

Vertigo and Dizziness: Common Complaints 3rd ed.
by Michael Strupp, Thomas Brandt, Marianne Dieterich

Chapter 13 – Central Vestibular Disorders

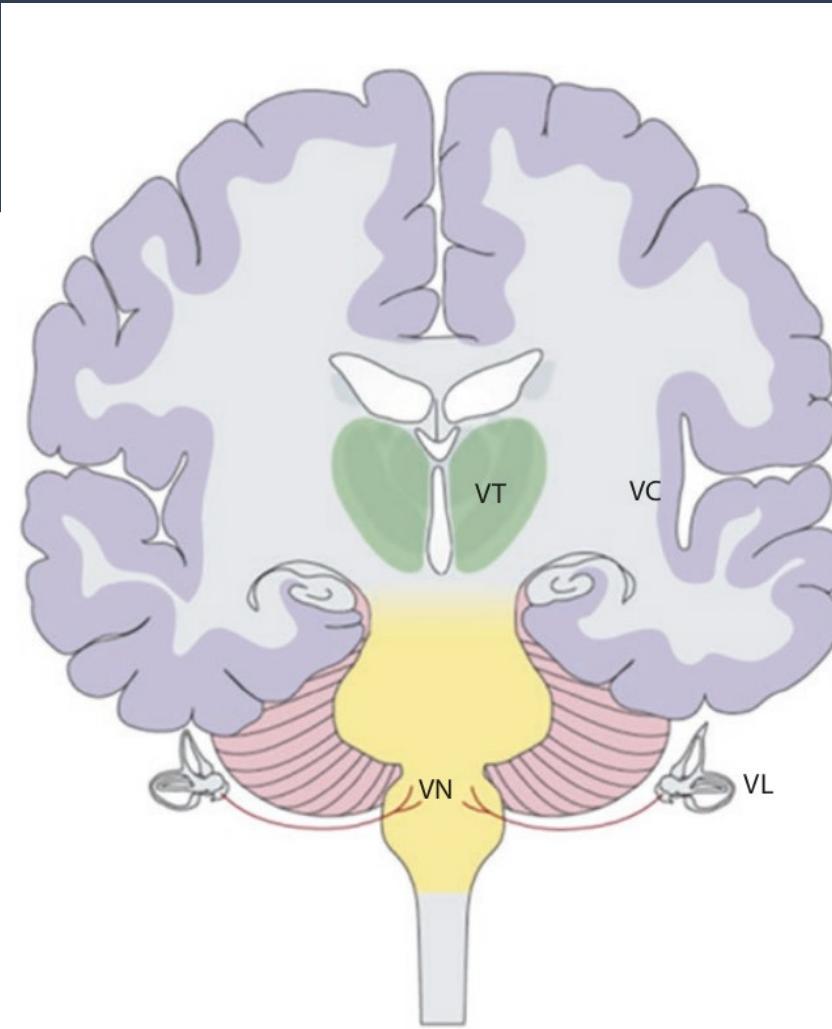
雙和醫院 神經科 陳致中

Outline

- 13.1 General survey of central vestibular syndromes
- 13.2 Clinical aspects of central vertigo and dizziness disorders
- 13.3 Higher (cognitive) vestibular syndromes
- 13.4 Cerebellar vertigo and dizziness
(共 44 頁 cf. AUVP 28 頁、BPPV 26 頁)

Central Vestibular Pathway

- Brainstem (vestibular nuc, ocular motor nuc)
- Vestibulo-cerebellum
- Thalamus
- Temporo-parietal cortex: parieto-insular vestibular cortex (PIVC); medial superior temporal (MST)



Cortex

- Cortical vertigo
- Pusher syndrome
- Room tilt illusion
- Spatial hemineglect
- Spatial memory deficit
- Vestibular epilepsy

Thalamus

- Thalamic astasia
- Pusher syndrome

Brainstem

- Lateropulsion
- Ocular tilt reaction
- Paroxysmal ataxia/ dysarthria
- Pseudoneuritis
- Room tilt illusion
- Skew-torsion
- Vestibular migraine

Cerebellum

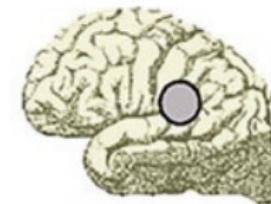
- Downbeat nystagmus
- Episodic ataxia type 2
- Ocular tilt reaction
- Positional vertigo/ nystagmus
- Pseudoneuritis
- Upbeat nystagmus

Course and Duration Help in the Differential Diagnosis

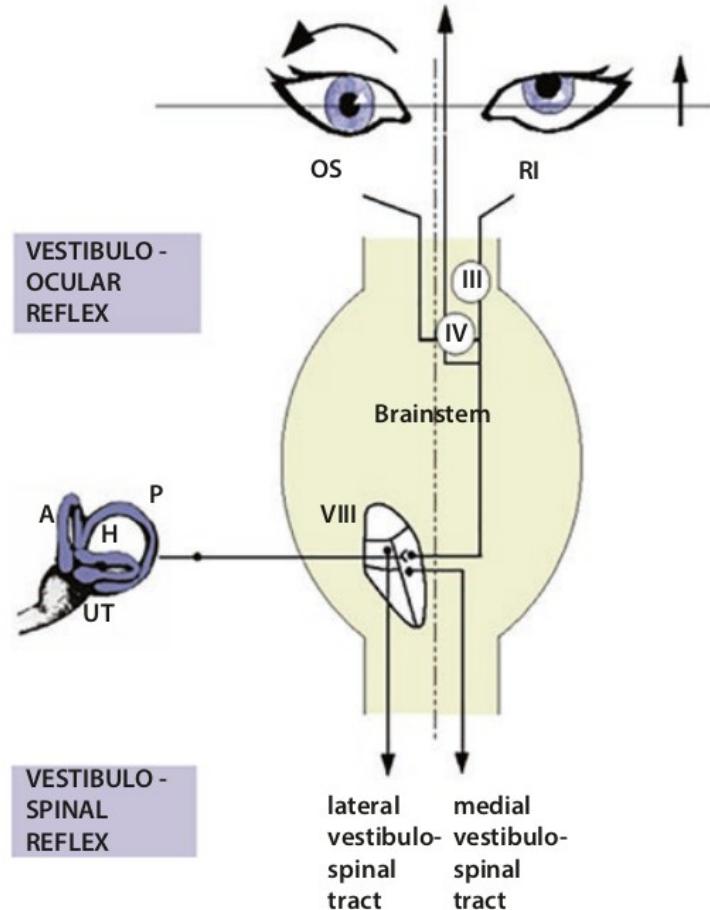
- **Seconds~Minutes**
 - TIA, vestibular migraine, EA-2, paroxysmal brainstem attacks (MS), vestibular epilepsy
- **Hours~Days**
 - Stroke, MS, vestibular migraine
- **Days, Weeks, Persistent**
 - Permanent brainstem or cerebellar pathology (degeneration, stroke, intoxication, tumor...)

Central Component of VOR

PERCEPTION

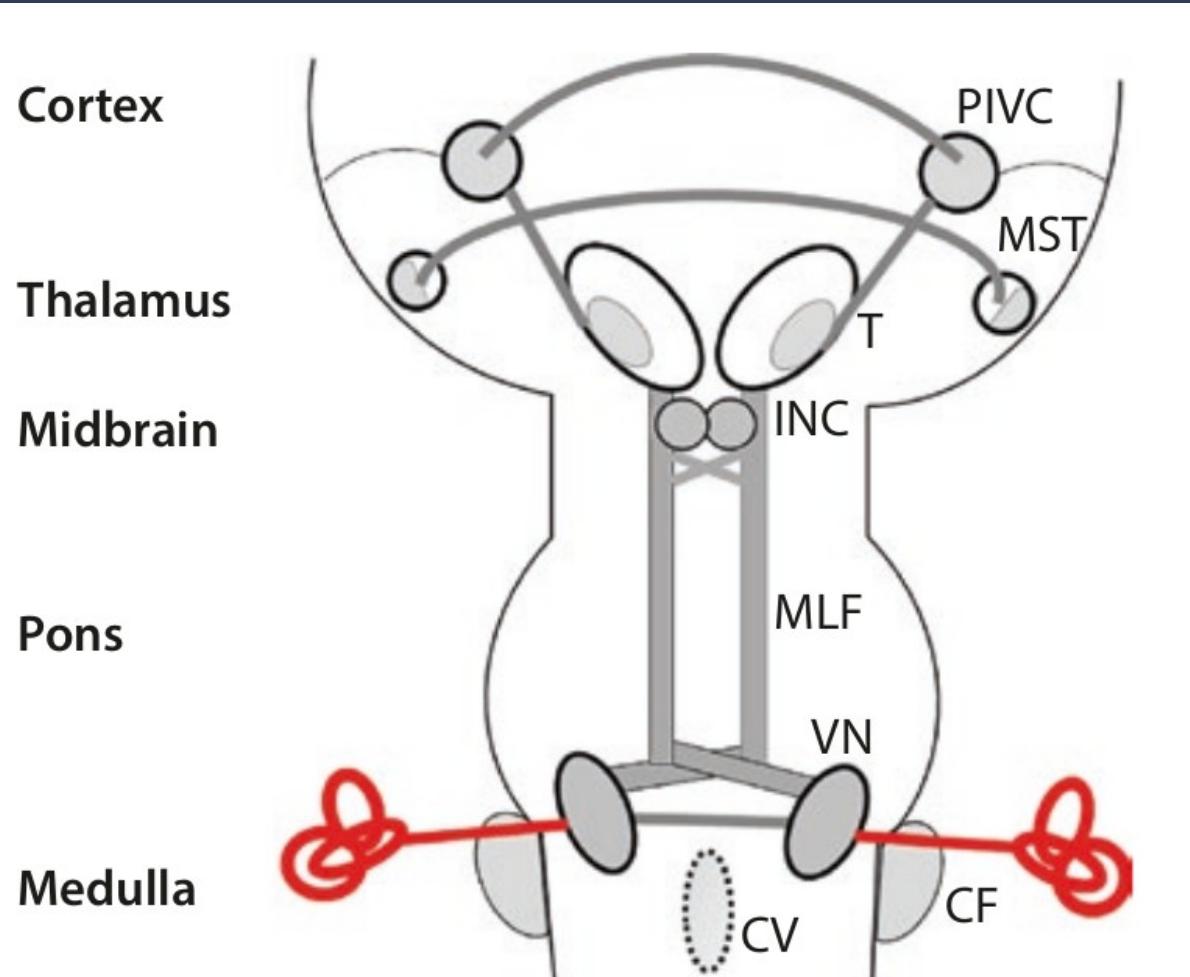


- **Vestibular nuc**
- **Ocular motor nuc, CN 3,4,6**
- **Integration centers**
 - **Midbrain (vertical/torsional eye movement): interstitial nuc of Cajal, INC and rostral interstitial nuc of MLF, riMLF**
 - **Pons (horizontal eye movement): nuc prepositus hypoglossi, NPH**
 - **Cerebellum**



Ascending Pathway to Cortex

- Thalamus
- Cortex: temporo-parietal cortex (parieto-insular vestibular cortex, PIVC; medial superior temporal area, MST)
- Perception
- Bilateral organization: at least 5 crossings (3 in brainstem, 2 between hemispheres)



