Clinical findings following Ahmed Glaucoma Valve™ implantation in pediatric glaucoma

Amir Pirouzian1
Joseph L Demer2

1Department of Ophthalmology, San Diego Children’s Hospital, San Diego, UCSD, San Diego, CA, USA; 2Department of Ophthalmology, Jules Stein Eye Institute, UCLA, Los Angeles, CA, USA

Purpose: To describe clinical findings after Ahmed valve drainage implantation in children.
Design: All records in one practice were reviewed to identify and describe clinical findings in all children who had undergone Ahmed Glaucoma Valve™ S2 model insertion for uncontrolled primary or secondary glaucoma.
Results: A total of 6 patients were identified, ranging in age from 2–15 years. Mean follow-up time averaged from 2–5 years from the time of tube insertion. Three patients exhibited pupillary peaking towards the tube of the valve. All patients required additional surgery or additional medications to control intraocular pressure. Lenticular opacification near the tube site developed in one patient. Gradual tube extrusion was also noted in another two patients.
Conclusion: Multiple clinical events follow the Ahmed valve insertion in children. Pupillary irregularity is the most commonly noted event in this series. To avoid or reduce the risk of this complication, additional or modification of surgical procedures could be considered. The mechanism of such occurrence will further be discussed.

Keywords: Ahmed Glaucoma Valve, children, pediatric glaucoma

Introduction
Management of pediatric glaucoma is a challenging and complicated process; significant devastating post operative complications may occur. Primary congenital pediatric glaucoma due to its aggressive nature has been traditionally controlled surgically by goniotomy, trabeculotomy, or trabeculectomy in conjunction with/without antimetabolite application (Mandal et al 1997; Beck et al 1998; Mullaney et al 1999). The combination of trabeculotomy/trabeculectomies has been reported to have a good success rate in the latest studies (Mullaney et al 1999). In primary congenital glaucoma, initial goniotomy or trabeculotomy is generally the preferred surgical approach. The application of glaucoma drainage devices with the introduction of Ahmed Glaucoma Valve™ (AGV) (New World Medical, Inc., Rancho Cucamonga, CA) has been gaining significant momentum in the surgical armamentarium of glaucoma management in selective clinical cases (Da Mata et al 1999; Morad et al 2003). There have been numerous publications stating the preferred use of Ahmed glaucoma drainage devices in comparison to other procedures such as trabeculotomy and/or filtering procedures under certain clinical indications (Englert et al 1997, 1999; Mandal et al 1997). Much of the recent reported literature has focused on the short and long term efficacy of AGV insertions (Coleman et al 1997; Morad et al 2003). In this study we have attempted to review and state some of the unreported incidents in association with the AGV insertion surgeries in pediatric patients and in the management of pediatric glaucoma.

Methods
Six consecutive patients who had undergone AGV implantation for control of glaucoma with or without previous glaucoma surgeries and with active follow up in both
glaucoma and pediatric divisions were identified from review of the total patients’ charts of the practice in the past one year. Four female and two male patients were identified. Complete review of each patient’s ophthalmologic chart was performed, including list of all previous ocular surgeries. Written informed consent according to a protocol conforming to the Declaration of Helsinki and approved by the Human Subject Protection Committee at the University of California, Los Angeles was waived. Institutional Review Board waiver was obtained and HIPPA regulations were followed for all the patients involved in this study.

Patients in this study had all undergone AGV placement by one surgeon (A.C) except one. Surgeries were based on limbal based incision and one step valve insertion. Post-operative IOP measurement were done by Tonopen (Mentor, Norwell, Massachusetts) or hand-held Goldmann applanation tonometer or Perkins applanation pneumotonometer. Second or third AGV insertions in the same eye were performed in different quadrant with the same technique if needed.

Full size AGV implant with dimension of $16 \times 13 \text{ mm}^2$ (model S-2, 185 mm$^2$) was used in all of the operated cases except in case 5 (model S-3, 96 mm$^2$).

Results

Case 1

An 8-year-old female with history of congenital glaucoma and multiple surgeries (Tables 1 and 2) including AGV insertion twice done OU by age of two had a penetrating keratoplasty (PKP) secondary to corneal decompensation from the tube to the cornea touch. Bilateral cataract extraction, intraocular lens implantation and anterior vitrectomy (CE, IOL, AV) at age 6 with an exchange of AGV implant OD at age 8 due to nonfunctioning AGV were performed. Patient’s final best corrected visual acuity (VA-snellen) is 20/40 OD and 20/100 OS. The patient’s refraction in spherical equivalent is $-0.50D$ OD and $-3.25D$ OS. Final axial lengths are 25.80 mm OD and 24.99 mm OS. Anterior segment examination revealed presence of two well positioned-tubes OD (at 10 and 8 clock-hour) with healthy clear corneal graft, peripheral iridectomy superotemporally and two well placed-tubes OS (at 11 and 1 clock-hour) with significant correctopia toward the 11-clock valve. Gonioscopy examination revealed no peripheral anterior synechia (PAS) surrounding an anteriorly-entered tube. The optic nerves have 0.50 round sharp cuppings with healthy neuroretinal rims. Intraocular pressures are in mid-teens on betoptic-S (Betaxolol, Alcon Lab) BID.

