Goniosurgery for Prevention of Aniridic Glaucoma

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Objective: To report the long-term success and complications of modified goniosurgery to prevent aniridic glaucoma, an entity that typically is difficult to control medically or surgically.

Design: A retrospective review of the medical charts.

Results: Fifty-five eyes in 33 patients who had aniridia without glaucoma and who underwent goniosurgery were identified. Ninety-one procedures were performed on the 55 eyes by 1 surgeon (D.S.W.). Each eye had an average of 1.65 procedures and an average of 200° of goniosurgery. Average age at time of initial goniosurgery was 36.6 months. There were no operative complications. No eye had a decrease in visual acuity at last follow-up. All eyes had a preoperative intraocular pressure (IOP) less than 21 mm Hg. At last follow-up (average, 9 years 6 months; range, 8 months to 24 years), 49 eyes (89%) had IOPs less than 22 mm Hg without medications. The remaining 6 eyes (11%) had IOPs of 22 mm Hg or less with up to 2 types of eyedrops.

Conclusions: Without prophylactic goniotomy, aniridic glaucoma may be expected in half of patients, and when it occurs, it is extremely difficult to control. Prophylactic goniosurgery in selected eyes of young patients with aniridia may be effective in preventing aniridic glaucoma.


Aniridia is a rare hereditary ocular disorder typically characterized by iris hypoplasia and the risk of glaucoma. This condition may show autosomal dominant transmission or be sporadic. Other clinical manifestations of aniridia may include decreased vision, cataracts, and nystagmus. Decreased vision can occur secondary to corneal opacities, cataracts, ectopia lentis, foveal and optic nerve hypoplasia, and glaucoma.

Although aniridic glaucoma at birth is rare, the incidence of glaucoma in late childhood or early adulthood ranges from 6% to 75%. Its development has been attributed to progressive changes in the glaucomatous angle occurring during the first 2 decades of life. The most common course of glaucomatous angle progression includes increased confluence of irregular attachments from the iris stroma onto the angle wall. These attachments migrate forward to obscure the scleral spur and posterior trabecular meshwork. This may be accompanied by tilting of the iris from the normal plane perpendicular to the axis of the eye to a plane parallel to the axis of the eye (Figure 1). Other possible mechanisms of increased intraocular pressure (IOP) may include absence of Schlemm canal or secondary angle closure after miotic therapy (D.S.W., unpublished observation, April 4, 1972).

Once glaucoma develops, medical therapy may prove inadequate. Surgical therapy also has an uncertain prognosis and has included argon laser trabeculoplasty, goniotomy, filtering procedures (trabeculectomy and full-thickness filters), and cyclocryotherapy. Other techniques, such as trabeculotomy or Molteno implants or internal sclerectomy with an automated trephine, may have more potential for success.

Because of the progressive nature of the angle changes associated with worsening glaucoma and because of the difficulties of medical and surgical treatment in advanced stages of glaucoma, a prophylactic modified goniosurgery described by Grant and Walton and Walton has been performed to prevent glaucoma in eyes showing early glaucomatous angle changes. This prophylactic surgery detaches the abnormal extensions of iris stroma from the angle wall.
PATIENTS AND METHODS

We reviewed all medical records of patients with aniridia who were seen by one of us (D.S.W.) from 1971 to 1997. These records included those who had prophylactic modified goniotomy as well as those who did not.

For all patients, IOP at time of initial examination was recorded. For patients who had prophylactic goniosurgery, the following information was reviewed: age at surgery, preoperative and postoperative IOP, corneal diameter, visual acuity, length of postoperative follow-up, complications, and gonioscopic observations.

Exclusion and inclusion criteria for prophylactic goniosurgery were as follows. Because of the normal shallow anterior chamber present during the first year of life, surgery was avoided during this interval. Office gonioscopy and Perkins tonometry, however, were performed in patients examined during their first year. After 1 year of age, gonioscopic abnormalities dictated the recommendation for surgery. When office examination produced uncertain observations, the patient was subsequently examined under general anesthesia. If the trabecular meshwork was found to be unobstructed for greater than half of its circumference, no surgery was recommended. If, however, the posterior trabecular meshwork was covered by apparent extensions of tissue from the peripheral iris for more than half of its circumference, then surgery was recommended. Anterior progression of peripheral iris tissue onto the trabecular meshwork over time was also an indication for prophylactic goniotomy. Patients were excluded from consideration for prophylactic surgery if their IOP was greater than 21 mm Hg. A second operation was done if there were any persistent iridotrabecular attachments or if there were continued progressive angle changes.