4 Functional Advantages of Bilateral Organization

- Optimal differentiation of active and passive head motion
- Spatial orientation
- Sensory substitution (of a unilateral peripheral failure)
- Central compensation

3 Major Functional Groups in Central Pathway

- **Reflex control of gaze, head and body in 3 spatial planes (yaw, pitch, roll); brainstem-cerebellar level**
- **Perception of self-motion and control of voluntary movement and balance; cortical-subcortical level**
- **Higher vestibular cognitive function: spatial memory and navigation**

Peripheral vs Central

Anatomically → simple

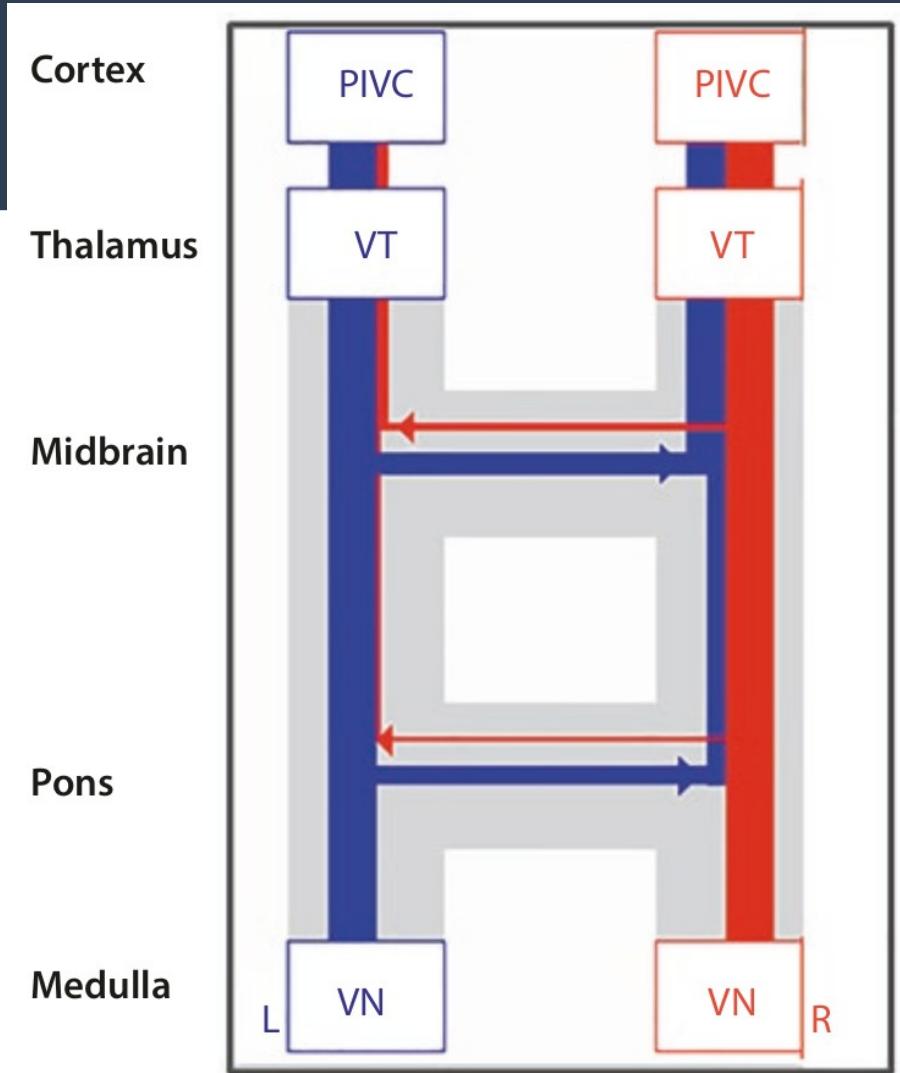
- Peripheral: labyrinth, vestibular nerve
- Central: vestibular nuc, brainstem, cerebellum, thalamus, cortex

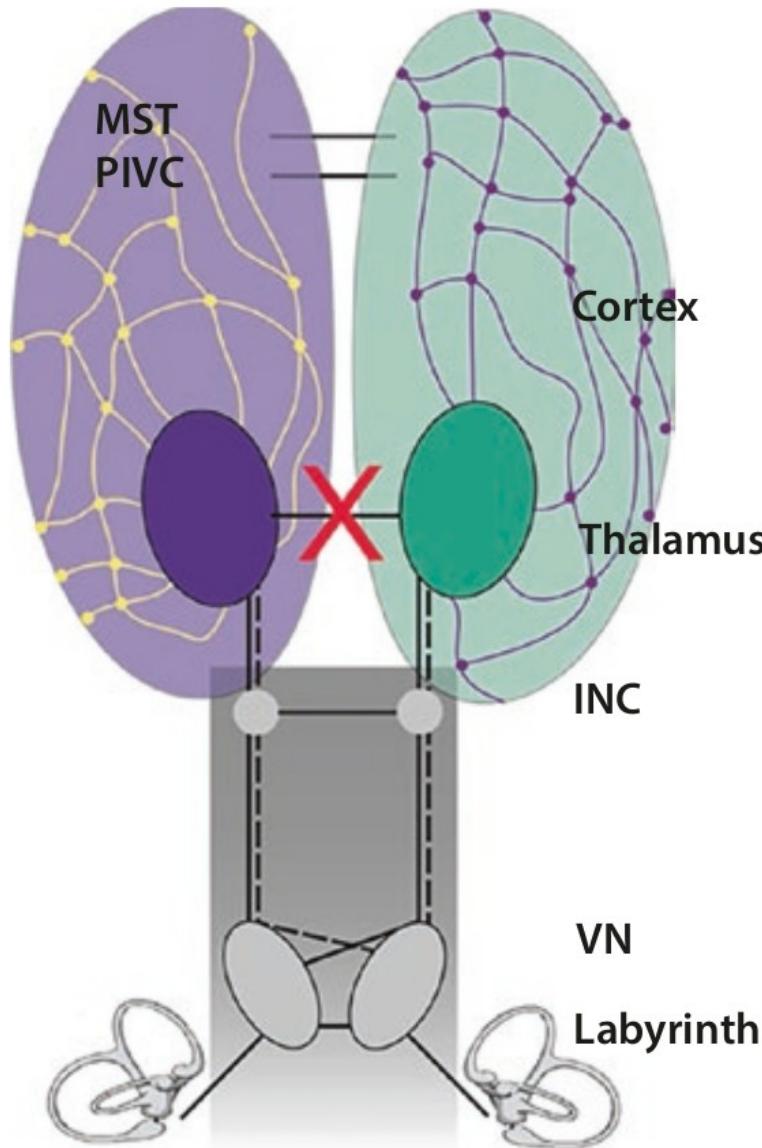
Clinically → some weaknesses

- Root entry zone of CN8, peripheral in anatomy, central pathology

Vestibular Cortical Network

- Right hemispheric dominance in right-handers
- Demonstrated by PET and fMRI via caloric, galvanic and auditory (otolith) stimulation
- Lateralization of spatial attention
- Clinical significance?





Cortex

vestibular dominance
2 crossings (PIVC, MST)

Thalamus

vestibular dominance
no crossing

Brainstem

INC vestibular dominance
 1 crossing

VN vestibular symmetry
 2 crossings

Labyrinth

vestibular symmetry

Reciprocal Inhibition between Vestibular, Visual and Somatosensory systems (cortex level)

- Activation of vestibular cortex → deactivation of visual and somatosensory cortex
 - Acute nystagmus → less amplitude of oscillopsia
- Visually induced self motion perception (parieto-occipital cortex activation) → deactivation of vestibular cortex
- Shifting the sensorial weight from the less to more reliable modality

Causes of Central Vestibular Disorders

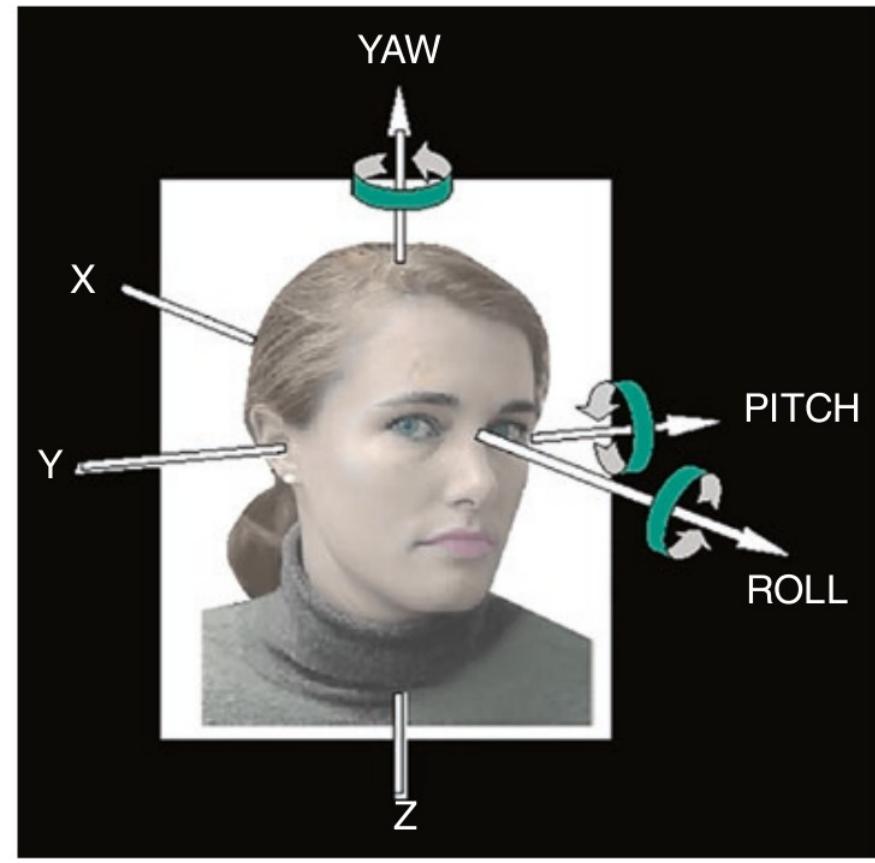
- Lesions
 - Infarction, hemorrhage, multiple sclerosis, tumor, degenerative
- Pathological excitation (rare)
 - MS, stroke, vestibular epilepsy

AVS: Central vs Peripheral

- 6-step procedure (for general neurologists)
 - Head impulse (not very specific, vHIT better)
 - Skew deviation (specific, not sensitive)
 - Spontaneous peripheral nys
 - Gaze-evoked nys, vertical nys
 - **Smooth pursuit, saccades**
 - **Romberg test**
- 6-step procedure is more sensitive than the 3-step HINTS test
- Cf: HINTS-plus (hearing)
- Cf: STANDING (Spontaneous Nystagmus, Direction, head Impulse test, standiNG)

