Factors Affecting the Outcome of Congenital Esotropia Surgery

by Amy R. Lay

Advisor: Mark Kosciuszko, O.D.

Optometry 799
Special Studies

INTRODUCTION

Congenital esotropia is considered to be the most common type of esotropia, accounting for 28-54% of all esotropia and having an incidence in the general population of about 1% [1]. It is defined as having onset within the first six months of life. However, several authors such as Costenbader [2] have questioned whether the condition referred to as congenital esotropia is truly congenital, that is, existing at birth.

Studies by Helveston [3] and Nixon et al [4] were designed to determine whether esotropia is ever present at birth. Helveston examined 500 infants between 3 hours and 3 days in a newborn nursery to determine alignment, and Nixon examined 1219 newborn infants in a hospital nursery. Helveston's group did not find any cases of esotropia, while Nixon found only three. Thus, many authors believe that true congenital esotropia is rare, and therefore prefer the term "infantile esotropia."

The classic characteristics of congenital esotropia are the following:

1) an inward eye turn documented during the first six months of life
2) a constant large angle deviation (usually 40-50 PD or more) which remains unchanged in the post-natal period
3) a primarily non-accommodative deviation having no more than mild to moderate hyperopic refractive error

Common associated conditions include amblyopia, dissociated vertical deviation, overaction of the inferior obliques, and latent nystagmus. These conditions have variable onset and generally occur in the second or third year of life [5]. In addition, many congenital esotropes demonstrate cross-fixation. They use a turned right eye to view the left field and a turned left eye to view the right field.

Competing theories concerning the etiology and development of congenital esotropia have resulted in significant controversy. Worth's theory stated that esotropia was a "wired-in" defect of the sensory fusion system [6]. Chavasse, on the other hand, rejected this concept and introduced the "motor" theory postulating that most congenital esotropes are capable of developing fusion if the deviation could be fully corrected before the age of two [7]. Still others believe that there are different subgroups of congenital esotropes. Nixon et al [4] hypothesized that there could be two subgroups of infants with esotropia: one with faulty, and one with normal central fusion potential. This theory was offered to explain the rare reports of bifixation and normal stereopsis in post-surgical cases.

Nonetheless, the two original opposing theories have distinct therapeutic implications. If esotropia is caused by an inborn defect in the fusion faculty, no treatment, no matter how early in life, will restore normalcy. If, on the other hand, no such defect exists, a cure could be accomplished by aligning the eyes as early as possible. It was this latter idea put forth by Chavasse that has dominated current therapeutic practices.

SUBJECT AND METHODS

A twenty-two year old white male presented to the Ferris State University Optometry Clinic for a routine examination. Ocular history included two extraocular muscle surgeries at the age of two for congenital esotropia. Previous therapy included patching at the age of five. Medical history included asthma and environmental allergies.

Refractive and binocular examination results are listed below:

		distance	near
Unaided VA:	OD	20/60	
	OS	20/70	
	OU	20/60	
Aided VA:	OD	20/20 +3	20/20
	OS	20/15 -1	20/20
	OU	20/15 -1	20/20
BVA:	OD	+4.25 -2.00 x 180	20/15
	OS	+4.50 -2.25 x 180	20/15 -1
Stereopsis:	200" with correction (Titmus Circles)		
Cover Test: (aided)	6 PD alternating esotropia at 6 meters 4 PD alternating esotropia at 40 cm dissociated vertical deviation (distance and near)		

This particular case of congenital esotropia was closely analyzed at the clinic because of the noteworthy monocular visual acuities of 20/20 or better following early surgical correction. A

literature study was then undertaken to investigate the variables affecting cosmetic and functional success with surgery for congenital esotropia. The primary purpose was to evaluate which factors enhance the prognosis for achieving a functional cure. A brief discussion of the optometrist's role in managing congenital esotropia cases has also been included.

A cure of strabismus may be defined as the following [9]:

1) restoration of single binocular vision in the practical field of gaze (orthotropia or asymptomatic heterophoria)
2) normal visual acuity in each eye
3) normal stereoacuity on random dot testing
4) normal retinal correspondence
5) stable sensory and motor fusion

There is agreement between strabismus surgeons that complete restoration of normal binocular vision with normal random dot stereopsis is unattainable in congenital esotropia [9]. In light of this statement, one may ask:

1) What preoperative treatment can be initiated to assist in attaining the best postoperative results?
2) What are the goals of treatment, if restoration of normal binocular vision is unattainable?
3) What is the nature of the subnormal or abnormal forms of binocular vision that may result?
4) Does the occurrence of these forms of binocular vision reliably relate to certain factors, such as age at surgery, postoperative angle, or surgical technique?

Five general issues must be considered for every patient presenting with congenital esotropia. These include:

1) treatment of any refractive error
2) treatment of amblyopia if present
3) consideration of nonsurgical treatment using lenses, prism, occlusion, and vision therapy
4) referral for surgery
5) postsurgical management

PRESURGICAL EVALUATION

In the evaluation of refractive error, Costenbader has reported that the spread of refractive error in children with congenital esotropia is the same as would be expected in a nonstrabismic group of children of similar ages [2]. Birch and Stager et al [10] emphasize the importance of prescribing spectacles preoperatively for hyperopia of three diopters or more. Christensen, Rouse, and Adkins [5] recommend prescribing the full cycloplegic refraction plus an overcorrection of +1.00 diopter. The overcorrection is suggested due to the strong possibility of a latent hyperopic component and the fact that infants' visual activities are generally confined to near and intermediate distances.

