Glaucoma in Childhood
A Review

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Introduction
Childhood or pediatric glaucomas are a heterogeneous group, comprising the true primary developmental glaucomas, associated with developmental anomalies in the eye since birth and other secondary glaucomas. Early onset primary open angle glaucoma, the so called juvenile glaucoma, as well as angle closure glaucomas both primary and secondary are also seen in the pediatric age group. Pediatric glaucoma is a potentially blinding disease accounting for about 4.2% of childhood blindness. Early diagnosis and prompt surgical intervention form the mainstay of management of this condition. The aim of treating pediatric glaucomas should be to lower the IOP permanently, to preserve visual acuity and field and help in the development of binocular single vision. Surgeries such as goniotomy, trabeculotomy and trabeculectomy formed the mainstay of surgical treatment and are still the commonest procedures performed today, albeit with modifications. Aqueous drainage devices and intra- and post-operative use of antimetabolites are the more significant recent advances in the surgical management of glaucoma. Medical management is only a temporizing measure while the child is awaiting surgery. However medical management does continue to play a role in older children with raised IOP or progression of glaucoma after failure or limited success of surgical procedures. Topical carbonic anhydrase inhibitors and prostaglandin analogues are the newer drugs tried in the management of paediatric glaucoma though beta blockers continue to be the most often prescribed. Continued long-term follow up and management of associated refractive errors and amblyopia are of prime importance. Early identification and prompt referral to ophthalmologists by paediatricians is very critical if functional, stable vision is to be achieved in the long term.

Classification and nomenclature
The term developmental glaucoma refers to glaucoma that is associated with developmental anomalies in the eye since birth. It may be associated with anomalies of the anterior chamber angle and trabecular meshwork alone or associated with other ocular or systemic/developmental anomalies. Hoskin classified developmental glaucomas based on the anatomy/structural abnormalities as:

1) Isolated trabeculodysgenesis
   A Flat iris insertion
      • Anterior insertion
      • Posterior insertion
   B Concave iris configuration

2) Iridotrabeculodysgenesis
   A anterior stromal defects
      • hypoplasia
      • hyperplasia
   B Anomalous iris vessels
      • Persistence of tunica vasculosa lentis
      • Anomalous superficial vessels
   C Structural anomalies
      • Holes
      • Colobomata
      • Aniridia

3) Corneotrabeculodysgenesis
   A Peripheral (Axenfeld anomaly)
   B mid peripheral (Reiger’s anomaly)
   C central (Peter’s anomaly)

Isolated congenital glaucoma refers to isolated trabecular maldevelopment without associated ocular or systemic anomalies. This entity has often been called primary congenital glaucoma (PCG). The terms congenital glaucoma and infantile glaucoma are often used synonymously with isolated congenital glaucoma but these could encompass other developmental glaucomas as well. Juvenile glaucoma is non-specific and is used for any type of glaucoma occurring in late childhood or teenage years for which no secondary cause is obvious. The term Juvenile Open Angle Glaucoma (JOAG) has been used to refer to chronic open angle glaucoma diagnosed between 10 to 35 years age. Thus, paediatric glaucoma includes developmental glaucoma as well as acquired glaucomas in children (secondary to tumors, inflammation, trauma) of which aphakic or pseudophakic glaucoma following cataract surgery is an important cause.

Epidemiology
Isolated congenital glaucoma occurs in about 1 in 30,000 live births. Various studies have shown varying prevalences. In India, according to a population based study in south India, the prevalence of PCG is 1 in 3,300. Prevalence has varied from 1 in 10,000-20,000 in the west to 1 in 1250 in the gypsy population of Slovakia. Pediatric glaucoma is a potentially blinding disease accounting for about 4.2% of childhood blindness. It is usually bilateral (75%) in most cases. Males constitute 65% of the cases. Most cases are sporadic. 10% show a hereditary pattern, usually autosomal recessive with variable inheritance.
Diagnosis and differential diagnosis

