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Perceived Vertical and Lateropulsion: Clinical Syndromes, Localization, and Prognosis

Thomas Brandt, MD, FRCP, and Marianne Dieterich, MD

We present a clinical classification of central vestibular syndromes according to the three major planes of action of the vestibulo-ocular reflex: yaw, roll, and pitch. The plane-specific syndromes are determined by ocular motor, postural, and perceptual signs. Yaw plane signs are horizontal nystagmus, past pointing, rotational and lateral body falls, deviation of perceived straight-ahead to the left or right. Roll plane signs are torsional nystagmus, skew deviation, ocular torsion, tilts of head, body, and perceived verrical in a clockwise or counterclockwise direction. Pitch plane signs are upbeat/downbeat nystagmus, forward/backward tilts and falls, deviations of the perceived horizon. The thus defined vestibular syndromes allow a precise topographic analysis of brainstem lesions according to their level and side. Special emphasis is placed on the vestibular roll plane syndromes of ocular tilt reaction, lateropulsion in Wallenberg's syndrome, thalamic and cortical astasia and their association with roll plane tilt of perceived vertical. Recovery is based on a functionally significant central compensation of a vestibular tone imbalance, the mechanism of which is largely unknown. Physical therapy may facilitate this central compensation, but this has not yet been proven in prospective studies. **Key Words:** Visual vertical—Lateropulsion—Vestibulo-ocular reflex—Central vestibular syndromes.

Vestibular pathways run from the eighth nerve and the vestibular nuclei through ascending fibers, such as the ipsilateral or contralateral medial longitudinal fasciculus (MLF), the brachium conjunctivum, or the ventral tegmental tract to the ocularmotor nuclei, the supranuclear integration centers in the rostral midbrain, and the vestibular thalamic subnuclei. From there they reach several cortex areas through the thalamic projection. Another relevant ascending projection reaches the cortex from vestibular nuclei via vestibular cerebellum structures, in particular the fastigial nucleus.

In the majority of cases central vestibular vertigo syndromes are caused by dysfunction or a deficit of sensory input induced by a lesion (1,2). In a few cases they are due to pathological excitation of various structures, extending from the peripheral vestibular organ to the vestibular cortex. As opposed to peripheral vestibular disorders, which are always characterized by a combination of perceptual, ocular motor, and postural signs and symptoms, central vestibular disorders may manifest as "a complete syndrome" (as in a peripheral lesion) or with only single components. The ocular motor aspect, for example, predominates in the syndromes of upbeat or downbeat nystagmus. Lateral falls may occur without vertigo in vestibular thalamic lesions (thalamic astasia) or as lateropulsion in Wallenberg's syndrome.

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Clinical Classification of Central Vestibular Disorders

The "elementary" neuronal network of the vestibular system is the di- or trisynaptic vestibulo-ocular reflex (VOR). There is evidence for a useful clinical classifica-

tion of central vestibular syndromes according to the three major planes of action of the VOR (Figure 1): yaw, roll, and pitch (1,3).

The plane-specific vestibular syndromes are determined by ocular motor, postural, and perceptual signs (Figure 2):

- **Yaw plane signs** are horizontal nystagmus, past pointing, rotational and lateral body falls, deviation of perceived straight-ahead to the left or right.
- **Roll plane signs** are torsional nystagmus, skew deviation, ocular torsion, tilts of head, body, and perceived vertical in a clockwise or counterclockwise direction.
- **Pitch plane signs** are upbeat/downbeat nystagmus, forward/backward tilts and falls, deviations of the perceived horizon.

Clinical examination with Frenzel's glasses (magnifying glasses with +16 diopters have light inside to prevent visual fixation, which could suppress spontaneous nystagmus) allows the clinician to better observe spontaneous eye movements. Upbeat and downbeat nystagmus, however, can be best seen with visual fixation without Frenzel's glasses. The alternating monocular cover test reveals horizontal or vertical misalignment of the visual axes, such as skew deviation. Determination of ocular torsion (measured as the angle formed by a straight line through the papilla and fovea and a horizontal line) requires fundus photographs or a laser scanning ophthalmoscope. Measurements of perceived vertical require special devices that do not give visual cues of true vertical. Head tilts, body lateropulsion, and the direction of vestibular falls can be observed at the bedside.

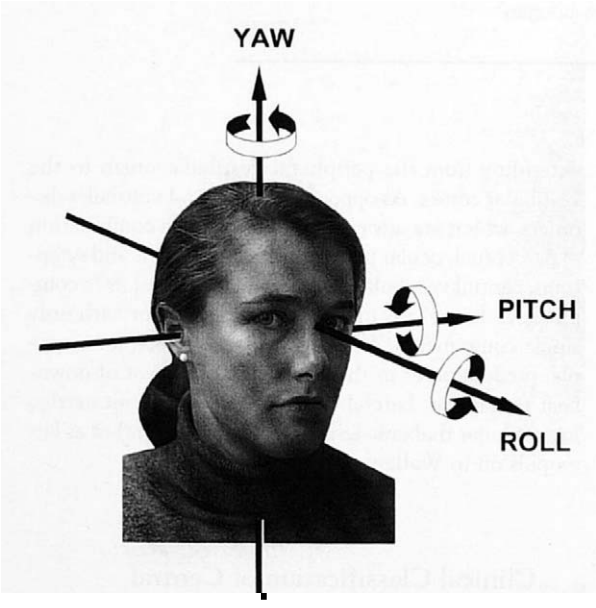

Figure 1. Schematic representation of the three major planes of action of the vestibulo-ocular reflex. Horizontal rotation about the vertical z axis = yaw; vertical rotation about the binaural y axis = pitch; vertical rotation about the x axis ("line of sight") = roll.

The thus defined VOR syndromes allow for a precise topographic diagnosis of brainstem lesions as to their level and side (Figure 3).

- **A tone imbalance in yaw** indicates lesions of the unilateral medulla including the root entry zone of the eighth nerve and/or the vestibular nuclei.
- **A tone imbalance in roll** indicates unilateral lesions (ipsiversive at pontomedullary level, contraversive at pontomesencephalic level).
- **A tone imbalance in pitch** indicates bilateral (paramedian) lesions or bilateral dysfunction of the flocculus.

Some vestibular disorders are characterized by a simultaneously peripheral and central vestibular involvement. Examples are large acoustic neurinomas, infarctions of the anterior inferior cerebellar artery, head trauma, and syndromes induced by alcohol intoxication. Others may affect the vestibular nerve root in the brainstem (lacunar infarction or focal demyelination in multiple sclerosis (MS) mimicking a peripheral disorder).

Cortical vestibular syndromes include vestibular seizures and lesional dysfunction with tilt of the perceived vertical, lateropulsion, and rarely rotational vertigo. There is no primary vestibular cortex, but the parietoinsular vestibular cortex (4) seems to act as a kind of main integration center. Dysfunction of this multisensory and sensorimotor cortex for spatial orientation and self-motion perception may be involved in spatial hemineglect and rare paroxysmal room-tilt illusions.

