Long-term Outcome of Trabeculotomy for the Treatment of Developmental Glaucoma

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Objective: To elucidate long-term outcome of trabeculotomy in primary and secondary developmental glaucoma.

Methods: One hundred forty-nine eyes of 89 patients with developmental glaucoma who underwent trabeculotomy were retrospectively studied. Intraocular pressure (IOP), success probabilities, visual acuities, and visual field were determined during follow-up and at the final visit.

Results: The mean±SD IOP of 112 eyes with primary developmental glaucoma at the final visit with a mean±SD follow-up period of 9.5±7.1 years was 15.6±5.0 mm Hg. The average IOP for 37 eyes with secondary developmental glaucoma was 16.7±4.2 mm Hg. One hundred eyes (89.3%) with primary developmental glaucoma were defined as achieving success at the final visit. Complete and qualified successes were achieved in 71 eyes (63.4%) and 29 eyes (25.9%), respectively. Visual acuities were 20/40 or better in 78 (59.5%) of 131 eyes examined and were poorer than 20/200 in 32 eyes (24.4%). The causes of poor visual acuities were mainly progression of glaucoma, including delay of detection of onset or surgery and amblyopia. Eyes with glaucoma that existed before 2 months of age or eyes that needed several trabeculotomies were considered to have poor visual acuity. Visual fields were classified as normal or almost normal in 21 (44.7%) of 47 eyes.

Conclusions: Trabeculotomy for developmental glaucoma is effective over a long time. There is a fairly good prognosis for visual function of eyes with developmental glaucoma with early detection of the onset, proper treatment, and proper management after trabeculotomy.

These eyes were classified into 2 groups according to the classification of Hoskins et al. One hundred twelve eyes of 64 patients were classified as having primary developmental glaucoma that had no secondary findings, and 37 eyes of 26 patients were classified as having secondary developmental glaucoma associated with congenital anomalies in the other portion of the eye. In 1 girl, we saw aniridia in her right eye but no secondary findings in her left eye. We classified her right eye as having secondary glaucoma and her left eye as having primary developmental glaucoma. Eyes with primary developmental glaucoma were further divided into 3 groups based on the age at onset or, if it was not clear, age at the first visit to the eye clinic. Twenty-five eyes of 14 patients were classified as having congenital glaucoma (existing before 2 months of age), 65 eyes of 36 patients were classified as having infantile glaucoma (occurring from 2 months until 2 years of age), and 22 eyes of 14 patients were classified as having juvenile glaucoma (existing after 2 years of age). Congenital anomalies included Axenfeld-Rieger syndrome or Axenfeld anomalies in 15 eyes of 8 patients, Sturge-Weber syndrome in 10 eyes of 10 patients, and aniridia in 6 eyes of 4 patients, and 6 eyes of 4 patients were studied after pars plana lensectomy had been performed for congenital cataract.

EXAMINATION AND SURGICAL PROCEDURES

The IOP was measured by a Perkins hand-held applanation tonometer (MK2; Clement Clarke, London, England) while the patient was under general anesthesia or in a near-anesthetic state from triclofos sodium (Triclofrin; Glaxo Inc, London, England) and by the Goldmann slitlamp applanation tonometer, if possible. Angle structure was examined with a Koepp or Goldmann gonioscopic lens, if possible. The visual field was examined by the Goldmann perimetry or a Humphrey field analyzer (field analyzer 750; Zeiss-Humphrey Systems, Dublin, Calif). Axial length was measured with 1-dimensional mode ultrasonography (Echo Scan US-2000; Nidek, Aichi, Japan).

Surgical procedures were performed as described previously. Briefly, a conjunctival incision and a 4 × 4-mm scleral flap are created at the limbus. The external wall of the Schlemm canal is opened, and U-shaped probes are inserted into both sides of the canal and rotated into the anterior chamber against the trabecular meshwork. The scleral flap is closed with 10-0 nylon sutures until the wound becomes watertight. The conjunctival flap is then closed.

