DISSOCIATED VERTICAL DEVIATION: AN EXAGGERATED NORMAL EYE MOVEMENT USED TO DAMP CYCLOVERTICAL LATENT NYSTAGMUS*

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ABSTRACT

Purpose: Dissociated vertical deviation (DVD) has eluded explanation for more than a century. The purpose of this study has been to elucidate the etiology and mechanism of DVD.

Methods: Eye movement recordings of six young adults with DVD were made with dual-coil scleral search coils under various conditions of fixation, illumination, and head tilt. Horizontal, vertical, and torsional eye movements were recorded for both eyes simultaneously. Analyses of the simultaneous vertical and torsional movements occurring during the DVD response were used to separate and identify the component vergence and version eye movements involved.

Results: Typically, both horizontal and cyclovertical latent nystagmus developed upon occlusion of either eye. A cycloversion then occurred, with the fixing eye intorting and tending to depress, the covered eye extorting and elevating. Simultaneously, upward versions occurred for the maintenance of fixation, consisting variously of saccades and smooth eye movements, leading to further elevation of the eye behind the cover. The cyclovertical component of the latent nystagmus became partially damped as the DVD developed.

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Conclusions: In patients with an early-onset defect of binocular function, the occlusion of one eye, or even concentration on fixing with one eye, produces unbalanced input to the vestibular system. This results in latent nystagmus, sometimes seen only with magnification. The cyclovertical component of the latent nystagmus, when present, is similar to normal vestibular nystagmus induced by dynamic head tilting about an oblique axis. Such vestibular nystagmus characteristically produces a hyperdeviation of the eyes. In the case of cyclovertical latent nystagmus, the analogous hyperdeviation will persist unless corrected by a vertical vergence. A normal, oblique-muscle-mediated, cycloversion/vertical vergence is called into play. This occurs in the proper direction to correct the hyperdeviation, but it occurs in an exaggerated form in the absence of binocular vision, probably as a learned response. The cycloversion/vertical vergence helps damp the cyclovertical nystagmus (a cyclovertical "nystagmus blockage" phenomenon), aiding vision in the fixing eye. But this mechanism also produces unavoidable and undesirable elevation and extorsion of the fellow eye, which we call DVD.

INTRODUCTION

The ocular motor phenomenon now known as dissociated vertical deviation (DVD) has eluded explanation since it was described by George Stevens in 1895.[1] He attributed "alternating vertical strabismus" to an error in declination (torsion) of the fixing eye.[2] Others have postulated vertical rectus muscle imbalance,[3] oblique muscle imbalance,[4,5] primitive vestibular reflexes,[6] or abnormal vertical motion processing[7] as the cause of this strange vertical strabismus that appears to violate Hering's law of equal innervation of yoke muscles.[8]

DVD is known to be strongly associated with early strabismus and with manifest latent and latent nystagmus,[9-11] especially latent nystagmus with a torsional component.[4,10,12,13] Many patients with DVD have anomalous head postures.[6,12,14-17] The torsional and horizontal movements that often accompany DVD, or sometimes predominate, have come to be known as "dissociated torsional deviation" and "dissociated horizontal deviation," respectively,[18,19] with the entire phenomenon referred to as the "dissociated strabismus complex."[20] But the ocular motor mechanism underlying DVD and its torsional and horizontal components is yet to be explained.

STUMBLING BLOCKS TO UNDERSTANDING

In the course of our investigation of DVD, we have identified several common teachings that have represented stumbling blocks to understanding. The first stumbling block has been the teaching that "the fixing eye does

not move when the other eye is covered." Using video recordings and scleral search coil recordings, we discovered early in our investigation that the fixing eye usually intorts when the other eye is covered, consistent with old observations buried in the literature.[12,21]

The second stumbling block has been the teaching that "DVD violates Hering's law of equal innervation of yoke muscles." Through a careful analysis of scleral search coil recordings, we have been able to identify the vergence and version components of DVD, each of which obeys Hering's law upon proper identification of the yoke muscles involved.

The third stumbling block has been the teaching that "DVD is brought about by inattention." While this may be true in many cases, there are clearly patients who develop manifest DVD when reading smaller and smaller letters on a visual acuity chart in the distance,[22] as if the development of the DVD enables better visual acuity in the fixing eye. We have observed this phenomenon especially in those DVD patients who have peripheral motor fusion under binocular conditions. As smaller and smaller letters are read, the fusion breaks, presumably because of more concentration with the fixing eye, with deeper suppression of the vision in the nonfixing eye, and DVD develops. An alternative explanation is that attempts to read small letters lead to increased manifest latent nystagmus from stress or anxiety,[23] and DVD develops to help damp the nystagmus (J. Pratt-Johnson, personal communication, 1998). The fact that active monocular fixation is linked to the appearance of DVD was pointed out as early as 1944 by Posner,[24] and elaborated upon in recent years by Spielmann.[25] Spielmann has nicely demonstrated that DVD disappears in the absence of fixation, either with bilateral occlusion or in the dark. The Bielschowsky phenomenon is another manifestation of the disappearance of DVD when active fixation is prevented, either by a dark filter before the fixing eye,[26] by a diffuser before the fixing eye,[21] or by an illuminated occluder before the fixing eye.[12]

The final stumbling block to understanding has been the teaching that DVD serves no obvious purpose. In 1938, Marlow[27] wrote, "Divergence from parallelism is quite purposeless. Any useful purpose served by it is unthinkable." Further review of the literature suggests otherwise. In 1954, Anderson[4] noted that manifest "rotary" nystagmus in patients with DVD often disappeared when one eye was covered. In 1981 Mein and Johnson[14] proposed that DVD is one of the compensatory mechanisms used to damp latent nystagmus. Scott McClatchey independently proposed this in a personal communication to one of us (D.L.G.) in 1992. Upon being alerted to this possibility, we began to notice various amounts of manifest latent and latent nystagmus in our patients with DVD. The nystagmus, both horizontal and torsional, often intensified immediately when one eye was covered, but tended to become damped as the DVD

movement fully developed. We documented this phenomenon with video recordings and then used scleral search coil recordings to analyze in detail the complex eye movements involved.

BACKGROUND

A major key to the understanding of DVD was the report by Enright[28] in 1992 that disparity-induced vertical vergence in humans is associated with binocular torsion. Using video oculography, Enright recorded the horizontal, vertical, and torsional eye movements induced when small vertical disparities in the range of 1.5 to 3 prism diopters were introduced by added prism or by a haploscopic method. The vertical fusional vergence that occurred was accompanied by a cycloversion, with the downward moving eye intorting and the upward-moving eye extorting. By careful analysis of the direction and magnitudes of these movements, he concluded that the oblique muscles in both eyes are largely responsible for vertical vergence movements induced by small vertical disparities. Van Rijn and Collewijn[29] confirmed Enright's findings regarding the relative directions of vertical and torsional movements with scleral search coil recordings, and in 1997 showed that the vergence movement occurring in DVD is similar to that seen in normal eyes when fusing small vertical disparities.[30] In 1993, Inoue and Kita[31] had observed intorsion of the fixing eye in patients with DVD using a bilateral fundus video haploscope. The covered eye elevated and extorted.

We also confirmed Enright's findings by using an afterimage technique.[32] Two parallel streak afterimages were created by using a taped-off electronic flash on a rotatable mount. Black-and-white text was placed surrounding the streak apertures, and a fixation point was placed midway between the two streaks. The normal subject fused the background text from 40 cm away, and a 3 or 4 prism diopter vertical prism was introduced. As soon as the subject could fuse the vertical disparity, the flash was fired, the prism was removed, and the subject re-fused the background letters surrounding the flash aperture. The afterimages usually appeared rotated, and the subject manually rotated the flash aperture to be parallel with the afterimages, thus enabling measurement, with a vernier protractor, of the amount of torsion that the eyes experienced upon recovery of vertical fusion. (Monocular afterimages, created using polarizing filters, behaved the same as the binocular afterimages.) In 93% of 55 trials, the direction of torsional movement was consistent with oblique muscle predominance in producing the vertical fusional vergence, with the mean (±SD) torsional shift being 1.15° (±0.76°) in response to the 3 or 4 prism diopters of vertical disparity fused.

To determine whether a given vertical movement of an eye is mediated

primarily by an oblique muscle or by a vertical rectus muscle, the previously mentioned studies have relied on detection of the direction of torsional movement occurring simultaneously with the vertical movement. For example, if an eye is moving downward and is simultaneously intorting, the superior oblique muscle must be playing a predominant role. If the eye is moving downward and is simultaneously extorting, the inferior rectus muscle must be playing a predominant role. This is not to say that the entire vertical movement is necessarily due to the identified muscle (although Enright's calculations[28] led him to claim that the superior oblique muscle is almost uniquely involved in vertical fusional vergence). The direct antagonist muscle generally relaxes, and the other cyclovertical muscles may also play a role, but the laws of mechanics dictate that the cyclovertical muscle identified from the simultaneous vertical and torsional directions of movement must at least be playing a significant role. As will be seen in the present study, this simultaneous vertical/torsional direction analysis is the key to understanding the component movements in DVD.

Another recent study is relevant to the eye movements occurring in DVD. In 1996, Jáuregui-Renaud and colleagues[33] reported that ocular counter-rolling in normal humans, induced by dynamic head tilting (actually whole-body tilting in the dark, either in the upright or supine position, stimulating the vertical semicircular canals), was accompanied by the development of a small vertical deviation. The extorting eye moved downward, and the intorting eye moved relatively upward. The counter-rolling (cycloversion) and vertical divergence were partly compensated by corrective cyclovertical saccades, with asymmetric vertical components, during the dynamic whole-body roll motion. If the effect of the corrective saccades was subtracted out, the cumulative vertical divergence with tilt to each side was about 2.9° in the vertical orientation, and about 2.2° in the supine orientation.

Of particular note is that when we performed simultaneous vertical/torsional direction analysis on their published tracings, with particular attention to the corrective saccades, it appeared that the cyclovertical movements of the intorting eye, during the dynamic roll motion, were being at least partly controlled by the oblique muscles. (In 1984, Kushner and colleagues[34] had noted that the torsional nystagmus seen on active head tilting is decreased or eliminated in the affected eye by superior oblique paresis or by surgical weakening of the inferior oblique muscle.) Simultaneous vertical/torsional direction analysis further showed that the cyclovertical movements of the extorting eye were being largely controlled by the vertical rectus muscles.

