Bilateral Vestibulopathy

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Epidemiology

- the most frequent cause of postural imbalance

  in the elderly requiring precise diagnosis.
- The relative frequency is 4–7%

(Rinne et al. 1998; Zingleret al. 2007; Kim et al. 2011)

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	

Diagnosis

combination with a clinical and laboratory examination

- patient history

  leading symptoms: chronic postural imbalance ,unsteadiness (walking) ,

  without symptoms under static conditions (sitting or lying down)

- video-HIT, caloric testing and/or rarely rotatory chair : bilateral deficit of the vestibulo- ocular reflex

Box 7.1 Diagnostic Criteria for Bilateral and *Probable* Bilateral Vestibulopathy According to the Classification Committee of the Bárány Society (Strupp et al. 2017)

Bilateral vestibulopathy

1. Chronic vestibular syndrome with the following symptoms:
   (a) Unsteadiness when walking or standing plus at least one of 2 or 3
   (b) Movement-induced blurred vision or oscillopsia during walking or quick head/body movements and/or
   (c) Worsening of unsteadiness in darkness and/or on uneven ground
2. No symptoms while sitting or lying down under static conditions
3. Bilaterally reduced or absent angular VOR function documented by:
   - Bilaterally pathological horizontal angular VOR gain <0.6, measured by the video-HIT or scleral-coil technique and/or
   - Reduced caloric response (sum of bithermal max. peak SPV on each side <6°/s) and/or
   - Reduced horizontal angular VOR gain≤0.1 upon sinusoidal stimulation on a rotatory chair (0.1 Hz, Vmax=50°/sec) and a phase lead≥15 degrees (time constant≤6sec)
4. Not better accounted for by another disease

Probable bilateral vestibulopathy

1. Chronic vestibular syndrome with the following symptoms:
   (a) Unsteadiness when walking or standing plus at least one of 2 or 3
   (b) Movement-induced blurred vision or oscillopsia during walking or quick head/body movements and/or
   (c) Worsening of unsteadiness in darkness and/or on uneven ground
2. No symptoms while sitting or lying down under static conditions
3. Bilaterally pathological horizontal bedside head impulse test
4. Not better accounted for by another disease

The diagnosis of "bilateral vestibulopathy" requires quantitative testing of vestibular function by means of video-HIT and/or caloric testing, since the bedside head impulse test has a low sensitivity and specificity (Yip et al. 2016). Only the diagnosis of "*probable* BVP" can be made using bedside testing.

Patient History

- postural imbalance, dizziness, and impaired gait (worsening in darkness and/or on uneven ground,no symptoms under static conditions.
- blurred vision(40–70%) (walking or during fast head movements)

