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Case Report

Mini Review on Ahmed Glaucoma Valve Implants/Tube Shunts in Treatment of Primary Congenital Glaucoma and Refractory Pediatric Glaucoma

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Abstract

Primary Congenital Glaucoma (PCG) contributes to marked visual morbidity in developing countries if not treated properly on time. There are various treatment modalities available to control the Intraocular Pressure (IOP) in PCG. Goniotomy, trabeculotomy and trabeculectomy though considered as better surgical methods to control the IOP in PCG, over the years, studies reported failure of these procedures with uncontrolled IOP in children. Glaucoma drainage devises (GDD) are the treatment of choice for such cases of PCG and those with refractory pediatric glaucomas. Various GDD's available in the management of PCG are Baerveldt, Molteno, Krupin and Ahmed implants. Of which, Ahmed glaucoma valve devises are commonly used by many pediatric ophthalmologist in the world. This mini review briefly discusses the risks and benefits of the Ahmed glaucoma valve implants in PCG and refractory pediatric glaucomas.

Keywords: Ahmed implant; Bleb; Glaucoma; Goniotomy; Trabeculectomy; Tube shunt

Abbreviations

AGV: Ahmed Glaucoma Valve; EUA: Examination Under Anaesthesia; GDD: Glaucoma Draining Device; IOP: Intraocular Pressure; MMC: Mitomycin C; OCT: Optical Coherence Tomography; PCG:Primary Congenital Glaucoma; RD: Retinal Detachment; RNFL: Retinal Nerve Fiber Layers; VA: Visual Acuity

Introduction

Primary Congenital Glaucoma (PCG) is a challenging condition to the pediatric ophthalmologists and glaucoma specialists due to the complexity of angle anatomy and the pathophysiology [1]. It is a form of developmental glaucoma that begins within the first five years of life and is characterized by isolated trabecular meshwork dysgenesis but not associated with any development ocular anomalies or ocular diseases that can raise the intraocular pressure (IOP). Because of the abnormal drainage of fluid from the eye, the accumulated fluid builds up the pressure in the eye leading to big eyes, cloudy eyes, epiphora, photosensitivity and decreased vision. It is more common among the population where consanguineous marriages are prevalent such as Middle Eastern countries and Southern India than the Northern America. The most common culprit gene in the aetiology of PCG is CYP1B1 gene. It is an autosomal recessive gene with mutations that show variable expressivity and almost complete penetrance. The diagnostic and therapeutic challenges pose a threat to the long-term vision in children and can even lead to blindness. Management of raised IOP in PCG is primarily by surgery that aims to open the draining system either from inside (goniotomy), outside (trabeculotomy, viscocanalostomy) or by making an alternate new drainage pathway (trabeculectomy, deep sclerectomy, implantation of devices) [2-5]. In this mini review, we describe briefly the role of Ahmed glaucoma valve (AGV) implants in the treatment of PCG and refractory pediatric glaucoma.

Clinical Presentation

If an infant or a child presents with the following symptoms, consider the diagnosis of glaucoma.

Common symptoms

Epiphora (tearing) enlarged eye (buphthalmos), blepharospasm (frequent eyelid blinking), light sensitivity, facial birthmarks, lack of eye contact, and corneal opacity [1].

Common signs

Increased IOP, increased horizontal corneal diameter, corneal edema, corneal haze, Haab'sstriae (breaks in the descemets membrane), deep anterior chamber, anterior iris insertion (on gonioscopy), anomalous iris vessels (rarely), enlarged cup with optic atrophy, thin sclera, and atrophic iris [1].

Diagnostic options

Examination underAnaesthesia(EUA) is crucial in the examination, to confirm the diagnosis and evaluate the response to the treatment. Common parameters to check upon EUA are the IOP, pupillary reflex, corneal diameter (vertical & horizontal), retinoscopy, keratometry, axial length, gonioscopy (angle structures), and fundus photography (Retcam) [1]. Advanced diagnostic modalities such as Optical Coherence Tomography (OCT) of the anterior segment (to evaluate the cornea & the angle) and optic nerve (to measure the retinal nerve fiber layer thickness [RNFL]) & macula are performed by many pediatric ophthalmologists to study the ocular anatomy both pre and post treatment to monitor the response to various therapeutic methods. B-scan Ultrasonogramis performed in cases with hazy view

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of the fundus to rule out the posterior segment pathologies [1].

