual potential. We also use it as adjunct in treating eyes with failed implants before a second implant is planned. The IOP control is not uniform, with a proportion of children requiring repeat interventions. In conditions where vitreoretinal surgery is needed with refractory glaucoma or those with ciliary staphylomas or in eyes with sclerocorneas, we prefer intraocular cycloablation with endocyclophotocoagulation. We use a straight or a curved laser probe with endoscopic visualization using much lower energy (250–300 mW) and treat 270° of the ciliary processes.

Surgical Outcomes of Primary Congenital Glaucoma in Developing Countries

There are very few reports on the outcomes of goniotomy in PCG eyes with corneal clouding. Bowman et al. presented the results of goniotomy to treat advanced PCG with late presentation in Tanzania, demonstrating a 60% success rate after one goniotomy and an 89% success rate after two goniotomies in 36 eyes followed up for only 12 months [41]. Al-Hazmi et al. reviewed 532 children with PCG and stratified them into mild, moderate, and severe disease based on IOP, corneal diameter, and corneal clarity [42]. Patients with milder form of the disease had good surgical outcomes with goniotomy; however, outcomes were poor in the advanced stage of the disease irrespective of the surgical procedure. They concluded that surgical success decreased with disease severity and that CTT with MMC would provide best results in advanced cases. Similar results have been published by several authors from different parts of United Arab Emirates, Africa, and India. Ben-Zion et al. reported that advanced stage of the disease and late presentation at surgery are related to poor surgical outcomes [43]. They used trabeculotomy and the AGV to treat 20 patients (38 eyes) with PCG in Ethiopia. They also detailed problems of surgery in the advanced stages of the disease with extremely buphthalmic eyes. Furthermore, authors reporting on a series of West African children with PCG described poor results from CTT,

with probability of success falling from 83% at 6 months to 44% at 1 year, possibly due to severe disease at presentation, racial influences, and no use of antiscarring agent [44].

Elder reported poor long-term success (IOP ≤ 21 mmHg and no medication) in Palestinian Arab children with PCG undergoing goniotomy and trabeculotomy as compared to primary trabeculectomy without antiscarring agents [45]. In a subsequent study by the same author comparing retrospective trabeculectomy data to prospectively collected CTT data, both without antiscarring agents, the cumulative chance of success for CTT was 93.5% compared to trabeculectomy, which was 72% after a 24-month follow-up [46]. For a similar follow-up in the same population, trabeculotomy had only a 51% cumulative chance of success. Mullaney et al. reported that CTT was superior to trabeculotomy or trabeculectomy alone; however, they did use MMC [47].

Whether primary CTT is superior to trabeculotomy or trabeculectomy alone is a subject of considerable debate. In a small series of seven Arab Bedouin infants, Biender and Rothkoff found no difference between trabeculotomy and CTT in patients with PCG [48]. Dietlein et al. investigated retrospectively the outcome of trabeculotomy, trabeculectomy, and combined procedure as initial surgical treatment in PCG [49]. Although the combined procedure seemed to have favorable outcome, the advantage of this procedure over trabeculotomy or trabeculectomy was not significant after 2 years when assessed by life table analysis. The authors concluded that the prognosis of primary ab externo glaucoma surgery in PCG seemed to be governed more by the individual course and severity of the disease than by modification of surgical techniques. In a retrospective, comparative study, Lawrence and Netland concluded that trabeculectomy and CTT with MMC were equally effective at lowering average IOP in children [50]. However, CTT was associated with greater long-term success. In a prospective comparative study conducted in Egypt, Khalil and Abdelhakim concluded that both primary trabeculotomy and CTT with MMC had similar outcomes, which could mean that trabeculotomy could be resorted to first [51].

The argument in favor of primary CTT in some ethnic populations is the higher incidence of successful IOP control with a single operative procedure, as has been reported from India and the Middle East (Fig 11.2) [42, 49, 52]. In the largest Indian series of 624 eyes of 360 consecu-

