# INFANTILE ESOTROPIA

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INFANTILE ESOTROPIA IS ANY ESOTROPIA which is known to be present before the age of one year. This knowledge may be from a history obtained from the parents or may be from examination by an ophthalmologist prior to the age of one year.

The objectives of the present study are (1) to determine the characteristics which are representative of infantile esotropia, (2) to determine the percentage of cures (if any) after treating these patients, and (3) to list and qualify the factors which are important in obtaining a cure.

In considering the first objective, we must recall that it has been stated[1] that infantile esotropia comprises a homogenous group characterized by a large deviation (50+ prism diopters), equal visual acuity, small to moderate hyperopia, and limitation of abduction O.U. However, two subsequently unpublished studies have stated that infantile esotropias run the gamut of motor and sensory findings. It seems important to obtain a more definitive answer to this question.

Berke[2] has stated that no congenital esotrope can obtain stereopsis, and thus a cure is not possible. This raises three questions. How should cure be defined? Is cure possible in congenital (and infantile) esotropia? Which of the following factors favorably or unfavorably influence cure, if cure is possible? Some of these factors are:—

(1) age of onset
(2) duration of squint
(3) age at first alignment
(4) size and nature of the initial deviation
(5) initial presence of hypertropia
(6) presence of a persisting deviation
(7) amblyopia—what degree?
(8) initial refractive error
(9) number of operations
(10) age at first operation

The question of definition of cure has been raised before. Berke[2] has stated that "the standard of perfection in squint surgery is the attainment of single binocular vision with good stereopsis" and with

this I agree. Some years ago,[3] however, and again in discussion of Berke more recently,[4] I suggested that "cure is not absolute, but may consist of several grades of binocular cooperation." While the attainment of a high degree of binocular vision with excellent stereopsis is a most desirable goal, lesser grades of stereopsis, or fusion without stereopsis, are highly desirable, if perfect results cannot be obtained. Thus, the degrees of cure possible, and the circumstances under which they can be obtained, should be determined.

It seems quite evident that many factors may be of great prognostic significance in attempting to obtain a cure. We should then determine what findings favorably or unfavorably influence a final good result, and in what manner they should be dealt with.

## LITERATURE

A vast amount has been written pertinent to this present study.

### POINTS OF EMPHASIS

CURE. Berke[2] has stated that congenital constant strabismus never attains a cure. Anderson,[5] Law,[6] Gibson,[7] Duggart [8] Douglas,[9] Kempe,[10] Worth,[11] Leahey,[12] McCann,[13] and others are in general agreement that the prognosis for cure of constant congenital strabismus is poor. They vary in their attitude toward early surgery from a position of antipathy to one of no urgency.

EARLY TREATMENT. However, many authors have urged early treatment, especially surgery, with an attempt at early alignment, stating that the early opportunity for binocular single vision is important to cure. Riise,[14] Costenbader,[3] Cooper and Evans,[15] Cashell,[16] Lyle,[17,18] Lyle and Foley,[19] Chavasse,[20] Cooper,[21] Leahey,[12] Duke-Elder,[22] Nordlow,[23,24] and many others[25–40] have stressed this point.

There has been a decided lack of agreement as to *what constitutes early surgery*. Several decades ago surgery was considered to be early if performed by the age of six to eight years, and in many countries, even today, surgery by school age is considered early. However, the view that very early surgery (six to eighteen months of age) is most important is becoming increasingly prevalent. Dowler, *et al.*[40] suggest surgery at three to six months of age.

On the other hand, Arruga and Downey[41] have strongly criticized the trend to early surgery in strabismus. They suggest that much early surgery causes or allows an insecure binocular vision at a small angle

of deviation, making further treatment and improved functional results impossible. Weve[42] also adds a word of caution since a large survey showed fifty percent less strabismus at age twelve to thirteen years than at five to six years. This suggests a considerable degree of spontaneous cure.

DEVIATION. It is well recognized[2,12,14,20–22] that the later the onset of the deviation and the more infrequent its occurrence (intermittent deviations), the better the prognosis. On the other hand, the persistence of a constant deviation following treatment makes cure impossible.[3,43–46]

HISTORY. While the question of reliability of history in determining the age of onset of esotropia has been raised,[47,2,12,19] no definitive study has been performed to qualify this reliability.

TEST FOR CURE. The tests used to establish the presence of, or degree of, cure have been the subject of some discussion. Berke[2] would test stereopsis on a stereoscope only. Nordlow[23] would use binocular fixation supported by fusion, fusional amplitude, and stereopsis on the major amblyoscope. Leahey[12] would use the Wirt tests if alignment is possible, while the major amblyoscope is used if manifest deviation is present, when the patient is at least four years of age.

## SIMILAR STUDIES

Many studies somewhat similar to the present one have been published.

Berke[2] investigated a series of 265 cases of strabismus, followed from six months to nineteen and a half years (average follow-up = 5.3 years). Using stereopsis on the stereoscope as a prime criterion, he concluded that (1) no patient having a visual acuity of 20/50 or less developed third degree fusion, (2) no patient having constant squint at birth developed third degree fusion, (3) the longer the duration of squint the poorer the chances of functional cure, and (4) prognosis is always better in intermittent than in constant squint.

Cooper,[21] in general, agreed with Berke's conclusions when reporting 483 cases of squint (382 non-accommodative esotropes and 101 intermittent esotropes).

Lyle and Foley,[19] when reporting 287 cases of comitant esotropia, agreed that early onset and prolonged duration offered a poor prognosis for functional cure. They, like others, suggested that early operation should produce better functional results but offered no statistical support.

Nordlow[47] cites the difficulty in accurately determining the age of onset. In a large series he finds that the onset was before the age of one year in fifty-one percent of all esotropia. A subsequent survey[23] of the long-range effect of early treatment is quite enlightening. Seventy patients whose esotropia became evident between the age of six months and four years were operated early (six months to four and a half years of age) and followed for an average of eleven years. Seventy-five percent obtained binocular fixation and fifty-seven percent had binocular single vision (measured on the major amblyoscope)—quite excellent results. Additionally helpful information might have been obtained from (*a*) cases of esotropia present from birth (rather than a minimum age of six months), (*b*) stating graphically the age of the patients at operation, (*c*) testing stereopsis on the stereoscope or similar device (more nearly simulating daily vision). A most important point was the emphasis upon "age at which binocular fixation was first obtained." It would have been helpful to relate this age to the attainment of binocular single vision.

Kennedy and McCarthy,[48] in reviewing 315 operations for esotropia performed at various ages, found that only twenty percent obtained stereopsis and that nine percent were overcorrected.

Keiner[49,50] reviewed 656 patients with esotropia, of whom 50 were less than one year of age, while 123 were between one and two years, and 400 over two years. Onset of esotropia was noted before one year of age in fifty-four percent. Keiner discards refractive errors absence of fusion faculty, convergence anomalies, paralysis, or other mechanical factors as etiologic agents other than as a trigger mechanism or adjunctive cause. He believes that a delay or failure in myelination (possibly due to an inherited endocrine imbalance) causes a disturbance in development of the optomotor reflexes. The monocular adductive reflexes predominate over the abductive and conjugate reflexes, resulting in esotropia.

Crone and Velzeboer[51] are in general agreement with Keiner concerning etiology. They have surveyed 914 patients (fifty-two percent of whom were over seven years of age) and noted the onset of strabismus before the age of eighteen months in forty-two percent. They concluded that all characteristics and types of strabismus are inherited in about the same percentage and that birth trauma or ocular paresis are precipitating factors, rather than primary causes, of strabismus.

Scobee[52] reviewed 422 esotropes of whom twenty-eight percent

were congenital (and 13.5 percent premature). He stressed the role of anatomical factors in etiology of the congenital strabismus. He proposed the somewhat unique thought of recessing the medial recti to the equator for esotropia of all degrees.

Leahey[12] agrees with some of the propositions previously stated, and disagrees with the thought[49] that basic information cannot be obtained on many children at a very early age. Reporting 1019 strabismus operations, he concludes that (*a*) very early surgery is most important to the infant esotrope—as young as ten months or younger, and stereopsis can be obtained, (*b*) duration of squint is most important and strongly influences the fusion potential, and (*c*) if the patient is not available until age three years, tests for stereopsis should be attempted. If no stereopsis exists, surgery should be deferred until age five and six years since only a cosmetic effect can be obtained. (*d*) Intermittent deviations offer a good prognosis at most any age.

