Skew deviation

Three dimensions of skew deviation

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Are all skew deviations the clinical expressions of central vestibular dysfunction in three dimensional space?

Skew means set, placed, or running obliquely; slanting. For a century, skew deviation has been the descriptive term for an acquired vertical deviation that slants the interpupillary axis to an oblique orientation. This condition has traditionally connoted a severe and debilitating neurological injury within the posterior fossa. Subsequent investigation has refined our understanding of this disorder and expanded the boundaries of this diagnosis. Working over the past two decades, the promethean team of Brandt and Dieterich have established that skew deviation has a clinical localising value within the posterior fossa; that it is a component of the ocular tilt reaction (a triad of skew deviation, binocular torsion, head tilt); and that it is associated with a tilt in the subjective visual vertical. Brandt and Dieterich have supplanted our basic descriptive definition of skew deviation with a mechanistic understanding of this complex disorder.

The underlying mechanism for skew deviation involves a unilateral lesion that inhibits (or occasionally stimulates) central otolithic pathways which run from the medulla to the mesencephalon. Since a physiological imbalance in central graviceptive tone is normally produced by a head or body tilt in the roll plane, this lesion produces the sensation of body tilt and evokes the same stimulus as is evoked by the Bielschowsky head tilt test. By activating prenuclear input to the superior rectus and superior oblique muscles of the ipsilateral eye, and the inferior rectus and oblique muscles of the contralateral eye, and by simultaneously inhibiting their antagonists, a cyclovertical divergence is produced, with intorsion of the higher eye, extorsion of the lower eye, and a corrective head tilt toward the side of the lower eye. This constellation of abnormalities is termed an ocular tilt reaction, which is considered to be compensatory for a tilt in the subjective visual vertical (that is, a perceived tilt of the body relative to the visual environment). Thus, even with the head held in its tilted position, the patient with an ocular tilt reaction perceives it to be upright. The beauty of this re-equilibration lies in way that components cancel each other out but the vertical components summate.

We have proposed that this summation can produce a bilateral alternating skew deviation, in which patients exhibit an alternating hypertropia of the abducting eye on gaze to either side. Because the otolithic pathways corresponding to the anterior and posterior canals are segregated within the brainstem, bilateral lesions tend to selectively affect those otolithic pathways corresponding to one set of canals. A bilateral lesion causing alternating skew deviation may selectively damage the otolithic pathways corresponding to the anterior canals, thereby activating the posterior canal otolithic pathways (which stimulate the superior oblique and inferior rectus muscles bilaterally). Since the superior oblique muscles are infrauctors in adduction and extorters in abduction, there is greater infraction of the adducting eye, producing a pattern of alternating hypertropia of the abducting eye. Conversely, a bilateral injury to otolithic pathways corresponding to the anterior canals would produce alternating hypertropia of the abducting eye.

If skew deviation can reflect a roll or pitch imbalance, can other prenuclear injury to the central vestibular pathways similarly alter horizontal alignment of the eyes? The answer is yes. Acquired comitant esotropia can be produced by structural lesions localised to the posterior fossa such as cerebellar vermal tumours or Arnold Chiari malformations. Since these prenuclear lesions presumably affect central vestibular subserving horizontal rather than vertical vergence, the resulting acquired comitant esotropia can be classified as a horizontal skew deviation. Although these cases do not conform to our original descriptive definition of skew deviation (requiring an oblique or slanted orientation of the visual axes), this nosology recognises a mechanistic overlap with other forms of skew deviation (Table 1).

The physiological underpinnings of horizontal skew deviation may lie in the translational vestibulo-ocular reflex. Primates have developed highly specialised vestibular mechanisms capable of eliciting robust short latency eye movements, known as translational vestibulo-ocular reflexes in response to linear head movements. These reflexes optimise binocular gaze stability during linear disturbances. In any different heading direction, vestibular signals from the otolith organs of the inner ear encode the direction of the movement and transmit that information to the brainstem, including the motor-neurons innervating the eye muscles. This information is used to generate conjugate version movements (during
lateral translation) and disconjugate vergence movements (during fore and aft translation) that serve to reduce retinal image slip and provide stable processing of the visual surround.\textsuperscript{21-25} According to Miles,\textsuperscript{25} these vestibularly driven eye movements use radial optic flow and binocular disparity vergence to minimise head movement.

Optic flow and binocular disparity vergence is of the order of seconds of arc.\textsuperscript{22} Convergence alone or with convergence can be important for the maintenance of the fixation plane.\textsuperscript{24} Convergence may also influence horizontal eye position. In rabbits, the cerebellar flocculus has an inhibitory projection to the medial rectus but not the lateral rectus muscles, so that a lesion in the flocculus could lead to a convergence bias.\textsuperscript{30} Some cells in the flocculus discharge in relation to the angle of vergence.\textsuperscript{15} Nevertheless, the selective motor effects of bilateral flocculocytome and paraflocculocytome on horizontal alignment have not been defined. Future study of the perceptual correlates of bilateral alternating skew deviation and acquired comitant esotropia should help to determine whether these vertical and horizontal vergence abnormalities indeed reflect central disturbances in pitch and surge, and whether all skew deviations are the clinical expressions of central vestibular dysfunction in three dimensional space.\textsuperscript{21}

**Table 1** The three dimensions of skew deviation

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<th>Laterality</th>
<th>Pathways</th>
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