The features with which the child with congenital glaucoma could present include photophobia, epiphora, blepharospasm, large eyes and hazy cornea. On examination there can be raised IOP and glaucomatous discs. The differential diagnoses for each of these symptoms and signs that have to be considered before making a diagnosis of congenital glaucoma include:

**Photophobia:** corneal infection, corneal epithelial defects, foreign bodies, ocular albinism

**Epiphora:** congenital naso-lacrimal duct block, congenital ectropion

**Large cornea:** megalocornea, high myopia

**Hazy cornea:** healed keratitis, corneal dystrophies, metabolic disorders like cystinosis congenital anomalies like Peter’s anomaly, Irido-corneo-endothelial (ICE) syndrome, Congenital Hereditary Endothelial Dystrophy (CHED)

Management

Management of a case of pediatric glaucoma begins with taking a good history regarding the onset of symptoms, treatment taken, birth history (for forceps delivery) and general activity of the child. Initial examination under sedation (Syrup trichlofos sodium- 0.75ml or 75 mg/kg body weight or Syrup chloral hydrate 25-50mg/kg body weight) to record corneal diameter, tonopen IOP and fundoscopy to evaluate disc is attempted in all children. Examination under anaesthesia with the idea of proceeding to a surgical solution is the definitive management. In older children who are cooperative for office examination, best corrected visual acuity, detailed slit-lamp examination including recording of IOP with an applanation tonometer, gonioscopy and fundus examination is done. Other tests including measurement of the corneal diameter, refraction, axial length and visual field assessment if possible should be done.

Examination under anaesthesia would include performing and documenting the following procedures:

1) Refraction (retinoscopy) if corneal clarity of either eye allows it

2) Intraocular pressure (using Tonopen or Perkins tonometer)

3) Corneal diameter – white to white (using corneal calipers)

4) Fundus examination (with indirect ophthalmoscope and 20D lenses, direct ophthalmoscope)

5) Examination of the cornea under operating microscope or, preferably, a portable slit lamp to look for Descemet’s tears or Haab’s striae. Haab’s striae are horizontal in the centre of the cornea and curvilinear at the limbus. This kind of break has to be differentiated from breaks due to birth trauma which are more oblique or vertical.

6) Direct gonioscopy (using an indirect gonioscope and operating microscope or Koepppe’s lens with a hand held slit lamp) if the cornea is clear. In normal newborns, the trabecular meshwork(TM) is transparent and iris is inserted posterior to the scleral spur but in PCG the iris insertion is anterior and directly into the TM.4

7) Pachymetry to measure corneal thickness (using ultrasound pachymeter)

8) Axial length measurement (ultrasound)

9) B –scan (if corneal haze does not allow any view to fundus)

Choice of anaesthetic agent and the stage or plane of anaesthesia during which IOP is checked is important. Cyclopropane and succinyl choline tend to elevate the IOP giving higher values of IOP.7 A rapid lowering of intraocular pressure occurs particularly with halothane; halothane also lowers IOP artificially during deep anaesthesia.7 Agents achieving lighter anaesthesia like ketamine and diethylether are suitable agents.7 It is important to check the IOP immediately after induction as deeper stages of anesthesia will give falsely low pressure. IOP of more than 20 mmHg should be viewed with suspicion, but there is a wide variation expected with the type of anaesthetic agent, the age of the patient and the stage of anaesthesia.8 IOP values cannot be alone used for a diagnosis of glaucoma but must be interpreted with co-existing signs like corneal diameters, optic disc cupping, axial length etc.

