Vertical Strabismus

Diagnosis From the Ground Up

In this issue of *Archives*, Parulekar et al[1] document the resolution of ocular torsion and skew deviation when patients are examined in the reclined position. While some of its methodology may strike the general reader as abstruse and academic, this study is original, clever, and innovative in its clinical application.

Until recently, ophthalmologists and neurologists lacked a mechanistic understanding of skew deviation. The term was generally used to describe a comitant vertical deviation that signified major injury to posterior fossa structures.[2,3] Skew deviation differed from other forms of vertical diplopia in that its size generally remained the same in different positions of gaze, it was unassociated with a primary or secondary deviation, and it did not change with head tilt.[4] As such, it was considered a diagnosis of exclusion that was confined to neurologic patients.

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Our concept of skew deviation was revitalized when Brandt and Dieterich[5-9] published several seminal articles that redefined this entity by characterizing its pathophysiology. They found that skew deviation results from a central or peripheral disruption of the vestibular pathways that transmit input from 1 utricle. The resulting asymmetry in utricular input simulated the imbalance in utricular tone that results when the head is tilted in space. Thus, if the left utricular pathways are injured, the right utricular pathways will be selectively excited, as they would if the patient were tilted to the right. Subjectively, this disturbance manifests as a leftward tilt of the visual world, evoking a compensatory triad of skew deviation, binocular torsion, and head tilt (termed *ocular tilt reaction*).[5-9] This oculocephalic synkinesis strives to realign the interpupillary axis of the eyes, the torsional position of the eyes, and the position of the head to an orientation that the patient erroneously perceives as vertical.[5-9] Evolutionarily, this elaborate cyclovertical deviation of the eyes is derived from the purely vertical skew deviation (depression of the higher eye, elevation of the lower eye) that compensates for postural tilt in lateral-eyed animals.[10] Following the studies of Brandt and Dieterich, skew deviation could be viewed as part of a more generalized dystonia.

Although most patients with skew deviation have a negative Bielschowsky head tilt test result, Donahue et al[11] recognized that this is not always the case and showed that some patients with skew deviation can simulate superior oblique palsy on the Parks-Bielschowsky 3-step test. They found intorsion of the higher eye to be a distinguishing clinical feature of skew deviation. This observation added a fourth step (examination of ocular torsion) to our clinical armamentarium. In retrospect, there is little doubt that many patients with ocular tilt reaction received diagnoses of superior oblique palsy based on their hypertropia and their head tilt to the side of the lower eye. But these 2 disorders are profoundly different. In skew deviation (a prenuclear disturbance), the head tilt is not compensatory for the vertical diplopia; rather, both are compensatory for a tilted visual world. In superior oblique palsy (a nuclear or infranuclear disturbance), a head tilt is used by a patient to evoke unequal utricular stimulation and thereby compensate for a vertical deviation.[4,12]

In the present study, Parulekar et al[1] show that skew deviation and ocular torsion disappear or diminish in the supine position, while the vertical deviation of superior oblique palsy does not change. At first glance, these findings may appear to conflict with a classic study by Sydnor et al,[13] which showed that the head tilt associated with traumatic superior oblique palsy resolves in the supine position. Pediatric ophthalmologists have since used this history to help differentiate congenital superior oblique palsy from congenital muscular torticollis by asking parents if their child's head tilt disappears during sleep. However, the authors provide a unifying explanation for all of these phenomena. They propose that head-dependent changes in torsion and vertical misalignment may be due to decreased activity of the utriculo-ocular reflex. In the patient with skew deviation, the baseline utricular imbalance that drives the cyclovertical deviation dissipates in the supine position. This mechanism also explains why the patient with superior oblique palsy can no longer use a head tilt to access compensatory utricular innervation to assist with vertical fusion in the supine position. Dissociated vertical divergence does not cause vertical diplopia, so it rarely produces diagnostic confusion. However, its persistence in the supine position[14] reflects the fact that it is driven directly by unequal visual input and not by the utricles.[15] It all fits together nicely.

But how does this supine cancellation effect occur? The answer to this question is a matter of speculation. The otoliths consist of the utricle and the saccule, which serve complementary roles in sensing gravitational and other linear forces applied to the head.[16] The utricular macula lies on the floor of the utricle, approximately in the plane of the lateral semicircular canal. The saccular

macula lies on the medial wall of the saccule, nearly parasagittal with respect to the head, and approximately orthogonal to the utricular macula.[16] The utricle is oriented to respond best to tilt (roll) and pitch (fore-and-aft) movements and side-to-side translations of the head. The saccule is oriented to respond best to up-and-down translations of the head.[16] In the upright position, the saccular hair cells are more stimulated by gravity, while in positions of pitch or tilt, the utricular hair cells are more stimulated.[17] In contrast to the semicircular canals, determining the appropriate response to a head translation, which is mediated by the otoliths, is computationally and behaviorally complex.[18] At the neurophysiologic level, it is not known what happens to the firing properties of regional otolithic afferents in different body positions or how this peripheral input is integrated centrally. It is certainly possible that saccular afferents help to modulate the supine cancellation effect observed in skew deviation. New observations create the feedback loop wherein clinical discovery redirects basic neurophysiologic investigation.

One methodological limitation of this study is that it uses a subjective response to estimate objective torsion. Double Maddox rods measure subjective visual tilt, while examination of retinal photographs are necessary to quantitate objective torsion.[5] Because ocular torsion (the compensatory response) should reduce the subjective visual tilt (the stimulus), there is no a priori reason why these 2 functions should be identical. Several studies have found that the measured deviations in subjective visual vertical and objective torsion are always ipsidirectional but rarely in lockstep.[5,19] However, retinal photographs from 1 patient with skew deviation shows resolution of binocular torsion in the supine position, supporting the authors' conclusion that this torsion indeed dissipates.

These findings raise the question of whether infants with esotropia should ideally be examined in the upright position. They also caution that it may be advisable to sit adults up when performing intraoperative suture adjustment during strabismus surgery. For future study, it will be interesting to investigate how prone positioning affects skew deviation and its associated ocular torsion. Marti et al[20] found the ocular drift velocity in cerebellar downbeat nystagmus to be minimal in the supine position and maximal in the prone position. Using the analogy of an airplane, in which activation of one brake flap causes the plane to tilt, while activation of both brake flaps results in downward pitch, Brandt and Dieterich[21] postulated that downbeat nystagmus may be a form of bilateral ocular tilt reaction in which the vertical components summate to produce the slow phase vertical drift in both eyes and the torsional components cancel. If this analogy holds, one would predict that skew deviation would also be maximal in the prone position.

Parulekar et al have discovered another critical step for distinguishing skew deviation from superior oblique palsy. More importantly, they have shown us that we are missing a lot by focusing on the effects of upright head tilt in the diagnosis of vertical diplopia. For skew deviation, the answer seems to be found not in tilt but in pitch. By incorporating supine examination into our clinical armamentarium, we can peer further into the crystal ball and determine whether a utricular imbalance is causing or compensating for the vertical deviations that produce diplopia. In the patient with vertical diplopia, it can be inferred that any cyclovertical strabismus that resolves in the reclined position indicates a prenuclear utricular disturbance, while one that persists in the reclined position indicates a nuclear or infranuclear disturbance. The authors are to be congratulated for a seminal study that opens up a new dimension into the clinical diagnosis of vertical strabismus.

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