Arruga and Downey,[41] in analyzing cases having early treatment (especially surgery), state that such early treatment often results in a very small deviation with either abnormal retinal correspondence or poor fusional amplitudes or both. Because of this small angle and unstable relationship, functional cure is made more difficult or impossible in the future. They urge that alternate or monocular occlusion (to prevent sensory anomalies) be used until the patient is old enough for accurate diagnosis (three years plus) and orthoptic treatment be given before any surgery is performed. However, they present no evidence that such delayed treatment produces a higher percentage of secure functional results. It seems preferable to obtain a significant number of less secure and less perfect results by early treatment, rather than delay and obtain a questionable number of theoretically more secure results by the late surgery. Years of patching an obvious strabismus are not well tolerated by many.

## PRESENT STUDY

In this study the current file of our private office was consecutively surveyed and all properly qualified cases retained for analysis. Of a total of 3157 patients having strabismus (esotropia 2393, exotropia 718, hypertropia 46), there were 1152 patients having infantile esotropia (onset before one year of age). Many were eliminated from the study:

| | | |
|---|---|---|
| Infantile esotropia | | 1152 |
| Excluded because of: | | |
| (1) insufficient findings | 277 | |
| (2) insufficient follow-up | 136 | |
| (3) gross incomitance | 36 | |
| (4) organic amblyopia | 74 | |
| (5) previous surgery | 38 | |
| (6) retardation | 33 | |
| (7) miscellaneous | 58 | 652 |
| Infantile esotropia retained for this study | | 500 |

Since this study included many cases who had been seen initially fifteen or more years ago, many tests now felt to be important for proper evaluation had not been done. For this reason, 277 patients were eliminated from the study.

One hundred and thirty-six patients were excluded because the period of follow-up and observation was less than two years from the beginning of definitive treatment. An elapsed interval of two years semed important, since many patients were first seen before the age of one year or soon thereafter, and the age of at least three years was necessary to obtain reasonable final fiindings. Because they had not been observed at least two years after the beginning of treatment, 136 patients were excluded.

Thirty-six patients were found to have gross incomitance (congenital paralysis of lateral recti, Duane's retraction syndrome, Brown's superior oblique sheath syndrome, congenital third nerve paralysis, gross trauma to central nervous system or orbits) and were excluded from the study because functional cure seemed impossible.

Seventy-four patients had intractable amblyopia, due to organic causes (congenital optic atrophy, retrolental fibroplasia, congenital cataract, gross microphthalmos, etc.). Central fixation in both eyes seems important to cure, and none of these patients qualified in this respect and were eliminated.

Because it seemed important to have full knowledge of all treatment, all cases who had been subjected to surgery prior to our observation (38 patients) were excluded. Mental retardation (33 patients) made the obtaining and evaluation of findings difficult and such cases were excluded. Because the age of onset was in doubt, especially those patients who were not seen until several years after the presumed

onset, 38 cases were excluded. An additional 20 cases were excluded because follow-up, while of sufficient length, was too sporadic.

It becomes at once apparent that the remaining 500 cases of infantile esotropia have been carefully screened as to presumed time of onset, adequacy of findings, and sufficient period of follow-up. Of 500 patients having infantile esotropia, 114 (22.8 percent) were examined prior to the age of one year (Table 1) by me or one of my associates and true strabismus was found to be present.

TABLE 1. AGE AT FIRST VISIT (500 PATIENTS)

| | | |
|---|---|---|
| 0–11 months | 114 patients | (22.8%) |
| 12–23 months | 162 patients | (32.4%) |
| 24–48 months | 180 patients | (36.0%) |
| 49+ months | 44 patients | (8.8%) |

In addition to 22.8 percent seen before the age of one year, 32.4 percent were seen between the ages of one and two years (a total of well over 50 percent seen before the age of two years), and only 8.8 percent were first seen after the age of four years. Thus the opportunity for early observation and treatment is readily evident.

The age of onset of the 386 patients (77.2 percent) seen after one year of age was obtained by history. It must be admitted that parents are not always good observers, and may, at times, imagine a strabismus which cannot be confirmed by the ophthalmologist. With this in mind, an analysis of 753 patients in whom the parents suspected the presence of strabismus was made (Table 2).

TABLE 2. TRUE VERSUS PSEUDO-SQUINT IN 753 PATIENTS WHOSE PRESENTING COMPLAINT WAS APPEARANCE OF STRABISMUS

| *Age at first visit* | *Esotropia** | *Pseudo-squint* |
|---|---|---|
| 0–11 months | 102 | 60 |
| 12–23 months | 118 | 126 |
| 24–35 months | 77 | 80 |
| 36–47 months | 53 | 50 |
| 48–59 months | 23 | 22 |
| 60+ months | 28 | 14 |
| TOTALS | 401 | 352 |

*Those patients on whom the diagnosis of esotropia had been established elsewhere were excluded.

Of 162 patients seen before the age of one year, 102 had true esotropia while 60 had straight eyes, though an appearance of strabismus. This finding had been anticipated. The appearance was usually due to one or more of several causes—epicanthus, excessively narrow interpupillary distance, broad nasal bridge, facial asymmetry, or a negative angle kappa.

In those patients who presented at a greater age, the percentage of pseudo-squint was even higher, and this finding had not been anticipated. Of those seen during the second year of life, slightly more pseudo-squints than true squint were seen (126:118). A somewhat similar pattern exists for those patients first seen through the fifth year of life, but thereafter, the incidence of true esotropia predominates over the pseudo-squints (28:14). It may well be true that parents continue to be concerned about the appearance of estropia up to the school age, but after that age the maturing of the child's features and the reassurance from school-screening procedures decreases the false complaint of squint.

A further factor possibly influencing the ratio of true and false squint in this series is the fact that the area in which the survey was conducted is squint conscious and any patient suspected of strabismus is rather promptly examined. While this causes a high rate of over-referrals, it also makes the early diagnosis and treatment of true strabismus possible.

## SURVEY METHOD

All patients entering the office were subjected to a careful history and comprehensive examination. The age of the patient at the initial examination was noted.

The age of onset was noted and all those whose onset was one year or older were excluded from this study. Those whose onset was below the age of one year were divided into three groups:

(1) Birth through three months (333 patients—66.6 percent). It was noted that few parents expected the baby's eyes to be straight before the age of three months and only when the strabismus persisted thereafter were they concerned. Thus, birth through three months was a distinctive onset period.

(2) Four through six months (52 patients—10.4 percent)

(3) Seven through eleven months (115 patients—23 percent)

Because only a part of the patients' age of onset was proven by examination, further analysis will attempt to show the influence of

factors upon the "examination" group (114 patients) and the "history" group (386 patients) separately. If the findings are similar in the two groups, the history of onset may be considered valid, while if the findings in the two groups are widely divergent, the validity of the history of age of onset can be questioned.

The examination included estimation of visual acuity, refractive error, measurement of the horizontal deviation, presence or absence of a vertical component, versions with special emphasis on ability to abduct, and the status of binocular vision. As many of these findings as possible were obtained at the initial visit, again just prior to the first surgery, six months after first surgery, two years after first surgery, and at the final visit. In addition, the first date of approximate binocular alignment was noted, as well as the first date at which any fusion was noted. When surgery was not performed, findings were obtained six months after the beginning of definitive non-surgical treatment, two years thereafter, and at the final visit.

## ACQUISITION OF INFORMATION

### HISTORY

A careful history of age of onset of strabismus was taken, and subsequent behavior of the deviation was noted. When previous treatment had ensued, this was noted, but if the patient had had previous surgery, he was excluded from the study. The history of familial strabismus was obtained, noting the presence or absence in siblings, parents, and other relatives. The information is interesting but not immediately germane to this study.

### EXAMINATION (TESTS, THEIR APPLICATION AND EVALUATION)

It is readily apparent that many tests cannot be applied at early age levels and that as the patient becomes older and more attentive, more exactly definitive tests may be applied.

VISUAL ACUITY. In determining visual acuity, the standard Snellen alphabet chart at twenty feet was used for all those who knew their alphabet. In general, this was possible at the age of six years and later. In like manner, the illiterate E chart was used for those who could comprehend, usually between the ages of three and six years. It was recognized that the use of isolated ABC or E would in general allow for two lines better achievement than when the full chart was visualized. However, since each eye was tested by the same method,

differences in visual acuity in the two eyes could be rather accurately determined. All tests were performed with full cycloplegic correction, if significant spherical or cylindrical correction was indicated.

