BOWMAN LECTURE

Current Concepts of Infantile Esotropia

G. K. VON NOORDEN

Houston, Texas

Summary

Several forms of esotropia with a different pathophysiology that meet the criterion of an onset early in life must be distinguished from essential infantile esotropia. A hypothesis is presented, according to which a delayed development or a congenital defect of retinal disparity sensitivity (motor fusion) in an otherwise normal infant with immature sensory functions causes esotropia under the influence of various strabismogenic factors. Some of these factors are genetically determined, hence the familial occurrence of essential infantile esotropia. The absence or marked decrease of stereopsis and the asymmetry of optokinetic nystagmus are interpreted as the consequence of ocular misalignment early in life rather than of structural anomalies in the afferent visual pathways of esotropic patients. The therapeutic results after surgery are classified into four groups: subnormal binocular vision, microtropia, small angle eso- or exotropia and large angle residual or consecutive eso- or exodeviations. Analysis of data from 358 operated patients with a documented onset of esotropia prior to the sixth month of life has shown that the probability of obtaining an optimal functional result is increased when surgical alignment is completed before completion of the second year of life. However, surgery after the age of two or even four years of life does not preclude the development of binocular vision on a subnormal or anomalous basis.

William Eddowes Bowman was born in 1816 in Nantwich, Cheshire. After schooling at Hazelwood college in Birmingham he was appointed, at the age of 17, apprentice to Joseph Hodgson, a well known surgeon at the General Hospital in Birmingham. As Hodgson was also the founder of the Birmingham Eye Infirmary we may assume that the young Bowman received an early exposure to the art of ophthalmic surgery. After five years of apprenticeship he joined Kings College in London as an anatomic demonstrator

under Robert Hentley Todd. Thus began a 5 year period during which Bowman, still in his early twenties, made an extraordinary series of anatomic discoveries while working with Todd on two monumental volumes, the *Physiological Anatomy and Physiology of Man* and the *Cyclopaedia of Anatomy and Physiology*. During this time occurred the description of the anterior limiting membrane which has immortalised his name. He made major contributions to the histology of striated muscle and of the inner ear, of the

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Correspondence to: G. K. von Noorden, M. D., Ophthalmology Service, Texas Children's Hospital, Box 20269, Houston, Texas 77225, USA

ciliary muscle and the musculature of the iris. Indeed, as Sir Arthur Keith put it in his 1930 Bowman Lecture: 'in the first President of this Society England had one of the greatest anatomists she had ever produced'¹. In 1884, at the age of 30, Bowman began his career as an ophthalmologist by joining the staff of Kings College Hospital in London. During his lifetime he advanced our science substantially, notably the surgery of the lacrimal system and of the anterior segment. He was a genius of great versatility and the praise of his work has been sung so many times and so competently that I shall not attempt to do it again. However, in reviewing his work I was astonished to learn that Bowman performed brow suspension of the upper lid nearly 100 years before the Friedenwald-Guyton suture and determined the angle of strabismus quantitatively long before this became a routine clinical procedure. In 1851 William Bowman, already famous and well established at Moorfields received the visit of two young colleagues from the Continent, Albrecht von Graefe and Frans Cornelius Donders. They were given a warm and hospitable welcome, staying as guests in Bowman's house. Thus began a close friendship that united the three greatest ophthalmologists of the 19th century for a lifetime. After a long, happy and fulfilled life Bowman died at the age of 76. As Frank Law commented in the history of this society, 'he was a whole man in every sense, thorough and painstaking, honest and fair, modest and abstemious, punctual and business-like. He was everybody's friend and a unique figure in many fields'². For Donders' obituary Bowman wrote: 'he had the fortune to have contributed considerably to the advancement of his specialty and lived long enough to see the fruits of his work universally recognised'³. Indeed, what he said about his great Dutch friend could well have been written about Bowman himself.

I have chosen the topic of infantile esotropia for three reasons. First, and foremost, because Brian Harcourt had suggested that I speak about this subject which was of special fascination to him while, at the same time, holding the interest of general ophthalmologists. Second, because much of the seri-

ous thinking about this entity began in this country and third, because there exists uncertainty in the ophthalmological community with regard to the aetiology, what may be expected from treatment and when to operate on children with this disorder.