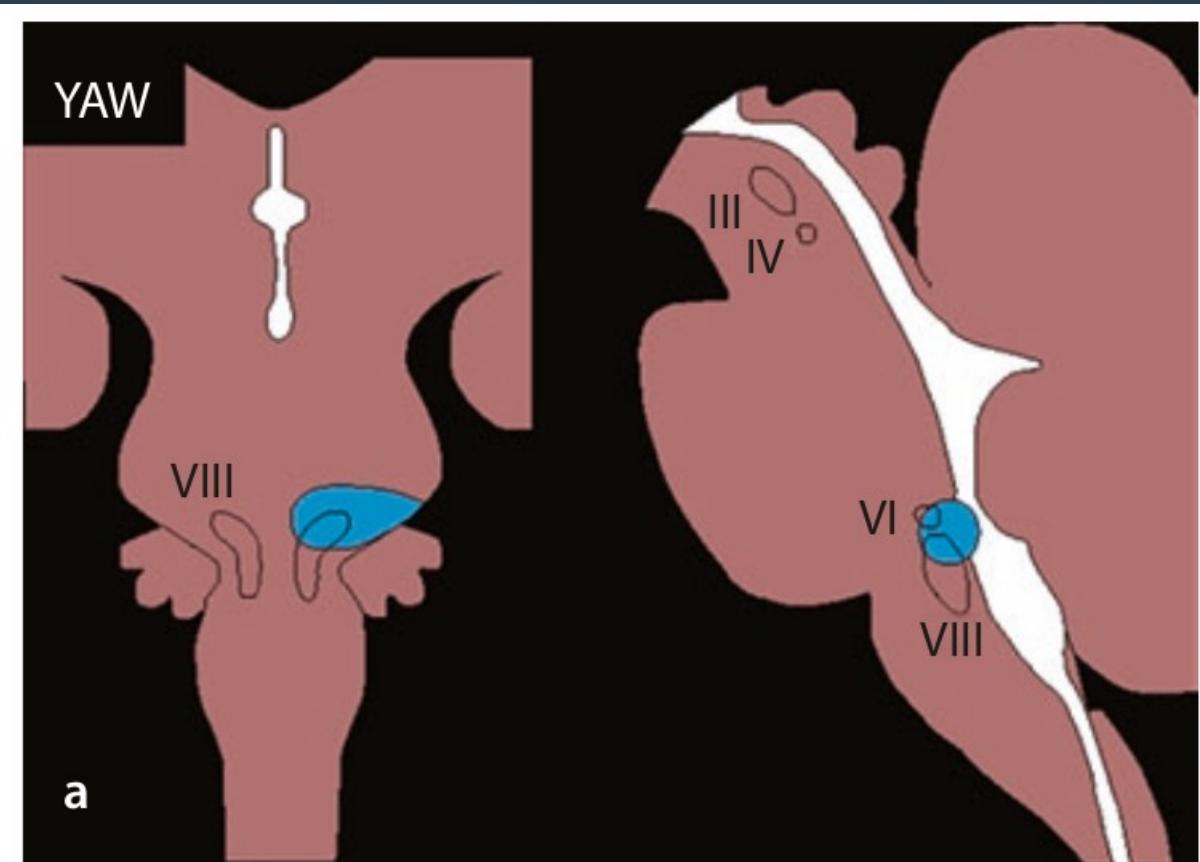
Central Vestibular Syndromes in 3 planes of VOR

- Yaw (horizontal) plane
 - Vestibular pseudoneuritis
 - Spontaneous horizontal nystagmus
- Pitch (sagittal) plane
 - DBN
 - UBN
- Roll (frontal) plane
 - OTR (ocular tilt reaction)



Horizontal (yaw) Plane

- Lesions in
 - Entry zone of vestibular nerve
 - Medial vestibular nuc
 - Integration center of horizontal eye movements: NPH, PPRF

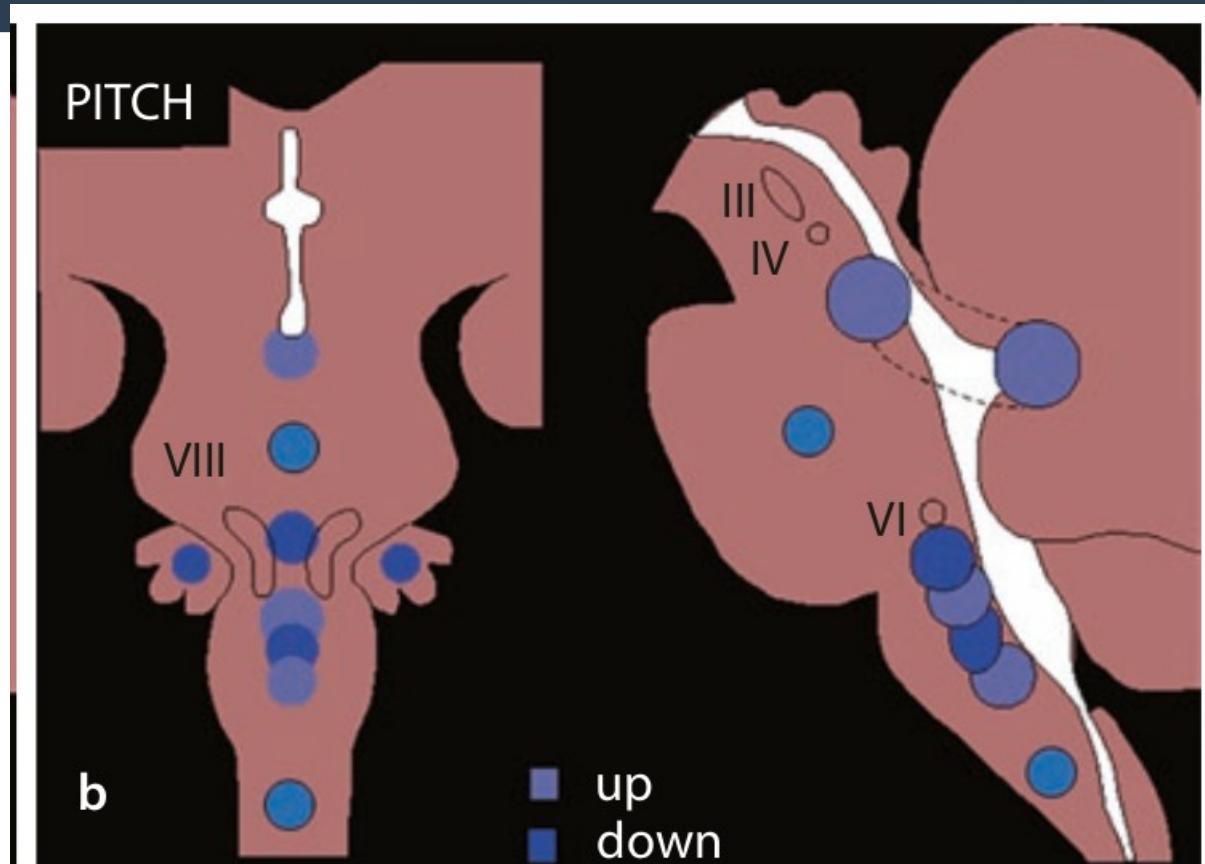


Horizontal (yaw) Plane - symptoms/signs

- Vestibular pseudoneuritis (skew is often present)
- Spontaneous horizontal nystagmus (not suppressed by visual fixation)
- Horizontal past-pointing
- Postural instability
- Lateropulsion
- Ipsilateral caloric unresponsiveness
- Horizontal gaze deviation
- Fall to affected side
- Deviation of subjective straight-ahead

Sagittal (pitch) Plane

- Lesions in
 - Bilateral paramedian medullary, pontomedullary, pons, pontomesencephalic brainstem
 - Bilateral flocculus, paraflocculus



Sagittal (pitch) Plane - symptoms/signs

- Downbeat nys, upbeat nys
- Subjective horizontal deviation, up- or downward
- Past-pointing to up/down
- Tendency to fall forward or backward

Downbeat Nystagmus Syndrome

- Most common acquired fixational nystagmus
- Nystagmus aggravated on lateral gaze and head-hanging position, morning > afternoon
- Tendency to fall backward
- Past-pointing upward
- Vertical pursuit abnormality
- OKN abnormality
- Abnormal visual suppression of VOR
- Mechanism: bilateral flocculus/PFL dysfunction
- Causes: idiopathic , degenerative (including CANVAS), lesions (vascular, MS..), AED intoxication, paraneoplastic...

Causes of DBN

Table 1. Causes of downbeat nystagmus

	Halmagyi <i>et al.</i> [6] (n=62)	Yee [5] (n=91)	Wagner <i>et al.</i> [1] (n=117)	Weighted frequency (of 270 patients)
Idiopathic	27	5	45	77 (28.5%)
Cerebellar degeneration, including spinocerebellar ataxia and acquired cerebellar degeneration	15	26	23	64 (23.7%)
Cerebellar ectopias, mainly Arnold–Chiari malformations	17	11	8	36 (13.3%)
Infarction and vascular lesions	0	25	10	35 (13.0%)
Multiple sclerosis	0	12	4	16 (5.9%)
Toxic	1	4	5	10 (3.7%)
Tumours	1	3	4	8 (3.0%)
Trauma	0	3	2	5 (1.9%)
Infectious	0	1	4	5 (1.9%)
Paraneoplastic	1	0	4	5 (1.9%)
Other	0	0	5	5 (1.9%)
Metabolic	1	0	3	4 (1.5%)

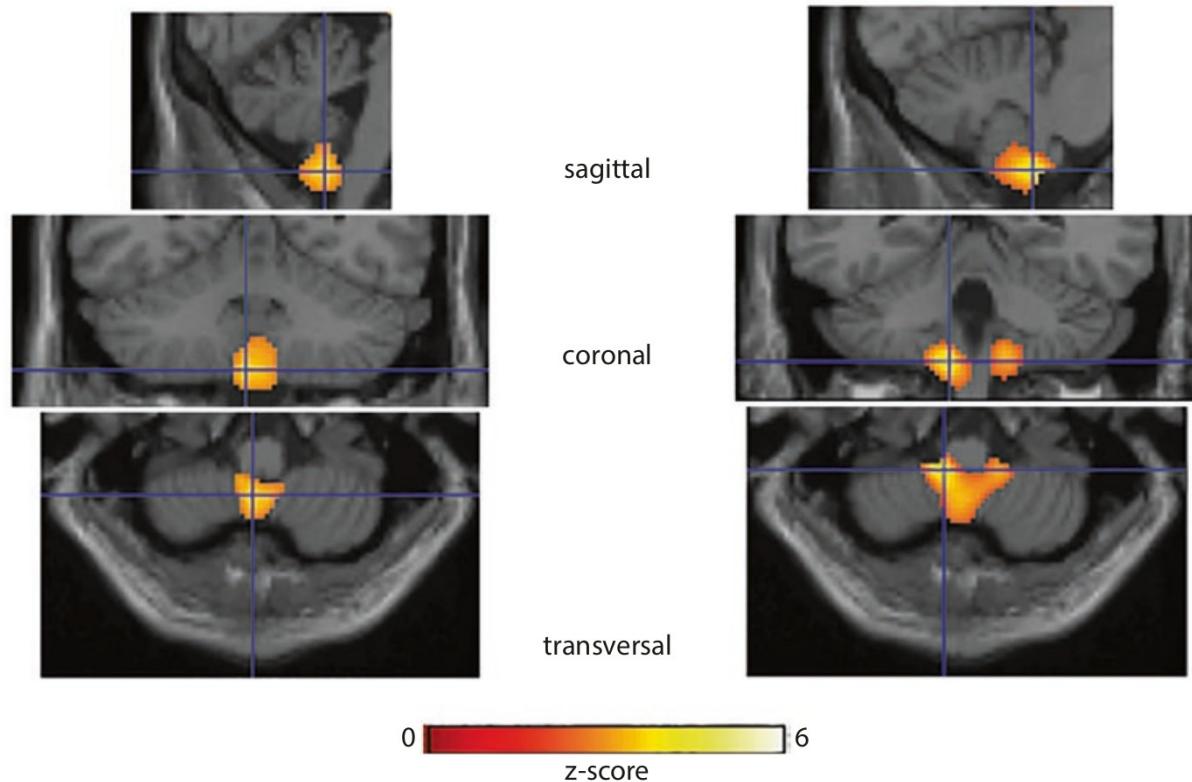
Tran, T. M., Lee, M. S., & McClelland, C. M. (2021). Downbeat nystagmus: A clinical review of diagnosis and management. *Curr Opin Ophthalmol*, 32(6), 504–514.

