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## UNTREATED EARLY ONSET ESOTROPIA IN THE VISUAL ADULT

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**Abstract**

*One hundred and forty two patients with a history of esotropia present before the age of six months who had had no previous ophthalmic examination, and who were aged eight years or more were examined in order to ascertain the presence or absence of DVD, manifest latent nystagmus and asymmetric OKN, the main features of essential infantile esotropia (ET).*

*Twenty nine patients showed no evidence of essential infantile esotropia, despite the history of onset of strabismus under the age of six months.*

*One hundred and thirteen cases were diagnosed as having essential infantile esotropia. There were 90 patients with manifest latent nystagmus, 67 with DVD and 97 with an asymmetric OKN response. Only eight cases had visual acuity in the non-fixing eye of less than 6/12. 'A' pattern esotropia was present in 72 cases and 'V' pattern elicited in 16 patients. There were no abnormal head postures.*

*It is suggested that the untreated condition of essential infantile esotropia should be studied before making definitive conclusions about the characteristics of the condition.*

**Key words:** *'A' pattern, amblyopia, manifest latent nystagmus, DVD, asymmetric OKN.*

## INTRODUCTION

Concomitant convergent strabismus with a reported onset before the age of six months is a frequently researched, well documented and easily identified phenomenon. The literature is full of erudite communications on the subject, investigating all aspects of the aetiology, and detailing the various manifestations and their implications for surgical and non-surgical management. The terminology has changed since it has been shown that the condition is not present from birth. The term congenital is discouraged and the terms early onset and infantile esotropia are variously used in the literature. In an attempt to further clarify the situation, von Noorden[1] has detailed the various types of early onset esotropia and suggested the preposition of 'essential' before infantile esotropia to describe those patients who conform to the syndrome described by Kommerell,[2] where a large esotropia is associated with manifest latent nystagmus and monocular opto-kinetic asymmetry. The presence of DVD is accepted as a further main feature of the syndrome. Previous clinical studies have concerned either neonates or infants in developed countries who have been documented from the age of presentation in the early days of their life, until their discharge as visual adults. These prospective studies have not taken into account several important factors; firstly that detailed assessment of eye movements in early infancy is not always possible; secondly that current methods of estimating the presence or absence of amblyopia in babies are less than accurate

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and finally whether the treatment which has been carried out has had any influence on the natural history of the condition.

## METHODS AND MATERIALS

Between October 1986 and October 1989, all new patients referred to Groote Schuur Hospital in Cape Town and St John's Eye Hospital, Baragwanath with concomitant esotropia which was said to have been present before the age of six months, who had had no previous ophthalmic examination and who were aged eight years or more, were entered into the study. Also included were patients seen during a tour of the Sightsavers Team of the Bureau for the Prevention of Blindness to the homelands of Transkei and Kwazulu. A full orthoptic examination including cover test and ocular movements, fixation preference assessment, and prism cover test for near and distance, and on up and down gaze in order to determine the presence of 'A' and 'V' patterns, together with special reference to testing for manifest latent nystagmus, DVD, and asymmetric optokinetic nystagmus. Any abnormal head posture was noted. OKN was tested with a hand held drum which was rotated at 20 cycles per minute at approximately 30 centimetres from the patient. A visuscope was used to check fixation and to elicit manifest latent nystagmus if it had not been seen on cover testing, and a synoptophore was used to search for DVD if distance cover test with Spielmann occluder had failed to reveal its presence. Each patient was refracted following the instillation of cyclopentolate drops, and the media and fundi were examined. Where possible the patient was checked by a second coworker, and random sampling was undertaken to ensure continuity of test standards. Subsequently patients were checked again when they attended for surgery and post-operative data was obtained. All follow-up information on the patients was checked in August 1992.

## RESULTS

One hundred and forty two patients fulfilled the criteria and were included in the series. There were 106 patients of African origin, 34 of mixed race, one Asian and one Caucasian.

The mean angle of deviation was 49.4 dioptres.

One hundred and thirteen (79.6%) patients showed one or more signs of essential infantile esotropia in that manifest latent nystagmus, DVD or asymmetric OKN had been elicited in the presence of a large esotropia of onset before six months of age.