The vertical divergence produced with whole-body roll motion was a relative hypodeviation of the extorting eye. This might be predicted because of the more effective vertical action of the vertical rectus muscles

in the extorting eye than of the oblique muscles in the intorting eye. Further studies by this group[35] have shown that whole-body roll motion about an oblique axis produces a form of vestibular nystagmus that is practically identical to the cyclovertical component of latent nystagmus. The accompanying vertical divergence is even more pronounced when the roll axis is oblique.

Jáuregui-Renaud and colleagues performed most of their experiments in the dark and did not identify a corrective vergence that might be assumed to occur during dynamic head tilting in ordinary lighted surroundings to compensate for the induced vertical divergence. They postulated, in fact, that the apparent vertical divergence might be an artifact of their measurement technique and that true vertical alignment might be maintained after all. Evidence for a true compensating vergence is present in their recordings, however, through asymmetry of the corrective vertical saccades. Such asymmetric vertical saccades contain a vergence component[36] in the proper direction to compensate partially for the induced vertical divergence.

If one can speculate that the pattern of cycloversion/vertical vergence that Enright described[28] normally occurs during dynamic head tilting to help maintain vertical fusion, the cycloversion/vertical vergence would involve further intorsion of the intorting eye, with accompanying depression, and further extorsion of the extorting eye, with accompanying elevation. As will be seen from the present study of patients with DVD, this normal cycloversion/vertical vergence, which appears to accompany dynamic head tilting in normal subjects, appears to be the primary vergence mechanism involved in the manifestation of DVD, often in a grossly exaggerated form.

SUBJECTS

Six adult patients with DVD were recruited for scleral search coil recordings. All were women, ranging in age from 18 to 43 years. All gave a history of esotropia in infancy. Five of the patients had undergone eye muscle surgery, with two patients' surgeries involving the vertical rectus muscles as well as the horizontal rectus muscles. The patient who showed the greatest torsional eye movements when occluded, chronologic patient 4, was the one patient who had not had previous surgery. Four of the six patients had peripheral fusion, with one of these having motor fusion only and the other three having 3,000 seconds of arc stereopsis on the stereo fly test. Three of the patients preferred the right eye for fixation, and three preferred the left eye. Visual acuities ranged from 20/15 to 20/25+ in the preferred eye and from 20/20 to 20/40 in the nonpreferred eye in five of the patients and 20/100 in the sixth patient. When dissociated, three of the

patients developed an esodeviation along with typical elevation of the occluded eye. One of the patients developed an exodeviation with the DVD, and two others showed no horizontal deviation with the DVD. Although the horizontal and vertical deviations generally changed on head tilt to either side, none of these patients had an anomalous head posture. Small "A" patterns were noted in three of the patients, and 2 to 3+ intorsion of the fundi[37] was noted in two of the four patients in whom fundus torsion was specifically assessed.

METHODS

Informed consent for the experimental recordings was obtained after the nature and possible consequences of the study were explained. The research protocol had been approved by the institutional human experimentation committee, and the procedures used followed the tenets of the Declaration of Helsinki. Horizontal, vertical, and torsional eye movements for both eyes simultaneously were recorded in Dr Zee's ocular motor and vestibular testing laboratory using dual-coil scleral search coils[38,39] obtained from Skalar Medical in Delft, The Netherlands. Analog signals were filtered to remove high-frequency noise above 90 Hz and then sampled and saved by a computer at 500 Hz with 12-bit resolution. The 3-dimensional eye positions were expressed as rotation vectors[40] and converted to degrees. Tracings were analyzed using custom Matlab programs.

Zero reference positions were determined for horizontal, vertical, and torsional recordings by occluding each eye in turn and having the other, viewing eye fix on an LED target straight ahead at a distance of 1.25 m. Accurate horizontal and vertical reference positions can be obtained in this way, but the absolute torsional position of each eye is unknown. As will be seen, monocular fixation by either eye in patients with DVD induces intorsion in that eye, and under binocular conditions the torsional positions of the two eyes vary depending on how much vertical phoria is being fused. Attempts were made to determine reference torsional positions of the eyes with the patient trying to look straight ahead in the dark or into illuminated empty space (a uniform white diffuser held before the eyes). Without control of the horizontal or vertical positions of the eyes, however, such attempts to obtain absolute torsional positions for the eyes cannot be considered accurate. The absolute amount of intorsion or extorsion can therefore not be determined from the recordings, but changes in torsion from one response to the next can be analyzed readily.

RESULTS

All six patients showed similar DVD movements, both clinically and on the

recordings, with the exception of chronologic patient 1, who had an upward drift to her latent nystagmus, which will be discussed later. A typical segment of recorded eye movements, from patient 5, is shown in Fig 1. Eye movement directions are from the patient's perspective looking forward. An upward deflection on the recording signifies an upward movement for the vertical movement recordings, a rightward movement for the horizontal movement recordings, and a clockwise (dextrocyclo) movement for the torsional movement recordings.

In Fig 1, patient 5 initially has both eyes open, peripherally fusing. A high-frequency cycloversional nystagmus (RET and LET) is present. The left eye is exactly aligned with the target, both horizontally (LEH) and vertically (LEV), except for a small amount of manifest latent nystagmus. The right eye is approximately 1° esodeviated (REH) and 1° hypodeviated (REV). (In the dark, this patient typically developed an 8° left hyperdeviation, which she was easily able to fuse under binocular conditions in the light. As explained by Enright's oblique muscle mechanism for vertical fusional vergence, both eyes are dextrocyclorotated under fusing conditions [RET and LET are above the baseline], bringing the right eye up and the left eye down in order to fuse the vertical phoria.)

Continuing with the recordings in Fig 1, the left eye is suddenly occluded, requiring the right eye (REH and REV) to take up fixation. Approximately 200 ms after occlusion (the exact moment of occlusion was not recorded in these tracings), the right eye begins to move downward (REV), adducts (REH), and intorts (RET). These initial movements rep-

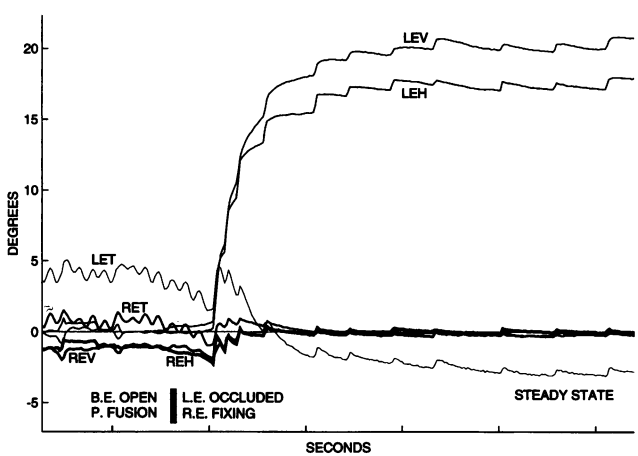

FIGURE 1
Typical eye movement recordings, horizontal (REH and LEH), vertical (REV and LEV), and torsional (RET and LET), when left eye of patient 5 is occluded and DVD develops.

resent the disruption of peripheral fusion and happen to be, in this instance, in the same direction as the slow-phase drifts of the latent nystagmus induced by occluding the left eye. The downward, inward, and intorting drift of the right eye takes the right eye away from fixation on the LED target, and a series of upward saccades occur, representing the fast phases of the latent nystagmus, eventually bringing the right eye upward and outward to fix on the target. Simultaneously, yoked upward saccades occur in the left eye (LEV), very much larger than occur in the right eye (REV), representing a vergence combined with the saccades.[36,41,42] The combination of the vertical vergence and vertical saccades results in a left hyperdeviation (LEV) of 20°. This is accompanied by a left esodeviation (LEH) of about 17°. The large nonconjugate saccades (saccades combined with a vergence) are complete in approximately 1 second, with baseline latent nystagmus continuing in the steady-state, left-eye-occluded condition. Note that the amplitude of this latent nystagmus is larger in the occluded left eye, horizontally, vertically, and torsionally, than in the fixing right eye (although such asymmetry was not present in all patients).

The directions of the horizontal, vertical, and torsional nystagmus are important in understanding the directions and types of eye movements that are occurring. It is the slow phase of the latent nystagmus that is particularly instructive to analyze, especially the relative directions of the vertical and torsional slow phases. Note that the corrective saccades, which occur in the opposite direction to the slow phase drifts, always occur simultaneously in the horizontal, vertical, and torsional directions. In other words, single multidimensional corrective saccades, representing the fast phase of the latent nystagmus, are simply the brain's response to the fixing eye's drifting off the intended fixation point. (Dell'Osso and colleagues[43] have shown that the corrective saccades in latent nystagmus actually move the fovea past the object of regard so that the decreasing-velocity slow phase of the nystagmus returns the fovea to the object of regard for a longer period of time before crossing over it and inducing another corrective saccade.)

To understand the eye movements that occur in DVD, the steady-state latent nystagmus will be analyzed first, followed by the vergence movement during the onset of the vertical deviation, followed by the versions that occur to acquire and maintain fixation by the fixing eye.

THE STEADY-STATE LATENT NYSTAGMUS

Fig 2 shows eye movement recordings of patient 4 when the nonfixing left eye is suddenly occluded. Because the right eye is already fixing, there is no fixation saccade at the beginning of the DVD movement. The left eye (LEV) elevates to a steady-state DVD posture, with striking latent nystag-

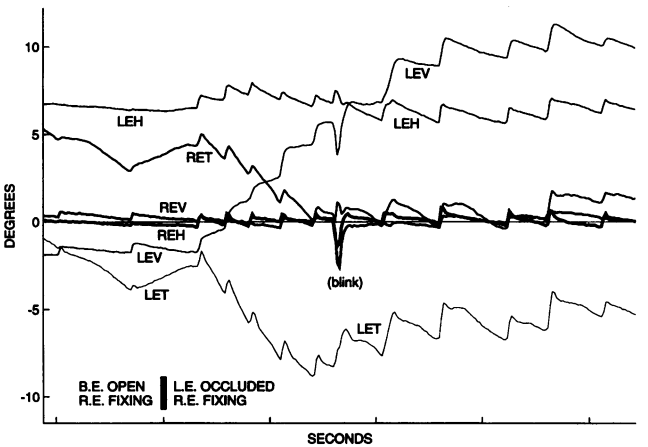

FIGURE 2
Striking latent nystagmus developing in patient 4 when the nonfixing left eye is occluded and left DVD develops.

mus seen horizontally, vertically, and torsionally for each eye once the left eye is occluded. The horizontal, vertical, and torsional tracings are plotted separately in Figs 3, 4, and 5. In Fig 3, both eyes are seen to drift to the left when the left eye is occluded. In Fig 4, both eyes are seen to drift

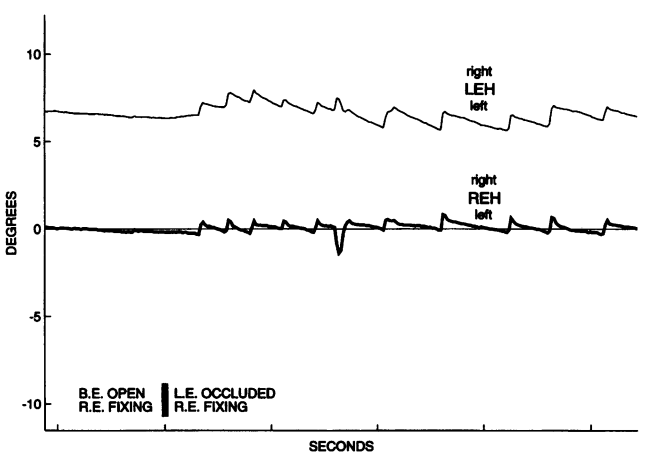

FIGURE 3
Horizontal eye movement tracings from Fig 2, showing both eyes drifting to left when left eye is occluded.

