Results of Filtering Surgery in Young Patients with Aniridia

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ABSTRACT

To evaluate whether filtering surgery is effective in controlling the intraocular pressure of young aniridic patients with glaucoma, we retrospectively reviewed the charts of aniridic patients with glaucoma under the age of 40 years. We defined a good intraocular pressure control period as the time from surgery until IOP exceeded 20 mm Hg, with or without glaucoma medication. Twenty filtering surgeries (17 trabeculectomies and 3 trabeculectomies with mitomycin C) were performed on 10 eyes in 6 patients for more than 20 years. The mean good intraocular pressure control period after the filtering surgery was 14.6 months (range, 2 to 54 months). Aside from mild choroidal detachment, no other serious complications were encountered. We believe that filtering surgery is efficacious for control of intraocular pressure of young aniridic patients with glaucoma.

Key words: Aniridia, Glaucoma, Filtering surgery, Mitomycin C

Aniridia is a rare malformation characterized by the absence of all but the most peripheral portion of the iris. Glaucoma often develops in aniridic patients, especially during childhood or adolescence, and carries a poor prognosis. Unfortunately, it has been reported that glaucoma in aniridic eyes is difficult to control even with surgical intervention. Moreover, when control of the IOP is achieved after the primary operation, it gradually becomes elevated and uncontrolled and further surgery will be required. In fact, many aniridic patients with glaucoma undergo several operations during their lifetime. Some reports of filtering surgery have described the result of surgery in aniridic patients with glaucoma. Several clinicians reported discouraging results because of the unfavorable influence of a previous filtering surgery, aphakia, or youth on the outcome of a subsequent trabeculectomy. In contrast, several ophthalmologists have recommended this procedure. It is still unknown whether filtering surgery is effective in controlling the intraocular pressure (IOP) in young aniridic patients with glaucoma. We retrospectively report here the follow up study of young aniridic patients treated with filtering surgery for glaucoma.

PATIENTS AND METHODS

All the charts of aniridic patients under the age of 40 years who had undergone filtering surgery for glaucoma at the Hiroshima University Medical Hospital between January 1979 and September 1999 were reviewed. The diagnosis of aniridia was made by slit-lamp examination, and all of the patients had the characteristic clinical manifestations of aniridia. The diagnosis of glaucoma was based on elevated IOP (> 21 mm Hg) in association with optic disc or visual field changes in young patients (older than one year) or elevated IOP with corneal opacity in infants (one year or younger). We did not measure subjective visual acuity in infants. Before the first surgery and after, young patients were treated with topical beta-blockers, pilocarpine and dipivalylepinephrine and oral acetazolamide. After surgery, infants were treated with topical beta-blockers, pilocarpine and dipivalylepinephrine only. All patients had elevated IOP with glaucoma medication at the time of surgery. Intraoperative mitomycin C treatment was included with all trabeculectomies performed after March 1994 (Table). We defined a good intraocular pressure control period as the time from surgery until IOP exceeded 20 mm Hg, with or without glaucoma medication.

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The IOP measurements were recorded during unseeded examinations or examinations in which chloral hydrate sedation was used. The measurements were taken with a Goldmann or pneumatic tonometer.

All surgeries were performed with the same operative technique. Briefly, after a limbal-based conjunctival flap was prepared, a 4 × 4 mm rectangular scleral flap of one-third to one-half scleral thickness was dissected into the clear cornea. A 1 × 3 mm trabeculectomy opening was then created. A peripheral iridectomy was performed and the scleral flap was loosely closed with 10-0 nylon sutures. The conjunctival incision was then closed with a running 8-0 absorbable suture. In the cases of trabeculectomy with mitomycin C, after the scleral flap preparation, a solution of 0.2 mg/ml mitomycin C was prepared, and a small surgical sponge soaked with the solution was held in direct contact under the scleral flap for 5 min. After that, the entire area was irrigated with 250 ml of saline and the scleral flap was sutured tightly. The conjunctival incision was then closed with a running 8-0 absorbable suture. Laser suture lysis was performed when it was deemed necessary.

The postoperative visits were set for 1, 2 and 4 weeks and then every month within the first 6 months, every 1 or 2 months within the second 6 months, and thereafter every 1 to 3 months.

