

EVIDENCE OF ABNORMAL OPTIC NERVE FIBRE PROJECTIONS IN PATIENTS WITH DISSOCIATED VERTICAL DEVIATION (DVD) — A PRELIMINARY REPORT*

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Abstract

Abnormal optic nerve pathways can be demonstrated by testing half field checkerboard VER's. This method has been used to detect abnormal optic nerve fibre projections in human albinos, a group who also have anomalous nystagmoid movements.

Patients with DVD have recently been observed to have abnormal nasotemporal OKN (Mein 1982). The hypothesis that these patients also have abnormal visual pathway projections was tested.

Patients with DVD (both unilateral and alternating), congenital esotropes without DVD and normal subjects were investigated. Results showed a very high incidence of abnormal VER recordings in patients with DVD while recordings from congenital esotropes and the control group were normal. As well as demonstrating abnormal optic nerve fibre pathway projections, the VER's showed a large increase in latency in all patients with DVD which was not present in the other two groups studied. These results occurred regardless of visual acuity or amblyopia in the eyes tested.

The high correlation between VER abnormalities and naso-temporal OKN anomalies indicates the importance of the latter in diagnosing DVD.

The possible significance of abnormal pathways in the presence of a DVD is discussed.

Key words: VER, albinos, abnormal projection, OKN.

Dissociated vertical deviation (DVD) was first described by Stevens in 1896. Bielschowsky (1932) was the first to give detailed observations of this disturbance.

Recently Mein and Johnson¹ have summarised the principal characteristics as:

1. Elevation of either eye when the amount of light entering the eye is reduced by occlusion or other means.
2. Extorsion as the eye elevates.
3. Latent nystagmus.

The aim of this paper is to show that patients with DVD have abnormal optic nerve fibre pathway projections. This was confirmed using visually evoked responses (VER's).

The importance of this finding to orthoptists is that patients with this abnormality also have

abnormal opto-kinetic nystagmus (OKN), the latter being an easily demonstrated clinical manifestation of the abnormality in retinal projections.

The VER is primarily used in the diagnosis of conditions affecting the optic nerve and its pathways. Half field VER's may be used to determine the relative contributions of optic nerve projections to the visual cortex. Neither nystagmus nor amblyopia affect the accuracy of recording these projections.

When stimulating temporal retinal fibres in the right eye the normal response (i.e. the response similar to that recorded at the central occipital electrode) is recorded from the right visual cortex (see Fig. 1). As there is virtually no input going to the left visual cortex the response from this

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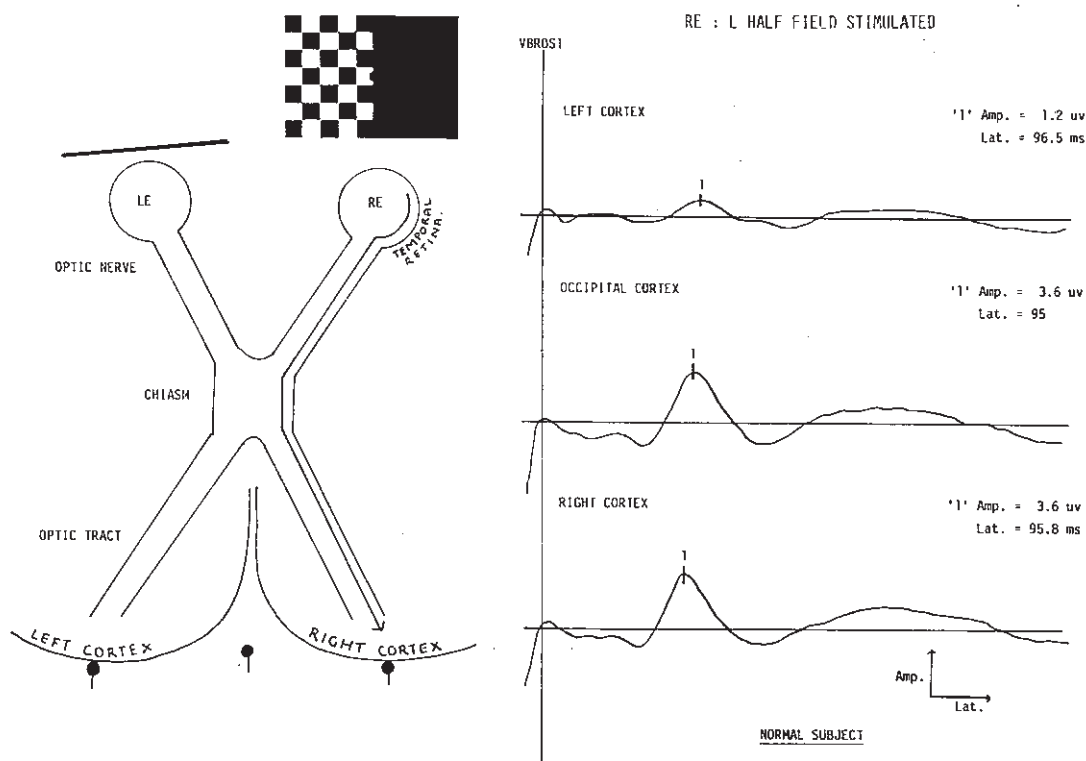