When the patient was too young for the Snellen charts, the ability to fixate and maintain fixation in each eye separately was used for estimation of visual acuity. It had been previously determined[53] that when an eye fixated a light or small picture well and centrally, the visual acuity was 20/20 to 20/70 (average 20/35); when central but not maintained, the visual acuity was 20/30 to 20/200 (average 20/70); when fixation was either roving or grossly eccentric, visual acuity was 10/200 or less.

Four gradations of visual loss were recorded for this study: (1) "none," indicating either good, central, and maintained fixation, or 20/40 or better acuity on the Snellen chart; (2) "mild," indicating good, central, but not maintained fixation, or a visual acuity between 20/40 and 20/70; (3) "moderate," indicating questionably central fixation, or a visual acuity of 20/70 to 20/200; (4) "severe," indicating either roving or grossly eccentric fixation, or a visual acuity of less than 20/200. A difference in visual acuity of the two eyes of at least one grade was considered a significant amblyopia, and amblyopia itself was graded as "none," "mild," "moderate," and "severe."

Visual acuity (and grade of amblyopia) was recorded at the initial visit, immediately before the first surgery (being the resultant of the passage of time and occlusion), six months after the first surgery (the result of passage of time, surgery, and at times further occlusion), and at the final visit (the result of time, occlusion, further surgery, and orthoptics). Improvement or deterioration of visual acuity was noted at each such visit and many intermediate visits.

REFRACTIVE ERROR. This was noted at least at the initial and final examinations, and often at six to twelve month intervals. Standard procedure was the use of homatropine hydrobromide 5 percent, gtt ii, and cyclogyl 1 percent gtt ii in each eye, and retinoscopy at the end of one hour. Many refractions were confirmed using one-half to one percent atropine sulfate twice a day for three days at home. Since less than +.50Sph difference was found in the two examinations, the homatropine and cyclogyl cycloplegia was thought to be acceptable for children of all ages.

The retinoscopic finding was accepted as final on all preschool children, but a further subjective examination was performed on all school age children. A hyperope of 1.50 diopters or more was cor-

rected by glasses, or in some instances, compensated for by the use of floropryl or phospholine iodide. Since the work of Brown,[54] and some unpublished work of our own had shown an increase in manifest hyperopia of 1.4 diopters from age one to age six years, there was no reluctance to prescribe the full cycloplegic acceptance in patients six years or younger, but to reduce the hyperopic correction slightly after the age of six years.

For purposes of this study, the effective refractive error was considered that of the least hyperopic eye since almost always the fixating eye is that having the least refractive error. The hyperopia most effective in influencing accommodation is that of the fixating eye. For example, if the patient accepts O.D. +2.50Sph and O.S. +3.25Sph, the O.D. is assumed to be the fixating eye (this was true in a partial review of the series) and thus the accommodative convergence is chiefly due to the +2.50Sph O.D. When cylinders of more than 0.5 diopter were present, the spherical equivalent (half the power of the cylinder) was added to or subtracted from the sphere (according to the appropriate sign).

HORIZONTAL DEVIATION. Estimates of the size of the horizontal deviation were determined in several ways. Those patients old enough (usually three years) and with sufficiently good fixation O.U. were examined by prism and alternate cover. Those too young or with too poor fixation (as in amblyopia or nystagmus) were measured by prism reflex (Krimsky—the centering of corneal reflexes by means of prisms). In patients three years and older, the measurement was taken at twenty feet and thirteen inches, without and with full correction (if worn). In those young patients whose attention for distance fixation was too poor, the measurement at thirteen inches was determined, while the deviation at twenty feet was estimated by appearance. The use of flashing lights and changing pictures and symbols greatly enhanced the attention factor and thus the accuracy of the measurements.

Measurement on the perimeter was found inaccurate and difficult to apply to the small child. Any measurements of the deviation on the major amblyoscope were felt to be too often influenced by attention and by instrument convergence to furnish valid findings.

The deviation was noted as E if latent, E(T) if intermittent (including periodic), and ET if constantly manifest even though variable in amount. That deviation remaining when measured with full hyperopic correction at twenty feet was considered non-accommodative

while that increase in deviation when the correction was removed was considered accommodative.

VERTICAL DEVIATIONS. For purposes of this study these were not measured precisely. It was carefully noted, however, whether the vertical deviation in the primary position was latent (H), intermittent (H(T)), or constantly manifest (HT). These were determined by the cover-uncover and the alternate cover tests.

Interest in hypertropia stemmed from the fact that the normal amplitude of vertical fusion is quite limited and thus, any manifest and possibly many intermittent vertical deviations may impose a definite bar to fusion.

VERSIONS. These were determined by (1) simple ocular rotations and noting fields of apparent overaction or underaction of the extraocular muscles, (2) prism reflex in the various fields of fiaze, and (3) prism and alternate cover in the various fields of gaze. Patients having a gross incomitance have been removed from this study.

Interest was focused mainly on the ability of the infant to abduct, it being noted that many initially had presumed paresis of the lateral recti O.U., but after patching, or after surgery on the medial recti only, the ability to abduct was normal or greatly improved.

FUSION. Tests for fusion presented several problems. First, various grades of binocular cooperation may be present, requiring a variety of tests. Second, the validity of certain commonly used tests may be in doubt. Third, even when considered valid, certain tests are impossible or difficult to perform by the very young or amblyopic patient.

The tests used in trying to determine the degree of fusion (or binocular cooperation) were: (1) the Wirt stereotest* to determine foveal or perifoveal fusion; (2) the Wirt "fly" test,* and (3) the Worth four-dot test to determine perifoveal fusion. In a few cases, fusional convergence, using a rotary prism, established the fact that fusion existed but the degree was not always apparent. Also in some cases, approximate binocular alignment was maintained, and restored itself after removal of cover. It was assumed that some degree of fusion existed even though none could be confirmed by ordinary testing methods.

It should be noted specifically that in conformity with Berke's thoughts, fusion or stereopsis on the major amblyoscope were not used as criteria. Such tests suggest the fusion potential of any patient, but do not show how this potential is translated into daily binocular vision.

*Stereo Optical Company, Chicago, Illinois

The tests used in this study more nearly fulfil this daily life requirement.

The most commonly used test in this series was the Worth four-dot test and it was applied to all 500 patients. In general, it was found applicable at twenty feet and thirteen inches to the average child over four years of age, and, at thirteen inches only, to many children as young as two years. When the child was too young to count or did not know his colors, he was asked to touch all the lights seen at thirteen inches. As is evident, the touching of four lights suggested fusion, five lights diplopia or alternate suppression, and two or three lights monocular suppression.

To cast some doubt on the validity of the Worth four-dot test as a measure of fusion, it should be noted that of the 500 patients, 44 seemed to demonstrate Worth fusion and yet had a manifest esotropia of five diopters or more. This occurred more frequently when the standard Worth rather than the reduced Worth test was used. This emphasizes the fact that the Worth four-dot test determines the presence or absence of perifoveal, not foveal, fusion, and even at times rapid alternation or abnormal retinal correspondence may be misinterpreted as fusion. None the less, the visualizing of four lights in the presence of bifoveal fixation, or even in the presence of a small manifest esotropia, suggests a more stable binocular relationship than if no fusion could be demonstrated.

The next most commonly used test for fusion was the Wirt "fly" test which in many cases could be applied to children as young as eighteen months, and routinely to those over the age of three years. It is a test for perifoveal fusion and, for the purposes of this study, along with "graded" stereopsis (see below) of thirty percent or less, has been termed gross stereopsis.

The Wirt "graded" stereopsis test is the most critical used in this series. It records degrees of stereopsis from five to 105 percent. For this study, 30 percent stereopsis or less was termed gross stereopsis and indicated perifoveal fusion, while stereopsis of 50 percent or more was termed refined and indicates foveal fusion. The test could occasionally be applied to the three-year-old, and almost routinely to those patients five years of age and older. Both because of the age factor and because the test has only recently been available, it was applied to only 95 of the 500 patients. Of the 86 patients demonstrating gross stereopsis, 15 had a manifest deviation of five prism diopters or more, while of those showing refined stereopsis, only one had a similar deviation.

## ANALYSIS AND CONCLUSIONS

Some of the many factors probably influencing cure of esotropia have been enumerated. In the analysis, an effort is made to show:

(1) The incidence of amblyopia; its influence on final cure; the results of treatment of amblyopia.

(2) The relationship of the age of onset to the initial refractive error, the size of the initial deviation, and the attaining of fusion at any time.

(3) The relationship of the initial and final horizontal deviation to final cure; the relationship of the initial deviation to the initial refractive error; and the improvement in deviation resulting from treatment.

