Vestibular paroxysmia

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Vestibular paroxysmia

Leading symptom:

Recurrent short-lasting spontaneous attacks of spinning or non-spinning vertigo

Vestibular paroxysmia

(each point needs to be fulfilled):

1. At least ten attacks of spontaneous spinning or non-spinning vertigo
2. Duration less than 1 min
3. Stereotyped phenomenology in a particular patient
4. Response to treatment with a sodium channel blocker
5. Not better accounted for by another diagnosis

Box 11.1 Modified Diagnostic Criteria for Vestibular Paroxysmia According to the Classification Committee of the Bárány Society (Strupp et al.2016b)

Probable vestibular paroxysmia
(each point needs to be fulfilled):

1. At least five attacks of spinning or non-spinning vertigo
2. Duration less than 5 min
3. Spontaneous occurrence or provoked by certain head movements
4. Stereotyped phenomenology in a particular patient
5. Not better accounted for by another diagnosis

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International classification of vestibular disorders (ICVD)

Vestibular symptoms	*J Vestib Res.*2009;19:1–13.
Vestibular migraine	*J Vestib Res.*2012;22:167–172.
Menière's disease	*J Vestib Res.*2015;25:1–7.
BPPV	*J Vestib Res.*2015;25:105–117.
Vestibular paroxysmia	*J Vestib* Res.2016;26:409–415.
PPPD	*J Vestib* Res.2017;27:191–208.
Bilateral vestibulopathy	*J Vestib Res.*2017;27:177–89.
Hemodynamic orthostatic dizziness/vertigo	*J Vestib Res.*2019;29:45–56.
Presbyvestibulopathy	*J Vestib Res.*2019;29:161–170.
Mal de débarquement syndrome	*J Vestib Res.*2020;30:285–293.
Vestibular migraine of childhood and recurrent vertigo of childhood	*J Vestib Res.*2021;31:1–9.

Epidemiology

- No data
- A rare disease
- The relative frequency of VP in a group of 39,918 patients with vertigo and dizziness in a tertiary care center was around 3.1%.

Chang TP, Wu YC, Hsu YC (2013) Vestibular paroxysmia associated with paroxysmal pulsatile tinnitus: a case report and review of the literature. Acta Neurol Taiwanica 22(2):72–75

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Table 1.1 Absolute numbers and relative frequency of the various vestibular syndromes and diseases in the multiregional specialized outpatient unit of the German Center for Vertigo and Balance Disorders and the Neurological Clinic of LMU Munich (2019–2021)

Diagnosis	Frequency	
	n	%
1. Functional dizziness	6854	17.2
2. BPPV	5518	13.8
3. Central vestibular vertigo	5067	12.7
4. Vestibular migraine	5012	12.5
5. Menière's disease	4047	10.1
6. Unilateral vestibulopathy	3659	9.2
7. Bilateral vestibulopathy	2584	6.5
8. Vestibular paroxysmia	1234	3.1
9. Third mobile window syndrome	185	0.5
Unknown vertigo syndromes	1823	4.6
Other disorders	3926	9.8
	39,918	

Tab. 1.1 Absolute (n) und relative (%) Häufigkeiten der verschiedenen Schwindelsyndrome in der überregionalen Spezialambulanz des Deutschen Schwindelzentrums und der Neurologischen Klinik der LMU München (1998–2020)

Diagnose	n	%
1. Funktioneller Schwindel	6465	17,3
2. BPPV	5228	14,0
3. Zentraler vestibulärer Schwindel	4977	13,3
4. Vestibuläre Migräne	4631	12,4
5. Morbus Menière	3772	10,1
6. Unilaterale Vestibulopathie	3406	9,1
7. Bilaterale Vestibulopathie	2461	6,6
8. Vestibularisparoxysmie	1180	3,2
9. Syndrome des 3. mobilen Fensters	178	0,5
unklare Schwindelsyndrome	1723	4,6
andere[a]	3307	8,8
Gesamtzahl	37328	

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Age

- Mean age:
  - 51 years (range 25–67 years) (Brandt and Dieterich 1994),
  - 48.0 ± 15.3 years (Hüfner et al. 2008a)
  - 47.2 ± 14.7 years (Best et al. 2013).
- In total, 63 patients, 32 were female
- In children(8,9,12y/o), features similar to adults (Lehnen et al. 2015)

Patient History

- Recurrent short-lasting spontaneous attacks of spinning or non-spinning vertigo
- Typically lasts seconds to a minute
- 5-30 attacks per day (up to 70)
- Posture trigger (head turning)
- Induced by hyperventilation