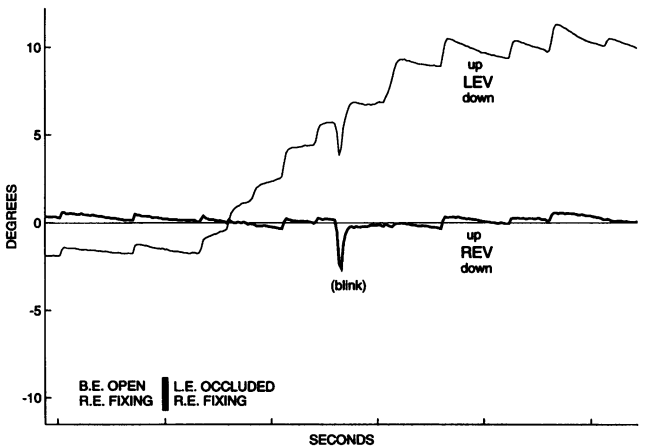

FIGURE 4
Vertical eye movement tracings from Fig 2, showing both eyes drifting down, in the steady state, after left eye is occluded.

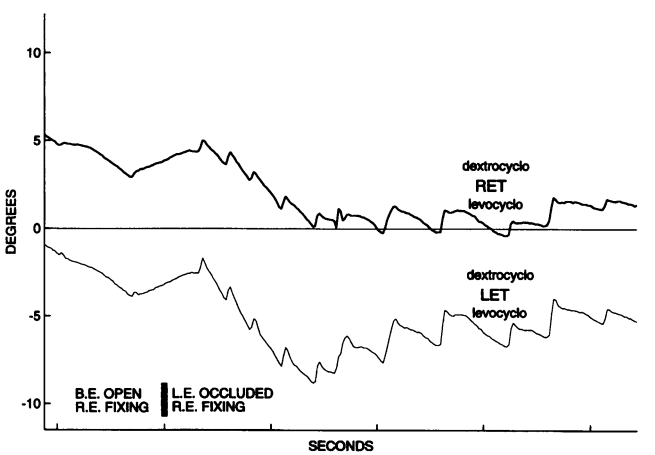

FIGURE 5
Torsional eye movement tracings from Fig 2, showing both eyes drifting counterclockwise (a levocycloversion), when left eye is occluded.

down, in the steady state, after the left eye is occluded. In Fig 5, both eyes drift counterclockwise (a levocycloversion) when the left eye is occluded. In summary, the fixing right eye drifts in, down, and intorts. The nonfixing left eye, behind the occluder, drifts out, down, and extorts. These movements of the 2 eyes together are conjugate, representing versions.

Note in Fig 2 that with the directional conventions chosen for plotting, all six eye movement tracings drift downward simultaneously, with the corrective saccades all occurring in the upward direction. This slow-phase pattern was seen not only in primary position, but also in 20° left gaze and 20° right gaze. Fig 6 shows tracings obtained in each of these gaze positions, showing this common directional pattern of latent nystagmus in the horizontal, vertical, and torsional directions. This common pattern, with each eye movement tracing drifting downward with the left eye occluded, can also be seen in tracings from patients 5 and 6 (Figs 7 and 8, respectively). Patients 5 and 6 show successively lower amplitudes of latent nystagmus than does patient 4, but the pattern of the tracings is the same with the left eye occluded.

Figure 9 shows the striking latent nystagmus that develops in patient 4 when the fixing right eye is suddenly occluded and right DVD develops.

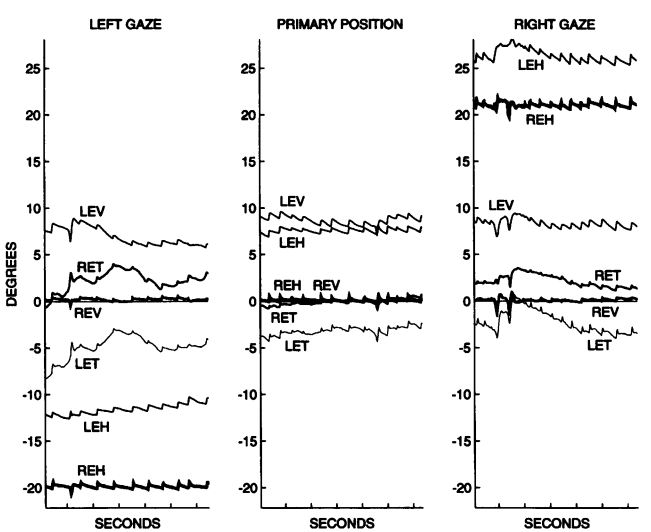

FIGURE 6

Common directional pattern of latent nystagmus in left gaze, primary position, and right gaze for patient 4 with left eye occluded. Each tracing drifts downward regardless of direction of gaze.

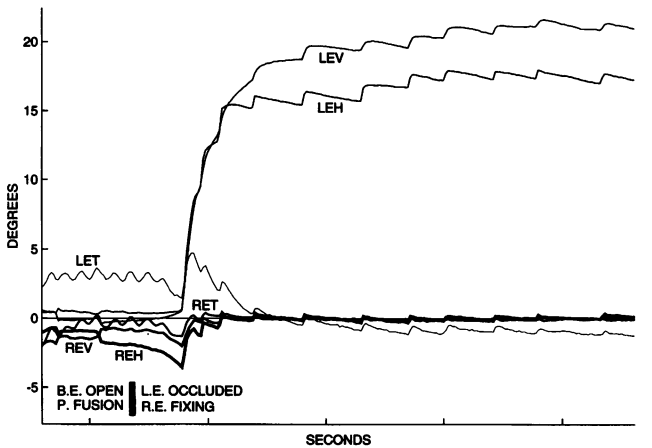

FIGURE 7
Tracings from patient 5 showing same directional pattern of latent nystagmus as in patient 4 when left eye is occluded.

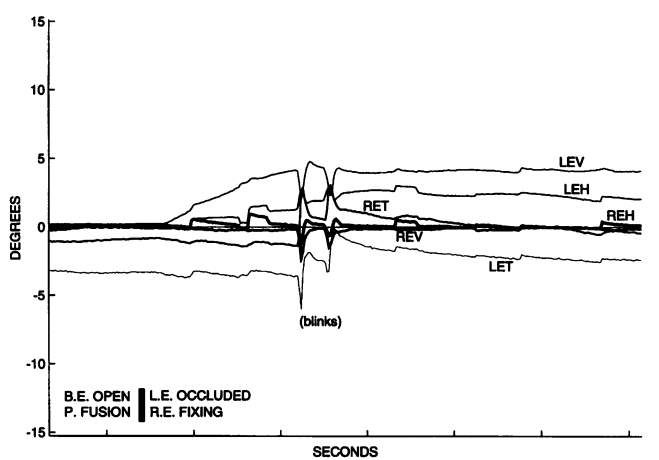

FIGURE 8
Tracings from patient 6 showing same directional pattern of latent nystagmus as in patient 4 when left eye is occluded, although with much lower amplitude nystagmus in all directions.

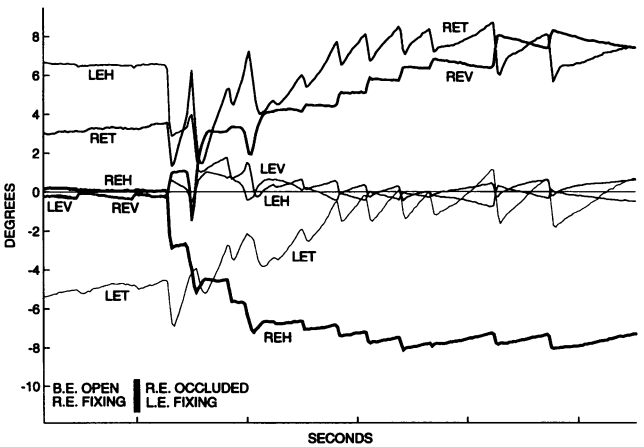

FIGURE 9

Striking latent nystagmus developing in patient 4 when fixing right eye is suddenly occluded and left eye takes up fixation, with right DVD developing.

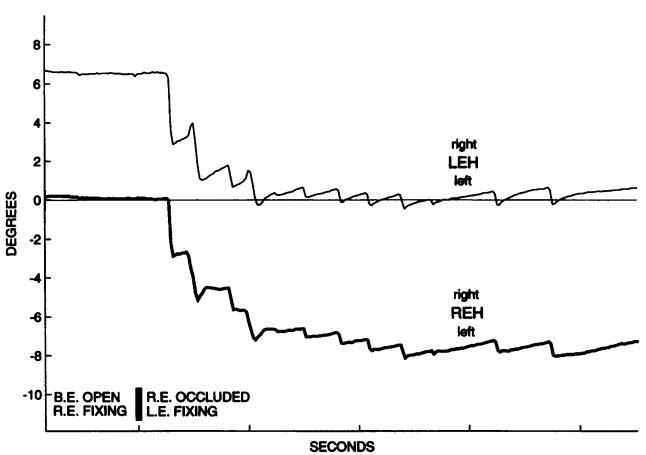

FIGURE 10

Horizontal eye movement tracings from Fig 9, showing both eyes drifting to right, in the steady state, when right eye is occluded.

