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Glaucoma drainage device implantation in glaucoma associated with aniridia

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ABSTRACT

Based on a retrospective analysis of the literature describing implantation of glaucoma drainage devices in patients with aniridia-related glaucoma, we review the results of glaucoma drainage device implantation in this group of patients. In addition, we present a case report of a 3-month-old infant with the WAGR syndrome (Wilms tumor, aniridia, genitourinary anomalies, mental retardation) and congenital glaucoma associated with aniridia, in whom the glaucoma drainage device was implanted. In our case, in a 3-month-old infant in whom implantation of an Ahmed valve (model FP8) was performed in the right eye, intraocular pressure decreased from 37.0 mmHg to 17.3 mmHg. One year after the surgery, a surgical revision was performed and the proper function of the Ahmed valve was restored – intraocular pressure decreased from 29.0 mmHg to 15.0 mmHg. At the last visit, 21 months after the valve implantation, pharmacotherapy was introduced and intraocular pressure was 15.6 mmHg. Cataract was the only observed complication and was operated on successfully at 7 months of age. To date the research results indicate that glaucoma drainage device implantation is an effective method of treatment of aniridia-related glaucoma.

Conclusions: Early diagnosis and glaucoma drainage device implantation as well as close monitoring of patients give a chance for prolongation of functional vision.

KEY WORDS: glaucoma associated with congenital aniridia, glaucoma drainage devices.

INTRODUCTION

Aniridia is a rare, congenital panocular disorder, in most cases caused by a mutation in the PAX6 gene on chromosome 11p13 [1]. The incidence of aniridia is between 1:64 000 and 1:100 000 [2]. It may be a component of WAGR syndrome (Wilms tumor, aniridia, genitourinary anomalies, mental retardation) [3] or Gillespie syndrome (aniridia, ataxia and mild to moderate developmental delay), which is not related to PAX6 gene mutation [4]. Aniridia is a partial or complete hypoplasia of iris. It can be accompanied by corneal opacification, cataract, lens dislocation, dry eye disease, meibomian gland dysfunction, foveal and optic nerve hypoplasia, nystagmus and glaucoma [5, 6]. Glaucoma associated with aniridia is one of the major therapeutic challenges, and its prevalence ranges from 6 to 75% [2, 7]. Pathophysiology of this type of glaucoma is multifactorial. Development of tissue between the iris rudimentary and angle wall causes anterior rotation of the iris which leads to closure of the anterior chamber angle. In addition, there are abnormalities of Schlemm's canal and trabecular meshwork [2, 6, 8, 9].

MATERIAL AND METHODS

We found 13 publications describing the results of aniridic glaucoma treatment. The research results show that in the treatment of aniridia-related glaucoma, the following methods are used: pharmacotherapy, argon laser trabeculoplasty, cyclophotocoagulation with diode laser, cyclocryotherapy, goniotomy, trabeculotomy, trabeculectomy, and implantation of glaucoma drainage devices (GDDs). In 7 papers, the treatment method was GDD implantation. In total, 37 patients who were implanted with GDDs were described: a Molteno implant in 17 of them, an Ahmed glaucoma valve (AGV) in 13, and a Baerveldt implant in 7.

Wiggins and Tomey reported 6 Molteno drainage device implantations [10]. Molteno and Cunliffe implanted 3 Molteno drainage devices [11]. Molteno *et al*. implanted 3 Molteno drainage devices [12]. In Adachi's research 6 Molteno drainage devices were implanted [9]. In the Demirok *et al*. study 5 AGVs and 1 Molteno implant were implanted [7]. In Almousa and Lake's study 8 AGV implantations were performed [13]. In our case the AGV was implanted. Arroyave

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et al. reported 7 Baerveldt implants and 1 Molteno implant [14].

Our Ahmed glaucoma valve (model FP8 designed for children) (Figures 1 and 2) implantation technique was as follows.

