

Skew deviation

Skew deviation may be a unilateral variant of [Parinaud's Syndrome](#).

Skew deviation is an acquired vertical misalignment of the eyes resulting from asymmetric disruption of supranuclear input from the otolithic organs (the utricle and saccule of the inner ear, both of which contain otoliths, which are tiny calcium carbonate crystals). These organs sense linear motion and static tilt of the head via gravity and transmit information to the vertically acting ocular motoneurons, as well as to the interstitial nucleus of Cajal (INC), all of which are in the midbrain.

Both peripheral and central lesions can produce skew deviation. Central causes of skew deviation are more common and can occur anywhere within the posterior fossa (brainstem and cerebellum). Skew deviation can be comitant or incomitant, and torsional abnormalities may be present. At times it may be difficult to distinguish some presentations of skew deviation from a fourth nerve palsy, and proper use of the Parks-Bielschowsky 3-step test (discussed later in the chapter) is essential. Skew deviation often produces diplopia and is an important exception to the general rule that supranuclear lesions do not produce double vision.

An alternating skew deviation on lateral gaze usually manifests as hypertropia of the abducting eye (ie, right hypertropia on right gaze) that switches when gaze is directed to the opposite side (ie, becomes left hypertropia on left gaze). Responsible lesions are located in the cerebellum, cervicomedullary junction, or dorsal midbrain. This disorder must be distinguished from bilateral fourth nerve palsies, which differ in that they produce a hyperdeviation that increases on gaze to the opposite side (eg, right hypertropia is larger on gaze to the left), in addition to excyclotropia.

Ocular tilt reaction is a combination of a head tilt, skew deviation, and cyclotorsional abnormalities of both eyes that can occur in tonic or, rarely, in paroxysmal fashion. This syndrome typically develops because of loss of otolithic input to the INC from a central lesion, which may be in the medulla, pons, or midbrain. Such a lesion can alter one's sense of true vertical, which in turn drives the head and rotates the eyes toward the same side in a compensatory response to correct to true vertical.

With an ocular tilt reaction, if the head is tilted to the left, the right eye is hypertropic, and the upper poles of both eyes rotate toward the lower ear. The opposite response of the eyes is present if the head tilt is to the right. The changes in head and eye position of an ocular tilt reaction should be distinguished from those of a normal response of head tilting as well as from those of a fourth nerve palsy; the normal ocular reflexes induced by tilting the head cause both eyes to rotate toward the higher ear (counterrolling), whereas the ocular tilt reaction causes the opposite response. With a fourth nerve palsy, the compensatory head tilt is typically contralateral to the side of the hypertropia (similar to ocular tilt reaction), but the higher eye is extorted—opposite the pattern of an ocular tilt reaction. Thus, in establishing the diagnosis of ocular tilt reaction, one must attend to both head position and ocular cyclotorsion.

Periodic alternating skew is a rare disorder producing alternating hypertropia, typically with a 30–60 second periodicity, indicative of a midbrain lesion ¹⁾

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Donahue SP , LavinPJ, HamedLM. Tonic ocular tilt reaction simulating a superior oblique palsy: diagnostic confusion with the 3-step test. Arch Ophthalmol. 1999;117(3):347–352.

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