Primary Oblique Muscle Overaction

The Brain Throws a Wild Pitch

Michael C. Brodsky, MD; Sean P. Donahue, MD, PhD

Background: Sensorimotor and orbital anatomical mechanisms have been invoked to explain primary oblique muscle overaction.

Methods: Review of primitive visuo-vestibular reflexes and neuroanatomical pathways corresponding to vestibulo-ocular reflexes, and correlation with known clinical abnormalities in patients with primary oblique muscle overaction.

Results: Bilateral superior oblique muscle overaction, which corresponds to a backward pitch in lateral-eyed animals, can occur when structural lesions involving the brainstem or cerebellum increase central otolithic input to the extraocular muscle subnuclei that modulate downward extraocular muscle tonus. Bilateral inferior oblique overaction, which corresponds to a forward pitch in lateral-eyed animals, may result from visual disinhibition of central vestibular pathways to the extraocular muscle subnuclei that modulate upward extraocular muscle tonus.

Conclusions: Primary oblique muscle overaction recapitulates the torsional eye movements that occur in lateral-eyed animals during body movements or directional luminance shifts in the pitch plane. These primitive ocular motor reflexes become manifest in humans when early-onset strabismus or structural lesions within the posterior fossa alter central vestibular tone in the pitch plane.


Primary oblique muscle overaction is a common ocular motility disorder characterized by vertical incomitance of the eyes in lateral gaze. In primary inferior oblique muscle overaction, an upshoot of the adducting eye occurs when gaze is directed into the field of action of the inferior oblique muscle, producing a greater upward excursion of the adducted eye than of the abducted eye. The opposite occurs in primary superior oblique muscle overaction. Although ductions appear to be normal and there is no evidence of yoke muscle paresis, alternate cover testing discloses a vertical tropia of similar magnitude in the abducting eye. Primary inferior oblique muscle overaction is usually associated with ocular extorsion and V-pattern strabismus, whereas primary superior oblique muscle overaction is usually associated with ocular intorsion and A-pattern strabismus. Superior oblique muscle overaction is often accompanied by other neurologic disease, whereas inferior oblique muscle overaction generally occurs in children who have congenital esotropia but no other overt neurologic abnormalities. Surgical weakening of the overacting oblique muscles improves versions, eliminates the associated A or V pattern, and reduces torsion.

In 1916, Ohm postulated that pattern strabismus and oblique muscle overaction may be due to abnormal vestibular innervation. Almost a century later, a unifying neurologic mechanism to explain primary oblique muscle overaction remains elusive. This ocular motor phenomenon seems to defy fundamental principles of physiology since nowhere else in the body do individual muscles bilaterally overact.

The primary function of the oblique muscles in lower vertebrates such as fish is to counterrotate the eyes torsionally in response to pitch (fore-and-aft) movements of the body. As a fish pitches its body to swim upward or downward, a compensatory "wheel" rotation of the eyes is produced by the oblique muscles in response to vestibular stimulation. The existence of this physiologic oblique muscle overaction in lower animals led us to ques-
The term *tonus* was originally coined by Ewald to describe the state of excitation of a living muscle during rest. In 1977, Meyer and Bullock advanced their tonus hypothesis, which states that neuronal tonus pools within the central nervous system receive multisensory input and that tonus asymmetries between antagonistic pools can produce tonic motor responses. According to this hypothesis, the eyes are not merely sensory organs but components of a multimodally driven tonus pool that calibrates baseline muscle tone (ie, tonus-inducing components of a multimodally driven tonus pool that influence eye position. The primary function of the vestibuloocular system is to maintain eye position and stabilize fixation during head movements. Vestibulo-ocular movements are the most primitive of all extraocular movements. As expounded by Walls, the primitive function of the eye muscles was not to aim the eyes at all. Their original actions were all reflex and involuntary, and were designed to give the eyeball the attributes of a gyroscopically-stabilized ship, for the purpose of maintaining a constancy of the visual field despite chance buffetings and twistings of the animals body by water currents.

