Dissociated Vertical Divergence
A Righting Reflex Gone Wrong

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Dissociated vertical divergence (DVD) is an ocular motor disorder characterized by a slow, upward drift of 1 eye when the other eye is fixating a target. I propose that DVD is a dorsal light reflex in which asymmetrical visual input to the 2 eyes evokes a vertical divergence movement of the eyes. This primitive visuo-vestibular reflex functions as a righting response to restore vertical orientation in lower lateral-eyed animals by equalizing binocular visual input. The dorsal light reflex is suppressed in humans but can manifest as DVD when early-onset strabismus precludes normal binocular development. Arch Ophthalmol. 1999;117:1216-1222

In the course of evolution, primitive responses to external stimuli are suppressed by newer neurologic reflexes. When newer systems fail to function properly, primitive reflexes that have been phylogenetically retained may reappear.1 The emergence of dissociated vertical divergence (DVD) in children with early-onset strabismus may signal this type of atavistic response. It is my hypothesis that DVD is a dorsal light reflex that produces a visually mediated modulation of central vestibular tone. This visuo-vestibular reflex, which is activated by fluctuations in binocular visual input, manifests in humans when binocular control mechanisms fail to develop in infancy.

WHAT IS DVD?

Dissociated vertical divergence is an enigmatic disorder characterized by a slow ascent of 1 eye that is followed, after a variable interval, by a slow descent of the higher eye back to the neutral position.2-4 The deviating eye frequently extorts during its ascent, then intorts as it descends to resume fixation. Dissociated vertical divergence manifests when binocular visual input is mechanically, optically, or sensorially preempted (Figure 1).5,6 During the period of vertical misalignment, visual input from the hyperdeviated eye is usually suppressed by the brain so that affected individuals do not experience diplopia.2-4 Since the intermittent hyperdeviation of 1 eye is unassociated with a corresponding hypotropia in the nondeviated eye on alternate cover testing, DVD is said to ignore Hering’s law and seems to defy explanation according to current concepts of neuroanatomy.4

A cogent neurophysiological explanation of DVD must account for the following observations:

1. Dissociated vertical divergence is a postscript to any early disruption of normal binocular interactions.6 It is seen most commonly in the setting of congenital esotropia but is also observed in association with congenital exotropia and after surgical realignment of the hypotropic eye in congenital “double elevator palsy.”6 As succinctly stated by Helveston,3 “… DVD is a reflex type of event that is programmed to occur if the appropriate mechanisms for nullifying its expression are not functional.”

2. A cogent neurophysiological explanation of DVD must account for the following observations:
1. Both eyes never drift up simultaneously (Figure 1).\textsuperscript{7}

2. An inverse form of DVD, in which 1 eye drifts downward below horizontal position, has only rarely been observed.\textsuperscript{8} Why should this intermittent vertical deviation manifest only as a hypertropia, and why should this hypertropia alternate between the 2 eyes?

3. DVD develops as a delayed phenomenon in children with infantile strabismus. It is usually first noted between 2 and 5 years of age.\textsuperscript{4}

4. The amplitude of the hyperdeviation is often asymmetrical in the 2 eyes, and DVD may be unilateral in amblyopic eyes.\textsuperscript{2-4}

5. The vertical amplitude of DVD is variable, making accurate measurement difficult.\textsuperscript{2,3}

6. The slow velocity of the upward and downward drift of the deviating eye does not resemble a saccade or pursuit movement but rather a slow divergence in which fixation is constantly maintained by the nondeviating eye.\textsuperscript{2,3}

7. Placement of a vertical prism before 1 eye will induce a corresponding vertical divergence.\textsuperscript{2}

8. The amplitude of the vertical deviation is incrementally related to the asymmetry of visual input in the 2 eyes.\textsuperscript{2,4} This effect is most clearly shown by the Bielschowsky phenomenon, in which filters of increasing density placed before the fixating eye cause the hypertropic eye to descend incrementally, sometimes into a hypotropic position.\textsuperscript{2}

9. In some individuals, the horizontal component of DVD predominates.\textsuperscript{9}

10. DVD is accompanied by a manifest head tilt in approximately 35\% of cases.\textsuperscript{10-12}

WHAT IS A DORSAL LIGHT REFLEX?

