The Variable Clinical Characteristics and Course of Early Infantile Esotropia

Richard M. Robb, M.D.
Dorothy W. Rodier, C.O.
Boston, Massachusetts

ABSTRACT

A study of 75 patients with early infantile esotropia treated by one of the authors (RMR) between 1965 and 1980 revealed a broad range of clinical findings and treatment modalities. The study group included nine children born prematurely and 16 with neurologic impairment; the balance had no apparent illness other than strabismus. Only 21 patients had spontaneously alternating fixation; the rest required some form of occlusion therapy. Glasses were prescribed for 75% of the patients sometime during their management, and changes in refractive error were common. One or more operations were performed on 56 patients, but 19 patients required no surgical intervention because of a spontaneous decrease in their deviation. Binocularity was found more frequently in patients straightened surgically before 2 years (50%) than in those straightened after that time (14%), but it was also present in five of the 19 patients who required no surgery. The presence of binocularity during the course of treatment did not insure stability of alignment.

INTRODUCTION

Much of the recent discussion of infantile esotropia has focused on the optimal time for surgical correction. Treatment recommendations have centered on this aspect of management, often in highly-select subgroups of patients. More broadly inclusive accounts of infantile esotropia have suggested a lack of uniformity of patients with the condition as well as variability in their course following surgical alignment. Because it may not be clear during early examination which patients will have spontaneous improvement in their misalignment, which will experience temporary or permanent realignment with glass correction for hyperopia, and what will be the response to efforts at surgical correction, it is important to examine the variability in characteristics and course of an inclusive group of patients with early infantile esotropia in order to put in perspective our efforts to achieve early surgical realignment. Although retrospective analyses of groups of patients are limited by incomplete and sometimes inaccurate data, they may help to define clinical experience and sharpen questions to be addressed in future investigations. This review of 75 patients with early infantile esotropia, treated and followed since 1965, was undertaken with these aims in mind.

PATIENTS AND METHODS

The records of 258 patients considered to have had early infantile esotropia when first examined by one of the authors (RMR) between 1965 and 1980 were reviewed. Patients were excluded from further study if they had been seen in consultation or for only a single visit (75 patients), if they had moved or were lost to follow-up (28 patients), or if they had had strabismus surgery elsewhere (45 patients). Also excluded were 30 patients whose initial ophthalmologic examination was after 14 months of age, because there was a clear decrease in the frequency with which the diagnosis of early infantile esotropia was made after that age, and because it was felt that the history of the onset of esotropia would be less reliable beyond that time. Five patients with ocular abnormalities that probably influenced their strabismus (oculocutaneous albinism, infantile hemangiomata of the eyelid, hypoplasia of the optic nerve, and cataracts) were also excluded, leaving 75 patients for study. All patients were seen and followed by a single
TABLE 1
Initial and Final Deviations in Various Groups of Patients

<table>
<thead>
<tr>
<th>Initial Deviation</th>
<th>Number of Patients</th>
<th>Esotropia in Prism Dipters*</th>
<th>Number of Patients</th>
<th>Final Deviation Type of Deviation</th>
<th>Prism Dipters</th>
</tr>
</thead>
<tbody>
<tr>
<td>All patients</td>
<td>75</td>
<td>44 ± 16.5</td>
<td>66†</td>
<td>ET</td>
<td>11.0 ± 8.2</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>7</td>
<td>XT</td>
<td>9.1 ± 6.6</td>
</tr>
<tr>
<td>Normal growth and development</td>
<td>42</td>
<td>46 ± 15.5</td>
<td>37</td>
<td>ET</td>
<td>10.2 ± 7.5</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>5</td>
<td>XT</td>
<td>9.6 ± 7.5</td>
</tr>
<tr>
<td>Premature birth</td>
<td>9</td>
<td>45 ± 14.9</td>
<td>9</td>
<td>ET</td>
<td>11.2 ± 7.9</td>
</tr>
<tr>
<td>Neurologic impairment</td>
<td>16</td>
<td>40 ± 17.3</td>
<td>13†</td>
<td>ET</td>
<td>12.5 ± 9.2</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>1</td>
<td>XT</td>
<td>4.0</td>
</tr>
</tbody>
</table>

*Mean ± standard deviation
†Two patients excluded because surgery declined by parents.
ET = esotropia
XT = exotropia

ophthalmologist. Patients were referred by personal contacts, pediatricians, and other physicians. The average age at the time of first ophthalmologic examination was 7.9 months. The onset of esotropia was before 3 months in 59 patients and between 3 and 6 months in 16 patients. The average length of follow-up was 8.7 years with a range of 2.7 to 19 years. Sixty-five of the 75 patients were followed longer than 5 years. Ocular deviations were measured with prism and alternate cover when possible, and otherwise by Kirmesky estimates. Refractions were done by retinoscopy 30 to 45 minutes after instillation of 2 drops of 1% cyclopentolate solution.

