it, and the outflow mechanism is under α-adrenergic influence. Further studies will be needed to confirm these ideas. From the clinical point of view, adrenaline, either alone or in combination with an adrenergic potentiating agent, is still the most useful drug in the group.

We should like to acknowledge assistance by members of the Glaucoma Unit, Moorfields Eye Hospital, High Holborn, and the Frost Trust for a grant in support of the work.

COMMENTARY

ADRENALINE AND GUANETHIDINE COMBINATIONS IN GLAUCOMA

Professor Drance commented that the use of a combination of adrenaline and guanethidine might be of great importance in reducing the side-effects due to adrenaline alone. He noted that the results from his own series were different from those reported by Dr. Paterson and Mr. Miller. In Professor Drance’s study the patients had been pretreated with guanethidine only for 3 days and were then studied with the instillation of a single drop of adrenaline. This was a single-dose response study, whereas in Dr. Paterson’s study the guanethidine was used for prolonged periods so that the sympathetic denervation had occurred which was obviously not complete in his own shorter studies.

Punctate epitheliopathy did occur on occasion with guanethidine therapy but was much more common when the 10 per cent. solution was used rather than the 5 per cent. or even 2 per cent. solution now in common use. The epitheliopathy always disappears if the drugs are discontinued.

Management of infantile glaucoma

N. S. C. RICE

London

The objectives in the continuing management of children with congenital glaucoma are twofold: namely, evaluation of control of the glaucoma and prevention of amblyopia; they are of equal importance.

EVALUATION OF CONTROL OF GLAUCOMA

Anaesthesia

To obtain the data needed to evaluate the control of glaucoma in infants it is necessary to make regular examinations under general anaesthesia. It had been our practice to use ether anaesthesia for this purpose, the advantages being that it appears to have a reasonably consistent effect on the intraocular pressure and that it is safe in infants. However, repeated inhalation anaesthesia in children does have important disadvantages, not the least of which are the psychological effects. Recently we have been using ketamine hydrochloride (Ketelar: Parke-Davies) for the routine examination of children with congenital glaucoma, and we have found it to have a number of advantages over ether anaesthesia. Ketamine hydrochloride is a potent analgesic agent. When given by intra-
Management of infantile glaucoma

muscular injection it is effective within 10 minutes; its action is short, lasting about 30 minutes, although if the children are left undisturbed they will usually sleep for a few hours. The pharyngeal and laryngeal reflexes are retained and there is minimal, if any, depression of respiration so that endotracheal intubation is not necessary. Ketamine hydrochloride usually causes transient elevation of the systemic arterial blood pressure and this may be reflected by a slight rise in intraocular pressure. It is not possible to perform intraocular surgery with ketamine hydrochloride alone but conventional anaesthetic agents can be used after its administration; however, a few children have exhibited respiratory depression under these conditions. Adults given ketamine hydrochloride can experience disturbing dreams but, as far as can be judged, this does not occur with children. It has been our experience that ketamine hydrochloride offers a very satisfactory method for the regular examination of children with congenital glaucoma.

Intraocular pressure

It is of course important to measure the intraocular pressure by tonometry since the first aim of treatment is to reduce the ocular hypertension; undue reliance on tonometry can be just as misleading as in adult glaucoma and it should never be the sole method by which control of the glaucoma is assessed. It is our practice to estimate the intraocular pressure by both Schiötz and applanation tonometry, using the Perkins instrument for the latter method.

The cornea

An increase in the corneal diameter can be an indication that the intraocular pressure is not controlled; the cornea retains the ability to expand in the presence of persistently raised intraocular pressure up to the age of approximately 18 months.

Continuing oedema of the cornea may indicate that the glaucoma is not controlled; however, in some cases, corneal oedema persists for a few weeks after the intraocular pressure has been reduced to normal levels and this is presumably a manifestation of delayed recovery of function by the damaged corneal endothelium.