Definition

Infantile esotropia may be defined as a constant esotropia with onset occurring during the first 6 months of life. Another term, often used synonymously is congenital esotropia which carries the implication that the esotropia is present at birth. In the light of present knowledge it is highly relevant whether a child is born with crossed eyes or whether the eyes cross after a period of initial alignment. Animal experiments have taught us, during the past 20 years, about the extraordinary sensitivity of the infantile visual system and the behavioural, physiological and structural consequences of abnormal visual stimulation during visual immaturity. It is reasonable to assume from these data that an infant whose eyes are crossed from birth has little chance of developing binocularity. On the other hand, an esotropia preceded by a brief period of normal eye alignment may cause less damage to a patient's ability to recover at least some binocular functions. Several authors have expressed doubt that the esotropia actually occurs at birth.^{4,p.87;5,6} However, it was not until Nixon and co-workers from Indiana University⁷ and Friedrich and de Decker from the University of Kiel⁸ published their observations in a total of 2200 physically normal and alert newborns that such suspicions were confirmed and became facts. These authors reported that while strabismus, usually intermittent exotropia but occasionally also a transient esotropia, was found in about a third of all newborn infants during the first few days of life the eyes of these infants had aligned themselves over an interval of three months. When re-examining the children in the series from Indiana at the age of six months only three who had been exotropic or orthotropic at birth had developed a clinical picture consistent with infantile esotropia.⁹ Thus, true congenital esotropia, if it exists at all, must be exceedingly rare and it is for this reason that we prefer the term 'infantile' to

describe this condition, and add the prefix 'essential' to distinguish it from other forms of esotropia that occur in infancy. However, we must not lose sight of the probability that congenital and hereditary factors contribute to the development of infantile esotropia.

Clinical Characteristics

Although certain variations are common the clinical findings in essential infantile esotropia are fairly consistent and are summarised in Table I. This Table is a modification of a consensus of opinions from European and North American strabismologists during an informal gathering in Modena, Italy in 1984 and to which I have added asymmetry of the optokinetic response and latent or manifest nystagmus.

Differential Diagnosis

When studying essential infantile esotropia it becomes mandatory to exclude similar conditions in which because of different

pathophysiology the clinical management and the treatment results cannot be expected to be the same. Next to obvious congenital esotropias such as the retraction syndrome of Stilling, Türk and Duane or Moebius syndrome there are other forms of infantile esotropia which must be distinguished from essential infantile esotropia. These conditions are listed in Table II and will be discussed here only as they pertain to the differential diagnosis of essential infantile esotropia.

Nystagmus dampening by convergence (nystagmus blocking or compensation syndrome)

The relationship between congenital nystagmus and infantile esotropia and a possible aetiological association between these two conditions has been mentioned often in recent years. However, the literature is often confusing because of failure to differentiate clearly between latent and manifest congenital nystagmus. Why is this distinction so important? Because the roles of manifest and of latent nystagmus in infantile esotropia are entirely different: in the case of manifest nystagmus the infantile esotropia may be a consequence of the nystagmus, in latent nystagmus both conditions though frequently occurring in the same patient have no apparent aetiological link.

Manifest nystagmus is of equal intensity regardless whether both eyes are open or either eye is covered (Fig 1). The nystagmus is called latent when it can be provoked only by covering either eye (Fig 2). Sorsby¹⁰ was first to note, however, that even in latent nystagmus the eyes are rarely entirely at rest under binocular viewing conditions. In other words, latent nystagmus often has a manifest phase and true latent nystagmus occurs infrequently. Dell'Osso and co-workers,¹¹ therefore, substituted the term latent nystagmus with 'manifest-latent' nystagmus, a term which in spite of its oxymoronic connotation has become firmly established in the nystagmus literature. Manifest and manifest-latent congenital nystagmus have uniquely distinctive waveform characteristics which suggest a different aetiology for each nystagmus type.¹¹ Since manifest-latent nystagmus occurs frequently and manifest nystagmus infrequently with infantile esotropia it is highly probable