(4) The influence of initial and final, latent and manifest vertical deviation to final fusion.

(5) The relationship of final fusion to age of onset, age at first surgery, number of surgical procedures, age at first alignment, and duration of squint.

While the results of treatment of amblyopia and strabismus will be considered, methods of treatment will not be discussed. Occlusion. spectacles, miotics, surgery, and orthoptics were all used when indicated.

### AMBLYOPIA

RESULTS OF TREATMENT OF AMBLYOPIA. Of 1152 patients having infantile estropia, 74 had intractable amblyopia due to organic defects and were discarded from this study. Of those 500 cases retained for study, 295* had no amblyopia at the initial or final visit (Table 3).

TABLE 3. RESULTS OF TREATMENT OF AMBLYOPIA

| | *Final amblyopia* | | | | *Total "initial"* |
|---|---|---|---|---|---|
| | *Severe* | *Moderate* | *Mild* | *None* | |
| Initial amblyopia | | | | | |
| Severe | 3 | 4 | 8 | 26 | 41 |
| Moderate | 0 | 7 | 21 | 54 | 82 |
| Mild | 0 | 3 | 6 | 57 | 66 |
| None | 0 | 4 | 12 | 295* | 311 |
| TOTAL (FINAL) | 3 | 18 | 47 | | |

Of those having amblyopia initially, 41 were severe (20/200 or less), and three of these persisted in spite of treatment. Four reduced to

moderate (20/70 to 20/200), and eight to mild amblyopia (20/40 to 20/70), while 26 (63.4 percent) had no amblyopia at the final visit.

Of those having moderate amblyopia initially, seven continued to have moderate amblyopia, 21 reduced to mild, and 54 (65.9 percent) had no amblyopia at the last visit.

Of those thought to have mild amblyopia at the first visit, three were found to be moderately amblyopic at the last visit, while six retained their mild amblyopia, and 57 (86.3 percent) were found to have no amblyopia.

Of 311 thought to have no amblyopia at the first visit, four were found to have moderate amblyopia, 12 mild amblyopia, and 295 no amblyopia at the final visit. It seems probable that those who seemed to deteriorate under observation had been diagnosed incorrectly in the first place and possibly were treated inadequately subsequently. We may conclude that contrary to usual thinking, *amblyopia is rather common in early esotropia, but that it responds well to treatment.*

RELATIONSHIP OF INITIAL AMBLYOPIA TO FINAL FUSION. Next, it is important to note in what fashion the initial amblyopia influences the final cure (Table 4). Does the presence of amblyopia at the initial visit have prognostic significance? Is it possible to obtain refined stereopsis or a lesser grade of fusion in the amblyopic patient, and to what extent?

TABLE 4. RELATIONSHIP OF INITIAL AMBLYOPIA TO FINAL FUSION

| | *No amblyopia* | *Moderate amblyopia* | *Severe amblyopia* |
|---|---|---|---|
| No fusion at any time | 88 (28.3%) | 40 (27.0%) | 19 (46.1%)* |
| Fusion not maintained | 61 (19.6%) | 33 (22.3%) | 8 (19.9%) |
| Worth fusion | 92 (29.6%) | 47 (31.8%) | 7 (17.0%) |
| Gross stereopsis | 58 (18.6%) | 21 (14.2%) | 7 (17.0%) |
| Refined stereopsis | 12 (3.9%) | 7 (4.7%) | 0 |
| TOTAL | 311 | 148 | 41 |

As would be anticipated, a higher percentage of severe amblyopes (46.1 percent)* never demonstrated fusion than was true with non-amblyopes (28.3 percent). About an equal percentage from each group attained fusion at some time, though it was not maintained at the final visit (19.6 percent, 22.3 percent, 19.9 percent). On the other hand, an appreciably higher percentage of non-amblyopes demonstrated Worth fusion and gross stereopsis, than in the amblyopic group. It is interesting to note that seven patients who had moderate amblyopia initially, finally attained a high grade of cure, that is, refined stereopsis.

We may conclude that, *while moderate amblyopia is not prognostically unfavorable, severe amblyopia noted at the first examination imposes an absolute bar to attaining good fusion, though lesser grades of fusion are attainable.*

RELATIONSHIP OF FINAL AMBLYOPIA TO FINAL FUSION. Finally, it is necessary to show the influence of final amblyopia upon final fusion. Is it possible to maintain fusion with any significant amount of amblyopia still present?

TABLE 5. RELATIONSHIP OF FINAL AMBLYOPIA TO FINAL FUSION

| | *No amblyopia* | *Moderate amblyopia* | *Severe amblyopia* |
|---|---|---|---|
| No fusion at any time | 121 (28.0%) | 24 (36.9%) | 2 (66.6%) |
| Fusion, not maintained | 88 (20.4%) | 13 (20.0%) | 1 (33.4%) |
| Worth fusion | 126 (29.2%) | 20 (30.8%) | 0 |
| Gross stereopsis | 80 (18.5%) | 6 (9.2%) | 0 |
| Refined stereopsis | 17 (3.9%) | 2 (3.1%) | 0 |
| TOTAL | 432 | 65 | 3 |

At the time of final examination (see Table 5) only three severe amblyopes were found and as would be expected, none of these showed any fusion at the final visit. However, about the same percentage of moderate amblyopes and non-amblyopes attained some fusion: (Worth 30.8 percent and 29.2 percent); (gross stereopsis 9.2 percent and 18.5 percent); (refined stereopsis 3.1 percent and 3.9 percent). It would seem then that *while final severe amblyopia* (20/200 or less) *is a decided bar to fusion, moderate amblyopia* (20/40 to 20/100) *is not*, two patients even attaining refined stereopsis. However, both patients had a final visual acuity of 20/50 or better.

DEVIATION

It is important to note the improvement in deviation as the result of treatment, and whether the size of the initial deviation is significant to the size and type of initial refractive error, and to the incidence of final cure.

IMPROVEMENT IN DEVIATION. It is worth noting (Table 6) that of 191* patients having a deviation of 50 prism diopters or greater, 104 (54.4 percent) had a heterophoria or intermittent heterotropia at the last visit, while 75 (39.2 percent) were undercorrected, and 12 (6.4 percent) were overcorrected. Those patients with a lesser deviation initially (133 having ET=25–49 prism diopters and 106 having ET=5–24 prism diopters) also had approximately the same percentage

TABLE 6. RELATIONSHIP OF INITIAL DEVIATION TO FINAL DEVIATION

| | *Final deviation* | | | | |
|---|---|---|---|---|---|
| | E,E(T) X,X(T) | ET = 5−24$^{\Delta}$ | ET = 25−49$^{\Delta}$ | ET = 50$^{\Delta}$+ | XT |
| Initial deviation: | | | | | |
| E or E(T) (70 pts.) | 59 (84.3%) | 9 | 0 | 0 | 2 |
| ET = 5−24$^{\Delta}$ (106 pts.) | 61 (57.5%) | 36 | 0 | 0 | 9 |
| ET = 25−49$^{\Delta}$ (133 pts.) | 66 (50.0%) | 56 | 3 | 0 | 8 |
| ET = 50+$^{\Delta}$ (191 pts.)* | 104 (54.4%) | 68 | 7 | 0 | 12 |
| TOTAL (FINAL) | 290 | 169 | 10 | 0 | 31 |

of straight eyes at the final visit (50 percent and 57.5 percent, respectively). As would be expected, a much higher percentage of those initially demonstrating an esophoria or an intermittent esotropia had good alignment at the final visit (84.3 percent).

From this, we may conclude that *the size of the initial deviation does not materially influence the final alignment, except for those cases who were well aligned in the first place.* The final deviation findings would be generally unsatisfactory if it were not recalled that all of these squints had a very early onset. Also, most of the residual deviations were of small to moderate amounts.

RELATIONSHIP OF INITIAL DEVIATION TO REFRACTIVE ERROR. Does the refractive error influence the size of the initial deviation in infantile esotropia?