Treatment

The goal of treatment is to keep the IOP within the normal limits, prevent corneal opacification, & damage to the retinal nerve fiber layers of the optic nerve (causes glaucomatous optic atrophy), preserve the visual acuity and prevent the loss of light perception (blindness) from PCG. Treatment options can be medical and surgical. Medical option gives temporary relief and controls the IOP prior to surgery or postoperatively to maintain the IOP within normal limits. Laser procedure has not much role in the treatment of PCG and is not considered as the best treatment option. Treatment by surgical methods is the best, effective and definitive option in the management of PCG. Pharmacological treatment with topical medicines has many challenges such as availability of suitable eye drop for the infants, efficacy, side-effects, compliance, and achieving the success. Surgical options in the treatment of PCG are goniotomy (draining from inside), trabeculotomy (draining from outside), trabeculectomy (with or without antifibrotic agents like mitomycin C) &Glaucoma Drainage Devices (GDD) (creating alternate drainage pathway), cyclodestructiveprocedures (cyclophotocoagulation) and combined surgeries (example-trabeculotomy with trabeculectomy ± MMC) [2-5]. GDD is considered in cases of refractory glaucoma (not responding to topical medications), prior failed goniotomy or trabeculectomy, significant conjunctival scar barring the filtration surgeries, poor success by prior filtration surgeries and buphthalmic eyes with thin sclera [2-7]. The GDD's can be either open tubed, non-restrictive devices such as Baerveldt&Molteno implants or valved, flow-restrictive devices such as Krupin& Ahmed implants [4-10]. Ahmed glaucoma valve implants are more commonly used than the other types of implants. The material used for making the Ahmed implants is either polypropylene or silicone (best) [11,12]. The size of the implant varies (96 to 250mm^2) with the size of the infant's eye. The common size of the silicone implants in children is S3 or FP8 and in adults is S2 or FP7 and the size of polypropylene is S3 in kids. It is controversial about the benefits of MitomycinC (MMC) while performing the GDD surgery [3-5,11]. MMC helps in preventing the closure of drainage pathway openings. Studies have reported that by using MMC there is development of fibrosis which encapsulates the AGV leading to failure of the valve function [4,5]. Complications encountered with the usage of GDD can be tube related or surgical [2-7,10,13-15]. Tube related complications are obstruction of the tube tip, tube exposure, tube malpositioning, and tube migration [2-7]. Others are transient hyphema, inflammation, corneal oedema, shallowing of the anterior chamber, choroidal effusion, exudative choroidal detachment with dropout of the plate, Retinal Detachment (RD), suprachoroidalhaemorrhage, fibrosis and encapsulation of the valve, vitreous haemorrhage, recurrent or persistent iritis, pupillary membrane, eye motility disorder, bleb failure, endophthalmitis (due to tube exposure) and phthisis bulbi [2-7,13-15].