Most central vertigo syndromes have a specific locus but not a specific etiology. The etiology may, for example, be vascular, autoimmunologic (e.g., in MS), inflammatory, tumorous, toxic, or traumatic.

Vestibular Disorders in (Frontal) Roll Plane

The "graviceptive" input from the otoliths converges with that from the vertical semicircular canals at the level of the vestibular nuclei (5) and the ocular motor nuclei (6,7) to subserve static and dynamic vestibular function

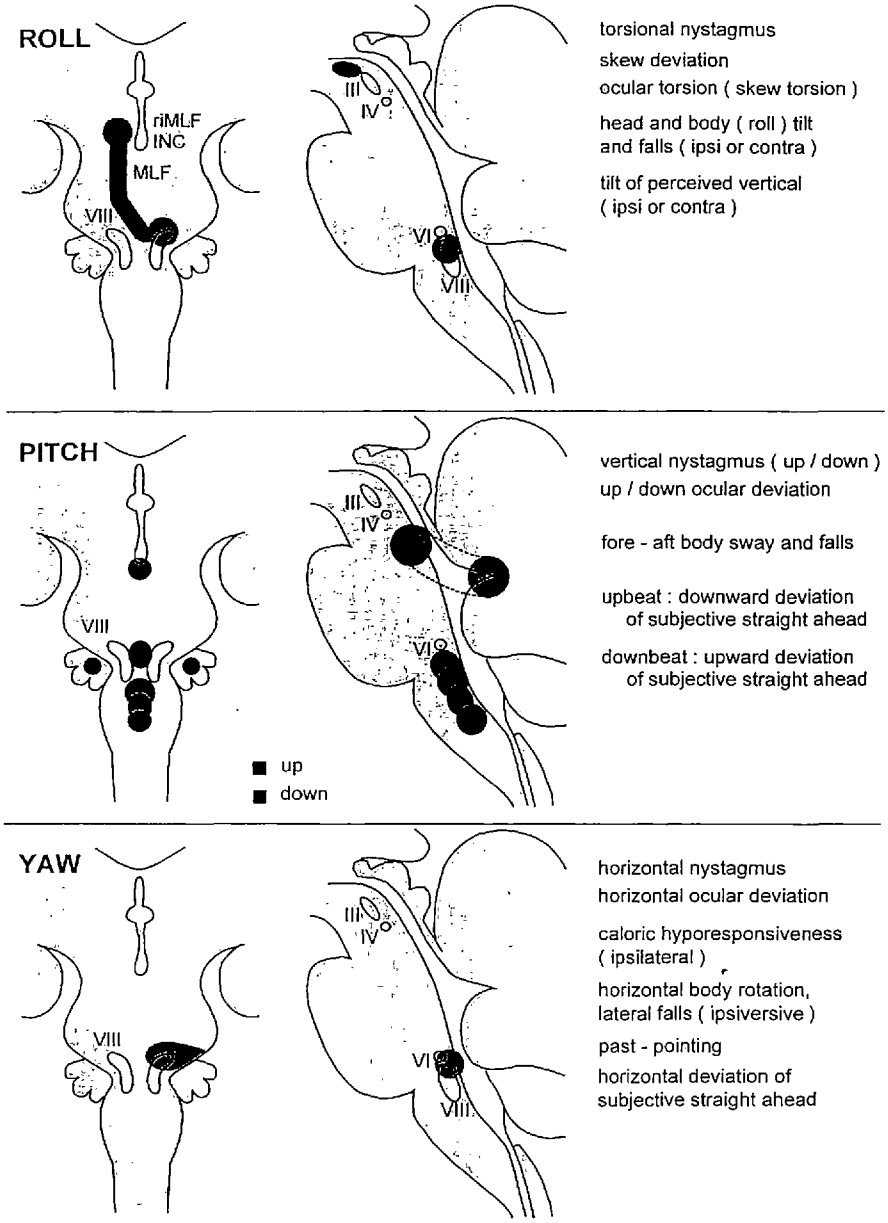

Figure 2. Topographic diagnosis of vestibular syndromes in roll, pitch, and yaw planes: schematic presentation of the distinct areas within the brainstem and vestibulo-cerebellum (frontal and sagittal views) in which a lesion induces a vestibulo-ocular tone imbalance in roll, pitch, or yaw plane. Typical ocular motor signs are torsional, vertical (up/downbeat), or horizontal nystagmus. A tone imbalance in roll indicates unilateral "graviceptive" pathway lesions from the medial or superior vestibular nuclei (inducing ipsiversive signs), crossing midline to the contralateral MLF and the rostral integration center for vertical and torsional eye movements, the INC (inducing contraversive signs). A tone imbalance in pitch indicates paramedian bilateral brainstem lesions at pontomesencephalic or pontomedullary level, the brachium conjunctivum, or the flocculi. It is striking that pontomedullary lesions may induce either upbeat or downbeat nystagmus or transitions between the two, whereas binocular flocculus lesions result only in downbeat nystagmus and a pontomesencephalic lesion only in upbeat nystagmus. A tone imbalance in yaw indicates a unilateral pontomedullary lesion involving the medial and superior vestibular nucleus. This area overlaps with roll and pitch function syndromes in more than one plane (riMLF = rostral interstitial nucleus of the medial longitudinal fasciculus, INC = interstitial nucleus of Cajal, III = oculomotor nucleus, IV = trochlear nucleus, VI = abducens nucleus, VIII = vestibular nucleus).

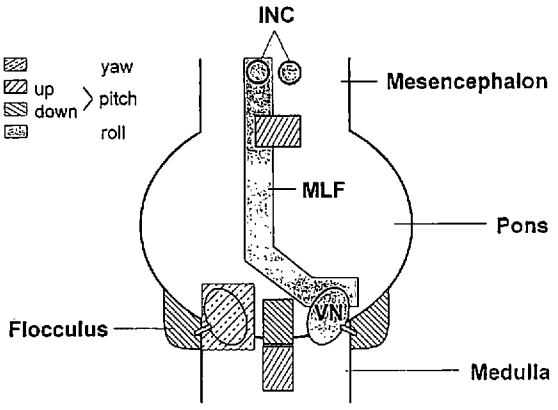

Figure 3. Vestibular syndromes in roll, pitch, and yaw planes: critical areas are schematically represented based on our current knowledge of vestibular and ocular motor structures and pathways, a lesion of which causes a vestibular tone imbalance in one of the three major planes of action. The mere clinical sign of a vertical, torsional, or horizontal nystagmus—if central-vestibular—allows a topographic diagnosis of the lesion, although the particular vestibular structures involved are still under discussion. Whereas a vestibular tone imbalance in the roll plane indicates unilateral brainstem lesions (a crossing in the pons), vertical nystagmus indicates bilateral lesions. Two separate causative loci are known for upbeat nystagmus: medullary or pontomesencephalic. Downbeat nystagmus indicates a bilateral paramedian lesion of the commissural fibers between the vestibular nuclei or a bilateral flocculus lesion. Horizontal nystagmus indicates unilateral pontomedullary lesions involving the vestibular nuclei. The differentiation of vestibular ocular motor signs according to the three major planes of action of the VOR and their mapping to distinct and separate areas in the brainstem are helpful for topographic diagnosis and for avoiding incorrect assignment of clinical signs to brainstem lesions identified with imaging techniques (INC = interstitial nucleus of Cajal, MLF = medial longitudinal fasciculus, VN = vestibular nucleus).

