Objectives: To evaluate the long-term efficacy of intraocular pressure reduction and complications of Ahmed glaucoma valve (AGV) implantation in children with primary congenital glaucoma.

Methods: The medical records of patients with primary congenital glaucoma who underwent AGV implantation with a minimum follow-up of 6 months were reviewed. The primary outcome measure was cumulative probability of success, defined as intraocular pressure greater than 5 mm Hg and less than 23 mm Hg and at least a 15% reduction from the preoperative intraocular pressure, without serious complications, additional glaucoma surgery, or loss of light perception.

Results: Thirty eyes of 19 children with primary congenital glaucoma who underwent AGV implantation with a minimum follow-up of 6 months were reviewed. The children had a mean (SD) age of 1.8 (2.6) years, a mean (SD) preoperative intraocular pressure of 28.4 (6.7) mm Hg, and a mean (SD) follow-up time of 57.6 (48.0) months. The cumulative probability of success was 63% in 1 year and 33% in 5 years. After a second AGV implantation, the cumulative probability of success was 86% in 1 and 2 years and 69% in 5 years. Hispanic ethnicity ($P = .02$) and being female ($P = .005$) were associated with increased risk of failure.

Conclusions: Thirty-three percent of AGV implantations in children with primary congenital glaucoma were successful after 5 years of follow-up. With the implantation of a second AGV, the 5-year success rate increased to 69%.


Primary congenital glaucoma (PCG) is present at birth but often goes unrecognized until increased intraocular pressure (IOP) damages ocular structures and produces vision loss. Primary congenital glaucoma is a potentially blinding disease that usually does not respond adequately to medical treatment. Although goniotomy and trabeculotomy are associated with good early success rates, eventually 20% of these procedures fail and many children with PCG require additional surgery to control IOP in the long-term. Various surgical approaches have been proposed, including trabeculectomy with or without adjunctive antimetabolites, nonpenetrating external trabeculectomy, combined trabeculotomy and trabeculectomy, glaucoma drainage devices, and cyclodestructive procedures. Trabeculectomy with mitomycin C has shown promise, with studies reporting 52% to 93% success rates. However, the increased success with the addition of antimetabolites has also resulted in well-recognized long-term complications such as bleb failure, bleb leak, and bleb-related endophthalmitis.

The Ahmed glaucoma valve (AGV) (New World Medical, Inc, Rancho Cucamonga, California) is a tube shunt device with a unidirectional flow-restrictive mechanism designed to prevent postoperative hypotony. The success rates reported for this device in the pediatric population range from 58% to 93% in achieving IOP of less than 21 mm Hg, but these rates have been in mixed populations of children with a variety of pediatric glaucoma diagnoses and with short-term follow-up. These reports include children with congenital glaucoma as well as children with secondary diagnoses including aphakia or pseudophakia, Sturge-Weber syndrome, uveitic glaucoma, aniridia, and anterior segment dysgenesis. Because of the heterogeneity in the mechanisms of glaucoma secondary to these various disorders, we evaluated the long-term surgical efficacy of AGV implantation in children with PCG.

METHODS

This was a retrospective study of patients with PCG who underwent AGV implantation from January 1, 1995, to December 31, 2007.
The eye was the unit of analysis. However, repeat analysis was performed with 1 eye per patient (n=19 patients), using the first eye that underwent AGV implantation. In cases where both eyes received AGV implantation simultaneously, the right eye was evaluated.

Kaplan-Meier survival analyses were used to assess the long-term success rates according to the criteria defined earlier. In addition, Cox proportional hazards regression analysis was used to determine the predictive factors for failure. The following potential predictors were evaluated in a univariate analysis: age, sex, ethnicity, baseline IOP, preoperative cup-disc ratio, presence of preoperative corneal edema, preoperative medications used, and number of previous glaucoma surgical procedures. In comparing Hispanic vs non-Hispanic patients, continuous variables were compared using the Kruskal-Wallis test, whereas categorical variables were compared using the Fisher exact test. The Cochran-Armitage test was used to evaluate the underlying trend in postoperative medication use and changes in cup-disc ratios. \( P < .05 \) was considered statistically significant. All statistical analyses were performed using SAS version 9.1 statistical software (SAS Institute, Inc, Cary, North Carolina).