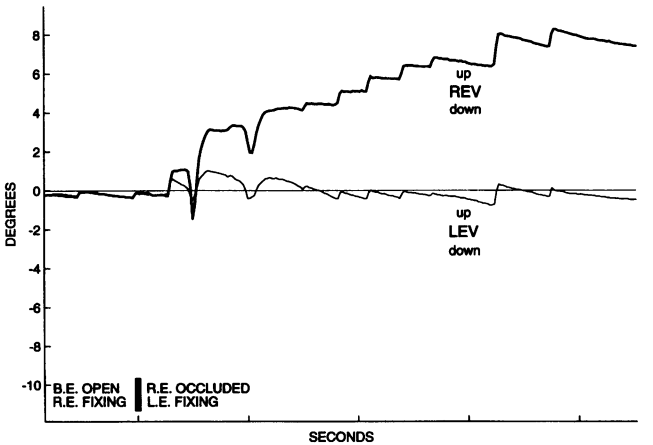

FIGURE 11
Vertical eye movement tracings from Fig 9, showing both eyes drifting down, in the steady state, after right eye is occluded.

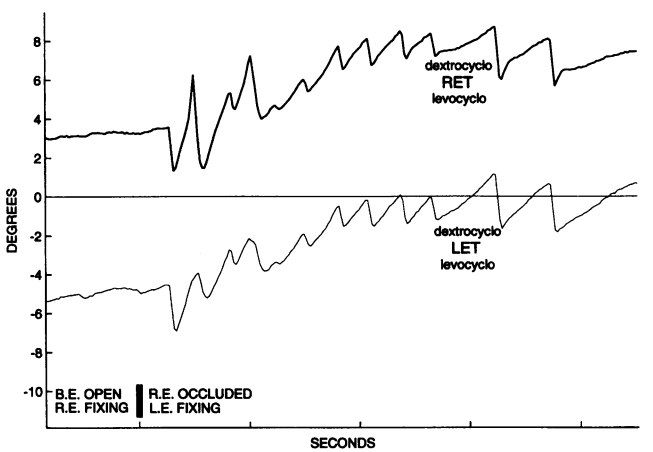

FIGURE 12
Torsional eye movement tracings from Fig 9, showing both eyes drifting clockwise (a dextrocycloversion), when right eye is occluded.

The horizontal, vertical, and torsional tracings are plotted separately in Figs 10 through 12, as before for the left eye occluded. As can be seen from Figs 10 through 12, when the right eye is occluded and the left eye is fixing, the slow phase of the latent nystagmus is to the right, down, and clockwise (a dextrocycloversion). Again, the fixing left eye is drifting in, down, and intorting, and the nonfixing right eye is drifting out, down, and extorting.

With the directional convention chosen for the tracings in Fig 9, the tracings all drift upward when the right eye is occluded, with the exception of the vertical eye movement tracings, which drift downward. This overall pattern in Fig 9 can be seen in both left gaze and right gaze as well (Fig 13, patient 4 with the right eye occluded). This same directional pattern of latent nystagmus, with the right eye occluded, can be seen in patients 5 and 6, in Figs 14 and 15, respectively, although with successively lower amplitudes of the nystagmus.

This same horizontal, vertical, and torsional conjugate pattern of latent nystagmus was described from purely clinical observations by Crone[12,44] in 1954. The downward drift of both eyes has recently been confirmed by video oculography[45] and can be seen in the published eye movement trac-

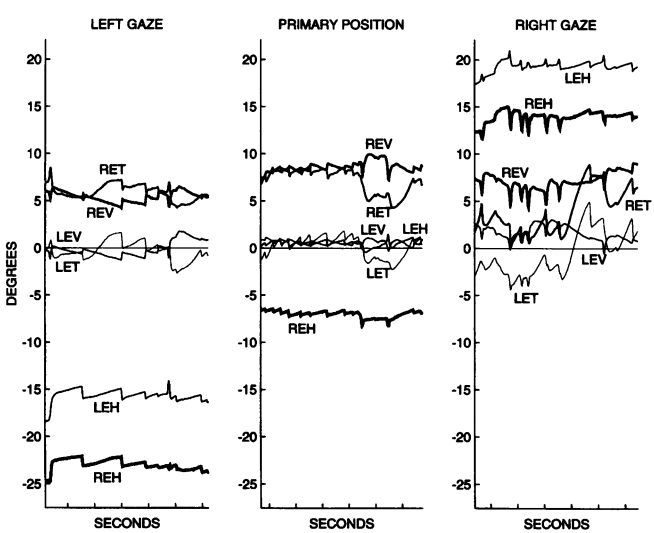

FIGURE 13

Common directional pattern of latent nystagmus in left gaze, primary position, and right gaze for patient 4 with right eye occluded. Each tracing drifts upward with exception of vertical eye movement tracings, which drift downward.

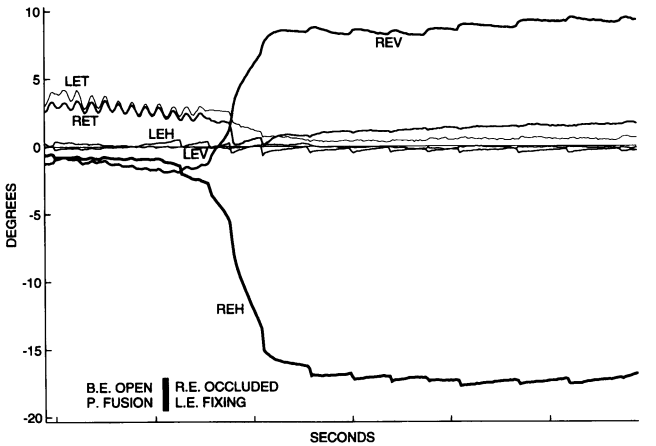

FIGURE 14
Tracings from patient 5 showing same directional pattern of latent nystagmus as in patient 4 when right eye is occluded.

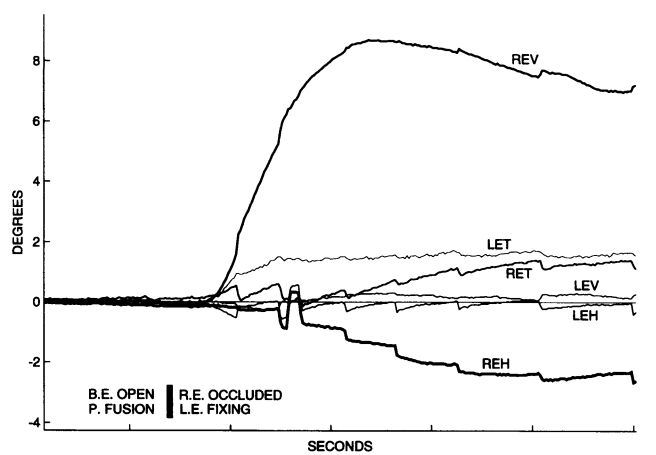

FIGURE 15
Tracings from patient 6 showing same directional pattern of latent nystagmus as in patient 4 when right eye is occluded.

ings of others.[30,41]

The muscles predominantly active in these characteristic directional patterns of latent nystagmus, with the right eye fixing, and with the left eye fixing, can be identified from the multidimensional eye movement recordings. Horizontally, the slow phase of the nystagmus is always away from the fixing eye and the fast phase is always toward the fixing eye, with these movements clearly being mediated by the horizontal rectus muscles.

Cyclovertically, it is possible to identify the predominant muscle active in each of the vertical movements by noting the direction of simultaneous torsional movement. For example, as previously described, an eye can be moved downward by either the superior oblique muscle or the inferior rectus muscle. If the eye intorts while moving downward, the superior oblique muscle must be predominantly active, whereas if the eye extorts while moving downward, the inferior rectus muscle must be predominantly active. Fig 16 shows the vertical and torsional movements of the right eye of patient 4 when the left eye is occluded. The fixing right eye can be seen to be drifting downward and intorting, indicating primary activity of the right superior oblique muscle. Fig 17 shows the vertical and torsional movements of the occluded left eye of patient 4. In the steady-state of the latent nystagmus, the left eye is drifting downward and extorting, implying primary action by the left inferior rectus muscle. The corrective saccades in both Figs 16 and 17 are in the opposite directions to the slow phase movements, implying primary action by the inferior

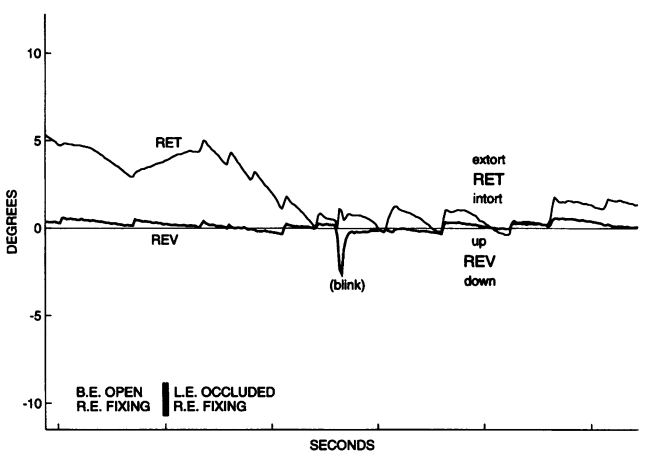

FIGURE 16

Vertical and torsional eye movements of fixing right eye of patient 4, with left eye occluded. Right eye drifts down and intorts, indicating primary action by right superior oblique muscle.

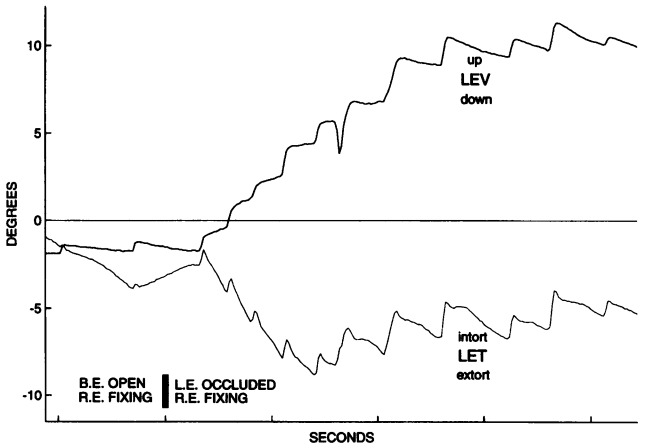

FIGURE 17

Vertical and torsional eye movements of occluded left eye of patient 4. Left eye drifts down and extorts, indicating primary action by left inferior rectus muscle.

	RIGHT EYE (fixing)	**LEFT EYE** (occluded)
Steady state drift:	Down / Intorts	Down / Extorts
Muscle:	SUPERIOR OBLIQUE	INFERIOR RECTUS
	(version - - - - - - - - - - - - - - - version)	
Corrective saccade:	Up / Extorts	Up / Intorts
Muscle:	INFERIOR OBLIQUE	SUPERIOR RECTUS
	(version - - - - - - - - - - - - - - - version)	

FIGURE 18

Summary of drifts and corrective saccades of each eye of patient 4 with right eye fixing; left eye occluded. Vertical movements of fixing right eye are controlled primarily by oblique muscles, and vertical movements of occluded left eye are controlled primarily by vertical rectus muscles.

oblique muscle in the fixing right eye and the superior rectus muscle in the occluded left eye.