The optic disc

The state of the optic disc is the most important parameter by which the control of the glaucoma is judged. Because it is not possible to test the visual acuity and visual fields in infants, examination of the optic disc is the only means available of assessing the functional status of glaucomatous infants' eyes. The degree of cupping should be recorded as the cup/disc ratio. An increase in the size of the cup always indicates that the glaucoma is not controlled and that further therapeutic measures are necessary; conversely, constancy of the size of the cup in the presence of possibly elevated intraocular pressure indicates that further measures need not be introduced precipitately. This concept is particularly valuable in the management of cases having a poor surgical prognosis such as aniridia and the Sturge-Weber syndrome.

Gonioscopy

Examination of the chamber angle is seldom helpful in assessing glaucoma control, since the appearances after goniotomy do not seem to correlate with the response of the intraocular pressure. Gonioscopy forms part of the routine follow-up examination of infants with glaucoma since the information it yields is both interesting and stimulating to the surgeon.
PREVENTION OF AMBLYOPIA

That amblyopia might be a significant cause of poor vision in cases of congenital glaucoma was suggested by Richardson, Ferguson, and Shaffer (1967), and results of a study of children under our care have confirmed this (Table I). Seventeen children were studied who were old enough to allow estimation of the visual acuity; none had had occlusion previously; the glaucoma was under good control in all of them. The glaucoma was unilateral in ten cases and bilateral in seven. In sixteen cases the visual acuity of one

Table I Visual acuity in seventeen children with congenital glaucoma (average age 3 yrs 6 mths)

<table>
<thead>
<tr>
<th>Squint</th>
<th>No. of cases</th>
<th>Visual acuity</th>
<th>Satisfactory occlusion</th>
<th>Improvement</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Equal</td>
<td>Unequal</td>
<td></td>
</tr>
<tr>
<td>Present</td>
<td>13</td>
<td>0</td>
<td>13</td>
<td>11</td>
</tr>
<tr>
<td>Absent</td>
<td>4</td>
<td>1</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>17</td>
<td>1</td>
<td>16</td>
<td>14</td>
</tr>
</tbody>
</table>

eye was worse than that of the other and the poor acuity of the worse eye could not be satisfactorily explained by ocular damage due to glaucoma. Thirteen cases were squinting and, in all of these, the squinting eye had the worse vision. In all the fourteen cases in which a satisfactory regime of occlusion of the better eye was achieved, the visual acuity of the worse eye improved (Figure). It seems a reasonable assumption that the improvement in visual acuity after occlusion indicates that amblyopia ex anopsia was a significant factor leading to poor vision in one eye.

Figure Response of visual acuity to occlusion in fourteen cases
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There are probably a number of inter-related factors which are relevant to the development of amblyopia in these children; two would seem to be of particular importance, namely squint and anisometropia.

Squint

Of the seventeen patients described above, thirteen were squinting. In a study of 54 cases of congenital glaucoma the incidence of squint was found to be 54 per cent.; 56 per cent. of the bilateral cases and 51 per cent. of the unilateral cases were squinting (Table II). It seems reasonable to assume that eyes with potentially good vision which develop squints are at risk of becoming amblyopic.

Table II  Incidence of squint in 54 children with congenital glaucoma

<table>
<thead>
<tr>
<th>Glaucoma</th>
<th>No. of cases</th>
<th>Squint</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Convergent</td>
<td>Divergent</td>
</tr>
<tr>
<td>Bilateral</td>
<td>25</td>
<td>11</td>
<td>3</td>
</tr>
<tr>
<td>Unilateral</td>
<td>29</td>
<td>10</td>
<td>5</td>
</tr>
<tr>
<td>Total</td>
<td>54</td>
<td>21</td>
<td>8</td>
</tr>
</tbody>
</table>

Anisometropia

In thirteen of the fourteen cases in which amblyopia was demonstrated, there was anisometropia of between 1.0 and 7.5 dioptres and the amblyopic eyes were the most ametropic. One patient had no anisometropia but the amblyopic eye was squinting. Thus it would seem that anisometropia may play some part in the development of amblyopia and also in the high incidence of squint. Because of the abnormal anatomy of buphthalmic globes, aniseikonia may also be relevant.