Figure 1: VER tracings recorded from temporal retinal stimulation in the right eye.

electrode should be very reduced. This is also clearly demonstrated in Fig. 1.

This method has been used to study the abnormal decussation of temporal retinal fibres² that has been found in 70% of human albinos.³ Thus, any marked abnormality in VER's recorded from patients with DVD would suggest an anomaly of decussation of optic nerve fibre pathways.

In normal subjects OKN is a jerk nystagmus with a slow phase in the direction of drum rotation and the fast phase in the opposite direction.

This paper reports on the VER and OKN findings of 20 patients with DVD, 13 normal subjects and six congenital esotropes ranging in age from four to 47 years.

METHOD

Three groups of patients were examined:

Group 1 consisted of 13 normal subjects. Each of these patients were ophthalmologically normal

with corrected visual acuity of 6/6 or better. Only the right eye was tested.

Group 2 consisted of 20 DVD patients. Eight had unilateral DVD and 12 had alternating DVD.

Group 3 consisted of six patients with congenital esotropia (i.e. a positive history of strabismus before six months of age). None of these patients had any demonstrable DVD.

All patients were given a full orthoptic examination independently by two orthoptists.

VER's were tested monocularly using a 16° half field check screen. The subjects were instructed to fix on the central spot all the time to ensure only hemi-retinal stimulation. Thus, when the right half of the screen is visible (when testing the right eye) only nasal retinal fibres receive stimulation (see Fig. 2A).

Three active electrodes were used to detect optic nerve projections. The occipital electrode (see Fig. 2A) mainly detects responses from the macular area. In hemi-retinal stimulation this

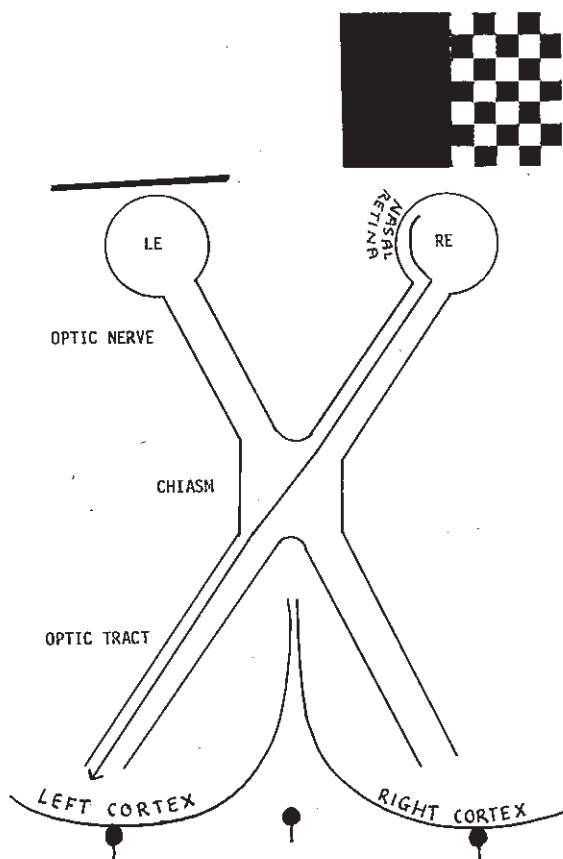


Figure 2A: Hemi-retinal checkerboard stimulation.