DBN – Pathomechanism not fully clarified...

- Hypotheses include:
**vertical saccadic system,
vertical pursuit system,
central connection of
otolith pathway, focculus
and paraflocculus....**
- **Focculus and
paraflocculus: inhibit the
pathway of superior
vestibular nuc**
- **Damage of focculus and
paraflocculus →
disinhibition of the
pathway of superior
vestibular nuc**

Idiopathic DBN syndrome

- Reduced FDG-PET in flocculus and paraflocculus
- Improved PET and S/S after 4-aminopyridine treatment

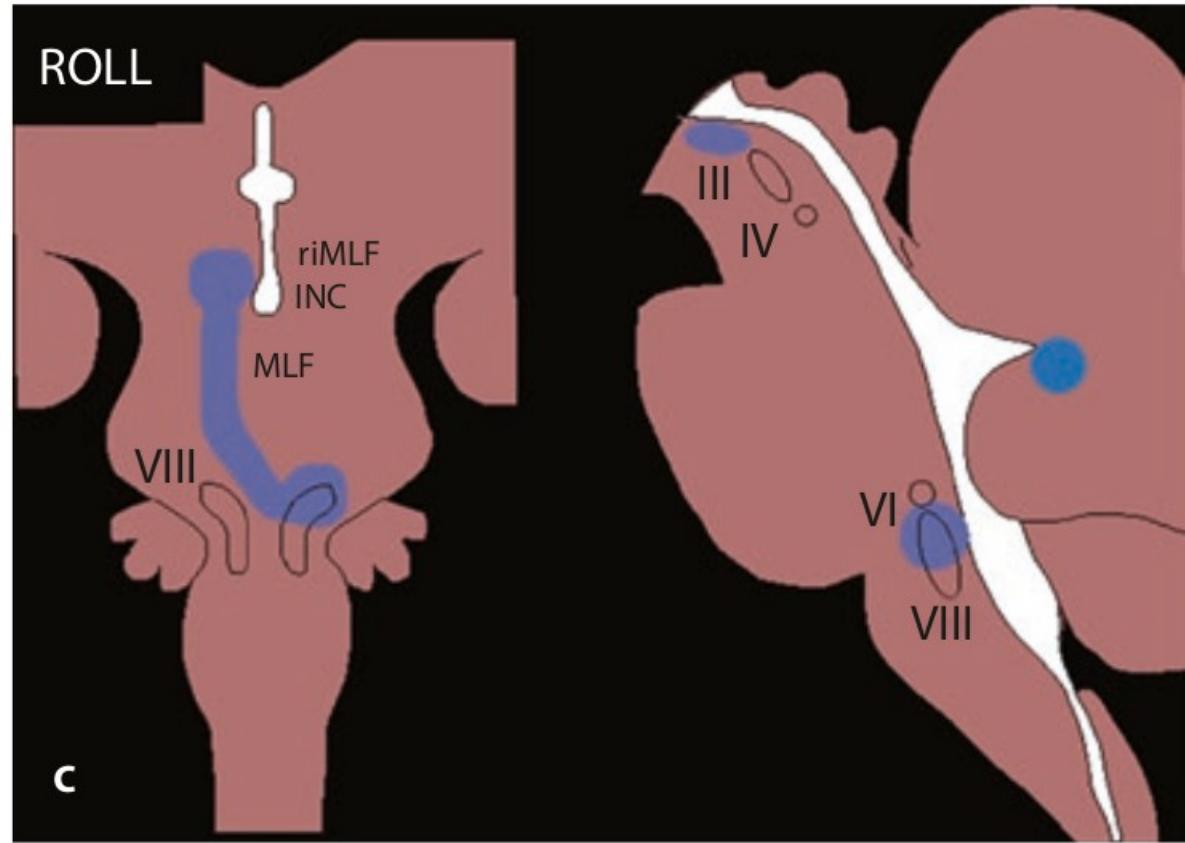


Treatment for DBN

- 4-aminopyridine (potassium channel blocker)
 - 3,4-diaminopyridine (potassium channel blocker)
 - Fampridine (CR of 4-AP)
 - Chlorzoxazone
 - Acetyl-DL-Leucine
 - Memantine
 - Gabapentin
- UBN is usually self-limited, drug treatment is not necessary

Frontal (roll) Plane

- Lesions in graviceptive pathway (utricle→vest nuc→MLF→riMLF, IN C..)



Frontal (roll) Plane – symptoms and signs

- SVV deviation (perceptive)
tested by bucket test or
half-spherical dome
- Ocular tilt reaction
 - Head tilt
 - Skew deviation
 - cyclotorsion

A sensitive sign for acute brainstem lesion!!
Infarction, hemorrhage 最常見

Physiologic OTR (ocular tilt reaction)

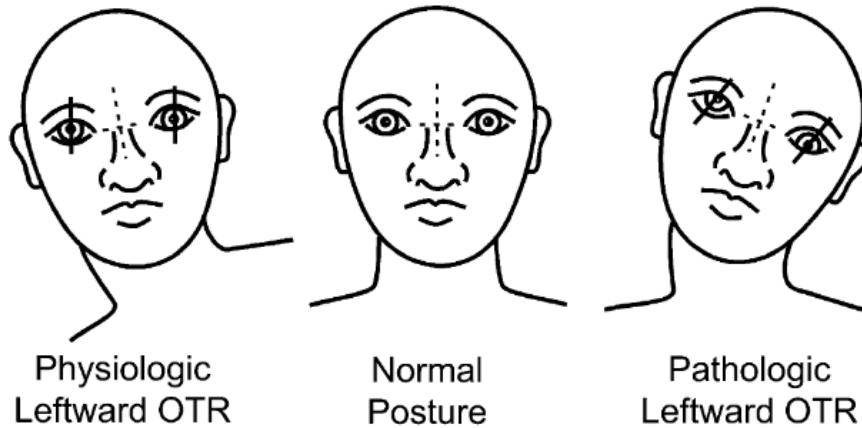
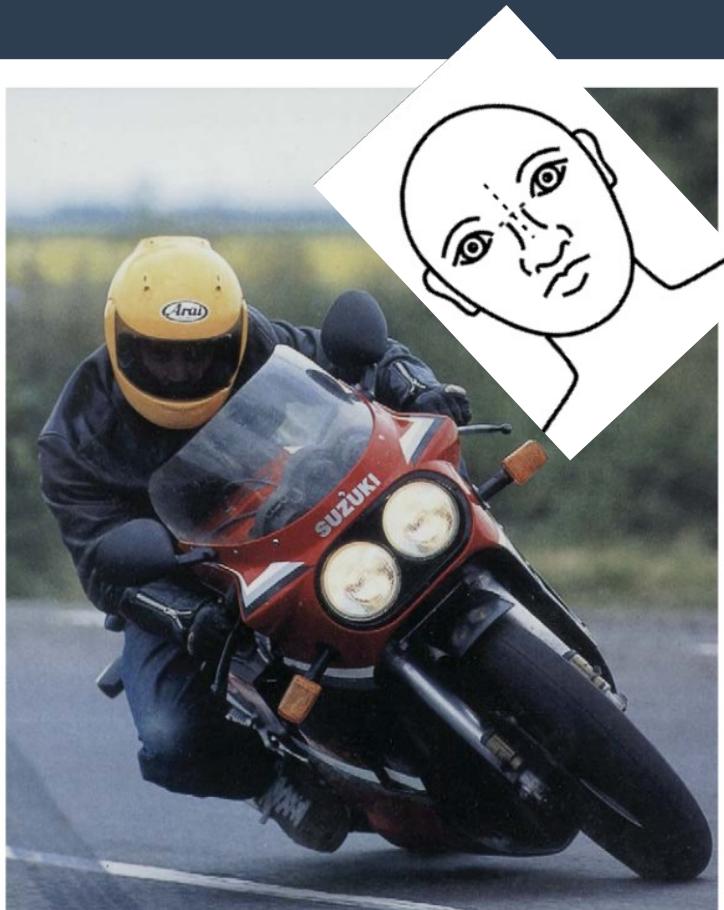


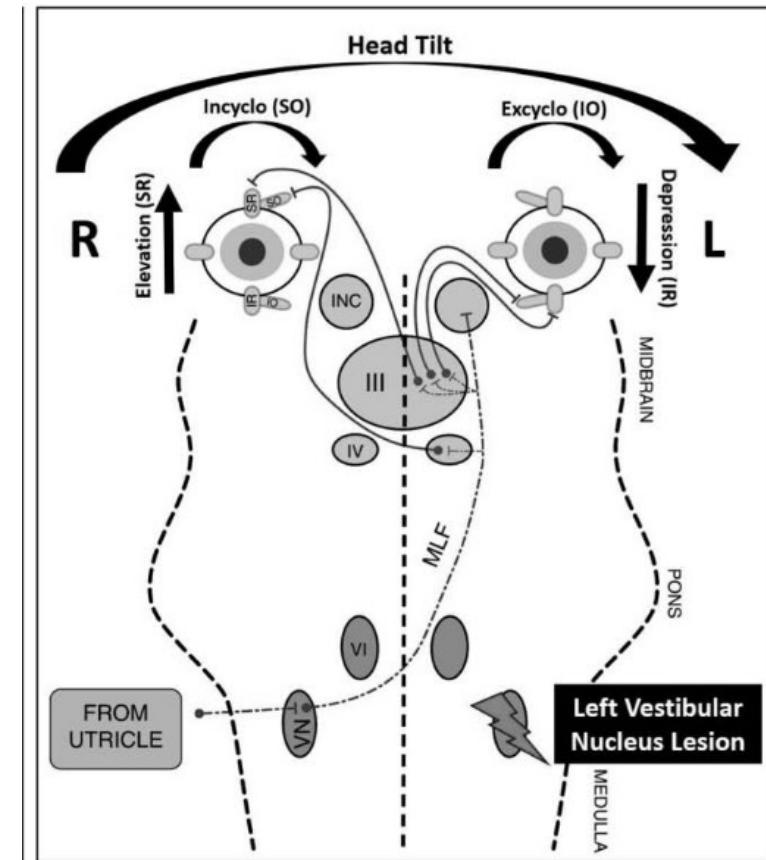
Fig. 2. Figure showing physiologic and pathologic skew deviation. In the physiologic ocular tilt reaction (*left*), the compensatory head tilt predominates, with only a small skew deviation or static ocular counterroll. In the pathologic ocular tilt reaction (*right*), all three components of the ocular tilt reaction are present. (Reprinted from Brodsky³⁷ with permission of the American Medical Association.)