Case 2

A 2½-year-old female with a history of congenital glaucoma and with early insertions of AGV tubes supero-temporally both in OD and OS in her first month underwent second AGV insertion in her left eye before six-months of age. PKP surgery for the left eye was performed at age of 1½ secondary to persistent corneal edema. Cataract extraction and IOL in the right eye was followed a few months later. Significant left iris dyscoria was noted with pupillary opening retracted towards the superior tubes. Progressive left eye cataract formation led to subsequent cataract extraction and IOL-placement surgeries. Anterior segment examination for the right eye revealed correctopia with the tip of the tube to be drawn back to the edge of corneal limbus in the right eye. The supero-temporal left Ahmed tube was retracted to outside of anterior chamber and laid on the scleral surface. The supero-nasal tube was still 1 mm into the anterior chamber. Gonioscopy revealed no PAS formation surrounding anteriorly entered-tubes. The intraocular pressures on Betoptic-S are around low teens with stable optic nerve cups of 0.2 OD/OS. The patient has good fixation in the right eye and no fixation in the left eye. The corneal graft on the left eye is clear. The axial lengths of 23.20 mm OD and 22.50 mm OS have been stable during the past one year’s examinations. Follow-up period from the cataract surgery was one year.

Case 3

A 15½-year-old female with history of juvenile rheumatoid arthritis (JRA), secondary iridocyclitis and steroid induced

<table>
<thead>
<tr>
<th>Table 1 Post-operative anterior segment findings in patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
</tr>
<tr>
<td>------</td>
</tr>
<tr>
<td>Patient 1</td>
</tr>
<tr>
<td>Patient 2</td>
</tr>
<tr>
<td>Patient 3</td>
</tr>
<tr>
<td>Patient 4</td>
</tr>
<tr>
<td>Patient 5</td>
</tr>
<tr>
<td>Patient 6</td>
</tr>
</tbody>
</table>
Clinical findings following Ahmed Glaucoma Valve \textsuperscript{TM} implantation

Table 2 Surgical data of our patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>Visual acuity (cc)</th>
<th>PKP</th>
<th>CE/IOL</th>
<th>Follow up complication to AGV</th>
<th>IOP control</th>
<th>Prior operations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>20/40 OD 20/100 OS</td>
<td>+</td>
<td>+</td>
<td>Correctopia</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Patient 2</td>
<td>Fix OD/No fix OS</td>
<td>+</td>
<td>+</td>
<td>Correctopia</td>
<td>Yes</td>
<td>None</td>
</tr>
<tr>
<td>Patient 3</td>
<td>20/30 OD &amp; 20/40 OS</td>
<td>-</td>
<td>-</td>
<td>None</td>
<td>Yes</td>
<td>None</td>
</tr>
<tr>
<td>Patient 4</td>
<td>20/50 OD &amp; 20/50 OS</td>
<td>-</td>
<td>-</td>
<td>Correctopia</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Patient 5</td>
<td>20/25 OD &amp; 20/40 OS</td>
<td>-</td>
<td>-</td>
<td>Correctopia</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Patient 6</td>
<td>20/160 OD and HM OS</td>
<td>-</td>
<td>+</td>
<td>Oval shaped</td>
<td>Yes</td>
<td>Yes</td>
</tr>
</tbody>
</table>

Abbreviations: CE/IOL, cataract extraction and intraocular lens implant; IOP, intraocular pressure; PKP, penetrating keratoplasty.

glaucoma had undergone bilateral trabeculectomies with Mitomycin-C OU and cataract extraction OU around age of seven. Subsequent trabeculectomy OS was done around age of 10 and AGV placement was done by age of 11 in the left eye due to an uncontrolled IOP. In addition, surgical peripheral iridectomies supero-nasally and supero-temporally had also been performed. No pupil irregularity was observed. The IOPs have been under control in both eyes on Betoptic-S. No intra-ocular inflammation was noted and JRA was under control on Methotrexate (Rheumatrex, Ben Venue Laboratories, Bedford, OH). Cupping of optic nerves is stable at 0.5 OD and 0.4 OS. Axial lengths of 22.45 mm OD and 23.12 mm OS have been noted. Follow-up period from the last tube insertion was about 4 years.