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), vertical misalignment of the visual axes (diplopia), ocular torsion, or a complete ocular tilt reaction (OTR), i.e., the triad of roll head tilt, skew deviation, and ocular torsion.

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. 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 (nystagmus) and dynamic (ocular deviation) 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 (8). Skew deviation was found in one-third and a complete OTR in one-fifth of these patients.

Current clinical data support the following preliminary topographic diagnostic rules based on vestibular signs and symptoms in roll (1) (Figures 3 and 4):

1. The fundamental pattern of eye-head tilt in roll—either complete OTR or skew torsion without head tilt—indicates a unilateral peripheral deficit of otolith and vertical canal input (which converge in the vestibular nuclei) or a unilateral lesion of "graviceptive" brainstem pathways from the vestibular nuclei (crossing midline at lower pontine level) to the interstitial nucleus of Cajal (INC) in the rostral midbrain.
2. Tilts of SVV, resulting from peripheral or central vestibular lesions from the labyrinth to the vestibular cortex, are the most sensitive sign of a vestibular tone imbalance in roll.
3. All tilt effects—perceptual, ocular motor, and postural—are ipsiversive (ipsilateral eye lowermost) and due to unilateral peripheral or pontomedullary paretic lesions below the crossing of the graviceptive pathways. They indicate involvement of the labyrinth, vestibular nerve, or medial and/or superior vestibular

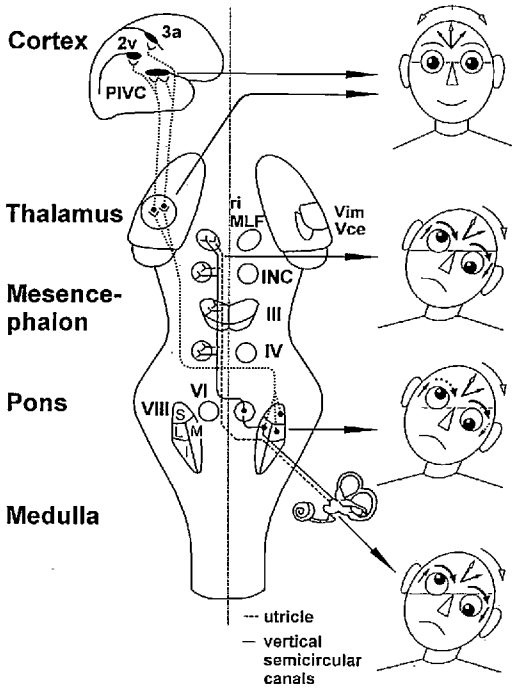

Figure 4. Vestibular syndromes in roll plane: "Graviceptive" pathways from otoliths and vertical semicircular canals converge and mediate vestibular function in roll plane. The projections from the otoliths and the vertical semicircular canals to the ocular motor nuclei (trochlear nucleus IV, oculomotor nucleus III, abducens nucleus VI), the supranuclear centers of the INC, and the rostral interstitial nucleus of the MLF (riMLF) are shown. They subserve VOR in three planes. The VOR is part of a more complex vestibular reaction which also involves vestibulo-spinal connections via the medial and lateral vestibulo-spinal tracts for head and body posture control. Furthermore, connections to the assumed vestibular cortex (areas 2v and 3a and the parieto-insular vestibular cortex, PIVC) via the vestibular nuclei of the thalamus (Vim, Vce) are depicted. "Graviceptive" vestibular pathways for the roll plane cross at the pontine level. OTR (skew torsion, head tilt, and tilt of perceived vertical, SVV) is depicted schematically on the right in relation to the level of the lesion: ipsiversive OTR with peripheral and pontomedullary lesions; contraversive OTR with pontomesencephalic lesions. In vestibular thalamic lesions, the tilts of SVV may be contraversive or ipsiversive; in vestibular cortex lesions they are preferably contraversive. OTR is not induced by supratentorial lesions above the level of INC. Dotted versus solid arrows around the eyes indicate direction and disconjugacy of skew deviation and ocular torsion.

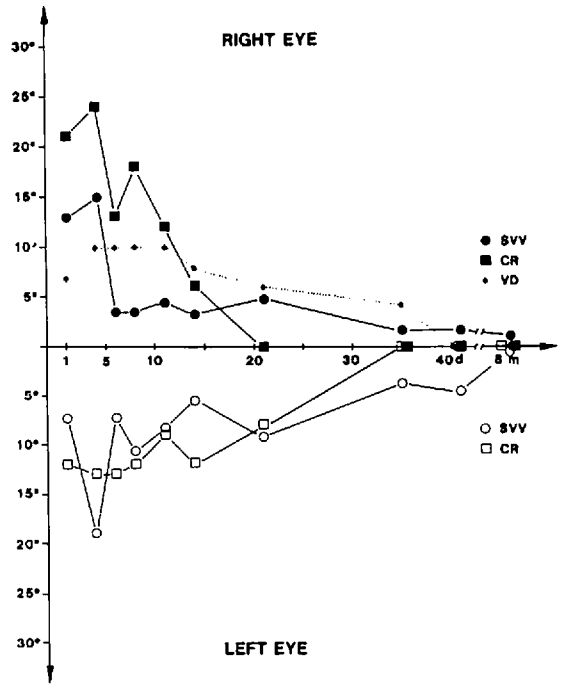

Figure 5. Spontaneous course of ocular torsion (cyclorotation, CR) skew deviation (vertical divergence, VD), and SVV tilt, in degrees, in a patient with a left paramedian midbrain infarction presenting with a complete OTR to the right. OTR consisted of contraversive head tilt of 20°, skew deviation of 10°, left eye over right eye, and ocular torsion of 10°–20°. Both directions along the y-axis are "rightward" and show disconjugate SVV and CR. All signs in the roll plane—CR, VD, and SVV tilts—show gradual recovery within 6 weeks.

nuclei; the latter are mainly supplied by the vertebral artery.