The normal horizontal corneal diameter is approximately 10.5 to 11 mm. At birth normal horizontal corneal diameter is considered to be 10 mm, up to 10.5 mm at 6 months of age and less than 11 mm at 1 year of age.9 Horizontal corneal diameter of more than 12 mm by one year of age must be treated as suspicious of glaucoma. Glaucomatous disc changes are often seen early in children; conversely, enlargement of the cup often reverses following reduction of IOP. Both phenomena are probably due to the increased elasticity of the connective tissues of the optic nerve head at this age.10 Cup-disc ratio (CDR) ≥ 0.3 and presence of asymmetry in cupping between the two eyes is suggestive of congenital glaucoma. The normal disc has a round central cup with surrounding pink healthy NRR. Normal axial length is about 22 mm by one year of age. It has been shown that nearly 90% of the growth of the eye takes place by 18 months of age and from this age to 11 years of age the eye grows by a maximum of 2 mm.11 Thereafter, the axial length of the eye stabilizes.
Therapy

In PCG the mainstay of treatment is surgery. Medical therapy is used as a temporizing measure while the child is awaiting evaluation by a paediatrician or anaesthetist to assess the child’s fitness for general anaesthesia. It reduces and stabilizes the IOP to a certain extent thus facilitating surgical intervention. However these drugs do have a role in the management of glaucoma in older children.

Medical therapy

1. Beta Blockers

Timolol (0.25%), a non-selective beta-blocker, is the most commonly used medication for treatment of developmental glaucoma. It acts by reducing aqueous inflow. Side effects such as respiratory distress or apnoea, hypothermia and bradycardia are well described. Punctal occlusion should be employed after instilling timolol in the eyes to reduce systemic absorption. Selective beta blockers are safer in children. Betaxolol (0.25%) is a selective β1 blocker and causes less respiratory problems. The gel form is preferred to the purely aqueous preparation in children as gels are less absorbed systemically; however, these are not very easily available. Local side effects of beta blockers include ocular burning, stinging and dryness but are usually not so severe as to cause discontinuation.

2. Carbonic Anhydrase Inhibitors (CAI)

Oral Acetazolamide (Diamox) in the dose 5-10mg/kg body weight every 6 hours is safe and well tolerated in infants. Side effects from short-term use of carbonic anhydrase inhibitors (CAIs) in infants and young children are rare. However side effects of oral CAIs like diarrhoea, hypokalemia, hyponatremia, asthenia, hypochloremic acidosis, hypersensitivity reactions, renal stones and growth suppression could occur. Systemic side effects are less common with topical CAIs. Topical CAIs have been shown to effectively lower IOP in children with few side effects. Systemic acidosis has been reported in a neonate on topical CAI alone. Dorzolamide (2%) and Brinzolamide (1%) are carbonic anhydrase inhibitors available for topical use. Dorzolamide is preferred to brinzolamide since it causes lesser ocular burning, stinging and itching. In a randomized controlled trial of 56 patients below 6 years of age followed up over 3 months, by Ott et al, IOP reduction of about 20% was noted with dorzolamide 2% tid with no serious systemic side effects. Ocular discharge, ocular hyperemia and ocular burning caused discontinuation. Portellos et al have reported an IOP-lowering effect of 27% in a 6-month study in children. Sabri et al found greater addictive effect in children compared to adults when oral CAI were added compared to topical dorzolamide. However, others have found more or equal efficacy of topical CAIs as compared to oral CAIs. Combination of the two forms is effective in reducing IOP in children more than in adults probably due to the presence of a higher level of carbonic anhydrase receptors in pediatric ocular tissue.

3. Prostaglandin analogues

Prostaglandin (PG) analogues are found to be less effective in children compared to adults. The exact reason for the poor response is not clear. Variation in the outflow pathways in these children as well as the alteration in these pathways post-surgery are considered to be the possibilities for poor response. Late onset glaucomas with normal angles as in Sturge Weber syndrome as well as older children have been found to show better response to latanoprost. In a study by Enyedi et al, latanoprost reduced the IOP by only 14.7% in 8 eyes in contrast to adults where PG analogues reduce IOP by as much as 30%.

In another study on 57 eyes of 48 pediatric patients with uncontrolled IOP with congenital glaucoma, aphakic glaucoma, and juvenile open-angle glaucoma (JOAG), 32% had an IOP reduction of at least 10% after the addition of latanoprost, whereas only 19% had at least a 15% decrease in IOP. The median age of responders and non-responders was 11 and 5 years respectively. In addition, responders were significantly more likely to carry a diagnosis of JOAG than non-responders.