Case 4
The patient is a 10-year-old myopic male with a history of prematurity and intracranial bleed with placement of VP shunts. The patient’s past ocular history is significant for congenital glaucoma and regressed ROP stage II/zone II without surgical treatment OU. The patient had undergone 180-degree goniotomy OD and 120-deg goniotomy OS around 2 months of age and a subsequent repeat goniotomy at age two (Tables 1 and 2). The patient underwent an AGV tube insertion for uncontrolled glaucoma at age 7 OD. The patient’s intraocular pressure has remained under control in mid 10’s on Betoptic-S 0.25% BID. The cup to disk ratio (C/D) is 0.9 for both OD/OS. The patient displayed iris peaking at 10 o’clock towards the tip of a well-positioned tube. No PAS was noted on gonioscopy examination. The patient’s refraction in spherical equivalent was −5.00D OD and −7.00D OS. Axial lengths of average 23.45 mm OD and 23.76 mm OS are noted.

Case 5
The patient was a 6-year-old male with history of Sturge-Weber syndrome (SWS) with secondary left eye open angle glaucoma (Tables 1 and 2). The patient had diffuse choroidal hemangioma OS. The patient’s seizures were controlled with tegretol (carbamazepine, Novartis Pharmaceuticals, East Hanover, NJ). The patient had undergone AGV drainage implant at age 2½ year in the left eye. During an examination under anesthesia, the intraocular pressures were measured to be in mid 10’s and a significant left correctopia was noted toward the anteriorly positioned AGV tube at the two o’clock position in the left eye. The cycloplegic refraction shows minimal myopia OU. The patient’s visual acuity is 20/25 OD and 20/40 OS (Allen-test). Left periorbital port-wine stain was also noted. The AGV tube was noted to be 2 mm within anterior chamber from the corneal limbus. No PAS is noted surrounding the anteriorly entered tube on gonioscopy examination. The cup to disk ratios is 0.2 OD and 0.7 OS.

Case 6
The 9-year-old female presented with history of severe congenital glaucoma OU and progressive glaucomatous optic neuropathy. The patient had failed goniotomy × 1 OU, trabeculotomy OU × 1 followed by trabeculectomy (with MMC OD) before age of one and placement of AGV OU × 1 around age of one. A repeat AGV insertion in the left eye was performed two months later. The extrusion of AGV tubes noted a few months after their initial placement OU. Subsequent removal of AGV was performed OU due to tube retraction and new AGV were inserted in both eyes for control of IOP. Significant corneal haze was present in the left eye. The corneal haze in the right eye was minimal and was confined to the superior one third of cornea in the location of the Ahmed tube. Penetrating keratoplasty procedures were performed OU a few weeks apart (secondary to corneal scar) with new AGV insertions in the pars-plana positions. Subsequent diode-laser endo-photocoagulation of the ciliary bodies (laser assisted-ciliary body ablation) 270 degrees OS was performed by age of two. Additional trans-scleral ciliary
body diode laser ablation was done 180 degrees for both eyes. Cataract extraction OU, in addition, was done around age of three. At the last examination under anesthesia, the AGV tubes were visible in good locations at 10 o’clock OD and at 5 o’clock at the pars-plana positions. The IOPs were controlled on two glaucoma medications: Betoptic-S and Trusopt (Dorzolamide HC1 2%, Merck and Co. Inc., Whitehouse Station, NJ). Patient is aphakic and has best corrected Snellen visual acuity of 20/160 OD and hand-motion OS. The irises of both eyes are oval-shaped without any direct peaking. Optic nerve cup/disk ratios are 0.8 for both OD and OS. Revision of the Ahmed tube OS due to exposure and conjunctival erosion (without overlying coverage) was done yet again one year ago by placement of additional tutoplast allograft (Tutogen, Fl) over the exposed tube.

**Discussion**

In our small retrospective review of pediatric glaucoma cases in our practice, we have found significant number of adverse post-operative outcomes requiring further surgical management subsequent to the AGV insertions. As in other published studies, we have observed similarly that congenital or infantile glaucoma is an aggressive disease typically requiring multiple surgeries with variable results.

To date, we have found only one previously reported case in the literature of pupillary abnormality in association with AGV drainage devices (Al-Torbak and Edward 2001). Based on numerous published data, Ahmed valves are found to be effective devices for controlling intraocular pressures when other surgical methods have failed (Englert et al 1999). Ahmed glaucoma drainage devices are rarely applied as a first line of surgical management for control of high intraocular pressures in association with pediatric glaucoma. Moreover, AGV have been shown to have less impact on the extra-ocular movements in comparison to the other larger plated devices.