It is widely believed that early onset of strabismus and/or delay in starting treatment have an adverse influence on amblyopia. Von Noorden found that amblyopia or a history of previously treated amblyopia occurred in 35% of 408 patients with congenital esotropia [11]. Other reports state that the prevalence of amblyopia in children with congenital esotropia is between 20% and 50% [12].

Therefore, amblyopia must be treated without delay prior to surgery. For best results, amblyopia prophylaxis must continue until the child is beyond the susceptible period of recurrence of amblyopia, which occurs between the age of eight and ten years [12].

There is a difference in opinion regarding which factors have the greatest association to severe amblyopia after conventional treatment. The three main factors considered are the age of onset of the congenital esotropia, the amount of ametropia, and the average duration of the strabismus before treatment. Ingram and Walker et al [13] studied 161 congenital esotropes and found no evidence to suggest that the age of onset or duration of the strabismus between onset and presentation significantly determined the visual outcome. They found that the only significant predictor of a poor final acuity was abnormal meridional hyperopia. They defined this as 3.5 diopters or more of hyperopia in the most hyperopic meridian of each eye.

Customarily, it is held that cross-fixation prevents the development of amblyopia. However, in 1961 Costenbader [2] stated that despite a high frequency of alternate fixation in 500 cases of congenital esotropia reviewed, developmental amblyopia occurred in approximately 40%. More recently, in 1987 Dickey and Scott [14] reviewed 217 children with congenital esotropia and reported amblyopia in 51%. Approximately half of these patients were cross-fixators (56/110). Dickey and Metz et al [12] state that the diagnosis of amblyopia is made based on the point at which alternation of fixation takes place. By this method, if there is equal visual acuity, alternation will occur at the midline with each eye. If amblyopia

exists, the sound eye will continue to follow the target beyond the midline, into abduction, before the poorer seeing eye picks up fixation. Successful amblyopic treatment occurs for these patients when alteration occurs at the midline following occlusion therapy.

The goals of surgical treatment can be described in terms of a cosmetic cure and a functional cure. Cosmetic cure is generally accepted to be a strabismus which measures ten prism diopters or less of esotropia or exotropia. No functional binocularity need be present. The term "functional cure" is more difficult to pinpoint since normal bifoveal binocular vision has rarely if ever been demonstrated after surgical intervention alone for congenital esotropia [15]. Von Noorden believes that the best outcome possible is subnormal binocular vision, in which the patient is ortho or has an asymptomatic heterophoria, motor fusion, unilateral foveal suppression under binocular conditions, reduced stereoacuity, and normal retinal correspondence [15]. He defines reduced stereoacuity as less than 120 seconds on the Randot stereo test, and he advocates testing retinal correspondence with Bagolini striated lenses. Moreover, von Noorden states that the most likely "functional" outcome is a small angle strabismus with peripheral fusion, gross stereopsis, and a monocular suppression scotoma under binocular conditions.

CLASSIFICATION OF SURGICAL RESULTS

In 1984 von Noorden suggested classifying the results of surgery for congenital esotropia into the following categories:

1) subnormal binocular vision

2) microtropia

3) small angle and cosmetically acceptable residual esotropia or consecutive exotropia

4) large angle residual esotropia or consecutive exotropia that requires additional surgery

Characteristics of each of these are detailed below.

SUBNORMAL BINOCULAR VISION:
orthotropia or asymptomatic heterophoria
normal visual acuity in both eyes
fusional amplitudes
normal retinal correspondence
foveal suppression in one eye in binocular vision
reduced or absent stereopsis
stability of alignment
optimal treatment result

MICROTROPIA:
inconspicuous shift or no shift on cover test
mild amblyopia frequent
fixation central or parafoveolar
fusional amplitudes
anomalous retinal correspondence
reduced or absent stereopsis
some stability of alignment
no further treatment except amblyopia prevention
desirable treatment result

SMALL ANGLE ESOTROPIA/EXOTROPIA:
cosmetically acceptable
80% have anomalous retinal correspondence
less stability of angle
no further treatment except amblyopia prevention
acceptable result

LARGE ANGLE ESOTROPIA/EXOTROPIA (>20 PD):
usually cosmetically unacceptable

less chance for anomalous retinal correspondence,
suppression prevails
unstable angle
unacceptable result

Generally, anomalous retinal correspondence (ARC) occurs after surgical treatment in patients with microtropia and with residual small angle esodeviations and exodeviations. This remains as a permanent residual sensory defect in most of these patients. Von Noorden [9] states that currently ARC is considered a functionally beneficial state of binocularity that should not be disturbed because it is superior to suppression. Functional benefits of ARC include simultaneous binocular perception, fusional vergences, an intact binocular visual field, normal distance judgment, and, in many instances, even gross stereopsis. In addition, treatment of ARC by orthoptic methods may yield intractable diplopia in a formerly asymptomatic patient.

PATIENT AGE AT TIME OF SURGERY

A significant question is at what age should alignment of the eyes be attained to achieve the best functional outcome? Most authors today agree that early surgery is preferable, due to better final motor and sensory results [16]. The motor alignment failure in cases operated on later in life may result from secondary changes in the extraocular muscles, conjunctiva, and Tenon's capsule. On the other hand, some surgeons still prefer to operate at an older age, since a better and more complete examination can be accomplished.