response is normal. The two other electrodes were placed 5 cms laterally on either side of the occipital electrode. Recordings were taken individually from lateral electrodes thus detecting both nasal and temporal halves of the retina. The reference electrode was placed on the forehead and an earth electrode was placed on the ear.

OKN was assessed monocularly using the opto-kinetic drum at 1/3 m. Assessments were made rotating the drum both naso-temporally and temporo-nasally (see Fig. 2B).

RESULTS

Group 1: Normal Subjects

Normal subjects showed the characteristic VER recordings expected from hemi-retinal stimulation (see Fig. 3). When stimulating nasal retinal fibres in the right eye the normal response (which

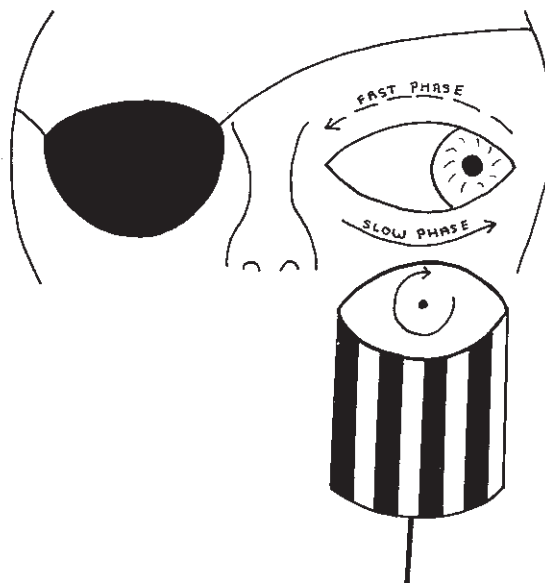


Figure 2B: Unilateral naso-temporal OKN.

was similar to the response recorded at the occipital electrode) was recorded from the left visual cortex (see Fig. 3). As there was virtually no input to the right visual cortex, the response recorded from this cortex was very reduced. This was clearly demonstrated in the VER tracing.

The latency of point "1", or the time taken for the impulse to travel from the retina to the visual cortex in all these patients was normal (approx. 96 ms). Amplitude recorded from the occipital electrode (3.6 uv) and the stimulated hemi-retina (3.6 uv) was also normal.

No abnormality in OKN was detected with naso-temporal or temporo-nasal drum rotation in normal subjects.

Group 2: DVD Patients

Seventy-nine per cent of the patients in the DVD group showed abnormalities in the VER tracings recorded from temporal retinal optic nerve fibres. In Fig. 4, recordings taken from temporal retinal stimulation of the right eye show that the normal response (i.e. the response most similar to the recording from the occipital electrode) was recorded from the left visual cortex. However, with temporal retinal stimulation (RE) there should be virtually no input to the left visual