In this study, we have observed that AGV insertion could create cosmetically significant albeit clinically insignificant pupillary irregularity, lenticular opacification as well as extrusion out of anterior chamber, particularly in the first year of life, as the globe is enlarging with age, ie, buphthalmic process. Though the mechanism of pupillary peaking towards the tube area is not well understood, we postulate through observation that outflow of anterior chamber fluid may create a turbulent pressure-flow wave which may in turn cause elevation of iris and gradual movement of iris towards the tip of the tube. Other potential explanations for iris peaking may include inflammatory response to presence of talc on the tube or local creation of peripheral anterior synechia. Simple eye rubbing could as well extend and retract the tube with secondary iris pull from a sudden gush of aqueous out of the sclerostomy fistula site. Occasionally lamellar iris incarceration due to acute decompression during a fistula creation may lead to iris entrapment and secondary iris elevation. To cope with this adverse outcome, in some subsequent cases, we have manipulated the tube and inserted the tube in a tangential method posterior to the grey surgical limbus line. In such succeeding cases, no pupillary peaking has yet been noted possibly due to the change in the mechanism of outflow direction. Additionally we propose creating a surgical peripheral iridectomy, albeit technically difficult, whenever possible at the entrance site of valve, which in addition may alter the fluid, outflow pathway as well. As an example, the two cases in our series (3 and 6) with co-existing surgical iridectomy did not display pupillary irregularity. It could be postulated that surgical iridectomy may divide the flow of aqueous to the tube entrance site into two separate streams emanating from posterior and/or anterior chamber.

In review of the literature, we have noted numerous published data on the Ahmed valve surgeries in the pediatric patients. In their consecutive large interventional case series of 60 pediatric eyes, Morad and colleagues (2003) reported that AGV is an effective treatment for pediatric glaucoma with long term success rate of 47% over three years; although high rate of post-operative complications including hypotony, tube exposure, tube retraction, endophthalmitis were reported (Al-Torbak and Edward 2001, 2002). A pupillary abnormality with ‘iris blocking the tube’ was noted in their study. In their retrospective review of 27 eyes of pediatric glaucoma, Englert and colleagues (1999) found AGV implantations to be useful therapy with success rate of 58% at 24 months. However, complications of hypotony, retinal detachment, corneal-tube contact and choroidal effusions were reported in their study (Englert et al 1999). In their retrospective review of 37 adult and pediatric patients who had undergone AGV insertion with MMC, Kook and colleagues (2000) reported a 77% success rate over 2-year period without significant rise in complication rate. In the first clinical study to evaluate AGV in pediatric glaucoma, Coleman and colleagues (1997) reported 60% success rate in the study of 21 consecutive pediatric patients at 24 months.

As a result of this study, surgeons have to be aware that as the globe enlarges, the tube is drawn posteriorly and occasionally completely out of the anterior chamber. We have observed through this clinical series that the earlier the AGV insertion is done in life of a child, the higher the probability of tube
extrusion may exist. To cope with this problem, we propose the tube insertion to be done through a tangentially curved corneal tunnel in the primary surgical insertion phase. Once a tube is retracted and/or extruded from anterior chamber, the tube could be re-inserted in a straight-line method directly into the chamber in a secondary procedure circumventing the requirement for an extender. Tube extenders are also currently available and could be added to the tip of the tube if needed.

Postoperative hypertensive phase after AGV implantation has also been described elsewhere along with number of other potential complications in the literature (Nouri-Mahdavi and Caprioli 2003). The mechanism of the late AGV occlusion as well as the surgical approach to reoperation technique with the aid of MRI has already been discussed elsewhere (Hill et al 2000; Pirouzian et al 2006). Furthermore, in our series, we did not observe any intra/postoperative choroidal hemorrhage. However to reduce such a potential complication, we propose decreasing intraocular pressure gradually in the pre-operative period by utilizing diuretics. Application of viscoelastic substances prior to insertion of the tube-shunt, most notably Healon 5 (Pharmacia and Upjohn), may lead to a decreased risk of sudden intra-operative hypotony thus decreasing incidence of choroidal effusion and cataract formation from inadvertent tube-lens touch.

Conclusion
In summary, multiple complications may follow the AGV insertion in children. Pupillary irregularity is most commonly noted in our series. To avoid or reduce the risk of this complication, angular tube placement or peripheral iridectomy could be considered.

Disclosure
No grants have been received for this study. Authors have no financial interest in any of part of this research.

References