4. All tilt effects in unilateral pontomesencephalic brainstem lesions are contraversive (contralateral eye lowermost) and indicate involvement of the medial longitudinal fasciculus (MLF) (paramedian arteries arising from basilar artery) or INC and riMLF (paramedian superior mesencephalic arteries arising from the basilar artery.
5. OTR with unilateral (ponto)medullary lesions (vestibular nuclei) indicates the "ascending" (reflexive) type of a tone imbalance of the VOR in roll.
6. OTR due to rostral midbrain lesions (INC) reflects the "descending" type of tone imbalance involving the neural integration center for eye-head coordination in roll. The functional concept is that there are two distinct and separate brainstem structures controlling eye-head coordination in roll and pitch planes by virtue of separate pathways: the bilateral caudal VOR and the bilateral rostral integration center.
7. Skew deviation is always combined with ocular torsion, i.e., skew-torsion sign. It manifests without head tilt if ascending pontomesencephalic "graviceptive"

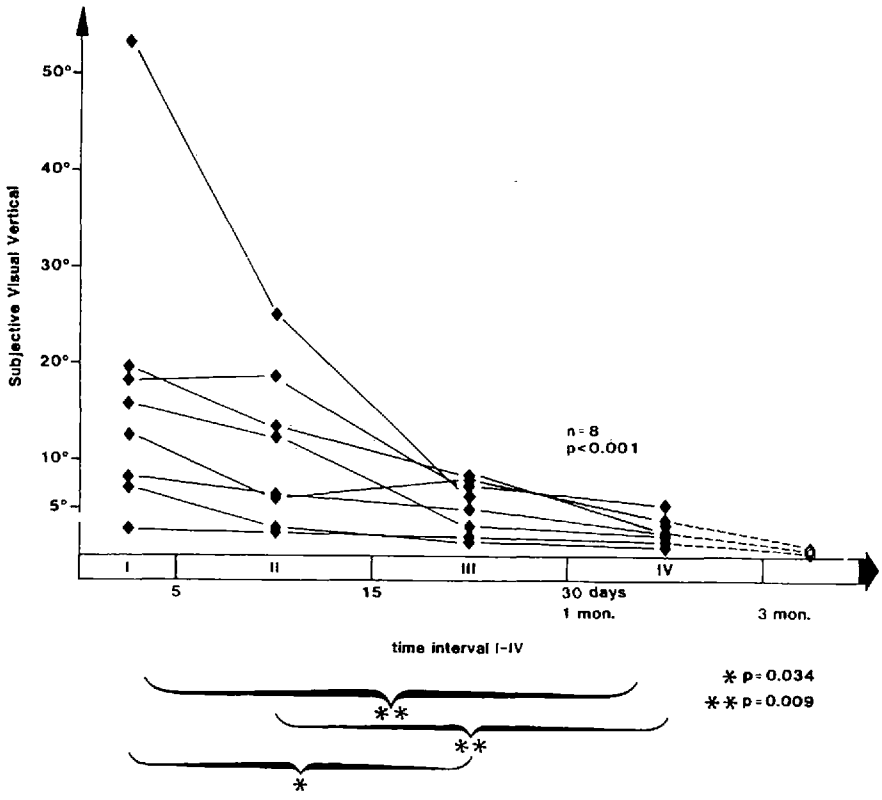

Figure 6. Time course of change in deviation of subjective visual vertical (SVV) after acute unilateral infarction of the lateral medulla oblongata in 8 patients with Wallenberg's syndrome. Friedman nonparametric analysis of variance shows a significant difference between SVV values for different time intervals ($p < 0.001$), and examination of the individual differences (Wilcoxon and Wilcox test) shows a significant decrease in SVV from time I to time III, time I to time IV, and time II to time IV. Gradual recovery is most pronounced in the first 30 days.

pathways are affected rostral to the downward branching of the vestibulospinal tract.

8. Unilateral lesions of ascending (sensory) vestibular pathways rostral to the INC typically manifest with deviations of perceived vertical without concurrent eye-head tilt.
9. OTR in unilateral paramedian thalamic infarctions (paramedian thalamic arteries from basilar artery) indicates simultaneous ischemia of the paramedian rostral midbrain including the INC.
10. 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 (thalamogeniculate arteries).
11. Unilateral lesions of the parieto-insular vestibular cortex (PIVC) cause moderate, mostly contraversive SVV tilts (temporal branches of the middle cerebral artery or deep perforators) and "cortical lateropulsion."
12. 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 (9).

Tilt effects caused by paroxysmal activation of "graviceptive" pathways point in the opposite direction of those caused by lesional inhibition, such as unilateral infarction (10–13). Thus all clinical signs of vestibular dysfunction in roll can be helpful when determining not only the level but also the side of the brainstem lesion. If the level of damage is known from the clinical syndrome, the vestibular syndrome indicates the more severely affected side. Conversely, if the side of damage is clear from the clinical syndrome, the direction of OTR, skew deviation, and SVV tilt indicates the level on the brainstem.

Etiology

The two most common causes of tonic OTR are brainstem ischemia (especially Wallenberg's syndrome and unilateral paramedian thalamic plus rostral mesencephalic infarctions) and brainstem tumors. We have also seen cases with unilateral thalamic hemorrhages, lower brainstem hemorrhages (cavernous angioma, lymphomas), after severe brainstem concussion, in multiple sclerosis, or associated with attacks of basilar migraine.

The paroxysmal OTR described in a patient with MS (12) may be a variant of the paroxysmal attacks assumed to arise from ephaptic spreading between adjacent demyelinated axons. We have observed repeated paroxysmal attacks (vestibular excitation rather than inhibition) of contraversive OTR with ipsiversive torsional nystagmus in the acute stage of Wallenberg's syndrome. The typical direction of torsional nystagmus is contraversive in the dorsolateral medullary syndrome.

Natural Course and Management

The natural course and management of OTR depend on the etiology. Deviations of eyes and perceived vertical are usually transient because of the unilateral lesion and the central compensation via the unaffected side; in cases of hemorrhage or infarction recovery occurs within a few days to weeks. However, it can be permanent in asymmetric bilateral lesions, as we observed in a patient with severe brainstem concussion. Here the possibility of central compensation by the pathways of the less affected brainstem side is reduced. Following unilateral brainstem infarctions, all features of OTR—postural, ocular motor, and perceptual—disappear naturally and gradually within 4-5 weeks to months (repeated measurements made in seven patients over a period of up to 1–3 years; (14) (Figures 5 and 6). Repeated measurements of skew deviation, ocular torsion (OT), and tilts of perceived vertical (SVV) made during a single day showed consistent tilts. Some patients, however, maintained a residual OT of a few degrees without a corresponding tilt of SVV for up to 2 years.

Recovery is based on a functionally significant central compensation of a vestibular tone imbalance induced by a unilateral central lesion. The mechanisms underlying central compensation of central lesions may be similar to those of central compensation of peripheral vestibular lesions. Recovery from vestibular lesions is neither a simple nor a single process; multiple processes are involved.