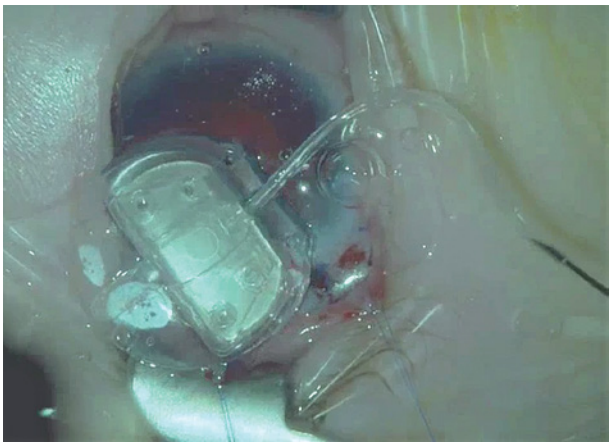

Figure 1. Ahmed valve FP8

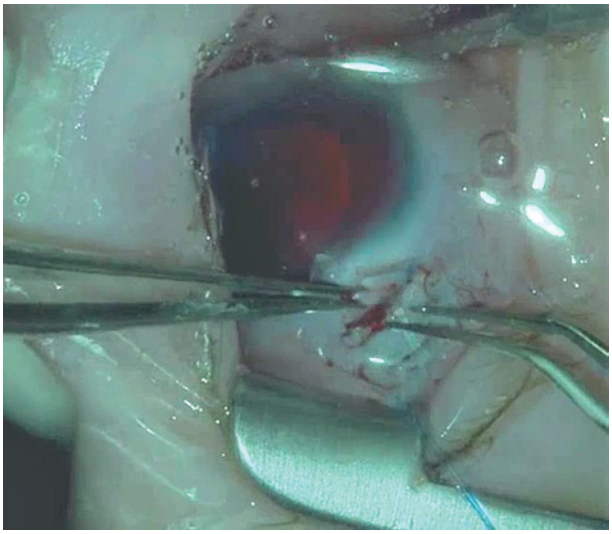

Figure 2. Fixation of the silicone tube in the anterior chamber of the eye

The surgery was performed using the NGENUITY 3D Visualization System. The patient was under general anesthesia. Initially, a perilimbal incision of the conjunctiva and Tenon's membrane, approximately 10 mm long, was made in the superior temporal quadrant, dissecting them posteriorly. Then, a 4 × 3 mm scleral flap was made. At a distance of 8 mm from the limbus, the AGV plate was fixed to the sclera with two 6-0 Nylon sutures. At a distance of 2 mm from the limbus, an intrascleral tunnel was made into the anterior chamber with a 22G needle. A viscoelastic was administered to the anterior chamber and a silicone tube approximately 3 mm long was inserted into it (Figure 2). The scleral flap was sutured with 2 sutures of 10-0 Nylon. The conjunctiva was sutured in the limbus with 10-0 Nylon continuous suture. Finally, cefuroxime was administered subconjunctivally.

RESULTS

In Almousa and Lake's research, after 12 months of follow-up, surgical success was achieved in 87.5% of patients (implantation of 8 Ahmed glaucoma valves) [13]. In the Demirok *et al*. study, after 12 months, the effectiveness of the procedure was 66.7% (5 AGVs and 1 Molteno implant) [7]. Arroyave *et al.* achieved the expected intraocular pressure (IOP) in 87.5% of patients after 12 months (7 Baerveldt implants and 1 Molteno implant) [14]. In the Wiggins and Tomey paper the procedure was successful in 5 eyes – 83% of patients (6 Molteno implants) [10]. Molteno and Cunliffe reported 3 aniridic eyes with Molteno implants with 100% success [11]. In the Molteno *et al*. study all 3 implantations of Molteno drainage devices were successful but the follow-up period was unknown [12]. In the Adachi *et al*. research 6 Molteno implant procedures were performed, and 2 of 6 succeeded [9].