Patient History

- (In some) associated with hyperacusis or hypacusis and/or tinnitus
  - Help to identify affected side

- Typewriter tinnitus, an analogous disease
  - Compression of cochlear nerve

Response to sodium channel blockers

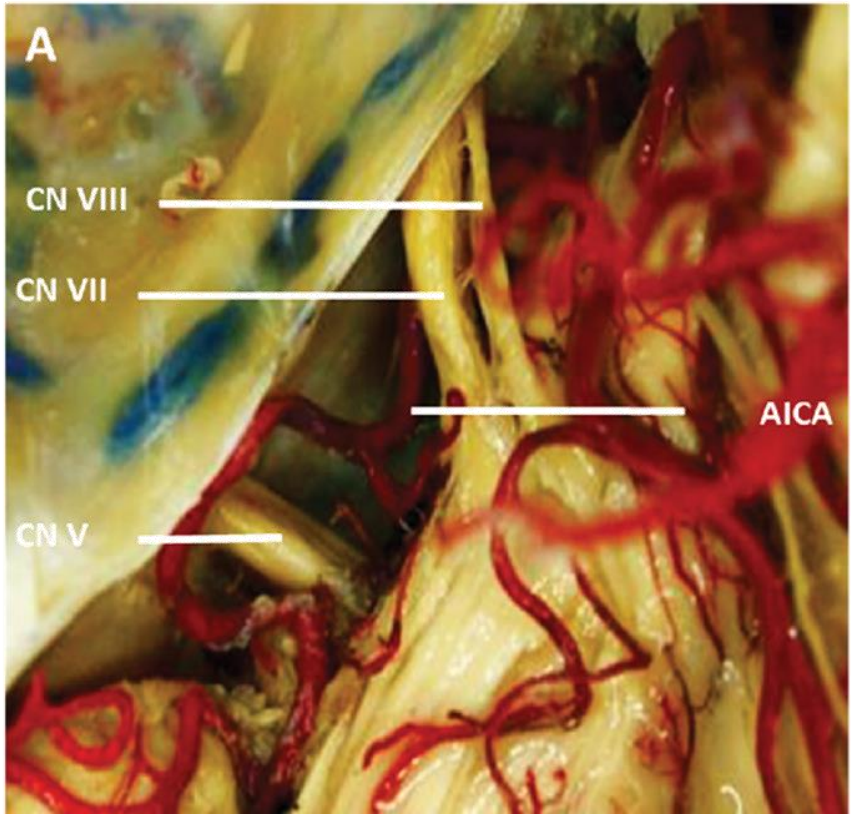
- Convincing response to sodium channel blockers, such *oxcarbazepine*, *carbamazepine*, or *lacosamide*, is important for the diagnosis
  - *Ex juvantibus*:指的是根據治療的效果來進行診斷或推論的原則

- If the response to treatment is not known yet: call probable VP
- If poor response to adequate treatment: diagnosis is doubtful