RESULTS

The study results are presented as mean±SD. The mean age at the first operation in the 64 patients with primary developmental glaucoma included in this study was 2.3±3.6 years (range, 0-15 years). During the follow-up period, 162 trabeculotomies were performed in eyes with primary developmental glaucoma. One trabeculotomy procedure was performed in 72 eyes, 2 procedures were performed in 30 eyes after an average period of 1.5±3.0 years, and 3 trabeculotomy procedures were performed in 10 eyes after an average period of 5.6±5.6 years. In 5 eyes that had uncontrolled IOPs even after the trabeculotomies, trabeculectomy with the use of antimetabolites or cyclotherapy was performed (2 eyes with infantile glaucoma, 3 eyes with juvenile glaucoma). These eyes were classified as failures at the point where trabeculectomy was required because of the uncontrolled IOP.

The mean age at the first operation in the total of 26 patients with secondary glaucoma in children who were included in this study was 2.3±3.3 years (range, 0-15 years). During the follow-up period, a total of 60 trabeculotomies were performed in eyes with secondary glaucoma. One trabeculotomy procedure was performed in 20 eyes, 2 procedures were performed in 11 eyes, and 3 trabeculotomy procedures were performed in 6 eyes. In 4 eyes that had uncontrolled IOPs even after trabeculotomies, trabeculectomies with the use of antimetabolites or cyclotherapy were performed (3 eyes with Axenfeld-Rieger syndrome and 1 eye with congenital cataract).

INTRAOCULAR PRESSURE

The mean IOP in primary developmental glaucoma before surgery was 29.3±8.6 mm Hg, and the mean number of antiglaucoma medications before surgery was 0.17±0.52 (Table 1). The mean IOP at the final visit was 15.6±5.0 mm Hg, and the mean number of medications was 0.63±1.0. The slight increase seen for the mean IOPs is partially due to the fact that we performed surgery soon after detection of the disease and before using drugs in many of the cases. At the final visit, the mean IOP of eyes with infantile glaucoma was lower than that for congenital (P=.01) and juvenile glaucoma (P=.001, Mann-Whitney U test). The mean IOP after trabeculotomy in primary developmental glaucoma ranged from 13.4 to 17.9 mm Hg during the 18 years of follow-up (Figure 1).

The mean IOP in secondary glaucoma before surgery was 26.9±8.3 mm Hg, and the mean number of antiglaucoma medications was 0.41±0.72 (Table 2). The mean IOP at the final visit was 16.7±4.2 mm Hg, and the mean number of medications was 0.94±1.07. Among the

Table 1. Average Intraocular Pressure (IOP) and Number of Medications Before and After Surgery for Primary Developmental Glaucoma

<table>
<thead>
<tr>
<th>Glaucoma Type</th>
<th>Eyes, No.</th>
<th>Follow-up Period,</th>
<th>Final Visit</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>IOP, Mean ± SD, mm Hg</td>
<td>Medications, No.</td>
<td>IOP, Mean ± SD, mm Hg</td>
</tr>
<tr>
<td>Congenital</td>
<td>25</td>
<td>32.0±11.6</td>
<td>9.6±6.3</td>
</tr>
<tr>
<td>Infantile</td>
<td>65</td>
<td>28.1±7.7</td>
<td>10.0±7.7</td>
</tr>
<tr>
<td>Juvenile</td>
<td>22</td>
<td>30.1±7.0</td>
<td>8.0±6.0</td>
</tr>
<tr>
<td>All Primary</td>
<td>112</td>
<td>29.3±8.6</td>
<td>9.5±7.1</td>
</tr>
</tbody>
</table>

*P<.05. †P<.01.
eyes with secondary glaucoma, the mean IOP of eyes with Sturge-Weber syndrome was higher than for eyes with congenital cataract ($P = .02$). The mean IOP after trabeculotomy in secondary glaucoma ranged from 15.4 to 21.5 mm Hg during the 18 years of follow-up (Figure 1). There was no significant difference in the average IOP at the final visit between primary and secondary developmental glaucoma ($P = .38$, Mann-Whitney U test).