In our case surgical success was achieved (IOP between 5 and 22 mmHg, regardless of pharmacotherapy – according to Demirok) [7] and the follow-up was 21 months. The mean IOP at the last visit was 15.6 mmHg. GDD implantation reduced IOP by 53% after surgery (17.3 mmHg) and by 58% (15.6 mmHg) at the last visit, compared to the highest IOP (37.0 mmHg).

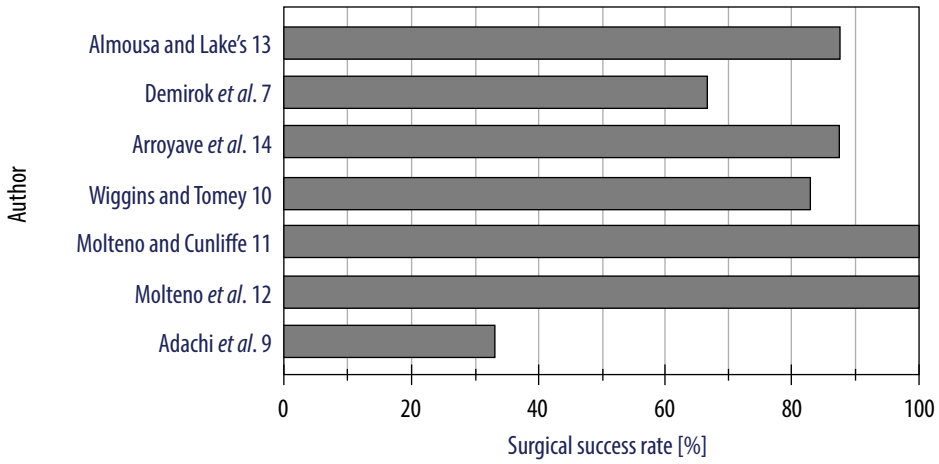

Figure 3. Surgical success rate of GDD implantation

CASE REPORT

In a newborn born in the 39th week of pregnancy with the WAGR syndrome associated with congenital glaucoma at 8 weeks of age trabeculectomy was performed in the right eye due to high IOP (mean 35.0 mmHg). Because of increased IOP (37.0) at 11 weeks of age fistula restoration was performed without success. The Ahmed glaucoma valve (AGV) (FP8) (Figure 1) was implanted in the right eye at 3 months of age and a decrease in IOP (average 17.3 mmHg) was achieved. The only observed complications were cataract in the right eye and decreased filtration efficiency of the AGV caused by scarring of the conjunctiva surrounding the plate of the AGV. Therefore at 7 months of age phacoaspiration with posterior capsulorhexis and anterior vitrectomy was performed. Additionally at 16 months of age a surgical revision of the GDD was performed and the proper function of the Ahmed valve was restored (IOP decreased from 29.0 to 15.0 mmHg). At the last visit at 24 months of age (21 months after GDD implantation) an anti-glaucoma drug was added (Xalacom) and IOP was 15.6 mmHg. Visual acuity could not be established due to the lack of patient cooperation. There was a subcapsular cataract in the left eye. Left eye IOP was stabilized on conservative treatment (Xalacom 1x/day). Binocular glaucoma neuropathy (c/d [cup to disc ratio] – 0.7) with the absence of other significant changes in the retina was diagnosed.

DISCUSSION

Treatment of glaucoma associated with aniridia is very difficult. Pharmacotherapy is not sufficient to obtain significant reduction of the intraocular pressure and it may be effective in only 38.7% of cases [8]. Laser trabeculoplasty and photocoagulation with a diode laser are ineffective because the changes in the anterior chamber angle are so extensive and there are abnormalities of Schlemm's canal and trabecular meshwork [10, 15].