Twenty nine (20.4%) cases reported a history of esotropia before the age of six months, but had none of the signs associated with essential infantile ET.

The incidence of amblyopia of more than two lines on the Snellen chart was 19.7% (28 cases).

The overall incidence of DVD was 47.1% (67 patients), of manifest latent nystagmus, 63.4% (90 cases) and asymmetric OKN was 68.3% (97 cases).

Seven and three-quarter per cent (11 patients) had an anomalous OKN response and large esotropia without manifest latent nystagmus or DVD.

'A' pattern esotropia occurred in 57% (81 cases) and 'V' pattern in 14.8% (21 patients).

No patient was reported to have an abnormal head posture, and there were no underactions of the lateral recti of more than minus one when assessed on a scale of one to four.

For further analysis the patients were then divided into two groups; Group A for those with one or more signs of essential infantile esotropia.

Group B for patients with other types of early onset esotropia. It cannot be assumed that the history was inaccurate in the 29 cases in Group B with no sign of essential infantile esotropia, and normal OKN. However, these patients did not fall into the well defined categories described by von Noorden[1] i.e. nystagmus blocking syndrome, VI nerve palsy, accommodative esotropia, infantile esotropia with CNS disorders, infantile esotropia with sensory disorders, Duane's retraction syndrome and Moebius syndrome. We suspect that they form a further group of early onset esotropes whose deviation occurs before the age of six months but after the maturation of the nasotemporal OKN response.

GROUP A

One hundred and thirteen cases with one or more signs of essential infantile ET were identified, of whom 90 were of African origin, and 21 of mixed race. There was one Asian and one Caucasian. The mean angle of deviation was 49.8 dioptres.

*Latent Nystagmus, Asymmetric OKN and DVD*

Ninety seven (85.8%) patients demonstrated an asymmetric OKN response, 67 (59.2%) showed DVD and in 90 (79.6%) latent nystagmus was elicited. Forty seven (41.5%) cases showed DVD, latent nystagmus and asymmetric OKN. Eighty (70.7%) patients with anomalous OKN were shown to have latent nystagmus, and 53 (47%) had DVD.

Fifty four (47.8%) patients had latent nystagmus and DVD but only eight of these (7.1%) cases were judged to have normal OKN responses. Eleven cases showed asymmetric OKN and esotropia without other signs of essential infantile ET.

*Amblyopia and Fixation Preference*

Fifty six (49.6%) patients showed no fixation preference and free alternation at first assessment.

Of the 57 (50.4%) cases with fixation preference, 22 (38.6%) had a difference in the visual acuity between the two eyes of more than two lines on the Snellen chart, but 49 (86%) of the 57 achieved vision in the non-preferred eye of 6/12 or better. There was significant unilateral refractive error in five of the eight amblyopic patients.

*'A' and 'V' Patterns*

Seventy two (63.7%) cases had 'A' pattern esotropia and 16 (14.1%) patients had a 'V' pattern.

GROUP B

Twenty nine cases were found to have no sign of essential infantile ET. Eight (27.5%) patients had amblyopia of more than two lines on the Snellen chart, but in only one case was there a relevant unilateral refractive error.

Nine (31%) cases demonstrated 'A' pattern ET and five (17.2%) had 'V' pattern ET.

DISCUSSION

Prospective studies of early onset esotropia in developed countries have a major advantage over a retrospective study such as this, in that the medical diagnosis is generally made very early in life and corroborated with photographic evidence. However, the great difficulty in accurate assessment of the young infant, and the natural desire to re-align the visual axes, at the earliest opportunity, for both cosmetic purposes and the possibility of obtaining some rudimentary post-operative binocularity, may well have obscured the natural history of the early onset esotropia.

