Vestibular disorders in (frontal) roll plane

The clinical signs, both perceptual and motor, of a vestibular tone imbalance in the roll plane include ocular tilt reaction (OTR), ocular torsion, skew deviation, and tilts of the perceived (subjective) visual vertical (SVV). Complete OTR or skew torsion without head tilt indicates either a unilateral peripheral deficit of otolith and vertical semicircular canal input or a unilateral lesion of “graviceptive” brainstem pathways from the vestibular nuclei (crossing midline at the pontine level) to the interstitial nucleus of Cajal (INC) in the rostral midbrain. SVV tilts are the most sensitive clinical sign of an acute unilateral brainstem infarction and signify a vestibular tone imbalance in roll. They occur with peripheral or central vestibular lesions from the labyrinth to the vestibular cortex.

All tilt effects – perceptual, ocular motor, and postural – are ipsiversive (ipsilateral eye undermost) when unilateral peripheral or pontomedullary lesions occur below the crossing of the graviceptive pathways. They are contraversive (contralateral eye undermost) in cases of unilateral pontomesencephalic brainstem lesions, and indicate involvement of the medial longitudinal fasciculus (MLF) or the rostral midbrain integration centres of the VOR in roll and pitch planes (INC; riMLF). Vestibular disorders in the roll plane may be distinguished by the predominant perceptual or motor signs and symptoms.

It is also possible to relate the clinical syndrome to a particular brainstem level, the lesioned vestibular structures, and the involved reflex (VOR) or integration (INC) mechanism. Three types of vestibular disorders in roll can be thus described (see also p. 181):

1. the “ascending” type of VOR-OTR with pontomedullary lesions of the medial or superior vestibular nucleus or graviceptive pathways which subserve the VOR in the roll plane,
2. skew torsion without head tilt, with unilateral lesions of the pontomesencephalic “graviceptive” pathways, and
3. the “descending” type of integrator-OTR with lesions of the rostral midbrain integration centres for eye-head coordination in the roll and pitch planes.

The “ascending, reflexive” type (lateral medulla) of OTR simply reflects a tone imbalance of the VOR, whereas the “descending integration” type (rostral midbrain tegmentum) integrates signals of eye-head velocity with those of holding position and involves cortical control of the fundamental pattern of OTR during active locomotion. The (pontomesencephalic) “skew torsion” type results from lesions of ascending “graviceptive” pathways but does not cause head tilt, because vestibulospinal tracts are not involved. Unilateral infarctions of the brainstem (see Chap. 19) cause dysfunction predominantly in the roll plane, less frequently in the yaw and pitch planes. Dysfunction in the pitch plane requires bilateral vestibular dysfunction.

Unilateral lesions of vestibular structures rostral to the INC typically manifest as deviations of perceived vertical without concurrent eye-head tilt. OTR in unilateral paramedian thalamic infarctions indicates simultaneous ischaemia of the paramedian rostral midbrain including the INC. Unilateral lesions of the posterolateral thalamus can cause thalamic astasia and moderate ipsiversive or contraversive SVV tilts, thereby indicating involvement of the vestibular thalamic subnuclei. Unilateral lesions of the parietoinsular vestibular cortex cause mostly contraversive SVV tilts. An SVV tilt found with monocular but not with binocular viewing is typical for a trochlear or oculomotor palsy rather than a supranuclear graviceptive brainstem lesion.

“Graviceptive” input converging from otoliths and vertical semicircular canals subserve vestibular function in the roll plane. A unilateral lesion of these pathways may result in static and dynamic effects, e.g., torsional nystagmus. Several distinct and separate lesions have been associated with torsional nystagmus: lesions of the vestibular nuclei, the lateral
The clinical syndrome

The “graviceptive” input from the otoliths converges with that from the vertical semicircular canals at the level of the vestibular nuclei (Angelaki et al. 1993) and the ocular motor nuclei (Baker et al. 1973; Schwindt et al. 1973) to subserve static and dynamic vestibular function in pitch (up and down in the sagittal plane) and roll (lateral tilt in the frontal plane). In the “normal” position in the roll plane, the subjective visual vertical (SVV) is aligned with the gravitational vertical, and the axes of the eyes and the head are horizontal and directed straight ahead.

Signs and symptoms of a vestibular dysfunction in the roll plane can be derived from the deviations from normal function. A lesion-induced vestibular tone imbalance should result in a syndrome consisting of a perceptual tilt (SVV), head and body tilt (OTR, lateropulsion), vertical misalignment of the visual axes (skew deviation), and ocular torsion (Fig. 10.1). This has been demonstrated in animal experiments by unilateral stimulation of the utricular nerve (cat: Suzuki et al. 1969), the utricular macula (guinea-pig: Curthoys 1987), and vertical canal nerves (cat: Cohen et al. 1964; Tokumasu et al. 1971), which resulted in either a complete ocular tilt reaction (OTR), i.e. the triad of head tilt, skew deviation, and ocular torsion, or in its single components. In humans, inadvertent damage to one utricle (Halmagyi et al. 1979) or inappropriate stimulation of the otoliths in patients with a Tullio phenomenon (Dieterich et al. 1989; p. 108) also caused OTR.

Not only the complete synkinesis of OTR but also one of its components (postural or perceived tilt, ocular torsion, or skew deviation) indicates a dysfunction in the roll plane. Skew deviation appears primarily as an ocular motor misalignment of the visual axes in the vertical plane, but its regular association with ocular torsion and SVV tilt (Brandt and Dieterich 1993) makes a vestibular roll plane dysfunction most likely.

We can establish clinical rules, which help our basic understanding and diagnostic routine, although exceptions may prove them inaccurate. The only difference between the ocular skew-torsion sign and OTR is head tilt. Whereas skew deviation does not manifest without ocular torsion (exceptions will probably be found in the future), monocular or binocular torsion is frequently seen without concurrent skew deviation. Finally, perceived vertical may be tilted with or without concurrent skew deviation, ocular torsion, or head tilt (Fig. 10.1, Table 10.1).

There is convincing evidence that all following signs and symptoms reflect vestibular dysfunction in the (frontal) roll plane:

- ocular tilt reaction (OTR)
- skew deviation (skew-torsion sign)
- spontaneous torsional nystagmus
- tonic ocular torsion (monocular or binocular), if not caused by infranuclear ocular motor disorders
- tilt of perceived visual vertical (SVV) (with binocular viewing)
- body lateropulsion

Ocular motor or postural tilts as well as misadjustments of subjective vertical point in the same direction, either clockwise or counterclockwise (as seen from the viewpoint of the examiner). The direction of all tilts is reversed if pathological excitation of unilateral “graviceptive” pathways is the cause of vestibular tone imbalance in roll rather than a lesional input deficit. The combination of static and dynamic signs is not surprising if one considers the functional cooperation of otoliths and vertical semicircular canals due to their neuronal convergence within “graviceptive” pathways. The above-listed signs and symptoms may be found in combination or as single components at all brainstem levels. A systematic study of 111 patients with acute unilateral brainstem infarctions revealed that pathological tilts of SVV (94%) and ocular torsion (83%) are the most sensitive signs (Dieterich and Brandt 1993a). Skew deviation was found in one-third and a complete OTR in one-fifth of these patients (see Table 10.1, Fig. 10.2).

Clinical evaluation of vestibular function in roll therefore includes psychophysical adjustments of the SVV, determination of the vertical divergence of the visual axes by means of prisms, and determination of ocular torsion by means of fundus photographs (for methods, see Dieterich and Brandt 1993a).