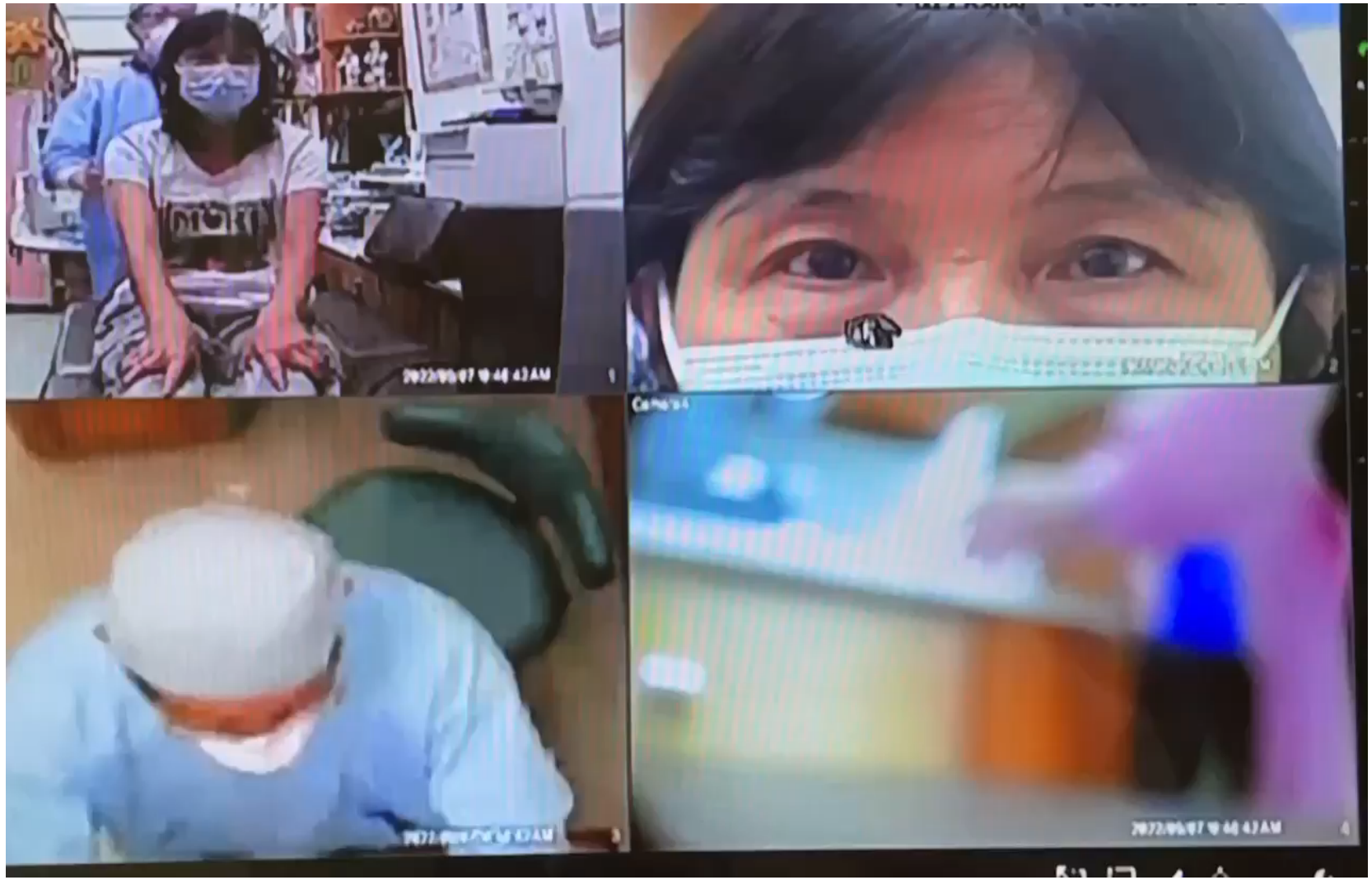
  pulse synchronous or chewing oscillopsia (In severe cases)
- vertigo (33–67%) recurrent spinning or non-spinning lasting for minutes to days. (sequential BVP)
- impairment of spatial navigation and memory, as well as impaired spatial learning