Analysis of the mechanisms of recovery requires a careful comparison of normalization between parallel phenomena at the behavioral level, on the one hand, and the neuronal level, on the other. Incongruencies in the time course and the magnitude of the changes in behavior and neuronal activity clearly indicate that multiple processes of compensation occur in distributed neuronal networks at different locations and at different times (15–17).

Physical therapy may facilitate this central compensation, but this has not yet been proven in a prospective study. Vestibular sedatives can be administered in exceptional cases of pronounced nausea. These drugs should not be given after nausea disappears, because they prolong the time required to achieve compensation of the vestibular tone imbalance.

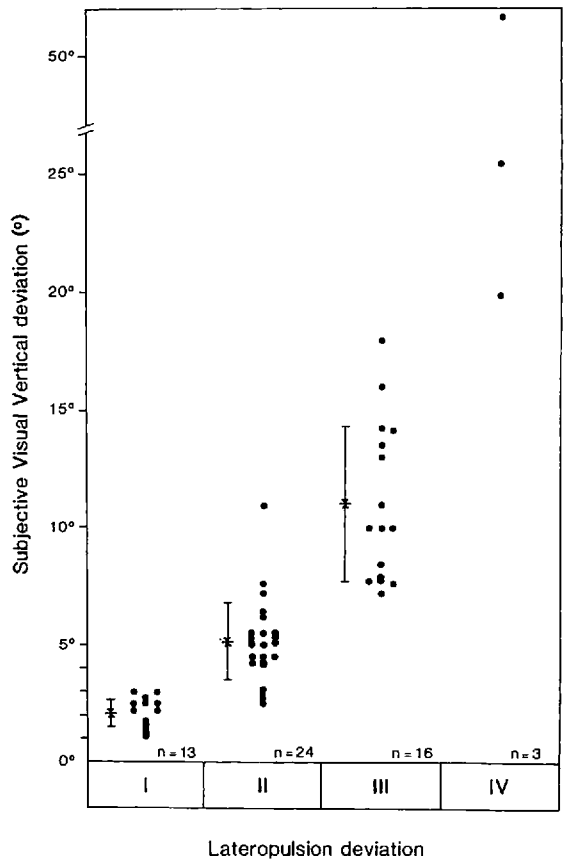

Figure 7. Deviations of the subjective visual vertical (SVV, tilt in degrees) in 36 patients with infarctions of the lateral medulla oblongata. Adjustments of the SVV (first measurements in the acute stage) are depicted in relation to the severity of body lateropulsion (abscissa). The dots represent single measurements (in some patients there were repeated measurements during the course of the disease); means and standard deviations are also depicted. The more pronounced the lateropulsion, the greater the deviations of SVV. SVV and lateropulsion are both ipsiversive to the side of the lesion.

Lateropulsion in Wallenberg's Syndrome

Lateropulsion of the body is a well-known transient feature of dorsolateral medullary infarction. These patients have irresistible, ipsiversive falls but generally no subjec-

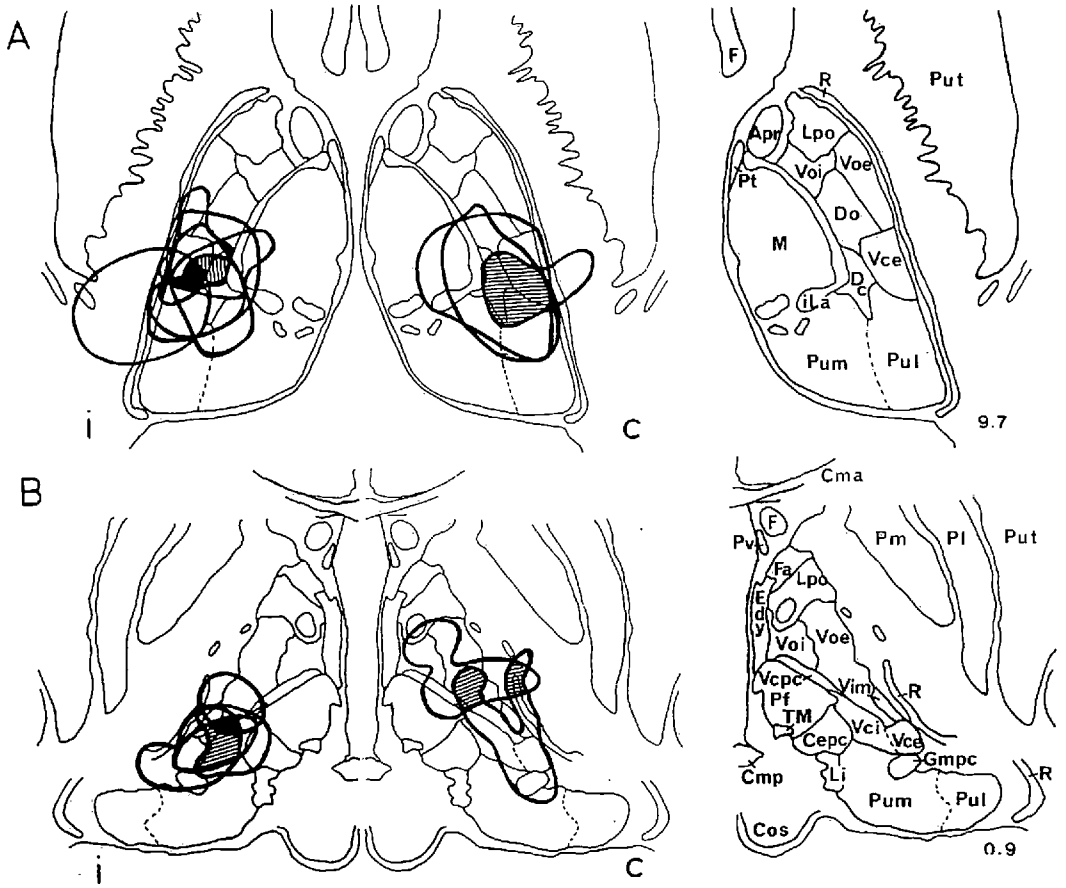

Figure 8. Overlap areas of 9 posterolateral infarctions which caused either ipsiversive (n = 6, left) or contraversive (n = 3, middle) tilts of SVV. As can be seen by comparison with the transverse section of the middle thalamic level (A; 9.7 mm above the AC-PC line) and the lower thalamic level (B; 0.9 mm above the AC-PC line) the overlap area involves the thalamic nuclei Vce, Dc, Vci, Vim, Voe, (Do), independent of the direction (i = ipsiversive; c = contaversive) of induced tilt of the internal representation of gravity. (Vce= nucleus ventrocaudalis externus; Dc= nucleus dorsocaudalis; Vci= nucleus ventrocaudalis internus;Voe= nucleus ventrocaudalis externus; Do= nucleus dorso-oralis).