TABLE 7. RELATIONSHIP OF INITIAL DEVIATION TO SIZE AND NATURE OF REFRACTIVE ERROR

| | E−E(T) | ET=5−24$^{\Delta}$ | ET=25−49$^{\Delta}$ | ET=50$^{\Delta}$+ | *Total (refractive errors)* |
|---|---|---|---|---|---|
| Myopia | 2 (3.0%) | 5 (4.7%) | 6 (4.5%) | 15 (8.1%) | 28 |
| 0 to +2.00Sph | 28 (40.0%) | 44 (41.5%) | 64 (48.1%) | 96 (50.0%) | 232 |
| +2.25Sph to +5.00Sph | 28 (40.0%) | 47 (43.3%) | 56 (42.2%) | 77 (40.2%) | 208 |
| +5.25Sph + | 12 (17.0%) | 10 (9.5%) | 7 (5.2%) | 3 (1.7%) | 32 |
| TOTAL (DEVIATION) | 70 | 106 | 133 | 191 | |

The percentage of small (0 to +2.00Sph) and moderate (+2.25Sph to +5.00Sph) hyperopia (see Table 7) is approximately the same in initial deviations of all sizes from esophoria to an esotropia of 50+ prism diopters. It should be noted that myopia is somewhat more

frequently seen in large rather than small deviations (e.g. E shows three percent of myopes, while ET=50+ prism diopters shows myopia in 8.1 percent of all patients). Conversely, high hyperopia is more frequently seen in esophoria or intermittent esotropia (17.0 percent) than in large esotropia (1.7 percent). *However, in general, the size and type of refractive error would seem to have little influence upon the size of the initial deviation.*

Recalling that initial examination may have taken place at any time from age six months to five years, it then seemed important to decide whether the initial refractive errors varied significantly depending upon the age at which the first examination took place. In other words, if the patient is not examined until one, two, or more years after the onset of esotropia, has the refractive error changed significantly and thus made the pooling of initial refraction (regardless of the age of the patient) invalid? Can we, by comparing those patients who were examined during the first year of life with those who were not seen until later, gain significant information? With this in mind, the refractive error was compared for those patients first examined during the first year, during the second year, and after the second year (see Table 8).

TABLE 8. RELATIONSHIP OF REFRACTIVE ERROR TO AGE AT FIRST EXAMINATION

| | *1st year* | *2nd year* | *After 2nd year* |
|---|---|---|---|
| Myopia | 4 (3.8%) | 9 (5.7%) | 15 (6.8%) |
| 0 to +2.00Sph | 58 (54.8%) | 70 (40.0%) | 96 (42.9%) |
| +2.25Sph to +5.00Sph | 42 (39.4%) | 68 (42.8%) | 93 (31.6%) |
| +5.25Sph + | 2 (2.0%) | 12 (7.5%) | 19 (8.7%) |
| TOTAL | 106 | 159 | 223 |

It would seem that a somewhat higher percentage (see Table 8) examined during the first year of life had a hyperopia of 0 to +2.00 diopters (54.8 percent) than when seen after the first year (40.0 percent and 42.9 percent). Those having a hyperopia of +2.25 to +5.00 diopters were almost identical in numbers regardless of the age of examination. A somewhat higher percentage of high hyperopes (over +5.00 diopters) were found after the first year (7.5 percent and 8.7 percent) than during the first year (2.0 percent). This tendency to increasing hyperopia has been demonstrated by Brown.[54] Since changes in refraction were small during the first several years, we may assume that the initial refractions used in Table 7 are valid for the conclusion arrived at.

RELATIONSHIP OF INITIAL DEVIATION TO FINAL FUSION. The influence of the size of the initial deviation upon the incidence of final cure is of considerable prognostic importance. Does a large deviation make the attaining of fusion more difficult than does a smaller deviation?

TABLE 9. RELATIONSHIP OF SIZE OF INITIAL DEVIATION TO FINAL FUSION

| | E−E(T) | ET=5−24$^{\Delta}$ | ET=25−49$^{\Delta}$ | ET=50$^{\Delta}$+ |
|---|---|---|---|---|
| No fusion | 7 (10.0%) | 24 (22.8%) | 40 (30.5%) | 76 (40.0%) |
| Fusion not maintained | 6 (8.6%) | 25 (23.8%) | 32 (24.4%) | 39 (20.0%) |
| Worth fusion | 34 (48.6%) | 32 (30.5%) | 32 (24.4%) | 48 (25.0%) |
| Gross stereopsis | 18 (25.7%) | 19 (17.2%) | 23 (17.0%) | 26 (13.5%) |
| Refined stereopsis | 5 (7.1%) | 6 (5.7%) | 5 (3.7%) | 3 (1.5%) |
| TOTAL | 70 | 106 | 132 | 192 |

It will be noted (Table 9) that 40 percent of the initial deviations of 50 prism diopters and greater showed no fusion while 30.5 percent of ET=25–49 prism diopters, 22.8 percent of ET=5–24 prism diopters, and 10 percent of E-E(T) showed no fusion—no fusion becoming progressively more frequent as the deviation increases in size. To put it another way, the smaller the initial deviation, the more often is there some evidence of final fusion. It will be noted that in E–E(T) a definitely higher percentage showed fusion (Worth 48.6 percent, gross stereopsis 25.7 percent, refined stereopsis 7.1 percent) than in other groups of initial deviations. However, there is a moderate difference in final fusion in the three groups of manifest initial deviation (ET=5–24 prism diopters = 53.4 percent; ET=25–49 prism diopters = 45.1 percent; ET=50 prism diopters + = 40.0 percent). This is also evident in determination of refined fusion, the higher percentage being present in the small deviation (7.1 percent) and the lowest percentage in the large deviation (1.5 percent).

We may conclude that *the prognosis for cure is best in the initial latent deviations, and less good in the larger manifest ones,* and *even refined stereopsis is possible and more often in the smaller deviations.*

There is the possibility that the true initial deviation may have been different from that determined at the first examination, that is, deviation of 25–50 prism diopters when first examined at the age of four years might well have been E–E(T) if it had been first examined before the age of one year. Such a patient, having had an early opportunity for binocular single vision, might reasonably be expected to have a better final fusion (as seemed indicated by Table 9). With this in mind, patients were divided into three groups (those first

examined during the first year of life, those examined during the second year, and those not examined until after the second year of life), and the cure rate for these groups determined. If cure rate is similar, it may be assumed that the size of the initial deviation and age of onset within the first year are valid.

TABLE 10. RELATIONSHIP OF AGE AT EXAMINATION TO FUSION

| | *Exam 1st year* | *Exam 2nd year* | *Exam 2+ years* |
|---|---|---|---|
| No fusion | 36 (31.6%) | 54 (33.5%) | 57 (25.3%) |
| Fusion not maintained | 23 (20.0%) | 35 (21.7%) | 44 (19.7%) |
| Worth fusion | 35 (30.1%) | 46 (28.6%) | 65 (29.0%) |
| Gross stereopsis | 16 (14.0%) | 24 (14.9%) | 46 (20.3%) |
| Refined stereopsis | 4 (3.7%) | 2 (1.3%) | 13 (5.7%) |
| TOTAL | 114 | 161 | 225 |

It will be noted (Table 10) that in the first two columns the percentages are almost identical, suggesting that *for the patient no older than two years, the history of the time of onset and size of the initial deviation are probably valid*, since the cure rate is constant within these ages. The third column shows a lower percentage of no fusion and somewhat higher percentage of gross and refined fusion than in the first two columns. The cure rate is higher in patients not seen until after the second year of life, thus suggesting that *if examination is deferred until after the second year of life, the history of age of onset and size of initial esotropia become less valid.*

RELATIONSHIP OF INITIAL HYPERTROPIA TO FINAL FUSION. What influence does the presence of an initial vertical deviation have upon the hope for eventual cure? Of 139 patients exhibiting some vertical deviation (always associated with an eso-deviation) at the first examination, nine had a hyperphoria, and of these three obtained no fusion, two Worth four-dot fusion, and two gross stereopsis. One hundred and thirty had a frank hypertropia at the first examination, and of these 51 never obtained fusion, 35 demonstrated fusion at some examination but not at the final one, while at the final examination 29 had Worth fusion, ten gross stereopsis, and five refined stereopsis. It may be said that *while the presence of a vertical deviation at the first visit is a bad prognostic finding, it is not an absolute bar to obtaining fusion, even a high degree of stereopsis.*

RELATIONSHIP OF FINAL DEVIATION TO FINAL FUSION. Implicit in some of the questions already asked is the question, what effect if any does the presence of a final deviation (horizontal or vertical) have

upon the possibility of demonstrating fusion? With this in mind, the cure relationships of final manifest and latent, horizontal and vertical deviations were investigated.

