Tonic Ocular Tilt Reaction
Simulating a Superior Oblique Palsy

Diagnostic Confusion With the 3-Step Test

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Background: The tonic ocular tilt reaction (OTR) consists of vertical divergence of the eyes (skew deviation), bilateral conjugate ocular torsion, and paradoxical head tilt. The head and superior pole of both eyes are rotated toward the hypotropic eye.

Objective: To describe ocular motility and torsion findings in 5 patients with OTRs that mimicked superior oblique palsies (SOPs).

Results: In 5 patients, results of the 3-step test suggested an SOP (bilateral in 1 patient); however, no patient had the expected excyclotorsion of the hypertropic eye. Two patients had conjugate ocular torsion (intorsion of the hypertropic eye and extorsion of the hypotropic eye), and 2 patients had only intorsion of the hypertropic eye. All had other neurologic features consistent with more widespread brainstem disease.

Conclusions: Vertical ocular deviations that 3-step to an SOP are not always caused by fourth nerve weakness. When a patient with an apparent fourth nerve palsy has ocular torsion that is inconsistent with an SOP, OTR should be suspected, especially if vestibular system or posterior fossa dysfunction coexists. The rules for the 3-step test for an SOP may be fulfilled by damaging the otolithic projections corresponding to projections of the contralateral anterior semicircular canal. Because results of the Bielschowsky head tilt test may be positive in patients with the OTR, the feature distinguishing OTR from an SOP is the direction of torsion.

We advocate use of a fourth step—evaluation of ocular torsion—in addition to the standard 3 steps.


The 3-step test was established by Parks\(^1\) to identify the paretic muscle responsible for an acute cyclovertical strabismus. Kushner\(^2\) recognized several pitfalls of the 3-step test and identified skew deviation as 1 disorder that can mimic a fourth nerve palsy.

The ocular counterrolling reflex is the basis for the third component of the 3-step test. When the head is tilted to 1 side, the lower eye supraducts and intorts and the fellow eye infraducts and extorts. This response occurs because of central projections from the semicircular canals and otoliths to the ocular motor subnuclei (Figure 1). For example, head tilt to the left excites the intorters of the left eye (superior rectus and superior oblique) and extortors of the right eye (inferior rectus and inferior oblique) while inhibiting their antagonists (extortors of the left eye and intorters of the right eye). The ocular counterrolling reflex is compensatory only for approximately 10° of roll; vertical and torsional fusional amplitudes are sufficient to prevent diplopia in the normal situation. A patient with a unilateral superior oblique palsy (SOP) develops an ipsilateral hypertropia because the depressing and intorting functions of the superior oblique are diminished (Figure 2). When the patient’s head is tilted toward the paretic side, the hypertropic eye supraducts because the remaining intorter—the superior rectus—is activated, producing elevation. Vertical semicircular canals provide phasic innervation during dynamic head tilting; ocular alignment is then tonically maintained by similar signals from the dependent (lower) otolith.

Skew deviation is a supranuclear or prnuclear vertical strabismus caused by dysfunction of these vertical vestibulo-ocular reflex (VOR) pathways. It occurs with posterior fossa lesions, particularly those affecting the brainstem tegmentum, cerebellum,\(^3,4,8\) and midbrain.\(^5,9\) Such lesions may be structural (stroke, trauma, or tumor), demyelinating, or neurodegenerative, although skew has been described with idiopathic intracranial hypertension.\(^9\) Ocular misalignment in skew
deviation may be comitant or incomitant, may simulate a paresis of an extraocular muscle,11,12 and may alternate with time13 or with gaze direction.14-16

The ocular tilt reaction (OTR)17 is a particular type of skew deviation. It consists of a triad of vertical deviation, head tilt in the direction of the lower eye, and bilateral ocular torsion in the direction of head tilt (Figure 2). The amount of ocular torsion may be symmetrical or asymmetrical. Halmagyi et al18 and others19 noted that a tonic OTR can be produced by injury to the utricle or the vestibular nerve or by stimulating the otoliths,20 utricular nerve,21 semicircular canals,22 or midbrain.23 Like other forms of skew deviation, OTR may be associated with diplopia or signs of brainstem dysfunction, such as pendular nystagmus. Patients with OTR may also perceive the subjective vertical as tilted.

We describe 5 patients with OTRs that simulated SOPs using the 3-step test. In each patient, however, ocular torsion was inconsistent with an SOP. We hypothesize that asymmetrical dysfunction of the otolithic vestibular projections, corresponding to projections of the anterior semicircular canal contralateral to the eye with apparent SOP, is responsible.