*Amblyopia and Fixation Preference*

The original hypothesis was that children with untreated infantile esotropia are less likely to become amblyopic than their contemporaries who have early surgery. This is completely at odds with all the current literature on infantile esotropia. Von Noorden[1] describes amblyopia as a characteristic of essential infantile esotropia with an incidence of 35%, Hiles[3] suggests that up to 75% of the cases may need occlusion; Pratt-Johnson and Tillson[4] report an incidence of amblyopia of 50%, with 30% having visual acuity of less than 6/12 in the strabismic eye; Walker and Taylor[5] found 45% of their patients still amblyopic at final assessment and Dickey and Scott[6] found more than two lines difference in the visual acuity of 51% of their patients. Few authors specify, however, whether there was amblyopia before therapy commenced, or on what basis the diagnosis of amblyopia in infancy was made. The only paper which addresses the subject of pre- and post-operative amblyopia is that of Hoyt[7] who compared visual acuity before and after surgery and found only four (12.9%) patients out of 31 to have been amblyopic pre-operatively but that 19 (61%) developed amblyopia within the year after operation, thus suggesting that the post-operative cases were considerably more at risk of amblyopia than the untreated cases. In our study the incidence of amblyopia with a visual acuity in the fixing eye of less than 6/12 was only 7% in the untreated essential infantile esotropes. Therefore the un-

treated cases had a significantly lower incidence of amblyopia than their treated counterparts. Hoyt[8] has recently confirmed these findings in a series of untreated infantile esotropes who are mainly of Asian origin, unlike those in our series who are mainly African. This refutes the suggestion that the racial basis in our series affects the conclusions and confirms that the low incidence of amblyopia is characteristic of the condition.

However, some of the patients in the study did become amblyopic and the reason for this has to be investigated. The original examination noted in all cases not just the visual acuity but also the fixation preference, since this is the method most commonly used in the diagnosis of amblyopia in the pre-verbal infant. A study of the 113 patients with essential infantile ET reveals that although 56 patients showed free alternation, 11 of these patients did have a difference between the visual acuity of the two eyes, although in each case the visual acuity of the less good eye was more than 6/9.

Of the 57 cases with a fixation preference, 24 had equal visual acuity or visual acuity of 6/6 or more in the non-fixing eye, and a further 14 achieved 6/9 in the strabismic eye.

Thus 19% of the alternators had an element of amblyopia and 66% of the cases with fixation preference had little or no amblyopia. This would suggest that fixation preference is not a reliable method of diagnosing amblyopia, and may well account for the seemingly high incidence of amblyopia suggested in studies conducted in infancy.

Nineteen patients failed to see 6/9 with the non-fixing eye, but 11 cases did achieve 6/12, and in six (54.5%) of the patients there was a significant degree of anisometropia.

There were eight patients with dense amblyopia ranging from 6/18 to light perception. Five (62.5%) cases had significant anisometropia, but there was no correlation between the amount of anisometropia and the depth of the amblyopia. There was significant astigmatic error in the strabismic eye in four of these patients.

Four patients had bilateral amblyopia due to bilateral uncorrected refractive error.

Uncorrected refractive error was therefore the cause of the amblyopia in 65% of the cases who achieved 6/12 or less in the non-preferred eye. The amblyopia in these patients would therefore appear to be of refractive rather than strabismic origin. It has been suggested that the patients with amblyopia in the absence of refractive error, may well have had an episode of infection or trauma during the sensitive period which could have resulted in stimulus deprivation amblyopia.

The important fact is, that out of 113 essential infantile esotropes, only 19 (16.8%) failed to achieve visual acuity of 6/9 in the squinting eye and only eight (7%) registered vision of less than 6/12 in the deviating eye.

*Manifest Latent Nystagmus and DVD*

The presence of latent or manifest latent nystagmus and DVD have long been recognised as integral elements of essential infantile ET. Lang[9] has suggested that the dominant feature of the infantile ET syndrome is latent nystagmus and Kommerell[2] and von Noorden[1] agree that it is a main feature of the condition. It is thought to originate as manifest nystagmus, often rotary in nature, and become latent with the passage of time, hence the manifest latent description. In this series there was an incidence of manifest latent nystagmus of 79.6%. Helveston[10] has suggested that manifest latent nystagmus is invariably found in conjunction with DVD, while Harcourt[11] describes an incidence of 73%. This study revealed DVD in association with manifest latent nystagmus in only 47.8% of the cases. This was due to a relatively low incidence of DVD in this group of patients, only 59.2%. It is possible that this low incidence is due to the difficulties of eliciting subtle amounts of DVD in the presence of very large esodeviations. DVD has been specifically searched for post-operatively where it was absent prior to surgery and four cases have been found. However, 18 patients with essential infantile ET without DVD did not attend for surgery or follow-up appointment.

