THE OKN RESPONSE AND BINOCULAR VISION IN EARLY ONSET STRABISMUS*

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Abstract

Asymmetry of uniocular opto-kinetic nystagmus (OKN) occurs when a normal response is elicited by moving the targets in a temporal-nasal direction but not by moving them in a nasal-temporal direction. This finding has been reported in normal infants and in those with congenital esotropia.

In this study OKN has been investigated in 69 patients with early onset strabismus and in 25 patients with later onset strabismus. Asymmetry has been found only in those with strabismus, nystagmus and dissociated vertical divergence (DVD) and not in other patients with early or later onset strabismus. Although superficial binocular vision was found in some cases it was insufficient to maintain peripheral fusion.

Key words: *Dissociated vertical divergence, opto-kinetic nystagmus, nystagmus, congenital esotropia.*

INTRODUCTION

Early onset strabismus is defined as a manifest deviation present at birth or occurring within the first six months of life. This paper is based on a further study of patients in two series previously reported. Series 1 comprised 100 patients with early onset strabismus and dissociated vertical divergence (DVD) and was presented at the 4th International Orthoptic Congress (Mein and Johnson 1981);[1] 88 patients were esotropic and 12 exotropic, all had nystagmus. Series 2 comprised 130 consecutive patients with early onset esotropia who were currently attending the Orthoptic Clinic at the General Infirmary at Leeds: these patients were studied to assess their characteristics, the incidence of nystagmus and DVD, and their natural history. The results were presented at a meeting of the Consilium Europaeum Strabismus Studio Deditum (CESSD) in 1981 (Harcourt and Mein 1982).[2] From this study three groups emerged:

Group 1	Esotropia + nystagmus + DVD	43
Group 2	Esotropia + nystagmus – DVD	13
Group 3	Esotropia – nystagmus – DVD	74
		130

Group 1

The main characteristics of patients in this group were:

1. large angle esotropia with crossed fixation in nearly all cases.
2. early presentation; the average age when first seen was 10 months.
3. jerky horizontal nystagmus with the fast phase to the uncovered eye. Nystagmus appeared latent in the majority but 30% of patients seen in infancy in Series 1 had manifest nystagmus on presentation, usually rotary or with a rotary component,

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which gradually changed to the horizontal nystagmus more typical of the condition. According to Dell Osso *et al* (1979)[3] this nystagmus should be classified as manifest latent since no patient had binocular single vision.

4. gradual onset of DVD between the age of 18 months to three years, with a few later exceptions.
5. a face turn to the side of the fixing eye with or without a head tilt. We believe that the face turn is adopted to compensate for nystagmus rather than limited abduction.
6. a preponderance of A pattern.
7. poor or no binocular vision.

Group 2

Approximately half the patients presented with the same characteristics as those in Group 1 except that they did not develop DVD. It is possible that the younger patients will still do so. The remaining patients presented with smaller and often variable unilateral strabismus. A compensatory head posture and poor binocular vision were the rule.

Group 3

The main characteristics of patients in Group 3 were:

1. smaller angle esotropia, usually unilateral, with consequent amblyopia.
2. later presentation, averaging two years four months when first seen, probably because of relatively good cosmesis.
3. no compensatory head posture.
4. a slight preponderance of V pattern (26), although the incidence of A pattern was high (23).
5. poor or no binocular vision in all but seven patients.

It was impossible to prove the early onset in all cases and it is probable that a few patients with early pseudo-strabismus developed a true strabismus later. The history is supported by the poor binocular vision found in 67 patients.

Recently Kommerell (1978)[4] stated that another characteristic of congenital esotropia is asymmetry of optokinetic nystagmus (OKN) when tested uniocularly. When the stripes of a drum or scarf are moved in a temporal to nasal direction a normal response is elicited but when they are moved in a nasal-temporal direction there is no response or only weak irregular saccades.

A similar finding has been reported in normal infants under the age of three to four months by Atkinson (1979).[5] She states that it is likely that symmetry of OKN provides an indicator of the maturation of the binocular cortex in infants and suggests that OKN may be usable as a test for anomalies of binocular development.

Recent papers have suggested that peripheral or even central fusion can exist in patients with strabismus and DVD. Sprague *et al* (1980)[6] stated that peripheral fusion will control the deviation once the horizontal angle is corrected. Helveston (1980)[7] found fusion for near using Worth's four-dot test in 23% of 111 patients, but could demonstrate fusion for distance in only one patient. He reported gross stereoacuity (Titmus fly) in 9% and a weak fusion range on the major amblyoscope in 7%. Cohen and Moore (1980),[8] reporting on primary DVD unassociated with other forms of strabismus, found peripheral fusion in 15 out of 23 cases and central fusion in one patient. In Series 1 we found evidence of superficial abnormal retinal correspondence (ARC) using Bagolini striated glasses in 23% but were unable to substantiate anomalous fusion in any other test. Only 2% of our patients claimed to see the Titmus fly stereoscopically. We did not consider these responses constituted useful peripheral fusion.

AIMS OF PRESENT STUDY

Because of this discrepancy and to assess the significance of the OKN response in relation to binocular function it was decided to investigate OKN and re-evaluate binocular vision in available patients in the two series and in new patients with early onset strabismus.

METHODS

1. *OKN*

Horizontal OKN was first tested with both eyes open using a hand-held drum, moving the stripes

in each direction. OKN was then tested with each eye in turn, moving the stripes first temporal-nasal and then nasal-temporal, repeating the manoeuvre several times. Occasionally it was easier to hold the attention of a young child by using a scarf instead of a drum.