### RESULTS

#### PREOPERATIVE CHARACTERISTICS

Thirty eyes of 19 patients were included in the study. Twelve patients (63%) were male. Eight patients (42%) were white, 8 patients (42%) were Hispanic, and 3 patients (16%) were Asian. The mean (SD) age at the time of AGV implantation for each eye was 1.8 (2.6) years, with a range of 0.02 to 9.9 years. Twenty-eight eyes (93%) underwent prior glaucoma surgery, with 7 (23%) undergoing a single angle surgical procedure (goniotomy or trabeculotomy) and 18 (60%) undergoing 2 or more angle surgical procedures. Among the 7 patients who had only a single procedure prior to AGV implantation, 2 were of older age (mean age, 8.4 years) at the time of implantation, suggesting that the single angle surgical procedure controlled the IOP for a relatively long time before further intervention was necessary. The remaining 5 patients had IOP uncontrolled after a single angle surgical procedure, and a decision was made to proceed to AGV implantation. Three eyes (10%) also underwent trabeculectomy in addition to angle surgery prior to drainage device implantation. The mean (SD) number of prior glaucoma surgical procedures was 1.8 (0.9). The mean (SD) preoperative IOP was 28.4 (6.7) mm Hg, while the mean (SD) IOP at the last follow-up was 17.7 (6.7) mm Hg. Data on glaucoma medications used prior to AGV implantation were not available for 3 eyes. Nine eyes (33%) did not receive any glaucoma medications prior to AGV implantation, 13 eyes (48%) received glaucoma medication prior to AGV implantation, and 5 eyes (19%) received 2 glaucoma medications prior to AGV implantation, with a mean (SD) number of glaucoma medications of 0.9 (0.7). **Table 1** summarizes the baseline characteristics of the study population.

#### IOP CONTROL

The mean (SD) follow-up time was 57.6 (48.0) months, with a range of 8 to 166 months. The mean (SD) IOP de-
increased from a preoperative value of 28.4 (6.7) mm Hg to 16.6 (4.3) mm Hg at 12 months and 17.7 (6.7) mm Hg at 60 months after AGV implantation. All intervening mean postoperative IOP measurements were decreased from baseline (P < .001). Figure 1 displays the course of the mean IOP during the study.

**NUMBER OF MEDICATIONS**

The proportion of eyes requiring IOP-lowering medications increased over 5 years of follow-up. Sixty-three percent of eyes did not require medications at 3 months, whereas 11% of eyes did not require medications at 5 years. This trend was found to be statistically significant using the Cochran-Armitage trend test (P = .002). Figure 2 displays the proportion of eyes requiring no medications vs 1 to 3 medications during the follow-up period.

**CUP-DISC RATIO**

The proportion of eyes that demonstrated no change or improvement in cup-disc ratio on follow-up examinations as compared with baseline increased over time. The Cochran-Armitage trend test demonstrated that the trend of worsening in cup-disc ratio over time was not statistically significant (P = .15). Figure 3 displays the proportion of eyes demonstrating no change or improvement vs worsening in cup-disc ratio over the follow-up period.

**SURGICAL COMPLICATIONS AND ADDITIONAL SURGICAL PROCEDURES**

While there were no intraoperative complications of AGV implantation, postoperative complications were as fol-
SURGICAL SUCCESS AND RISK FACTORS FOR FAILURE

In Kaplan-Meier survival analysis, there was a 63% success rate at 1 year, a 50% success rate from years 2 to 3, a 41% success rate at year 4, and a 33% success rate at 5 years (Figure 4). Six eyes (20%) failed according to IOP criteria, with 5 eyes failing due to IOP greater than 23 mm Hg and 1 eye failing due to IOP less than 5 mm Hg. An additional 10 eyes (33%) failed because a second AGV was implanted, although 8 of these eyes did not fail secondary to IOP criteria. There were no failures due to severe complication or loss of light perception, although 1 eye did have endophthalmitis after tube revision that resulted in a phthisical eye. However, this eye had already failed secondary to IOP criteria prior to the tube revision surgery. When this analysis was repeated using only the first eye of each patient to receive an AGV (19 eyes), similar success rates were found: 68% at 1 year and 31% at 5 years (data not shown).

In the Cox proportional hazards model, Hispanic ethnicity was associated with a 3-fold increased risk of failure ($P = .02$) in univariate analysis. Being male had a decreased risk of failure ($P = .005$). Age, preoperative IOP, preoperative cup-disc ratio, presence of preoperative corneal edema, preoperative medical use, and number of prior glaucoma surgical procedures were not associated with increased risk of failure (Table 2). We repeated the analysis using only the first eye of each patient to receive an AGV, and both Hispanic ethnicity ($P = .02$) and being female ($P = .04$) remained statistically significant risk factors for failure.