tive vertigo. Different brainstem lesions from midbrain to medulla cause deviations of the subjective vertical (8,18). Transient ocular-tilt reaction and ipsiversive deviations of the subjective vertical, which indicate a pathological shift in the internal representation of the gravitational vector, are typically found in Wallenberg's syndrome (Figure 6) (19,20). We hypothesized that the subjective vertigo is missing in these patients (despite a striking tendency to fall sideways), because individual multisensory postural regulation is adjusted to the deviated vertical (Figure 7). Lateropulsion then represents postural compensation of an apparent body tilt contraversive to the lesioned side. Despite the resulting postural imbalance and the conflicting true vertical, the body is continuously adjusted toward what the central nervous system erroneously computes as vertical (19). This could explain why patients fall without vertigo or warning signals from the multisensory spatial orientation system. Lateropulsion in dorsolateral medullary lesions (Figure 6) exhibits spontaneous recovery within days to weeks. This process might be facilitated by physical therapy. Lateropulsion without hemiparesis also occurs in cortical lesions.

Thalamic and Cortical Astasia Associated with SVV Tilts

An association of SVV tilts with falls is also typical for posterolateral (vestibular subnuclei) thalamic lesions.

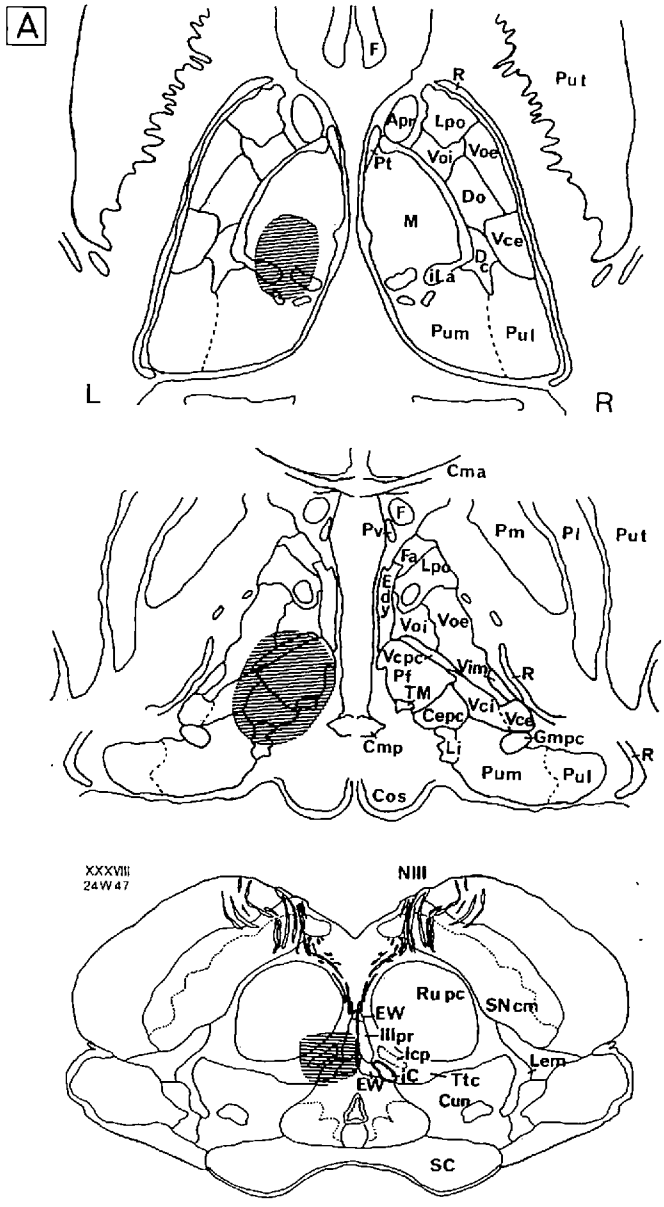

Figure 9. Typical lesioned area in a paramedian thalamic infarction with contraversive OTR that is caused by the simultaneous involvement of the rostral midbrain. Infarcted areas were taken from MRI scans and projected onto the appropriate transverse thalamic and midbrain sections of the stereotaxic atlas (9.7 mm and 0.9 mm above the anterior commissure-posterior commissure line; Van Buren and Borke, 1972) (top and middle) and the midbrain atlas (Olszewski and Baxter, 191982; plate XXXVIII). Complete contraversive OTR is due to the lesion of the rostral midbrain tegmentum including the region of the INC (iC) (bottom). Abbreviations: Apr = nucleus anterior principalis; Cepc = nucleus centralis parvocellularis; Cma = anterior commissure; Cmp = posterior commissure; Cos = superior colliculus; Cun = nucleus cuneiformis; Dc = nucleus dorsocaudalis; Do = nucleus dorsooralis; Edy = nucleus endymalis; EW = Edinger-Westphal nucleus; F = fornix; Fa = nucleus fascicularis; Gmpc = medial geniculate body, pars parvocellularis; Hl = nucleus habenularis lateralis; Hm = nucleus habenularis medialis; iC = interstitial nucleus of Cajal [INC]; Icp = nucleus intracapsularis; IIIpr = nucleus oculomotorius principalis; iLa = nucleus intralamellaris; Lem = medial lemniscus; Li = nucleus limitans; Lpo = nucleus lateropolaris; M = nucleus medialis; NIII = oculomotor nerve; Pf = nucleus parafascicularis; Pl = lateral pallidum; Pm = medial pallidum; Pt = nucleus parataenialis; Pul = lateral pulvinar; Pum = medial pulvinar; Puo = oral pulvinar; Put = putamen; Pv = nucleus paraventricularis hypo-thalami; R = reticular nuclei; Ru pc = red nucleus, pars parvocellularis; SC = superior colliculus; Smth = stria medullaris thalami; SN cm = substantia nigra, pars compacta; TM = tract of Meynert; Ttc = central tegmental tract; Vce = nucleus ventrocaudalis externus; Vci = nucleus ventrocaudalis internus; Vcpc = nucleus ventro-caudalis parvocellularis; Vim = nucleus ventro-oralis intermedius; Voe = nucleus ventro-oralis externus; Voi = nucleus ventro-oralis internus.

Thalamic astasia (21) is a condition in which patients without paresis or sensory or cerebellar deficits are unable to maintain an unsupported, upright posture. Postural imbalance with a transient tendency to fall has been reported following therapeutic thalamotomy and thalamic hemorrhage (22). According to our experience in some 30 patients with thalamic infarctions, the posterolateral type may cause contraversive or ipsiversive postural instability with SVV tilts (Figure 8), whereas the paramedian type (if it extends into the rostral midbrain) always causes contraversive falls (Fig. 9). Astasia and gait failure with damage of the pontomesencephalic (locomotor) region was described by Masdeu and coworkers (23). Although not discussed, it could also be explained in part by a vestibular tone imbalance in roll, especially since skew deviation was described as a feature of the syndrome.