Table I *Characteristics of essential infantile esotropia*

Onset birth – 6 months
Large angle (>30 prism dioptres)
Stable angle
Normal central nervous system
May be associated with
–asymmetric optokinetic nystagmus
–defective abduction
–excessive abduction
–dysfunction of oblique muscles
–dissociated vertical deviation (DVD)
–latent or manifest-latent nystagmus
Initial alternation with crossed fixation
Potential for normal binocular vision limited

Table II *Esotropia with onset during infancy*

Essential infantile esotropia
Nystagmus blocking syndrome
Abducens palsy
Refractive accommodative esotropia
Infantile esotropia with central nervous system disorders
Sensory esotropia
Duane's syndrome type I
Moebius syndrome

that many esotropes who in the past were classified as having manifest nystagmus actually had the manifest-latent form.¹²

The possibility that congenital nystagmus may cause an esotropia was first suggested by Adelstein and Cüppers¹³ who coined the term nystagmus blockage syndrome. These authors proposed that the esotropia occurs from hypertonicity of the medial recti, caused by sustained adduction. Adduction of both eyes is provoked by a convergence impulse, the purpose of which is to decrease (or to 'block') the nystagmus and thus to increase visual acuity. This blockage syndrome is characterised by manifest congeni-

Fig. 1. Binocular electronystagmogram of manifest nystagmus. Note mostly pendular wave form and no significant change in intensity upon covering either eye. From von Noorden, Munoz and Wong¹².

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tal nystagmus, an onset of esotropia within the first few months of life, a variable angle, pseudoabducens paralysis and an inverse relationship between the angle of strabismus and the intensity of the nystagmus (Fig 3). Because of the rather unique aetiology of the esotropia in these cases, nystagmus blocking syndrome must be considered an entity different from essential infantile esotropia.

Fig. 2. Binocular electronystagmogram of manifest-latent nystagmus. Note minimal nystagmus with both eyes open and marked increase of jerk nystagmus upon covering either eye. The nystagmus with both eyes open was not seen on clinical examination in this patient. From von Noorden, Munoz, and Wong¹².

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Fig. 3. Binocular nystagmogram showing dampening of manifest nystagmus by convergence with improvement of visual acuity during esotropic phase. From von Noorden, Munoz and Wong¹².

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Unfortunately, Adelstein and Cüppers did not distinguish clearly between manifest and latent congenital nystagmus. Their nystagmus blockage syndrome has become confused with a much more common condition described by Ciancia¹⁴ and consisting of infantile esotropia associated with manifest-latent nystagmus. In this syndrome the relationship between the esotropia and the nystagmus is, as far as we know, purely coincidental. Such patients may present with a head turn toward the side of the fixating, adducted eye and have a jerky nystagmus with a neutral point in adduction and increasing amplitude as the eye abducts (Fig. 4). Whether esotropia associated with manifest-latent nystagmus as described by Ciancia represents a nosologically viable subgroup in the essential infantile esotropia syndrome remains an intriguing subject for future research.

Abducens paralysis

A unilateral or bilateral abducens paralysis in

Fig. 4. Monocular electronystagmogram of manifest-latent nystagmus in different gaze positions. Note increase of jerk nystagmus in abduction with fast phase beating toward side of fixating eye and improvement of visual acuity when each eye fixates in adduction. From von Noorden, Munoz, and Wong¹².

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infancy is in my experience exceedingly rare although I find, not infrequently, that patients with essential infantile esotropia are referred to us with this diagnosis. In most instances the apparent inability to abduct either eye beyond the midline is caused by crossed fixation, the right eye fixating in left gaze and the left eye in right gaze. After occlusion of either eye for a few hours or when applying the doll's head manoeuvre abduction becomes normal. Pseudoparalysis of the lateral recti is also a frequent finding in the nystagmus blocking syndrome¹³ and in esotropia with manifest-latent nystagmus.¹⁴

Refractive accommodative esotropia

Even though the majority of children with refractive accommodative esotropia develop their squint between the ages of 2 and 3 years a refractive accommodative esotropia fully correctible with glasses may occasionally occur as early as during the first 6 months of life^{15,16} and thus be confused with essential infantile esotropia. The earlier view that accommodation is inactive in infants can no longer be upheld, as anyone can attest to who has done refractions in newborn infants. In fact, accommodation may reach adult capacity by the 4th month of life.¹⁷

Infantile esotropia with central nervous system disorders

The infantile esotropia occurring in patients with Down's syndrome, hydrocephalus, ocular or oculo-cutaneous albinism, cerebral palsy, meningocele etc. must be considered separately from essential infantile esotropia as different aetiologies may be involved. It is noteworthy, however, that these conditions are frequently associated with manifest nystagmus. We have recently examined 15 children with strabismus and ocular or oculo-cutaneous albinism. All had manifest nystagmus and all but three had a nystagmus blocking syndrome! It seems, therefore, that esotropia, seen so frequently with the central nervous system anomalies just mentioned may often have a common cause, namely the manifest nystagmus.