These steady-state primary muscle actions are summarized in Fig 18 for both the steady-state drift and the corrective saccades with the right eye fixing (left eye occluded). It is immediately noted that the cyclovertical components of the latent nystagmus in the fixing right eye are mediated primarily by the oblique muscles, and in the occluded left eye, by

the vertical rectus muscles. This is a typical pattern of vestibular nystagmus, the slow phase of which can be elicited by stimulation of the posterior semicircular canal on the right.[46] The oblique muscles in one eye, cooperating with their yoke vertical rectus muscles in the opposite eye, produce conjugate eye movements, in this case a downward drift of each eye accompanied by a levocycloversion. The corrective saccades are upward in both eyes and produce rapid dextrocycloversions.

The apparent result of this analysis, referring back to Fig 6, is that the cyclovertical components of the latent nystagmus of the fixing eye are mediated primarily by the oblique muscles, not only in the straight-ahead position, but also in left gaze and in right gaze. Common teaching holds that the vertical rectus muscles should have more to do with the vertical movements of the eye in abduction. But the vertical component of the latent nystagmus of the fixing right eye in this case is mediated primarily by the oblique muscles, in abduction as well as in adduction. If this interpretation from the eye movement recordings is correct, it is likely that the "pulley" system within the supporting connective tissues of the eye[29,47,48] enables the oblique muscles in the fixing eye to have such comitant vertical action from left gaze to right gaze, and enables the vertical rectus muscles in the occluded eye to have such comitant vertical action from left gaze to right gaze. Alternatively, because of the head-fixed coordinate system used for the recordings, there is some degree of cross-talk between the vertical and torsional recordings in 20° of side gaze. The extent of this cross-talk, and its effect on the above analysis, are yet to be determined.

A similar analysis of the eye movement recordings from patient 4 with the right eye occluded (Figs 19 and 20) results in the summary shown in Fig 21. In this case, again, the cyclovertical components of the latent nystagmus in the fixing (left) eye are mediated primarily by the oblique muscles, and in the occluded (right) eye, by the vertical rectus muscles.

COMPENSATORY EYE MOVEMENTS

The overall latent nystagmus that occurs when either eye is occluded, as just explained, results from the fixing eye's drifting inward, downward, and intorting. The inward and downward drifts are not compatible with continued fixation, so it is not surprising that compensatory eye movements occur to maintain fixation. In addition to the corrective saccades (versions) seen in the steady-state tracings, both horizontal and vertical vergences occur. The typical response that occurs horizontally is an asymmetric convergence, representing the combination of convergence with a horizontal version. The convergence tends to decrease the velocity of drift, thereby damping the

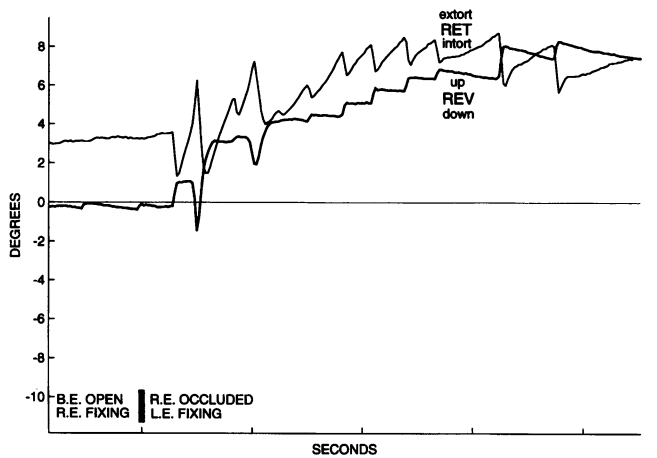

FIGURE 19
Vertical and torsional eye movements of occluded right eye of patient 4. Occluded eye drifts down and extorts, indicating primary action by right inferior rectus muscle.

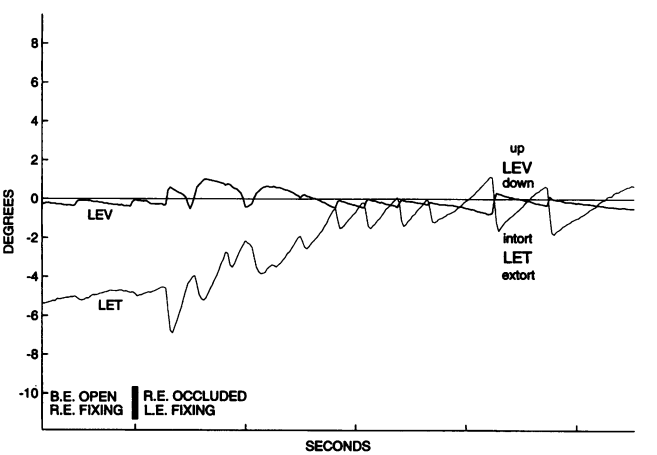

FIGURE 20
Vertical and torsional eye movements of fixing left eye of patient 4, with right eye occluded. Fixing left eye drifts down and intorts, indicating primary action by left superior oblique muscle.

	RIGHT EYE (occluded)	LEFT EYE (fixing)
Steady state drift:	Down / Extorts	Down / Intorts
Muscle:	INFERIOR RECTUS	SUPERIOR OBLIQUE
	(version - - - - - - - - - - - - - - - - version)	
Corrective saccade:	Up / Intorts	Up / Extorts
Muscle:	SUPERIOR RECTUS	INFERIOR OBLIQUE
	(version - - - - - - - - - - - - - - - - version)	

FIGURE 21

Summary of drifts and corrective saccades of each eye of patient 4 with left eye fixing; right eye occluded. Vertical movements of fixing left eye are controlled primarily by oblique muscles, and vertical movements of occluded right eye are controlled primarily by vertical rectus muscles.

horizontal nystagmus and improving vision.[19,49-51] The horizontal version must occur to maintain fixation by the fixing eye, which would otherwise be driven from the object of regard by the convergence.

An analogous vergence and version occur vertically. The vergence appears to decrease the velocity of the vertical slow phase of the latent nystagmus, helping to improve vision. The vertical version must occur to maintain vertical fixation by the fixing eye, which would otherwise be driven from the object of regard by the vertical vergence. It is instructive to examine separately the vertical vergence and the vertical version that occur, and to identify the muscles in each eye that primarily mediate these two eye movements.

THE VERTICAL VERGENCE PRODUCING DVD

As noted earlier, with whole-body roll movement about an appropriate oblique axis, Jáuregui-Renaud and colleagues[35] produced vestibular nystagmus in normal subjects that had the same cyclovertical characteristics as the cyclovertical component of the latent nystagmus in our patients with DVD. The slow phases of the vestibular nystagmus of both eyes were downward, with one eye intorting and the other eye extorting.

The primary muscles acting in each eye during the vestibular-induced movement could be determined from the geometry of the semicircular canals in relation to the oblique roll axis and from the known neural projections of the semicircular canals to the extraocular muscles. The downward-moving, intorting eye was moved primarily by its superior oblique muscle, and the downward-moving, extorting eye was moved primarily by its inferior rectus muscle. The additional observation was that the extorting eye moved down farther than the intorting eye, presumably because of

the more effective vertical action of the inferior rectus muscle, creating a vertical misalignment.

Let us speculate, for the moment, that the cyclovertical component of the latent nystagmus in our patients with DVD involves the same vestibular pathways as the above roll-induced vestibular nystagmus. If so, then the vertical vergence creating DVD may have its roots in the vertical vergence that is normally used to correct the hyperdeviation created by the vestibular-induced counter-roll. If so, and if the vertical vergence is mediated by the normal oblique muscle mechanism,[28] then the downward-moving, intorting eye will be driven farther downward, and intorted more, by its superior oblique muscle, and the downward-moving, extorting eye will be pulled upward, and extorted more, by its inferior oblique muscle.

Evidence for this cycloversion/vertical vergence mechanism is seen in Figs 22 and 23, which show the vertical and torsional movements of the left and right eyes, respectively, of patient 4, when the left eye is occluded and left DVD develops. In Fig 22, as the left eye begins to move upward (LEV) after it is occluded, it also extorts (LET). Such an upward, extorting movement must be mediated primarily by the left inferior oblique muscle, consistent with the proposed mechanism.

In Fig 23, the right eye shows no net vertical movement, because it must maintain fixation, but does show intorsion as the DVD on the left develops. This is consistent with the proposed mechanism, but with the

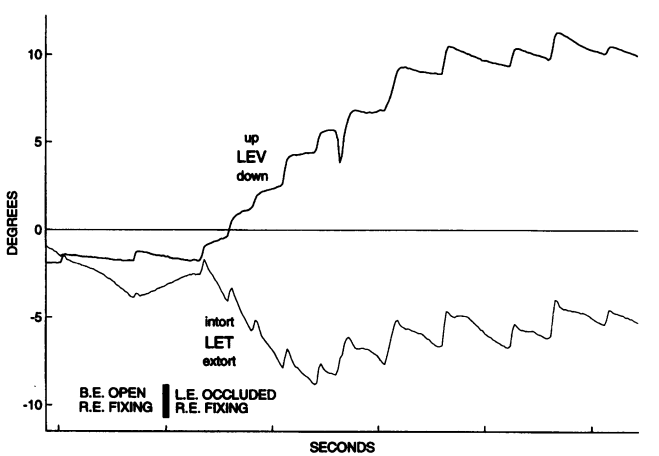

FIGURE 22

Vertical and torsional eye movements of left eye of patient 4 when left eye is occluded suddenly and left DVD develops. Left eye moves up and extorts, indicating primary action by left inferior oblique muscle.