Pathologic OTR – L utricle-vest nuc (below decussation)



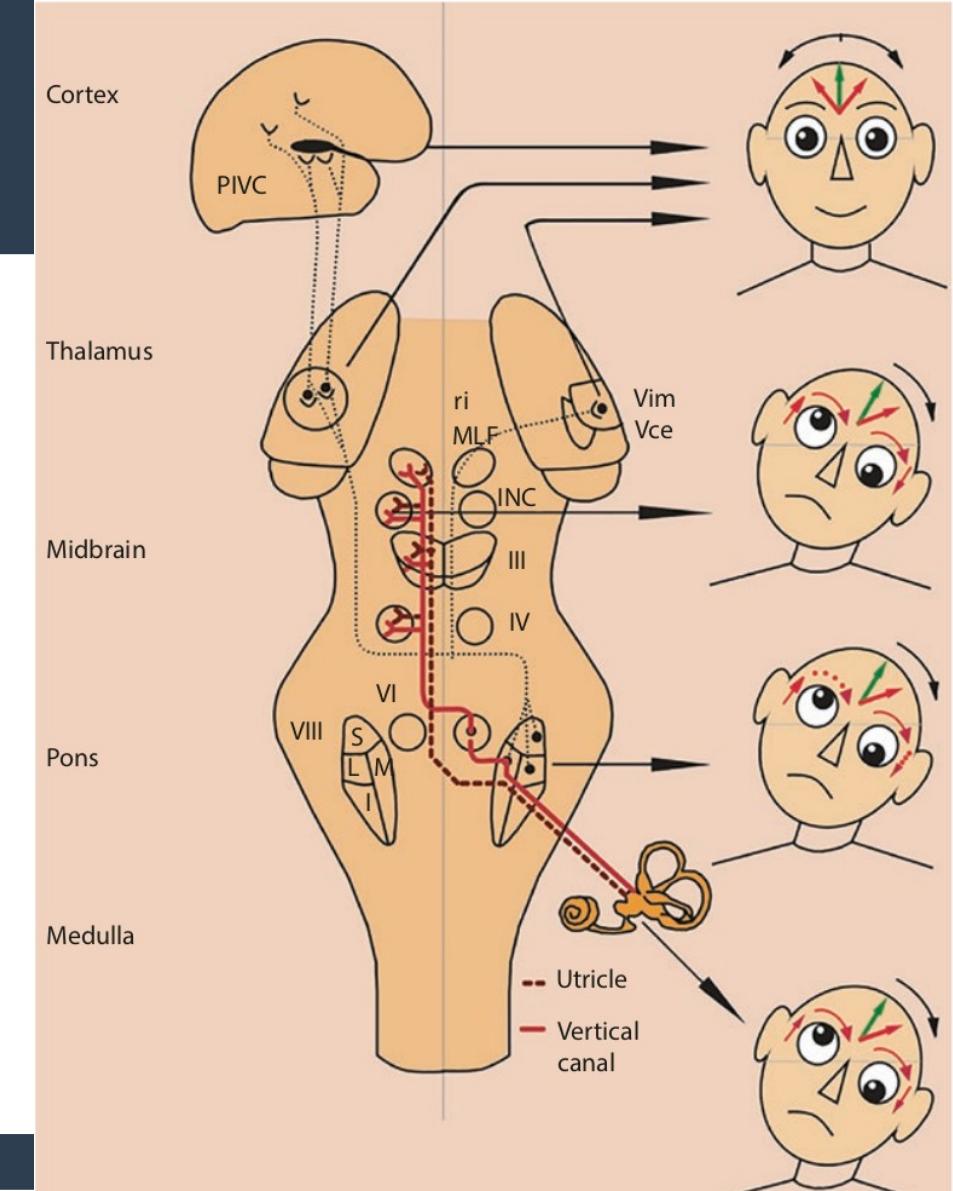
Pathologic
Leftward OTR

偏向病灶 (中樞 >> 週邊) 同側



Summary of Pathologic OTR

- Peripheral and vest nuc (medulla): 偏向病灶側
- MLF, INC, riMLF (pons, midbrain): 偏向健康側
- 合併 CN3,4: 可能出現 mixed pattern (兩眼 oculotorsion 方向相反)
- Thalamus-cortex: 僅出現 SVV deviation , 無 skew, head tilt, cyclotorsion
- Vestibulocerebellum: 偏向健康側或病灶側



OTR due to Vestibular Nerve Pathology (VN, neurectomy...)

In this setting, skew deviation generally disappears within several days while the conjugate cyclotorsion and tilting of the static visual vertical may persist for weeks to months.

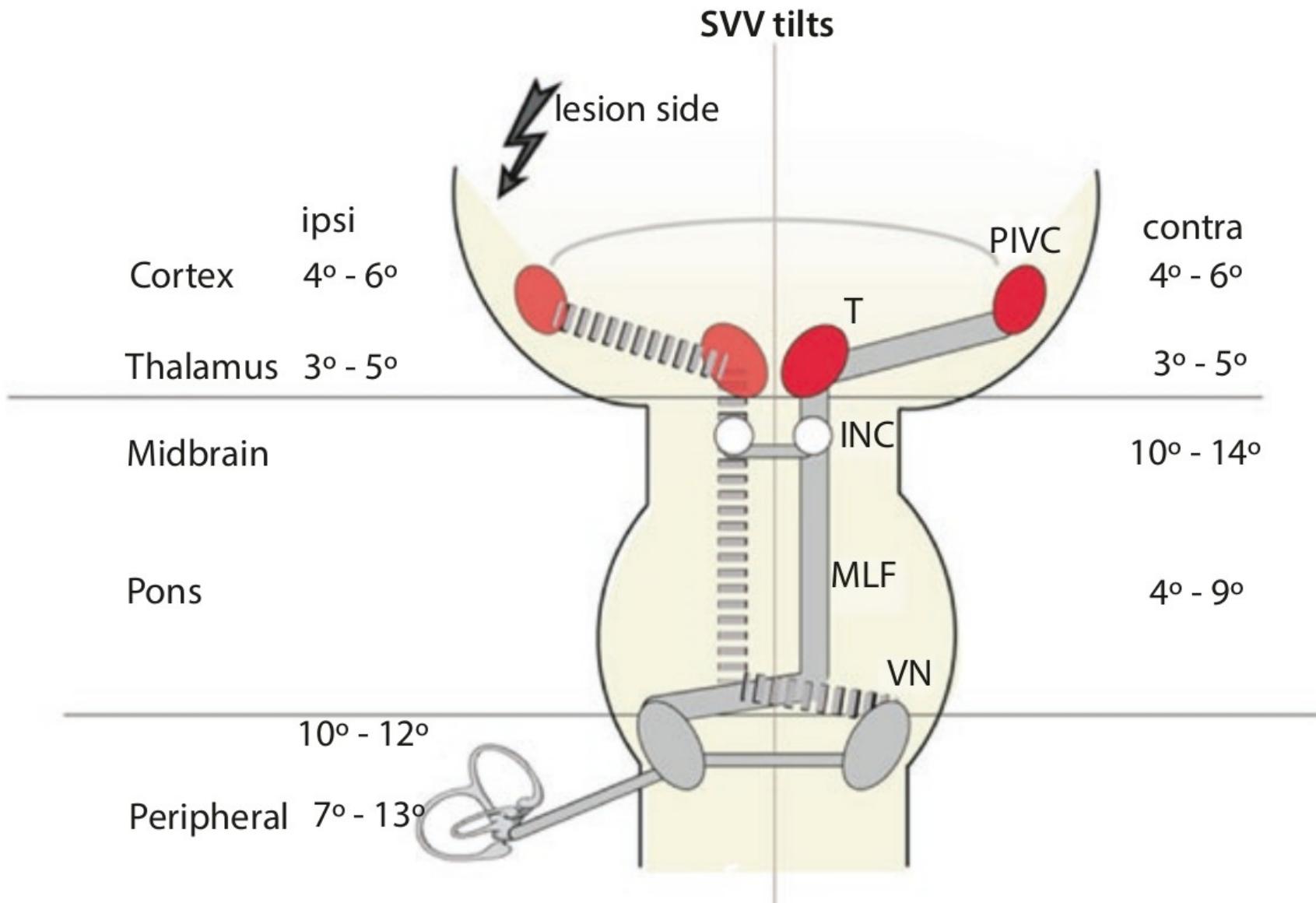
Brodsky, M. C., Donahue, S. P., Vaphiades, M., & Brandt, T. (2006). Skew Deviation Revisited. Survey of Ophthalmology, 51(2), 105–128.
<https://doi.org/10.1016/j.survophthal.2005.12.008>

Bucket test of SVV

(subjective visual vertical)