The age when surgery is performed depends on several factors. The most obvious is the age of the patient at the initial presentation to an ophthalmologist. Helveston and Ellis et al [17] state that other factors depend on the experience of the surgeon and include the following:

1) safety factors associated with anesthesia
2) perceived importance of early surgery in producing better binocular vision
3) reliability of diagnosis and measurement in the very young child
4) anatomical maturity of the infant eye

Helveston and Ellis et al support the practice of performing surgery as early as 4 months of age. They reviewed all charts of patients who had bimedial rectus recessions before entering their fifth month during the period January 1986 to December 1988. Eleven patients met the criteria for inclusion in the study. The age at first presentation averaged 3 1/2 months, and the age at which surgery was performed averaged 4 1/3 months. The interval between first examination and surgery averaged 21 days. The preoperative angle ranged from 30 to 90 prism diopters. Postoperatively at the last visit (four to eight weeks after surgery), 9 patients were straight or within ten prism diopters of orthotropia. The remaining two had 20-30 prism diopters residual esotropia requiring additional surgery. Both were straight postoperatively. Moreover, no complications were noted in any of the patients.

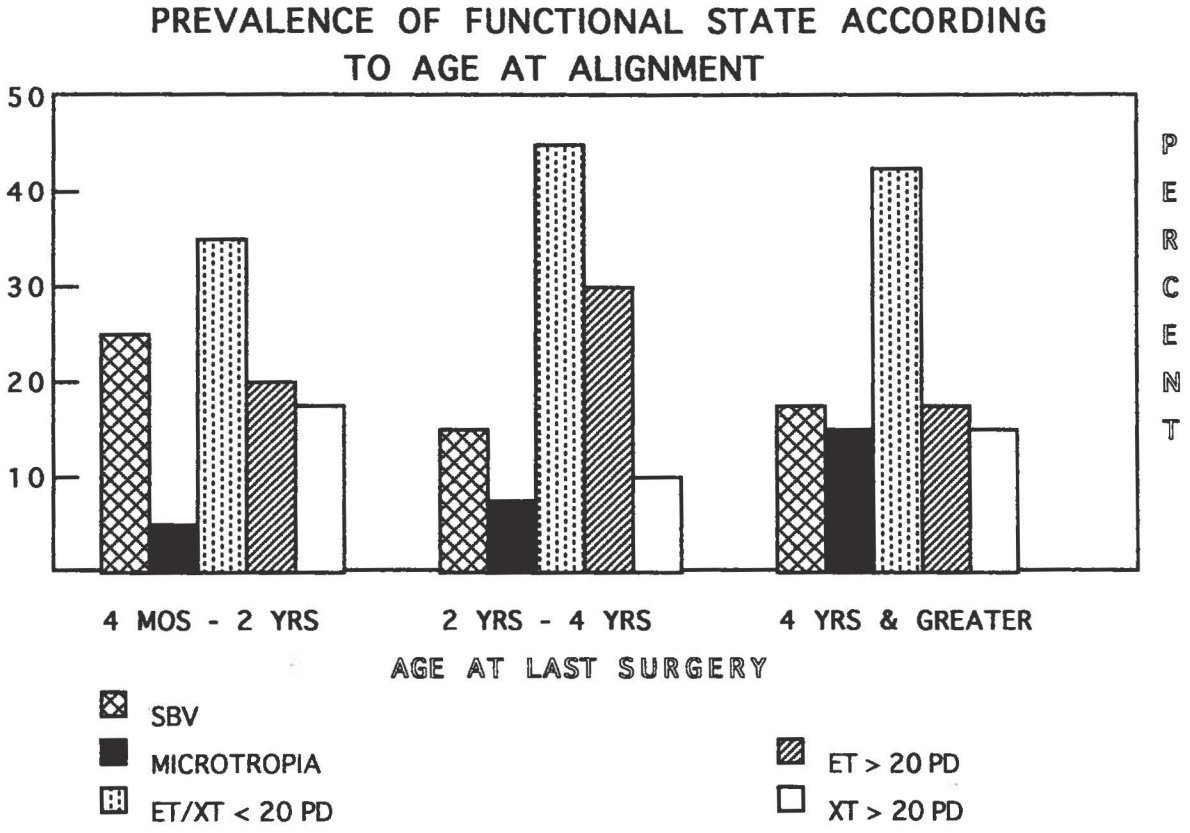
Helveston and Ellis et al believe that 4 months is the earliest surgery should be performed because the 2 to 4 month period is a

state of flux and maturation. Infant studies indicate that between 2 and 4 months of age, infantile strabismus often disappears, normal alignment takes its place, and stereo acuity, visual acuity, and optokinetic symmetry develop normally [17]. That is, strabismus occurring in infancy often gradually disappears with maturation. Esodeviations tend to disappear by two months of age and exodeviations by 6 to 10 months of age [17]. Therefore, those esodeviations still present at four months of age can be reliably confirmed as needing treatment. Helveston and Ellis et al also state that measurements and anatomy appear to be adequate by four months, and anesthesia is safe in experienced hands.

Spierer [16] reviewed fifty patients with congenital esotropia who were randomly selected and operated on at different ages. Patients were divided into two groups and evaluated in regard to their final alignment and sensory results. One group included patients that were operated on and aligned by 36 months of age, and the other group included patients operated on and aligned after the age of 36 months. Spierer found that a similar percentage of patients was aligned in both groups, regardless of whether the operation was performed by 36 months (78.6%) or after 36 months of age (77.7%). However, a higher percentage of the first group aligned by 36 months of age enjoyed binocularity (45.5%) as compared with the second group (28.6%). Binocularity was assessed by the Worth 4-dot test, major amblyoscope, or the Polaroid Titmus 3 D Vectograph test.

Von Noorden [11] examined whether the quality of sensory results depends on the age at which surgical treatment is completed. In a study of 358 surgically treated congenital esotropes, the

prevalence of the various functional end stages of therapy were grouped according to whether therapy was completed between 4 months and 2 years, 2 to 4 years, or older than 4 years. Results shown in the table below demonstrate that as the age of completion of treatment increased, the probability of an optimal outcome (subnormal binocular vision) decreased.



