THE OCULAR TILT REACTION AND SKEW DEVIATION

D. Zackon
Ottawa

The ocular tilt reaction composes a triad of ipsilateral head-tilt, ipsilateral conjugate ocular torsion, and skew deviation with ipsilateral hypotropia. It results from a disruption of central vestibular pathways conveying information from the semicircular canals and otoliths. Skew deviation is one component of the syndrome which may or may not be associated with the concomitant torsion and head-tilt of the ocular tilt reaction. Because of the frequent clinical presentation of skew deviation it merits special attention.

Westheimer and Blair (1975) produced the ocular tilt reaction in monkeys by unilateral electrical stimulation of a region of the brainstem tegmentum dorsolateral to the oculomotor and trochlear nuclei from the caudal wall of the third ventricle to the rostral floor of the fourth ventricle. This area encompasses, but does not consist entirely of, the interstitial nucleus of Cajal. They observed 1) a skew deviation of the eyes resulting from an equal and opposite movement of the globes, 2) a conjugate torsional movement ipsilaterally to the stimulated side, and 3) an ipsilateral head tilt (Westheimer and Blair, 1975). Generally the eye ipsilaterally to stimulation becomes hypotropic. Rarely the opposite is seen with contralateral hypotropia and contralateral torsion. Further experimental studies in animals (Hyde and Eason, 1959; Hyde and Toczek, 1962; Mabuchi, 1970) and in humans (Sano et al, 1972) have confirmed that the rostral midbrain and around the interstitial nucleus of Cajal is the crucial area for eliciting the ocular tilt reaction.

The presence of ocular counter-roll in humans, although disputed in the past, has unquestionably been confirmed. In an elegant study utilizing the search coil technique, Collewijn et al (1985) documented the degree of counter-roll present under static conditions, during a roll movement of the head, and under conditions of voluntary pseudo-sinusoidal head rotation. Confirming previous observations dating back to the 19th century (Nagel 1896) they found static counter-roll to be primarily an otolith function though likely with a small additional input from neck proprioception. Static counter-roll compensates only slightly for the degree of head rotation in roll with a gain on the order of ten percent. Gain is increased during an active step in head roll in which the torsional eye movements consist of conjugate smooth cyclorotary movements opposite to the direction of head roll interspersed with conjugate cyclorotary saccades (approximately two to four per second) in the direction of head roll.

As the saccadic eye movements compensate largely, though not fully, for the preceding smooth eye movements, this results in an accumulation of torsional deviation which, at the end of the head roll, represents the static torsional component of counter-roll. Continuous voluntary pseudo-sinusoidal oscillation of the head in roll again shows an increase in gain of the counter-roll with increased frequency of the oscillation and with the amount of visual information. This study documents the progressive increase in gain of counter-roll eye movements secondary to canal input (rotary head acceleration) and visual input.

The role of the utricle in the generation of the ocular tilt reaction has been confirmed by animal studies. (Suzuki et al, 1969; Curthoys, 1987). The utricle projects to both the lateral vestibular nucleus and the superior vestibular nucleus. From the lateral vestibular nucleus there are descending vestibulo-spinal projections presumably contributing to head tilt. An ipsilateral projection from the superior vestibular nucleus follows the medial longitudinal fasciculus and terminates in the ipsilateral trochlear nucleus, the inferior rectus subdivision of the oculo-motor nucleus bilaterally, the ipsilateral interstitial nucleus of Cajal and the rostral interstitial nucleus of the MLF. Contralateral projections of the superior vestibular nucleus terminate in the trochlear nucleus and the superior rectus and inferior oblique subdivisions of the oculo-motor nucleus. There are scattered cross projections to the interstitial nucleus of Cajal. The lateral vestibular nucleus projects ipsilaterally to the oculo-motor nucleus via the ascending tract of Dieters. Brant and Dieterich (1987) propose that the typical eye movements of the ocular tilt reaction may be due to activation of the ipsilateral inferior rectus and inferior oblique (via ipsilateral MLF or ipsilateral ascending tract of Dieters) as well as excitation of the contralateral superior rectus and superior oblique (via the contra-lateral ascending tract of the MLF). It goes without saying that these proposed pathways are purely speculative. The ascending tract of Dieters may in fact not be involved in vertical eye movements.