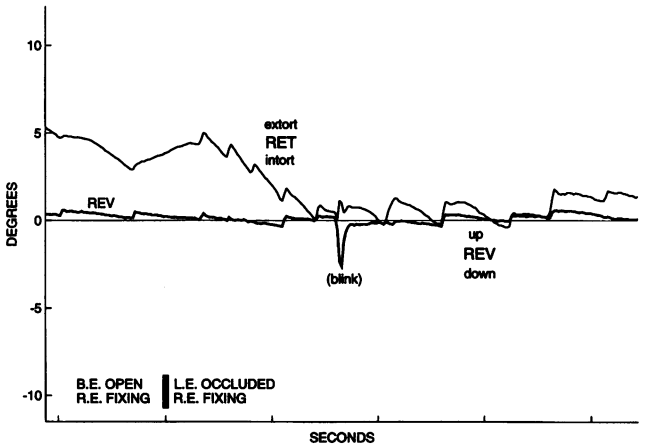

FIGURE 23

Vertical and torsional eye movements of fixing right eye of patient 4 when left eye is suddenly occluded and left DVD develops. Right eye shows no net vertical movement, because it must maintain fixation vertically, but it does intort.

added constraint that fixation with the right eye must be maintained. The pure intorsion of the right eye in Fig 23 cannot be produced by a single cyclovertical muscle acting alone, so at least two cyclovertical muscles must be acting to produce this intorsion. It is tempting to speculate that both the superior oblique muscle and the superior rectus muscle might cooperate in producing the net intorsion, without a net vertical deviation, consistent with the commonly taught physiology underlying the Bielschowsky head-tilt test. It is probably not this simple, however, as can be appreciated by the following analysis of a fixation saccade occurring in patient 5.

THE UPWARD VERSION FOR MAINTENANCE OF FIXATION

The fixing eye must resist being driven downward by the vertical vergence that produces DVD, because it must maintain fixation. An additional eye movement, therefore, most likely an upward version, must occur to maintain fixation.

The nature of the upward version is best examined in those eye movement tracings that show fixation saccades at the beginning of the DVD movement. Fixation saccades are versions themselves, and if it can be determined which muscles are primarily involved during the simultaneous vertical and torsional movements occurring during the upward saccades, then the nature of the version that keeps the fixing eye level in DVD can

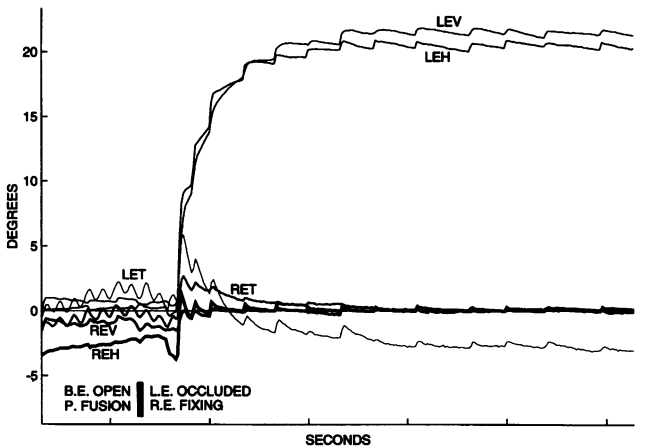

FIGURE 24
Eye movements occurring in patient 5 when left eye is suddenly occluded. Vertical saccade represents a version that raises right eye to take up fixation when left eye is occluded, and resulting downward drift of latent nystagmus begins to drive right eye farther downward.

be better understood.

Fig 24 is an eye movement recording from patient 5 showing asymmetric vertical saccades occurring at the beginning of the DVD movement after the left eye is suddenly occluded. The vertical and torsional movements of the right eye during the saccades are plotted separately in Fig 25, and the vertical and torsional movements of the left eye during the vertical saccades are plotted separately in Fig 26. In Fig 25, the right eye extorts as it saccades upward, indicating primary action by the right inferior oblique muscle. In Fig 26, the left eye intorts as it saccades upward, indicating primary vertical action by the left superior rectus muscle. When the left eye is suddenly covered, therefore, the saccades that occur to raise the right eye to take up fixation are versions that are mediated primarily by the inferior oblique muscle of the right fixing eye and the superior rectus muscle of the left occluded eye.

As is the case with the cyclovertical component of the latent nystagmus, the rapid version (saccade) that is used to establish vertical fixation by the fixing eye involves an oblique muscle in the fixing eye and a vertical rectus muscle in the nonfixing eye. It is likely, therefore, that other vertical versions during the DVD movement, such as slow vertical-pursuit-type movements, which help keep the fixing eye level during the vertical vergence, are also mediated largely by the inferior oblique muscle of the fixing eye and the superior rectus muscle of the occluded eye.

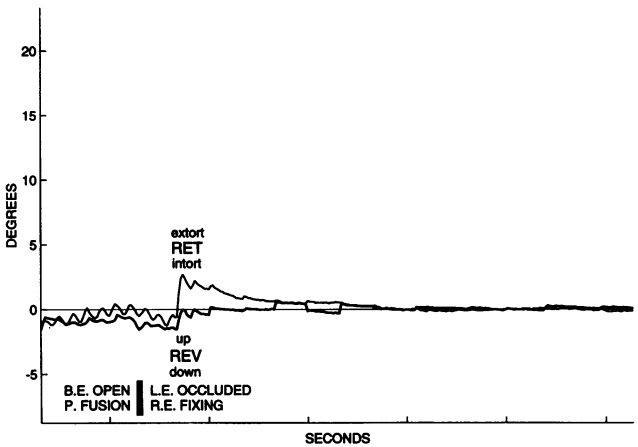

FIGURE 25
Vertical and torsional movements of right eye from Fig 24. In the vertical saccade that occurs shortly after occlusion of left eye, right eye moves up and extorts, indicating primary action by right inferior oblique muscle.

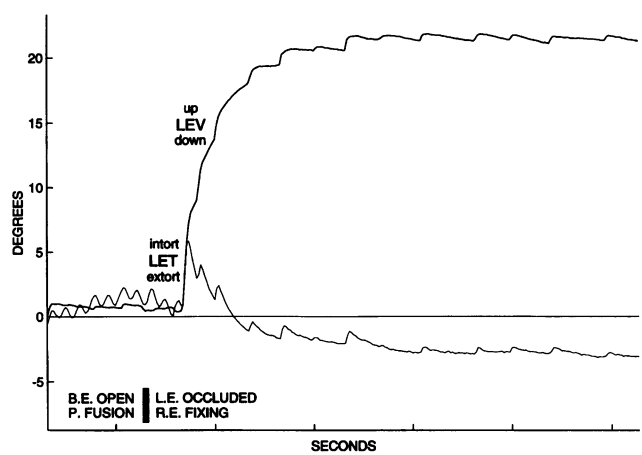

FIGURE 26
Vertical and torsional movements of left eye from Fig 24. In the vertical saccade that begins shortly after occlusion of left eye, left eye moves up and intorts, indicating primary action by left superior rectus muscle.

There is probably some contribution to maintenance of vertical fixation in the fixing right eye by the vertical rectus muscles also (contraction of the superior rectus muscle and/or relaxation of the inferior rectus muscle), because otherwise the inferior oblique muscle would totally negate the torsional action of the superior oblique muscle as well as its vertical action. However, the hypothesis that the inferior oblique muscle is the primary restorer of vertical fixation, when the superior oblique muscle is activated in the fixing eye as DVD develops, helps to explain the clinical observation that the net intorsion of the fixing eye is usually minimal compared with the net extorsion of the occluded eye.

SUMMARY OF THE CYCLOVERTICAL MOVEMENTS

Fig 27 summarizes the primary cyclovertical muscles that appear to act on the two eyes when the left eye is covered, as the right eye continues fixing, and as left DVD develops. For the right eye, the cyclovertical movement is pure intorsion, because fixation must be maintained vertically. The superior oblique muscle is the primary intorter of the right eye, serving as part of the vergence mechanism that comes into play to help damp the cyclovertical latent nystagmus. The inferior oblique muscle is the primary elevator of the right eye, as part of the version that maintains fixation by the right eye despite the tendency for the right superior oblique muscle to push the right eye downward. (The superior rectus muscle is probably a secondary elevator of the right eye, helping to produce the net intorsion that develops.)

The primary muscles acting on the left eye, when it is occluded, and as left DVD develops, are the left inferior oblique muscle and the left supe-

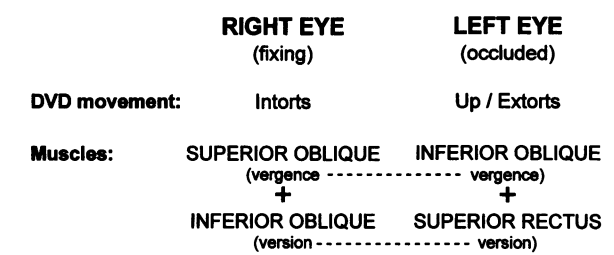

FIGURE 27

Summary of primary cyclovertical muscles acting in each eye as left eye is occluded and DVD develops on the left. DVD represents sum of a vertical vergence mediated by oblique muscles of both eyes and a version mediated by oblique muscles of fixing eye and vertical rectus muscles of occluded eye.

rior rectus muscle. The left inferior oblique muscle acts as part of the vertical vergence that occurs to help damp the cyclovertical component of the latent nystagmus. The left superior rectus muscle acts as part of the upward version to maintain fixation by the fixing right eye. Thus both elevators of the occluded eye are activated simultaneously, driving the eye upward (and extorting it because of a predominance of the vergence movement involving the inferior oblique muscle over the version movement involving the superior rectus muscle). Elevation and extorsion are the characteristic cyclovertical components of the DVD movement. In summary, the mechanism is an exaggerated combination of normal eye movements. A vertical vergence produced by the oblique muscles causes an accompanying cycloversion. An upward version is added to prevent the fixing eye from being driven downward, and this upward version drives the occluded eye up still farther.

An exactly symmetric analysis can be performed with the eye movement tracings in Fig 9, which shows the eye movements in response to occluding the right eye in patient 4, with DVD developing on the right. Again, the fixing eye is intorted by the superior oblique muscle and is maintained level primarily by activation of the inferior oblique muscle of the fixing eye. The occluded eye is driven upward by a combined action of its inferior oblique and superior rectus muscles.

THE HORIZONTAL MOVEMENTS

The analysis thus far has concentrated primarily on the cyclovertical movements of the two eyes during the DVD response. The cyclovertical muscles also have tertiary, horizontal actions, and these should be considered in helping to explain the horizontal movements of the occluded eye in the typical DVD response.

The oblique muscles that become activated in the fixing eye both have tertiary, abducting effects. The fixing eye will, therefore, tend to abduct as the DVD develops. Thus, for maintenance of fixation, a horizontal version away from the fixing eye will have to occur. This horizontal version toward the occluded eye, in cooperation with the abducting effect of the active inferior oblique muscle in the occluded eye, drives the occluded eye outward, as a rule. In those cases where there is no compensatory horizontal convergence to help damp the horizontal component of the latent nystagmus, an abduction of the eye under cover is noted, completing the familiar triad of elevation, extorsion, and abduction that is seen in the occluded eye of many patients with DVD.