**LIFE-TABLE ANALYSIS**

The procedure was regarded as a success in cases in which all of the following postoperative criteria were met: (1) IOP was less than 21 mm Hg with 1 or more trabeculotomies; (2) there were no other surgical treatments for glaucoma, such as trabeculectomy and cyclocryotherapy; (3) there was no progression of cupping of the optic nerve disc; and (4) there was no enlargement of the corneal diameter. The success probability was determined with the Kaplan-Meier method and then compared using the Breslow-Gehan-Wilcoxon test.

A life-table analysis showed that the success probabilities at 5, 10, and 20 years after the first trabeculotomy in primary developmental glaucoma were 94.3%±2.5%, 87.7%±3.9%, and 80.8%±6.1%, respectively (Figure 2). Among the eyes with primary developmental glaucoma, the success probabilities at 5, 10, and 15 years after the first trabeculotomy were 100%, 85.7%±9.4%, and 77.9%±11.3% in patients with congenital glaucoma, 96.6%±2.4% for all follow-up years in patients with infantile glaucoma, and 81.2%±9.8%, 67.7%±11.9%, and 67.7%±11.9% in patients with juvenile glaucoma, respectively (Figure 3). Patients with infantile glaucoma had a better prognosis than those with juvenile glaucoma for the outcome of trabeculotomy ($P = .005$).

At the final visit, 100 (89.3%) of 112 eyes with primary developmental glaucoma had IOPs less than 21
mm Hg, no progression of disc cupping, and no enlargement of the corneal diameter. Of these, in 71 eyes (63.4%) there was no medication administration (complete success), and in 29 eyes (25.9%) medications were required to control the IOP (qualified success). To achieve successful control, 67 eyes (59.8%) required 1 trabeculotomy, 21 eyes (18.8%) required 2 trabeculotomies, and 8 eyes (7.1%) required 3 trabeculotomies. Three eyes (2.7%) required trabeculectomy in addition to the trabeculotomies, and 1 eye (0.9%) required cyclocryotherapy. Eight eyes (7.1%) had IOPs of 21 mm Hg or more, progression of disc cupping, or worsening of the visual field but did not undergo further surgical treatment due to poor visual function in 4 eyes and an irregular corneal surface after penetrating keratoplasty for corneal opacity in 1 eye.

A life-table analysis showed that the success probabilities at 5, 10, and 20 years after the first trabeculotomy in secondary glaucoma were 82.2%±7.3%, 77.1%±8.5%, and 64.2%±13.7%, respectively (Figure 2). The success probabilities of trabeculotomy in primary developmental glaucoma were slightly higher than those in secondary developmental glaucoma (P=.047). At the final visit, 32 (86%) of 37 eyes with secondary glaucoma had IOPs less than 21 mm Hg, no progression of disc cupping, and no enlargement of the corneal diameter. Of these, in 15 eyes (40.5%) there was no administration of medicatations (complete success), and in 17 eyes (45.9%) medications were required to control the IOP (qualified success). To achieve successful control, 17 eyes (45.9%) required 1 trabeculotomy, 9 eyes (24.3%) required 2 trabeculotomies, and 4 eyes (10.8%) required 3 trabeculotomies. Two eyes (5.4%) required trabeculectomy in addition to the trabeculotomies. Four eyes (10.8%) had IOPs of 21 mm Hg or more, progression of disc cupping, or worsening of the visual field but did not undergo further surgical treatment due to poor visual function in 2 eyes and disagreement by the parents in 2 eyes. In 1 eye (2.7%), we could not judge the success because of phthisis after incurable retinal detachment.