Preoperative and postoperative office IOP measurements were recorded from unsedated examinations by means of a Perkins tonometer (Clement Clarke International Ltd., London, England). Under general anesthesia and before prophylactic goniotomies, all patients underwent Schiötz tonometry, anterior segment examination, and funduscopy by direct ophthalmoscopy. A handheld microscope and light source were used for Koeppen gonioscopy. All prophylactic modified goniotomies were done by 1 surgeon (D.S.W.) with the same technique, which is described below.

Surgical success was defined as IOP maintained at less than 22 mm Hg without medications and as no increase in optic nerve cupping. Control was defined as IOP less than or equal to 22 mm Hg with medications. Failure was defined as IOP greater than 22 mm Hg despite medications or as a need for further surgery. The postoperative course and repeated gonioscopy were described. Poor fixation or poor cooperation precluded adequate visual field examination in most of our patients.

All surgeries were performed with the patient under general endotracheal anesthesia. Goniotomies were done with an operating gonioscopy lens, loupes, and a nontapered knife. The eye was pretreated with 1 drop of 0.5% atropine sulfate to produce cycloplegia to prevent rotation of the iris stroma anteriorly and with 1 drop of 0.5% apraclonidine to decrease postsurgical bleeding.

The globe was entered nasally and temporally. The operating lens was smaller than that usually used in nonaniridic patients with larger corneas, to allow easy entry through peripheral clear cornea. The initial direction of entry was slightly more posterior than is customary, to avoid a lengthy corneal passage. Rotation of the globe by the assistant during the procedure allowed a larger arc of surgery to be performed.

The tip of the gonioscopy knife engaged the abnormal tissue extending anteriorly over the trabecular meshwork (Figure 2). Gentle pressure posteriorly was used to begin removing this tissue from the meshwork. Although such tissue was frequently vascularized with visible red vessels, bleeding was not encountered. Posterior movements of the knife tip continued as the blade was rotated circumferentially for approximately 3 clock hours. The knife was then returned to the starting meridian and was used to sweep the sulcus that had just been created to eliminate any persistent iridotrabecular attachments. The operation was then continued for the full extent of the angle that could be easily visualized. The knife was then removed and the anterior chamber reformed with balanced saline solution. Reflux of blood into the anterior chamber after surgery was minimal. The anatomical result of this technique was to produce a permanent separation between the iris and filtration tissue. Inspection of the eyes 6 and 24 hours after surgery showed little evidence of any procedure. Compared with the fellow eye, some shallowing of the anterior chamber was commonly observed. A final administration of 0.5% atropine sulfate was given on the first morning after surgery.

This prophylactic procedure departed from the technique of goniotomy for glaucoma by intentionally avoiding injury to the trabecular tissue. In contrast, when goniotomy is performed for treatment of aniridic glaucoma, a cleft is produced in the trabecular tissue.

In the present study, we report the long-term results in patients with aniridia who have undergone prophylactic goniosurgery.

RESULTS

Medical record review disclosed 145 patients with aniridia. Of these patients, 112 were seen for eye care and did not have prophylactic goniosurgery. None of these patients met the criteria for prophylactic goniosurgery. The other 33 patients had prophylactic goniosurgery in at least 1 eye. Some of these patients have been included in 2 previous publications.3,6

Of the 33 patients, 55 aniridic eyes met the inclusion criteria for prophylactic goniosurgery. All patients who were offered surgery underwent prophylactic goniosurgery. All patients preoperatively had IOPs lower than 21 mm Hg without glaucoma medications. Mean age at the time of initial prophylactic goniosurgery was 36.6 months (range, 10-113 months). There were 18 boys and 15 girls. Twenty patients showed autosomal dominant inheritance, and 13 patients showed sporadic inheritance. Eleven of the 55 eyes were myopic, with an average spherical equivalent of −2.23 diopters.

Of these 33 patients, 11 aniridic eyes did not meet the inclusion criteria for prophylactic goniosurgery. Six of these 11 eyes developed glaucoma at an average time of 7 years 11 months (range, at birth to 20 years 9 months). The other 5 of these 11 eyes had not developed glaucoma at the time of last follow-up.
Ninety-one procedures were performed on the 55 eyes by 1 surgeon (D.S.W.). Nineteen eyes had 1 procedure done, and 36 eyes had 2 procedures done. Each eye had an average of 200° of goniosurgery. There were no operative complications. The follow-up ranged from 8 months to 24 years (average follow-up, 9 years 6 months).