The age at which DVD occurs is said to be between the ages of 18 months and three years, and it would be reasonable to surmise that many patients will have had surgery by this juncture. It is impossible to say whether the four patients

in our series who demonstrated DVD post-surgery did so as a reaction to the operation, or whether the removal of the gross esotropia revealed a pre-existing DVD. Parks[12] has suggested that DVD may diminish or disappear before the age of eight years. This could be an explanation for the low incidence in this series, although the suggestion has been refuted by most other authors.

*Asymmetric OKN*

The asymmetric OKN response in essential infantile ET has become accepted as an integral aspect of the condition. The aetiology is far from clear. Fitzgerald and Billson[13] found a high correlation between abnormalities of VERs suggestive of abnormal visual pathway projection and anomalous nasotemporal OKN. They suggested that patients with asymmetric OKN might be at risk of developing DVD. Atkinson,[14] amongst others has found that infants of two to three months of age demonstrate asymmetric OKN, which would suggest that the anomalous OKN response is a consequence of the strabismus disrupting the normal development of the pursuit system, rather than of a primary structural defect of the visual pathways. Nevertheless, as Demer and von Noorden[15] have shown, the presence of an asymmetric OKN response suggests an 85% chance that the onset of the strabismus was before the age of six months. We found an 85.8% incidence of asymmetric OKN in our study, which would appear to confirm the accuracy of the history as well as emphasising the importance of this phenomenon in essential infantile ET. With an incidence of only 59.2% of DVD, and only a 47% incidence associated with asymmetric OKN, it was not possible to confirm a relationship between asymmetric OKN and DVD. This study of the natural history seems to confirm that manifest latent nystagmus (79.6% incidence) and asymmetric OKN (85.8% incidence) are the two main features of essential infantile ET but that the signs of manifest latent nystagmus, DVD and asymmetric OKN, while frequently present, are not invariably linked.

*'A' and 'V' Patterns*

The association of 'A' pattern with essential infantile ET was first noted by Lang,[16] and the prevalence of an 'A' phenomenon has been reiterated on many occasions by Mein,[17] although most authors persist in attributing the upshoot in adduction seen in these patients to primary overaction of the inferior oblique. von Noorden[1] cited an incidence of 68% of inferior oblique overaction in patients examined during infancy, while Hiles[18] found 78% of his patients with overaction of one or both obliques. We found an incidence of 63.7% of 'A' patterns compared with an incidence of 14.1% of 'V' patterns. These figures correlate well with those of Harcourt and Mein[19] who cite an incidence of 60.4% of 'A' patterns and 25.5% of 'V' patterns in patients with DVD. It seems likely that in those series citing a high incidence of inferior oblique overaction and 'V' patterns, the upshoot on adduction which occurs with DVD has been attributed to a primary overaction of the inferior oblique muscle, and that the diagnosis of 'V' pattern has been made without comparison of the cover test, or prism cover test measurements, on up and down gaze. It should be noted therefore that an upshoot on adduction is not necessarily an inferior oblique overaction, but may be the first manifestation of DVD, and 'A' and 'V' patterns diagnosed only when accurate assessment of the esotropia on up and down gaze can be made. The fact that the upshoot in adduction may be elicited at an early age may necessitate also reconsideration of the age at which DVD occurs. It is unlikely that the high incidence of 'A' patterns in this group of essential infantile esotropes is due to the 74.6% incidence of patients of African origin. We have not been able to elicit clinical evidence to support this hypothesis, and it is likely that there could be wide variations in the types and manifestations of strabismus in Africa. The IOA survey in 1983 suggested a higher incidence of esodeviations in patients of African origin, but while in southern Africa there does appear to be a prevalence of esotropia, patients originating from other areas often present with large exodeviations, with associated 'V' patterns.