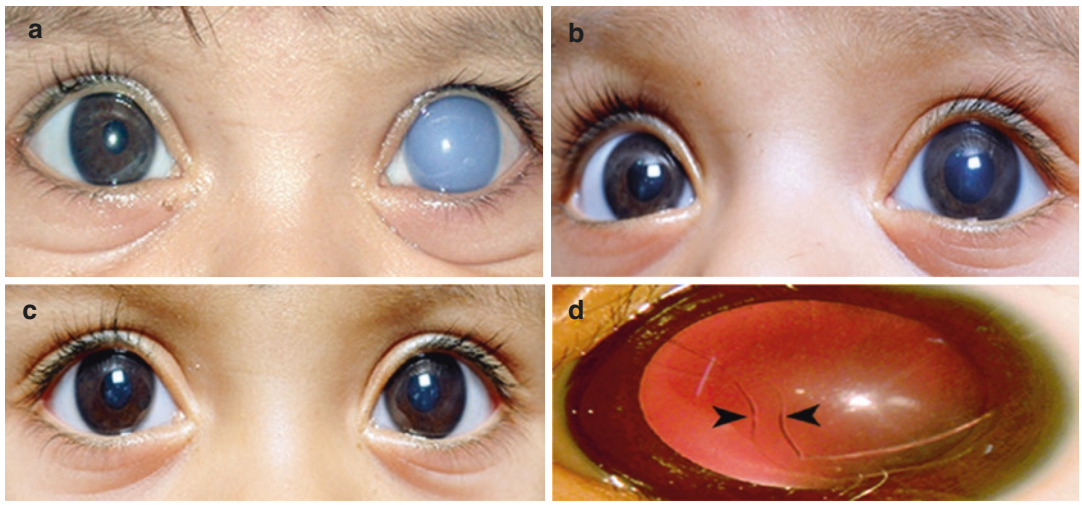

Fig. 11.2 (**a**) Preoperative appearance of the left eye in a 3-month-old child with primary congenital glaucoma who presented with acute corneal hydrops. (**b**) Two-week postoperative appearance of the left eye showing dramatic improvement of corneal edema. (**c**) Six-month postoperative appearance showing normal corneal clarity of the left eye. (**d**) Photomicrograph of the left cornea showing Haab striae (*arrows*)

tive Indian children with PCG undergoing CTT without MMC, IOP control was achieved in 85% of children after 1 year, reducing to 58% after 6 years. Forty-two percent of patients achieved vision of ≥20/60 [52]. Furthermore, primary CTT is a promising surgical technique for the management of different forms of childhood glaucoma in the developing world.

Although CCT offers the advantage of the dual mechanism of IOP control (trabeculotomy-trabeculectomy) and good long-term results have been reported by authors from India and various other parts of the developing world, there is always a search for the best surgical technique that should be employed in the given patient. Recently 360° trabeculotomy has emerged as an attractive choice that can be performed with a blunted 6/0 Prolene suture or with the help of an illuminated microcatheter. Shi et al. retrospectively studied 43 consecutive cases of Chinese eyes with childhood glaucoma undergoing trabeculotomy performed using either an illuminated microcatheter or a rigid probe trabeculotome [53]. They found microcatheter-assisted circumferential trabeculotomy to be significantly more effective at 1 year and as safe as traditional trabeculotomy with a rigid probe. Similarly, Shakrawal et al. from India performed a randomized clinical trial and concluded that in PCG, illuminated microcatheter-assisted circumferential trabeculotomy performed better than conventional partial trabeculotomy at 1-year follow-up and resulted in significantly lower IOP measurements [54]. However, a steep learning curve and the cost of therapy using disposable illuminated microcatheter is a limiting factor, especially in the context of the developing world. In a prospective study from India, illuminated microcatheter-assisted circumferential trabeculotomy achieved comparable results to primary CTT with MMC [55]. Hence, it appears that primary CTT may be the procedure of choice for advanced cases of PCG in developing countries.

Following failed angle surgery investigators have compared CTT to GDD. Helmy et al. reported their results of CTT versus AGV (FP8) in children with PCG (80% parental consanguinity) who had failed previous goniotomy or trabeculotomy [56]. They included 66 eyes with 33 eyes in each group. Success rates were 97% at 1 year in both groups, which decreased to 61% in the CTT group and 66% in the AGV group at 4 years. They concluded that AGV had slightly better long-term survival; however, doing a CTT first would allow a future implant in these patients. In their series 24.2% of the eyes in AGV group had hypertensive phase. Our results show a 90% success rate in PCG with the Ahmed device following failed CTT at 1 year [57].