This data is in accordance with the current view that surgery before the age of two years yields superior results [9]. However, it also demonstrates that many patients still achieved a useful form of

binocularity when surgical treatment was concluded after the age of two or even four years.

In a similar review, Scheiman, Ciner, and Gallaway [1] summarized data from seven studies investigating the functional results of surgery related to time of surgery. Data was divided into the following categories: results for restoration of "subnormal binocular vision" when surgery was performed before 12 months, between 13 and 24 months, and after 24 months of age. Surgery before 12 months led to a success rate of 71%; between 13 and 24 months the combined success rate was 43%; and after 24 months of age the success was 12%. They concluded that the most reasonable approach appears to be the concept of performing surgery as soon as an accurate determination can be made of the deviation and associated characteristics, and only after attention has been directed to any accommodative component and treatment of amblyopia.

Teller and Savir et al [18] examined a very important aspect of surgical outcome-- results four years after surgery. They evaluated surgical outcome in 46 children, and considered surgical "success" a postoperative deviation of 10 prism diopters or less in primary position. Periodic evaluations occurred 6 weeks, 6 months, 2 years, and 4 years after surgery. The children were divided into three age groups: less than 2 years, 2 to 3 years, and more than 3 years of age at the time of the operation. After four years of follow-up, 86.7% of the first group (less than 2 years of age at surgery) had deviations of 10 prism diopters or less. In addition, 75% of group two (2-3 years of age at surgery) and 70.9% of group three (more than 3 years of age) had deviations in this range. On the other hand, Teller and

Savir et al point out that only 20% of all of these patients had binocular function as demonstrated by the Worth 4-dot test and Bagolini striated lenses at the four year follow-up visit. Unfortunately, the authors did not give any comparison of the three groups in terms of binocularity, or any further classification of functional outcome.

PREFERRED ALIGNMENT SHORTLY AFTER SURGERY

Another important question regarding surgical results is what is the preferred alignment shortly after surgery? Caputo and Guo et al [19] reviewed 117 patients with congenital esotropia who were followed for a minimum of three years. Only those patients who were orthotropic or were within 10 prism diopters of orthotropia six months postoperatively were included. Their results indicated that 86% remained orthotropic or within 10 prism diopters of orthotropia three years postoperatively, but 11% (13 patients) developed consecutive exotropia greater than ten prism diopters. Five years after surgery, 25% had consecutive exotropia. They concluded that despite satisfactory postoperative alignment, there is a steady progression toward exotropic drift over long-term follow-up. They also suggested that the preferred alignment shortly after surgery is within 10 PD of esotropia since all patients who demonstrated a consecutive exotropia were either orthotropic or had less than ten prism diopters of exotropia six months after surgery. In a similar study, Hiles and colleagues [30] found 25% (eight of 54) of their patients developed consecutive exotropia, and seven had second

surgery even though their deviation had been reduced to within 10 PD of orthotropia soon after esotropia surgery.

Maruo and Kubota et al [20] reviewed 307 records of congenital esotropes with four or more years of postoperative follow-up. They determined preferred post-operative alignment to be 7 PD esodeviation to 2 PD exodeviation. They found that 84.4% of postoperative patients measured in this range one month after surgery, while 70.4% were still in this range four years after surgery. They also investigated the angle of strabismus four years after surgery as it related to simultaneous perception, fusion, and stereopsis. 39.7% obtained simultaneous perception and fusion with the major amblyoscope and 14.8% demonstrated stereopsis on the Titmus stereo test. Those attaining stereopsis were predominantly in the range of 7 PD esodeviation to 2 PD exodeviation, leading the authors to their conclusions about preferred alignment following surgery.

SURGICAL TECHNIQUE

Another factor relating to surgical success is the introduction of newer surgical approaches. The more traditional surgical approach has been referred to as uniform, or conventional two muscle surgery. In this approach a bilateral medial rectus recession of approximately 5 mm, or a 5 mm medial rectus recession combined with an 8 mm or 9 mm lateral rectus resection, is performed on all patients regardless of the initial deviation [1]. A variation of this procedure called graded recession has become more popular to decrease the need for multiple surgeries. In this approach, a graded recession that exceeds

the traditional 5 mm is used. The specific amount of recession chosen for any patient is based upon the size of the strabismus. Bimedial recessions as large as 6 to7 mm are performed.

The final surgical approach is selective surgery. Surgery is performed with the objective of eliminating all of the horizontal and vertical deviation in one operation. Advocates of selective surgery propose operating on two, three, or four muscles depending on the preoperative angle.

Scheiman, Ciner, and Gallaway [1] compiled the results of fifteen studies that clearly stated the surgical approach utilized to evaluate whether the newer surgical approaches are any more effective than the traditional uniform procedure. The data for the traditional two muscle, uniform procedure consisted of 343 patients and yielded a combined cosmetic cure rate of 53% (range from 33-84%). The graded recession group consisted of 358 patients and the combined cosmetic success rate was 83% (range from 73-91%). Finally, the selected surgery data included 173 patients and the combined cosmetic cure rate was 69% (range from 61-79%).

Weakley and Stager et al [21] advocate the technique of graded recession. They reviewed the surgical records of thirty-six patients with large angle congenital esotropia averaging 74 prism diopters. All patients underwent 7 mm bilateral medial rectus recessions. Successful horizontal alignment at the most recent follow-up examination (average 18.2 months postoperatively) was achieved in 75%. 14% were undercorrected and 11% were overcorrected. In addition, they state that the advantages of this method, as opposed to three or four muscle procedures, are that it is quicker, simpler, and a

less traumatic procedure which leaves the lateral rectus muscles unoperated for future surgeries if necessary.