Patient history

- Facial hemispasm

  VIII and VII in the meatus acusticus internus lie close

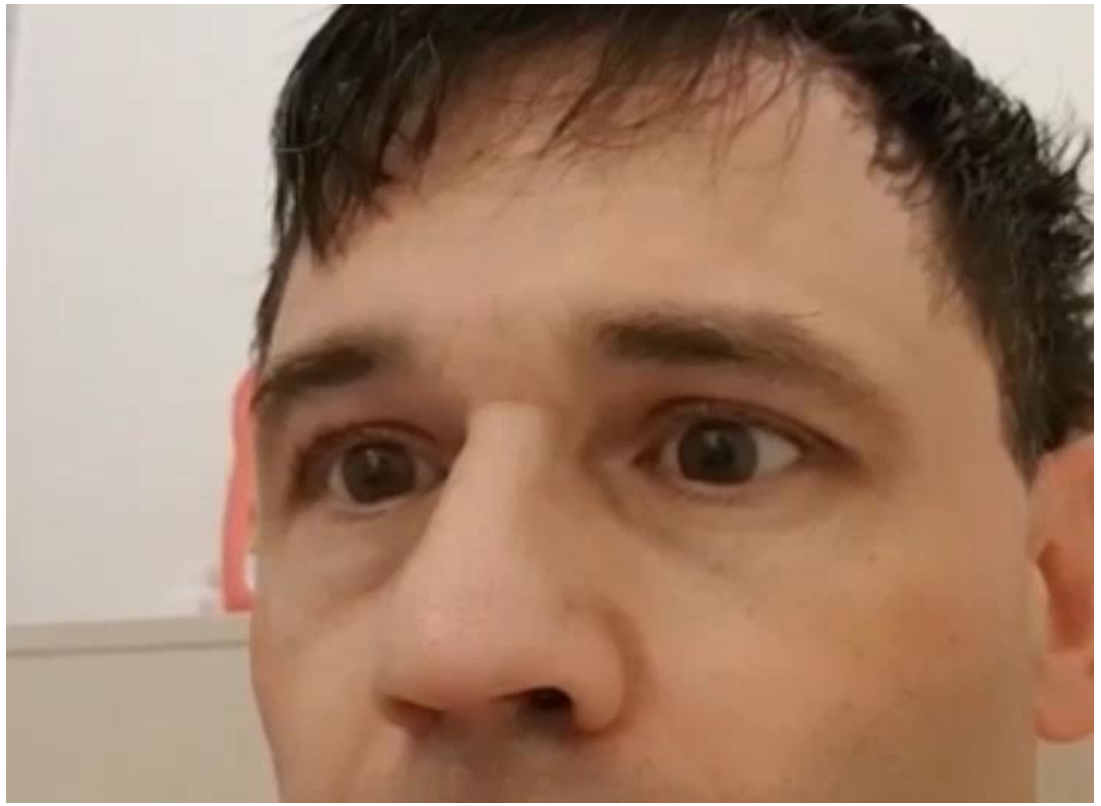


Bedside Examination
(between attacks)

- 20% mild unilateral peripheral vestibular hypofunction
  - HIT, head-shaking nystagmus
- Slight hearing impairment
- Hyperventilation induce vertigo and nystagmus (change direction)

Bedside Examination
(during attack)

- Spontaneous nystagmus



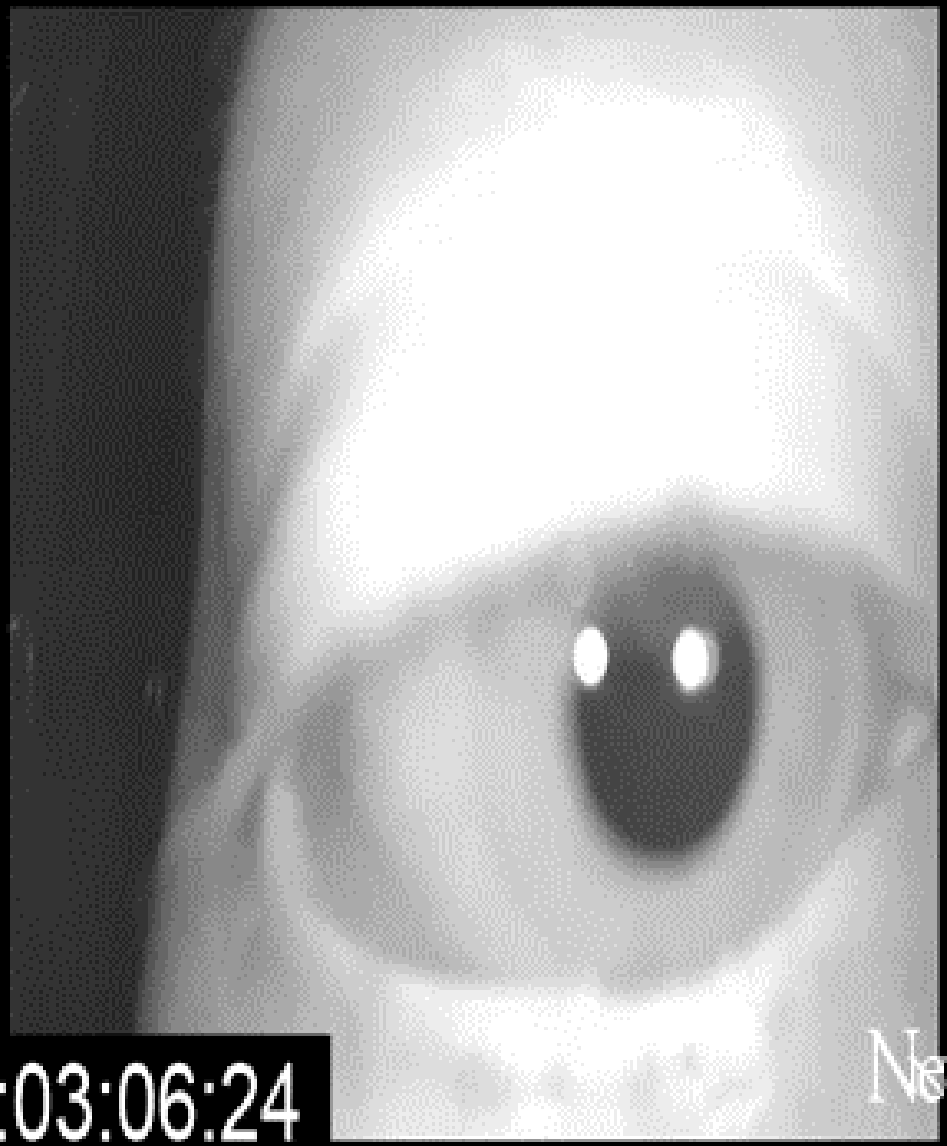
Laboratory Examinations
(between attacks)