The ocular tilt reaction has been reported following both peripheral and brainstem lesions. Halmagyi et al (1979) reported an ipsilateral ocular tilt reaction following a destructive lesion of the vestibule. They attributed the reaction to the unopposed action of the normally functioning utricle. Deecke et al (1981) reported the Tullio phenomenon in a patient with an irritative lesion of the vestibule. This patient developed a skew deviation with contralateral hypotropia, counter torsion of both eyes, and a contralateral head tilt when uttering the vowels u or e. They speculated that the otoliths were stimulated by a
hypermobile luxated stapes.

A similar case was described by Dieterich et al. (1988). Sound stimulation in their patient produced a paroxysmal ocular tilt reaction with both phasic and tonic components. Following stimulation, there was an initial rapid and large ipsilateral counter-clockwise rotary upward deviation followed by a smaller tonic effect which persisted as long as stimulation was maintained.

Rabinovitch et al. (1979) were the first to report a paroxysmal ocular tilt reaction in a patient with MS and brainstem demyelination. They postulated either excitation of the contralateral central utricular pathways or inhibition of ipsilateral central utricular pathways. Hedges and Hoyt (1982) reported a patient with a focal rostral midbrain abscess who demonstrated a paroxysmal ipsilateral ocular tilt reaction. How this lesion may have resulted in stimulation of ipsilateral central utricular pathways is unknown.

Brandt and Dieterich (1988) described four patients with Wallenberg's syndrome who demonstrated a tonic ipsilateral ocular tilt reaction with contralateral hypertropia, ipsilateral head tilt, and monocular excyciotropia of the ipsilateral hypotropic eye. In addition, three patients with upper brainstem midbrain lesions were described with an ipsilateral tonic ocular tilt reaction. All seven patients demonstrated a deviation of the subjective visual vertical towards the side of head tilt. Such deviation is presumed to reflect a deviation of the internal representation of the gravitational vector. They explained the ipsilateral hypotropia and excyciotropia in Wallenberg's syndrome by interruption of ascending posterior canal excitatory input to the ipsilateral superior oblique muscle and ascending inhibitory input to the contralateral inferior rectus. The three patients with midbrain lesions showed no resolution of the ocular tilt reaction whereas patients with the lateral medullary syndrome showed a gradual resolution over days to months.

Morrow and Sharpe (1988) described three patients with lateral medullary syndrome who manifested ipsilateral hypotropia and torsional nystagmus. The multiple areas involved in interruption of the lateral medulla include the vestibular cerebellum, caudal portion of the vestibular nuclei, and connections between the vestibular nuclei and the cerebellum. The complexity of the lesion precluded identification of the specific pathways involved in torsional nystagmus.

Morrow and Sharpe felt that the ocular tilt reaction in their patients represented an imbalance of central projections from the vertical canals and otoliths. A saccadic bias torsional pulsion analogous to the lateropulsion of vertical and horizontal saccades in the lateral medullary syndrome was seen in two of their patients. Roper (1983) reported a series of five patients, one with a labyrinthine lesion and four with pontine or ponto-medullary lesions, who experienced episodic tilting of the visual environment. These episodes were brief - usually one to five seconds in duration. There was no predictable relation between the direction of visual tilting and the side of the brainstem lesion.

Although the neuro-ophthalmic exam is not detailed in their cases it appears that at least two of their cases had skew deviation. There was no mention made regarding possible ocular torsion or head tilt. He suggested that lesions of the otoliths or their central connections were responsible for the visual tilting and suggested the term tortopia be used to describe the symptom of tilting of the visual environment.