Some surgeons have avoided large recessions for treating large angle congenital esotropia because of concerns of adduction deficiency, convergence weakness, or consecutive exotropia with prolonged follow-up [21]. On the other hand, there are high rates of undercorrection in large angle congenital esotropia with traditional bimedial rectus recessions. As a result, some surgeons prefer three or four muscle procedures as initial treatment in these cases. However, more recent studies have suggested that results comparable to three or four muscle procedures can be obtained in large angle esotropia with 6 mm and 7 mm bimedial rectus recessions. In addition, Weakly and Parks [22] found that postoperative adduction and convergence deficits have not been associated with recessions up to and including 7mm. Szymd et al [23] reported a 91% success rate at six weeks using 6 mm and 7 mm recessions in 45 congenital esotropes with deviations exceeding 50 prism diopters. Moreover, in a follow-up study Nelson et al [24] reported a 83.5% success rate in 97 congenital esotropes with deviations greater than 50 prism diopters undergoing 6 mm and 7 mm recessions with a mean follow-up of two years.

COSMETIC AND FUNCTIONAL SUCCESS RATES

Despite newer surgical techniques, multiple surgeries for congenital esotropia are often needed to achieve acceptable cosmetic results. Ing reported that 45% of his sample of 106 congenital esotropes required two or more operations [31]. Hiles found in his

study of 54 patients that an average of 1.8 operations were needed to achieve a cosmetic cure [30].

While many authors provide cosmetic success rates, the functional results are even more important. Scheiman, Ciner, and Gallaway [1] reviewed seventeen studies of congenital esotropia to determine both cosmetic and fuctional cure rates. 1185 patients were included in the study of cosmetic success, and a cosmetic cure rate of 64% resulted with a range from 29-91%. Furthermore, ten studies met specific criteria about subnormal binocular vision cure rates. A cure rate of 22% resulted, with a range from 4-66%. Similarly, Willshaw and Keenan [25] found that for 26 children aligned by 18 to 24 months, 31% achieved stable subnormal binocular vision. Interestingly, they also found that the presence of dissociated vertical deviation before the age of two years to be incompatible with the development of worthwhile subnormal binocular vision.

Von Noorden [9] classified functional results in his study of 358 patients. He found that 20% (71 patients) achieved subnormal binocular vision, and only three patients in this group had stereopsis on the Titmus test (one with 200 seconds of stereopsis and two with 3000 seconds of arc). Furthermore, none had stereopsis when tested with random dots. Seven percent were microtropic and 39% had a residual, cosmetically insignificant esotropia or exotropia. A residual esotropia was three times more common than a residual exotropia. An esotropia of more than 20 PD remained in 20%, and an exotropia of more than 20 PD remained in 15%. In summary, some form of binocular cooperation, ranging from near normal (subnormal

binocular vision) to anomalous (microtropia and small angle esotropia or exotropia), occurred in 66% of patients.

Van Selm [26] investigated a crucial aspect of functional success by reviewing 42 cases of congenital esotropia 20 years after surgery. Of these, 23 had no recognizable stereopsis, 12 had 100-800 seconds of arc, and 7 had 80 seconds of arc or better. Of the 23 cases with no recognizable stereopsis, 12 had no stereopsis after the initial 3 to 4 year review period, but nervertheless remained cosmetically acceptable. Of the remaining eleven, five had a subsequent surgery for diplopia, all exhibiting a residual cyclotropic deviation. Four of the other six developed monocular amblyopia with consecutive divergence. Of the 19 cases with stereopsis, all but four maintained their initial degree of stereopsis, the majority meeting the conditions for monofixation syndrome. These conditions typically include a small angle strabismus with mild manifest amblyopia, peripheral normal retinal correspondence, peripheral fusion, and a suppression scotoma including the macular field of the non-fixating eye [5]. The remaining four, who initially achieved 80 seconds of arc, remarkably improved to 40 seconds of arc [26].

Van Selm's [26] thorough investigation pointed to two main factors limiting the outcome of useful stereopsis: parental/patient cooperation and surgical incompetence. He stated that parental non-cooperation resulted in delayed investigation after the onset of strabismus, poor compliance with the prescribed pre- and postoperative treatment plan, or refusal of further surgery when, in the opinion of the parent, cosmesis was satisfactory. Surgical incompetence was due to failure to recognize and correct vertical

deviations early and to an over-enthusiastic initial amount of muscle adjustment with little regard to anatomical muscle insertions. All of the children who developed useful stereopsis and who maintained their initial stereopsis into adulthood enjoyed parental cooperation and improved surgical competence. These children returned for re-examination regularly, at six month intervals, until the age of at least 16 years.

Hiles [30] also completed a comprehensive study of the long term stability of surgical correction of congenital esotropia. All 54 patients studied had surgery before the age of one and were followed postoperatively for a minimum of ten years. Hiles found that he could divide the patients into three groups: 39% retained stable ocular alignment of +/- 10 prism diopters throughout the postoperative observation period; 31% demonstrated a deterioration of the strabismic angle after early initial alignment; and 30% showed variable or unstable ocular alignment throughout the entire period of follow-up care. Additional surgery for esotropia, consecutive exotropia, or overacting inferior obliques was necessary for all of the patients in the second category. Of this group, more than half deteriorated after the age of four.

SEQUELAE OF SURGICAL TREATMENT

The studies of long term results demonstrate that early successful alignment does not guarantee long-term stability. The need for repeat examinations throughout the first decade of life is based upon the fact that there are a number of associated conditions and sequelae which tend to result in instability after surgery. These

factors include over and under corrections, accommodative esotropia following congenital esotropia, dissociated vertical deviation, inferior oblique muscle overaction, amblyopia, and nystagmus.