Dissociated vertical divergence recapitulates a primitive visuo-vestibular righting response in lateral-eyed animals (termed the dorsal light reflex or Lichtrückenreflex).\textsuperscript{13} To determine the role of DVD in humans, one must first understand the nature and function of the dorsal light reflex in lower animals. Animals need to know which way is up, since maintaining vertical orientation is important for balance, navigation, and survival. Primates rely predominantly on graviceptive input to the vestibular system (ie, gravity receptors within the 2 labyrinths) to maintain verticality. The need to use visual input is evident when one considers that a fish swimming in turbulent waters is subjected to mechanical forces that produce constant fluctuations in vestibular input.\textsuperscript{14}

In 1935, von Holst\textsuperscript{15,16} discovered that visual and otolithic signals are yoked within the central vestibular system to establish postural orientation in the roll plane. In the restrained, labyrinthectomized fish, labyrinthine input can no longer curb this visually induced postural reflex, and the vertical divergence re-
response to a lateral light stimulus is approximately doubled.\textsuperscript{17} The enhancement of this visual righting reflex in the absence of vestibular input demonstrates that the dorsal light reflex is a visually mediated ocular tilt reaction that is counterbalanced by the otoliths.\textsuperscript{14} If the dorsal light reflex in fish resulted from otolithic imbalance, ablation of the otoliths would abolish it rather than increase it. When the right labyrinth and left eye are left intact, utricular and visual innervation oppose each other and normal postural responses are again observed.\textsuperscript{14,16} This variability is also a prominent feature of DVD. In DVD, the observed asymmetry in the hyperdeviation of the 2 eyes may reflect the momentary visual advantage of 1 eye, as determined by the degree of amblyopia or by fluctuations in the level of suppression.\textsuperscript{8} In the dorsal light response and in DVD, the eyes diverge slowly and the divergence persists for a variable period after the inciting stimulus is removed.\textsuperscript{14} The rate of each reaction shows a decreasing exponential waveform, suggesting that the driving force for both reactions is proportional to the deviation from an altered postural equilibrium.\textsuperscript{14,20}

That DVD is a dorsal light reflex should not be taken to imply that it is also dependent on the direction of incoming light. von Holst observed that a fish that has had bilateral labyrinthectomy and 1 eye removed will commence permanent rolling rather than orient the remaining eye with reference to the light source.\textsuperscript{16} This experiment demonstrates that the dorsal light response is a tropotactic response (ie, one that functions to reestablish binocular equilibrium rather than to directionally orient an eye toward incoming light) rather than a telotactic response (ie, a direct orientation toward or away from light.

The dorsal light reflex bears a profound resemblance to DVD (Table 1). (For purposes of comparison, dorsal can be conceptualized as the direction from which vertical sunlight illuminates the labyrinths in upright fish and humans.) In fish, the amplitude of the dorsal light response increases with the intensity of illumination. In DVD, the finding of variable vertical amplitudes in the 2 eyes must also be a function of the degree of visual input asymmetry, as evidenced by the Bielschowsky phenomenon in which the amplitude of the DVD can be titrated by placing filters of varying density before the fixating eye.\textsuperscript{2} von Holst devoted considerable discussion to the phenomenon of Umstimmung (change of bias), which is the variability of response from one trial to the next depending on internal and external factors (length of time in darkness, mood, hunger, immediate visibility of prey).\textsuperscript{16,19}
without the necessity of maintaining bilateral balance). In humans with DVD, diffusion of light into 1 eye can produce a hyperdeviation similar to that which occurs during occlusion of that eye. That DVD persists in the supine position suggests that the dorsal light reflex in humans and lower animals functions as a binocular disparity signal that produces a preprogrammed neural output to the extracocular muscles. This neural output retains its innervational characteristics regardless of light direction or body position.

The role of the otoliths in inhibiting this visual postural reflex in humans is unknown. The reversal of vertical amplitude asymmetry induced by reclining in a head-hanging position supports the notion that otolithic input can modulate DVD to some degree. Utricular counterbalancing might explain why the higher eye, in which vision is suppressed, eventually descends to its neutral position.

Neuroanatomical studies have shown that direct retinofugal projections to the pretectal accessory optic nuclei and the lateral valvula cerebelli control the dorsal light reflex in goldfish. Unilateral lesions of the ipsilateral pretectal nucleus or lateral valvuli cerebelli selectively abolish responses to light stimulation of the contralateral eye. Bilateral lesions completely abolish this visually guided response, while lesions of the optic tectum have no such effect. In goldfish, only the caudal portion of the lateral valvuli cerebelli receives visual input from the contralateral retina via the ipsilateral pretectal nucleus. The rostral portion receives sensory vestibular (ie, utricular) input, which is integrated with visual input from the caudal portion to maintain optimal roll orientation. The valvula cerebelli has no analogous structure in the mammalian cerebellum, but it is likely that visuo-vestibular information contributes to postural adjustment through the cerebellum.