With regard to the surgical outcomes of GDD surgery in the developing world, Al-Mobarak et al. have reported their results with AGV in PCG and secondary glaucoma and found success rates of 54.8% at 2 years with polypropylene implant versus 90.9% with silicone implant [58]. They also found poor success rates with AGV when MMC was used (31.3% at 2 years), although there may have been selection bias between the two groups. Better outcomes have been reported by Balekudaru et al. from India with AGV (both silicone and polypropylene) in refractory pediatric glaucoma, with cumulative success rates of 97% at 1 year and 80% at 2 years [59]. Reoperations for tube-related complications were needed in 12.6% of eyes and 13% of eyes needed repeat surgery for IOP control. In their study, post-keratoplasty glaucoma was a significant risk factor for failure. In our unpublished data, the success rates with silicone AGV implantation are 91% in refractory PCG and 83% in secondary glaucomas with 4-year follow-up; our success rates with AADI are 91% at 1 year and 81% at 2 years (unpublished data). A recent study by Kaushik et al. from India has prospectively studied the outcomes of the AADI implant in refractory pediatric glaucomas and reported success rates of 81.7% at 2 years [60].

Although PCG accounts for the majority of pediatric glaucomas, glaucoma following congenital cataract surgery is the next most common childhood glaucoma and is important in the developing world [61–64]. Mandal et al. reported complete success probability with trabeculectomy with or without MMC (IOP between 6 and 21 mmHg without medication/additional surgery/sight-threatening complication) in only

37% eyes, and an additional 21% eyes had qualified success after a mean follow-up of 2 years [65]. So this group of patients has a higher risk of failure of trabeculectomy and may need GDD or other surgery.

Refractory Childhood Glaucomas in Developing Countries

Some children with glaucoma have a poor surgical prognosis with uncontrolled IOP despite surgery and/or with maximum tolerated medical treatment. The majority of studies show surgical failure is higher in eyes with greater severity of disease at presentation and increases with time. However, in the developing world, this is further compounded by the fact that most children with PCG are born to consanguineous parents [66] with more severe disease at presentation and usually with delayed diagnosis [42, 67]. Causes of late presentation in the developing world are thought to include delayed diagnosis (lack of awareness of disease) and the limited access to and unaffordability of health care. Furthermore, the majority of cases require repeat surgical intervention to control IOP. Hence, it is not surprising that children in the developing world who are blind from glaucoma have a high degree of consanguinity [56, 68]. Furthermore, the initial benefits of glaucoma surgery may be lost if there is no continuity of care for these children when they fail to attend follow-up appointments. There may also be issues with continuing medical therapy due to the inability to prescribe or afford medications [41, 43]. Therefore, the burden of blindness due to pediatric glaucoma in the developing world is significant with three-fourths of the world's blind children living in developing countries [69].

The above points are illustrated by a number of studies. Ben-Zion et al. reported the average age at diagnosis of congenital glaucoma in Ethiopia to be 3.3 years [43] compared to Western counterparts, where the average age was less than 1 year of age [6, 70]. Close to 50% of the eyes continued to have elevated IOP beyond 22 mmHg even after surgery, and the majority needed second intervention for IOP control. Also Mandal et al. showed that the success rate of CTT in advanced glaucoma (with corneal diameter $\geq$ 14 mm) was 75.5% at 1 year but decreased to 60.5% at 6 years [67]. Furthermore, Al-Hazmi reported a retrospective review of 820 eyes of 532 PCG patients with variable degrees of severity and less than 1 year of age who underwent goniotomy, trabeculotomy, or CTT with MMC [42]. The mild form of PCG had high success rates with all techniques (81–100%). Eyes with moderate glaucoma following CTT with MMC had an 80% success rate, and those with advanced PCG (corneal diameter of >14.5 mm) 70% at 1–3 years. In moderate to advanced PCG, the other techniques had poor outcomes. A study from Northern Tanzania reporting the outcomes in secondary childhood glaucoma of TSCPC (18%) compared to trabeculectomy (48%, with higher rate of complications) noted that a third of children did not return for follow-up after 1 year [71]. Distance to the hospital of greater than 100 km was a significant risk factor for trabeculectomy failure and was also speculated to contribute to late presentation and erratic follow-up. The same group published results in children with PCG comparing various surgical techniques. Their conclusion recommended that for early disease, goniotomy and trabeculotomy were options and for severe disease trabeculectomy showed moderate success. TSCPC had poor success rate of 17% at 1 year [72].