Given that 10 eyes (33%) from 6 patients required a second tube, survival analysis was performed using the same failure criteria to examine the success of the second AGV implantation in these eyes. In Kaplan-Meier survival analysis, the success rate was 86% at 1 and 2 years and 69% at 5 years (Figure 5). Two eyes failed because a third AGV was implanted at 27 months and 69 months after the second AGV implantation, although they did not fail due to IOP criteria. The third eye failed due to IOP greater than 23 mm Hg on 2 consecutive visits 12 months after the second AGV implantation, and a third AGV was implanted 14 months after the second AGV implantation. Because of the small number of eyes, no risk factor analysis was performed.
In this study, 3 eyes (10%) required a third AGV implantation, with 2 eyes (66%) maintaining adequate IOP control at the last follow-up (11 months and 48 months). One eye (33%) failed due to IOP greater than 23 mm Hg on 2 consecutive visits 6 months after the last AGV implantation.

**COMMENT**

In PCG, the cumulative probability of success with a single AGV implant was 63% at 1 year of follow-up but decreased to 33% at 5 years of follow-up. However, with a second AGV implantation, the success was 86% at years 1 and 2 of follow-up and 69% at 5 years. Hispanic ethnicity increased the risk of failure of AGV implantation by 3-fold in our study.

The safety and efficacy of aqueous shunt devices in pediatric glaucoma have been established in prior studies.\(^8,11,18,20\) However, PCG is a disease distinct from pediatric glaucomas, which is a term that may include PCG but also includes secondary glaucomas such as aphakic glaucoma or glaucomas associated with increased episcleral venous pressure. Currently, there is no consensus as to the preferred surgical algorithm after failure of angle surgery and there may be wide variation in recommended treatment by centers experienced in the treatment of PCG. Owing to the rarity of the disease, randomized prospective trials have not been performed. We are unaware of other studies evaluating the long-term outcomes of AGV implantation performed for PCG or studies that report the long-term outcomes of a second AGV implantation.

Although aqueous shunt devices have demonstrated considerable success in pediatric glaucoma, it is difficult to compare surgical success rates because prior studies examining surgical outcomes of drainage devices vary in the populations studied, lengths of follow-up, and types of devices implanted. The cumulative probability of success varies widely in different studies, ranging from 42% at 10 years of follow-up in a retrospective case series by O’Malley Schotthoefer et al\(^21\) comparing glaucoma drainage device surgery in congenital glaucoma and aphakic glaucoma to a 93% success rate of AGV implantation in pediatric glaucoma at 12 months in a retrospective case series by Morad et al.\(^20\) Our short-term (63% at 1 year) and long-term (33% at 5 years) survival rates are slightly lower than the rates reported in the literature with similar success criteria, although the studied populations included children with other types of glaucoma, variability in phakic status, and variability in type of glaucoma drainage device implanted.

There is controversy as to whether AGV implantation has a higher rate of failure in patients with PCG compared with patients with other diagnoses.\(^8,9\) While to our knowledge no studies have examined the outcomes of AGV implantation in children with exclusively PCG, Djodeyre et al\(^19\) reported shorter mean survival time with AGVs implanted in 17 eyes with congenital glaucoma compared with 18 eyes with other diagnoses (Sturge-Weber syndrome, postoperative glaucoma, etc). Having more than 2 previous glaucoma interventions was also a risk factor for time to failure. However, the criteria for failure included tube malposition, which was defined as any change in the optimal position of the tube or valve plate that resulted in uncontrolled IOP or other complications (tube retraction, tube-endothelium contact, tube-iris contact, valve plate extrusion). In their study, of the 11 eyes that were considered failures, 9 were eyes with congenital glaucoma, with 56% failure secondary to tube malposition. However, in comparison with our study, the follow-up time was shorter at 12 and 24 months. In contrast, other studies have not found any correlation between surgical failure and glaucoma type in the pediatric population.\(^8,9,20,21\) One of the largest studies, published by O’Malley Schotthoefer and associates, examined the long-term outcome of aqueous drainage devices in 38 eyes with congenital glaucoma and 32 eyes with aphakic glaucoma. One-year success rates were 92% and 90% in the congenital and aphakic groups, respectively, but these decreased to 42% and 55% after 10 years. In the congenital glaucoma group, AGVs were implanted in 27 of the 38 eyes (71%) and Baerveldt implants were placed in the remainder. In addition, 10 of the 38 eyes (26%) had other nonglaucoma surgical procedures prior to aqueous drainage device placement, including cataract surgery, penetrating keratoplasty, and vitrectomy. Thus, there were significant differences between the patients with congenital glaucoma examined in the study by O’Malley Schotthoefer and associates compared with our study, which included patients with PCG who had only glaucoma surgical procedures previously and had only AGVs implanted.