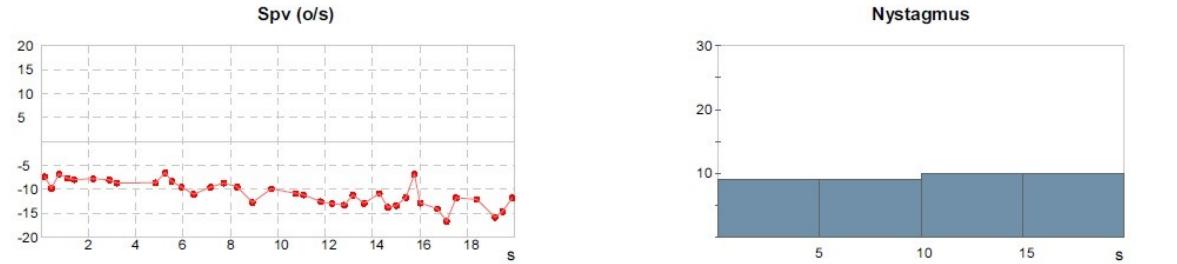


Utriculo-ocular pathway, normal deviation $\leq 2.5^\circ$

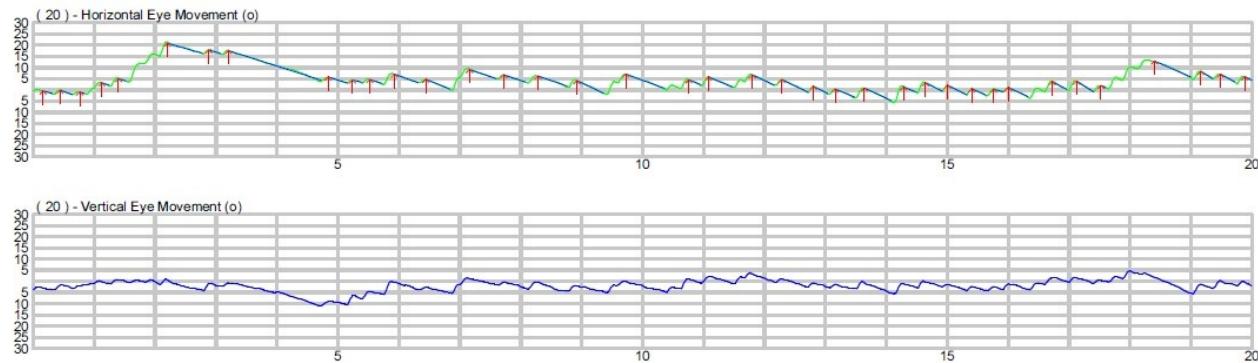


44 y/o, AVS, vomiting and unsteadiness, HA-, aural-

[Day 5]
VNG: upright
sitting,
complete
darkness

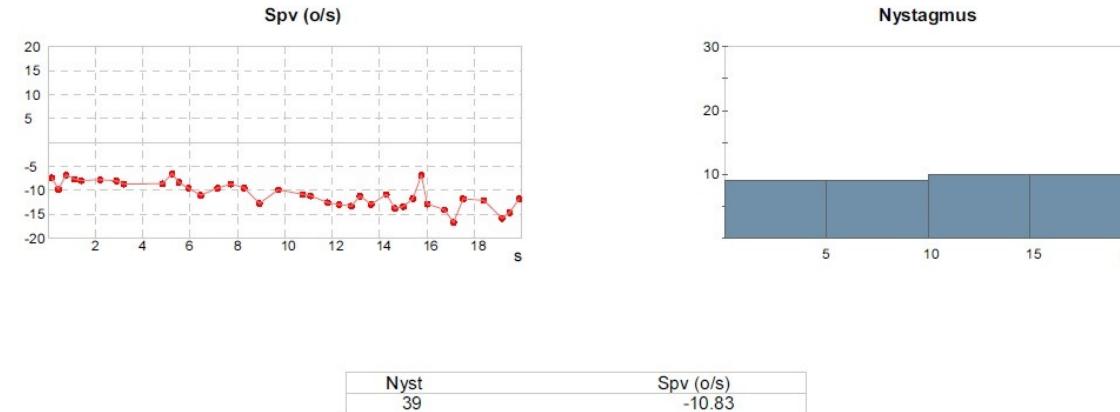


Nyst	Spv (o/s)
39	-10.83

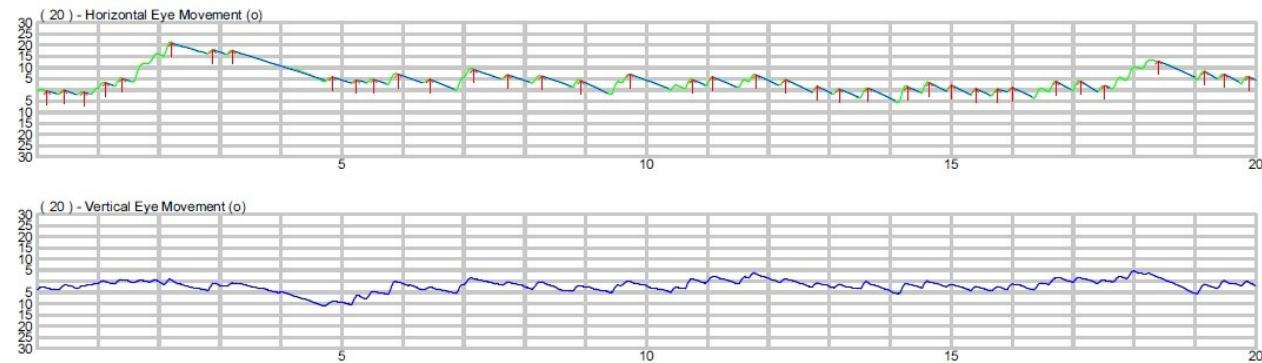


44 y/o, AVS, vomiting and unsteadiness, HA-, aural-

RBN with
upbeating
component
→ left sup. VN



(supported by
left HIT+, no
apparent skew,
Fukuda fall to
left)



Qizz: What is the most likely result of bucket test?

僅示意圖非當事人



?

- (1) no SVV tilt (<2°)**
- (2) SVV tilt to right**
- (3) SVV tilt to left**

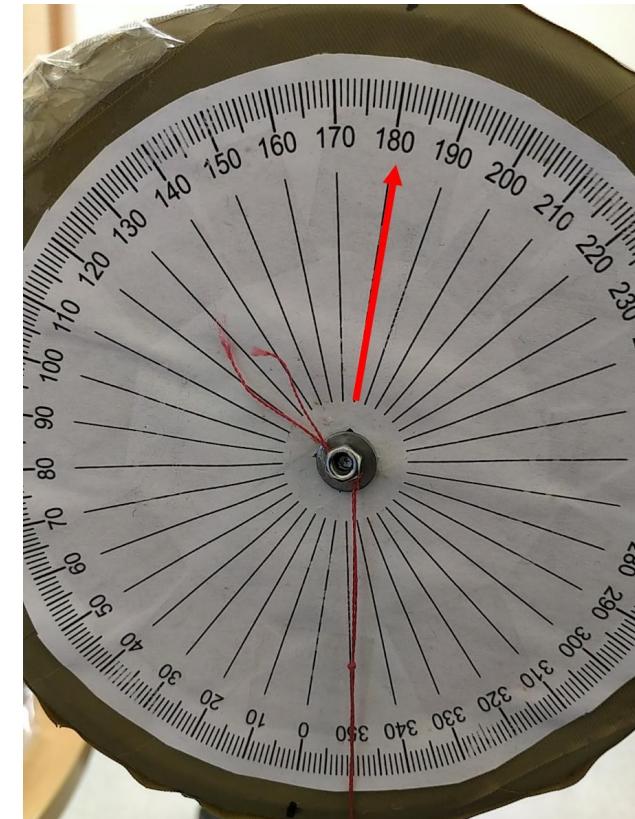
Qizz: What is the most likely result of bucket test?

僅示意圖非當事人

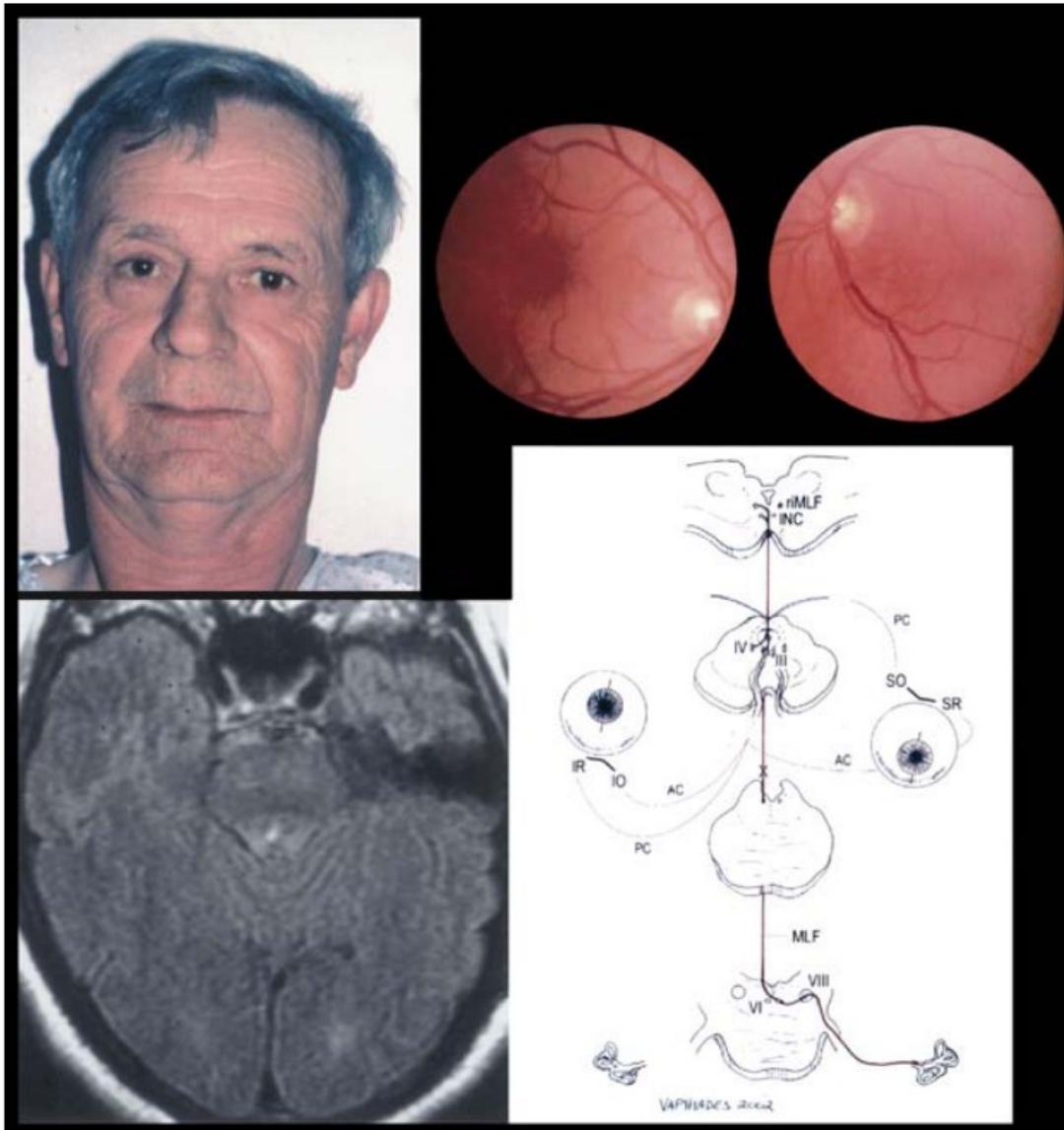


?

- (1) no SVV tilt ($<2^\circ$)
- (2) SVV tilt to right
- (3) SVV tilt to left
 10°



Right MLF lesion
→ Left OTR



Brodsky, M. C., Donahue, S. P., Vaphiades, M., & Brandt, T. (2006). Skew Deviation Revisited. Survey of Ophthalmology, 51(2), 105–128.

<https://doi.org/10.1016/j.survophthal.2005.12.008>

CN3,4 problem 有時看起來像 skew ，也出現 cyclotorsion... 作單眼 SVV 可以鑑別！

day 2

Right Eye

Left Eye

SVV Tilt

(mono / bino)

mono +**17°**

bino +**19°**

mono +**15°**

Vestibular disorder

Torsion: **15°**

pathological
excyclotropia



Normal: excyclotorsion 0-10 degree

Torsion: **7°**
pathological
incyclotropia

day 3

Paretic Eye

Non-paretic Eye

SVV Tilt
(mono / bino)