2. *Binocular vision*

Binocular vision was assessed using

(a) Bagolini striated glasses for near and distance.
(b) Lang's 2-pen test.
(c) stereoacuity test, TNO if possible.
(d) 15Δ or 20Δ base out prism to assess motor fusion.
(e) major amblyoscope to diagnose retinal correspondence and fusion.

These tests were chosen because, with the exception of the major amblyoscope, they are simple and non-dissociating.

PRELIMINARY RESULTS

Sixty-nine patients with early onset strabismus have been tested grouped as in Series 2 but including some patients with exotropia. A control group of patients with later onset strabismus has been added and a small number of patients with nystagmus who maintain binocular single vision (BSV) has been assessed, making a total of 99 patients divided into five groups:

Group 1 strabismus + nystagmus + DVD	34
Group 2 strabismus + nystagmus – DVD	17
Group 3 strabismus – nystagmus – DVD	18
Group 4 later onset strabismus	25
Group 5 BSV + nystagmus	5
	99

Results of OKN testing

Group 1

When the stripes were moved temporal-nasal all patients showed a normal response. When the stripes were moved nasal-temporal no patient showed a normal response, either nystagmus was absent or weak intermittent saccades were seen. This occurred whether latent (or manifest latent) nystagmus was obvious or minimal. Latent nystagmus was not demonstrable in the primary position in two patients but one, included in Series 1, was known to have had latent nystagmus in the past. The second patient, aged nine years, remained in the group because nystagmus could not be excluded in the absence of EOG studies. All patients showed a normal response when tested with both eyes open.

Group 2

No clear cut pattern emerged. Five patients showed the same response as those in Group 1, four are still young enough to develop DVD. Four patients with latent nystagmus showed a normal response. Eight patients showed a variety of responses, OKN was reduced in both directions, more reduced on temporal-nasal movement or was, e.g., consistently better on R. gaze with either eye. Further study is needed to determine the exact nature of the nystagmus in each case.

Groups 3 and 4

There was a normal response in all but one patient in each group. Both patients showed a reduced response in a severely amblyopic eye.

Group 5

Normal OKN was elicited in two patients. It could be demonstrated in the remaining three patients when the fast phases of nystagmus and OKN coincided but was reduced in amplitude in the opposite direction. All three were albinos with severely impaired vision.

The youngest patient tested was 18 months of age. Problems in eliciting OKN were encountered if the visual acuity was less than 6/60.

Binocular vision

Group 1

Six patients showed a superficial ARC (Bagolini glasses and a crossing angle on the major amblyoscope) but only three demonstrated a weak fusional response to a prism. No patient had stereopsis. Eight now have consecutive exotropia.

Group 2
Two patients showed superficial ARC, none demonstrated motor fusion or stereopsis. Four have consecutive strabismus.

Group 3
Two patients showed ARC, one demonstrated a weak fusional response and three had some stereopsis. Five have consecutive strabismus.

Group 4
Binocular vision varied from good (60″ stereoacuity) to non-existent.

Group 5
The presence of BSV was proven in all cases. The poor binocular responses and the high incidence of consecutive strabismus do not support the view that there is sufficiently strong peripheral fusion to control the residual deviation in early onset strabismus.

DISCUSSION

Animal studies suggest that temporal-nasal movement can be controlled by a direct pathway from the retina to the pretectum but nasal-temporal movement requires a more advanced pathway via the cortex (Hoffmann 1979).[9] The fact that human infants demonstrate temporal-nasally induced OKN three months before nasal-temporal OKN suggests that two pathways may exist in man. Kommerell (1978, 1982)[4,10] has proposed that OKN remains at an atavistic level in congenital esotropia and that this is a factor in the production of nystagmus. He suggests that these patients can be compared with animals lacking binocular stereopsis, for example the rabbit, which shows the same asymmetry of OKN (Collewijn 1975).[11]

In the rabbit and in other non-binocular animals both nasal and temporal retina are represented in the contralateral cortex. It is now established that up to 20° of temporal retina in albino animals such as the Siamese cat is projected contralaterally (Guillery 1979).[12] VEP studies have provided evidence of a similar pathway abnormality in human albinos (Jay and Carroll 1980).[13] Guillery has stated that the nystagmus and strabismus in albinos can reasonably be ascribed to this defect: it could also account for the asymmetry of OKN. It is tempting therefore to speculate whether a retino-geniculate pathway anomaly could be present in cases of early onset strabismus showing only temporal-nasal OKN. VEP studies have not as yet been carried out on patients in this study but Tsutsui and Fukai (1978)[14] reported on three patients in whom they found evidence of a pathway defect. All had latent nystagmus and "alternating hyperphoria". They found supporting evidence from VEP in one patient. Inverse nystagmus was demonstrated in two out of their three cases but was not observed in our study.

Asymmetrical OKN has been found in both eyes of uniocularly deprived cats by van Hof-van Duin (1976, 1978).[15,16] She suggested that lack of binocularly driven cells accounted for this finding. Our results do not support this view since symmetrical OKN has been seen in many patients lacking binocular vision.

Crone (1977)[17] reported asymmetry of OKN in amblyopic adults but we have found that amblyopia is not the significant factor, both symmetrical and asymmetrical OKN have been seen in different patients with moderately severe amblyopia.

CONCLUSIONS

1. Only patients with the triad of early onset strabismus, nystagmus and DVD consistently demonstrated asymmetry of uniocular OKN.
2. Symmetry of OKN was the rule in patients with early onset strabismus uncomplicated by nystagmus or DVD.
3. No evidence was found to support the value of uniocular OKN testing as a means of assessing binocular vision but it is suggested that it has a place in the detection of nystagmus and DVD in early onset strabismus.

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