PROGNOSIS OF GONIOTOMY IN PRIMARY INFANTILE GLAUCOMA (TRABECULODYSGENESIS)

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THE EARLIER THAT ANY GLAUCOMA IS DIAGNOSED AND BROUGHT UNDER CONTROL, the better the prognosis. This is particularly true of primary congenital (infantile) glaucoma. Although an occasional spontaneous remission or cure may occur, most of these infants become blind unless successful surgery is performed. Statistics on the success rate of surgery are usually unreliable for several reasons. In the world literature there is no clearly acceptable classification of the congenital glaucomas. Statistics usually include various unrelated congenital anomalies, operated upon at varying ages, and often in small numbers.

The developmental glaucomas are separated into three general groups in the Hoskins' Classification.[1] These anomalies involve the three anatomic structures of the anterior segment, the iris, the trabeculum and the cornea. Iridotrabecular dysgenesis includes aniridia and familial hypoplasia of the iris. Iridocorneotrabeculodysgenesis includes such anomalies as Axenfeld's, Rieger's and Peter's. Trabeculodysgenesis alone refers to primary infantile glaucoma. It is this group of patients which should be treated primarily by goniotomy.

SURGICAL COMPLICATIONS OF GONIOTOMY

The surgical complications of goniotomy in competent hands have been minimal. The operation is safer, easier and less traumatic than trabeculotomy and is equally effective in appropriate cases. As shown in Table I, in 577 consecutive goniotomies there were 13 ocular complications, an incidence of 2%. Only one of these resulted in loss of vision in an eye which developed a blood-stained cornea. There were six cardiopulmonary arrests, all of which recovered. This led to our present policy of performing bilateral goniotomy in appropriate cases rather than risking two anesthetics in these tiny patients.

TABLE I: COMPLICATIONS IN 577 CONSECUTIVE GONIOTOMIES

Hemorrhage	2 severe, one lost vision
Iridodialysis	4 small, without sequelae
Cyclodialysis	2 small, without sequelae
Shallow chamber	5 lasting one to two days
Lens injury, infection	None
Cardiopulmonary arrests	6 all recovered

PRESSURE CONTROL AFTER GONIOTOMY

We have followed 287 eyes long enough to be quite certain of the success or failure of the operation. The glaucoma was considered cured if the intraocular pressure (IOP) remained below 20 mm Hg without medication for at least six months and cupping of the optic nerve was either the same or improved. Overall, the success rate was 76.7%. However, when signs and symptoms of glaucoma were present at birth or over the age of 24 months, the success rate was close to 30%. In contrast, one or two goniotomies cured 94% of the cases diagnosed between the ages of 1 and 24 months. Many of the cases present at birth could have been diagnosed as iridotrabecular dysgenesis as many of them have hypoplasia and often increased vascularity of the iris. Such eyes always have a poor prognosis. Those over the age of 24 months are known to have less cellularity and more collagenous tissue according to Alvarado et al.[2] Such changes may be responsible for the decreasing effectiveness of goniotomy.

In patients in whom the symptoms appeared between the 1st and the 24th month and whose examination revealed trabeculodysgenesis, we would always perform two or three goniotomies before resorting to more drastic surgery. In eyes diagnosed at birth or after 24 months, if the corneas are reasonably clear, one goniotomy would be tried before resorting to trabeculotomy (Table II).

LONG-TERM PROGNOSIS OF GONIOTOMY

The mobility of a young population makes a long-term follow-up extremely difficult. In general, patients which were not controlled were returned to our care. Those with a favorable surgical result were not seen again. When follow-up letters were sent to the referring ophthalmologist,

TABLE II: EFFICACY OF 287 GONIOTOMIES BY AGE

	BIRTH TO 1 MONTH	1 TO 24 MONTHS	OVER 24 MONTHS
No. of cases	50	205	32
One goniotomy	8 (16%)	129 (75%)	11 (35%)
Two goniotomies	5 (10%)	66 (19%)	1 (3%)
	26%	94%	38%
Multiple operations (various types)	37 (74%)	10 (6%)	20 (64%)

quite frequently the family had moved and left no forwarding address. Satisfactory follow-up was possible in 82 eyes which had had successful goniotomies with IOPs between 10 and 20 mm Hg without therapy. Thirty-two of these eyes had been followed between 5 and 15 years. None of them had had complications. Fifty had been followed between 15 and 25 years.

Some complications began to appear after the 15th year (Table III). Three had an increase in IOP into the mid-20s to 30s, requiring medical therapy. Two had mild endothelial dystrophy with corneal edema; two developed neovascular glaucoma following trauma; and two had retinal detachments. No change in disc cupping occurred. Most had full visual fields except those with severely damaged optic nerves at the time of operation.

Unfortunately, the visual results have been less favorable. Visual acuity was determined in 52 eyes. Twenty-eight had vision of 20/20 to 20/40; 11, 20/50 to 20/200; and 13, less than 20/200. The decreased vision was caused by breaks in Descemet's membrane in the visual axis and by amblyopia due to anisometropia and marked astigmatism. Patching of these eyes was generally unsuccessful.