Of the 75 patients reviewed, nine were born prematurely at an average gestational age of 34.3 weeks (range 32 to 36 weeks) and had an average birth weight of 4.9 pounds. Two of the premature patients were also in a group of 16 with neurologic disease or impairment. These children had one or more of the following: mental retardation (ten patients), cerebral palsy (seven patients), hydrocephalus (three patients), seizures (two patients), and Down's syndrome (one patient). Fifty-two patients were born at term and appeared to have normal growth and development except for their strabismus. Nearly one-third of the group had a known family history of strabismus.

RESULTS

Initial Deviation: The average initial esotropic deviation for the 75 patients was 44 ± 16.5* prism dipters (Table 1), a value that did not vary appreciably for those who were born prematurely (45 ± 14.9 prism dipters) and those with normal growth and development (46 ± 15.5 prism dipters). The average initial deviation for the 16 patients with neurological impairment was 40 ± 17.3 prism dipters, slightly less than the other groups, but not statistically different because of the large standard deviation.

Fixation Pattern: Twenty-one patients had an alternating fixation pattern and required no patching. Fifty-four patients (72%) had a monocular fixation preference that prompted occlusion therapy. Of the latter, 29 patients were patched only during the first 3 years and for periods of less than 18 months; 25 patients required some patching beyond 3 years of age or for periods longer than 18 months. Four patients had a reversal of fixation preference while being occluded, but none was rendered permanently amblyopic in the occluded eye.

Refractive Correction: Significant refractive errors requiring glass correction were found in 56 of the 75 patients (75%). Twenty-nine patients had more than 2 dipters of hyperopia, but there were notable variations within this group: 16 patients had relatively stable hyperopia that was corrected with glasses at the time of initial refraction and continued to warrant correction throughout the follow-up period; 11 patients had hyperopia that increased with time, and five of these patients were given glasses after initial surgery had been performed; two patients had decreasing hyperopia for which glasses were initially prescribed and subsequently discontinued. Four patients had myopic anisometropia greater than 2 dipters, all had persistent monocular fixation, and two developed deep ambylophia in the more myopic eye despite occlusion therapy. Another two patients had symmetrical high myopia that increased during the first 4 years.

Approximately 40% (23) of the patients who had strabismus surgery wore glasses prior to their first surgery, while an additional 32% (18) were given glasses after an initial operation had been performed. Fifteen of 19 patients (79%) for whom surgery was not required wore glasses at some
time during the course of their management.

Strabismus Surgery: Strabismus surgery was performed on 56 of the 75 patients with infantile esotropia. All but one patient had horizontal muscle surgery; this patient, who had only bilateral inferior oblique myectomies, was excluded from calculations of pre-and postoperative horizontal alignment. One additional patient was excluded from these calculations because the severe degree of his mental retardation became increasingly apparent after an initial operation on the horizontal rectus muscle of one eye, and additional surgery was declined by the parents in spite of a residual 40 prism diopter esotropia. The remaining 54 surgical patients had an average preoperative deviation of 50.5 ± 12.9 prism dipters of esotropia. The surgery was performed generally on two horizontal rectus muscles at each operation, usually a recession of one medial rectus muscle combined with a resection of the lateral rectus muscle of the same eye (50 patients), and only occasionally recessions of both medial rectus muscles (four patients). Additional horizontal muscle surgery was carried out when a significant residual esotropia remained. Using this approach, 36 patients with an average preoperative deviation of 46.4 ± 12.2 prism dipters of esotropia had one operation, and 18 patients with an average preoperative deviation of 58.9 ± 10.4 prism dipters of esotropia had more than one operation on the horizontal muscles. In 48 patients who were esotropic at the end of the follow-up period, the average final deviation was 10.8 prism dipters, and in six patients who were exotropic the average residual deviation was 10.5 prism dipters. The final deviations were slightly larger in patients who had more than one surgical procedure than in those who had a single operation (Table 2).

There was no significant difference in the final deviations of patients in the groups with premature birth, neurologic impairment, and normal growth and development. Different numbers of operations were required, however, to reach this endpoint (Table 3). In the prematurely born group, seven of nine patients required nine operations, an average of one operation per patient; in the neurologically impaired group, six of 14 patients required a total of seven operations, an average of 0.5 operations per patient. In the group with normal growth and development, 42 of 52 patients had 57 operations, an average of 1.1 operations per patient.