**REPORT OF CASES**

**CASE 1**

A 52-year-old man had a history of intractable diplopia after 2 severe closed-head injuries sustained 3 years apart and 6 years before evaluation. He had a left head tilt of approximately 10° and a slight left face turn (Figure 3). Visual acuity was 20/40 OD and 20/60 OS. He had bilateral torsional nystagmus, worse on side gaze. His pupils were normal.

Ocular motility demonstrated +2 overaction of the right inferior oblique with −1 underaction of the right superior oblique (Figure 4).

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12 Δ in left gaze and 6 Δ in right gaze. The right hypertropia was 15 Δ on right head tilt and 5 Δ on left head tilt. However, results of double Maddox rod testing demonstrated 5° of intorsion of the right eye and 5° of extorsion of the left eye. Results of dilated fundus examination confirmed the torsional findings; the right eye was intorted 22° and the left eye was extorted 15° as measured by fundus photography (Figure 5). A small horizontal incomitancy may have explained the face turn. Because of the torsion and nystagmus, the patient was unable to fuse with press-on prisms. Baclofen use did not relieve his oscillopsia.

CASE 2

A 4-year-old boy had a left hypertropia noted by his parents for 2 years but worsening recently. His developmental milestones were delayed. Visual acuity was 20/30 OD and 20/80 OS as a result of strabismic amblyopia. Lid and pupil examination findings were normal, and he was not dysmorphic. He had an intermittent right head tilt.

Results of an ocular motility examination demonstrated a +3 overaction of the left inferior oblique with a −3 underaction of the left superior oblique. He had a comitant 35-Δ esotropia and a 6-Δ left hypertropia in primary position that increased to 12 Δ on right gaze and to 20 Δ on left head tilt (Table 1). The left hypertropia measured 18 Δ on up and right and down and right gaze. At near, he had a 12-Δ left hypertropia in addition to 35 Δ of esotropia. He had no right hypertropia on up and left gaze. Cycloplegic refraction was normal. Dilated fundus examination findings were normal except for moderate intorsion of the left fundus. There was no significant extorsion of the right fundus. A magnetic resonance image of the brain demonstrated old diffuse white matter lesions. Genetic investigation revealed an XXY karyotype (Klinefelter syndrome). Strabismus surgery consisting of bimedial rectus recession and left superior rectus recession brought the eyes into monofixation range and eliminated the objective fundus torsion.

CASE 3

A 76-year-old man developed vertical diplopia after suboccipital craniotomy for a posterior fossa meningioma that had caused imbalance and incoordination. His visual acuity was 20/50 OD and 20/25 OS. He had bilat-

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*OTR indicates ocular tilt reaction; L, left; R, right; RHT, right hypertropia; LHT, left hypertropia; and NA, not applicable.
eral cataracts, more dense on the right. Results of his pupil examination were normal.

The man had a moderate right head tilt. Results of orthoptic examination demonstrated a left hypertropia measuring 14 Δ in primary position that increased on right gaze and left head tilt (Table 1). He did not complain of subjective image tilt but reported 4° of excyclodeviation in the right eye when tested with a double Maddox rod. Results of indirect ophthalmoscopy demonstrated moderate incyclodeviation of the left fundus and moderate excyclodeviation of the right fundus.

Magnetic resonance imaging demonstrated a left infratentorial mass that extended anteriorly to the left cerebellopontine angle and around anteriorly to the brainstem. The mass effect enhanced and extended throughout the left cerebellar hemisphere, suggesting tumor infiltration. Extra-axial compression of the brainstem and midline shift were present.

CASE 4

A 35-year-old man awoke with dizziness and blurred vision 6 weeks before examination. His dizziness improved in 2 weeks, but his blurred vision became frank vertical double vision that varied with gaze. He had unsteadiness and a drunken feeling, slurred speech, and transient difficulty swallowing. He had 2 previous episodes of similar dizziness 3 to 4 years earlier, each lasting only a few days.

His visual acuity was correctable to 20/15 OU with normal color vision and confrontation visual fields. His pupil responses were mildly sluggish, with an equivocal left relative afferent pupillary defect and mild light-near dissociation. Versions were full.

Ocular motility demonstrated a 22 Δ comitant esotropia with a left hypertropia that fulfilled the 3-step criteria for a left SOP (Table 1). He had a right hypertropia on right head tilt suggesting a bilateral SOP, but he had 5° of incyclotorsion of the left eye measured with a double Maddox rod. General neurologic examination findings revealed absent abdominal reflexes, hyperreflexia of the left upper limb with finger-nose ataxia on the left, a wide-based unsteady gait, and an inability to perform tandem gait.