A number of strategies have been initiated in the developing world to expedite diagnosis and improve prognosis. For example, in India the training of pediatricians and neonatologists for timely referral is in progress, along with public education and childhood glaucoma awareness programs through the media. To alleviate the fear surrounding the diagnosis of glaucoma, potential blindness, and surgery, various education strategies and special support groups for children with glaucoma and families have also been implemented.

Quality of Life of Children Afflicted with Glaucoma and that of their Caregivers

Good quality of life (QoL) is a key outcome for children afflicted with glaucoma, an outcome parents want most for their children. Patient-based assessment of the impact of disease is now widely advocated [73, 74]. However, little is known about the QoL of children with congenital glaucoma in the developing world. It is necessary to assess the health-related QoL (HR-QoL) of the children with congenital glaucoma who undergo surgery and to compare it with an age-matched control group.

Children with PCG undergo surgery at a very early age and undertake multiple, frequent hospital visits over a long period of time, during which there can be uncertainty about visual prognosis. In advanced cases, children with glaucoma require several surgical interventions to achieve satisfactory IOP control, and treatment of such cases is often challenging. In a cross-sectional study at LV Prasad Eye Institute, Hyderabad, India, we assessed the HR-QoL of 121 children with PCG (mean age, 11.8 years) who underwent glaucoma surgery (mean time since surgery, 10 months–2 years) by a single surgeon and 204 age-matched healthy controls. Despite undergoing successful glaucoma surgery, children with PCG reported significantly compromised HR-QoL as compared with their typically developing peers (unpublished data). Younger children with PCG and those belonging to families from lower socioeconomic status were more likely to experience decreased HR-QoL. Ophthalmologists need to be alert to the impact of glaucoma on the QoL of children and efforts must be made to improve it.

Caregivers of children with PCG have significant emotional and psychological burdens; moderate to severe depression may be present in a third of such caregivers [75].

Recently Gothwal et al. designed, developed, and validated the first novel Caregivers Congenital Glaucoma Quality of Life (carCGQoL) questionnaire for use in helping caregivers of children with PCG. It is psychometrically robust, short (20 items), and relatively easy to complete [76]. It has excellent psychometric properties: unidimensionality, good reliability, good targeting, and well functional rating scale. In a cohort, which included 111 consecutive caregivers, Gothwal et al. demonstrated a large improvement in the caregivers QoL scores from preoperative (baseline) to the 6–8-week postoperative period [77]. In a subsequent study, the same authors demonstrated that this significant improvement in the caregivers QoL was maintained up to 2 years following surgery (unpublished data). Hence, successful PCG surgery can improve not only the child's clinical condition but also the caregiver's quality of life (see Chap. 11).

Conclusion

The prevalence and severity of childhood glaucoma are higher in developing countries and a significant number of these children are blind. Delayed diagnosis from lack of awareness (among other factors leading to late presentation), more severe disease, and poor follow-up are probably the reasons for this increased blindness rate. Surgery is the primary treatment option in managing childhood glaucomas. However, due to the severity of disease at presentation, repeat interventions with a combination of different surgical techniques may be needed to manage these complex glaucomas.

The responsibility of the surgeon does not stop with surgery and good surgical control of IOP. Visual rehabilitation is as important in the management of the disease as is IOP control. Visual rehabilitation involves correction of refractive errors, correction of opacities in the media (e.g., corneal scarring and cataract), and amblyopia therapy (when necessary). Amblyopia must also be aggressively managed to give these children the best chance for good vision in both eyes. These measures should be undertaken as early as possible.

One of the major concerns in the developing countries, despite improvement with early recognition and appropriate treatment, is the problem of poor follow-up. The emphasis on childhood glaucoma as a serious chronic dis-

ease with need for lifelong follow-up is needed. Creating awareness is the first step to tackling this problem and patient support groups play an important role in helping this cause. Furthermore, to provide appropriate care as well as to follow these children, pediatric glaucoma specialists must be appropriately trained and their skill and knowledge updated periodically.

It must be recognized that in some eyes the prognosis for long-term vision is poor regardless of the technical skill and heroic efforts of patients, caregivers, and physicians. Preservation of even limited amounts of vision in these children can improve their ability to function as adults. However, children with glaucoma resulting in moderate to severe visual loss require the combined resources of a team that extends well beyond the patient-caregiver-physician triad, including those who can provide counseling and help with training, education, integration into the schools, and later into society. These needs are great in all societies but perhaps greatest in developing countries.

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