**VISUAL ACUITY AND ORTHOPTIC STATUS**

Final visual acuity in 131 eyes, in which we could obtain reliable data, was 20/40 or better in 78 eyes (59.5%), 20/200 to 20/40 in 21 eyes (16.0%), and poorer than 20/200 in 32 eyes (24.4%) (Figure 4). There was no significant difference in the ratio between eyes with primary and secondary developmental glaucoma. In primary developmental glaucoma, eyes with congenital glaucoma had a poorer prognosis for visual acuity than eyes with infantile (P=.01) and juvenile glaucoma (P=.004, Mann-Whitney U test). The visual acuities of eyes in which the IOP was controlled by only 1 trabeculotomy were better than those for eyes controlled by 2 or 3 trabeculotomies (P<.001, Mann-Whitney U test). Of 23 unilateral cases, visual acuities for 20 eyes were available. In these eyes, 12 eyes (60%) had visual acuities of 20/40 or bet-
ter, and 8 eyes (40%) had visual acuities of less than 20/200. There was no significant difference in visual acuity prognosis between bilateral and unilateral cases ($P = .79$, Mann-Whitney $U$ test). The reasons for the poor visual acuity (<20/200) were progression of glaucoma (including delay of detection of onset or surgery) in 14 eyes, corneal opacity in 2 eyes, and retinal detachment in 4 eyes. In 12 eyes, amblyopia was the most likely cause of the poor visual acuity.

Refraction was estimated in 92 eyes with the use of a refractometer or skiascopy. Six eyes had more than a 6-diopter (D) spherical equivalent of myopia, 14 eyes had between 6 and 2 D of myopia, 38 eyes had between 2 and 0 D of myopia, 12 eyes had 0 D of hyperopia, 13 eyes had between 0 and 2 D of hyperopia, 3 eyes had between 2 and 6 D of hyperopia, and 6 aphakic eyes had more than 6 D of hyperopia. Seven patients had anisometropia greater than 2 D. Among this group, a unilateral decrease of vision was seen in 4 patients. Thirteen patients had exotropia, and 6 patients had esotropia. Eighteen eyes had nystagmus. Although patching was performed in 6 patients to prevent amblyopia, 4 patients had differences in the final visual acuity of both eyes.

**Table 3. Aulhorn Classification of Visual Fields**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-1</td>
<td>Relative small GVFD with an intensity of 0.6 log unit up to 1.0 log unit</td>
</tr>
<tr>
<td>1</td>
<td>Small GVFD with an intensity of more than 1.0 log unit</td>
</tr>
<tr>
<td>2</td>
<td>Incomplete NFBD</td>
</tr>
<tr>
<td>3</td>
<td>Complete NFBD or incomplete NFBD with nasal breakthrough</td>
</tr>
<tr>
<td>4</td>
<td>Complete NFBD with nasal breakthrough involving less than 1 quadrant</td>
</tr>
<tr>
<td>5</td>
<td>Complete NFBD with nasal breakthrough involving more than 1 quadrant</td>
</tr>
<tr>
<td>6</td>
<td>Temporal island</td>
</tr>
</tbody>
</table>

Abbreviations: GVFD, glaucomatous visual field defects; NFBD, nerve fiber bundle defect.

*As modified by Greve and Geijssen.*

**FINAL VISUAL FIELD**

Visual fields were able to be examined in 47 eyes by Goldmann perimeter or Humphrey perimeter and classified as described by the Aulhorn classification as modified by Greve and Geijssen (Table 3 and Figure 5A-C). Twenty-one eyes (44.7%) were classified as normal, stage 0 to 1, and stage 1 (almost normal), whereas 13 eyes (27.7%) were classified as stages 5 and 6 (advanced stage) (Figure 5D).

**CORNEA**

The horizontal corneal diameters before surgery in 130 eyes ranged from 9.5 to 15 mm (average, 12.4±1.3 mm), and those at the final visit ranged from 11 to 14.5 mm (average 12.9±0.95 mm). Eighty-eight eyes had corneal edema before surgery, and in 15 eyes the corneal opacity was so severe that the iris was hardly visible. Thirty-two eyes had Haab striae (Descemet membrane rupture). At the final visit, partial corneal opacity was seen in 10 eyes, and total corneal opacity was found in 1 eye.