Magnetic resonance imaging demonstrated hyperintense lesions in the lower brainstem, near the region of vestibular nuclei and suggestive of demyelinating disease (Figure 6).

CASE 5

A 47-year-old man complained of intermittent vertical diplopia, progressively worsening, for 5 years. He also noted cycloplegia, intermittent oscillopsia, poor balance, and vertigo. He had no history of trauma. Medical history included childhood polio that had recovered fully. His mother had vertigo, ataxia, nystagmus, and other cerebellar findings, suggesting dominant spinocerebellar ataxia.

Visual acuity was 20/15 OU. Color vision and confrontation fields were normal. The pupils were normal.

Ocular motility was full. He had gaze-evoked nystagmus in all directions. On right and left gaze, the nystagmus had a torsional and downbeating component. A small right hypertropia in primary position increased on left gaze and with right head tilt (Table 1). The right hypertropia was greatest in up and left gaze. Saccadic dysmetria was present, and adducting saccades were slowed bilaterally. Results of double Maddox rod testing demonstrated no cyclotorsion. We did not evaluate the fundi for objective torsion because we were unaware of a relation between skew deviation, ocular torsion, and SOPs when this patient was first seen.

Results of general neurologic examination demonstrated incoordination of both upper limbs with finger-nose ataxia, dysdiadochokinesia, and difficulty with tandem gait. Vestibulo-ocular reflexes were tested by measuring visual acuity during head shaking at 1 Hz and were markedly impaired. Dilated fundus examination findings were normal.

Infrared eye movement recordings demonstrated primary position upbeat nystagmus that increased on up gaze. Recordings also demonstrated small-amplitude left-beating nystagmus primary position and gaze-evoked horizontal nystagmus. Saccades were hypermetric with rebound nystagmus.

Magnetic resonance imaging of the posterior fossa demonstrated marked midline cerebellar atrophy that was most pronounced in the superior vermis.

COMMENT

All 5 patients had orthoptic measurements suggestive of an SOP, but the fundus torsion was inconsistent with that diagnosis in the 4 patients in whom it was evaluated. Patients with a true SOP have extorsion of the hypertropic eye.24 Our patients had intorsion of the hypertropic eye. Our patients had intorsion of the hypertropic eye or extorsion of the hypotropic eye. Surgery for SOP (either a weakening procedure for the antagonist inferior oblique or a strengthening procedure for the parietal superior oblique) would aggravate torsion in these pa-

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tients, and therefore, other surgical methods of addressing torsion should be considered. Thus, determining the direction and pattern of ocular torsion is crucial to establishing the correct diagnosis.

Our patients also had other neurologic signs, although subtle, which were attributable to posterior fossa disease, suggesting that the strabismus was not the result of an isolated fourth nerve palsy. Patients 3, 4, and 5 had poor coordination and balance. Patients 1 and 5 had torsional nystagmus. Thus, findings from a careful history and clinical examination are also important when evaluating ocular motility disorders.

The relationship between skew deviation and ocular torsion is well documented. In 1984, Trobe proposed that patients with skew deviation do not have ocular torsion and that this feature distinguishes them from patients with SOP. However, he excluded patients with positive Bielschowsky head tilt test results, which probably biased the results by excluding those patients with ocular torsion. Halmagyi and Hoyt and others reported that patients with the OTR may have positive head tilts and torsion. Brandt and Dieterich reported some degree of ocular torsion in all their patients with skew deviation, in whom torsion was toward the hypertropic eye when evaluated with fundus photography. Unilateral or bilateral torsion is now considered to be a necessary and sufficient condition for diagnosing OTR. The double Maddox rod, indirect ophthalmoscopy, fundus photography, and evaluation of the subjective visual vertical are all reliable methods of detecting ocular torsion.

In our experience with these patients, the double Maddox rod gives reproducible measurements (within 3°) of subjective ocular torsion. We place the rods at oblique angles and ask the patient to make the rods parallel to each other and to the horizon.

Objective measurements of ocular torsion are difficult to make because of variability in the normal relationship between the optic disc and fovea. Extorsion may often be overlooked because the healthy fundus typically has the fovea lower than the center of the disc by 2° to 7°. However, clear intorsion of the hypertropic eye was evident in 4 of our patients. The fifth patient had subjective torsion that was not quantified. Objective torsion using fundus photography was measured only in patient 1.

We believe that all 5 patients had an OTR caused by unilateral damage to the vertical VOR pathways that subserve the normal ocular counterrolling reflex. These pathways originate in the vestibular end organ (utricle, saccule, and semicircular canals) and travel through the brainstem to the cyclovertical ocular motor subnuclei in the midbrain (Figure 1); their injury causes skew deviation.