About 45–50% of patients have evidence of mild to moderate peripheral vestibulo- cochlear deficits

- Hearing impairment less pronounced than Menière’s disease
- Excitation or a combination of excitation and inhibition in various tests (SVV, measurement of ocular torsion, caloric testing, or VEMP)
- A study with ABR showed that patients with VP had longer interpeak latency I-III and wave III latency compared to non-VP patients which also supports the pathophysiology

Laboratory Examinations
(during attacks)

- In a well-documented case with right-sided neurovascular cross-compression, initially a left-beating nystagmus was recorded by videooculography, which, after 47 s, beat to the right for 10 s
  - compatible with an inhibition and excitation of the vestibular nerve
- In another case, an excitatory nystagmus was recorded, which supports the assumed pathophysiology of VP and the response to treatment with sodium channel blockers.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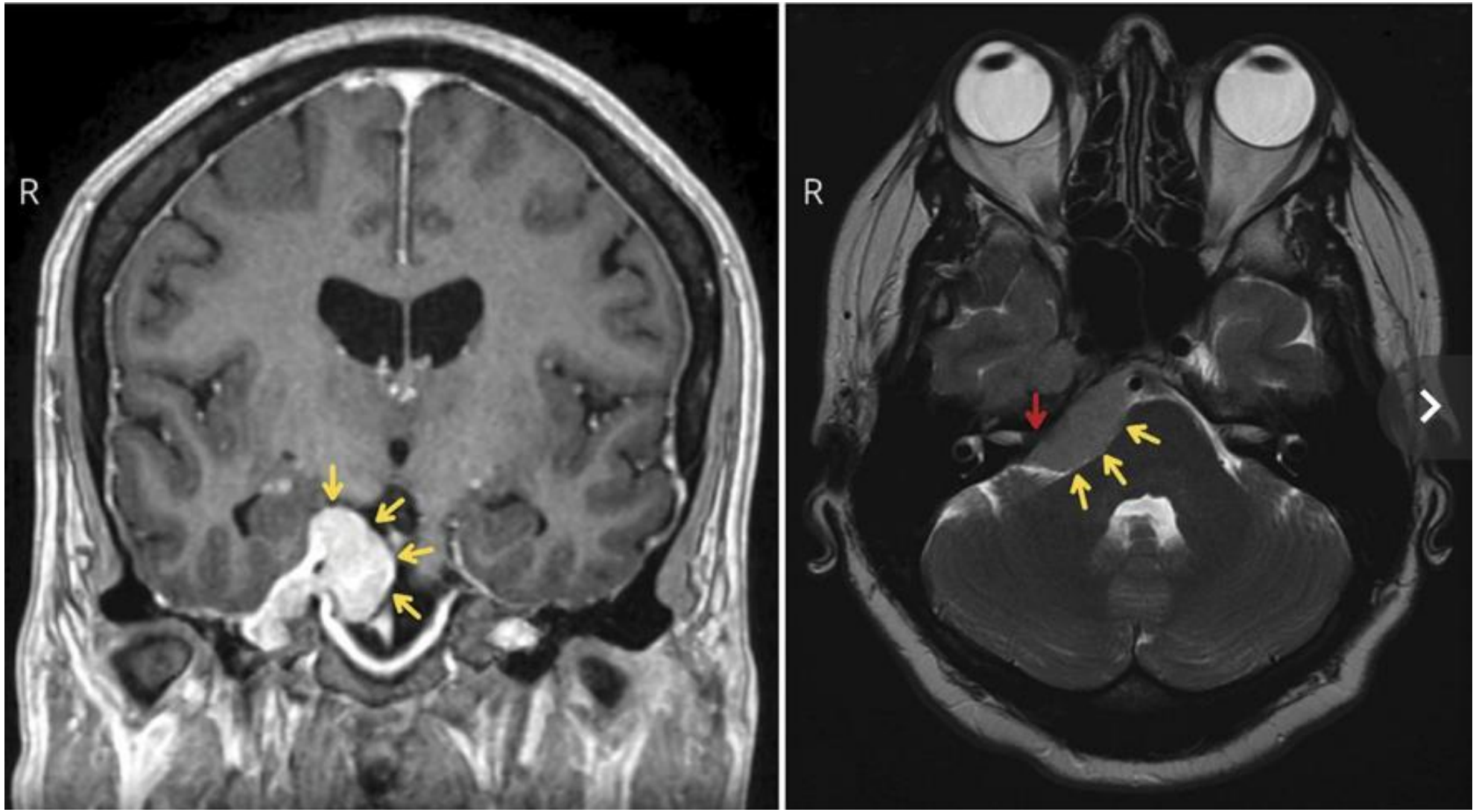
Neurology



