underscore the importance of careful comparative ophthalmoscopy in infantile glaucoma.

REFERENCES

Discussion

Dr. Daniel Snyder. Daniel Snyder, M.D., agreed with the publication that asymmetry of optic discs was most unusual. When present at any age, it should lead the ophthalmologist to most careful examinations to determine the presence or absence of glaucoma. John Hetherington, M.D., reported the observation that the size of a glaucomatous cup can become much smaller after normalization of pressure in an infant with glaucoma.

Principles and problems of therapy in congenital glaucoma

Joseph Haas

It is proper and fitting to begin this discussion by acknowledging the immense debt of infants with congenital glaucoma and their ophthalmologists to the late Dr. Otto Barkan. In spite of the problems that will be discussed, his application of the goniotomy operation has favorably reversed the prognosis of this disease.

Suspicion of congenital glaucoma may result from the symptoms of epiphora, photophobia, and blepharospasm, but unfortunately the majority of cases are seen when the mother notices a sudden clouding of the cornea in eyes which were previously considered big and beautiful. The infant is then referred to the ophthalmologist for further care.

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At the initial consultation it is unusual to be able to conduct more than a cursory examination, but as much should be accomplished as possible. Rarely in small infants can a satisfactory presumptive diagnosis be made by external examination, tonometry, and even gonioscopy and ophthalmoscopy. Since this is usually impossible, nevertheless, it affords an excellent opportunity to obtain a comprehensive history relative to the pregnancy with emphasis on rubella, a good family history, and the knowledge of whether associated congenital defects are present. At this point, the ophthalmologist must skillfully blend the art of medicine with the science of ophthalmology, for in addition to the eye in question, there are widespread social ramifications involved.
Primarily, treatment is required for the parents of the child. They are usually quite young, and often emotionally and economically ill-equipped to cope with the problems that have suddenly and dramatically occurred. Their guilt problems must be assuaged, particularly if a family history of glaucoma exists.

Then an attempt should be made to familiarize the parents with the protracted nature of the illness, the prognosis, the frequent necessity for repeated surgery, and the life-long necessity for continued examinations. The parents should be familiarized with the various agencies that will afford financial assistance when necessary, for it is difficult to obtain retroactive help. Time and effort well-spent will reward the ophthalmologist many times over at a later date, particularly when medical or surgical progress is not going well.

Eventually, the social aspects of the glaucomatous child will require repeated counseling. His poor vision will need explanation to the parents and schools to avoid excessive dependence, he may require the help of the visual aids, his cosmetic blemishes will need correction where possible, and even his selection of a mate may require advice, for visually handicapped individuals often practice nonran-dom mating simply on the basis of geographical proximity, to start the sequence all over again.

After the ophthalmologist has explored the social aspects of congenital glaucoma, he then must concentrate on the medical and surgical aspects of the disease. The initial step is the admission of the infant to the hospital for examination under general anesthesia.

Preoperatively, the infant should have a general physical examination plus blood count, hemoglobin tests, and urinalysis. The urine should be tested for amino acids, as well as mucopolysaccharides. If a history of rubella exists, throat and nasal cultures should be obtained as well as determining the antibody titer of the mother. When there are no physical contraindications, the patient is taken to the operating room for examination under general anesthesia.

Diagnosis

The following steps are conducted and recorded.

1. The corneal diameter is measured horizontally with a millimeter caliper. When the horizontal end points are obscure, a vertical measurement should be included. The average corneal diameter at birth is approximately 10 mm., but it grows rapidly, reaching a measurement of 12 mm. at one year of age.

In Fig. 1, an attempt to correlate corneal diameter with initial intraocular pressure has been

![Fig. 1. The intraocular pressure of 181 eyes under miotic therapy for infantile glaucoma. The rarity of control is obvious. The corneal diameters are scattered throughout the entire range.](http://iovs.arvojournals.org/pdfaccess.ashx?url=data/journals/iovs/933000/)
made without an obvious relationship. Although these intraocular pressures have been altered by the use of miotics, it appears that the increased corneal diameter is not a function of the intraocular pressure per se.

However, in Fig. 2, the corneal diameter when compared with the age of the infant at the time of the diagnosis, demonstrates a closer relationship. These charts suggest that in the presence of elevated tension the increased corneal diameter represents a sign of elapsed time. Thus, a rapidly increasing corneal diameter, or 1 over 12 mm., should arouse suspicion of glaucoma. Megalocornea, an inherited corneal enlargement, without associated signs of glaucoma may rarely cause confusion.10

II. Corneal cloudiness is the commonest presenting complaint of infantile glaucoma. This usually does not represent neglect, for clouding of the cornea in infantile glaucoma is not the result of age, but of the intraocular pressure (Figs. 3 and 4).

Haab's striae, which represent tears in Descemet's membrane, occur as a more localized and dense form of opacity, but they do not seem to occur in the corneas less than 12.5 mm. in diameter.