After initial surgical correction of congenital esotropia, an accommodative component may become manifest with time. Nirenberg [27] reported that 87% of the patients who re-developed esotropia did so because of an accommodative component. The rate of correction of the subsequent esotropia with glasses alone was 73%. In Hiles' ten year follow-up of 54 congenital esotropes, he found that glasses were necessary in 65% of the children to maintain cosmetic alignment [30].

Baker and De-Young Smith [28] undertook a study to look specifically at the development of accommodative esotropia following the surgical correction of congenital esotropia. The criterion of a minimum of three years postsurgical follow-up was met by 101 patients. Of these patients, 52 developed accommodative esotropia, 25 within 3 months of surgery and 27 from 3 to 60 months after surgery. The average preoperative refractive error in the early group was +3.90 and +1.95 in the later group. The authors concluded that if the preoperative refractive error is +3.00 or more, and especially if the preoperative deviation responds to spectacles with a reduction of fifteen prism diopters or more, then it is very likely that glasses will be required for accommodative esotropia very soon following surgery. They stated that there appear to be no clues for detecting the group that will develop accommodative esotropia at some time in the future.

Other factors resulting in instability after the initial surgical procedure are the presence of inferior oblique overaction and dissociated vertical deviations. They may not be present at the time the first operation occurs, but develop or become manifest afterwards. Hiles [30] reported that 78% of the sample of infantile esotropes that he followed had an overaction of one or both inferior obliques. The most frequent age of onset was the second year of life for both inferior oblique overaction and dissociated vertical deviations. Hiles also found that dissociated vertical deviations are the single most common defect associated with the need for further surgery. Therefore, the presence of this condition in particular should be regarded as a factor limiting the prognosis for a functional cure.

Maruo and Kubota et al [20] examined results four years after surgery by dividing the cases as follows: 385 congenital esotropia cases not combined with vertical deviation, 109 cases combined with dissociated vertical deviation, and 78 cases combined with overaction of the inferior oblique muscles. Adequate correction for the angle of strabismus was achieved for 81.6% of those not combined with vertical deviation, 61.5% of those with dissociated vertical deviation, and 70.5% of those with overaction of the inferior oblique muscles. Those cases in which simultaneous perception and fusion were proved were 62.4% of those with no vertical deviation, and 49.2% of those with overaction of the inferior oblique muscles. They concluded that the prognosis for binocular vision after treatment of congenital esotropia and dissociated vertical deviation is not good, and yields unsatisfactory results.

The development of amblyopia postsurgically is an area of concern especially because some studies indicate that amblyopia may be more likely after surgery than before surgery. Hoyt et al [29] used visually evoked potentials to assess the visual acuity of 42 congenital esotropes before and after surgery. Thirty-one infants with congenital esotropia did not have nystagmus, while eleven patients demonstrated abducting nystagmus. While only four of the infants in the first group were amblyopic before surgery, 23 infants developed amblyopia during the follow-up period after the first surgical procedure. In contrast, all 11 patients with esotropia and abducting nystagmus were amblyopic at some time during the study. Nine of the 11 were amblyopic before surgery, and the other two developed amblyopia in the first three postoperative months. Hoyt et al concluded that amblyopia occurs infrequently in patients with congenital esotropia and no nystagmus until surgery reduces the angle of deviation and breaks down the cross-fixation reflex. In its place, the establishment of a monocular fixation preference with resulting amblyopia of the nonfixating eye frequently occurs. Similarly, Scheiman, Ciner, and Gallaway [1] found in their literature search that most authors believe the postsurgical increase in amblyopia is due to the change from a large angle, alternating esotropia to a small angle, unilateral esotropia.

Forms of nystagmus associated with congenital esotropia include latent nystagmus, rotary nystagmus, end-point nystagmus, and horizontal pendular nystagmus. Hiles et al [30] found rotary nystagmus to be the most common form of nystagmus in their study of 54 congenital esotropes. The onset of rotary nystagmus occurred

with greatest frequency during the first year of life, and its presence diminished over the next several years. Interestingly, the authors speculated that rotary nystagmus may be a precursor to the development of dissociated vertical deviation. While the occurrence of rotary nystagmus diminishes progressively over the first decade of life, dissociated vertical deviation increases in occurrence over the same time period.

SUMMARY AND CONCLUSIONS

Restoration of some level of binocular vision is the most desirable outcome of congenital esotropia surgery. This study of which factors enhance the likelihood of achieving this goal shows that certain peoperative treatment regimens can assist the surgical outcome. For example, Birch and Stager et al pointed out the importance of prescribing spectacles preoperatively for hyperopia of three diopters or more. In addition, various reports list the prevalence of amblyopia between 20% and 50%. Amblyopia must be treated without delay prior to surgery, and post-surgical follow-up care needs to be continued until the mid-teens to monitor for its development or recurrence. Special care must be taken to evaluate for amblyopia even in cross-fixators, since cross-fixation does not necessarily prevent the development of amblyopia.

This review of the literature also sought to define the goals of treatment, since restoration of normal binocular vision is considered to be unattainable. It is important to note that many authors

reported only cosmetic success rates, and failed to relate these results to functional outcome. Cosmetic success is generally accepted to be a strabismus measuring ten prism diopters or less. Overall results from the reports indicate that approximately two-thirds of patients achieve a cosmetic cure after one surgical procedure. Many authors, such as Von Noorden, believe that the best possible functional outcome is subnormal binocular vision. Restoration of subnormal binocular vision generally occurs in about one out of five cases. Some form of binocular cooperation, ranging from near normal (subnormal binocular vision) to anomalous, occurs in about two-thirds of patients. This review found that the most likely outcome is a small angle esotropia. Anomalous retinal correspondence typically occurs in patients with microtropia and residual small angle esodeviations and exodeviations. In most cases, this state should not be disturbed because of its superiority to suppression, and treatment may yield diplopia.

