NEW ASPECTS OF DISSOCIATED VERTICAL DEVIATION

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Abstract

Dissociated vertical deviation (DVD) is a condition where an eye deviates upwards upon reduction of illumination. The precise characteristics, differential diagnosis and aetiology of this condition however remain very confusing.

Forty-five patients with hyperdeviation under cover were divided into two groups:
(1) those with marked updrift
(2) those with small updrift
and reviewed with special reference to optokinetic nystagmus (OKN) and the visual evoked response (VER) in an effort to clarify thoughts on this condition.

Results indicate that a high percentage of patients with DVD show asymmetry of monocular OKN possibly indicating anomalies of visual fibre projection as evidenced by abnormalities on the VER. OKN may therefore be a useful and simple clinical test to assess the presence of DVD.

Key words: *Optokinetic nystagmus, visual evoked response, Bielschowsky phenomenon.*

INTRODUCTION

The phenomenon of either eye deviating upwards on occlusion was described as early as 1895 by Stevens.[1] Since then an amazing variety of terms have been used to describe this entity including: alternating hyperphoria, double hyperphoria, alternating hypertropia, occlusion hypertropia, anophoria, alternating sursumduction, dissociated vertical divergence and dissociated vertical deviation.

A great deal of the early analysis of this phenomenon was carried out by Bielschowsky[2] who used the term "dissociated vertical deviation", now usually abbreviated to DVD.

Most reports in the literature describe DVD as a slow rotation[3,4] of the occluded eye progressively upwards to a moderate to marked degree,[5] often associated with latent nystagmus and extorsion.[3-13] Prolonged occlusion may be required to elicit the deviation.[5,12,14] Past reports have emphasised that this vertical deviation is not constant but rather, is very variable in nature.[2-4,6,7,9,11,12,14-16] Although DVD may frequently be associated with an overaction of the inferior obliques or underaction of the superior obliques, these ocular muscle imbalances actually have no causal relationship with the DVD.[2,4,5,11,12,16,17] Much confusion still remains however as to the precise characteristics which constitute this phenomenon.

In an effort to clarify our thoughts, it was decided to carry out an investigation of patients with DVD especially with respect to optokinetic nystagmus (OKN), following Mein[13] who stated that these patients show asymmetrical responses on this test. A total of 45 patients were gathered who exhibited presence or increase of a hyperdeviation, either unilateral or bilateral, on dissociation. Some of these patients exhibited marked deviation under cover while in others this

deviation was only small. The patients were therefore divided into two groups:

Group 1 patients showed a significant updrift of an eye on dissociation.

Group 2 patients showed only a slight updrift.

Marlow[18] in his investigation into the use of prolonged occlusion (24 hours to one week) as a diagnostic tool for heterophoria found, in the normal population, a high incidence of hyperdeviation of the covered eye. In view of this it was queried as to whether those patients in Group 2 actually showed DVD or only a latent vertical component after prolonged occlusion.

Therefore in order to determine if these two groups were actually demonstrating the same ocular condition, the characteristics of these patients were examined more closely with several features in mind:

Sex Distribution

This was of interest as it showed twice as many females as males (30: 15). Only two other reports on DVD were found in the literature with an analysis of sex distribution: Sprague *et al.*[3] and MacLellan.[5] This tendency for a preponderance of females was also found, to an even more marked degree, the proportions being 75% female: 25% male. This possibly suggests that DVD is a condition which is more likely to be found affecting females than males.

Type of Deviation

It is well documented that DVD most frequently occurs in the presence of a constant convergent deviation of congenital origin.[3-5,8-13,16] Evidence from this series agrees with this statement: 31 of 45 patients having an esotropia, the majority of which had an onset of less than six months of age. It is of interest to note however, that in this series, eight patients showed only an intermittent deviation.

Visual Acuity

Bielschowsky[2] and Jones[14] have stated that the anomalous vertical movements as found in DVD may also be seen in markedly amblyopic eyes. Visual acuity in this series did not appear to be significant. Only two eyes showed marked amblyopia of less than 6/60, the majority of eyes having vision of 6/12 or better (74 of 94 eyes).

Binocular Vision

As previously stated, DVD is most commonly found in association with a constant squint, usually of longstanding, and consequently it is not surprising that most patients with DVD are found to have poor or no binocular function. This was the case in this series: no patient demonstrated good binocular single vision (BSV); fair BSV was found in only eight patients, the remaining 37 demonstrating a poor level or total absence.

Nystagmus

It is well documented that there is an association between DVD and nystagmus.[5,7-9,13] In fact Anderson[7] found that in his series of patients, every case of true latent nystagmus with one exception, showed a varying degree of "alternating hyperphoria". Mein[9] has postulated that DVD may be a further manifestation of the Nystagmus Blockage Syndrome. Latent nystagmus was present in only 53% of cases in this series however, if nystagmus is fine it may only be demonstrated on visuscopy and not all patients in this series had this test performed.

Ocular Movements

Mein[9,13] in her series of patients with DVD has found a high incidence of A pattern. This phenomenon has also been described in other reports.[4,16,17] MacLellan[5] however, found that the incidence of A, V, or X pattern in her series was about equally distributed. In this present series overall, there was a slight preponderance of V pattern (17V: 11A). If the two groups are examined separately, this preponderance of V pattern is found to be more marked in Group 2 (Group 1 — 10V: 7A; Group 2 — 7V: 4A).

