Congenital Esotropia: Definition, Course, and Management

Paul E. Romano, M.D.
Chicago, Illinois

The purpose of this paper is to consider three aspects of congenital esotropia: first, the definition of "congenital" esotropia; second, the etiology, course, and prognosis of this condition; and third, its management, especially the When? and Why? of its surgical correction.

There are many truly congenital conditions, but the term congenital strabismus is misleading. Congenital means "present at birth," but no child has straight eyes and normal binocular vision at birth. Most children's eyes are divergent at birth. The rest may be close to parallel, but normal eye movements are present in none. Nor is normal monocular fixation or binocular cooperation present. Therefore, literally all children are strabismic at birth. "Congenital" strabismus, rather than implying a pathologic defect, may be said to be the normal state of affairs.

By common usage, however, the term congenital esotropia is usually meant to designate or isolate those children who have an esodeviation and have never had, by virtue of the early onset of their deviation, any opportunity for normal binocular vision. The term is used to separate them from those who acquire a deviation after having had binocular vision for a period of time, or whose deviation is the result of accommodative factors.

Most of the components of normal binocular vision and motility develop in the first three months of life. First there are conjugate searching movements, then conjugate fixation saccades, then conjugate pursuit movements. During this period the eyes become parallel if they were divergent at birth. It is the infants who do not follow this pattern but who become esotropic instead that we wish to specify with this term. The development of these conjugate and parallel eye movements is well advanced by four months of age. Normal responses to the four diopter prism test at four months indicate a high degree of binocular cooperation at this age as well. Soon after the fourth month, however, the accommodation and convergence mechanisms start to develop rapidly and are well established by six months.

It is appropriate, therefore, to define congenital esotropia as the presence of a constant esodeviation at four months of age. Such a definition has been implied by Lang. Such a child has had no real opportunity for normal binocular vision and the deviation is not the result of hyperopia or accommodation.

Since this definition is not generally accepted, the following remarks on the condition and its management must be based...
on previous definitions of congenital esotropia. Most workers in this area have used as a definition “a constant esotropia at six months of age,” as proposed in 1950 by Costenbader. This group may be contaminated by some cases of accommodative esotropia. Occasional authors may not be so strict in their case selection or in their historical proof of the deviation.

What is the etiology, course, and prognosis of this disease, congenital esotropia?

An occasional child is esotropic at birth due to a specific neurologic disease or from abducens paralyses incurred during birth. For the majority of congenital esotropes, however, the specific etiology is essentially unknown. The condition is strongly hereditary but no specific gene or chromosome defect has been found responsible.

Characteristically, congenital esotropes have a large deviation of 50 or more prism dipters. Cross fixation is usually present and, since this is an alternating fixation, amblyopia is usually not a problem. Some do, however, have varying degrees of preference for fixation with one eye, and consequently varying degrees of amblyopia in the nonpreferred eye. They usually have a small hyperopic refractive error typical for their age. Overacting inferior oblique muscles may be seen, but more striking and consistent is a marked limitation of abduction bilaterally.

This limitation of abduction is often mistaken for bilateral sixth nerve paralyses but it is only a manifestation of cross fixation. Full abduction can often be demonstrated by occlusion of one eye at a time, or by passive “doll’s head” manipulation, or the induction of nystagmus, rotary or caloric. Personally, I have found most of these children to demonstrate good abduction, but often not really full abduction. This may be due to contracture of the medial recti muscles. Proof that mild palsies does not in fact exist is exceedingly difficult in these children.

Alternating sursumduction, latent nystagmus, and abnormal head posture may also be present. The course of congenital esotropia is not well documented. Many adults claim to have “grown out of” crossed-eyes in childhood. Most of these have probably only outgrown a pseudoesotropia from prominent epicanthal folds. Under age two, pseudoesotropia is quite common. In regard to true congenital esotropia, however, there are no studies which indicate how often the condition is present at birth, or how soon after birth the deviation becomes manifest, if it was not present at birth. Nor are there data to indicate the subsequent course of the deviation. There are no data to show whether the esodeviation remains the same or changes with the passage of time. Overacting inferior obliques do seem to become either more frequent or perhaps just more obvious with increasing age.