Cyclocryotherapy is usually used as the last line of treatment due to the high incidence of severe complications (phthisis bulbi) and low efficiency. An additional reason for using cyclodestructive treatments mainly as the last line of treatment is the irreversible destruction of ciliary body functions by these methods [10, 18].

There is no consensus as to the choice of the best method of surgical treatment. In some cases, prophylactic goniotomy can prevent or delay aniridic glaucoma. However, performing prophylactic goniotomy in each patient with aniridia raises doubts (prevalence of glaucoma associated with aniridia ranges from 6 to 75%) [2, 10]. There are only a few studies describing trabeculotomies as an effective treatment for aniridia-associated glaucoma. It requires a lot of experience and skill from the surgeon. Additionally, changes in the anterior chamber angle occurring in the aniridia can significantly impede the performance of this procedure. Many authors have failed to replicate the results presented by Adachi. "Ten (83%) of 12 eyes obtained IOP control after first (6 eyes) or second (4 eyes) trabeculotomy with a mean follow-up period of 9.5 years" [6, 9].

The results of the studies on the effectiveness of trabeculectomy in aniridic glaucoma are not conclusive. Some of the research (Wiggins and Tomey, Grant and Walton, Adachi and coworkers) results indicate low effectiveness of trabeculectomy in maintaining IOP < 21 mmHg [8-10]. Nelson *et al*. and Okada *et al*. had good results that differed from other authors [2, 16]. This may be due to the specific selection of patients as well as the improvement of the trabeculectomy technique.

The most commonly used types of GDDs are the Molteno and the Baerveldt (open drain devices) and the Ahmed (restricted drain), which is the only one among the mentioned devices that has a filter valve [6]. Complications of GDD implantation occur with a frequency of 12 to 33% within 12 months after surgery. Mild complications include hyphema, bleb leakage, flat anterior chamber, contact of the tube with the cornea, hypotony and cataract. Possible severe complications are phthisis bulbi (long-term occurrence in up to ¼ of patients and resulting from ciliary body hypoplasia), vitreous hemorrhage, tube exposure or its obstruction, endophthalmitis, corneal opacification and retinal detachment [6, 7, 13, 17].

Over time, as a result of fibrotic processes, the drainage efficiency of the GDDs decreases. Implantation technique and the type of covering tissue may also probably influence the decrease in drainage efficiency [7, 13, 17]. In addition, patients with aniridia may have aniridic fibrosis syndrome, which is a rare complication of invasive intraocular surgery (especially cataract surgery with implantation of an artificial lens, as well as corneal transplantation and GDD implantation). It is characterized by progressive fibrosis in the anterior chamber. Early changes involve the formation of a thin membrane covering the anterior and posterior surfaces of the IOL (intraocular lens), fibrosis and capsular contraction causing slight anterior displacement to the posterior chamber of the IOL. This membrane may also cover the anterior retina and ciliary body, leading to hypotony [19, 20].

The research studies to date show that GDD implantation is one of the most effective methods of treatment of glaucoma associated with aniridia, usually used secondary to pharmacotherapy and filtration treatments, when they did not bring the expected decrease in IOP [10]. However, GDD implantation is increasingly used as the primary surgical procedure [7, 13, 14].

Out of 37 GDD implantations described in the mentioned studies only 9 failed (75.7% of procedures achieved surgical success). Our case also shows the effectiveness of Ahmed valve implantation in glaucoma associated with aniridia. Use of the Ahmed valve probably reduced the risk of serious complications such as hypotony, compared to open drain devices. To our knowledge, this patient is the youngest, among patients described in the literature, whose Ahmed valve was implanted. Other methods in long-term observations are not as effective as GDD implantation but future studies on a large group of patients are necessary to prove our thesis.

CONCLUSIONS

Glaucoma drainage device implantation is an effective method of treatment of aniridia-related glaucoma. Early diagnosis and GDD implantation as well as close monitoring of patients give a chance for prolongation of functional vision.

DISCLOSURE

The authors declare no conflict of interest.

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