Of the 55 eyes that had prophylactic goniosurgery, 49 eyes (89%) had IOPs less than 22 mm Hg without medications and were therefore successful at last follow-up (Figure 3). None of these 49 eyes had an increase in optic nerve cupping. The remaining 6 eyes (11%) of 3 patients had IOPs less than or equal to 22 mm Hg with 1 or 2 topical glaucoma medications and were therefore controlled (Figure 3). The average time after prophylactic goniosurgery at which IOP increased above 21 mm Hg was 13 years (range, 10-17 years). In these 6 eyes, the average age at which IOP increased above 21 mm Hg was 16 years 3 months. No patients needed further surgery or had IOP greater than 22 mm Hg despite medications; therefore, there were no failures. At last follow-up, no patients had a decrease in visual acuity.

### COMMENT

The reported incidence of glaucoma in aniridia varies from 6% to 75%, but the approximate risk of glaucoma is generally conceded to be 50%. In our office, 53% of 224 eyes (112 patients) had glaucoma. All of these 112 patients either already had glaucoma at the time of initial examination or did not otherwise qualify for prophylactic goniosurgery. Glaucoma was defined as an IOP greater than 21 mm Hg. The average age at which glaucoma developed in these 112 patients who did not have prophylactic goniosurgery was 8 years 1 month (range, at birth to 60 years 5 months). Of the patients who did not develop glaucoma, the average age at time of last follow-up was 16 years 1 month (Figure 4).

The mechanism of aniridic glaucoma is often related to progressive angle changes that occur during the first decade of life. Glaucoma usually appears in childhood or early adulthood and does not typically manifest with enlarged corneas or myopia. Aniridic glaucoma in infancy is uncommon and has been associated with abnormalities of Schlemm canal or angle function. The angles of these young patients typically do not show progressive angle changes. Once glaucoma develops, the condition often becomes refractory to medical therapy.

Surgical management of aniridic glaucoma is difficult. Blake reviewed the experiences of 41 ophthalmologists who had operated on aniridic patients with glaucoma; he concluded that there was no convincing opinion as to the best surgical treatment.

Results of argon laser trabeculoplasty have been unsatisfactory. Goniotomy has also been tried for the treatment of aniridic glaucoma. Although Barkan reported a case of successful goniotomy at 9 months of follow-up, other authors with larger numbers of patients reported lower success rates (20% or less).

Trabeculectomy may also be disappointing. Nelson et al noted that 5 of 14 patients either needed reoperation or had failure with primary trabeculectomy. Others reported success rates for initial filtering procedures (partial- and full-thickness) only ranging from 0% to 9%.

Cyclocryotherapy does not show significantly better results. Higher rates of successful IOP control have been reported (17% to 66%), however, because of significant rates of phthisis (up to 50%) and loss of vision (up to 57%) with cyclocryotherapy, this cannot be recommended as a primary procedure for aniridic glaucoma. Strasser suggested that argon laser cyclophotocoagulation may even accelerate the preexisting tendency of tissue proliferation within the anterior chamber angle.

Molteno et al reported 3 aniridic eyes with glaucoma that achieved an IOP less than 20 mm Hg without medications after Molteno implantation. Billson et al used a 2-stage Molteno implant, which lowered IOP to less than 21 mm Hg in 2 eyes in 1 patient with aniridia. Wiggins and Tomey reported successful IOP control in 5 (83%) of 6 eyes. Although the success rate of Molteno implant surgery is favorable, most authors do not recommend this as an initial procedure because of the higher potential for complications.

Wiggins and Tomey noted that both trabeculectomies performed in their study were unsuccessful. Tra-
beculotomy, however, has been suggested as the preferred initial operation for uncontrolled glaucoma with aniridia. In a report of 12 eyes, 10 (83%) obtained IOP control after first (6 eyes) or second (4 eyes) trabeculotomy, with a mean follow-up of 9.5 years. If only eyes that had IOP controlled without medications were counted as surgical successes, only 25% (3 eyes) of the 12 eyes were successful. Trabeculotomy was possibly more successful in these patients because these eyes were in young patients (average age, 4 years 9 months) that, in general, did not show progressive angle changes. Of the 9 eyes that had gonioscopy, only 1 eye showed trabecular meshwork that was covered with iris stump. Of the 2 eyes that failed initial trabeculotomy, 1 eye was noted to have the later-onset type of glaucoma associated with progressive angle changes.