No significant side effects have been reported with PG analogues in children. A case of uveal effusion has been reported in a child with Sturge Weber syndrome on travoprost. PG analogues thus possibly have a role in the management of late onset glaucoma in older children and in secondary glaucomas like aphakic and pseudophakic glaucomas in the paediatric age group.

Alpha 2 agonists (brimonidine, apraclonidine) have significant central nervous system toxic side effects like somnolence, respiratory depression and apnoea due to the drug crossing the BBB easily in neonates and children and are best avoided.

Miotics like pilocarpine have not demonstrated much effect due to abnormal iris insertion. They are not currently recommended in the routine management of paediatric glaucoma.

Surgical Management

Goniotomy

This procedure consists of incising the obstructing structures in the angle which allows separation of the trabecular sheets and subsequent flow of aqueous into Schlemm’s canal. It is done by entering the anterior chamber and sweeping the angle with a blade under direct visualization using an indirect
gonioscope such as a Barkan lens under the operating microscope. The conjunctiva is not opened. It can be done only in clear corneas where angle visualization is possible. Shaffer\(^3\) reported a success rate of 94% with goniotomy in children aged between 1 and 24 months. The outcome was poorer in neonates less than 1 month (26%) as well as in children older than 2 years (38%). The main inhibition to using this procedure is that most paediatric glaucomas in our country are seen at an advanced stage when the cornea is hazy. Further, lack of experience with the technique means that this is a rarely used surgical option.

**Trabeculotomy**

In this procedure, a trabeculotome is inserted from the external aspect under a partial thickness scleral flap and incision into Schlemm’s canal and swept across the angle with the inner blade of the trabeculotome entering the anterior chamber. As with goniotomy the aim is to remove the obstruction to aqueous outflow across the anomalous angle. The advantage over goniotomy is that the procedure can be done irrespective of corneal clarity. However, considerable expertise by the surgeon is required. Success rates of 80-90%\(^3\) have been reported. Hyphaema, stripping of Descemet’s membrane and iridodialysis are some of the associated complications.

Ikeda et al\(^3\) in a study of 112 eyes with primary developmental glaucoma with trabeculotomy followed up over 9.5 ± 7.1 years, reported good long term success of 89.3% with trabeculotomy.

Debnath et al\(^3\) in a retrospective study compared trabeculotomy (31 eyes of 16 children) with trabeculectomy (30 eyes of 16 children) and found success of 67% and 54% respectively. The difference was not statistically significant. Trabeculectomy was associated with more complications.

Tamcelik et al\(^3\) compared viscotrabeculotomy (group1, n=58) with standard trabeculectomy (group2, n=51) in a long term study in children with at least 3 years of follow up. At the last visit, the success rates of group 1 and group 2 were 91.3% and 68.6% respectively, and the difference was statistically significant (p = 0.02). The mean number of anti-glaucoma medications used after surgery was significantly lower in the viscotrabeculectomy group They concluded that the use of viscoelastic materials during trabeculotomy may increase the success rate of the procedure by prevention and inhibition of postoperative haemorrhage, adhesion of the incision lips and fibroblastic proliferation.

**Trabeculectomy**

Trabeculectomy creates a fistula draining aqueous from the anterior chamber into the subconjunctival space and bypasses episcleral venous pressure. Thus, combining a trabeculotomy and trabeculectomy would work through separate mechanisms and produce additional lowering of IOP. Trabeculectomy in children has lower success rates due to the thick Tenon’s capsule and exaggerated healing. There are varying reports of success (50% to over 90%) of trabeculectomy in paediatric glaucoma depending on the definition of success, length of follow up and variations in surgical techniques.

