A Baby with Primary Congenital Glaucoma Not Responding to Trabeculotomy

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CASE PRESENTATION

A 3-month-old girl was diagnosed with primary congenital glaucoma (PCG) at birth. She was born via normal vaginal delivery and was the first child of healthy non-related parents. Ocular findings at birth are listed in Table 1. She underwent trabeculotomy in the superonasal and inferotemporal quadrants of both eyes in one session. The results of examination six weeks after the operation are shown in Table 1 and Figure 1. Timolol 0.5% and dorzolamide 2% eye drops were started for both eyes and the examination was repeated 6 weeks later which showed no specific change except for intraocular pressure (IOP) reduced to 30 and 28 mmHg in the right and left eyes, respectively. There is no data on axial length, but B-scan sonography of the right eye revealed no posterior segment abnormality or retinal detachment.

- What would your management be for this patient?
- Do you recommend latanoprost or brimonidine eye drops?
- What would your next operation be for controlling IOP?
- Do you recommend trans-scleral cyclophotocoagulation?
- If corneal opacity does not improve, do you suggest corneal grafting? If so, at what age?

Table 1. Ocular findings at birth and 6 weeks after trabeculotomy

|          | Right Eye | Left Eye |
|----------|-----------|----------|
|          | IOP (mmHg) | Corneal diameter | Corneal clarity | Fundus | Refraction | IOP (mmHg) | Corneal diameter | Corneal clarity | Fundus | Refraction |
| At Birth | 45        | 15       | Cloudy Opacity | NV     | NP         | 37         | 15       | Cloudy Opacity | NV     | NP         |
| 6 wk after Tx | 35   | 15       | Cloudy Opacity | NV     | NP         | 30         | 15       | Cloudy Opacity | 80% cupping | -8.00      |

IOP, intraocular pressure; NV, not visible; NP, not possible; Tx, Trabeculotomy

George L. Spaeth, MD

The case in question is a 3-month-old girl who was noted to have cloudy corneas and markedly elevated IOP at birth. If at all possible, I would have tried hard to get an axial length at that time, would have had the child in the operating room as promptly as possible, as soon as the anesthesia people were comfortable in putting her to sleep. A trabeculotomy is a reasonable first procedure. At the time of the trabeculotomy, the surgeon

Figure 1. Megalocornea and corneal opacity in a baby with primary congenital glaucoma.
can get a good idea whether Schlemm’s canal is present and whether the surgery went as hoped. One could use probes, as apparently was done in this case, or a technique in which a suture is threaded through the Schlemm’s canal for 360 degrees, entering in one side of Schlemm’s canal and exiting in the other, and then pulled up so that the entire trabecular meshwork is torn open. Because the corneas were cloudy, a goniotomy would not be possible. If the corneas were cloudy due to elevated IOP, then they should rapidly clear. If not, either the procedure has not succeeded or some other congenital abnormality is present.

Following the trabeculotomy, assuming that it was uncomplicated and Schlemm’s canal was found and opened well into the anterior chamber, I would not have used any anti-glaucoma drops. Probably I would have checked the pressure again under anesthesia in about two weeks, and certainly if the corneas did not clear promptly and completely. At that point, IOP in this particular child would almost certainly have been found to be elevated and the next step would be the implantation of a tube shunt device. The child would then be followed primarily by measurement of axial length. If the axial length was increasing we would know that IOP was too high for that particular eye.

The eyes of infants with high IOP can deteriorate rapidly. Moving ahead promptly with successful pressure-lowering procedures is a high priority.

Ching Lin Ho, MD

This baby girl with PCG had a typical presentation at birth and an expected poor response to angle surgery, in this case trabeculotomy, as well as to topical glaucoma medications. The angle anomaly in newborn glaucoma, unlike PCG of infantile onset, is usually more severe with poorer success rates and less favourable IOP control with conventional angle surgery.

In this baby, initial trabeculotomy in both eyes failed to control IOP and clear the corneas despite addition of topical timolol and dorzolamide. Further surgery is needed for glaucoma control while topical latanoprost or another prostaglandin analogue, as well as systemic acetazolamide may be used to temporarily reduce IOP and clear the corneas until further surgery; but these are not long-term treatment options. Brimonidine eye drops are contraindicated in infants and young children due to the risk of central nervous system suppression and potentially life-threatening apnea.

With this fairly severe presentation of newborn glaucoma with opaque corneas, a combined trabeculotomy-trabeculectomy with mitomycin C as the initial surgery choice may have yielded a better outcome. If the superior conjunctiva has been spared in the initial trabeculotomy, combined surgery may still be used as the next option where expertise for glaucoma drainage devices (GDDs) is unavailable. My own immediate management for this patient after failed trabeculotomy will be a GDD followed by trans-scleral cyclophotocoagulation with transillumination later as adjuvant therapy should IOP control be unsatisfactory despite addition of topical medications. The effects of cycloablation alone in the absence of filtration in children are often inadequate and unpredictable. The resilience and recovery of aqueous producing ciliary epithelium means that the procedure often needs to be repeated multiple times with consequences of recurrent intraocular inflammation which may ultimately limit vision with resultant cataract formation and posterior segment changes.

Adequate IOP control clears the corneal haze in most cases of PCG cases with no corneal anomalies. In some cases of newborn glaucoma with central corneal hydrops, significant corneal scarring precluding clear vision may persist even after successful IOP control. In Peters anomaly with iridocorneal or keratolenticular adhesion, or certain subtypes of congenital glaucoma with CYP1B1-related mutations, corneal opacities unrelated to raised IOP will persist despite successful IOP control. These patients will need referral to a corneal surgeon with expertise in pediatric corneal grafting for further management. In the presence of dense corneal opacity in the visual axis, corneal grafting should be considered as early as possible after
successful IOP control to prevent amblyopia. However, the parents and caregivers will need to be counseled regarding the lower success rate and higher complication rate of pediatric corneal transplants. Other than IOP measurement and observation of corneal clarity, which can be difficult in the presence of corneal anomalies, adequacy of IOP control should also be assessed during follow-up with axial length and corneal diameter measurements.