right petroclival meningioma

Neurology. 2019;92:723-724.

Imaging
(MRI)

- In a study of 32 VP, neurovascular compression of the eighth cranial nerve was detected in 95% of the patients; bilateral neurovascular compression was found in 42% of the patients.

Imaging
(MRI)

- In another study of 20 patients with VP, neurovascular compression of the eighth cranial nerve was found in all patients, but also in 7 out of 20 control subjects (sensitivity: 100%, specificity: 65% for the diagnosis of VP by MRI).
- AICA in 15 cases (75%), the PICA in one (5%), a vein in two (10%), and VA (10%) in another two cases
- The distance between the brainstem and compressing vessels varied between 0.0 and 10.2 mm. This part of the nerve is proximal to the transition zone (up to 15 mm) and covered by oligodendrocytes.

Imaging
(MRI)

- In a third case series, a neurovascular compression of the eighth cranial nerve was also found in 45% of healthy subjects; however, this study included only 20 healthy controls and 9 patients with VP.
- Nerve angulation at the point of contact occurred in 5 of the cases, but in none of the controls. This study supports the concept of neurovascular compression in VP and suggests that nerve angulation may be a specific feature.

Imaging
(MRI)

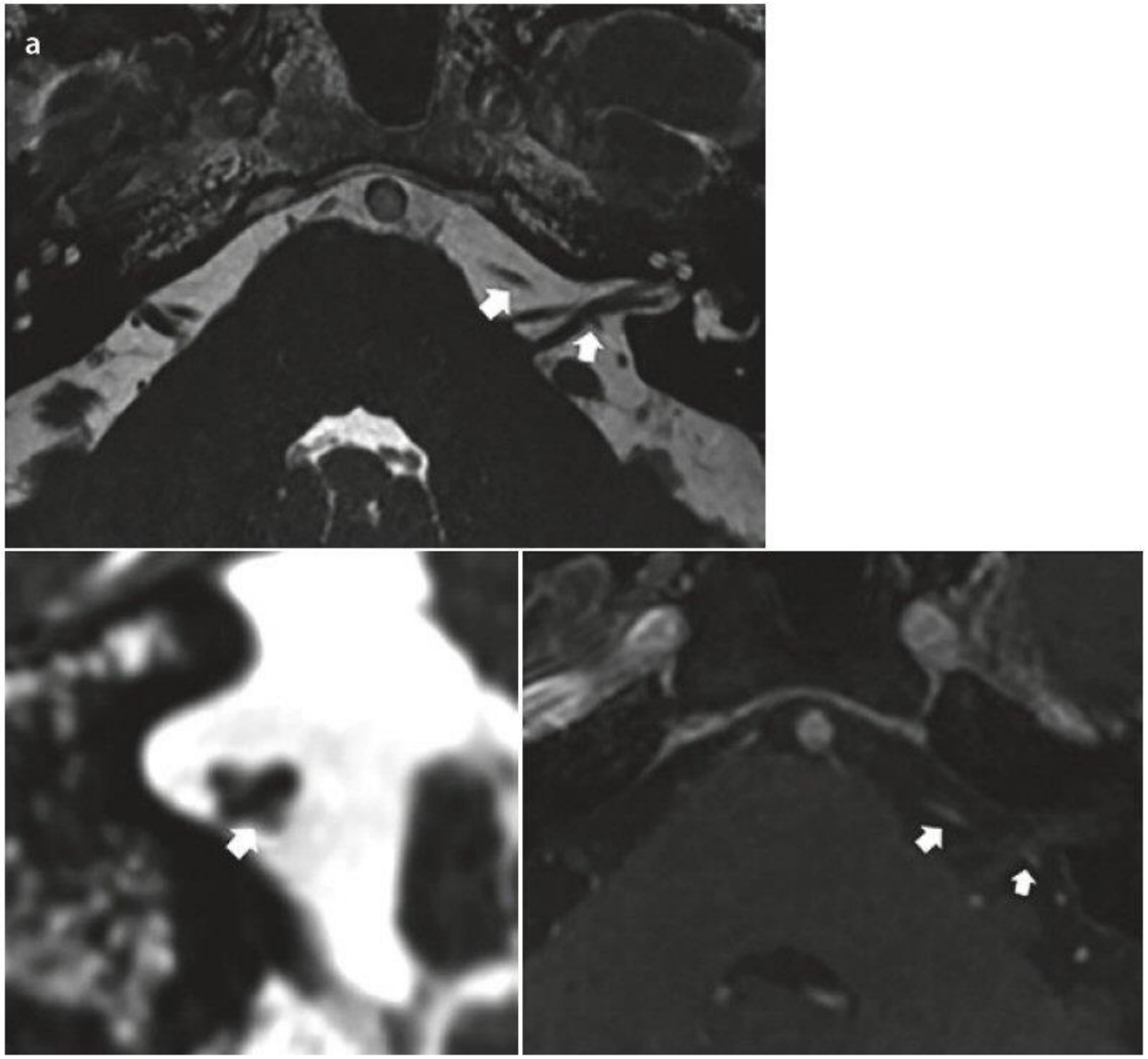
- A fourth imaging study with a total of 470 MRI scans, the authors also concluded that the vascular loop is a normal variant which may or may not give rise to audiovestibular symptoms or VP and that clinical assessment is still the most important tool in deriving a diagnosis of VP and MRI may be useful to rule out other central causes