*Compensatory Head Posture*

Harcourt and Mein[18] report an incidence of compensatory head posture in essential infantile

ET of 75%. It is thought to be motivated either by the attempt to fix in adduction to obtain the best visual acuity by minimising the nystagmus, or possibly to compensate for the restriction of abduction, or DVD. It is frequently of variable nature and is often seen only on testing the visual acuity. We found no patient with a compensatory head posture. The only possible explanation is that whilst the majority of our patients were literate, education probably commenced at a much later age than in the developed countries, and there would have been much later exposure to TV and reading matter. It is possible that in these patients the manifest component of their manifest latent nystagmus had disappeared long before they needed accurate vision.

*Post-Operative Results*

Of the complete group of 142 patients a total of 35.2% did not attend for surgery, and in the essential infantile ET group this rose to 37.2% (42 cases). This may be attributed to the fact that in this patient group cosmesis is not a problem and that the family may have been reassured with regard to the vision at the first visit. Of the patients who did consent to operation, a satisfactory cosmetic result was claimed in all cases. The final angle of deviation varied from 10 dioptres of exotropia to 25 dioptres of esotropia, the majority falling within the five to 15 dioptre range. No patient complained of diplopia following operation. No patient achieved binocular vision following surgery. The incidence of binocularity in patients who have had early surgery is low, and although there have been reports in the literature of adults with long-standing esotropia achieving binocularity with so called peripheral fusion, and monofixation syndrome following surgery for strabismus as adults, it is very unlikely that any worthwhile binocular vision could be elicited in patients who have squinted since the first few months of life.

Whilst accepting that the patients in this group, by having late surgery, have foregone any chance of developing binocularity, it has to be remembered that the underprivileged child or the child in Third World with essential infantile esotropia is being put at risk of developing amblyopia if operated early, and improving cosmesis is of secondary importance to the retention of good visual acuity in each eye.

## CONCLUSIONS

The cases of essential infantile ET who remained untreated until visual adulthood demonstrated a high incidence of manifest latent nystagmus and asymmetric OKN, which appear to confirm these phenomena as the characteristic features of the condition. The correlation between manifest latent nystagmus and DVD of more than 70% is not confirmed by this study. Examination of post-operative data has shown that subtle amounts of DVD have been elicited after surgery but the high failure rate of patients in attending for operation means that conclusions cannot be drawn over the appearance of DVD post-operatively. It is not possible to hypothesise as to whether, in some cases, the DVD had been present in the past and had disappeared before the initial examination. The reported age of onset of DVD may well have to be reconsidered if the upshoot on adduction which has been attributed to inferior oblique overaction in infancy, can be proved to be due to the first manifestation of the DVD.

This series has demonstrated clearly that amblyopia is not a characteristic of essential infantile ET. The amblyopia which has occurred in this series has been the result of uncorrected refractive error in over 65% of the cases. It is possible that, with a population such as we have studied, there may well be an element of stimulus deprivation amblyopia in some of the amplyopes without refractive error. The amblyopia which occurs in the treated cases is a direct result of the surgical intervention.

The incidence of 'A' patterns in 63.7% compared with 'V' patterns in only 14.1% confirms that an 'A' pattern is an important feature of the condition and re-emphasises the fact that the upshoot in adduction should not be attributed to inferior oblique overaction, and a 'V' pattern should be diagnosed only when confirmed by measurements in up and down gaze.

The absence of a compensatory head posture and lateral rectus weakness are interesting. The

head posture described by other authors is invariably associated with visual tasks; it is possible that a lack of specific visual stimulation at an early age has resulted in its non-appearance.

Finally it is to be hoped that this study will have been seen to emphasise that the management of essential infantile ET in the developed countries which is based on the necessity of removing the cosmetic aberration as soon as viable, should be modified when treating patients in the developing countries, where access to medical facilities is limited. For the underprivileged, cosmesis may not be a problem and retention of good visual acuity in both eyes, is the first priority. This study has shown that patients who can demonstrate one or more characteristics of essential infantile ET, have an alternating esotropia or equal visual acuity and absence of unilateral or high bilateral refractive error are at very low risk of developing amblyopia if left untreated. Whilst they may not achieve binocular single vision if the surgery is carried out later in life there is a low risk of diplopia.

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