The endothelial cells of the cornea at the final visit were counted in 25 eyes. The average was 2153±926/mm² (range, 664-4149/mm²). Two eyes had less than 1000/mm² but did not have corneal edema. The larger the corneal diameter was before surgery, the fewer the number of endothelial cells seen at the final visit ($p = -0.581$, Spearman rank correlation coefficient). The average for the eyes with Haab striae was 1239±364/mm², and the numbers were fewer than that seen without Haab striae (2763±799/mm²; $P < .001$, Mann-Whitney $U$ test).

**AXIAL LENGTH**

Axial length was measured in 30 eyes before surgery. The average was 21.9±1.9 mm (range, 19.4-27.4 mm). Compared with average axial length of healthy children at each age, 21 eyes (70%) were outside the average ±2 SDs, which means those eyes were significantly larger than the

![Figure 5. Final visual fields in 47 eyes were classified as described by the Aulhorn classification as modified by Greve and Geijssen.* A-C, Sample fields that were classified as stage 4 (A), stage 5 (B), and stage 6 (C). D, Distribution of the visual field.](image-url)
Developmental glaucoma is caused by malformation of the anterior chamber angle or damage to the outflow system caused by maldevelopment of some other portion of the eye.\(^1\) To reduce the outflow resistance, surgical procedures are regarded as a necessary treatment. In our large series, we have demonstrated that trabeculotomy for the treatment of developmental glaucoma is safe and effective over a long duration. The visual function of eyes with developmental glaucoma is deemed to be fair with early detection of onset, proper treatment, good control of IOP, and proper management for amblyopia.

Among 112 eyes with primary developmental glaucoma, the average IOP at the final visit with an average follow-up period of 9.5±7.1 years was 15.6±5.0 mm Hg. At the final visit, trabeculotomies in 100 (89.3%) of the 112 eyes were defined as successes (IOP less than 21 mm Hg, no progression of cupping of the optic disc, and no enlargement of the corneal diameter). Complete success was achieved in 71 eyes (63.4%), and qualified success was achieved in 29 eyes (25.9%). McPherson and Berry\(^3\) have reported that 22 (96%) of 23 eyes were controlled under 21 mm Hg at the final visit (mean follow-up, 5.6 years). In a study by Quigley,\(^6\) it was revealed that 17 (80%) of 22 eyes achieved success after 1 year, and a study by Meyer et al\(^7\) found that 31 (79%) of 39 eyes had IOPs less than 21 mm Hg at the end of the follow-up (mean, 24.7 months). When considering the long period of follow-ups, our results are comparable to those in the other studies. To achieve successful control, 67 eyes (59.8%) needed 1 trabeculotomy, 21 eyes (18.8%) needed 2 trabeculotomies, and 8 eyes (7.1%) needed 3 trabeculotomies. Because our past experience has demonstrated that additional trabeculotomy is also effective in lowering IOPs, we usually conduct second (or third) trabeculotomies in eyes with uncontrolled IOPs even after the first unsuccessful trabeculotomy. A life-table analysis showed that the success probabilities at 20 years after the first trabeculotomy in eyes with primary developmental glaucoma were 80.8%. Eyes with infantile glaucoma had the best success rates and mean IOPs at the final visit.

Among 37 eyes with secondary developmental glaucoma, the average IOP at the final visit with an average follow-up period of 8.1±5.4 years was 16.7±4.2 mm Hg. A life-table analysis showed that the success probability at 20 years after the first trabeculotomy in eyes with secondary glaucoma was 64.2%. Eyes with Sturge-Weber syndrome had the worst success rates and mean IOPs. Although there was no statistically significant difference between the average IOP of primary and secondary glaucomas, the success probabilities of trabeculotomy in primary developmental glaucoma were slightly higher than those in secondary glaucoma (\(P=0.047\)).