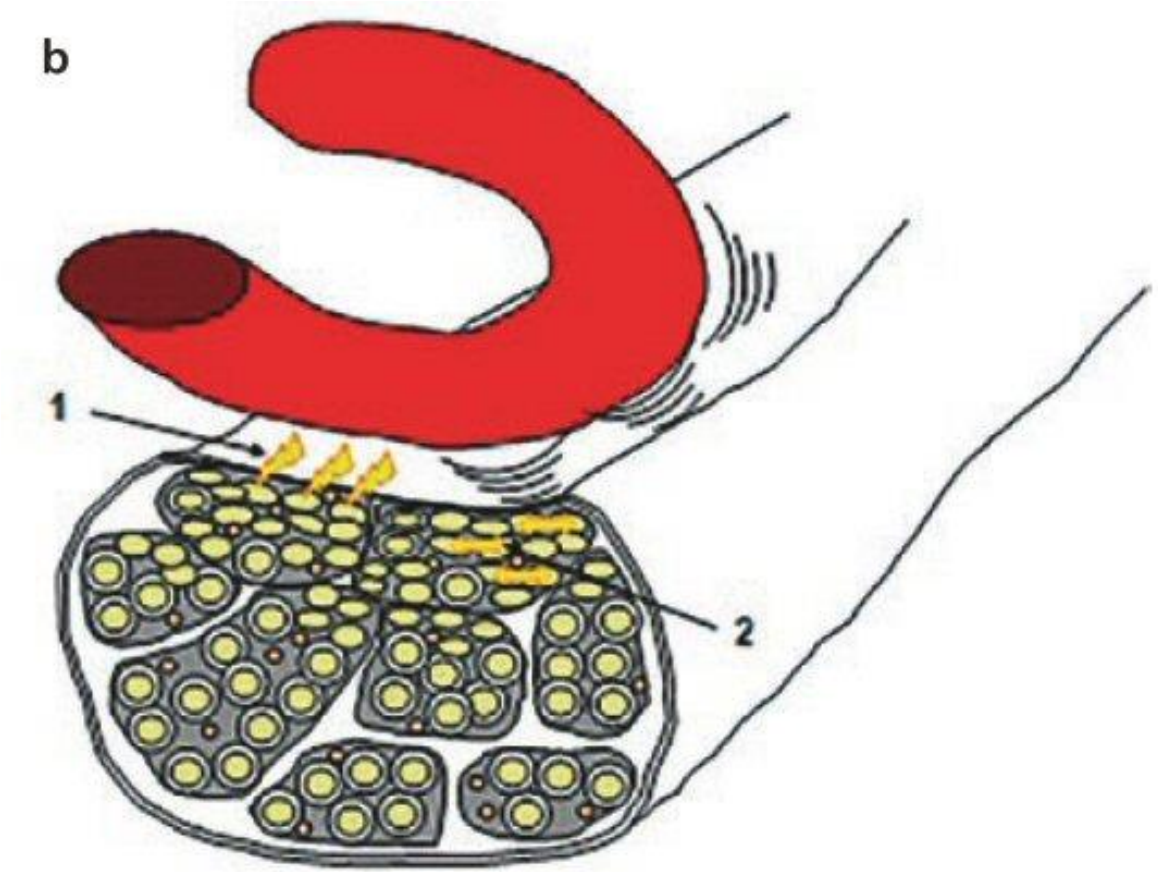
Imaging
(MRI)