Sensory esotropia

Before returning to the principal subject of

this discussion we must consider sensory esotropia because it also may have its onset in infancy. It is a common misconception originating with Chavasse^{18,p.172,19} that an eye blind at birth, or blinded during the first few weeks or months of life, diverges. Actually, eso- and exotropias are encountered with equal frequency if the onset of the visual loss is between birth and the age of 5 years.²⁰ Thus, a careful fundus examination, if necessary under anaesthesia, belongs at the very beginning of an evaluation of any esotropic infant. Let us remember the observation of Ellsworth that esotropia is the second most common sign of retinoblastoma.²¹

Aetiology

The aetiology of essential infantile esotropia has been the subject of much speculation. Our current concepts have been substantially moulded by the work of two great English strabismologists, Claud Worth and Francis Bernard Chavasse. Worth, a native of Lancashire who founded the first Orthoptic Clinic at Moorfields, instilled sound scientific thinking and meticulous observation into the practice of strabismology at a time when squint was still considered by some as an affliction brought on by malignant spirits or as a sign of an evil disposition. His classical text, published first in 1903 has passed through nine editions, was re-written after his death by Chavasse and survived into our time through its latest edition by Keith Lyle and Bridgeman. Worth's concept of a congenital defect of the fusion faculty was derided by Chavasse of Liverpool, an equally profound and enthusiastic strabismologist, who saw the aetiology of squint in obstacles preventing the development of binocular reflexes in an otherwise normal visual system. In his delightfully poignant prose Chavasse considered the defect in 'fusion faculty' 'a superstition that may have had its uses in the past', or 'a fireless altar before which we need no longer vainly gesticulate'.^{18,pp.viii,2} The theories of Worth and Chavasse have often been contrasted with regards to their therapeutic implications which may be summarised as follows: an inborn defect in the fusion faculty precludes normalisation of binocular vision and banishes the surgical

treatment of infantile esotropia to creating, at best, a cosmetic improvement. On the other hand, removal of the obstacles that prevent the development of normal binocular reflexes by surgery at a young age improves the chances for a functional cure. Indeed, it was the teaching of Chavasse that has triggered a current trend, which began in the United States with Costenbader,²² to operate on infantile esotropes before completion of their second year of life to prevent deterioration of binocular functions.

In examining the writings of Worth and Chavasse the contrast between their theories appears less profound than generally assumed and their views have held up remarkably well in the light of current knowledge. When speaking of a congenital fusion defect Worth does not clarify, however, whether he is referring to a sensory or motor fusion defect, nor whether he was aware that such a difference existed. Yet, a clear distinction between these two important components of normal binocular vision is indispensable in the evaluation of treatment results. As such distinction is often lacking in the modern literature but essential for communication I must digress for a moment and clarify our terminology which is based on the classical concepts of sensory physiology and thus on the foundations laid down by Hering, Helmholtz, Tschermak and Bielschowsky. I define sensory fusion as the unification of visual excitations from corresponding retinal elements of similar size and brightness into a single mental percept. These excitations arise from the two foveas but also from the retinal periphery. Thus, one cannot, as is commonly done, equate sensory fusion with 'central' fusion and 'peripheral' fusion with motor fusion because the retinal periphery is also involved with sensory fusion. The term central fusion, so frequently encountered in the current literature, should be avoided altogether since the entire process of fusion, sensory and motor, is a central one. For stable sensory fusion to come about motor fusion is essential. Motor fusion is evoked by retinal image disparity outside Panum's area. Such disparity elicits fusional vergence movements with the purpose to keep the eyes aligned so that sensory fusion can be main-

tained. It is important to note here that sensory and motor fusion may be independently impaired. We have observed patients with the ability to have momentary sensory fusion with normal stereoacuity but with defective motor fusion because of a post-traumatic motor fusion deficiency.²³ Likewise, motor fusion may be normal and stable in the absence of bifoveal sensory fusion but not in the absence of peripheral sensory fusion.