Tube-related problems were the most common complication after AGV implantation in this study, most of which were due to tube-corneal apposition, which necessitated tube trimming in 4 eyes (13%). Additionally, reoperation secondary to tube exposure was required in 2 eyes (7%). Tube migration is thought to be associated with shrinkage of the sclera and globe after IOP reduction, with the tube repositioning more anteriorly and closer to the corneal endothelium. Additionally, it is postulated that vigorous eye rubbing can move the tube forward toward the cornea. Lastly, as the child’s eye grows, the initially well-positioned tube may rotate more anteriorly over time. We recommend that the tube is placed at least 1 mm posterior to the limbus and that the tube is positioned parallel to the iris. Our tube revision rate is similar to other studies that examined glaucoma drainage device implantation in pediatric glaucomas.\(^9,20-22\)

Trabeculectomy with or without antimetabolites is another option in the management of PCG, although the introduction of antifibrotic agents has resulted in higher rates of late-onset bleb-related endophthalmitis compared with the rates in adults.\(^16-18\) A retrospective, comparative case series published by Beck et al\(^22\) comparing aqueous shunt devices with trabeculectomy with mitomycin C reported a cumulative probability of success of 87% in the aqueous shunt group and 36% in the trabeculectomy group during the first year of follow-up. This decreased to 53% and 19% for the aqueous shunt and trabeculectomy groups, respectively, at 6 years of follow-up. While that study included only children aged 24 months or younger, the study population was heterogeneous.
neous and consisted of patients with congenital glaucoma, anterior segment anomalies, varying phakic statuses, and persistent fetal vasculature. In addition, the patients received either the Ahmed or Baerveldt implant.

Kaplan-Meier survival analysis performed on 2 subgroups of eyes demonstrated a less favorable surgical outcome for those eyes in patients of Hispanic ethnicity compared with those of non-Hispanic ethnicity. To our knowledge, this difference has not been reported in prior studies, most likely owing to the rarity of the disease. A review of the literature also did not identify studies demonstrating worse surgical outcomes for adult Hispanic patients receiving glaucoma drainage devices. In this study, a comparison of various preoperative characteristics (ie, age, preoperative IOP, preoperative cup-disc ratio, preoperative corneal edema, preoperative medication use, and previous glaucoma surgery) did not reveal any differences between Hispanic and non-Hispanic patients. Since being female is also associated with increased risk of failure in this study, a comparison of preoperative characteristics was performed and did not show any differences between male and female patients, including those of Hispanic ethnicity (data not shown).

To our knowledge, no studies report the long-term outcome of a second glaucoma drainage device implantation in children with PCG. Given the wide variety of surgical algorithms used and the rarity of this disease, a clinician counseling a family might find it helpful that based on this study, a second AGV implantation for further IOP control in children with prior AGV implantation appears to be safe and effective with a 5-year probability of success of 69%. However, the small number of eyes that underwent a second AGV implantation in this study precluded an analysis of risk factors for subsequent failure.

Our study is limited by its retrospective design because of the differences in preoperative characteristics and lack of a prespecified interval of follow-up and data collection. For example, patients who have undergone prior glaucoma surgery or multiple surgical procedures may have a more severe course of glaucoma and may be more susceptible to surgical failure. In addition, because quantitative visual acuity testing could not be done at the time of AGV implantation, our ability to comment on visual outcomes is limited. The study’s strengths are its uniformity in PCG population, type of glaucoma drainage device used, and its relatively large number of eyes.

In summary, we report moderate success with IOP control using the AGV in patients with PCG. However, Hispanic ethnicity was a risk factor for surgical failure and a more severe course of glaucoma and may be more susceptible to surgical failure. In addition, because quantitative visual acuity testing could not be done at the time of AGV implantation, our ability to comment on visual outcomes is limited. The study’s strengths are its uniformity in PCG population, type of glaucoma drainage device used, and its relatively large number of eyes.

Submitted for Publication: December 23, 2008; final revision received March 12, 2009; accepted March 24, 2009.

Correspondence: Joseph Caprioli, MD, Glaucoma Division, Jules Stein Eye Institute, David Geffen School of Medicine, 100 Stein Plaza, Los Angeles, CA 90095 (caprioli@sei.ucla.edu).

Financial Disclosure: None reported.

Funding/Support: This work was supported by an unrestricted grant from Research to Prevent Blindness.

Previous Presentation: This study was presented at the annual meeting of the Association for Research in Vision and Ophthalmology; April 28, 2008; Fort Lauderdale, Florida.

REFERENCES