The abduction that tends to occur in the occluded eye can obviously be reduced, or even reversed, by any convergence that occurs as a compensatory mechanism to damp the horizontal component of the latent

nystagmus. The net horizontal movement of the occluded eye represents the horizontal component of the dissociated strabismus complex. This movement may well be asymmetric, with one eye fixing versus the other eye fixing, because of different amounts of latent nystagmus induced when one or the other eye is covered. The net horizontal component of the DVD (DHD) can obviously be an abduction or an adduction of the covered eye, and can be symmetric or asymmetric with occlusion of the two eyes, respectively.

COMPENSATION FOR CYCLOVERTICAL LATENT NYSTAGMUS

Some patients show very little latent nystagmus but can show large amounts of DVD. One must therefore postulate that if the DVD is occurring to help damp cyclovertical nystagmus, it is occurring in an anticipatory fashion, happening so quickly that the increased nystagmus is immediately damped and never has a chance to show itself. On rare occasions, careful analysis of an eye movement tracing will appear to show increased latent nystagmus occurring shortly after occlusion of one eye, just prior to the DVD compensatory movement, but this is most unusual. It is more common to see gradual damping of the latent nystagmus in the fixing eye as the DVD develops, as in Figs 1, 7, 14, and 15. The slope of the slow phases corresponds to the amplitude of the latent nystagmus, because the steeper the slow phases, the more frequent and the larger the corrective saccades have to be to return the fixing eye toward (or actually past) proper fixation. Note in Fig 1, for example, how the initial vertical and horizontal nystagmus of the right eye becomes quickly damped as the DVD develops, with the fixing eye showing a low amplitude of horizontal and vertical nystagmus within 1 second after the onset of the DVD movement. The elevated, nonfixing eye still shows a moderate amount of vertical and horizontal latent nystagmus.

The neurologic mechanism by which a superimposed vergence damps nystagmus is not clear. For convergence damping of horizontal nystagmus, some evidence suggests that the damping is roughly proportional to the amount of convergence[19] and is not related to the symmetry or asymmetry of the convergence.[51]

There is no question that patients often use head posture to damp the horizontal component of latent nystagmus by rotating the fixing eye into adduction. Alexander's law, which states that jerk nystagmus is damped with gaze in the direction of the slow phase, is relatively well explained by ocular motor control theory.[52,53]

The cyclovertical component of latent nystagmus also appears to be damped in some instances by compensatory head postures: chin up, chin down, or head tilt. It may be that even in straight-ahead gaze, some of the

damping effect produced by DVD is related to a simulated head-tilt mechanism. Because the torsional position of the fixing eye is not critical for fixation, the eye can assume an intorted posture via the cycloversion/vertical vergence mechanism of DVD. This intorted posture is in the direction of the slow phase of the torsional component of the latent nystagmus and may therefore help damp the latent nystagmus via the neurologic mechanism underlying Alexander's law.

SUMMARY OF EYE MOVEMENTS

Based on the above step-by-step analysis, the eye movements occurring in DVD include the following:

1. *Latent Nystagmus*

a. A comitant drift of both eyes, with the fixing eye adducting, depressing, and intorting. The horizontal muscles mediate the horizontal component of the drift, while the oblique muscles primarily produce the cyclovertical drift in the fixing eye and the vertical rectus muscles primarily produce the cyclovertical drift in the occluded eye.

b. Comitant saccades, compensating for the drift of the fixing eye, representing attempts to restore useful fixation. These involve the same muscles that produce the comitant drift, but in the opposite direction. These saccades are the quick phases of the latent nystagmus, with the comitant drift representing the slow phases.

2. *Vertical Vergence*

A combination cycloversion/vertical vergence, with the fixing eye depressing and intorting, and with the occluded eye elevating and extorting, mediated primarily by the oblique muscles in both eyes. This combination movement tends to damp the cyclovertical component of the latent nystagmus. Because of the tertiary horizontal actions of the oblique muscles, a certain amount of horizontal divergence of the two eyes is produced as well. Both the fixing and occluded eyes tend to abduct when the superior oblique muscle in the intorting fixing eye and the inferior oblique muscle in the extorting occluded eye become active.

3. *Vertical Version*

An upward version, necessary to compensate for the depression of the fixing eye caused by the vertical vergence. This upward version, in the form of saccades or smooth eye movements, is probably mediated largely by the inferior oblique muscle of the fixing eye and the superior rectus muscle of the occluded eye.

4. *Horizontal Version*
A horizontal version, away from the fixing eye, to compensate for the collective tertiary abduction effects of both oblique muscles as they become active in the fixing eye. This horizontal version helps to maintain horizontal fixation by the fixing eye and causes net abduction of the covered eye.

5. *Convergence*
An asymmetric convergence, mediated by the horizontal rectus muscles, and variable in amount, serving to damp the horizontal component of the latent nystagmus.

6. *Fixation Saccades*
Transient fixation saccades when taking up fixation with one eye or the other. These are roughly comitant. The horizontal saccades are mediated by the horizontal muscles in both eyes. Some of the cyclovertical saccades are probably mediated largely by the oblique muscles in the fixing eye and by the vertical rectus muscles in the nonfixing eye. (It is not always an oblique/rectus muscle pair that produces saccades. Large voluntary upward saccades often cause transient, simultaneous intorsion of both eyes, indicating primary action by both superior rectus muscles, while large downward saccades often cause transient, simultaneous extorsion of both eyes, indicating primary action by both inferior rectus muscles.)

DISCUSSION

With the great variety of versions and vergences occurring simultaneously and in varying amounts, it is not surprising that the summed DVD movement has been confusing. When the total movement is separated into its components, however, one can determine that Hering's law, when applied to each component, appears to be obeyed.[54]

What causes the inward, downward, and intorting drift of the fixing eye that leads to the fast-phase corrective movements that we label as latent nystagmus? Because similar cyclovertical drift components can be produced in normal subjects by dynamic head tilting about an oblique axis, an imbalance in the vestibular system is likely the culprit. To counter the vertical deviation that such dynamic head tilting produces, normal subjects appear to use a vertical vergence during dynamic head tilting to help maintain vertical fusion. This vertical vergence is in the same direction as, and appears to be analogous to, the vertical vergence that creates the exaggerated DVD response in patients with a defect in binocular vision and resulting latent nystagmus. In these patients the vertical vergence of the DVD response serves to damp the drift that causes the latent nystagmus. The vertical vergence producing the sustained hyperdeviation of the occluded eye in DVD

may thus simply be considered to be an exaggeration of the normal compensating vertical vergence that occurs with dynamic head tilting.

One is led to speculate that covering one eye in patients with DVD, or simply willful monocular fixation[21] (even with a blind eye or even in the dark[44,55,56]) leads to unbalanced input to the vestibular system, producing the eye movements of latent nystagmus. Such a mechanism may help explain the DVD-like movements noted by Bielschowsky[26] after prolonged unilateral occlusion of normal subjects. Furthermore, because inputs to the vestibular system would rarely be expected to be precisely equal in normal subjects, it should not be surprising that minimal DVD movements have recently been recorded in normal subjects.[57]

Upon realizing the connection between dynamic head tilting and cyclovertical latent nystagmus, we began to question patients with DVD regarding the subjective straightness of their heads when either eye was covered. Indeed, some patients with DVD report that the head feels as if it is tilted toward the side of the fixing eye as soon as the other eye is occluded, even though the head remains entirely straight.

The above analogy between the movements occurring in dynamic head tilting and the movements of DVD will undoubtedly help us understand the changes in DVD that occur with head posture. Also, we should be able to understand more clearly the contribution of the otolith system to the anomalous head postures that we often see in patients with DVD.

As is often the case in biologic systems, there are exceptions to the rule. One of our six patients, patient 1, showed what first appeared to be typical DVD movements, but the slow-phase drift of her eyes was always in the upward direction rather than in the downward direction. Because the recordings were not complete, however, we have not been able to establish the exact eye movement mechanisms in her particular variant of DVD. Second, there is an occasional patient with inverse DVD — in other words, a downward-deviating eye on occlusion or on spontaneous suppression of the vision in that eye.[2] Such movements can likely be explained by similar analyses as performed above. The drift directions, either horizontal, vertical, or torsional, will probably be different in these patients. The usual inward, downward, and intorsion drift of the fixing eye, which has just been analyzed in detail, appears to be the most common eye movement pattern in patients with DVD.

The analogy of the eye movements occurring in dynamic head tilting to the cyclovertical eye movements occurring in DVD may also help us devise noninvasive methods for minimizing or preventing the exaggerated DVD eye movements. If such efforts are not successful, then at least the understanding of the eye movements occurring in DVD should help us devise better surgical procedures for correcting this disfiguring form of strabismus.

One of our younger associates, Cameron Parsa, suggested that ablation of all four oblique muscles might be the best solution. We are concerned that such a procedure might lead to secondary complications, or perhaps loss of normal oblique muscle functions that we have not yet identified. A safer, and perhaps more logical, procedure will be to recess and anteriorly transpose the insertions of all the oblique muscles. This should minimize their vertical actions and thereby decrease or even prevent the vertical vergence that is the most objectionable component of the dissociated strabismus complex.

Unilateral superior rectus muscle recessions in the past have often led to worsening of the DVD on the contralateral side. This is undoubtedly because recession of the superior rectus muscle makes it easier for the ipsilateral superior oblique muscle to drive the ipsilateral eye downward, resulting in a hyperdeviation on the contralateral side when the ipsilateral eye takes up fixation.

In conclusion, the component eye movements that produce DVD and its associated torsional and horizontal deviations can be identified by careful directional analysis of the simultaneous vertical and torsional eye movements that occur in this disorder. As the component eye movements are identified, Hering's law appears to be obeyed after all, but for each movement separately, not for the DVD movement as a whole. Finally, DVD seems to have a purpose, providing a cyclovertical "nystagmus blockage" function. The vertical vergence component of DVD appears to be the cyclovertical analogue of the horizontal convergence that is used by some patients to help damp the horizontal component of latent nystagmus. Mediated primarily by the oblique muscles and probably being a learned response, the vertical vergence of DVD appears to at least partially damp the intensity of the cyclovertical component of the latent nystagmus that is associated with DVD, thereby preventing a decrease in vision that the latent nystagmus would otherwise cause.

Much remains to be discovered about the dissociated strabismus complex. The analysis above, however, should provide a good foundation for rapid progress.

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DISCUSSION

DR GUNTER K. VON NOORDEN. Bielschowsky[1] proposed in 1931 that dissociated vertical deviations (DVD) are caused by a vertical vergence signal of unknown origin that elevates the occluded eye. The impulse to depress the fixating eye is neutralized by a supraversion innervation in the opposite direction that is transmitted to both eyes and elevates the occluded eye even further. Bielschowsky reported also that DVD may be associated with incycloduction of the fixating eye, excycloduction of the elevating eye, and a cyclovertical latent nystagmus. Highly accurate eye movement recording techniques used in this and another study[2] are in support of this classic view.