When speaking of 'the instinctive tendency to blend the images formed in the two eyes' Worth may have referred to sensory fusion. However, when he described 'the desire for binocular vision' that keeps the eyes straight,^{24,p.54} or when he comments that he could not elicit fusional vergence movements on a haploscope in an esotropic patient^{24,p.60} it appears that he was referring to motor fusion instead. Be this as it may, according to Worth the development of this fusion faculty in squinters is either delayed, functions imperfectly, or not at all. The eyes are then 'in state of unstable equilibrium, ready to squint either inwards or outwards on slight provocation',^{24,p.55} The factors mentioned by Worth as causing strabismus in the presence of defective fusion, such as uncorrected hypermetropia or anisometropia are similar to those which, according to Chavasse, pervert the development of normal binocular reflexes.

On the basis of current knowledge it has become possible to add to the theories of Worth and Chavasse. Worth already suspected that vergence movements lack precision in newborns and modern psychophysical research in infants has shown indeed that the motor and sensory components of binocular vision are only incompletely developed at birth.^{25,26} For instance, visual acuity,²⁷ contrast sensitivity^{28,29} and stereopsis^{30,31} during the first few months of life are far from having reached their adult levels of accuracy. Since the development of stable ocular alignment may depend on a high quality visual input²⁶ binocular vision may be vulnerable during this period of immaturity. This may explain the transient strabismus so frequently observed in normal infants.^{7,8}

I would like to propose a working hypothesis according to which a delay in the

development or a permanent defect of motor fusional vergences in a sensorially normal infant causes esotropia during the vulnerable first three months of visual immaturity under the influence of factors that destabilise the oculomotor equilibrium. What is the nature of these factors? In addition to uncorrected hypermetropia and anisometropia already mentioned by Worth, excessive tonic convergence, an abnormally high AC/A ratio, or anomalies of the neural integrators for vergence movements which were recently discovered in the brainstem³² need to be considered here. Claud Worth already recognised fully that once fusion is firmly established none of these potentially strabismogenic factors short of an extraocular muscle paralysis can cause squint.^{24,p.54} In view of the frequent occurrence of infantile esotropia in members of the same family some of these components or, perhaps, the defect of motor fusion itself must be hereditary.

Two findings in essential infantile esotropia are seemingly at odds with our working hypothesis, namely the defective or absent stereopsis in the treated patient and the asymmetry of the optokinetic nystagmus.

The fact that not a single patient with essential infantile esotropia has ever been recorded as having normal stereopsis when measured with random dot tests has often been taken as evidence for a primary sensory defect that precludes the development of normal binocular vision. Since stereopsis has been linked to the presence of binocular neurons in the visual cortex^{33,34} we must at least consider the possibility that a congenital absence of binocular cells at birth may lead to instability of binocular vision which, in turn, causes the strabismus. In the absence of a valid animal model for essential infantile esotropia this argument is difficult to prove or disprove but there are several observations that cannot be reconciled with this theory. First, recent data suggest that random dot stereopsis does, at least transiently, occur in esotropes during infancy when the deviation is neutralised with prisms³⁵ or after surgical alignment of the eyes.³⁶⁻³⁸ Second, every clinician knows that there are many patients whose eyes are perfectly aligned in spite of stereoblindness. Third, we have

shown that artificial strabismus produced in infant monkeys for as briefly as one week duration decimates the number of striate neurons that normally receive input from both eyes.^{39,40} This loss of binocular neurons is irreversible even after realignment of the eyes and subsequent exposure to a normal visual environment for as long as two years.⁴¹ Since these binocular neurons have been linked to stereopsis^{33,34} it is of special relevance for this discussion that we have not observed a single instance of strabismus in more than 30 monkeys that had unilateral lid suture or artificial anisometropia during infancy and consequently, had lost all their binocular cells and their stereopsis. In view of these findings we propose that defective stereopsis in infantile esotropes is an irreversible consequence of strabismus early in infancy rather than the manifestation of an obscure primary sensory defect.