Each vertical semicircular canal has an excitatory projection to 1 cyclovertical extraocular muscle subnucleus for the ipsilateral eye and to the corresponding yoke muscle subnucleus for the fellow eye (Figure 1); inhibitory projections travel to the corresponding cyclovertical antagonist muscles. (The horizontal canals project only to the horizontal recti and therefore, are not involved in the vertical VOR.) Projections from the oto-liths (utricle and saccule) converge with the semicircular canal pathways at the vestibular nuclei and ascend together. Otoliths and semicircular canals together provide information for the dynamic VOR, whereas the oto-liths alone predominantly affect the static VOR.

Disruption of the semicircular canal inputs to the cyclovertical extracocular muscle subnuclei will produce an OTR that is phasic and associated with nystagmus. Because all our patients had sustained OTRs, the responsible lesions must have involved the otolithic projections—predominantly the utricular component—that parallel those of the semicircular canals but mediate static vestibular input. Thus, we hypothesize that OTR in our patients resulted from asymmetrical or unilateral damage to the otolithic-ocular projections.

In 1994, Brandt and Dieterich postulated that damage to the anterior semicircular pathways could produce intorsion, but they did not address the mechanism by which it could mimic an SOP. We postulate the following mechanism to explain how, using the 3-step test, OTR could produce an apparent SOP. Step 1: A patient with right-sided otolithic projection damage will have decreased tonic excitatory input to the right superior rectus and left inferior oblique (Figure 1) that results in a right hypotropia (left hypertropia) with incyclotorsion of the left eye. Disinhibition of projections to the right inferior rectus and left superior oblique will worsen the right hypotropia and left incyclotorsion. Step 2: The left hypertropia increases in right gaze because this is the field of action of affected rectus muscles. Step 3: Left head tilt stimulates the left utricle and its excitatory projections to the left superior oblique, left superior rectus, contralateral right inferior oblique, and right inferior rectus. In the absence of compensatory effects from the damaged right utricular projections, a large left-over-right vertical skew (the OTR) occurs, as is seen experimentally with left-sided utricular nerve stimulation. The left eye supraducts and intorts and the right eye infraducts and extorts, worsening the right hypotropia and paradoxical torsion.

Right head tilt fails to excite the damaged right otolithic projections, causing the vertical deviation to be less than with left head tilt. Thus, the net motility disturbance is a left hypertropia that fulfills the 3-step criteria for a left SOP but has paradoxical incyclotorsion of the hypertropic eye. In fact, the “parietic” left superior oblique actually “overacts,” because of the unopposed left-sided utricular excitation, to cause paradoxical intorsion. Related disturbances might cause vertical deviations that mimic inferior oblique palsies or pattern strabismus; we believe we have seen such patients.

Injury to the vertical VOR projections that cause such a skew deviation can occur anywhere from the vestibular end organ to the ocular motor subnuclei in the midbrain. Four of our patients had posterior fossa or brainstem pathology that produced such lesions, but the lesions were widespread and had poor localizing value.

An alternative explanation for the pattern of motility defects in our patients could be a midbrain lesion involving the VOR projections and the adjacent trochlear nucleus. An isolated midbrain skew deviation would produce a relatively comitant vertical deviation, with the hy-
pertropic eye on the side of the lesion. Because the fourth nerve crosses to the other side as it leaves the midbrain, extension of such a lesion into the trochlear nucleus would produce an SOP in the hypotropic eye, but it would be “masked” and, with the 3-step test, seem to be in the hypertropic eye. (We doubt that this is the mechanism responsible for most masked bilateral SOPs, however.) Another alternative explanation in the patient with trauma (patient 1) is a traumatic skew deviation with a separate traumatic SOP in the hypotropic eye.

This report demonstrates that objective assessment of ocular torsion with indirect ophthalmoscopy, the double Maddox rod, or fundus photography is necessary to differentiate SOPs from the OTR because both disorders have similar hypertropias and head tilts (Table 2). Patients with bilateral conjugate ocular torsion toward the lower eye or incyclodivergence in the hypotropic eye probably have skew deviation (Figure 2, lower right) rather than an SOP (Figure 2, middle right). Careful assessment for brainstem signs such as nystagmus is imperative in patients suspected of having an SOP. Objective assessment of ocular torsion should become the “fourth step” to complement the 3-step test. Surgically weakening the inferior oblique (or strengthening the superior oblique) may be contraindicated in these patients because the procedure will have a paradoxical effect on ocular torsion. Whether this iatrogenic worsening of torsion will be tolerated after surgery is unknown. Surgical treatment based on weakening relatively “overacting” cyclovertical muscles should theoretically be more helpful and was successful in 1 of our patients.

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