There are few patients who have had successful goniotomies who have reached the third or fourth decades of life, so we still cannot be sure of the long-term prognosis of their seemingly cured glaucoma. There is no doubt that periodic re-examination of all cases of developmental glaucoma is necessary if unsuspected loss of sight is to be avoided.

TABLE III: 52 SUCCESSFUL GONIOTOMIES (IOP 10 TO 20 mm Hg) COMPLICATIONS NOTED AFTER 15 TO 25 YEARS

Amblyopia (vision 20/50 or less)	20	46%
IOP 24 to 32 mm Hg after 18 years	3	6%
Endothelial dystrophy after 17 years	2	4.6%
Retinal detachment after 20 years	2	4.6%
Visual loss from trauma after 20 years	2	4.6%

SUMMARY

In developmental glaucoma with trabeculodysgenesis (primary infantile glaucoma), goniotomy is a safe and highly successful operation and should be used in preference to trabeculotomy. Prognosis varies markedly with the age of onset of signs and symptoms. When present at birth or after the age of two years, the success rate is only about 30%. Between the ages of 1 and 24 months, success following one or two goniotomies is 94%.

Eyes with successful goniotomies have now been followed for over 25 years with maintenance of pressure control without medication in most cases. The major complication has been amblyopia due to breaks in Descemet's membrane in the visual axis and to marked astigmatism and anisometropia. After 15 years, two cases developed an endothelial dystrophy and three had an IOP elevation. Obviously, periodic examination of such eyes will be necessary for life.

REFERENCES

1. Hoskins H, Hetherington J, Shaffer R, et al: Developmental glaucomas: diagnosis and classification. *Symposium on Glaucoma*. Tr New Orleans Acad of Ophthalmology. St Louis, CV Mosby Co, 1981, p 172.
2. Alvarado J, Murphy C, Polansky J, et al: Age-related changes in trabecular meshwork cellularity. *Invest Ophthalmol* 1981; 21:714-727.

DISCUSSION

DR MARSHALL M. PARKS. This past year the results of my last 20 years' experience with goniotomy used to treat congenital glaucoma were published. (*Am J Ophthalmol* 1981; 91:566-572). Although the number of my cases are less than Doctor Shaffer's, 50 eyes in 34 patients, the results of normalizing the intraocular pressure with goniotomy were practically identical to his.

An overlooked feature in the management of congenital glaucoma is amblyopia. The two causes are blurred images resulting from corneal edema and Haab stria and anisometropia secondary to induced myopia caused by the increased pressure stretching the infant's globe. Fortunately, following normalization of the intraocular pressure, the myopia tends to decrease. Among the 50 eyes the myopia decreased from a preoperative average of 6.25 diopters to 4.25 diopters six months after surgery.

Frequent repeat cycloplegic refractions starting as soon as the ocular media is sufficiently clear to permit frequent adjustments in lens power as the anisometropia changes, and appropriate occlusion therapy starting as soon as amblyopia is recognized become the second step in the management of congenital glaucoma. The second step must be pursued as aggressively as the first step which normalizes the intraocular pressure, if two good seeing eyes are to result.

DR PAUL R. LICHTER. I would like to congratulate Doctor Shaffer on an excellent paper and for giving us this huge volume of patients to indicate the great success of goniotomy. I would like to ask him about cases of 0 to 1 month of age. It is my experience and probably most of us who do congenital glaucoma that those cases often have cloudy corneas at the time of birth, often making it difficult or impossible to see into the angle. I wonder if he looked at those cases with regard to the status of the cornea as to whether he thought that a perfect goniotomy was obtained such that it is obtained in those patients with corneas that are clear or can be made clear by removing the epithelium? Also, I would like to ask him with regard to the particular time he operates those patients. Again, patients who have glaucoma in utero have had a chance to have severe damage to the disc and to the cornea at the time of birth and if there is delay in referring those patients and getting them operated upon, this may certainly contribute to the poor results of this group.

DR ROBERT SHAFFER. The iris hypoplasia with prominent radial iris vessels is commonly seen in blue-eyed infants. These vessels are more engorged when the pressure is increased. The abnormal vessels we were describing are not radial vessels. They meander over the peripheral iris surface like a neovascularization. The stroma is always hypoplastic with absence of the collarette. The prognosis in such cases is very poor.

Since these infant eyeballs are quite elastic, the reduction in myopia pointed out by Doctor Parks is logical. Doctor Sampaolesi in Buenos Aires emphasizes ultrasonography in these cases. If the pressure is not controlled, he thinks an increase in size can be shown by ultrasonography.

Doctor Hoskins is beginning to think that early goniotomy in the first week may improve the chances of cure in these difficult cases. The numbers are much too small to be sure. By preventing deprivation amblyopia, Doctor Parks is able to give 80% of these cases good vision. Our success has not been quite so good, so we must try harder.

I appreciate the comments of all the discussants.