Corneal clouding may occur without raised intraocular pressure in Hurler’s disease4 (mucopolysaccharides present in urinalysis), trauma, and a few rarer dystrophic conditions. It may occur in rubella syndrome with or without pressure.5-7 Finally, clouding may occur not only in primary infantile glaucoma, but in all of the secondary infantile glaucomas as well.

III. The lens may reveal subluxation in Marfan’s and Marchesani’s syndrome, as well as in homocystinuria.8 Cataract is commonly associated with
Fig. 4. The relation of patient's age at the time of diagnosis, to the intraocular pressure without treatment. The elevated pressures are similar and correspond to the degree of corneal clouding (Fig. 3).

IV. The intraocular pressure of normal infants is uncertain because it usually cannot be measured without the use of general anesthesia. It is apparently similar, or slightly higher, than the normal adult. Kornbleuth10 found the average value during the first year of life to be 20.9 mm. Hg with a standard deviation of 2.8 mm. Hg. These findings were obtained under diethyl ether anesthesia. Thus, the upper limits of normal for infants, under diethyl ether, should be about 27.5 mm. Hg on the 1955 scale.

Fig. 4 reveals the initial untreated intraocular pressure in 75 patients with definite infantile glaucoma. It is apparent that pressures with early onset glaucoma are as high as those of later onset (Fig. 4).

With popularity of other anesthetic agents after World War II, initial tonometric readings found in infantile glaucoma have tended to be less constant than those shown in Fig. 4, reducing the significance of intraocular pressure readings. In general, the effect of the anesthetic agent upon the intraocular pressure tends to parallel their effect on the cardiovascular tonus. Hence, Fluothane (halothane)11 tends to lower the pressure, and cyclopropane tends to elevate the pressure. In addition, succinylcholine12 temporarily elevates the pressure. Obviously then, standardization of anesthesia for diagnosis and follow-up of congenital glaucoma is highly desirable, and inconsistent readings should be evaluated in light of the patients general state, as well as the agent in use.

It is of interest that in a small group, comparison of applanation tonometry with indentation tonometry has to date been unrewarding. Tonography has been of almost no help.

V. The angle of the anterior chamber characteristically reveals a flat insertion of the iris plane to the ciliary body. The insertion rarely is high enough to obscure a gonioscopic view of the functioning portion of the trabeculum, but frequently one can visualize a mesodermal veil extending from the root of the iris along the angle wall.

Commonly, the iris vessels are hyperemic, and their branches may run tortuously through the angle sulcus. Through the iris stroma, pigmented arcades are seen.

When the corneal diameter is not over 14 mm., the canal of Schlemm may spontaneously fill with blood, or this phenomenon may be produced by compression of the jugular veins. With the canal of Schlemm filled with blood, the flat insertion of the iris does not seem high enough to block the outflow.

Although many patients with infantile glaucoma do not appear to have a diagnostic angle deformity, in unilateral cases a decided difference in iris insertion, as well as mesodermal content of the angle is seen.

VI. The optic nerve should be drawn and the relative size of the optic cup recorded. Surprisingly, the incidence of glaucomatous cupping in my patients has exceeded 50 per cent at the time of initial examination.

The rest of the fundus should be examined for tumor, retinitis, or associated congenital anomalies. When the corneal irregularities obscure the fundus, it is quite helpful to view it through the goniolens.

Treatment

Upon completion of the examination, a decision as to the type of treatment is made. Since the systemic effects of anesthesia make reliance upon numerical values of tonometry or tonography uncertain, this decision is based upon the entire clinical picture, including symptomatology, the physical findings, and the results of tonometry. If there is any doubt as to the condition or indicated treatment, continued observation should be substituted for active therapy.

It is generally accepted that in infantile glaucoma, surgery is the treatment of choice (Fig. 1). It is also generally accepted that angle surgery produces the highest incidence of intraocular pressure normalization.3, 7, 11 Therefore, when the diagnosis is certain and the pressure elevated, one should proceed with angle surgery at the completion of the examination.

The exception to this rule, in the writer's opinion, occurs when the corneal clouding precludes adequate examination of the eye. Here, it...
has been my practice to treat the eye with miotics (pilocarpine, 2 per cent every 6 hours) plus Diamox (5 to 10 mg per pound in divided doses) until visualization is either adequate or not improving. In the patient with rubella, it appears helpful to procrastinate as long as possible.

Because of the known advantages of a nonfiltering result, the writer's preference for initial surgery is the goniotomy operation with Barkan's technique. If two properly placed goniotomy operations fail, this operation is combined with the goniopuncture operation of Scheie. The combined operation of goniotomy and gonio puncture is also used initially when corneal clouding precludes the use of the surgical goniolens.

The most frequent complication of angle surgery is hyphema, which usually is innocuous. However, rarely it may fill the entire chamber and cause blood staining earlier than in the usual situation because of the pre-existing corneal damage. Early aspiration and lavage are therefore indicated.

Postoperative medication in the goniotomy operation is governed by the response of the eye to surgery. When surgery is successful, there is usually a rapid subsidence of symptoms, as well as return of the corneal luster. When this occurs, nothing locally is needed. If the presenting symptoms persist, the antiglaucomatous medication should be reinstituted, or if evidence of inflammation occurs, the local anti-inflammatory drugs should be used.