### Table 1

<table>
<thead>
<tr>
<th>Study</th>
<th>Mitomycin C (mg/ml)</th>
<th>Number of eyes</th>
<th>Age</th>
<th>Success at follow up (% or success rate)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Giampi et al (2008)*</td>
<td>0.4 mg/ml</td>
<td>75</td>
<td>0-14 years</td>
<td>70% at 1 year</td>
<td>Endophthalmitis in 4/18; Flat AC; bleb leak were other complications</td>
</tr>
<tr>
<td>Odenent et al (2005)#</td>
<td>0.2 mg/ml</td>
<td>25</td>
<td>48-26 months</td>
<td>70% at 1 year</td>
<td>No difference in success or complications with two concentrations of mitomycin</td>
</tr>
<tr>
<td>Eirlich et al (2005)$</td>
<td>0.2-0.4 mg/ml</td>
<td>19</td>
<td>44±29 months</td>
<td>70% at 1 year</td>
<td>No bleb-related complications</td>
</tr>
<tr>
<td>Mandal et al (1997)**</td>
<td>0.4 mg/ml</td>
<td>8</td>
<td>8 months to 18 yr</td>
<td>75% at 1 year</td>
<td>No bleb-related complications</td>
</tr>
<tr>
<td>Sasseau et al (1995)##</td>
<td>0.2 mg/ml</td>
<td>70</td>
<td>1 year</td>
<td>75% at 1 year</td>
<td>No bleb-related complications</td>
</tr>
</tbody>
</table>

# JPOS 2005; 42;97-102
$ BJO 2005; 89(2) 165-168
**Ophthalmology 1997;104:996-1001
##JGlaucoma 1995;4:151-157

Beauchamp et al\(^3\) (n=26) reported overall success of 50% in controlling IOP. Burke et al\(^3\) (n= 21) reported 87% success, Rao et al\(^3\) (n=25) 75% success and Debnath et al\(^3\) (n=30) 54% success. Fulcher et al\(^3\) reported success of 92.3% with trabeculectomy in primary infantile glaucoma. Detry Morel
et al45 (n=46) reported success rates of almost 90% following trabeculectomy as the initial procedure of choice.

There are numerous studies looking at the safety and efficacy of antimetabolites in congenital glaucoma. Some studies have found more bleb related complications while other studies have not. Table 1 summarises some relevant studies of trabeculectomy with mitomycin in paediatric glaucomas.

Rodrigues et al43 in a retrospective study of 91 patients compared success of trabeculectomy without (61) and with (30) mitomycin 0.3mg/ml for 3-4 minutes and found no difference (p = 0.97) in the success. Both age and the presence of previous trabeculotomy did not influence the success of trabeculectomy through time. The complication rate (shallow anterior chamber and hypotony) was higher among the patients who received mitomycin C (p = 0.01). There were no cases of bleb related infection. Yalvac et al44 also did not find any difference in the outcome with and without mitomycin.

Mandal et al45 in a retrospective review of refractory congenital glaucomas (n=19) undergoing mitomycin augmented trabeculectomy( 0.4 mg/ml for 3 minutes) found good success(94.74%) with no case of bleb related complications(follow up 19.52 ± 2.65 months) and considered it a viable option in failed conventional trabeculectomy.

Al-Hazmi et al46 noted that bleb related complications with the use of mitomycin C increased with age. The complication rate was 0% for eyes younger than 6 months of age to 37% for eyes after 4 yearsof age and hence additional complications could be anticipated on prolonged follow up. Children less than 2 years are probably less susceptible to the effects of MMC due to the large population of multiplying cells.

Low et al47 in a retrospective case review of 30 patients, evaluated childhood filtration surgery using releasable sutures, anti-metabolites, and bleb-needling with 5-fluorouracil (SFU). Patients had undergone either trabeculectomy or combined trabeculotomy-trabeculectomy using anti-metabolites, releasable sutures, and bleb modification was performed. There were no major or sight-threatening complications in their series; no eyes developed cystic avascular blebs or bleb-related infections.

Snir etal48 have used 5-FU alone as well as with mitomycin and found no significant drug induced complications. Up to 6 injections of 5-FU (5mg) were used.