Certain surgical factors do reliably relate to improved functional prognosis. Investigations into what age surgical alignment of the eyes should occur revealed that four months of age is the earliest surgery should be performed. Prior to this age, ocular alignment is still in a state of flux and maturation. Spierer's study found that similar percentages of patients were cosmetically aligned regardless of whether surgery occurred before or after 36 months of age. However, a much higher percentage of the group aligned by 36 months achieved binocularity. Von Noorden reported similar results in his study that looked more in depth at categorizing functional results according to age at alignment. He found that as the age of

completion of treatment increased, the probability of achieving subnormal binocular vision decreased. The trend by most studies indicates better functional results when surgery is performed before two years of age.

An investigation into the preferred alignment shortly after surgery revealed similar results between the studies reviewed. Caputo and Guo defined the preferred alignment to be within 10 PD of esotropia, while Maruo and Kubota determined it to be 7 PD esodeviation to 2 PD exodeviation. The rationale for these findings was that there is a steady progression toward exotropic drift over long-term follow-up, and patients in this range achieve improved stereopsis.

An encouraging finding was that newer surgical approaches appear to result in considerably better success rates. In particular, graded recession appears to be superior to the more traditional uniform, bimedial recession. In addition, the advantages of the graded method, as opposed to three or four muscle procedures, are that is a quicker, simpler, and less traumatic procedure which leaves the lateral rectus muscles unoperated for future surgeries if needed. Cosmetic success rates of 83% have been reported with the graded procedure. This information suggests that it may be important for optometrists to select a surgeon based upon a knowledge of the surgical approach being used. Unfortunately, none of the articles reviewed compared functional success rates between the traditional method and the newer procedures.

Another advantage of graded recession is the use of large recessions for treating large angle congenital esotropia. There are

high rates of undercorrection in large angle congenital esotropia with the traditional, uniform procedure. Bilateral medial rectus recessions of 6mm or 7mm are now being utilized for deviations exceeding 50 prism diopters, with no evidence of significant adduction or convergence deficits. Thus, the graded technique appears to be the superior method for enhancing the prognosis of successful alignment. On the other hand, almost half of all congenital esotropes still require two or more operations to achieve cosmetic success.

It appears that the achievement of greater than 80 seconds of arc stereopsis in cases of congenital esotropia is possible provided that certain criteria are fulfilled. The delay between onset of esotropia and the examination should be as short as possible. Full parental cooperation, especially in the immediate postoperative period, is essential. In addition, the surgeon's understanding of the special anatomical relationships of the extraocular muscles and their application to surgery in infants is critical. Moreover, Van Selm pointed out that the initial binocularity can deteriorate or be lost with the passage of time , despite initial successful attainment of motor parallelism, if regular examination until the mid-teens is neglected. Hiles' study of long term stability also concluded that long-term follow-up care is essential. One third of patients in his study demonstrated deterioration of the strabismic angle after initial alignment. Additional surgery was necessary for all of these patients, and more than half of this group deteriorated after the age of four.

The need for careful follow-up throughout the first decade of life is also based on the fact that sequelae such as accommodative

esotropia, dissociated vertical deviation, inferior oblique muscle overaction, and amblyopia may become manifest with time and result in instability. Patients who redevelop esotropia may do so because of an accommodative component, which in many cases may be corrected with glasses alone. Baker and De-Young Smith found that if the preoperative refractve error is +3.00 or more, and especially if the deviation responds to this correction with a reduction of 15 PD or more, it is very likely that glasses may be required for accommodative esotropia following surgery. Therefore, it is very important for both the surgeon and the optometrist to be aware of the possibility of an accommodative component, and to treat it accordingly with spectacles if needed.

Detecting dissociated vertical deviation is especially critical because authors such as Hiles have found that it is the most common defect associated with the need for further surgery. Results by Maruo and Kubota et al showed that adequate alignment occurred in approximately 80% of congenital esotropes not combined with vertical deviation, 61% with vertical deviation, and 70% with inferior oblique overaction. Moreover, prognosis for binocular vision in patients with dissociated vertical deviation is lower than that in the other two groups listed above. Unfortunately, only minimal speculation was found in the literature as to why this condition often becomes manifest with time. It is also interesting to note that the twenty-two year old patient examined in the Ferris State Primary Care Clinic demonstrated dissociated vertical deviation, but yet had acceptable cosmetic alignment, no evidence of amblyopia, and 200 seconds of stereopsis.

Amblyopia prevention after surgery is also an area of concern. Some studies believe that amblyopia may be more likely after surgery than before, due to the change from a large angle, alternating esotropia to a small angle, unilateral esotropia. Amblyopia prevention should continue until the patient is beyond the susceptible period for recurrence.

Several important conclusions from these results can be applied to the optometric management of congenital esotropia. After examining a patient and reaching a diagnosis of congenital esotropia, the task for every optometrist is to develop a sound management plan. This includes treatment of any refractive error, treatment of amblyopia if present, consideration of nonsurgical treatment if applicable, and referral for surgery. Appropriate surgical referral includes discussion about risks, success rates, complications , and need for postsurgical care. Follow-up optometric care is critical to manage any developing accommodative component, to treat amblyopia, and to prescribe vision therapy as indicated. Moreover, it is important to accurately assess and evaluate the need for additional intervention for inferior oblique and dissociated vertical deviations. Moreover, only through regular follow-up examinations into the mid-teens can the results of congenital esotropia surgery be further studied to evaluate factors favoring long term cosmetic and functional success.