A similar cause and effect relationship may exist in regard to the frequent occurrence of asymmetric nystagmus in infantile esotropes.^{42,43} In normals the optokinetic response evoked by a rotating nystagmus drum consists of a smooth pursuit movement in the direction of the moving stripes, followed by a corrective saccadic movement in the opposite direction (optokinetic nystagmus). Normally, the pursuit movement occurs with equal facility, regardless of whether the drum is moved into a naso-temporal or temporo-nasal direction. In infantile esotropia, however, this symmetry is grossly disturbed and the optokinetic response becomes asymmetric, i.e. irregular or difficult to elicit when the stripes move in a naso-temporal direction (Fig. 5). This naso-temporal directed anomaly of pursuit movements in infantile strabismus has been interpreted as a defect in visual motion processing due to a maldevelopment of the visual cortex.⁴⁴ We believe that there exists a more plausible explanation for optokinetic asymmetry. Naso-temporal optokinetic asymmetry is a consistent feature in normal infants during the first 3 months of life after which the optokinetic responses become symmetric.^{45,46} Disruption of binocular vision because of strabismus,^{42,47,48} anisometropia,⁴⁹ lid closure at birth,⁵⁰ or congenital cataracts⁵¹

cause an arrest of the development of the pursuit system at an infantile stage⁴² and thus optokinetic asymmetry persists beyond visual immaturity into adulthood. Indeed, in a preliminary study carried out by my collaborator, Dr. John Davis,⁵² a good correlation existed between the onset of strabismus and the presence of optokinetic asymmetry (Fig. 6).

We propose, therefore, that optokinetic asymmetry, similar to defective stereopsis, is a consequence of strabismus rather than of a

Fig. 5. Binocular nystagmogram during optokinetic stimulation from nasal to temporal (upper tracings) and temporal to nasal (lower tracings) in a normal subject (left) and a patient with essential infantile esotropia (right). Note poor optokinetic response when the drum is moved in a naso-temporal direction in the esotropic patient.

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Fig. 6. Results of electronystagmographic recordings of optokinetic nystagmus in 26 patients with a documented onset of esotropia at various ages.

primary structural defect in the visual pathways. Our working hypothesis of the pathogenesis of essential infantile esotropia, which will have to be modified as further information becomes available, is summarised in Figure 7.

In following this discussion of the aetiology of essential infantile esotropia one may wonder who was right, Worth or Chavasse? Worth was wrong in considering fusion a 'psychical' process, as there is abundant evidence that it is a physiological process instead. Considering the high degree of functional restoration that can be achieved in essential infantile esotropia by surgery and in view of the arguments just presented we also can no longer speak of a primary fusion defect, if this defect is defined as an absence or delayed development of sensory fusion. On the other hand, if Worth had made it clear that he was talking about a motor fusion defect the hypothesis I presented would be quite compatible with his view. Chavasse was correct in drawing attention to the many factors that may interfere during infancy with the development of normal binocular

reflexes although he overemphasised, in my opinion, the roles of refractive errors and of anisometropia. Thus both of the old masters may have been on the right track and their names have taken new lustre from the touch of time.

Preoperative Evaluation and Treatment

Although the treatment of essential infantile esotropia is ultimately surgical a few practical comments are in order regarding the preoperative management. Amblyopia or a history of previously treated amblyopia occurred in 35% of 408 patients with essential infantile esotropia.⁵³ Amblyopia must be eradicated without delay, and prior to surgery, by the appropriate treatment.⁵⁴ After surgery amblyopia prophylaxis must continue until the child is beyond the susceptible period of recurrence of amblyopia, which occurs usually between the age of eight and ten years. In addition to periodic reinforcements of the formerly amblyopic eye by part-time patching of the dominant eye, alternating penalisation has been proven to be effective in this regard.⁵⁵ We use two pairs

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Fig. 7. Working hypothesis of aetiology of essential infantile esotropia. For explanation see text.

of spectacles on alternative days, blurring either the right or the left eye with a +3.00 spherical lens.

Once infantile esotropia has become firmly established correction of a mild or moderate hypermetropic refractive error is rarely of benefit to the patient. However, we do recommend a trial with spectacles or miotics prior to surgery when the refractive error exceeds two and a half dioptres.