**RESULTS**

The review of all patients under the age of 40 years who had undergone filtering surgery resulted in 10 eyes from 6 patients with aniridia. All patients had at least 6 months’ follow up after the last surgery. Five eyes from three patients dropped out during the follow-up period, at an average of 97.6 months after the first surgery (all 3 moved out of Hiroshima). All of these patients, except for one, had not been subjected to any other operation at the time of the initial examination. One eye had undergone a pars plana lensectomy and a trabeculotomy. Nine eyes were phakic and did not have cataract. The ages of these patients at the first operation ranged between 0.4 to 278 months. The mean first preoperative IOP was 33.5 ± 6.9 mm Hg. The 20 filtering surgeries consisted of 17 trabeculectomies and 3 trabeculectomies with mitomycin C. The mean postoperative follow-up period after the first operation in our clinic was 85.6 months (range, 6 to 188 months). Complications included; mild choroidal detachment in 1 out of 19 (5%) trabeculectomies and in 1 out of 3 (33%) trabeculectomies with mitomycin C. The mean good IOP control periods after the whole filtering surgery, a trabeculectomy, and a trabeculectomy with mitomycin C were 14.6 months (range: 2 to 54 months), 14.2 months (range: 2 to 54 months), and 16.7 months (range: 2 to 42 months), respectively. The IOPs of 4 eyes remained at or below 20 mm Hg throughout the entire follow-up period after the first operation.

**DISCUSSION**

We have demonstrated that filtering surgery can be successful in young aniridic patients. There have been some reports of trabeculectomies performed on aniridic patients. Grant & Walton described 11 filtering operations performed on 7 aniridic patients (9 eyes) with glaucoma, and reported that they were unsuccessful. Beauchamp & Parks reported that only 40% of the 5 eyes in 3 aniridic children were controlled by the trabeculectomies even though the IOP control was defined as the last recorded IOP being 24 mm Hg or less. Wiggins & Tomey noted that 10 (91%) of 11 trabeculectomies were unsuccessful because many patients were young or aphakic. Adachi et al reported that in the four eyes out of five, failure occurred soon after the initial trabeculectomy. On the other hand, Nelson et al commented that trabeculectomies were likely to succeed because 11 (76%) out of 14 aniridic eyes had succeeded with follow-up periods exceeding one year. Susanna et al first illustrated that a trabeculectomy with mitomycin C in one eye of an aniridic infant lowered the pressure from 32 to 22 mm Hg, 11 months later, and suggested that this seemed to be a reasonable approach in congenital or developmental glaucomas eyes with a poor surgical prognosis or otherwise candidates for setons. A few years later, a couple of reports on trabeculectomy with mitomycin C performed on a small number of pediatric patients with aniridia were published stating that satisfactory outcomes were obtained. In this context, we found our results encouraging when they were compared with those of the above reports because 10 previously failed filtering surgeries were included in our series and our criteria of a good intraocular pressure period seems to be somewhat more stringent than others.

Several potential mechanisms that can produce an elevated IOP in patients with aniridia have
been proposed\(^6\) but the details are still under discussion. A histopathologic study\(^13\) of aniridic children with glaucoma demonstrated that the trabecular meshwork and the canal of Schlemm could not be identified in two patients with abnormal chromosomes. Moreover, the angles in other two children over the age of 10 years were closed by extensive peripheral anterior synechiae. A trabeculectomy for the angle closure glaucoma was effective in controlling IOP\(^2,19\). Therefore, it appears to be reasonable to perform filtering surgery on young aniridic patients with glaucoma.

First introduced in the early 1980s, the trabeculectomy with mitomycin C has largely superseded the trabeculectomy alone as the preferred treatment. The rationale for the use of mitomycin C was that mitomycin C would be effective in promoting bleb formation. The advantages of a trabeculectomy with mitomycin C include the fact that intraoperative mitomycin C has been shown to improve filtration in high-risk cases\(^19\), and that a history of previous surgery does not correlate with the postoperative IOP\(^2,19\) or the incidence of complications\(^9\). On one hand, we believe that we must take the toxic effects of mitomycin C into consideration. There are several negative views against use of mitomycin C. A long term outcome of infantile glaucoma\(^10\) suggested that it was not necessary to use antiproliferative agents for trabeculectomies in infantile glaucoma in view of postoperative complications. On the contrary, histopathologic specimens have demonstrated that the toxic intraocular changes due to mitomycin C were dose dependent, and did not recover overtime\(^10\). Encouragingly, a recent study using human tissue\(^16,30\) has indicated that exposure to mitomycin C at lower doses was relatively nontoxic. We believe that the use of mitomycin C in trabeculectomy for young aniridic patients is acceptable because young age is a significant negative factor in bleb survival\(^16,30\) and, moreover this type of glaucoma is considered to be intractable. We have the impression that the trabeculectomy with mitomycin C seems to be more effective in controlling the IOP than the trabeculectomy in the present patients, although the number of procedures which we performed in aniridic glaucoma is small.

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