Thus, additional use of mitomycin (0.2-0.4mg/ml for 3-4 minutes) may be beneficial in paediatric glaucomas. However complications in the form of hypotony and shallow AC are commoner with mitomycin, though the risk of bleb related endophthalmitis is not increased in children, unlike in adults. A well planned RCT would help quantify the risks and benefits.

**Trabeculectomy and trabeculotomy**

Good success has been reported with combined trabeculectomy and trabeculotomy, with or without antimetabolotes. Mandal et al49,50 and Elder et al51 have reported high success of more than 90% with the combined procedure.

Mullaney et al52 in a retrospective review of 60 patients (100 eyes) with congenital glaucoma, reported a 78% success rate for combined trabeculectomy and trabeculotomy for PCG. The success dropped to 45% when associated anterior segment anomalies were present.

Al-Hazmi et al53 in a retrospective review of 532 paediatric glaucoma patients (820 eyes) compared the outcomes of goniotomy, trabeculotomy, or combined trabeculotomy-trabeculectomy with mitomycin C in children less than 1 year old with minimum 1 year post-operative follow up. They found that all three surgical procedures resulted in high success rates of 81–100% for the mild form of PCG. Eyes classified with moderate glaucoma had a 13%, 40%, and 80% success rate respectively for goniotomy, trabeculotomy, and combined trabeculotomy-trabeculectomy with mitomycin C. The success rate for severe PCG was 10% and 70% for trabeculotomy and combined surgery respectively. Combined trabeculotomy-trabeculectomy with mitomycin C (CTTM) gave the best results for moderate and severe cases of PCG. Complications were most common when the CTTM technique was undertaken (31/73 cases).

Agarwal et al54 (n=30) compared mitomycin C 0.2 mg/ml vs mitomycin C 0.4 mg/ml and found no difference in the success rates of combined trabeculectomy and trabeculotomy with the two concentrations.

**Aqueous drainage devices**

Aqueous drainage devices are usually used in refractory glaucoma in patients in whom the other above mentioned surgeries have failed. Molteno implant, Barveldt glaucoma implant and the Ahmed Glaucoma Valve (AGV) have been used in paediatric glaucoma. Adult size implants and even double plate implants are usually suitable in paediatric patients as well due to the larger size of the globe in these children with glaucoma. Smaller sized (paediatric AGV implants) may be indicated in microphthalmic eyes. Tube-cornea touch with consequent corneal decompensation is the most common complication (5.7- 20%).55 requiring surgical repositioning of the tube. Changing dimensions due to growth and expansion of the globe and limbus leads
to angling of the tube towards the cornea. More posterior placement of the tube, 1-2 mm behind the limbus under a partial thickness scleral flap has been recommended to avoid this problem.55

Success rates of 44-100%56 have been reported depending on the age, diagnosis, definition of success, length of follow up, type of implant.

Table 2 shows details of some of the studies with aqueous drainage devices in children

Table 2

<table>
<thead>
<tr>
<th>Study</th>
<th>Implant</th>
<th>Number of eyes</th>
<th>Age (years)</th>
<th>Success at average follow up survival (months)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Khan et al (2009)*</td>
<td>AGV</td>
<td>42</td>
<td>≤2</td>
<td>73.8% at 12 months, 63.3% at 24 months</td>
<td>Tube repositioning in 26.2%; RDS, endophthalmitis other complications</td>
</tr>
<tr>
<td>Schotthoefer et al (2008)**</td>
<td>BGI</td>
<td>31 (2 congenital glaucomas)</td>
<td>≤2</td>
<td>92% at 12 months, 42% at 10 yrs, 90% at 12 months</td>
<td>Re-surgery needed in 50% cases</td>
</tr>
<tr>
<td>Budenz et al (2004)*</td>
<td>AGV</td>
<td>62</td>
<td>≤18 yrs</td>
<td>80% at 12 months, 67% at 24 months</td>
<td></td>
</tr>
<tr>
<td>Beck et al (2003)**</td>
<td>BGI</td>
<td>48</td>
<td>≤2</td>
<td>87% at 12 months, 53% at 72 months</td>
<td>45% needed re-surgery. Tube repositioning commonest indication</td>
</tr>
<tr>
<td>Dodds et al (2001)**</td>
<td>AGV</td>
<td>35</td>
<td>≤2</td>
<td>68% at 24 months</td>
<td></td>
</tr>
<tr>
<td>Hill et al (1991)**</td>
<td>SP</td>
<td>65</td>
<td>≤21 yrs</td>
<td>62% at 22 months</td>
<td></td>
</tr>
<tr>
<td>Munoz et al (1991)**</td>
<td>Moleto</td>
<td>35</td>
<td>≤12 yrs</td>
<td>68% at 18 months</td>
<td></td>
</tr>
</tbody>
</table>