Therefore, a review of the literature on the surgical treatment of aniridic glaucoma reports poor control of IOP with argon laser trabeculoplasty, goniotomy, and trabeculectomy. Although cyclocryotherapy and Molteno implants may also be used, their higher rates of complications make these treatments a poor choice for initial therapy. Because aniridic glaucoma is difficult to treat surgically, we propose a prophylactic goniosurgery that decreases the risk of glaucoma in select aniridic patients with progressive angle changes.

Grant and Walton noted that aniridic patients with blockage of the trabecular meshwork by anterior extensions of iris tend to have glaucoma. At the suggestion of Drs Chandler, Grant, and Johnson, a program of prophylactic goniosurgery was initiated for young aniridic patients without glaucoma, and the favorable initial results in 16 children were reported. This care has continued, and the present report describes the results of this effort to lessen the risk of glaucoma in young aniridic patients.

New patients seen for care most frequently come to our office either at an early age for consultation in regard to the diagnosis and care of aniridic or later in childhood with glaucoma resistant to medical therapy. Patients with glaucoma consistently demonstrate obstruction of the trabecular meshwork with an abnormal iris-like tissue circumferentially.

When a young child is seen, the program of careful observation is initiated. During the first year of life, Koeppe gonioscopy is done in the office. During the first 18 months of life, the anterior chamber progressively deepens, which facilitates goniosurgery when necessary. After the first year of life, regular examinations under general anesthesia are performed to better assess the filtration angle conditions. Depending on the findings, such examinations are repeated every 3 to 9 months. If gonoscopic findings qualify a patient for prophylactic surgery, surgery should be done or an appropriate referral to an experienced goniotomy surgeon should be made.

On gonioscopy, striking circumferential asymmetry is often present. For example, goniosurgery has not been done superiorly since findings there have not dictated surgery. In any patient, the nasal and temporal angles may be quite different. Surgery is only done in nasal, temporal, or inferior quadrants where an obstructive angle abnormality is present (Figure 1).

It is thought that the iridotrabecular attachments cause progressive compromise of angle function. Because prophylactic goniosurgery frees the trabecular meshwork of iris attachments, trabecular meshwork damage is retarded or stopped. Although no patients who had prophylactic goniosurgery had reattachment or revascularization of iris tissue onto the trabecular meshwork, 3 patients still developed an increase in IOP at an average of 13 years after prophylactic goniosurgery. The exact mechanism of this angle compromise is unknown. Perhaps filtration function was critically compromised at the microscopic level, or perhaps previous iridotrabecular attachments had already caused irreversible damage to the trabecular meshwork.

Although this study strongly suggests that prophylactic goniosurgery prevents or delays the onset of aniridic glaucoma, this study is limited by lack of an ideal control group. A randomized evaluation of 2 groups of patients with identical criteria would be helpful to confirm the value of this procedure. However, in our study, since all patients who qualified for prophylactic goniosurgery wanted to have the surgery done, there was no control group of patients who qualified for prophylactic goniosurgery and who did not have the surgery done. Also,
patients who had glaucoma at initial consultation obviously were not eligible for this prophylactic surgery. It would have been interesting to know whether glaucoma could have been prevented in any of these patients had they been offered prophylactic surgery. Also, some patients in our study had a shorter follow-up period after prophylactic goniosurgery. Perhaps if they had been followed up longer, these patients would have developed glaucoma.

CONCLUSIONS

Selected aniridic eyes with early progressive angle changes that had prophylactic goniosurgery demonstrated a significantly lessened risk of glaucoma. There were no failures of IOP control. Aniridic glaucoma is difficult to control with medical treatment, trabeculotomy, goniotomy, and trabeculectomy. Surgical treatment with cyclocryotherapy and Molteno implants is associated with high complication rates. Trabeculotomy or even goniotomy may be helpful in young aniridic patients with glaucoma with no angle closure or only a small, low iris residual. Prophylactic goniosurgery for selected young children with progressive angle changes appears to be effective in preventing or delaying the onset of aniridic glaucoma. Without prophylactic goniosurgery, aniridic glaucoma may be expected in half of affected children and is extremely difficult to control.

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REFERENCES


From the Archives of the ARCHIVES

A look at the past . . .

In this paper Howe again makes a strong plea for legislation compelling the use of Crede’s method in public institutions, producing statistics showing how, by this means, the disease can be practically wiped out. A committee was appointed by the Society, of which Dr. Howe was made chairman, to report on the subject at the next meeting of the Society.