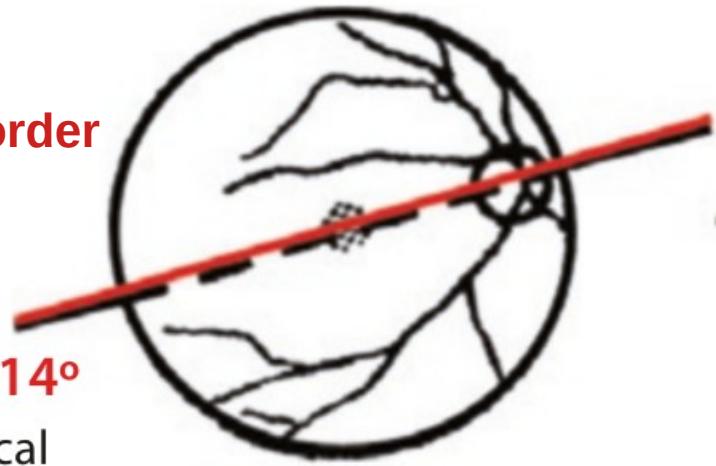
mono +**7.5°**

bino +0,7°

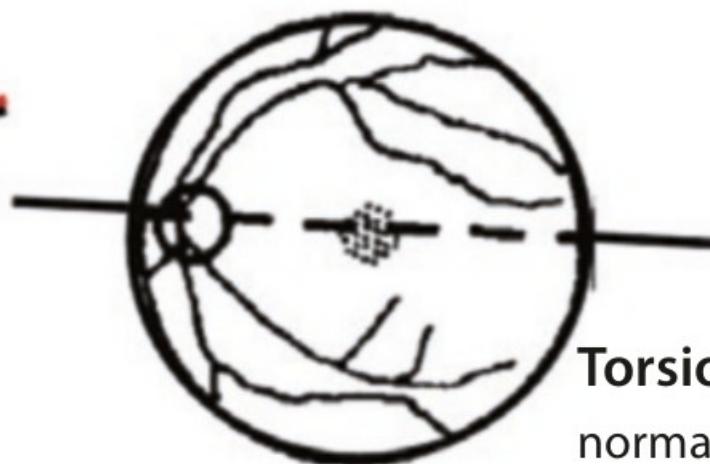
mono +1.9°

oculomotor disorder

Torsion: 14°
pathological
excyclotropia



Normal: excyclotorsion 0-10 degree



Torsion: 3°
normal
excyclotropia

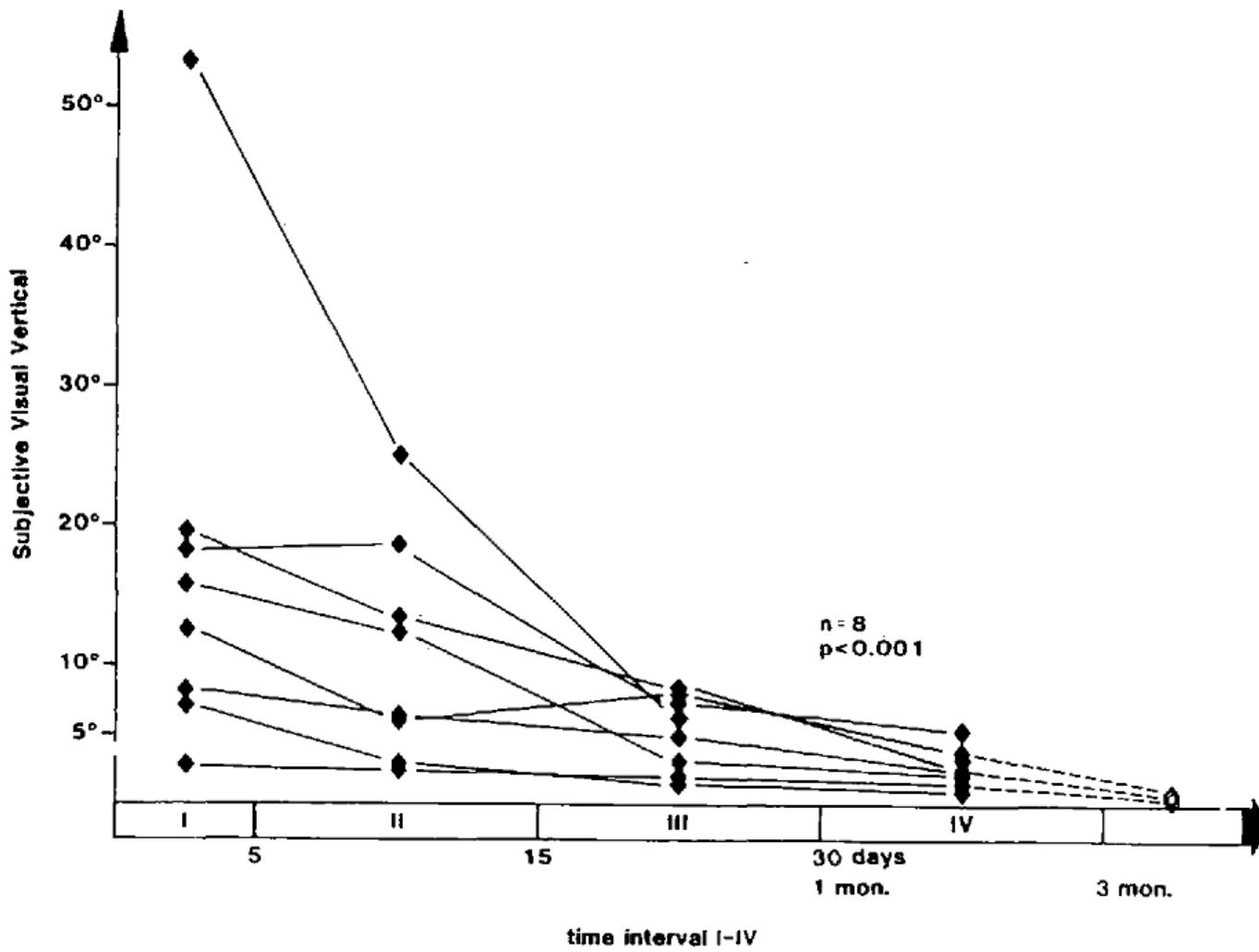
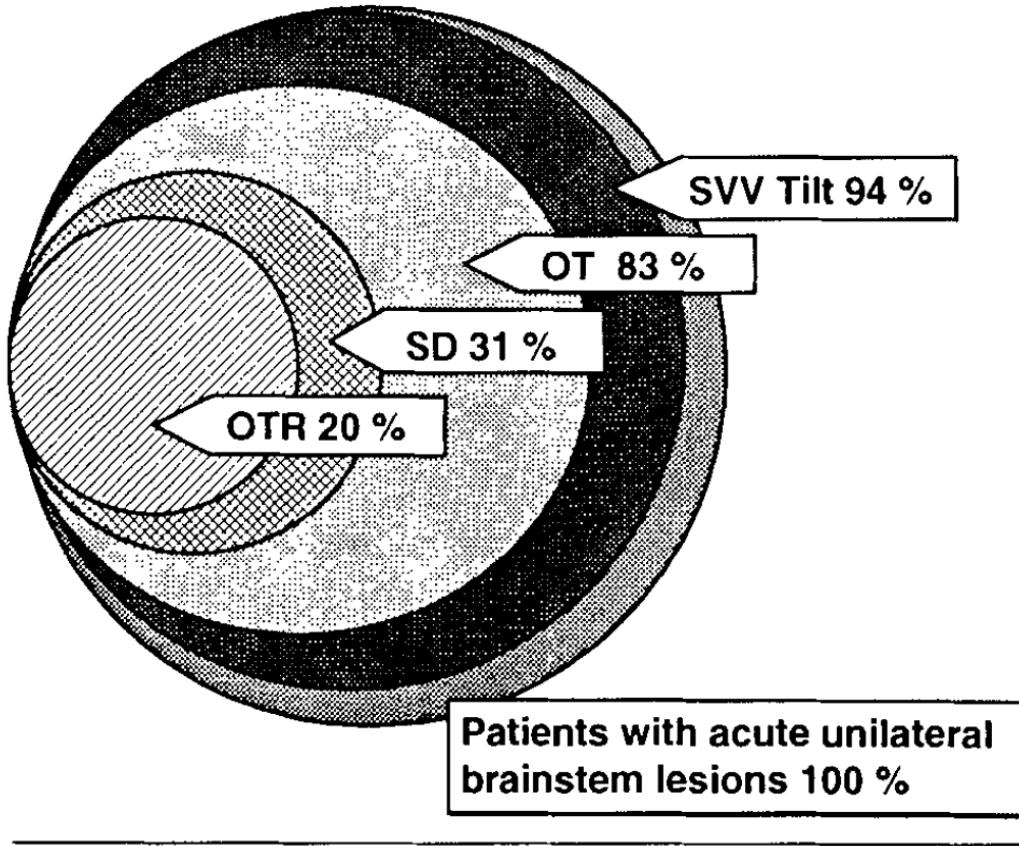


Figure 6. Time course of change in deviation of subjective visual vertical (SVV) after acute unilateral infarction of the lateral medulla oblongata in 8 patients with Wallenberg's syndrome. Friedman nonparametric analysis of variance shows a significant difference between SVV values for different time intervals ($p < 0.001$), and examination of the individual differences (Wilcoxon and Wilcoxon 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.

111 acute unilateral brainstem infarction



For acute brainstem stroke

Complete OTR (triad): high specificity, low sensitivity

Skew deviation: high specificity, low sensitivity

SVV tilt: high sensitivity, low specificity

Brandt, T., & Dieterich, M. (1994). Vestibular syndromes in the roll plane: Topographic diagnosis from brainstem to cortex. *Annals of Neurology*, 36(3), 337–347.
<https://doi.org/10.1002/ana.410360304>

Central dizziness - features

- **Labyrinth, vestibular nerve, vest nuc, vestibulocerebellum**
- → **sustained spinning + nystagmus**
- **Above nuc (midbrain, thalamus, cortex)**
- → **to and fro, swaying, disorientation, postural instability, OTR**
- **Exceptions exist, but are very rare**

Dizziness due to thalamic lesion

- Thalamus is a portion of central vestibular pathway
- Swaying dizziness (rarely rotational)
- Postural instability
- SVV deviation, ispi- or contralateral (no head tilt, skew, ocular torsion)
- Thalamic astasia: transient tendency to fall

Higher (cognitive) vestibular syndromes

- **Hemispatial neglect**
- **Room tilt illusion**
 - Paroxysmal transient upside-down vision
 - Caused by vestibular dysfunction (brainstem or peripheral) → transient mismatch of vestibular and visual coordinate system
 - Ex: BV, MD
- **Pusher syndrome (figure):**
 - 10% of stroke pt; thalamus, posterior insula involvement (cortical vestibular network)
- **Impaired spatial memory and navigation**
 - Central: dementia, TGA; Peripheral: BV, chronic unilateral deficit