**Table 1. Similarities Between Dorsal Light Reflex in Goldfish and Dissociated Vertical Divergence in Humans**

<table>
<thead>
<tr>
<th>Dorsal Light Reflex</th>
<th>Dissociated Vertical Divergence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Evoked by asymmetrical light input to the eyes</td>
<td>Evoked by asymmetrical visual input to the eyes</td>
</tr>
<tr>
<td>Visually deprived eye shifts dorsally</td>
<td>Visually deprived eye shifts dorsally</td>
</tr>
<tr>
<td>Tropotactic response*</td>
<td>Probably tropotactic</td>
</tr>
<tr>
<td>Magnitude of response dependent on strength of light stimulus</td>
<td>Magnitude of response dependent on degree of binocular visual disparity</td>
</tr>
<tr>
<td>Magnitude of response dependent on “mood” and external factors</td>
<td>Vertical amplitude variable</td>
</tr>
<tr>
<td>Long latency of vertical reequilibration after prolonged lateral illumination</td>
<td>Slow reversal of hyperdeviation after cessation of monococular occlusion</td>
</tr>
<tr>
<td>Decreasing exponential waveform</td>
<td>Decreasing exponential waveform</td>
</tr>
<tr>
<td>Body tilts in roll plane toward light (ie, toward the side of the preferred eye)</td>
<td>Presence and direction of head tilt variable</td>
</tr>
<tr>
<td>Right utricular output counteracts right eye illumination</td>
<td>Undetermined</td>
</tr>
</tbody>
</table>

* Functions to reestablish binocular equilibrium rather than to directionally orient the eyes toward incoming light.

**IS DVD A DORSAL LIGHT REFLEX?**

How would a dorsal light reflex manifest in humans, who have frontally placed eyes, predominantly binocular visual fields, and stereopsis? Binocular vision must function to suppress this reflex, since a vertical divergence of the eyes would effectively abolish binocularity and stereopsis. When binocularity is poorly developed, however, this primitive reflex can reemerge if binocular visual input fluctuates. If reduced visual input to 1 eye is interpreted by the brain as visual tilt, then the dorsal light reflex should serve to equilibrate visual input between the 2 eyes. For this to be the case, the central vestibular system must interpret decreased visual input to 1 eye as being equivalent to a hypotropia, and respond with a vertical vergence signal. In humans, the oblique muscles have been found to play the predominant role in producing visual vertical divergence movements (Figure 4), as elegantly demonstrated by Enright and elaborated on by others. Clinical observation and scleral search coil recordings in individuals with DVD have confirmed that extorsion and elevation of the deviating eye are accompanied by a reciprocal intorsion and depression of the fixating eye, which conforms to the visual vertical divergence response in humans. The prominent role of the oblique muscles in restoring vertical fusion is evidenced by the dynamic extorsion of the rising eye that accompanies DVD, as well as the horizontal divergence of the nonfixating eye, which reflects the tertiary abducting function of these muscles.

In the case of DVD, reduced visual input in the left eye would activate the left inferior oblique and right superior oblique muscles to produce a cyclovertical divergence movement (Figure 5). Contraction of these muscles produces a clockwise torsional movement of both eyes, together with infraduction of the right eye and supraduction of the left eye. Maintenance of fixation with the right eye would require simultaneous innervation to the elevators (superior rectus and inferior oblique muscles) of the right eye to counteract the infraducting action of the superior oblique muscle. By Hering law, compensatory fixational innervation to the elevators of the fixating eye recruits the same muscles contralaterally, which would augment vestibular innervation to the superior rectus and inferior oblique muscles of the higher eye and actively drive the vertical component of the deviation (Figure 5). Thus, the observed hyperdeviation in DVD is the composite of 2 visual righting reflexes: a visuo-vestibular reflex that activates a torsional divergence of the eyes and a compensatory fixational reflex that maintains fixation with the visually advantaged eye and produces an upward movement of the other eye. The pivotal role of the monococular fixation in DVD explains the nonexistence of simultaneous bilateral DVD and dictates that an upward movement of 1 eye never
begins while the other eye is higher. It is only after the higher eye has completed its descent to the midposition before ascending and extorting.