Mandal et al\(^10\) reported the effectiveness of trabeculectomy with mitomycin in refractory congenital glaucoma after failed trabeculectomies. In our follow-up, the IOP in 2 eyes with infantile glaucoma, 3 eyes with juvenile glaucoma, 3 eyes with Axenfeld-Rieger syndrome, and 1 eye with congenital cataract were not able to be controlled by trabeculectomies and needed further trabeculectomy and/or cyclocryotherapy. Although 5 of these severe and refractory cases were controlled with several trabeculectomies with mitomycin and/or cyclocryotherapy treatments, the others did not achieve a stable success state even with several applications of these procedures. The reasons for failure in these cases seemed to be difficulties with the adjunctive treatments such as massage, vigorous wound healing even with antimetabolites, and scarring of the conjunctiva partly due to prior surgical procedures.\(^11,12\)

Richardson et al\(^13\) reported that 34 (39%) of 88 eyes with congenital glaucoma were well controlled by goniotomy, with the maintenance of good vision at greater than 20/50, and 35 (40%) of 88 eyes had vision poorer than 20/200. In a study by Morgan et al,\(^14,15\) 7 (58%) of 12 eyes had visual acuities of 20/50 or better. Our rate of 78 (60%) of 131 eyes with visual acuities of 20/40 or better compares favorably with their series. As in our former studies\(^13,15\) and that reported in other studies,\(^16\) eyes that had congenital glaucoma existing before the age of 2 months had a poorer prognosis than in the other groups. As for the number of operations performed, visual prognosis was better in eyes controlled with fewer trabeculectomies.
Amblyopia might be a major obstacle to overcome in developmental glaucoma and is due to a combination of strabismus, corneal scarring, and anisometropia. In our series, amblyopia might be the cause of poor vision in 12 eyes. Rice has reported that all of his patients improved their visual acuity in the amblyopic eye with use of the occlusion regimen. In another study, Biglan and Hiles reported that half (6 of 12) of the patients improved their visual acuity with the use of occlusion therapy, although the others did not. In our series, some of the patients exhibited an improvement in the visual acuity in the amblyopic eye after their use of occlusion therapy. However, we were unable to elucidate the effectiveness of complete optical correction of refractive errors and intensive therapy for the prevention of amblyopia. Thus, further studies on this important issue are required.

Although complications of the trabeculotomies were not serious, it is noteworthy that retinal detachment occurred in 5 eyes (3.4%) with developmental glaucoma during the follow-up. This ratio seems to be high. The cause of the retinal detachment is thought to be due to an enlarged globe related to the high IOP. For example, the corneal diameter of one detached eye before surgery was 13.5 mm, and the axial length of another eye before surgery was 27.5 mm. Four of the eyes had poor visual function due to incurable retinal detachment. We believe that careful fundus examination of eyes with developmental glaucoma is necessary during the follow-up periods.

Because most patients with developmental glaucoma are too young to be examined, prognosis of the visual field has seldom been reported. In our study 21 (45%) of 47 eyes examined had normal or almost normal visual fields, whereas 13 (28%) of 47 of the eyes were at an advanced stage, most of which could be attributed to a delay in the diagnosis.

In our series, the cup-disc ratio in 19 eyes got smaller after successful surgery, similar to that seen in other studies. Quigley reported that the responses of the optic head in infants were more elastic to elevations and normalization of IOP due to the relative lack of collagen in the connective tissues. However, reversal of observed cup size enlargement did not occur once the nerve fibers were damaged by long-term exposure to high IOP.

In conclusion, our study shows that trabeculotomy is a safe and effective treatment for primary developmental glaucoma. Also, it is effective in lowering IOPs in infantile and juvenile patients with secondary glaucoma. Although useful visual functions may remain after successful surgical treatments, more vigorous management for the prevention of amblyopia needs to be considered.

Submitted for publication April 21, 2003; final revision received January 15, 2004; accepted January 15, 2004.

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