Finally, we must draw attention to the frequent occurrences of a dissociated vertical deviation (DVD) and/or upshoot of the adducted eye, apparently caused by a primary overaction of the inferior oblique muscle(s). DVD occurred in 51% and upshoot in adduction in 68% of our patients.⁵³ These figures are in accord with prevalences reported by other authors.⁵⁶ The results of quantitative measurements obtained in 170 of 408 patients with essential infantile esotropia and DVD are presented in Table III. The reason for the frequent occurrence of DVD or of inferior oblique overaction in patients with essential infantile esotropia is not at all clear. It is of interest to note, however, that overacting inferior obliques, usually associated with a V-pattern, are commonly detected during the initial examination of the child. DVD, on the other hand, is rarely present in infancy and occurs, as a rule, between the ages of 3-5 years and often several years after surgical alignment of the eyes.

We now treat a cosmetically significant DVD with a 7-8 mm recession of the superior rectus muscle. This amount is varied with asymmetric involvement.

Surgical Treatment

The aim of surgery for essential infantile esotropia is to align the eyes as closely to the orthotropic position as possible with the least number of operations. My own surgical

approach has undergone periodic changes over the years. Initially, we favoured a recession/resection operation on the non-dominant eye, combined with inferior oblique myectomies if indicated, to be followed, if necessary, by a recession/resection on the fellow eye. The amount of surgery varied according to the size of the deviation and on the basis of observations made during examination of the ductions of the eyes⁵⁷ and ranged from 3-5 mm recessions and 5-8 mm resections. However, the number of reoperations required to gain or maintain alignment was discouraging. In 1972 we reported that an average of 2.1 operations per patient was required to align the eyes.⁵⁸ Ing and coworkers,⁵⁹ who used 3-5 mm recessions of both medial recti required as many as 2.6 operations per patient to achieve this goal. In recent years we have changed our method and now employ recessions of the medial recti ranging from 5-8 mm.⁶⁰ This more aggressive approach has decreased the need for additional surgery and, contrary to our initial concern, does not cause limitation of adduction. Initial reports show that by doing these unconventionally large recessions of both medial recti 73-84% of the patients are successfully aligned with one operation.⁶¹⁻⁶³

Botulinum injection into the medial recti has been mentioned as an alternative to surgery.^{64,65} While chemodenervation with Botulinum has been most effective in the treatment of essential blepharospasm and may also be useful in certain cases of paralytic strabismus caution is advised with regard to its role in infantile esotropia. The need for frequent injections under anaesthesia, the common complication of ptosis and transient vertical deviations, the conspicuous lack of prolonged follow-up and of controlled studies comparing the results of surgery versus injections⁶⁶ have kept us from accepting Botulinum as a valid treatment option at this time.

Results of Treatment

What can be expected from surgery in terms of restoration of binocular vision and how critical is it to perform surgery in early infancy? These questions are difficult to answer from the literature because of a deplora-

Table III *Characteristics of DVD in essential infantile esotropia*

(n=170)			
Unilateral	24(14%)	symmetrical	13 (9%)
Bilateral	146 (86%)	asymmetrical	133 (91%)

ble lack of precise terminology in describing treatment results. Most current authors use one or several of the following findings as criteria for the restoration of binocular functions: the presence of gross stereopsis,^{67,68} fusion on the Worth 4 dot test,⁶⁷⁻⁶⁹ appreciation of both stripes on the Bagolini striate lens test^{61,67,69}, alignment of within 10 prism dioptres of the orthotropic position^{56,59,67} or the presence of a fusional vergence movement on the cover-uncover test.⁷⁰ However, there is no single test that provides all the answers about the extent of recovery of motor and sensory binocular functions. Nor are any of the tests just mentioned, without the application of additional tests, capable of detecting subtle but functionally important variations between normal, subnormal, abnormal or absent binocular vision.⁷¹

The current lamentable practice of grouping together normal and abnormal retinal correspondence has done little to advance our knowledge. By distinguishing clearly, whenever this is possible, between the various types of binocular cooperation in the treated patient a more meaningful analysis of treatment results becomes possible.