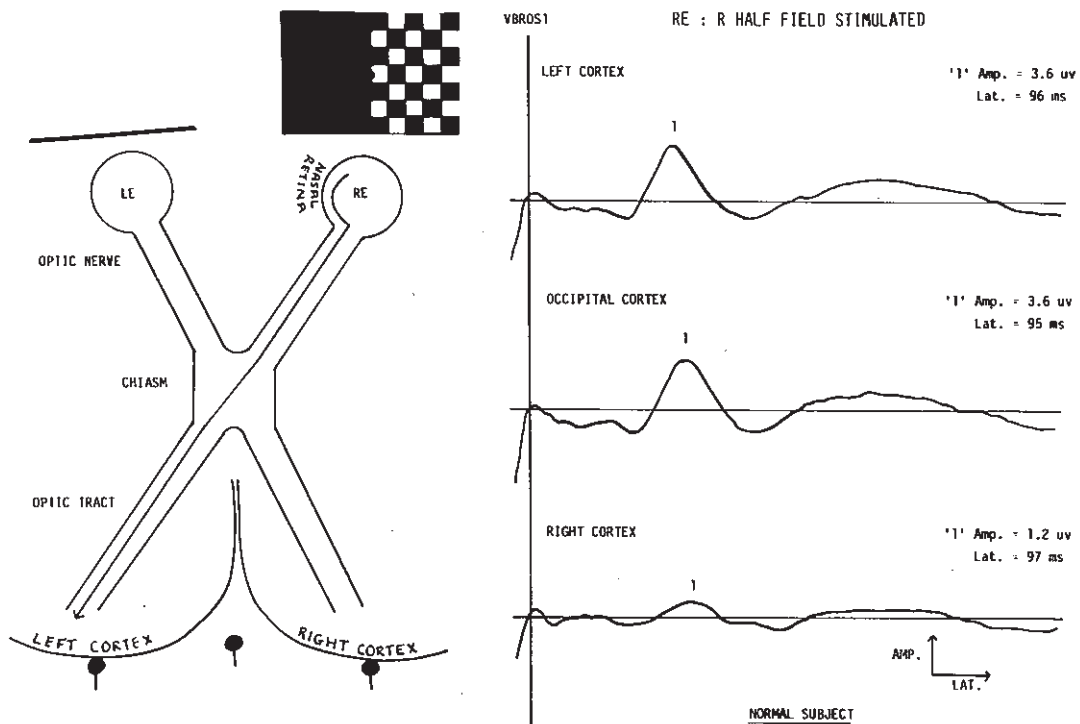


Figure 3: VER tracing from normal subject (nasal retinal stimulation).

cortex. Conversely, the right cortex should be receiving a normal input but the response recorded was greatly reduced.

Abnormal responses were recorded from nasal retinal stimulation in 37% of the patients with DVD.

In the patients with unilateral DVD temporal retinal stimulation of the eye without the DVD was abnormal in only 25% of the cases.

The latency of the point "1" was increased in all patients with DVD. This increase was most marked in the tracing from the abnormal temporal retinal fibres.

OKN was abnormal when the drum was rotated naso-temporally in 82% of the DVD patients while temporo-nasal rotation of the drum produced OKN anomalies in 12%.

Group 3: Congenital Esotropes

Eight per cent of the congenital esotropes in the study showed abnormal temporal or nasal retinal optic nerve fibre projection. There was a slight increase in latency of point "1" in all these patients.

OKN was normal in all but one eye of one patient.

DISCUSSION

The abnormal VER's recorded in patients with DVD constitute evidence of abnormal decussation of temporal retinal fibres at the chiasm. This is so because (as is seen in Fig. 5) the most normal response was recorded from the left cortex when only temporal retinal fibres were stimulated in the right eye, so some of the stimulated temporal fibres may cross over at the chiasm and go to the left cortex. Also, the decreased response at the right visual cortex is indicative of a decreased input to this area which can only be explained by misdirection of the fibres.

Several different studies conducted on human albinos have shown that optic nerve fibres originating in the temporal retina actually decussate at the chiasm in 70% of cases.³ This abnormal decussation produces characteristic anomalies in VER's similar to those found in 79% of the DVD eyes in this study. No sig-