Sasan Moghimi, MD

This is a six-month-old baby with PCG who has had more than 270 degrees of angle surgery three months ago which was not successful even after using timolol 0.5% and dorzolamide 2% eye drops. Classically, angle surgery, trabeculotomy or goniotomy, is the first line of treatment in PCG. I prefer to do 360 degree suture trabeculotomy if the cornea is cloudy, and canaloplasty if a catheter is available. In this procedure the entire circumference of the angle is treated using modified trabeculotomy in which a nylon or prolene suture is threaded into the Schlemm’s canal with the guide of the catheter. The suture is then tightened until it enters the anterior chamber 360° by pulling through the trabecular meshwork. This procedure has a higher success rate compared to conventional trabeculotomy or goniotomy with success rates of 87–92%.1

If more than 270° angle surgery, two sessions of angle surgery, 360° trabeculotomy, or surgery at two sites fail and pressure cannot be controlled with eye drops, we should proceed to filtering surgery. In the present case, the pressure is not controlled; right eye has corneal scar and edema, and the cornea in the left eye which seems to be the better eye, is large and edematous. The two options that we might consider are trabeculectomy with MMC and a shunt procedure. For patients less than 2-3 years of age or those with severe megalophtalmos, my first option is a drainage device. Although shunt procedures are not normally the first procedure in adult cases and are usually reserved for refractory cases or those with conjunctival scar, the success rate of trabeculectomy in pediatric cases is not as high as adults (50-70%)2 which may be the reason why it is not the first option after angle surgery in these cases. In a retrospective comparative case series, the success rate was significantly higher with Ahmed or Baerveldt implantation (87%) versus trabeculectomy with MMC (36%) in children under 2 years of age.3 Lower scleral rigidity, thinner sclera, buphthalmos with distortion of intraocular anatomy, and aggressive healing response in younger patients are some reasons for low success rates. Additionally, the postoperative management and long-term risks of bleb-associated leaks, dysesthesia, trauma and endophthalmitis are all serious considerations in children. Trabeculectomy with adjunctive mitomycin C is more likely to succeed in older phakic patients, in whom frequent postoperative follow-up is feasible.

I normally prefer valved implants in children to reduce the risk of hypotony and minimize post-operative manipulations that are often needed for nonvalved implants. To decrease the risk of flat anterior chamber after drainage device implantation I tend to inject high molecular weight viscoelastic in the anterior chamber at the end of surgery. Using these measures, the risk of hypotony is very low in my hands.

Complications of GDD implantation in children is unique such that excessive scarring in children often leads to failure of IOP control due to fibrous encapsulation around the plate. Anterior migration of the tube with growing the eye may result in tube-corneal touch, corneal edema, and tube exposure. Frequent eye rubbing may contribute to this complication, as well as externalization of the tube or plate exposure which are hard to manage.

In one study examining the long-term outcomes of Ahmed glaucoma valve (AGV) implantation in patients with PCG, the success rate at one year was 63% which decreased to 33% at 5 years.4 But after implantation of a second AGV, the 5-year success rate increased to 70%.

Although brimonidine is effective in lowering IOP, it has serious adverse effects on the central nervous system such as lethargy and drowsiness in pediatric patients. Therefore, other medications should be considered especially in
children weighing <20 kg and those younger than 6 years.

Latanoprost has become the most commonly prescribed first-line glaucoma medication in adults. It has an excellent safety profile in children but the response of PCG eyes to treatment with latanoprost is not predictable. A prospective study showed that after adding latanoprost, 32% of children eyes had IOP reduction more than 10% and only 19% had reduction exceeding 15%. Based on some recent studies, latanoprost showed a clinically relevant IOP-lowering effect in the PCG subgroup, but the magnitude of IOP-lowering effect may be less than that of the non-PCG subgroup. I usually reserve it as third glaucoma eye drop in children, but will examine the patient one month after starting it to check if it is effective.

I perform cyclophotocoagulation only for those refractory cases in which 2 or more drainage device have failed and visual potential is low. The overall success rate in pediatric glaucoma is 50% but the procedure carries risks of chronic postoperative hypotony or phthisis, uveitis, retinal detachment, cataract formation and vision loss. Although endoscopic diode laser cyclophotocoagulation, alone or combined with phacoemulsification, is one of my favorite procedures in adults with mild to moderate glaucoma, I do not believe that it is helpful in pediatric cases.

It has not been clearly established in the literature whether corneal transplantation is indicated in pediatric patients with corneal opacity due to congenital glaucoma. Corneal transplantation in patients below 4 years of age is associated with severe postoperative inflammation, high rates of secondary glaucoma and a high incidence of graft failure. This is especially true if age is less than 2 years such that almost all subjects develop graft rejection or failure.

Although some authors have reported a fair outcome for corneal grafts in their pediatric glaucoma patients and suggested that it can be considered in bilateral, visually debilitating disease, factors other than the indication for surgery can affect the graft outcome.

Regular follow-up and timely intervention can significantly alter the results. Loss of IOP control after surgery should be considered. Penetrating keratoplasty for the left eye of this patient, if IOP is well controlled, can be considered after 4-6 years if the child is cooperative and has a dedicated care-taker. Corneal transplantation in these eyes requires team work including glaucoma, anterior segment and pediatric specialists to have a 50% chance of graft clarity after surgery.

Suggested Readings

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Conflicts of Interest

None.
Consultants

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