Approximately 3 weeks after the operation, the patient is re-examined under general anesthesia. When surgery has been unsuccessful, it should be repeated according to the principles previously outlined. When the entire available angle has been utilized without success, filtration is considered.

The most successful filtration operation reported is scleral cautery with peripheral iridectomy, which controlled the intraocular pressure in 54 per cent, but with a much higher rate of complication than with angle surgery.

When pressure normalization is obtained, the child is then re-examined in 6 weeks, and if still normalized, 12 weeks later. It does not appear that intervals longer than 4 months should be utilized, for recurrence of the glaucoma may occur rapidly and with fewer than original symptoms.

When the patient reaches a cooperative age (usually about 4 years), examination can be performed in the office quite satisfactorily.

Results

Intraocular pressure. Approximately 80 per cent of eyes operated upon by goniotomy will have normalized intraocular pressure. An additional percentage will be added by the use of filtering operations.

Of 69 eyes, in which the onset of glaucoma occurred before the third month after birth, the total percentage of cure was only 55. Of greater significance is the fact that it required 149 goniotomies to normalize tensions in the eyes of these 38 patients.

When the onset occurred, after the second month, 97 per cent were normalized, and the incidence of success per goniotomy had risen to 72 per cent. Whether this distinction merely represents a difference in the severity of the disease or actually represents a different disease, remains to be seen (Table I).

Once successfully normalized for one year, the recurrence rate is about 10 per cent according to Barkan. Shaffer reports a group of 88 eyes in which 92 per cent had maintained tensions below 24 mm. Hg for from 5 to 26 years. Of considerable etiological interest is the report that some patients who, as children, had successful operations for monocular glaucoma, developed open angle glaucoma in the uninvolved eye 20 years later.

The writer's recurrence rate after one year is higher than either of the above quoted figures, averaging 18 per cent. Although angle surgery is indicated when sufficient area remains, it is less likely to succeed than primarily, and this decrease in the success rate appears more pronounced as the patient's age increases. Filtration then becomes increasingly neces-

<table>
<thead>
<tr>
<th>Age at onset</th>
<th>No. of eyes</th>
<th>No. normalized</th>
<th>% normalized</th>
<th>No. goniotomies to normalize</th>
<th>% success per goniotomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>From birth through 2 months</td>
<td>69</td>
<td>38</td>
<td>55</td>
<td>149</td>
<td>25</td>
</tr>
<tr>
<td>From 2 months through 4 months</td>
<td>38</td>
<td>36</td>
<td>97</td>
<td>50</td>
<td>72</td>
</tr>
<tr>
<td>From 2 months through 7 months</td>
<td>58</td>
<td>53</td>
<td>91</td>
<td>77</td>
<td>69</td>
</tr>
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sary, but fortunately is better tolerated as the patient becomes older.

It is obvious that these patients' intraocular pressures require close supervision for the rest of their lives.

**Visual results.** In spite of the control of intraocular pressure in greater than 80 per cent of eyes with infantile glaucoma, only 39 per cent of my patients, over the age of 5 years, have vision correctible to better than 20/50. This corresponds almost identically with those previously reported by Shaffer15 (Fig. 5).

Further analysis of Shaffer's group of 60

![Graph showing visual results of successful operation on eyes with infantile glaucoma.](http://iovs.arvojournals.org/pdfaccess.ashx?url=/data/journals/iovs/933000/)

Fig. 5. Analysis of visual results of successful operation on eyes with infantile glaucoma.

per cent with poor visual acuity, revealed that 65 per cent were due to a marked anisometropia of at least 2.0 D sphere or 1.5 D of cylinder. Fifty-eight per cent of these anisometropic patients had a heterotropia, usually an exotropia. Obviously then, even before the age at which subjective testing is possible, if a heterotropia develops, occlusive therapy should be instituted (Fig. 6).

The unilateral infantile glaucoma in my patients constitutes a different situation, for here the anisometropia is usually excessive, and I have no patients in which the affected eye has better than 20/50. Furthermore, in no instance have I been able to successfully maintain occlusion in a child with one normal eye and one enlarged eye.

Thus, in addition to constant vigilance regarding the intraocular pressure, the ophthalmologist must remain alerted to the possible prevention of amblyopia.

**Sociological results.** It is often reported that repeated exposure to hospitals, operating rooms, anesthetics, and doctors have severe psychological results. Also, when one considers the severe visual handicaps present in these children, one would anticipate an even poorer over-all result.

In my experience, with the exception of those children with either associated organic brain damage or complete parental rejection, I have been delighted with their progress. It should be emphasized that their ability to overcome their limited vision is remarkable, and usually results in their ability to live a normal life. Even when blind, the majority of these children do well.

**Summary**

1. A detailed summary of the principles of the treatment of congenital glaucoma has been attempted.
2. The diagnostic findings under general anesthesia have been evaluated.
3. The results of treatment are discussed.

**REFERENCES**


(End of Symposium)