Of 31 patients with cortical infarctions of the middle cerebral artery territory, 21 showed significant, mostly contraversive, pathological SVV tilts (24). The overlapping area of these infarctions centered on the posterior

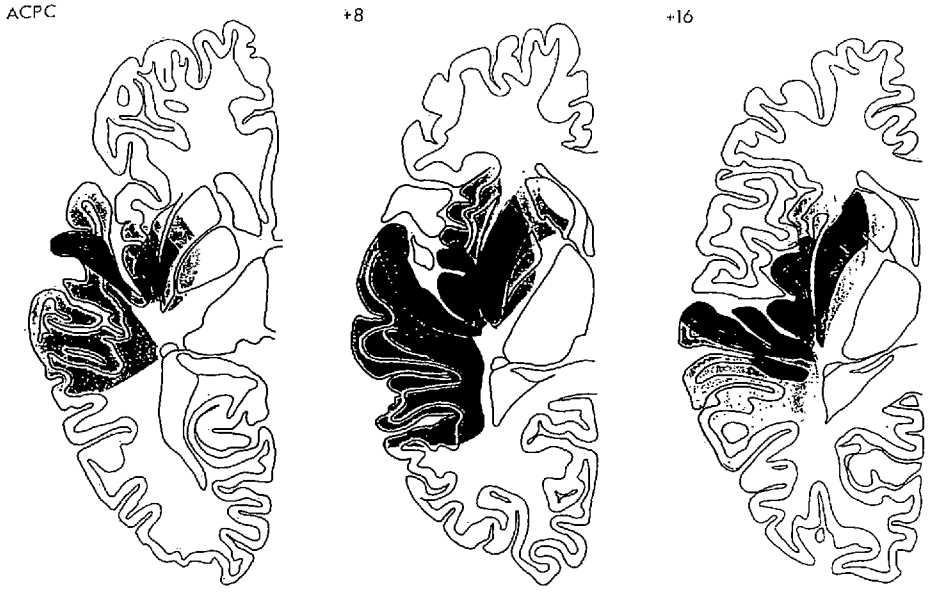

Figure 10. Collective presentation of infarcted areas taken from MRI scans and projected onto the appropriate transverse sections of the atlas of Duvernoy (1991) in 7 patients with clearly demarcated infarctions of the middle cerebral artery territory which caused significant contraversive tilts of perceived vertical. Overlapping areas of infarctions (7 of 7 in black) in three sections (0 mm AC-PC line; +8 mm above; +16 mm above) are centered at the posterior part of the insula, involving the long insular gyrus with the adjacent short insular gyrus, the transverse temporal gyrus, and the superior temporal gyrus.

insula, which is probably homologous to the parietoinsular vestibular cortex (Fig. 10). SVV tilts caused by vestibular cortex lesions may also be associated with (a compensatory) body lateropulsion. This explains the cortical phenomenon "pusher," which physical therapists readily recognize.

Cortical Spatial Disorientation

An inappropriate vestibular input due to peripheral or central dysfunction in patients can cause paroxysmal "room-tilt illusions." Tilts of SVV and room tilt illusions are obviously vestibular signs that indicate misperception of verticality. They both involve "graviceptive" pathways that extend from the otoliths and the semicircular canals through the vestibular nuclei and the thalamus to the parietoinsular vestibular cortex. The question arises as to whether both phenomena are the same, since their net tilt angles differ only in size. They are not, however, because of a few other typical differences (25):

1. SVV tilts are usually stable (chronic) signs and recovery occurs gradually, within days to weeks; room tilt illusions are paroxysmal or transient phenomena.
2. SVV tilts manifest as a continuum of angle of tilt up to a maximum of about 30°; room tilt illusions occur in 90° steps as a lateral, fore-aft tilt or upside-down vision.
3. SVV tilts are usually not associated with the perception of room tilt.

In brief, room tilt illusions are, in our opinion, transient mismatches of the visual and vestibular 3-D map coordinates that occur in 90° or 180° steps as the erroneous result of the attempted match of the spatial planes (25). Furthermore, a plane- and direction-specific tilt of static spatial orientation occurs in disorders of the vestibulo-ocular reflex, such as downbeat and upbeat nystagmus. Adjustments of subjective straight ahead exhibit an upward shift in downbeat nystagmus and a downward shift in upbeat nystagmus (26). Here the tilt of perceived straight-ahead is elicited by the asymmetric vestibular tone in the pitch plane in the brainstem which reaches the cortex by ascending projections. Vestibular syndromes caused only by cortical lesions have not yet been well defined:

Static cortical spatial disorientation may occur as paroxysmal room-tilt illusion in parietal or frontal lobe lesions, contralateral spatial hemineglect in inferior parietal or frontal lobe lesions, vertical neglect below the hor-

Table 1. *Central vestibular syndromes*

Site	Syndrome	Mechanism/Etiology
Vestibular cortex (multisensory)	Vestibular epilepsy	Vestibular seizures are auras (simple or complex partial multisensory seizures)
	Volvular epilepsy	Sensorimotor "vestibular" rotatory seizures with walking in small circles
	Non-epileptic cortical vertigo	Rare rotatory vertigo in acute lesions of the parieto-insular vestibular cortex
	Spatial hemineglect (contraversive)	Multisensory horizontal deviation of spatial attention with (right) parietal or frontal cortex lesions
	Transient room-tilt illusions	Paroxysmal or transient mismatch of visual- and vestibular 3-D spatial coordinate maps in vestibular brainstem, parietal, or frontal cortex lesions
	Tilt of perceived vertical with body lateropulsion (mostly contraversive)	Vestibular tone imbalance in roll with acute lesions of the parieto-insular vestibular cortex
Thalamus	Thalamic astasia	Dorsolateral vestibular thalamic lesions
	Tilt of perceived vertical (ipsiversive or contraversive) with body lateropulsion	Vestibular tone imbalance in roll
Mesodiencephalic brainstem	Ocular tilt reaction (contraversive; ipsiversive if paroxysmal)	Vestibular tone imbalance in roll (integrator-OTR with INC lesions)
	Torsional nystagmus (ipsiversive or contraversive)	Ipsiversive in INC lesions Contraversive in riMLF lesions
Mesencephalic brainstem	Skew torsion (contraversive)	Vestibular tone imbalance in roll with MLF lesions
	Upbeat nystagmus	Vestibular tone imbalance in pitch in bilateral brachium conjunctivum lesions
Ponto-medullary brainstem	Tilt of perceived vertical lateropulsion, ocular tilt reaction	Vestibular tone imbalance in roll with medial and/or superior vestibular nuclei lesions
	Pseudo "vestibular neuritis"	Lacunar infarction or MS plaque at the root entry zone of the eighth nerve
	Downbeat nystagmus	Vestibular tone imbalance in pitch
	Transient room-tilt illusion	Acute severe vestibular tone imbalance in roll or pitch
	Paroxysmal room-tilt illusion in MS	Transversally spreading ephaptic axonal activity
	Paroxysmal dysarthria/ataxia in MS	Transversally spreading ephaptic axonal activation
	Paroxysmal vertigo evoked by lateral gaze	Vestibular nuclei lesion?
Medulla	Upbeat nystagmus	Vestibular tone imbalance in pitch? (nucleus prepositus hypoglossi)
Vestibular cerebellum	Downbeat nystagmus	Vestibular tone imbalance in pitch caused by bilateral flocculus lesions (disinhibition)
	Positional downbeat nystagmus	Disinhibited otolith-canal interaction in nodullus lesions?
	Familial episodic ataxia (EA1 with myokymia and EA2 with vertigo)	EA1=autosomally dominant inherited potassium channelopathy EA2=autosomally dominant inherited calcium channelopathy
	Encephalitis with predominant vertigo	Viral infection of cerebellum
	Epidemic vertigo	Viral infection of cerebellum