Thus there is good evidence that the frequently successful results of surgery in congenital glaucoma are marred by the development of amblyopia. If eyes which are at risk of becoming amblyopic could be identified early, then it would seem reasonable to initiate an appropriate regime of occlusion however young the child. All children with congenital glaucoma should be examined regularly to determine if they have developed a squint. Refraction should be part of the routine periodic examination of these children and it is now our practice to perform retinoscopy when the glaucoma has been satisfactorily controlled. If a child develops a squint or has a significant degree of anisometropia, we institute a regime of occlusion which is continued intermittently until the child is old enough to co-operate with visual acuity testing.

It has been our experience that an orthoptist can be an exceedingly valuable member of the team caring for children with congenital glaucoma; her expertise enables her to identify squints in very young children, by supervising occlusion she can take a great load off the surgeon, and she is able to estimate the visual acuities of quite young children.

Miss M. E. A. Wakefield, D.B.O.T., of the Orthoptic Department, Moorfields Eye Hospital, has given invaluable help. My thanks are due to Arthur Lister, F.R.C.S., my mentor and guide to congenital glaucoma.
COMMENTARY

(1) EXAMINATION OF INFANTS

Dr. Anderson stated and Professor Goldmann confirmed that examination of infants can often be performed without general anaesthesia. All that is necessary for very small children up to the age of 4 months is to starve the child and keep it awake for the period before the procedure. Just immediately before the investigation, the child is given a bottle of milk which keeps it so detached that tonometry can be performed without difficulty. Local anaesthetics, of course, should be used to do pressure readings.

In slightly older children chloral hydrate syrup will put the child to sleep for 5 to 10 minutes. Ketamine has been found to be very satisfactory but this is only an anaesthetic agent and not an analgesic agent so that a topical local anaesthetic must be used for applanation tonometry. In adults ketamine gives very unpleasant hallucinations and should not be used. All the precautions necessary as a preparation for general anaesthesia should be observed because of the salivation which occurs with the use of ketamine, but provided a sucker is readily available it is a very safe agent.

(2) SIZE OF THE EYE IN INFANTILE GLAUCOMA

The normal eye increased in size up to the age of 18 months, the growth being very rapid in the first year and then slower over the next half year. When calculated in terms of percentage, the rate of increase of glaucomatous eyes was about the same as that of non-glaucomatous eyes, but once congenital glaucoma was controlled the increase in size of the cornea ceased even though the child had not attained the age of 18 months.

(3) DEVELOPMENT OF THE ANGLE OF THE ANTERIOR CHAMBER AND THE AETIOLOGY OF CONGENITAL GLAUCOMA

Dr. Hansson showed scanning electronmicrographs of foetal angles and an angle from a case of congenital glaucoma, feeling that he had shown in the normal embryonic angle a membrane that becomes perforated near term. He thought that this membrane persisted in congenital glaucoma but was destroyed by the preparation techniques used. This is why neither he nor Dr. Anderson had found a membrane in the angles of eyes from children with congenital glaucoma.

(4) OCCLUSION AMBLYOPIA IN THE AFTERCARE OF PATIENTS WITH CONGENITAL GLAUCOMA

While it is most important to correct the anisometropia in congenital glaucoma, great care should be taken in occluding children as young as 3 months of age continuously, for up to the age of 6 months total occlusion might lead to neuronal damage. This is analogous to the situation in animals where it has been shown that even short periods of occlusion can lead to permanent neuronal loss. Anisometropic amblyopia does not occur in myopia, only in hypermetropia and astigmatism, so that the squint found in so many cases of congenital glaucoma is most likely to result from corneal opacities or field loss or astigmatism but not from the myopia. Professor Richardson said that he had reviewed Dr. Barkan's cases. All of 55 patients with between 5 and 20 years' follow-up had good surgical control, but the statistics were strikingly similar to those found by Mr. Rice in that two-thirds of the patients had developed crossed eyes, two-thirds had significant anisometropia, and two-thirds had reduced vision in one eye. The groups did not in fact quite overlap, but there was enough overlap to suggest that there was a correlation between crossed eyes, poor vision, and anisometropia.

* See p. 146
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N S Rice

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