TABLE 11. RELATIONSHIP OF FINAL DEVIATION TO FINAL FUSION

| | *Horizontal phoria* | *Horizontal tropia* | *Hypertropia only* | *Horizontal tropia and HT* |
|---|---|---|---|---|
| No fusion | 18 (7.7%) | 30 (34.5%) | 19 (34.0%) | 80 (65.0%) |
| Fusion, not maintained | 25 (10.7%) | 29 (33.3%) | 15 (26.8%) | 33 (22.6%) |
| Worth fusion | 102 (43.6%) | 19 (21.8%) | 16 (28.6%) | 9 (7.4%) |
| Gross stereopsis | 71 (30.3%) | 8 (9.2%) | 6 (10.6%) | 1 (0.8%) |
| Refined stereopsis | 18 (7.7%) | 1 (1.2%)* | 0 | 0 |
| TOTAL | 234 | 87 | 56 | 123 |

Reference to Table 11 demonstrates clearly that, as might be expected, the presence of a final horizontal heterophoria was no bar to cure, 43.6 percent showing Worth fusion, while 30.3 percent had gross stereopsis, and 7.7 percent had a high degree of refined stereopsis (fusion in a total of 81.6 percent). However, when a residual horizontal or vertical heterotropia was present, the percentage of fusion (32.2 percent and 39.2 percent respectively) is definitely less, and when both horizontal and vertical heterotropia are present in the same patient, the results (fusion in 8.2 percent) are even less good.

It is interesting to note that a number of frank tropias seemed to show Worth fusion and gross stereopsis. Most of these were very small manifest deviations. As pointed out under "Examination (IV.B.6)," tests for Worth fusion and gross stereopsis may not require bifoveal fixation, but rely rather on peripheral fusion or even small angle abnormal retinal correspondence. One such patient* even seemed to demonstrate a high degree of stereopsis. This can hardly be accounted for except as an error in interpreting the test.

We may conclude that *the presence of either a horizontal or a vertical heterotropia at final examination makes fusion very difficult to obtain and when both horizontal and vertical deviation are present, any stereopsis is impossible.*

TIME

Certain factors related to time are of interest: time of onset of squint, duration of squint, age at first alignment, and age at first surgery. A very vital factor, the age of onset of strabismus, brings up important questions. How valid is the history of age of onset obtained from parents? Within the first year of life, is there a difference in

prognosis depending upon the age of onset? Is the duration of squint truly important or is the absolute age of onset more so?

RELATIONSHIP OF AGE OF ONSET TO INITIAL DEVIATION. As previously stated, within the first year three age periods might seem significant: 0 through three months, four through six months, and seven through eleven months. What relationship is there between age of onset and the size of the deviation?

TABLE 12. RELATIONSHIP OF AGE OF ONSET TO INITIAL DEVIATION

| | *Onset* 0–3 *mos.* | *Onset* 4–6 *mos.* | *Onset* 7–11 *mos.* |
|---|---|---|---|
| E or E(T) | 44 (13.2%) | 3 (5.9%) | 23 (20.0%) |
| ET = 5 – 24$^{\Delta}$ | 57 (17.1%) | 16 (30.7%) | 33 (28.7%) |
| ET = 25 – 49$^{\Delta}$ | 81 (24.3%) | 13 (25.0%) | 38 (33.0%) |
| ET = 50$^{\Delta}$ + | 151 (45.4%) | 20 (38.4%) | 21 (18.3%) |
| TOTAL | 333 | 52 | 115 |

Reference to Table 12 shows that even when onset is during the first three months of life, 13.2 percent of 333 patients would seem to have an esophoria or intermittent esotropia, 17.1 percent deviate 5–24 prism diopters, 24.3 percent deviate 25–49 prism diopters, and 45.4 percent deviate more than 50 prism diopters. This finding is contrary to the common thinking that all congenital esotropes have large deviations. Not quite one-half of all congenital esotropes deviate 50 prism diopters or more, while one-seventh have eyes approximately straight at times (esophoria or intermittent esotropia). Contrasted with this is the older group (seven through eleven months) of whom more are approximately straight (20 percent) than have a large esotropia (18.3 percent). It becomes then apparent that *even within the first year of life, the later the onset, the more moderate is the initial strabismus.* Also, *congenital esotropia does not always show a large deviation.*

Again the question arises—Is the history of the age of onset of strabismus valid? In an effort to redetermine this validity, all patients whose onset was presumably during the first three months of life were divided into three groups: (1) those first examined during the first year of life; (2) those first examined during the second year of life; and (3) those first examined after the second year of life.

Referring (see Table 13) only to those patients whose onset was presumed to be during the first three months of life, it will be noted that the percentage of large deviations (50 prism diopters or more) decreases from 70.5 percent for those examined during the first year

TABLE 13. INTERRELATIONSHIP OF AGE AT FIRST EXAMINATION, HISTORY OF THE AGE OF ONSET, AND THE SIZE OF THE INITIAL DEVIATION

| | *Examined first year* | | | *Examined second year* | | | *Examined after second year* | | |
|---|---|---|---|---|---|---|---|---|---|
| | *Onset* 0–3 *mos.* | *Onset* 4–6 *mos.* | *Onset* 7–11 *mos.* | *Onset* 0–3 *mos.* | *Onset* 4–6 *mos.* | *Onset* 7–11 *mos.* | *Onset* 0–3 *mos.* | *Onset* 4–6 *mos.* | *Onset* 7–11 *mos.* |
| E–E(T) | 11 (11.7%) | 1 | 2 | 9 (8.4%) | 1 | 9 | 24 (18.3%) | 1 | 12 |
| ET = 5–24Δ | 8 (8.3%) | 2 | 0 | 7 (6.6%) | 6 | 5 | 42 (32.1%) | 8 | 28 |
| ET = 25–49Δ | 9 (9.5%) | 4 | 1 | 36 (33.6%) | 3 | 17 | 36 (27.5%) | 6 | 20 |
| ET = 50Δ+ | 67 (70.5%) | 7 | 1 | 55 (51.4%) | 5 | 11 | 29 (22.1%) | 8 | 9 |
| TOTAL | 95 | 14 | 4 | 107 | 15 | 42 | 131 | 23 | 69 |

of life to 51.4 percent for those not seen until the second year, and further to 22.1 percent for those not examined until after the second year of life. This discrepancy could only be accounted for if the esotropia spontaneously has decreased in size as the patient grows during the first few years of life, but this has not been our experience. Esotropia rarely improves spontaneously during these years. The other possible explanation is that onset of true esotropia was not as early as the history would suggest and that a smaller percentage of high deviations has developed. This is borne out by the increasing percentage of moderate and smaller initial deviations when the patient is first examined after the first year of life.

Using only the size of deviation at the first examination as a standard, *history alone would seem not to be a valid criterion for establishing the age of onset of the deviation.*

RELATIONSHIP OF INITIAL REFRACTIVE ERROR TO AGE OF ONSET. Does the size and type of refractive error influence the age of onset of strabismus? It has been previously noted that the size and type of refractive error varies little with the size of the initial deviation (p. 413). Table 14 would suggest that the relationship between refractive error and the age of onset (within the first year) is also rather constant.

TABLE 14. RELATIONSHIP OF INITIAL REFRACTIVE ERROR TO AGE OF ONSET

| | *Onset* 0–3 *mos.* | *Onset* 4–6 *mos.* | *Onset* 7–11 *mos.* |
|---|---|---|---|
| Myopia | 17 (5.0%) | 4 (7.7%) | 7 (6.0%) |
| 0 to +2.00Sph | 160 (48.1%) | 22 (42.3%) | 47 (40.9%) |
| +2.25Sph to +5.00Sph | 141 (42.4%) | 23 (44.2%) | 45 (39.2%) |
| +5.25Sph + | 15 (4.5%) | 3 (6.8%) | 16 (13.9%) |
| TOTAL | 333 | 52 | 115 |

There is a slightly higher percentage of low hyperopes in the 0-to-three months onset than later, and slightly higher percentage of high

hyperopes in the seven-to-eleven months onset, though the differences are not great. Accommodation, with its attendant accommodative convergence is probably slightly more of a factor in causation of esotropia in the seven-to-eleven months group than earlier. We may conclude, however, that *the size and type of refractive error plays little part in the age at which esotropia appears during the first year of life.*

RELATIONSHIP OF AGE OF ONSET TO FINAL FUSION. The significance of the age of onset to the final cure has been presumed to be of prime importance. When the 0-to-three month and the seven-to-eleven month groups (Table 15) are considered, it is noted that 31.2 percent of the