Bielschowsky Phenomenon

This phenomenon, first described by Bielschowsky,[2] states that when the illumination of the fixing eye is sufficiently reduced, the upward deviated eye under cover will show an

isolated downward movement which will bring it down to or even below the midline as the fixing eye remains in the primary position. The Bielschowsky phenomenon may be used as a diagnostic test to differentiate between a true DVD and a basic hyperphoria, the phenomenon not being demonstrated in the latter.

It has been stated however, that this phenomenon can only be demonstrated in about 50% of those patients with DVD.[11] As such it would not seem a reliable test to use for differential diagnosis.

It is interesting to compare the results of this test as found in Group1 and 2 of this series. Of the eyes suspected of having a DVD and that were tested for the Bielschowsky phenomenon, in Group 1 69% (11 of 16 eyes) showed a positive response, whereas in Group 2 54% (6 of 11 eyes) showed a positive response.

Optokinetic Nystagmus (OKN)

Researchers[19-21] have noted asymmetry of monocular OKN in animals and humans where there is a lack of binocularity, for example, in congenital esotropia. This asymmetry is found when, with movement of stripes in a temporal to nasal direction, a normal OKN response is elicited however, with stripe movement in a nasal to temporal direction the response is either very erratic or totally absent.

Mein[13] in her investigation of monocular OKN in congenital esotropia found that only those patients with DVD showed an asymmetrical response. She therefore suggested that OKN might be a useful diagnostic tool in the detection of DVD in early onset strabismus.

With this in mind, the patients in this series were examined to determine if the two groups could be differentiated. Of those eyes suspected of having DVD that were tested for OKN responses, in Group 1 97% (37 of 38 eyes) showed asymmetry with poor nasal to temporal response whereas in Group 2 91% (10 of 11 eyes) showed this asymmetry.

Visual Evoked Response

Mein[13] and Fitzgerald[22] have suggested that this asymmetry of monocular OKN may occur as a result of an abnormality in the pathway of visual fibres where some of the fibres from the temporal retina actually decussate at the chiasm and are therefore represented in the contralateral visual cortex. This may be detected by the use of the visual evoked response (VER).

Of the eyes in this series suspected of having DVD that were tested for their VER, in Group 1 81% (22 of 27 eyes) showed abnormal VER indicating anomalous projection of the temporal retinal fibres whereas in Group 2 83% (five of six eyes) showed this abnormality.

DISCUSSION

The testing of the Bielschowsky phenomenon, OKN and VER was found to be of particular interest in these patients as these are put forward as differential diagnostic tests to distinguish between true DVD and a basic hyperphoria such as may be found in a primary overaction of the inferior oblique muscle.

The Bielschowsky phenomenon is known to be an inconclusive test even in the presence of true DVD. Responses to this test in this series could indicate that those patients in Group 2 are less likely to be true DVD. Conversely, it may only indicate that a positive response is more likely to occur in the presence of the more marked cases of updrift and therefore its value as a differential diagnostic test could still be questioned.

Testing for abnormalities of responses with OKN and/or VER would appear to be a much more reliable indicator as to the presence or absence of true DVD.

Clearly there has been confusion in the past as to the precise characteristics which constitute DVD. No doubt it is this confusion that has led to the plethora of terms that have arisen.

The confusion over this phenomenon has also led to the great variety of attributed aetiological factors. Although previously cited as an aetiological factor,[10,23] simple underaction of the superior obliques and corresponding overaction of the inferior obliques is now well accepted as not being the cause of DVD.[2,4,5,11,12,16,17] In fact, it has been documented that DVD may occur in the presence of overaction of the depressor

muscles, for example, the superior obliques.[12,16,17]

It is well known for there to be a disturbance of binocularity in patients with DVD but, which of these two comes first? Verhoeff[10] suggested that there may be arrested development or abnormality of the monocular conjugate mechanism of eye control such that certain movements of each eye then become independent. Posner[24] suggested that with weak binocular linkage there is apparent dissociation between the two eyes. This then gives rise to an aberration of the tonus-regulating mechanism of the eye muscles such that the eye favours the position of rest — elevation. Bielschowsky[2] suggested that these anomalous eye movements are caused by intermittent excitations of the "subcortical vertical divergence centres" and may become manifest when there is a lack of homogeneous retinal stimulation, for example, when there is reduction of the illumination to one eye. It is currently accepted that the aetiology is an innervational cause of supranuclear origin.[4]

This series suggests that these patients have some abnormality of visual fibre projection to the higher centres of eye movement control. This may then predispose these patients to squint, particularly of congenital origin, and also to the ensuing phenomenon of DVD.

CONCLUSION

In conclusion, it would appear after overall assessment that the majority of patients in both Group 1 and Group 2 do have a true DVD but to differing degrees.

The testing of optokinetic nystagmus and the visual evoked response are put forward as being useful diagnostic procedures in the differentiation of patients with this entity and maybe also as a method of predicting which of those patients with congenital esotropia will, at a later time, go on to develop this phenomenon of dissociated vertical deviation.

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