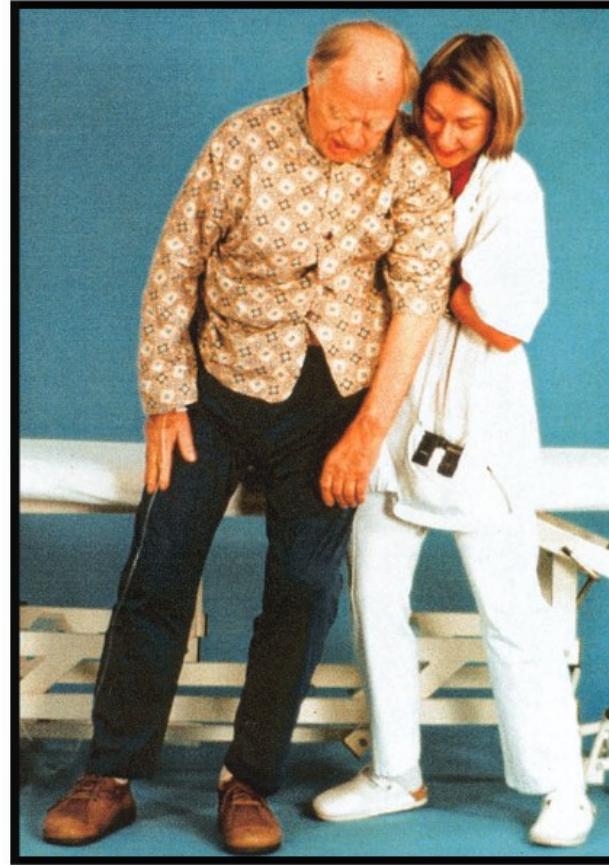
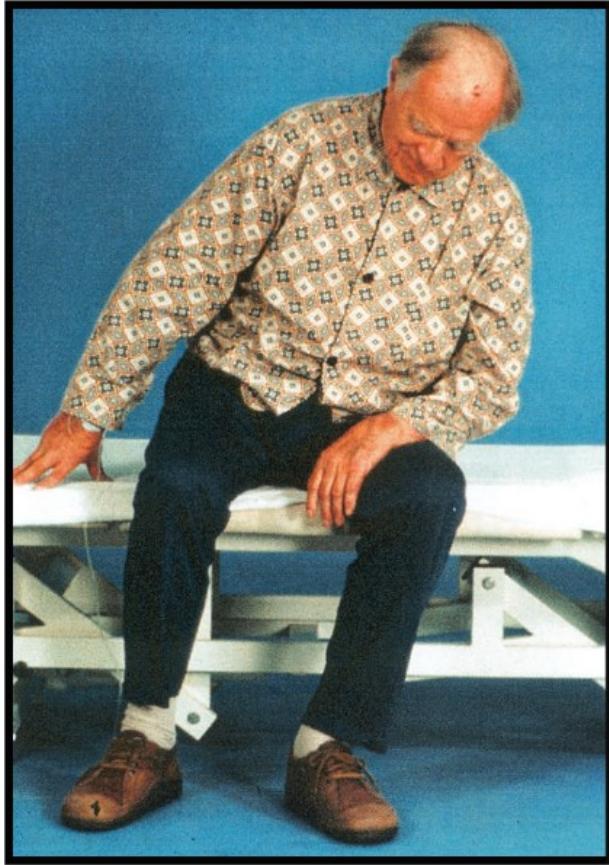


Figure 1.

A patient with right-side brain damage and pusher syndrome. The characteristic feature of the disorder is that these patients, while sitting (left) or standing (right), spread the nonparetic extremities from the body to push away actively from the nonparetic side. The result is the typical tilted body posture of these patients. If not assisted by the examiner, the patients push themselves into a lateral inclination until they fall toward the hemiparetic side.

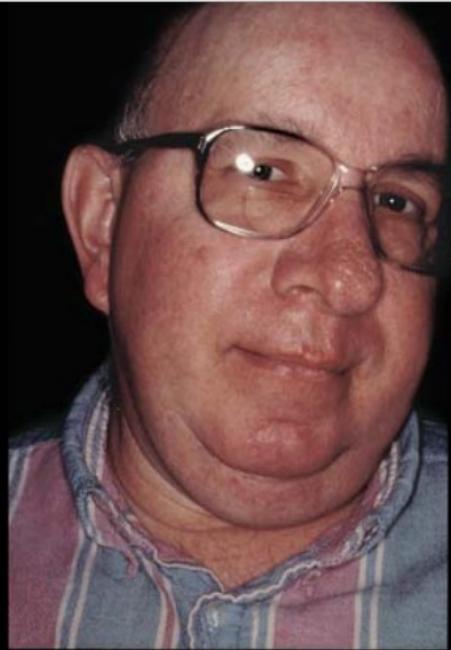
Cerebellar Dizziness/Vertigo

- Symptoms: swaying dizziness, postural imbalance, fall, dysarthria, dysphagia, limb ataxia
- Oculomotor signs: impaired saccades, pursuit, visual suppression of VOR; fixational nystagmus (DBN..), ocular misalignment
- Causes: degenerative, hereditary, vascular, paraneoplastic, autoimmune, inflammatory, toxic...

Table

Brain site/clinical syndrome	Artery
Medulla oblongata	<p>Wallenberg's syndrome (DVD) with OTR and its features (head tilt, vertical deviation of the eyes, ocular torsion, deviation of the SVV) ipsiversive: lesion of the medial vestibular nuclei</p> <p>Branches of the vertebral artery or the posterior inferior cerebellar artery (PICA) Rare: posterior spinal artery</p>
	<p>Acute vestibular syndrome (DVD) = “Vestibular pseudo-neuritis”: lesion of the vestibular fascicle.</p> <p>Branches of the vertebral artery or PICA</p>

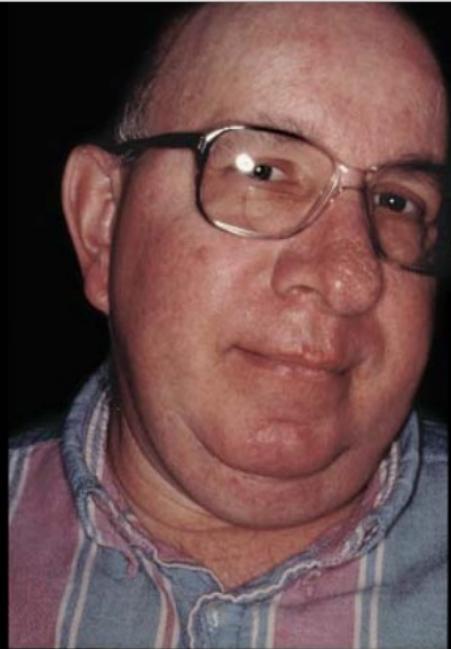




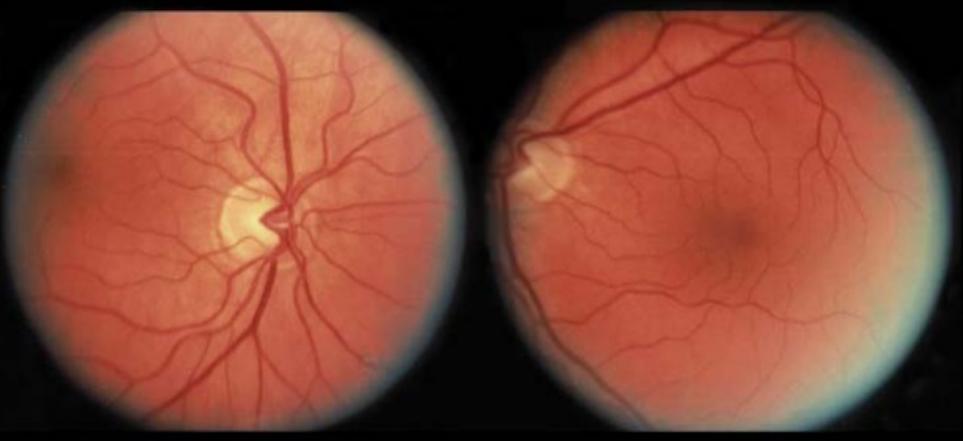
Field Measurements



Right CN4 palsy or Left OTR?



Field Measurements



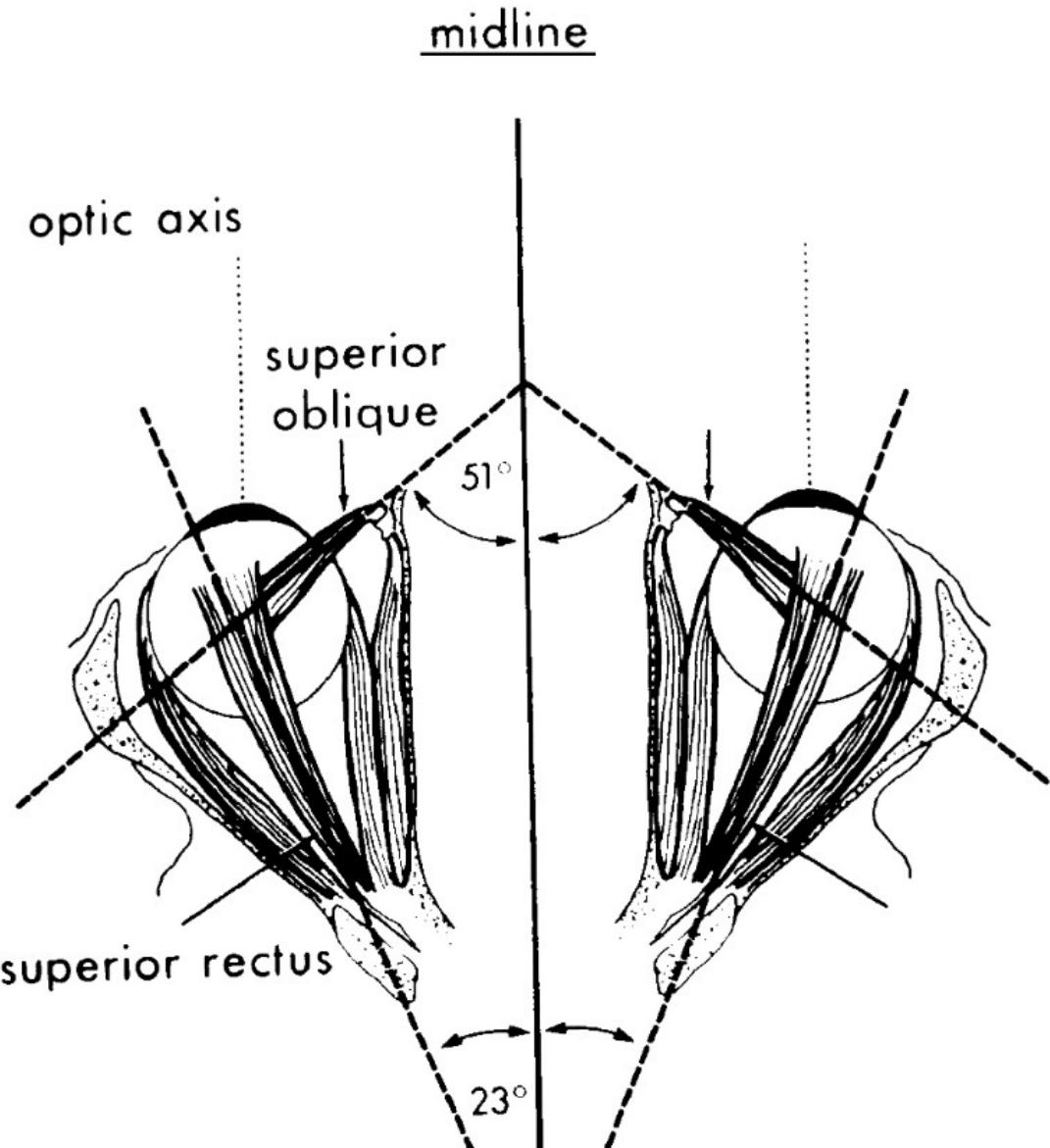
Right CN4 palsy or Left OTR?

TABLE 2

Ocular Tilt Reaction Versus Superior Oblique Palsy

Prenuclear Lesion (Ocular Tilt Reaction)	Superior Oblique Palsy
Intorsion of higher eye/ extorsion of the lower eye	Extorsion of higher eye
Binocular tilt of subjective visual vertical	Monocular tilt of subjective visual vertical
Head tilt compensatory for altered subjective visual vertical	Head tilt compensatory for vertical diplopia

- CN4→ sup. obliq (SO₄)
- SO 作用：depression, intorsion
- CN4 palsy→ elevation and extorsion
- Cf: in OTR, intorsion of higher eye



THE END