Nineteen of the 75 patients with infantile esotropia did not require strabismus surgery. Nine of these had neurologic impairment. The average initial esotropia in this nonsurgical group was 27.5 ± 10.0 prism dipters. Seventeen patients had an average final esotropia of 12.2 ± 8.6 prism dipters and one patient had an esotropia of 12 prism dipters. One patient was excluded from these calculations of initial and final deviations because his level of retardation led his parents to decline surgery despite a 50 prism dipters deviation that ordinarily would have placed him in the surgical group. Nine of the remaining 18 patients in this group had hyperopic refractive errors for which spectacle correction had a beneficial effect on the deviation, five were myopic, one had mixed astigmatism, and three had no significant refractive error. None of these patients was felt to have purely accommodative esotropia.

Consecutive Exodeviations: Seven of the 75 patients became exotropic while they were followed. One of these diverged spontaneously from an initial esotropia of 25 prism dipters. He had straight eyes at age 6 years with some binocular vision and gross stereoopsis, but by age 9 years had developed an intermittent exotropia of 12 prism dipters, the eyes being constantly exotropic with distant fixation. The six other patients developed exotropia at varying times following strabismus surgery: two following bilateral recess-resect operations, two following a recess-resect operation on one eye, one after a bimedial recession, and one after recession of both medial recti combined with a resection of one lateral rectus muscle. Three patients
required reoperation for their exodeviations; the others had a small esotropia of little cosmetic significance. Six of the seven patients who developed exodeviations had evidence of binocular vision at some time during the course of their strabismus, measured with Worth lights and the Titmus stereo test (Titmus Optical Company, Petersburg, Virginia). Four of the six had lost this evidence of binocularity at their last follow-up examination. Only one patient in the group with consecutive exotropia had retarded mental development; the others were felt to have normal growth and development.

Vertical Imbalance, Abduction Deficiency, and Nystagmus: Several additional features of ocular misalignment present in our 75 patients are worthy of consideration. Twenty patients (27%) had oblique muscle imbalance that warranted surgery. Seventeen bilateral and four unilateral inferior oblique myectomies were performed, either at the time of horizontal rectus muscle surgery or as a separate procedure.

The superior oblique muscles were found less often to be overacting, and only one bilateral and one unilateral superior oblique tenotomy were performed. The average age at the time of oblique muscle surgery was 4.5 years, considerably later than the time of horizontal muscle surgery. Dissociated vertical deviations were recognized in 30 of 75 patients (40%), but no surgery was specifically directed to this variable vertical misalignment. Abduction deficiency was noted in 41 patients (55%), a figure that must be considered approximate because the amount of deficiency of abduction was variable and may not have been recorded in all records. Although incomplete abduction was often associated with increasing jerk nystagmus in lateral gaze, no patient was felt to have the nystagmus blockage syndrome as presently described. The deficiency of abduction improved with time. It was often less apparent after surgery on the horizontal muscles, but record review did not establish accurately the timing of this improvement. No patient was left with a persistent limitation of abduction. Nystagmus in the primary position was recorded in 10% of patients, and occlusional nystagmus was present in 16% of the group.

Binocularity: Binocular responses on the Titmus stereo test and the Worth lights were not found in any patient in the premature group and in only three patients with neurologic impairment. Of the patients with normal growth and development who had surgical alignment to less than 10 prism diopters of esotropia, four of eight (50%) aligned prior to 2 years of age had some binocular vision. Only three of 22 patients (14%) who were similarly aligned after 2 years of age had evidence of binocular vision. Five patients with final alignment greater than 10 prism diopters of esotropia had no binocular vision. In the nonsurgical group, including those with neurologic impairment, three of five patients who reached an alignment of less than 10 prism diopters of esotropia by 2 years of age had evidence of binocular vision, whereas only one of eight patients who achieved this alignment after 2 years had binocularity. None of the five unoperated patients with exodeviations greater than 10 prism diopters had any evidence of binocular vision. The presence of binocular vision in six of seven patients who developed exodeviations and its loss in four of these patients has been described above. Five patients in the operated group and two in the unoperated group who had binocular vision had lost binocularity by the time of their last follow-up examination.

DISCUSSION

The composition of any group of patients with early infantile esotropia will vary with the setting in which the patients are seen and the referral patterns in a given community. Accordingly, different proportions of infants with premature birth or neurologic impairment will be found in reports on infantile esotropia, and the characteristics of the esotropic deviation itself may vary in different studies. It is probable that most of the variations in infantile esotropia are represented in this study. There is remarkable similarity in the breadth of the clinical picture reported here to that described by Costenbader in a larger group of patients in 1961. In the time between these two studies attention has been focused on the optimal time for surgical correction of infantile esotropia, often in selected subgroups that retrospectively exclude patients with large or asymmetrical refractive errors, amblyopia,
variable deviations, mental retardation, or cerebral palsy. Spontaneous divergence to an acceptable alignment has seldom been mentioned.\textsuperscript{5,6}

Recommendations for surgery have, therefore, been based on examination of a limited population, and may not be appropriate for all patients. It is not always apparent at initial examination which patients will have important refractive errors, which will require prolonged amblyopia therapy, which will have evidence of neurologic impairment, and which will experience a spontaneous change in the angle of misalignment. Repeated examinations help to resolve these uncertainties.