- Finally in a study with 18 patients with VP and 18 age-matched healthy controls even the combination of various imaging techniques did not suffice to diagnose VP on a single-subject level due to individual variability and lack of diagnostic specificity

Imaging
(MRI)

- All in all, it is important to note that neurovascular contact is a common phenomenon in asymptomatic patients, has a low specificity and therefore the correlation with symptoms and signs remains essential.





intraaxonal ephaptic transmission

Imaging
(MRI)

- Since recurrent attacks of vertigo can also have other causes, a cranial MRI should be performed to exclude the presence of a tumor in the area of the cerebellopontine angle, arachnoid cysts, megalodolichobasilaris, brainstem plaques in multiple sclerosis, brainstem infarctions (both can lead to paroxysmal brainstem attacks), or other brainstem lesions such as brainstem melanocytoma

Differential Diagnosis

- BPPV
- Central positional vertigo
- Episodic ataxias
- Functional dizziness
- Menière's disease
- Orthostatic dizziness
- Panic attacks
- Paroxysmal brainstem attacks
- Syndrome of the third mobile windows
- Tumarkin's otolithic catastrophe
- Tumors in the brainstem (e.g., melanocytoma) or the cerebellopontine angle (e.g., vestibular schwannoma or meningioma)
- Vertebral artery compression/occlusion syndrome
- Vertebrobasilar TIAs
- Vestibular epilepsy
- Vestibular migraine

Diagnosis

- *Ex juvantibus*
- Recurrent, most often spontaneous, short attacks of vertigo, which are intraindividually uniform and respond to treatment with a sodium channel blocker.
- Can be helpful:
  - Patient videos his eye movement during attack
  - NE and lab exam
- MRI: high sensitivity, low specificity (45% healthy subjects)

Course of Disease

Two studies

- 3.4y f/u (n=61) Hanskamp 2022
  - 72% still
  - 71% limited quality of life
- 4.8y f /u (n=146) Steinmetz 2022
  - 75% attack free
  - >half no medication

Recommend regular f/u

Pathophysiology and Etiology

- Neurovascular cross-compression in patients with hyperactive dysfunction symptoms of the eighth cranial nerve (1975).
- In analogy to the brief recurrent symptoms in trigeminal neuralgia, hemifacial spasm, glossopharyngeal neuralgia, or myokymia of the superior oblique muscle.

Pathophysiology and Etiology

- Compression cause partial demyelination of axons, lead to **ephaptic discharges**
- Triggered by the **pulsations of the artery** and by **sensory input** during head movements

- Ephaptic discharges: pathological paroxysmal inter-axonal transmissions between neighboring axons