What is the ultimate prognosis for these children with regard to achieving normal binocular single vision, to be “cured” of their strabismus? For this, the prognosis is poor and Worth’s original opinion* that these children have a congenital defect for central fusion seems to prevail in spite of all our efforts. Several reports in the past decade regarding the result of treatment of congenital esotropia indicate that no form of therapy currently available can produce truly normal binocular single vision in these children. Reported “cures” are either limited to cosmetic results or functional results at standards considerably below normal binocular single vision. It seems the best one can hope for, for these children, is a negligible residual deviation and a less than perfect degree of binocular cooperation in the form of a microstrabismus, microtropia, or monofixation syndrome.

The studies mentioned seem to point to the possibility of obtaining better results by operating at an earlier and still earlier age. Surgery has been done as early as six months of age without significantly better results than those obtained by operating a year or two later. Surgery before six months of age might bring a better functional result. Dr. Marshall Parks has such a group of patients, small in number, on whom he has operated for congenital esotropia at the age of four months or so. Although these children are still too young for complete functional testing, he has found no evidence yet that the results in this group are going to be any better than previous groups.*

Better functional results in congenital esotropia must await a better understanding of the etiology or the development of new methods of treatment. For the present we seem

---

to be stuck with Worth's original guarded prognostication.

Keeping in mind this prognosis, as well as our lack of knowledge concerning the etiology and course of congenital esotropia, let us turn now to its management.

The first, and perhaps most important, principle of management of congenital esotropia, is to find such children as early as possible. This facilitates (1) early and accurate diagnosis as a "congenital" deviation; (2) early treatment of amblyopia which is easier and more successful the younger the child is; and (3) makes early surgery, if indicated, possible. (Many serious ocular conditions present with esotropia, and their early detection is even more important).

If manpower permitted, in keeping with the proposed definition of congenital esotropia, it would be desirable for all children to be screened by an ophthalmologist at the age of three-and-one-half to four months. In the absence of such a screening program, this can only be accomplished by educating physicians and especially pediatricians in the area to the importance of referring such children as soon as possible. One should, of course, have a method of seeing these patients as soon as possible after they are referred. There is no sense in asking for immediate referral and then their having to wait two or three months or more for an appointment.

Presuming that the patient has been referred and seen at an early age, one must first confirm the diagnosis. It has been shown that historical information from parents is notoriously unreliable for observations more than a week or so old. Information regarding ocular deviations from referring physicians is often less reliable, for examination of these small children is anything but easy. Previous photographs of the child may assist in establishing the time of onset of the deviation so parents should be asked to bring any available prints with them. A complete and thorough eye examination is necessary to confirm the deviation and to rule out organic lesions responsible for poor vision in one or both eyes and congenital abducens paralyses or other neurologic problems. An accommodative component to the deviation must be ruled out or treated if the child is more than four months old. Significant anisometropia must also be taken into account.

After these points have been dealt with, the treatment of amblyopia is next. If the fundi are normal and the child does not spontaneously alternate or readily cross-fixate, amblyopia should be assumed present and should be treated until such fixation behavior is obtained.

The best method is full-time conventional occlusion of the preferred eye. In young children, occlusion amblyopia and even eccentric fixation may develop rapidly beneath the patch. Therefore, the child under a year of age should be checked three to four days after the initiation of patching and at weekly intervals thereafter. Elbow splints should be used if required. Other methods of amblyopia therapy are inferior and have no place in this situation. If alternating fixation cannot be obtained, amblyopia therapy may have to be continued intermittently or on a part-time basis until late childhood or adolescence.

It is important to obtain as much resolution of amblyopia as possible prior to attempting surgery as surgery is less predictable in the presence of a markedly amblyopic eye. When a plateau is reached, or alternating fixation obtained, correcting the deviation surgically is the next step.

There is little question that muscle surgery is needed in the vast majority of congenital esotropes. There is, however, a fair amount of controversy as to just When? and Why? In this regard there are five factors which are considered: functional results, cosmesis, secondary muscle contractures, diagnostic and surgical accuracy, and anesthetic morbidity and mortality.

With respect to functional results, the apparent congenital defect for central fusion in these children must be recognized. Lesser forms of binocular cooperation, based on peripheral fusion, such as the monofixation syndrome, can be achieved if surgical correction of the deviation is accomplished prior to age two or two and a half. Correction after this age produces a significantly poorer level of binocular cooperation. Surgical correction prior to the age of one or one and a half does not produce significantly better results."