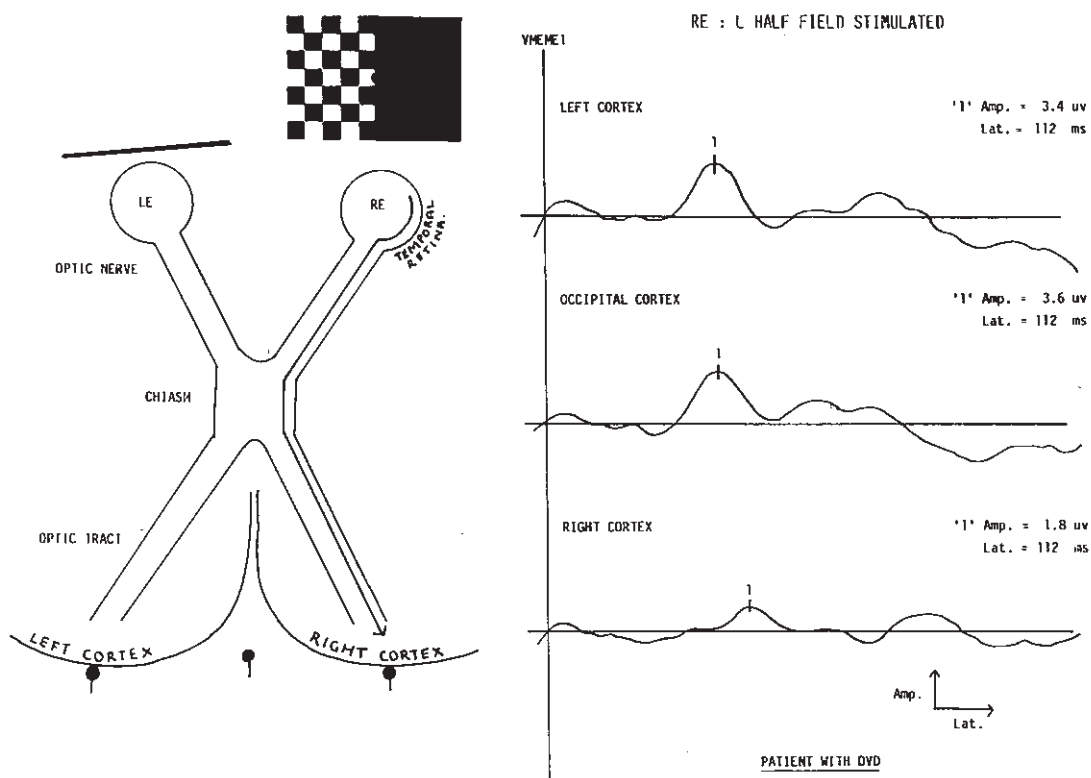


Figure 4: DVD patient: right eye temporal retinal stimulation.

nificant abnormality in nasal retinal projection pathways in albinos have been reported in the literature, however, in this study 37% of the DVD patients also show anomalies in the nasal retinal fibre projection.

In the unilateral DVD sub group 75% of the patients had normal VER tracings in the non DVD eye and anomalous recordings in the DVD eye, indicating that DVD does indeed occur unilaterally in some cases. The other 25% of the subgroup demonstrated temporal retinal fibre projection anomalies in both eyes. Thus, they

may have a bilateral DVD which is more marked in one eye.

Latency, or the time taken for the response to reach the visual cortex was markedly increased in patients with DVD. The misdirection of the optic nerve fibres at the chiasm could cause interference to the normal conduction along the nerve thus it takes longer for the response to reach the cortex.

Ninety-two per cent of the congenital esotropes in the study showed no abnormalities in VER's. This finding is in keeping with a study

TABLE 1: Summary of Results

Patient group	Abnormal VER tracings with		Abnormal OKN (N→T drum rotation)
	Temporal retina fibre stimulation	Nasal retina fibre stimulation	
Normals	0	0	0
Congenital esotropes	8%	8%	8%
DVD	79%	37%	82%