TABLE 15. RELATIONSHIP OF AGE OF ONSET TO DEGREE OF FINAL FUSION

| | *Onset* 0–3 *mos.* | | *Onset* 4–6 *mos.* | | *Onset* 7–11 *mos.* | |
|---|---|---|---|---|---|---|
| No fusion | 104 (31.2%) | | 21 (40.0%) | | 22 (19.2%) | |
| Fusion, not maintained | 56 (16.8%) | | 14 (27.0%) | | 32 (27.8%) | |
| Worth fusion | 108 (32.5%) | 173 | 9 (17.3%) | 17 | 29 (25.2%) | 61 |
| Gross stereopsis | 58 (14.4%) | (52.0%) | 5 (10.0%) | (32.9%) | 23 (20.0%) | (53.0%) |
| Refined stereopsis | 7 (2.1%) | | 3 (5.7%) | | 9 (7.8%) | |
| TOTAL | 333 | | 52 | | 115 | |

first group and only 19.2 percent of the second group had no fusion suggesting that the early opportunity for fusion in the older group is helpful. It is interesting that 16.8 percent of the younger group gained but did not maintain fusion, while 27.8 percent of the older group fell in the same category. Practically equal percentages of the two groups showed evidence of fusion (52.0 percent to 53.0 percent) at the final visit though a significantly higher percentage of the older group demonstrated gross or refined stereopsis. We may assume that (1) *some evidence of fusion may be obtained in about equal percentage in the earliest and later onset groups*, and (2) *stereopsis is possible to attain even in the esotropes of very early onset but the prognosis is better in later onset.*

RELATIONSHIP OF DURATION OF SQUINT TO FINAL FUSION. When the subject of duration of squint and its relationship to final cure is investigated, the results are equivocal. It has been said that if a squint endures more than ten percent of life in the one-year-old and twenty percent of life in the two-year-old, fusion is difficult or impossible to attain.

Of 356 infantile esotropes (Table 16) who had been operated and had attained alignment, 74 did not show fusion, 97 fused but did not maintain it, 108 obtained Worth fusion, 63 gross stereopsis and 14 refined stereopsis. If these various categories are spotted with reference

TABLE 16. RELATIONSHIP OF DURATION OF SQUINT TO FUSION

| | | *Years of duration of squint* | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | ½ | 1 | 1½ | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | 11 |
| No fusion | (74 pts) | | 2 | 10 | 17 | 12 | 11 | 9 | 4 | 2 | 1 | 2 | 2 | 1 |
| Fusion, not maintained | (97 pts) | 1 | 6 | 8 | *21* | *24* | 15 | 11 | 11 | | | | | |
| Worth fusion | (108 pts) | | 13 | 14 | *22* | *19* | *21* | 13 | 3 | 7 | 1 | | | |
| Gross stereopsis | (63 pts) | 1 | 2 | 5 | 12 | *21* | 11 | 5 | 2 | 1 | 2 | | | 1 |
| Refined stereopsis | (14 pts) | | | 1 | 1 | 2 | *4* | *4* | | 1 | 1 | | | |
| TOTAL | 356 patients | | | | | | | | | | | | | |

to the duration of the squint, it will be noted that the peak of incidence of Worth fusion spreads from the second through the fourth year after onset. Gross stereopsis had its greatest incidence at a duration of squint of three years and refined stereopsis seemed greatest at the fourth and fifth year. It will be noted also that several squints endured five years or more and subsequently demonstrated stereopsis.

From this we may conclude that *duration of squint, either in terms of a finite period or of a percentage of life, is not a valid prognostic criterion for cure.*

RELATIONSHIP OF AGE AT FIRST ALIGNMENT TO FINAL FUSION. When the visual axes have been aligned and amblyopia does not exist, the opportunity for fusion may be presumed. However, it is not known whether the early alignment actually favorably influences final fusion.

TABLE 17. RELATIONSHIP OF AGE AT FIRST ALIGNMENT TO FINAL FUSION

| | *Age at first alignment* | | | | |
|---|---|---|---|---|---|
| | 0–1 *yrs.* | 1–3 *yrs.* | *over* 3 *yrs.* | *None* | *Total* |
| No fusion | 1 (9.1%) | 34 (21.7%) | 39 (20.9%) | 62 | 136 |
| Fusion, not maintained | 3 (27.3%) | 40 (25.5%) | 53 (28.3%) | 1 | 97 |
| Worth fusion | 6 (54.5%) | 51 (32.5%) | 51 (27.3%) | 2 | 110 |
| Gross stereopsis | 1 (9.1%) | 29 (18.4%) | 33 (17.6%) | 0 | 63 |
| Refined stereopsis | 0 | 3 (1.9%) | 11 (5.9%) | 0 | 14 |
| TOTAL | 11 | 157 | 187 | 65 | |
| Not operated: 80 | | | | | |

Unfortunately, too few esotropes (Table 17) had been aligned before the age of a year to have statistical significance. On considering those patients first aligned at ages one to three years, and those not aligned until after the age of three years, there would seem to be no significant difference in those showing no fusion (21.7 percent and 20.9 percent) and those gaining but not maintaining fusion (25.3 per-

cent and 28.3 percent). Those aligned at one to three years did show 32.5 percent Worth fusion as compared with 27.3 percent for those aligned later. Gross stereopsis was about equal in the two groups, but the later alignments showed more refined stereopsis than the earlier. These statistics *would not suggest that the age at alignment is an important factor in final fusion.*

RELATIONSHIP OF AGE AT FIRST SURGERY TO FINAL FUSION. If early alignment does not give a definitely better percentage of cure than when aligned later, is there any merit to early surgery for strabismus?

TABLE 18. RELATIONSHIP OF AGE AT FIRST SURGERY TO FINAL FUSION

| | *Age at first surgery* | | |
|---|---|---|---|
| | 0–1 *yr.* | 1–3 *yrs.* | *over* 3 *yrs.* |
| No fusion | 5 (16.2%) | 71 (36.6%) | 60 (30.9%) |
| Fusion, not maintained | 6 (19.3%) | 46 (23.7%) | 45 (23.2%) |
| Worth fusion | 13 (41.9%) | 53 (26.8%) | 44 (22.7%) |
| Gross stereopsis | 7 (22.6%) | 23 (11.9%) | 33 (17.0%) |
| Refined stereopsis | 0 | 2 (1.0%) | 12 (6.2%) |
| TOTAL | 31 | 195 | 194 |
| Not operated: 80 | | | |

Note (Table 18) that when surgery is performed before the age of one year, even though alignment was not attained, fewer patients showed no fusion (19.3 percent), and more showed Worth fusion (41.9 percent) and gross stereopsis (22.6 percent), than when surgery was performed later. However, no patient operated before the age of one year showed refined stereopsis, while 6.2 percent of those operated after the age of three years had refined stereopsis. Recall that all patients operated before the age of one year had an onset of esotropia within the first three months of life, while many of those operated later had their onset during the last half of their first year.

There would seem to be *some merit in performing early surgery, even though alignment is not attained at the first operation.* The early changing of alignment by surgery seems to favorably influence final fusion when alignment is finally attained. Further, the very considerable psychological benefit to the patient and parents by the improved appearance must not be overlooked.

SURGERY

In at least one series,[2] patients who had been operated more than once were discarded from further consideration in determining the prerequisites for cure of squint. It is, now, important to show the

TABLE 19. RELATIONSHIP OF NUMBER OF OPERATIONS TO INCIDENCE OF CURE

| | *No. cases* | *Alignment and fusion* | *No alignment but fusion* | *Alignment but no fusion* | *No alignment or fusion* |
|---|---|---|---|---|---|
| No operation | 80 | 59 (76.0%) | 4 (5.0%) | 5 (6.0%) | 12 (13.0%) |
| 1 operation | 228 | 87 (38.4%) | 16 (7.0%) | 40 (17.4%) | 85 (38.2%) |
| 2 operations | 145 | 52 (36.0%) | 16 (11.0%) | 22 (15.0%) | 55 (38.0%) |
| 3 operations | 41 | 13 (32.0%) | 2 (4.9%) | 13 (32.0%) | 13 (31.1%) |
| 4 or more operations | 6 | 3 (50.0%) | 0 | 1 (16.6%) | 2 (33.4%) |

relationship of final fusion to the number of operations performed. Reference to Table 19 will show that of 500 patients, 80 were unoperated, 228 were operated once, 145 operated twice, 41 operated three times, and only six operated four or more times. Following final surgery, three combinations of relationship existed: some were aligned and had fusion; some were aligned but demonstrated no fusion (either because of the age of the patient or because fusion did not exist); and some seemed to have a small heterotropia but still had fusion on some tests (see page 417).