In both instances, a fixation shift provides a momentary glimpse into an underlying bias in central vestibular tone.

IS DVD A SKEW DEVIATION?

Skew deviation is a descriptive term used to denote an acquired, supranuclear, vertical misalignment of the eyes that fails to conform to known innervational patterns of the extraocular muscles. It is seen primarily in patients with unilateral brainstem lesions, particularly those involving the brainstem tegmentum in the mesodiencephalon or the medulla, although injury to the peripheral vestibular system or the cerebellum can also cause it. It is now accepted that such lesions inhibit or activate unilateral graviceptive output in the roll plane, allowing utricular innervation from one side to predominate, which evokes an ocular tilt reaction. This utricular ocular tilt reaction produces a tonic or paroxysmal vertical divergence of the eyes (ie, skew deviation), which differs from DVD in that the higher eye intorts, the lower eye extorts, and the head tilts toward the lowermost eye

Brandt and Dieterich have determined that skew deviation is usually accompanied by binocular torsion, that both findings are components of the ocular tilt reaction, and that the only difference between skew deviation and an ocular tilt reaction is the presence of a head tilt. This utricular ocular tilt reaction is associated with a pathologic shift in the internal representation of the gravitational vector, and functions as a righting reflex to adjust the eyes, head, and body to a position that the central nervous system erroneously computes as being vertical.

The interplay between visuo-vestibular and fixational innervation explains the perplexing observation that, after removal of a cover from an eye of a patient with DVD, the higher eye will sometimes descend below the neutral position before resuming fixation. A similar phenomenon is observed when occlusion of the fixating eye induces a downward refixation movement in the hyperdeviated eye, and the covered eye makes a simultaneous downward movement below midposition before ascending and extorting.

Figure 4. Vertical divergence of the eyes in DVD. Vertical divergence produced by decreased visual input to the right eye (A) and left eye (B) show opposite vertical and torsional components. These divergence movements enable normal individuals to fuse small vertical image disparities.

Figure 5. Visuo-vestibular and fixational innervation in dissociated vertical divergence with hyperdeviation of the left eye. In accord with the dorsal light reflex, decreased visual input in the left eye is equivalent to a hypotropia, which activates a vertical vergence mechanism to equilibrate the 2 eyes. (Arrow size denotes relative magnitudes of innervation.) A. In humans, vertical vergence is mediated primarily by the oblique muscles, causing contraction of the inferior oblique muscle on the left and the superior oblique muscle on the right. Visuo-vestibular innervation, if unaffected by fixational innervation, would cause simultaneous elevation and extorsion of the left eye and depression and intorsion of the right eye. B. Compensatory fixational innervation to the elevators is necessary to maintain a steady vertical position of the right eye. By Hering law, fixational innervation to the right eye will augment the effects of vestibular innervation to the inferior oblique and superior rectus muscles of the left eye, actively driving the hyperdeviation. Compensatory fixational innervation negates infraduction of the right eye but leaves it with a small intorsional predominance.

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tial.41 Under physiological conditions, this ocular tilt reaction functions as a righting reflex that enables an organism to maintain gravitational orientation during tilt in the roll plane (as occurs when skiing or motorcycle riding). Tilting in the roll plane evokes a physiological skew deviation in lower animals and, to a lesser degree, in humans (Figure 6).32 However, the purely vertical skew deviation in lower animals is supplanted by a torsional divergence movement in humans, since the eyes are frontally placed.

No physiological position of body tilt evokes DVD, and no neurologic lesion has produced it.40 The intermittent forms of acquired skew deviation that have been documented in patients with midbrain lesions have not been associated with extorsion of the ascending eye or intorsion of the descending eye, as occurs in DVD.30,47-49 Vertical divergence of the eyes with the “inverse” torsional characteristics of DVD is a signature of abnormal binocular vision. Evolutionary conservation of the dorsal light reflex as the visual counterpart of the utricular ocular tilt reaction (Figure 3) would explain the acquisition of these inverse torsional movements in frontal-eyed animals.