In the rabbit, for example, a rightward body tilt along its long axis causes the right eye to be lower in space than the left eye. This tilt elicits a compensatory vertical divergence of the eyes to elevate the right eye and depress the left eye, thereby stabilizing the eyes in space. A pitch forward of the body would produce a compensatory extorsional movement of both eyes.

Now consider the same pitch-down body movement in a rabbit that is fixating with the right eye maximally abducted and the left eye maximally adducted. Since the eyes are laterally placed in the rabbit, this position of gaze would direct the left visual axis anterior to its neutral position and the right visual axis posterior to its neutral position. A forward pitch in the body plane with...
the eyes in this position would tilt the left visual axis to a lower position in space than the right visual axis (Figure 2). This tilt would necessitate compensatory vestibulo-ocular innervation to increase upward tonus in the left eye and increase downward tonus in the right eye, while extorting both eyes in response to the body pitch. Conversely, if the body were pitched back during dextroversion, the higher visual axis of the adducted left eye would necessitate increased downward tonus in the left eye and increased upward tonus in the right eye to stabilize the position of the eyes in space. The necessary vestibulo-ocular movements, which correspond to the vertical divergence in lateral gaze seen in humans with primary oblique muscle overaction, are programmed at an early evolutionary stage to assure stability of the visual field in all fields of gaze. In 1996, Zee formulated this hypothesis to explain how the eyes move downward in space, the left visual axis (which is directed toward the nose) rotates downward, while the right visual axis (which is directed toward the tail) rotates upward (curved arrows). This divergence of the visual axes corresponds to a right hypertropia that must be neutralized by vestibular innervation to elevate the lower left eye and depress the higher right eye. The compensatory vertical divergence for a pitch-forward position corresponds to primary inferior oblique muscle overaction.

In lateral-eyed animals and in humans, the semicircular canals respond to angular acceleration and produce dynamic vestibular plane, a semicircular canal within the labyrinth detects acceleration and sends excitatory innervation to the extraocular muscle(s). Within the brainstem and cerebellum, peripheral vestibular input is summated to produce appropriate innervation to the extraocular muscle subnuclei and maintain the position of the eyes in space (Figure 4). Each anterior semicircular canal provides excitatory innervation to the ipsilateral superior rectus and the contralateral inferior oblique muscles while inhibiting the yoked ipsilateral inferior rectus and contralateral superior oblique muscles (Figure 4). Likewise, each posterior semicircular canal system provides excitatory innervation to the ipsilateral superior rectus and the contralateral inferior oblique muscle while inhibiting the ipsilateral inferior oblique and the contralateral superior rectus muscles. In humans, a pitch-up movement of the head (as occurs when raising the chin) activates both posterior semicircular canals, which send excitatory innervation to both depressors in both eyes. Like their target extraocular muscles, the semicircular canal pathways have a push-pull (yoke) relationship, so that activation of one canal inhibits the antagonist canal. Thus, the pitch-up movement that excites both posterior canals also inhibits both anterior semicircular canals, which send inhibitory innervation to the ocular elevators. The result is an equal contraversive movement of both eyes to adjust for the pitch-up head movement. Injury to or inhibition of anterior canal pathways suberving upward eye movements causes a functional activation of the posterior canal downgaze pathways and produces downward eye movements.

In addition to the semicircular canals, each labyrinth contains otolithic sensors consisting of the utricle and the saccule. While the semicircular canals respond to angular acceleration and produce dynamic ves-
tibuloocular movements, the parallel otolithic system responds to linear acceleration and is sensitive to changes in static head position.19 Damage to the semicircular canal pathways produces phasic ocular deviations and nystagmus, while damage to the otolithic projections corresponding to the semicircular canal pathways causes tonic ocular deviations (strabismus).19,23-25 The otolithic pathways are not as well studied, but are believed to have similar projections to the corresponding canal pathways.19 For the sake of simplicity, we refer to the otolithic pathways corresponding to a particular canal pathway simply as the anterior canal or posterior canal system, recognizing the similarity in projections between the otoliths and semicircular canals.19 The anterior canals receive inhibitory innervation from the cerebellar flocculi, while the posterior canals do not. Thus, a structural lesion or metabolic abnormality that inhibits output from the cerebellar flocculi can also disinhibit the anterior canals, resulting in an upward deviation of the eyes.21 Conversely, bilateral lesions of the ventral tegmental tract or brachium conjunctivum can injure central pathways from the anterior semicircular canals and produce a posterior canal predominance, resulting in tonic downgaze. Maturation of cerebellar floccular inhibition to anterior canal pathways may be dependent on normal visual experience early in life. Ocular stabilization is normally modulated by visual and vestibular input. When binocular visual input is preempted, this multisensory mechanism may fall under greater weight of labyrinthine control, allowing excitatory anterior canal output to predominate.28