It becomes immediately apparent that alignment and fusion may be attained by a second (36 percent) or third (32 percent) operation in at least one-third of the cases operated, and that alignment may be obtained even though without fusion in an additional 15 percent by a second operation, and 31.6 percent by a third operation. It is interesting to note that in 80 patients not operated, only 76 percent eventually had straight eyes and fusion. *We may conclude that multiple operations are quite worthwhile when indicated and offer a considerable additional chance for cure.*

## COMMENT

Some additional discussion of several points seems wise.

### AGE OF ONSET

Many authors have admitted that the history of the age of onset of strabismus as obtained from parents or other relatives is far from reliable. None the less, most authors give great prognostic importance to this same age of onset. If the history is not reliable, of what value is the presumed age of onset as a prognostic criterion? Many small children have the appearance of esotropia when none may exist. One parent may state that the onset was at ten months, while the other will claim six months, and the grandparents are equally emphatic that the child was born cross-eyed.

With this problem in mind, 753 patients, whose presenting complaint

was the appearance of strabismus, were carefully examined (see page 403). Almost 50 percent of such patients were found to have straight eyes, although an appearance of esotropia was present because of epicanthus, narrow interpupillary distance, facial asymmetry, or a negative angle kappa. As previously stated, a squint-conscious community and an ophthalmic office treating a high percentage of strabismus may have encouraged an unduly large number of pseudo-squints to be presented for examination.

None the less, even if a lesser percentage of all patients presenting because of the appearance of strabismus were found to have only pseudo-strabismus, the problem of dating the onset of true strabismus becomes a major one. Superimposed on this is the fact that many early esotropes have a small or intermittent deviation (see page 413), making the date of onset even less absolute.

In an effort to clarify this matter, several studies were done, comparing the findings and results of treatment in those 114 patients who were examined before the age of one year (and thus the diagnosis of infantile esotropia established), and those results obtained on 386 patients who were examined after the age of one year but with the onset presumed from the history alone.

Using the cure rate as a criterion (page 416), it was concluded that if the patient was examined before the age of two years, the presumed age of onset would seem valid, while if after the age of two years, the diagnosis was less reliable. However, if the size of the initial deviation was used as a criterion (page 418), then those patients examined by the age of a year showed a much higher percentage of high esotropia, while those not examined until the second year or later (and thus the age of onset only obtained by history) showed a much smaller percentage of high esotropia, and a much larger percentage of small and moderate deviation.

It must be generally concluded, then, that *the age of onset of esotropia is in doubt unless the patient is examined at a very early age.* Even then, unless the deviation is large, there is the possibility that esophoria or intermittent esotropia existed at first (and thus the early opportunity for binocular single vision) and that the manifest deviation came on at some later time. This is very discouraging if much prognostic significance is placed upon the age of onset, or the duration of the squint.

The age of onset within the first year would seem to influence the incidence of cure very little, though I had fully expected that it would

(see page 420). Presumably a period of established binocular single vision longer than several months is necessary before the onset of esotropia, if the chances for cure are to be favorably influenced.

The presumed age of onset is influenced only slightly by the size of the initial refractive error as might be expected. The child of six months or less is unable to use his accommodation even though a considerable hyperopia may be present, and thus little accommodative convergence is evidenced. Many infantile esotropes having a hyperopia of three to five diopters showed no improvement in the esotropia when given spectacles or miotics. However, after surgical correction of the deviation (and the passage of several weeks or months), a very large accommodative element could be demonstrated and controlled by spectacles or miotics. The patient had had an accommodative element masked by non-use before surgery, but becoming manifest after surgery and the passage of time. It is a mistake to suggest that the size of the refractive error is not important in the young squinter.

### FACTORS INFLUENCING CURE OF INFANTILE ESOTROPIA

What factors other than the age of onset seem to influence the cure of strabismus?

Mild or moderate amblyopia usually responds well to treatment (page 411) and its presence at the first examination does not unfavorably affect final cure. On the other hand, severe initial amblyopia (20/200 or less) responds moderately well to treatment but gives a poor prognosis for final fusion, and if the severe amblyopia persists, any fusion is precluded (page 412).

A final visual acuity of 20/200 or less precludes any fusion, while a visual acuity of 20/100 or better may allow Worth fusion and gross stereopsis. A visual acuity of 20/40 or better is necessary for refined stereopsis. It should be noted, however (page 411), that even if no amblyopia exists, only about 30 percent of all infantile esotropes show Worth fusion, 18 percent show gross stereopsis, and less than 4 percent obtain refined stereopsis.

When the influence of the deviation on final cure is investigated, it is noted that the presence of a large esotropia or any manifest hypertropia (page 417) makes the attaining of final fusion difficult. When a manifest deviation persists to the final examination, no patient attained refined stereopsis and only about ten percent demonstrated gross stereopsis. It can be said that a large initial devia-

tion or the presence of a final manifest deviation makes cure very difficult.

It was felt that if a patient could obtain alignment very early in life as the result of surgery, the opportunity for binocular single vision would then be present and a higher percentage of cure result. This does not seem to be true unless such alignment can be attained during the first year of life and maintained thereafter. Of course, any patient whose deviation was latent at first and continued so has a much better prognosis. It was interesting to note that occasionally even though alignment was not attained, the mere fact of changing alignment by early surgery seemed to improve the prognosis.

### CHARACTERISTICS OF INFANTILE ESOTROPIA

It is quite evident that all infantile esotropes do not adhere to a single pattern.

Contrary to some opinions, amblyopia is not uncommon, about eight percent having severe amblyopia while 30 percent had mild to moderate amblyopia. However, most of these responded rather well to treatment.

Refractive errors encompass almost the normal range for strabismus in general. There were 5.6 percent myopia, 46.4 percent low hyperopia, 41.6 percent moderate hyperopia, and 6.4 percent high hyperopia. The number of myopes is somewhat greater and high hyperopes somewhat fewer than the average found in older esotropes.

The initial deviations varied from esophoria to large esotropia (80+ prism diopters). A surprising number had a latent or intermittent deviation (14.0 percent) at the first visit. Small deviations were found in 21.2 percent, and moderate deviations in 26.4 percent. There were 38.4 percent esotropias of over 50 prism diopters. Thus, it is apparent that the range of initial deviation is considerable, though the large deviations predominate.

Weakness of abduction has been cited as a characteristic of infantile esotropia and, at times, it has been ascribed to pareses of the lateral recti. Of 165 patients on whom such a finding was noted, 81 had definitely poor abduction initially, but as the result of the passage of time or surgery on the medial recti, only 12 showed a residual weakness of abduction. It would seem that cross fixation and an habitual lack of use of abduction may well have been at fault.

Thus, infantile esotropes encompass the range of visual acuity, refractive errors, and initial deviations, and do not adhere to a constant pattern.

## SUMMARY AND CONCLUSIONS

(1) The records of 2,393 esotropes were reviewed, 1,152 infantile esotropes were found and of these 500 were satisfactory for analysis. The diagnosis of infantile esotropia was established by examination of 114 patients, and by history of onset in 386 patients.

(2) The defining of cure and methods of testing are described.

(3) An attempt has been made to demonstrate the lack of uniformity of characteristics of infantile esotropia, and to evaluate the factors relating to the cure of such esotropia.

(4) It is shown that approximately 30 percent of infantile esotropes may attain and retain Worth fusion, 15 to 20 percent attain gross stereopsis, and 3 to 5 percent refined stereopsis. About 70 percent of all infantile esotropes demonstrate fusion at some time during treatment but 20 percent fail to maintain fusion.

(5) The validity of the history of age of onset was challenged because of (*a*) the fact that 50 percent of another series was found to have pseudosquint rather than true squint, and (*b*) the size of the initial deviation differed considerably when presumed infantile esotropes were first examined at various ages—the first recorded deviation being smaller as the age at first examination increased.

(6) The most favorable findings for eventual cure of infantile esotropia are (*a*) a latent or intermittent deviation or a larger esotropia reduced in amount by early surgery, (*b*) the absence of any vertical component, and (*c*) an initial visual acuity of at least 20/100 in the poorer eye.

(7) Those factors seeming to be of little consequence in the cure of infantile esotropia are (*a*) the age of onset (within the first year of life), (*b*) the duration of the squint, (*c*) the age at first alignment, and (*d*) the size of the refractive error.

(8) Those factors found to be most unfavorable to cure were (*a*) severe amblyopia, especially if it persists, (*b*) an initial manifest hypertropia, and (*c*) a final manifest deviation.

(9) Esotropia of very early onset can be cured but only if all factors are favorable.

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