As a final note, it would be interesting for an follow-up study to be performed in the future to reevaluate the patient seen at Ferris State University who sparked the initial interest in performing this literature review. It would be of great value to review his pre-

surgical, surgical, and postsurgical data and to compare it to the findings in this study. A long-term follow-up evaluation of stability of the angle, type of correspondence used, stability of stereopsis, and parental/patient cooperation would provide useful insight into how this patient has achieved 20/20 or better visual acuities, some stereopsis, and cosmetic success despite having conditions such as dissociated vertical deviation and a highly hyperopic refractive error.

References

1. Scheiman M, Ciner E, Gallaway M. Surgical success rates in infantile esotropia. J Am Optom Assoc 1989; 60: 22-31.

2. Costenbader FD. Infantile esotropia. Trans Am Ophthalmol Soc 1961; 59: 397-429.

3. Helveston EM. Origins of congenital esotropia. Am Orthopt J 1986; 36: 40-48.

4. Nixon RB, Helveston EM, Miller K, et al. Incidence of strabismus in neonates. Am J Ophthalmol 1985; 100: 798-801.

5. Christenson GN, Rouse MW, Adkins, DA. Management of infantile-onset esotropia. J Am Optom Assoc 1990; 61: 559-572.

6. Worth C. Squint: Its causes, pathology, and treatment. 5th ed. Philadelphia: P. Blakiston Sons and Co., 1921.

7. Chavasse FB. Worth's squint or the binocular reflexes and the treatment of strabismus. 7th ed. Philadelphia: P. Blakiston Sons and Co., 1939.

8. Nixon RB, Helveston EM, Miller K, et al. Development of normal ocular alignment. J Pediatr Ophthalmol Strabismus 1988; 25: 210-211.

9. von Noorden GK. A reassessment of infantile esotropia. Am J Ophthalmol 1988; 105: 1-10.

10. Birch EE, Stager DR, Berry P, et al. Prospective assessment of acuity and stereopsis in amblyopic infantile esotropes following early surgery. Invest Ophthalmol Vis Sci 1990; 31: 758-765.

11. von Noorden GK. Current concepts of infantile esotropia. Eye 1988; 2: 343-357.

12. Dickey CF, Metz HS, Stewart SA, et al. The diagnosis of amblyopia in cross-fixation. J Pediatr Ophthalmol Strabismus 1991; 28: 171-175.

13. Ingram RM, Walker C, Billingham B, et al. Factors relating to visual acuity in children who have been treated for convergent squint. Br J Ophthalmol 1990; 74: 82-83.

14. Dickey CF, Scott WE. Amblyopia- the prevalence in congenital esotropia versus partially accommodative esotropia. In: Lenk-Schafer (ed). Orthoptic Horizons- Transactions of the Sixth International Orthoptic Conference. Harrogate, Great Britain; 1987: 106.

15. von Noorden GK. Infantile esotropia: a continuing riddle. Am Orthopt J 1984; 34: 52-62.

16. Spierer A. Binocular function after surgical alignment of infantile esotropia. Metab Pediatr Syst Ophthalmol 1988; 11: 35-36.

17. Helveston EM, Ellis FD, Plager DA, et al. Early surgery for essential infantile esotropia. J Pediatr Ophthalmol Strabismus 1990; 27: 115-119.

18. Teller J, Savir H, Yelin N, et al. Late results of surgery for congenital esotropia. Metab Pediatr Syst Ophthalmol 1988; 11: 115-118.

19. Caputo AR, Guo S, Wagner RS, et al. Preferred postoperative alignment after congenital esotropia surgery. Ann Ophthalmol 1990; 22: 269-272.

20. Maruo T, Kubota N, Iwashige H, et al. Long-term results after strabismus surgery. Graefe's Arch Clin Exp Ophthalmol 1988; 226: 414-417.

21. Weakley DR, Stager DR, Everett ME. Seven-millimeter bilateral medial rectus recessions in infantile esotropia. J Pediatr Ophthalmol Strabismus 1991; 28: 113-115.

22. Weakley DR, Parks MM. Results from 7-mm bilateral recessions of the medial rectus muscles for congenital esotropia. Ophthalmic Surg 1990; 21:827-830.

23. Szymd S, Nelson LB, Calhoun JH, Spratt C. Large bimedial rectus recessions in congenital esotropia. Br J Ophthalmol 1985; 69: 271-274.

24. Nelson LB, Calhoun JH, Simon JW, et al. Surgical management of large angle congenital esotropia. Br J Ophthalmol 1987; 71: 380-383.

25. Willshaw HE, Keenan J. Strabismus surgery in children: the prospects for single binocular vision. Eye 1991; 5: 338-343.

26. van Selm JL. Primary infantile-onset esotropia- 20 years later. Graefe's Arch Clin Exp Ophthalmol 1988; 226: 122-125.

27. Nirenberg N, Mazow ML. The frequency and correction of accommodative esotropia following surgical correction of congenital esotropia. Binocular Vis 1985; 1: 71-76.

28. Baker JD, DeYoung-Smith M. Accommodative esotropia following surgical correction of congenital esotropia, frequency and characteristics. Graefe's Arch Clin Exp Ophthalmol 1988; 226: 175-177.

29. Hoyt CS, Jastrzebski GB, Marg E. Amblyopia and congenital esotropia: visually evoked potential measurements. Arch Ophthalmol 1984; 102: 58-61.

30. Hiles DA, Watson BA, Biglan AW. Characteristics of infantile esotropia following early bimedial rectus recession. Arch Ophthalmol 1980; 98: 697-703.

31. Ing MR. Early surgical alignment for congenital esotropia. Trans Am Ophthalmol Soc 1981; 79: 625-663.