SUPERIOR OBLIQUE MUSCLE OVERACTION AND A-PATTERN STRABISMUS IN NEUROLOGIC DISORDERS

A bilateral lesion that injures both anterior canal pathways or disinhibits both posterior canal pathways will increase prenuclear innervation to the superior oblique and
inferior rectus subnuclei, resulting in a posterior canal predominance and increased downward tonus to both eyes. This downward tonus must be overcome by fixational innervation (Figure 6). Since the inferior rectus muscles retain their vertical field of action in abduction while the superior oblique muscles have minimal vertical action in abduction, this downgaze predominance would produce a relative overdepression of the adducting eye in lateral gaze (Figure 7). Activation of both superior oblique muscles produces bilateral intorsion in the primary position and an A pattern due to the tertiary abducting action of the superior oblique muscles in downgaze. In addition, binocular intorsion rotates the inferior rectus insertions laterally and reduces the adducting action of the inferior rectus muscles in downgaze.

The vestibuloocular pathways pass through the posterior fossa and are susceptible to injury when structural abnormalities involve the brainstem or the cerebellum. In children with hydrocephalus and myelomeningocele, the constellation of A-pattern strabismus, bilateral superior oblique muscle overaction, and bilateral intorsion is often associated with tonic downgaze early in life.28-38 Children with myelomeningocele not only have hydrocephalus but also frequently have an associated Chiari II malformation.35,36 Since prenuclear input to the vestibular system from the vestibulocerebellum is primarily inhibitory, bilateral compression of or injury to those vestibulocerebellar pathways activating the anterior canals would disinhibit the posterior canals and increase extraocular muscle tonus in their target muscles.

Previous investigators35-40 have speculated that bilateral superior oblique muscle overaction may be supranuclear or prenuclear in nature, citing the frequency with which it accompanies defective upgaze. Biglan37,38 attributed the overacting superior oblique muscles, A pattern, and chronic downward deviations of the eyes in children with myelomeningocele to defects in the vertical gaze pathways producing either a failure to inhibit the downgaze pathways or excessive stimulation of downward gaze. Acute comitant esotropia caused by neurologic disease such as hydrocephalus or Chiari malformation is often associated with bilateral superior oblique muscle overaction.39 Although orbital anatomical factors have also been implicated as a cause of superior oblique muscle overaction in hydrocephalus,1,29 the high frequency of structural abnormalities within the posterior fossa led Hamed35,36 and colleagues to propose that superior oblique muscle overaction and alternating skew deviation in lateral gaze may share a common neuroanatomical substrate. Recently, Hoyt41 has observed that premature infants with periventricular leukomalacia or intraventricular hemorrhage may initially manifest a tonic downgaze that evolves into an A-pattern esotropia and bilateral superior oblique muscle overaction.

Clinical observations and eye movement recordings have documented abnormal ocular responses to vestibular stimulation in children with strabismus.42-45 Gait and pos-
Primary oblique muscle overaction appears to defy Hering’s law, which dictates that, in any volitional conjugate movement, both eyes receive equal innervation. As summarized by Bielschowsky, “all of the muscles of both eyes always participate in each movement; one half experiences an increase in tonus and the other half a decrease.” This control system optimizes binocular vision in all positions of gaze. Although Hering’s law requires that the ocular motor system synthesize a conjugate signal to the motor neurons involved in the execution of any ocular movement, it should be evident from the previous discussion that equal innervation to any set of vertical yoke muscles would produce dissociated movements of the 2 eyes. To execute conjugate vertical eye movements, the extraocular muscles of both eyes must receive appropriate innervation to move the eyes equally rather than receiving equal innervation.