Should one strongly recommend surgery before age two for this reason? Such forms of

*Nadler, H.: personal communication.
binocular cooperation as monofixation syndrome, microtropia, or microstrabismus are frequently accompanied by gross stereopsis and good peripheral fusion. This is nice, but just how important is it to vision, to visual comfort, or to the ultimate success or accomplishment of the patient? The absence of normal binocular cooperation has never been shown to be a real handicap in life, unless one wishes to be an airline pilot or an orthoptist.

On the other hand, these forms of binocular cooperation are rather stable: vision in the amblyopic eye does not deteriorate, and the eyes retain their parallel or almost parallel position. Although I am not aware of any study which has been done to prove this stability, my own observations support this hypothesis. Surgery can be recommended before the age of two or two and one half so that the patient will have a stable negligible deviation, minimal amblyopia, and secondarily, better binocular cooperation for what it may be worth.

What about cosmesis as an indication for the timing of corrective surgery? It is important that a child have a good self-image so that he can have good self-acceptance. This is also related to acceptance by his peers. Such factors become significant at age three to four, and large deviations should be corrected before that time.

More important is the acceptance or rejection of a deformed child by the parents. The higher the socioeconomic standing of the parents, the more likely that congenital esotropia will be seen as a significant deformity. There may be guilt, however inappropriate, or the blame may be placed on the partner who has had or whose family has had crossed eyes. These problems may be accompanied by the frank rejection of the child by one or both parents. Psychotherapy is perhaps needed, but surgical correction of the deformity is easier, quicker, and a more permanent solution for all.

Cosmesis, therefore, is an indication for surgery before age four in all children with a significant deviation, and can be an indication for surgery as early as one is willing to do it when the cosmetic deformity is hampering interpersonal relationships in the family.

Another factor sometimes raised to provide an indication for surgery before the age of one is secondary contractures of the medical rectus muscles. This and other anatomic changes in the fascial layers of the globe secondary to the chronically adducted position are postulated to make surgical correction more difficult at a later date. As attractive as this hypothesis is, I am unaware of any evidence that this does occur in fact. By itself, it cannot be considered a good indication for surgery before the age of one.

Difficulty in evaluating the deviation in a very young child is a factor upon which some advocate a delay of surgery, even to the age of four or five. Difficult, yes, but each surgeon must decide this issue for himself in any given case. Doctors differ in their ability to handle children and obtain information from them, and children differ in their degree of cooperation both individually and with respect to different doctors. It is a matter of rapport and this cannot by itself give rise to a general rule about when to do surgery. If sufficient information can be obtained to ensure choosing the proper procedure, then other factors previously mentioned will help in the decision of When? If the required information cannot be obtained, then there are no indications for surgery in this condition which are worth the risk of performing the wrong operation.

Anesthesia, as a factor in deciding when to operate, has become less and less important in the past two decades. This is especially true if one is fortunate enough to have anesthetologists who work with children all the time. In such hands, a six-month-old is as safe as a 6- or 16-year-old. Even halothane hepatitis is a negligible risk as not a single case of this anesthesia complication has been reported in children. Thus far, they do not seem to be susceptible to it as adults are. Anesthesia morbidity and mortality may have to be considered in some hospitals but should no longer influence general recommendations as to when to do eye muscle surgery.

Summary

The term congenital esotropia should be reserved for those children with an esodeviation who have never had an opportunity for normal binocular vision. In clinical terms, any child with a proven constant esodeviation at four months of age should be considered to have congenital
esotropia. This condition is hereditary but otherwise of unknown etiology. It is typically characterized by a large deviation, cross fixation, and limited abduction bilaterally. There is, in addition, an apparent congenital defect for central fusion, and the best functional result which can be obtained is a monofixation syndrome, microtropia, or microstrabismus. Although less than perfect, such a result is desirable because it is stable and is accompanied by a fair degree of binocular cooperation. Management starts with early and accurate diagnosis and early treatment of amblyopia to establish alternating or cross fixation. To obtain maximal functional results, surgical correction of the deviation should be completed by age two to two and a half. Earlier surgery will not produce better functional results but may be indicated for cosmetic reasons.

Children's Memorial Hospital
2300 Children's Plaza
Chicago, Illinois 60614

References