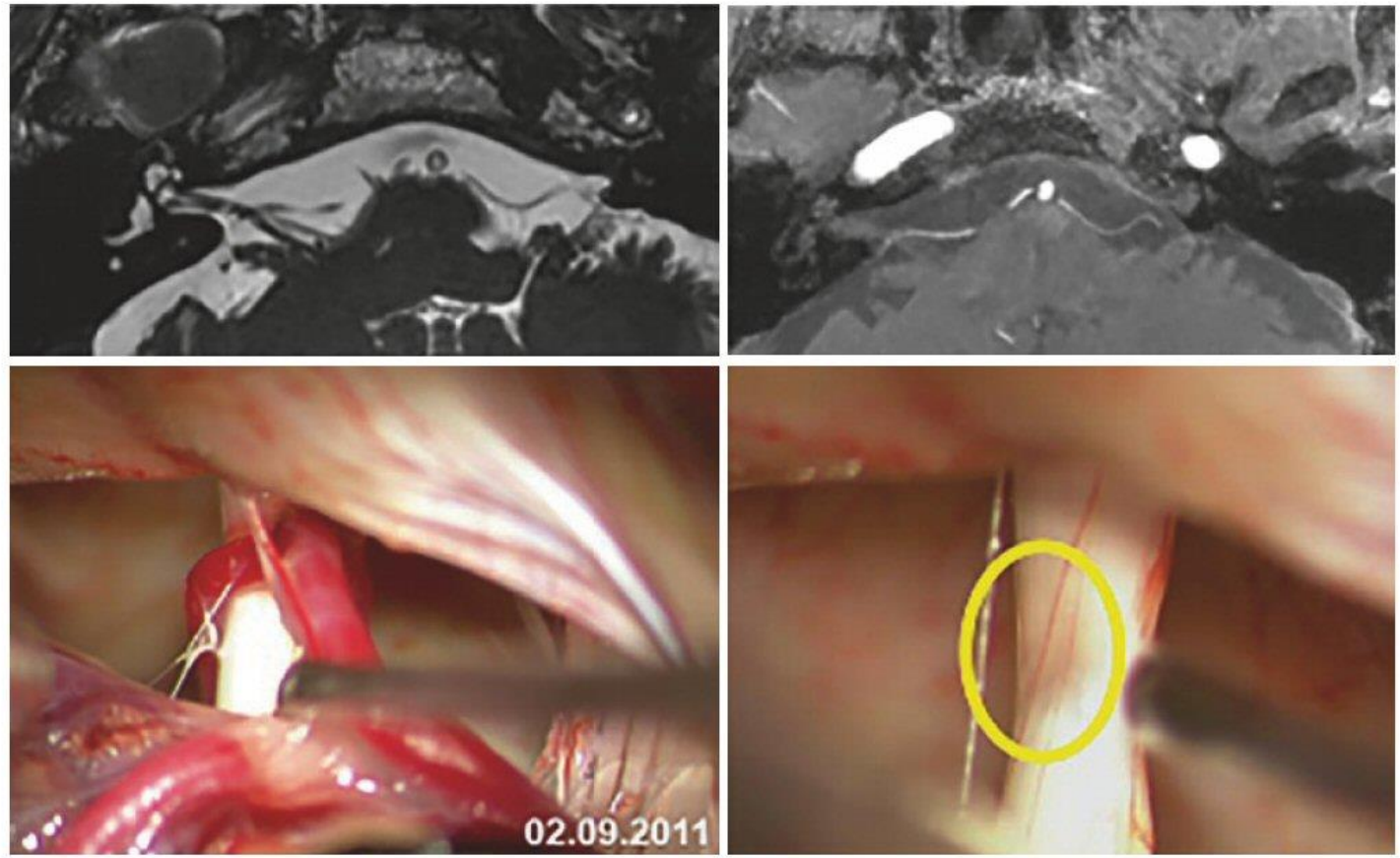


Fig. 11.5 Patient with right-sided vestibular paroxysmia with attacks of vertigo and buzzing in the right ear. Cranial MRI (left upper corner: constructive interference in steady-state sequence, right upper corner: time-of-flight) shows contact between the right eighth cranial nerve and the anterior inferior cerebellar artery. Intraoperative micrographs demonstrate vascular contact (left lower corner) and considerable compression of the eighth nerve after removal of the arteries (right lower corner, circle). The patient has been free of symptoms since surgery in 2011. (Adapted from Strupp et al. (2013))

Therapy

- Pharmacotherapy

  Sodium channel blockers reduce the excitability of the peripheral nerves

  - CBZ: Brandt 1994, Kanashiro 2005
  - OXC: Bayer 2018; 0.62 vs 0.41/d (reduced to 0.67)
  - CBZ/OXC : Hüfner 2008; frequency reduced to 10%
  - LCM: Strupp 2019; 13->3/m
- Surgical Treatment
  - Møller MB. (n=16, 41, 207; 1986, 1990, 1993; success rate 73-80 %)

CBZ: carbamazepine
OXC: oxcarbazepine
LCM: lacosamide

Pharmacotherapy

Sodium channel blockers

- Reduce the excitability of the peripheral nerves
- At least 4 weeks

- Carbamazepine (200-800 mg/d)
- Oxcarbazepine (300-900 mg/d)

- Lacosamide (100-400 mg/d)
- Theoretically effective (no clinical study)
  - Phenytoin (300-900 mg/d)
  - Lamotrigine (25 ->100-200 mg/d)

Surgical Treatment

- Microvascular decompression
- Prerequisites:
  1. the diagnosis is certain with a convincing response to pharmacotherapy
  2. various medications are not tolerated
  3. the affected side is clearly identified
- Risk of a brainstem and/or cerebellar infarction
  - Vasospasm risk (1-3%)
  - AICA(75%)->labyrinthitis artery

Thank you!