Hering made reference only to voluntary eye movements as conforming to his law of equal innervation. Since the semicircular canals and their corresponding otolithic pathways segregate innervation to each set of yoke muscles, it is not surprising that dissociated eye movements of central origin are generally associated with vestibular disease. Paradoxically, these dissociated movements may reflect the fact that the vertical yoked muscles receive roughly equal innervation rather than the necessary innervation to rotate the eyes equally in one plane.

Our model of primary oblique muscle overaction as a pitch plane imbalance predicts that oblique muscles overact bilaterally in conjunction with rather than relative to their yoke vertical rectus muscles. In primary gaze, the torsional action of the overacting oblique muscles predominates in both eyes, producing the bilateral extorsion observed in primary inferior oblique muscle overaction and the bilateral intorsion observed in primary superior oblique muscle overaction. When both sets of elevators or depressors receive excessive central vestibular innervation, addition of either eye produces excessive vertical excursion of the adducting eye as it moves into the vertical field of action of the overacting oblique muscle (Figure 7). In this context, an upward tonus imbalance to both eyes manifests as bilateral overrelevation of the adducting eye, and a downward tonus imbalance manifests as bilateral overdepression of the adducting eye. Volitional gaze out of the vertical field of action of the overacting yoke muscles recruits physiologic innervation to counterbalance the vertical tonus imbalance, while gaze into the vertical field of action of the overacting yoke muscles allows this underlying tonus imbalance to predominate, producing the A and V patterns observed clinically (Figure 7). The ocular torsion produced by primary oblique muscle overaction also initiates a cascade of secondary mechanical events, including rotational displacement of the rectus muscle insertions, oblique muscle length adaptation, and mechanical tightening of the oblique muscles, as detailed elegantly by Guyton and Weingarten. These peripheral responses augment the

**Figure 8.** Primary inferior oblique muscle overaction. A, Visuovestibular innervation. Failure to develop normal binocular vision is associated with increased upward tonus to the eyes, perhaps through reduced anterior canal inhibition from the cerebellar flocculi. A central vestibular tonus imbalance corresponding to bilateral anterior canal predominance would produce tonic upgaze, horizontal divergence, and extorsion of the eyes if unopposed by fixational innervation. B, Visuovestibular plus fixational innervation. Fixational innervation recruits equal innervation from the inferior rectus and superior oblique muscles to negate the vertical component of the upgaze bias, and allows the disconjugate extorsional bias to persist. PC indicates posterior canal; HC, horizontal canal; and AC, anterior canal.
overerelevation or overdepression of the adducting eye and the corresponding A and V pattern observed clinically.

This neurologic model would also explain why primary oblique muscle overaction is usually associated with a negative Bielschowsky head-tilt test.1,79 A head tilt to either side recruit ipsilateral otolithic innervation to stimulate 1 of the 2 overacting vertical muscles in each eye while inhibiting the other. The net result for each eye is a minimal change in vertical tonus in the primary position. However, this model would predict that pitching the head forward and backward (ie, a vertical head-tilt test) would superimpose a physiologic tonus imbalance on the underlying central vestibular tonus imbalance in the pitch plane and thereby alter the amplitudes of an existing A or V pattern and the amplitudes of the associated hyperdeviations in lateral gaze. Accordingly, the clinical practice of pitching the patient’s head forward and backward to obtain strabismus field measurements in upgaze and downgaze would augment an existing A or V pattern.