In humans, both ocular tilt reactions are conserved as complementary “mirror-image” righting reflexes, as evidenced by the observed extorsion of the rising eye in DVD and the intorsion of the rising eye in the utricular ocular tilt reaction (Table 2). In this context, DVD can be conceptualized as an inverse skew deviation. It is the visual counterpart of neurologic skew deviation; the former is a visuo-vestibular ocular tilt reaction, while the latter is a utricular ocular tilt reaction. An imbalance of visual input from the 2 eyes modulates the afferent limb of DVD, while an imbalance in graviceptive input from the utricles modulates the afferent limb of neurologic skew deviation. If the utricular ocular tilt reaction and the dorsal light reflex function together as complementary righting responses in lower lateral-eyed animals (Figure 3), it is difficult to imagine that one mechanism (the utricular ocular tilt reaction) would be retained to such a degree in frontal-eyed animals and the other (the dorsal light reflex) completely discarded. Indeed, small degrees of DVD can be evoked by monocular occlusion in some normal individuals.42,43

CONCLUSIONS

Dissociated vertical divergence is a dorsal light reflex that utilizes binocular visual input to calibrate central vestibular tone. In lower animals, this dorsal light reflex functions to equilibrate visual input by simultaneously increasing dorsal light input to one eye and decreasing it to the other. When binocular control mechanisms are poorly developed in humans, this dorsal light reflex evokes a phylogenetically newer vertical vergence movement of the eyes to produce an inappropriate vertical divergence with torsional characteristics opposite to those required to neutralize a body tilt (“a righting reflex gone wrong”).

In the upright human, the observed ocular movement is upward because the eye with greater visual input is generally used for fixation and the movement of the visually disadvantaged eye is dorsally directed. The absence of a corresponding hypodeviation on alternate cover testing reflects the instantaneous shift in visual advantage to the uncovered eye that occurs with monocular occlusion. This hypothesis explains the reciprocal nature of the observed hyperdeviation, the dynamic torsion that distinguishes DVD from other forms of skew deviation, and the tight link between DVD and visual fixation.

Table 2. Complementary Ocular Tilt Reactions in Humans

<table>
<thead>
<tr>
<th>Age at onset</th>
<th>Dissociated Vertical Divergence</th>
<th>Acquired Skew Deviation</th>
</tr>
</thead>
<tbody>
<tr>
<td>2-4 y</td>
<td>2-4 y</td>
<td>Any age</td>
</tr>
<tr>
<td>Gradual</td>
<td>Gradual</td>
<td>Acute</td>
</tr>
<tr>
<td>Intermittent</td>
<td>Intermittent</td>
<td>Usually constant</td>
</tr>
<tr>
<td>Extorsion of higher eye, intorsion of lower eye</td>
<td>Extorsion of lower eye</td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>Unilateral utricular, brainstem, or cerebellar lesion</td>
<td></td>
</tr>
<tr>
<td>Undetermined</td>
<td>Variable</td>
<td>Toward side of lower eye</td>
</tr>
<tr>
<td>Variable</td>
<td>Rotated in direction of ocular torsion</td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>Vertical</td>
<td>Seesaw or hemi-seesaw</td>
</tr>
</tbody>
</table>

Figure 6. Utricular ocular tilt reaction in humans. When the eyes are frontally placed, a torsional component becomes necessary to neutralize a tilt in the roll plane. A, Physiologic ocular tilt reaction. A rightward body tilt activates right utricular and inhibits left utricular pathways subserving graviceptive tone in the roll plane, resulting in vertical divergence with conjugate torsion of the eyes and head tilt toward the lowermost eye. B, Normal eye position with head upright. C, Pathologic ocular tilt reaction. A leftward ocular tilt reaction can be caused by an inhibitory lesion of the left utricular pathways or an excitatory lesion of the right utricular pathways.
The existence of DVD provides testimony to the duality of the ocular tilt mechanism in humans, to the interplay between visual feedback and vestibular modulation of extraocular muscle tone, and to the evolutionary role of binocular vision in the suppression of this visuo-vestibular response.

Accepted for publication May 21, 1999.

This study was supported in part by a grant from Research to Prevent Blindness Inc, New York, NY.

I thank John Flynn, MD, Thomas Brandt, MD, and Werner Graf, MD, PhD, for their invaluable assistance, encouragement, and tutelage during the preparation of this article.

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REFERENCES


Correction

Error in Figure Legend. In the Special Article by Brodsky titled “Dissociated Vertical Divergence: A Righting Reflex Gone Wrong,” published in the September issue of the ARCHIVES (1999;117:1216-1218), proper acknowledgment of the source of Figure 2 on page 1218 was inadvertently omitted from the legend. The acknowledgment is as follows: “Reprinted with permission from Graf and Meyer.” Copyright 1983, Springer-Verlag.” The journal regrets the error.