From the direction of the cycloversion during DVD, the essayists and others[3] have concluded that vertical vergence movements must be predominantly mediated by the oblique muscles, because vertical rectus contraction would produce a cycloversion in the opposite direction. The authors then propose that this oblique-muscle-generated cycloversion is a purposeful eye movement that damps a latent cyclovertical nystagmus to improve visual acuity. The accompanying elevation of the nonfixating eye, the so-called DVD, is interpreted as an unavoidable side effect of this nystagmus damping mechanism.

I find it difficult to envision the oblique muscles as the principal elevators and depressors during vertical vergences. Because of the angle formed between the muscle planes of the oblique muscles and the visual axis in an abducted eye, the vertical effect of the obliques is practically negligible in abduction. A vertical vergence mechanism that would function only in primary position but not in lateral gaze would not be of much use to us in overcoming vertical retinal disparities. Surely, only the oblique muscles can produce a cycloversion in the direction recorded by the authors. However, this finding could also be explained by a more powerful torsional action of the oblique muscles that overrides the less powerful torsional effect of the vertical rectus muscles in the opposite direction, while

both cyclovertical muscle pairs function in unison in elevation and depression. The frequent findings that DVD continues unabated after a myectomy of the ipsilateral inferior or a tenectomy of the superior oblique muscle[4] and that elevation of the occluded eye may actually be greater in abduction than in adduction[4] also militate against a predominant role of the oblique muscles in its causation.

Several observations cannot be reconciled with the proposed damping mechanism of latent nystagmus by a vertical vergence/cycloversion innervation. In my own experience and that of others,[1,2,5-8] neither the latent nystagmus nor the extorsion of the elevated eye or the intorsion of the fixating eye is a consistent feature of DVD. Moreover, the only known trigger for a nystagmus damping effort is a decrease of visual acuity caused by the nystagmus.[7] It has not been shown that visual acuity first decreases with the onset of the nystagmus and then improves again as DVD develops and the nystagmus diminishes. Also, the mechanism of a purported cycloversion/vertical vergence damping sequence is unclear, since the only known stimulus for a cycloversion is vestibular excitation and for a vertical vergence is vertical retinal disparity. Neither of these events is known to precede the development of DVD.

A cause-effect relationship different from that proposed by the authors deserves consideration. Could it not be that an exaggerated vertical vergence movement of unknown origin causes the DVD and that this vergence innervation, superimposed on the nystagmus innervation, damps the nystagmus? We know that convergence innervation has this damping effect on nystagmus. In other words, could the nystagmus damping not be a consequence of DVD rather than its cause?

The essayists are to be congratulated on a thought-provoking contribution. I hope they forgive me if I cannot yet embrace their explanation of DVD as a side effect of a nystagmus damping effort.

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EDWARD L. RAAB, MD. This is a very interesting concept, and I have some questions. What will be of importance is to confirm your observations in right gaze and in left gaze. At least clinically, that seems to be the requirement for the clinical diagnosis of DVD. I would also expect that even though different innervations may be the same, this should be proved in straight ahead up and straight ahead downgaze. I wonder if you have looked at that, Dave?

And, my next question has to do with the Bielschowsky darkening phenomenon, not the Bielschowsky head-tilt test, but the darkening phenomenon, which as far as I know is associated only with DVD. You say in your abstract that DVD and nystagmus diminish in the dark. Yet in trying to demonstrate the Bielschowsky darkening phenomenon, particularly as I had occasion to do once in a film, I found that I couldn't elicit it unless the flood lighting that I used to produce the film was considerably diminished. It made for a poor quality film, but it brought out the phenomenon much better. The Bielschowsky darkening phenomenon is a characteristic of DVD as opposed to any other vertical deviation. I think I am correct about that. Why does that behave differently in dimmer lighting than the phenomenon which you describe?

JOHN FLYNN, MD. I would like to make the audience aware that David gave this talk as a 45-minute talk as the Costenbader Lecture. It was wonderful to be in the audience. I'm sorry that all of our colleagues here today could not have been exposed to his analytical thought processes coupled with the video displays of eye movements that he showed. When you have a chance to review the published paper, ladies and gentlemen, I think you will see that David has given us another major contribution to this topic. I have a problem with the basic hypothesis that DVD blockades the nystagmus. Let me present it in the form of a patient we are examining together. The child has latent and manifest latent nystagmus. The nonfixing eye will spontaneously go into DVD and at the same time nothing that I can detect has happened to the latent nystagmus in the fixing eye. If this, in fact, is a vergence type of movement to blockade latent nystagmus, why doesn't this mechanism produce a perceptible change in the latent nystagmust. Thank you.

ALAN B. SCOTT, MD. That's an elegant analysis, David. And, I am sure there is some truth in it. EMG of the elevating eye shows, as you indicated, that both vertical muscles, elevator muscles, increase their innervation to drive the eye upward. And importantly, as we have discussed between us, EMG of the fixing eye has not really been done systematically in this situation and would be very valuable in analyzing these cases.

I'd like you to comment further on the ARVO poster that was recently presented, from Holland, showing half a dozen normal young male indi-

viduals with 20/20 vision and perfect binocularity — again fitted with search coils, and one eye was covered, and, of small amplitude, there were elevation and torsional movements systematically in all of these normal people, but no nystagmus. So I'm wondering whether there is a necessary correlation between nystagmus as you have implied and the elevation vertical behavior of the eye.

DAVID L. GUYTON, MD. I thank the discussants for their excellent comments. I am particularly honored that Gunter von Noorden, my fellowship mentor, agreed to open the discussion, for it was he, 22 years ago, who introduced me to the relatively unexplored area of ocular torsion. I thank him especially, though, for sharing his intended discussion with me yesterday, enabling me to make my response much more coherent. Dr von Noorden is also one of the few members of our Society who could have ferreted out Bielschowsky's 61-page German treatise on the subject. I was delighted to learn that Bielschowsky's observations were consistent with the eye movement recordings that we have documented. Robert Crone in Holland also described the same eye movement patterns back in 1954.

Dr von Noorden finds it difficult to envision how vertical vergence is normally mediated largely by the oblique muscles, especially because of the incomitance that might be expected in side gazes. I had difficulty with that too, until I realized that maintenance of a vertical vergence requires one oblique muscle to be active in each eye, acting in opposite directions. On side gaze, therefore, one of the oblique muscles will become less vertically effective, and the other will become more vertically effective, thus maintaining the same vertical vergence. It's actually a beautiful design to maintain vertical conjugacy in side gazes.

I agree that various forms of oblique muscle surgery in the past have not been very effective in reducing DVD. This is partly, I suspect, because reattachment practically always occurs after myectomy or tenectomy, either to the globe, or via Tenon's capsule to the globe, as confirmed by exaggerated traction testing in those patients requiring muscle surgery for other reasons later. More importantly, though, I would not expect significant improvement of DVD unless all four oblique muscles are weakened and anteriorly transposed. Even the bilateral inferior oblique recession and anterior transposition procedure, which has now become popular, is not fully effective. Nor should it be expected to be, because weakening the inferior oblique muscle in the fixing eye makes it easier for the superior oblique muscle to drive that eye downward, with the DVD persisting. In my opinion, therefore, all four oblique muscles must be addressed. The proof will rest with the patients in whom this is done, such as the patient I showed as part of the presentation.

The nystagmus-damping aspect of our thesis is still somewhat specula-

tive. The best evidence we have is the patient who develops DVD when reading down the visual acuity chart to smaller and smaller letters. A third of our patients showed this phenomenon when we began looking for it.

Our eye movement tracings show that when one eye is covered, the DVD and the latent nystagmus develop simultaneously, not one before the other. Chronologically, though, Mein and Johnson showed nicely in their large, longitudinal study that latent nystagmus develops at an earlier age than DVD, and that it practically always, if not always, was present in their patients who went on to develop DVD. The latent nystagmus often diminished with time. They, and we, feel that this is strong evidence that DVD is a learned, usually unconscious, compensatory response that develops immediately upon unilateral fixation and damps the latent nystagmus that would otherwise persist.

As for the variation in the relative amounts of torsion, vertical deviation, and horizontal deviation seen in DVD, we suspect that this represents variation in the different types of latent nystagmus that are being compensated, as explained further in the written manuscript.

Dr Raab encouraged us to look at what is happening in right and left gaze, and in up and downgaze. Indeed we are doing that, because the findings will give beautiful clues as to what is going on in DVD. We have the tracings, and that analysis is ongoing; we simply have to look at the tracings more carefully. He also asked about the Bielschowsky darkening phenomenon. Bielschowsky described how he could get the same effect by reflecting a strong light into the eye. Also, Verhoeff described how he could do the same thing by placing a diffuser in front of the eye. Robert Crone described the same phenomenon if he put an illuminated occluder in front of the fixing eye, so it appears to be an interference with fixation rather than a darkening effect if one can get the same phenomenon with these different types of "occlusion."

Dr Flynn has a patient with manifest latent nystagmus who does not seem to change his nystagmus as DVD comes and goes in the fellow eye. I suspect that if we did eye movement recordings on the patient, we would indeed see a slight increase in the latent nystagmus when one eye is covered or when one eye is spontaneously concentrating on fixing. It is unusual to see the nystagmus increase and then become damped. I think that the DVD and the damping occur simultaneously, and that the latent nystagmus or manifest latent nystagmus would otherwise increase, but that needs to be looked at further.

Dr Scott referred to the recent report at ARVO (that was also published in the European Strabismological Association Transactions last fall), showing that normal subjects can show small amounts of DVD. And he commented that the tracings that were shown in the published figures and on the ARVO poster showed no latent nystagmus at all. I questioned the

authors about this as well, and the author manning the poster pointed out that the tracings that were shown were vergences. Latent nystagmus is a comitant eye movement, and will disappear when plotting vergence. So we would not expect any latent nystagmus to show in those tracings at all. The author stated that he would have to go back and look for latent nystagmus in the raw tracings.

It is fascinating that you can see these DVD movements in normals. That tells me, you see, that in the same way that our two index fingers are not the same number of microns in length, the inputs from the two eyes to the vestibular system are not exactly balanced in individuals. And when you go to finer and finer eye movement tracings and cover one eye versus the other, this small imbalance will show up. This appears as clinically-obvious DVD in our patients with congenital esotropia, but one can also see small amounts of DVD in normals because of that slight imbalance that exists in all biologic systems.