AGV: Ahmed Glaucoma valve implant
BGI: Baerveldt Glaucoma Implant
SP: Single plate
DP: Double plate
*BJO2009;93 (6) 795-798
#JAAPPOS 2008;12(1) 33-39
S Ophthalmology 2004;111(12):2204-2210
** AJQ 2003;136:994-1000
*** JPOS 1991; 28:68-72

Khan et al** in a retrospective analysis compared the survival rates of polypropylene AGV (31) and silicone (11) AGV implantation during the first 2 years of life in children with 2 years’ postoperative follow-up. They found better average survival (p = 0.001) in the silicone group (23.36 months; CI 20.16 to 24.00 months) compared to the polypropylene group (19.10 months CI 16.1 to 22.12 months). Cumulative probabilities of survival at 2 years by Kaplan to Meier analysis were 90.9% and 54.8% respectively (p = 0.001). All eyes implanted with silicone AGVs had the diagnosis of congenital glaucoma, which was independently associated with better 2-year survival. They concluded that AGV implantation in other paediatric glaucoma diagnoses is needed to determine whether or not silicone AGVs independently have a better survival after implantation in the first 2 years of life.

Cyclodestructive surgeries

Cyclodestructive procedures like cyclocryotherapy and trans scleral cyclophotocoagulation are currently considered only for symptomatic children (pain) with refractory glaucomas with no visual potential. Complications like hypotony and phthisis leading to poor cosmetic outcome need to be kept in mind.

Follow up

Follow up is an essential part of glaucoma management and is particularly important in children. The treating surgeon needs to be very clear in his mind that the aim of treatment is to provide the child with good vision that will last out its lifetime, not just control of IOP. This means that refractive errors, media opacities and other factors affecting visual development must be actively looked for, and monitoring of vision to detect amblyopia is mandatory. Repeated examinations under sedation (or EUA if required) initially monthly and if stable after 5-6 months, at less frequent intervals of 2-3 months and later every 6 months are usually needed. All the parameters as detailed earlier for EUA must be recorded every time. Additionally, visual assessment by Sheridan’s or preferential looking tests as well as ocular alignment and media clarity must be specifically assessed. High IOP and increased disc cupping indicate uncontrolled glaucoma. Enlarging corneal diameters and axial length with increasingly myopic refractions are indirect clues to inadequate IOP control. Re-surgery may be needed in as many as 20% cases.55 Appropriate refractive correction and occlusion therapy if needed for amblyopia are very essential at every stage.

Conclusion

Goniotomy and trabeculotomy have yielded good results. A combined trabeculotomy and trabeculectomy procedure has the best success. All surgeries have the best outcome in isolated congenital glaucoma. Success is lesser in other types of developmental glaucomas. Primary trabeculectomy alone without trabeculotomy is not usually preferred. Mitomycin used intraoperatively has questionable benefits with some studies reporting increased incidence of bleb related complications. Aqueous drainage devices are useful for refractory paediatric glaucomas. Silicone implants (AGV) may
have better success, though more data is needed to confirm this. Medical management is restricted to patients awaiting surgery. However topical CA inhibitors and PG analogues may have a role in older children or as an adjunct to partially successful surgery.

Early diagnosis, prompt surgical treatment and prolonged life long follow up with amblyopia management is necessary in all cases for optimal long term visual outcomes.

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