We classify our results into four categories: (1) subnormal binocular vision, (2) microtropia, (3) residual small angle eso- or exotropia and (4) large angle esotropias or consecutive exotropias that may require further surgery and have described elsewhere the combination of sensory and motor tests required to diagnose these conditions.⁷¹ The clinical characteristics of each of these four end stages of surgical therapy are listed in Tables IV-VII. While there is unanimous agreement that a complete normalisation of all binocular functions including normal random dot stereoacuity is an unattainable goal in essential infantile esotropia we shall see that a state of near normalcy is by no means beyond the reach of the therapist. Subnormal binocular vision, a term introduced into the strabismus literature by the late Keith Lyle⁷² must be considered as an optimal outcome of treatment. Retinal correspondence is normal and with the exception of normal random dot stereopsis and bifoveal sensory fusion, such patients have nearly normal binocular vision. More common endstages of treatment are

microtropias or residual small angle eso- or exodeviations. These patients maintain binocular vision on the basis of anomalous retinal correspondence,⁷³⁻⁷⁵ require no further treatment except for amblyopia and are considered to have an acceptable treatment result.

Anomalous retinal correspondence, was once seen as an unsurmountable obstacle to normal binocular vision and treated with

Table IV *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
No treatment

Table V *Microtropia*

Inconspicuous shift or no shift on cover test
Mild amblyopia frequent
Fixation parafoveal
Fusional amplitudes
Anomalous retinal correspondence
Reduced or absent stereopsis
Some stability of alignment
Amblyopia treatment up to school age

Table VI *Small angle eso/exotropia (<20 prism dioptres)*

Cosmetically acceptable
80% have anomalous retinal correspondence
Less stability of angle
No treatment except for amblyopia

Table VII *Large angle eso/exotropia (>20 prism dioptres)*

Usually cosmetically unacceptable
Less chance for anomalous retinal correspondence, suppression prevails
Unstable angle
Further surgery often required

great zeal and conviction by many orthoptists. It is now viewed in an entirely different light. The patient with anomalous correspondence gains many functional benefits from this sensorial adaptation to a residual angle of squint. These benefits include an intact binocular field of vision, stability of the residual angle, fusional vergences,⁷⁶⁻⁷⁸ normal distance judgement^{79,80} and even gross stereopsis.^{73,75,81} Above all, it permits a patient with a residual manifest strabismus to enjoy single binocular vision. This is the principal reason why I am opposed to treating anomalous correspondence by orthoptics, a practice still rampant in various parts of the world. Anyone who has ever had to deal with the unfortunate patient who by lengthy and often costly eye exercises is now incapacitated by constant diplopia will agree with this assessment.

Finally, we must discuss the optimal age at which surgery should be performed. In my professional lifetime I have seen a progressive shift towards early surgery. In the sixties,

and in Europe largely under the influence of Lyle, Hugonnier and Arruga, surgery was usually performed between the ages of two and five years of age. The current tendency to attempt alignment earlier and preferably prior to the age of 18 months began in the United States with Costenbader and his school.^{59,82} Many authors claim that only by operating at this age can a functionally useful result be obtained^{59,68,69} and that surgery performed later precludes such an outcome.^{4,p.89} We have recently published our results in 358 patients operated on for essential infantile esotropia⁵³ and I will only summarise these data here. The mean follow-up was 39.4 months (1-203 months). The onset of the squint prior to the age of six months was confirmed by an ophthalmological examination or, in the few cases where this was not possible, documented on photographs supplied by the parents. The results are presented in Figure 8 and show that, as the age at completion of surgery increases, the probability of achieving subnormal binocular vision,

PREVALENCE OF FUNCTIONAL STATE ACCORDING TO AGE AT ALIGNMENT (358 Patients)

Fig. 8. Prevalence of surgical results in 358 patients with essential infantile esotropia according to age at alignment. SBV, subnormal binocular vision, ET, esotropia, XT, exotropia. From von Noorden⁵³.

defined by us as an optimal result decreases ($p=0.002$). Increasing age at the completion of treatment tends to move the patients into the less favourable though still acceptable categories of microtropia or small angle esotropia. Thus our findings are in support of the current tendency to complete surgical alignment prior to the age of two years. However, they also show that many patients achieved satisfactory results when surgery was delayed until after the ages of two or even four years.

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