It was surprising that 72% of the patients with infantile esotropia in this group required patching for monocular fixation and presumed amblyopia, because alternating fixation has been said to characterize the group.\textsuperscript{18} Our patients could be divided roughly into thirds; one third with alternating fixation, one third with monocular fixation requiring limited occlusion, and one third requiring extended occlusion for persistent amblyopia. Spectacle correction was also necessary for 75% of the patients. Approximately 40% of those who had strabismus surgery wore glasses preoperatively, and an additional 32% were given glasses after at least one operation. In this latter group were patients who had only moderate hyperopia associated with large initial deviations. Whereas spectacle correction was not felt to offer a significant reduction in the larger initial esotropia, it did make a significant reduction in the smaller residual esotropia following surgery, especially if the amount of hyperopia had increased with time. This circumstance has been recognized by Raab in a more general discussion of accommodative esodeviations.\textsuperscript{19} Nearly 80% of the patients who did not require strabismus surgery wore glasses. Some, but not all, were hyperopic, and none was felt to have early onset of purely accommodative esotropia.\textsuperscript{15-16}

We have not related the exact amount of strabismus surgery performed on our patients to the initial and final deviations. It is interesting, however, to note that the average initial esotropic deviation of those patients who had one operation (46.4 $\pm$ 12.2 prism dipters) was less than that of those patients who had more than one operation (58.9 $\pm$ 10.4 prism dipters); the average initial deviation of those who had no surgery (26.8 $\pm$ 10.2 prism dipters) was lowest of all. The standard deviations in all groups are large, however, and it would have been difficult at the outset to accurately place individual patients in their proper categories. One difficulty in categorization is the uncertainty as to whether the initial angle of esotropia will increase or decrease spontaneously over the first 12 to 18 months of life. Another problem is the potential inaccuracy of prism measurements for angles of deviation over 50 prism dipters.\textsuperscript{20-21} The final deviations in our patients were generally small and cosmetically apparent, perhaps reflecting the observation that the amount of correction obtained from strabismus surgery is directly related to the magnitude of the initial deviation.\textsuperscript{22-23} The larger residual deviations in those with neurologic impairment reflect a more conservative approach to surgery in these patients and may reveal their neuromuscular instability.\textsuperscript{12}

Those patients who became exotropic following strabismus surgery were not characterized by any single preoperative feature or by the type or amount of strabismus surgery they underwent. Only one had cerebral palsy, a condition generally associated with surgical overcorrection of congenital esotropia.\textsuperscript{24} The development of exodeviation did not seem to be prevented by prior fusion. Most patients who became exotropic had some evidence of binocular vision between their initial alignment and their subsequent exotropia. The instability of surgical correction in some patients with infantile esotropia has been well documented by Hiles.\textsuperscript{14}

Binocular vision, measured by two commonly used sensory tests, was found in only 13% of the total group of patients at last examination. Another 9% had evidence of binocular vision at one time, but had lost it by the end of the follow-up period. As might be predicted on the basis of earlier studies,\textsuperscript{2,8} binocularity was found only in those patients who were well aligned, and it was more common in those whose alignment was achieved by 2 years of age (50%) than in those who were straightened later (14%). These results are similar to those reported by others in retrospective studies of selected patients.\textsuperscript{2,7} The age factor in determining binocularity held true for patients whose esotropia had lessened spontaneously as well as for patients who were straightened surgically. That no patients in the premature group developed binocular vision might reflect their final alignment, the age at which alignment was achieved, or the small number of patients in this group. The tendency of a significant number of patients to lose evidence of binocularity in the course of the first decade has been noted previously,\textsuperscript{14} and raises a question about the ability of fusional responses to maintain ocular alignment once alignment has been achieved.

**CONCLUSION**

Early infantile esotropia is a disorder with a broad clinical spectrum. It requires careful diagnostic efforts and usually both surgical and non-surgical care over most of the first decade. The timing of possible surgical intervention is an important consideration, but is only one factor in many needed to provide optimal care for this condition. A better understanding of the cause of the disorder and the variations in its course should improve our current treatment.

**REFERENCES**