OBLIQUE MUSCLE OVERACTION AND DISSOCIATED VERTICAL DIVERGENCE

Dissociated vertical divergence may coexist with primary oblique muscle overaction.60 Dissociated vertical divergence has been attributed to a central vestibular tonus imbalance in the roll plane induced by fluctuations of binocular visual input.60,61 This hypothesis is based on physiologic studies60,61 in fish that show that unequal visual input to the 2 eyes induces a reflex body tilt in the roll (frontal) plane toward the side with greater visual input. This dorsal light reflex is a balancing movement that uses light from the sky as a visual reference to maintain vertical orientation by equalizing luminance input to the 2 laterally placed eyes. In a vertically restrained fish, unequal visual input induces a vertical divergence of the eyes, with depression of the eye that has greater visual input and elevation of the eye that has lesser visual input. This vertical divergence of the eyes corresponds to the dissociated vertical divergence seen in humans who fail to develop single binocular vision secondary to early-onset strabismus.

In humans with dissociated vertical divergence, suppression or mechanical occlusion of one eye induces upward tonus to the extraocular muscles of that eye and downward tonus to the extraocular muscles of the opposite eye.60,61 Simultaneous recruitment of central vestibular innervation to both elevators in the visually deprived eye has been invoked to explain the spontaneous overerelevation in adduction that can be observed with dissociated vertical divergence, when no V pattern or baseline excursion is present.60 The observation that decreased visual input increases upward tonus to one eye (in the case of dissociated vertical divergence) and to both eyes (in the case of inferior oblique muscle overaction) attests to the retention of primitive vision-induced tonus mechanisms60,61 in humans, and to the atavistic resurgence of these primitive subcortical reflexes when strabismus precludes the development of binocular vision. Our neurologic model of primary oblique muscle overaction as a central vestibular tonus imbalance in the pitch plane complements the recently proposed theory of dissociated vertical divergence as a central vestibular tonus imbalance in the roll (frontal) plane, and begs the question of whether latent nystagmus might be similarly driven by a central vestibular tonus imbalance in the yaw (axial) plane.

NONNEUROLOGIC CAUSES OF OBLIQUE MUSCLE OVERACTION

Most of the mechanisms invoked to explain the existence of A and V patterns with oblique muscle overaction have described orbital anatomical abnormalities that could account for the abnormal movements on a biomechanical basis.63-75 It is beyond the scope of this article to review and critique all of them. In some patients, neurologic and anatomical causes of oblique muscle overaction may coexist. Children with hydrocephalus and tonic downgaze, for example, may also have frontal bossing with anterior displacement of the trochlea, which can increase tension on the superior oblique muscles and produce a mechanical superior oblique muscle overaction.1,29 Recently, Clark76 and Demer77 and their colleagues have used magnetic resonance imaging to demonstrate that heterotopia of extraocular muscle pulleys within the orbits can also produce overerelevation or overdepression of the adducting eye and simulate oblique muscle overaction. Orbital pulley malposition may account for some children who have superior oblique muscle overaction and A-pattern strabismus in the absence of neurologic disease. Since orbital anatomical abnormalities can produce excessive vertical excursion of one or both eyes in the field of action of the oblique muscles, many authorities advocate use of the descriptive terms overerelevation and overdepression of the adducting eye rather than the diagnostic term overaction of the oblique muscles to characterize these movements.1,76

CONCLUSIONS

Lower lateral-eyed animals use light from the sky above and gravity from the earth below as major sources of sensory input to neuronal tonus pools within the central vestibular system. These neuronal tonus pools calibrate extraocular muscle and postural tonus to maintain vertical orientation. In lower animals, oblique muscle tonus is determined by luminance and gravitational input in the pitch plane. In humans, the brain leverages visual and gravistatic sensory input to calibrate extraocular muscle tonus in the pitch plane. Early loss of single binocular vision is treated by the central vestibular system as forward pitch, necessitating increased upward tonus to the extraocular muscles and manifesting as primary oblique muscle overaction. Neurologic lesions within the posterior fossa can produce the opposite central vestibular imbalance, in which a backward pitch evokes increased downward tonus to the extraocular muscles and produces primary superior oblique muscle overaction. This duality reflects an ancestral bimodal tuning of central vestibular output to the extraocular muscles that is subordinate to binocular vision in humans.

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