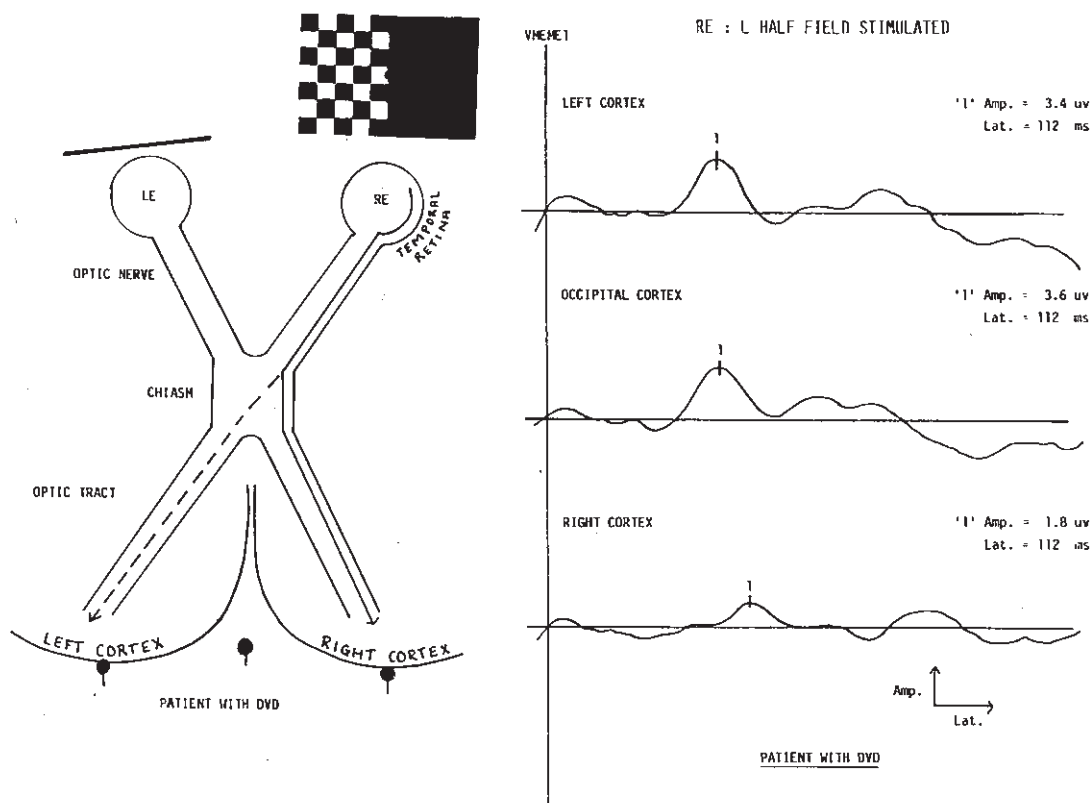


Figure 5: Abnormal decussation of temporal retinal fibres in a patient with DVD (re temporal fibres stimulated).

by McCormack⁴ who studied normally pigmented squinters (without DVD) and concluded that they had no visual pathway anomaly. The one patient that had abnormal VER's in this group may go on to develop a DVD at a later age.

The abnormal OKN which occurred in 82% of the DVD patients in this study is a significant finding. These results support those in a study of Mein⁵ who reported that all patients with DVD had abnormal OKN with naso-temporal drum rotation whereas congenital esotropes who did not have DVD (and were "too old to develop DVD") had normal OKN. Thus OKN is shown to be a useful clinical test in the diagnosis of DVD.

In order to have normal BSV there must be normal input from corresponding retinal layers to the lateral geniculate nucleus (LGN). Only

three out of the 20 DVD patients in the study had any demonstrable BV and none had BSV. The abnormal fibre projections in DVD patients would disrupt the ordered structure of the LGN hence these patients would not have BSV.

CONCLUSIONS

This study demonstrates evidence of anomalous crossing of temporal retinal fibres in patients with DVD together with evidence of some anomalous projection of nasal retinal fibres.

The results have demonstrated that misdirection of optic nerve fibres may vary between the two eyes and that in apparently unilateral cases of DVD the VER may be abnormal in only one eye, supporting the premise that DVD is not necessarily a bilateral condition.

There is a high correlation between anomalous naso-temporal OKN and abnormalities in VER

recordings. The VER recordings have shown that abnormal retinal projections occur in these patients. Therefore, an abnormal naso-temporal OKN response in the absence of a demonstrable DVD may anticipate the later development of this defect.

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