Slavin and LoPinto (1987) reported a patient with left medullary compression secondary to a dolichoectatic vertebral artery who experienced ipsilateral tilt of the environment associated with contralateral ocular torsion affecting both eyes. The environmental tilt was present under both monocular and binocular conditions. There was no head tilt. There was no skew deviation. They raised the question as to whether there is a separation of function of central utricular projections governing position of the head with respect to the gravitational vector, vertical alignment of the eyes, and alignment of the globes.

**Skew Deviation**

Skew deviation refers to an acquired vertical dissociation of the eyes of supranuclear etiology. It was first described by Magendie in 1825 and later reported in the cat by Hertwig in 1826 following an incision coming in laterally from a cerebellar hemisphere toward the middle of the medulla. Skew deviation has also been produced experimentally in animals with lesions of the middle cerebellar peduncle, the cerebellum, the restiform body, the inferior olive, and the vestibular nucleus outflow pathways. As pointed out by Cogan (1956) however, one cannot generalize from animals with laterally placed eyes to man in that skew deviation is a normal otolith response to head tilt in the animal with laterally placed eyes.

The first large series of cases of skew deviation was published by Keane (1975). In an analysis of one hundred cases of skew, approximately sixty percent had evidence of pontine damage, twenty percent had evidence of midbrain or pretectal lesions, and twenty percent demonstrated medullary lesions. Patients with lower brainstem lesions were somewhat more likely to show minor vertical separation which varied with head position but there were no hard differential features of skew resulting from lesions at various levels of the brainstem. Patients with internuclear ophthalmoplegia are more likely to show an ipsilateral hypertropia. Patients with lateral medullary lesions are more likely to show an ipsilateral hypotropia. Having said this however there are exceptions and skew deviation secondary to brainstem lesions is not of reliable value in lateralizing the lesion.

Skew deviation has also been reported secondary to increased intracranial pressure. Baker and Buncic (1985) reported three cases of presumed skew deviation as a nonlocalizing sign of increased intracranial pressure in pseudotumor cerebri. Skew deviation secondary to increased intracranial pressure had previously been reported by Merikangas (1978) and by Keane (1975). Frohman and Kupersmith (1985) reported a further five patients with skew deviation - three with cerebral tumors and two with benign intracranial hypertension. They proposed that the skew deviation resulted from the effect of increased intracranial pressure on the labyrinth or its' central projections.

As is the case with ocular bobbing, all sorts of weird
Paroxysmal skew deviation was first described by Alter and Hoyt (1962) in a patient with a brainstem glioma. Hedges and Hoyt (1983) reported paroxysmal skew deviation as part of the ocular tilt reaction in a patient recovering from a zona incerta brain abscess. Fixation suppressed any vertical movement of the fixating eye. Slowly alternating skew deviation was described by Corbett et al. (1981) in three patients who also demonstrated rotary nystagmus and a partial dorsal midbrain syndrome. It was characterized by an alternating vertical dissociation in which one eye rose as the other fell. Positions were maintained for thirty to sixty seconds with a transition period lasting from ten to thirty seconds.

Periodic alternating skew deviation was described by Mitchell et al. (1981). In this case the transition phase lasted approximately ten seconds. The duration of the right hypertropia was four to five minutes while that of the left hypertropia was forty to fifty seconds. The skew was comitant in both phases. This patient had a presumed infarct of the interstitial nucleus of Cajal. Lewis and Kline (1983) reported a patient with cerebellar degeneration who had both periodic alternating nystagmus and periodic alternating skew deviation. In this patient the right-beating nystagmus was associated with a comitant right hypertropia while the left-beating nystagmus was associated with a comitant left hypertropia. They attributed the disorder to disruption of cerebellar inhibitory input to the vestibular nuclei. Greenberg and Dewitt (1983) reported a case of periodic non-alternating skew deviation in a patient with an infarct in the region of the interstitial nucleus of Cajal.

This patient had a paroxysmal skew deviation, unaffected by fixation, associated with an ipsilateral head tilt and an intermittent counterclockwise torsional nystagmus - both of which were present only during the period of skew. The skew deviation lasted two to three minutes and alternated with one to two minute period of orthophoria.

REFERENCES