izontal meridian in bilateral parieto-occipital lesions, and roll of perceived vertical (mostly contraversive) and body lateropulsion in unilateral PIVC lesions. Dynamic cortical spatial disorientation with apparent motion or rotational vertigo may occur in vestibular epilepsy with temporoparietal foci and rarely as a transient vertigo in acute lesions of the vestibular cortex.

Conclusion

In this short review we have presented evidence showing that most central vestibular syndromes can be attributed to a particular spatial plane of the VOR and specific locus of vestibular structures in the brainstem, cerebellum, thalamus, or the cortex. Table 1 summarizes our current knowledge of central vestibular syndromes, their localizing value for topographic diagnosis, and their different mechanisms and etiologies.

References

1. Brandt T, Dieterich M. Vestibular syndromes in the roll plane: Topographic diagnosis from brainstem to cortex. *Ann Neurol* 1994; 36:337–347.
2. Brandt Th. *Vertigo, its multisensory syndromes*, 2nd ed. London: Springer-Verlag, 1999.
3. Brandt T, Dieterich M. Central vestibular syndromes in the roll, pitch, and yaw planes: Topographic diagnosis of brainstem disorders. *Neuro-ophthalmology* 1995; 15:291–303.
4. Guldin W, Grüsser O-J. The anatomy of the vestibular cortices of primates. In: Collard M , Jeannerod M, Christen Y (eds). *Le cortex vestibulaire*. Ipsen: Boulogne, 1996:17–26.
5. Angelaki DE, Bush GA, Perachio AA. Two-dimensional spatiotemporal coding of linear acceleration in vestibular nuclei neurons. *J Neurosci* 1993; 13:1403–1417
6. Baker R, Precht W, Berthoz A. Synaptic connections to trochlear motoneurons determined by individual vestibular nerve branch stimulation in the cat. *Brain Res* 1973; 64:402–406
7. Schwindt PC, Richter A, Precht W. Short latency utricular and canal input to ipsilateral abducens motoneurons. *Brain Res* 1973; 60:259–262.
8. Dieterich M, Brandt Th. Ocular torsion and tilt of subjective visual vertical are sensitive brainstem signs. *Ann Neurol* 1993a; 33:292–299.
9. Dieterich M, Brandt Th. Ocular torsion and perceived vertical in oculomotor, trochlear and abducens nerve palsies. *Brain* 1993c; 116:1095–1104.
10. Halmagyi GM, Brandt Th, Dieterich M, et al. Tonic contraversive ocular tilt reaction due to unilateral meso-diencephalic lesion. *Neurology* 1990; 40:1503–1509.
11. Lueck CJ, Hamlyn P, Crawford TJ, et al. A case of ocular tilt reaction and torsional nystagmus due to direct stimulation of the midbrain in man. *Brain* 1991; 114:2069–2079.
12. Rabinovitch HE, Sharpe JA, Sylvester TO. The ocular tilt reaction. A paroxysmal dyskinesia associated with elliptical nystagmus. *Arch Ophthalmol* 1977; 95:1395–1398.
13. Hedges TR, Hoyt WF. Ocular tilt reaction due to an upper brainstem lesion: Paroxysmal skew deviation, torsion, and oscillation of the eyes with head tilt. *Ann Neurol* 1982; 11:537–540.
14. Dieterich M, Brandt Th. Thalamic infarctions: differential effects on vestibular function in the roll plane (35 patients). *Neurology* 1993b; 43:1732–1740.
15. Straka H, Dieringer N. Spinal plasticity after hemilabyrinthectomy and its relation to postural recovery in the frog. *J Neurophysiol* 1995; 73:1617–1631.
16. Dieringer N. "Vestibular compensation": Neural plasticity and its relations to functional recovery after labyrinthine lesions in frogs and other vertebrates. *Prog Neurobiol* 1995; 46:97–129.
17. Curthoys IS, Halmagyi GM. Vestibular compensation: A review of the oculomotor, neural, and clinical consequences of unilateral vestibular loss. *J Vestib Res* 1994; 5:67–107.
18. Friedmann G. The judgement of the visual vertical and horizontal with peripheral and central vestibular lesions. *Brain* 1970; 93:313–328.
19. Brandt Th, Dieterich M. Pathological eye-head coordination in roll: Tonic ocular tilt reaction in mesencephalic and medullary lesions. *Brain* 1987; 110:649–666.
20. Dieterich M, Brandt Th. Wallenberg's syndrome lateropulsion: lateropulsion, cyclorotation and subjective visual vertical in 36 patients. *Ann Neurol* 1992; 31:399–408.
21. Masdeu JC, Gorelick PB. Thalamic astasia: Inability to stand after unilateral thalamic lesions. *Ann Neurol* 1988; 23:596–603.
22. Verma AK, Maheshwari MC. Hyperesthetic-ataxic-hemiparesis in thalamic hemorrhage. *Stroke* 1986; 17:49–51.
23. Masdeu JC, Alampur U, Cavaliere R, Tavoulareas G. Astasia and gait failure with damage of the pontomesencephalic locomotor region. *Ann Neurol* 1994; 35:619–621.
24. Brandt Th, Dieterich M, Danek A. Vestibular cortex lesions affect perception of verticality. *Ann Neurol* 1994; 35:528–534.
25. Brandt Th. Cortical matching of visual and vestibular 3-D coordinate maps. *Ann Neurol* 1997; 42:983–984.
26. Dieterich M, Grünbauer WM, Brandt Th. Direction specific impairment of motion perception and spatial orientation in downbeat and upbeat nystagmus in humans. *Neurosci Lett* 1998; 245: 29–32.