Evidence that GDD's are successful in PCG and refractory pediatric glaucoma's

Success after GDD implantation is reported by many authors as control of IOP within the normal range postoperatively with or without minimal usage of topical medication, absence of complications or loss of visual acuity & light perception and no further requirement of additional glaucoma surgeries [2-7,10]. Failure is considered if there is need for further glaucoma surgeries and loss of light perception. In a study by Daniel MC et al, GDD's were implanted within the first two years of life on 60 eyes of 43 children (median age 11.5 months) with childhood glaucoma [6]. The success rate at a median follow-up of 48 months was 93% in PCG group, 59% in patients with glaucoma after cataract surgery and 59% in patients with anterior segment dysgenesis group. They reported that GDD implantation was safe and effective in children with less postoperative complications and less requirement of examinations under general anaesthesia[6]. Ou Y et al did a chart review on 19 patients (n=30 eyes) with PCG who underwent AGV implantation [15]. They reported that patients after AGV implantation there was a statistically significant ($p<0.001$) decrease in mean pre-operative IOP from 28.4mm Hg to 16.6 at 12 months and 17.7 at 60 months post-operatively. Very few patients required topical medications to control the IOP postoperatively. The success rate after AGV implantation their study was 63% at 1 year and 33% at 5 years. Post-surgical complications noted were tube-endothelial touch that required tube trimming in 4 eyes (13%), and tube exposure in 2 eyes (7%) that required repeat scleral patch graft. Female gender and Hispanic ethnicity were the main risk factors contributing to the failure of AGV implants in PCG in this study. Six patients (n=10 eyes, 33%) required second AGV implantation due to primary AGV implantation failure. Following second AGV implantation, the success rate was 86% at 1 year and 69% at 5 years of follow-up. Approximately 3 eyes (10%) required third AGV implantation in this study with good control of IOP in 66% (n=2 eyes) at 48 months of follow-up. They summarized that there was moderate success rate with good IOP control in patients with PCG following AGV implantation [15]. Interestingly in another study by Pakravan M et al, the success rate after Ahmed glaucoma implantation was also moderate in refractory primary congenital glaucoma patients (n=62 eyes, group 1) than those with aphakic glaucoma patients (n=33 eyes, group 2) [2]. At 1-year follow-up the cumulative probability of success was 90% in both groups whereas at 5 years of follow-up the success rate was 52.5% in group1 and 71.5% in group 2. They noted more tube related complications in patients with refractory PCG than those with aphakic glaucoma [2]. Senthil S et al reported the reasonably good outcomes following implantation of silicon Ahmed valve devices in refractory pediatric glaucoma patients (n=65 eyes) [5]. Their success rate was 91% in only PCG cases (n=24 eyes) and 83% in all pediatric glaucoma cases (n=41) at 1 and 4 year follow-ups. The post-operative tube related complications were similar in both the groups. They concluded that though AGV implants have good success rate in refractory PCG, failure following GDD was attributed to the number of prior intraocular surgeries [5]. Baerveldt implants were also used safely in the treatment of primary congenital and refractory childhood glaucoma patients [8-10]. Rolim de Moura C et al performed a retrospective noncomparative study on 48 patients (mean age of 4.1 years) of pediatric glaucoma that are treated by the Baerveldt implants [9]. Though the cumulative success rate was 95% at 6 months, it dropped to 58% at 48 months. At a mean follow-up of 5.6 years following Baerveldt implant surgery, approximately 30% had failed implants (11/48 eyes) due to post-surgical complications such as uncontrolled IOP, RD, and no light perception. They concluded that in patients with pediatric glaucoma refractory to medical therapy, Baerveldt implant can be safe and effective treatment method [9]. Among the AGV implants, with silicon made implants

there was good control of IOP and lesser complications than the polypropylene made AGV's [11]. In a retrospective chart review by El Sayed Y et al, out of 50 eyes (n=33 patients, mean age 34.6 months), 25 eyes with pediatric glaucoma were treated by silicon AGV's and 25 with the polypropylene AVG's. At all follow-up visits, the mean postoperative IOP was lower in patients treated by silicon AGV's than polypropylenes AGV's. The cumulative probability of survival at the 2 years was 80% in silicon group and 56% in the polypropylene group. The mean survival time with silicone implants was 22.84 months, whereas with polypropylene implants was 18.36 months (P=0.001). There was no statistically significant difference in the rate of complications with either material. They concluded in children younger than 10 years of age with pediatric glaucoma, complete success with good IOP control was more in the silicon AGV group eyes than the polypropylene AGV group eyes [11]. Almost similar conclusions were reported by Khan AO et al when they compared silicone AGV's (n=11 eyes, FP7) with the polypropylene AGV's (N=31 eyes, 6S1, 25 S2) that were implanted in the first 2 years of life [12]. The average survival was longer with silicone type (23.36 months) than the polypropylene implants (19.10 months) with maintenance of IOP ≤ 22mmHg with or without topical medications and without significant complications. The cumulative probability of survival at 2 years was 90.9% in silicon group and 54.8% polypropylene group (p=0.001). They concluded that though the silicone AGV survived longer than polypropylene AVG's, silicone AGV's had better survival in patients with congenital glaucoma than patients with other pediatric glaucoma diagnoses [12].

Conclusion

Preservation and restoration of vision is important in children with pediatric glaucoma's. Management of PCG is always challenging despite of availability multiple treatment options that are proven to be safe and effective. Ahmed glaucoma implants are considered as a safe treatment modality with success rate of 50-95% in the treatment of primary congenital glaucoma and refractory pediatric glaucoma's. Like any other surgical procedures for PCG, the risks, benefits and efficacy of AGV implants should be weighed for each patient prior to the treatment.

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