oscillopsia

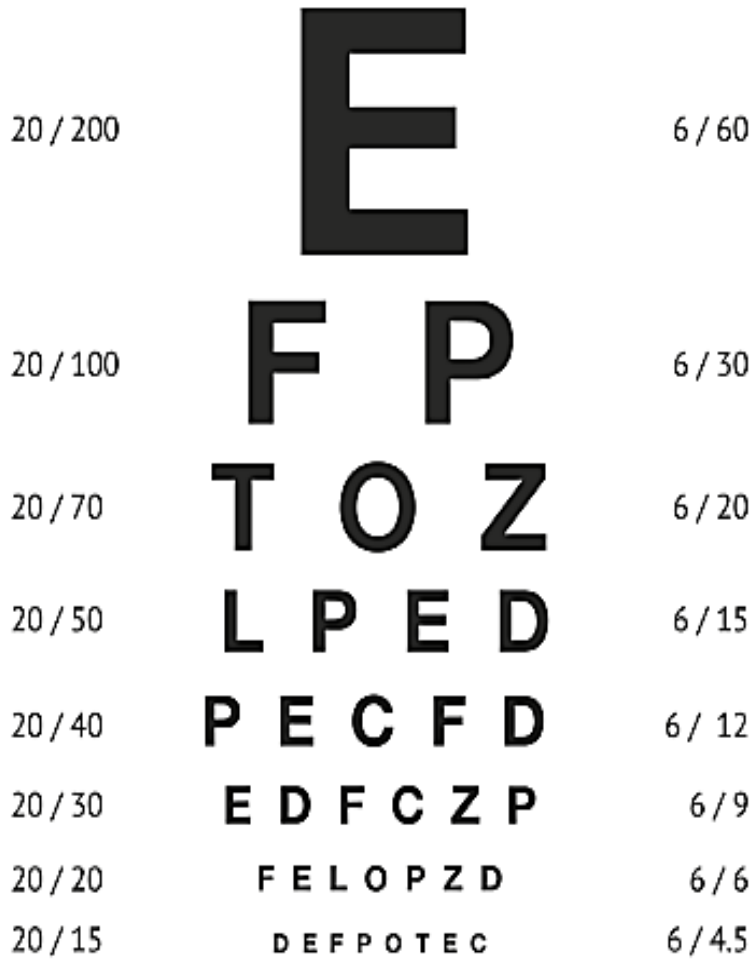
Bedside Testing

- “Bedside head impulse test” (bHIT) (Halmagyi 1988) : bilateral refixation saccades (65%)
- Dynamic visual acuity (DVA): decreases by > 0.2 , (Vital et al.2010).
- Romberg test : increased sway (eye closed) : a deficit of vestibulo-spinal reflexs
  standing on foam is evidently more sensitive (Sprenger et al. 2017).
  but the Romberg test is not specific
- Asymmetries of vestibular function (walking with the eyes closed)

Bedside head impulse test” (bHIT)



Dynamic visual acuity (DVA): decreases by > 0.2



Snellen Line	Acuity	LogMar
8	20/20	0
7	20/25	0.097
6	20/30	0.176
5	20/40	0.301
4	20/50	0.398
3	20/70	0.544
2	20/100	0.699
1	20/200	1

Laboratory Testing

- The video-HIT (Halmagyi et al. 2017) in the high- frequency range
- Caloric irrigation with testing of the VOR in the low-frequency range
- The rotatory chair in the middle-frequency range (0.1 Hz) (rarely necessary).

The video-HIT

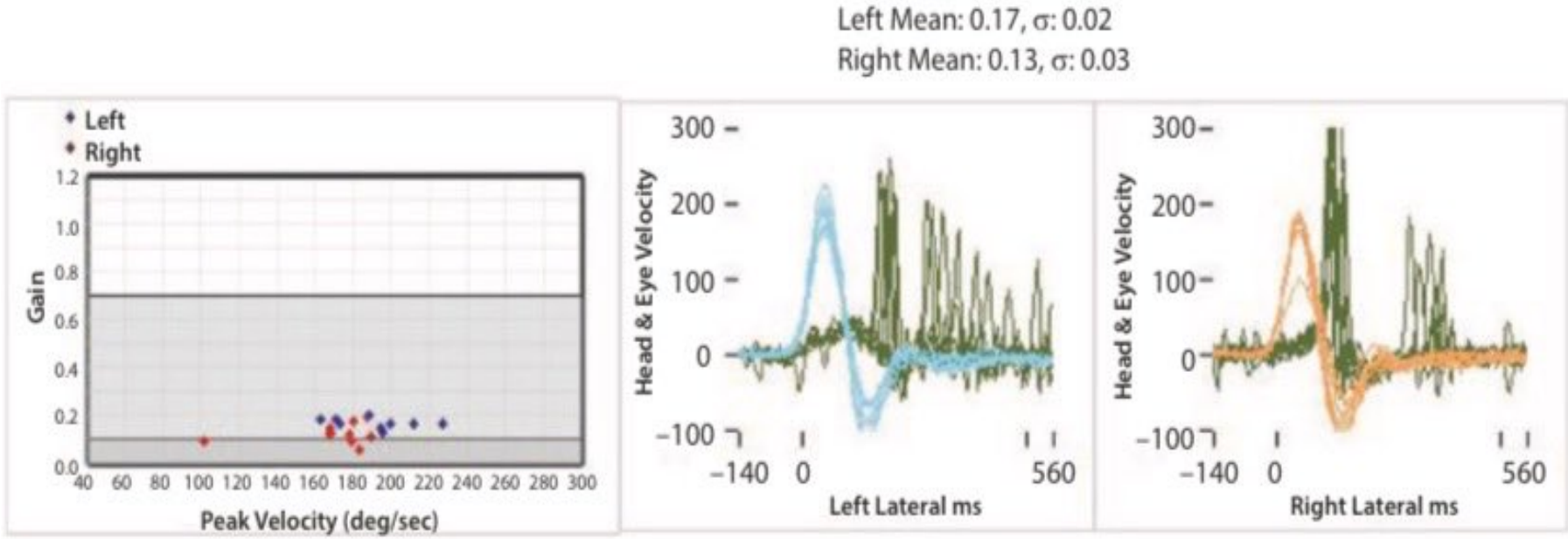
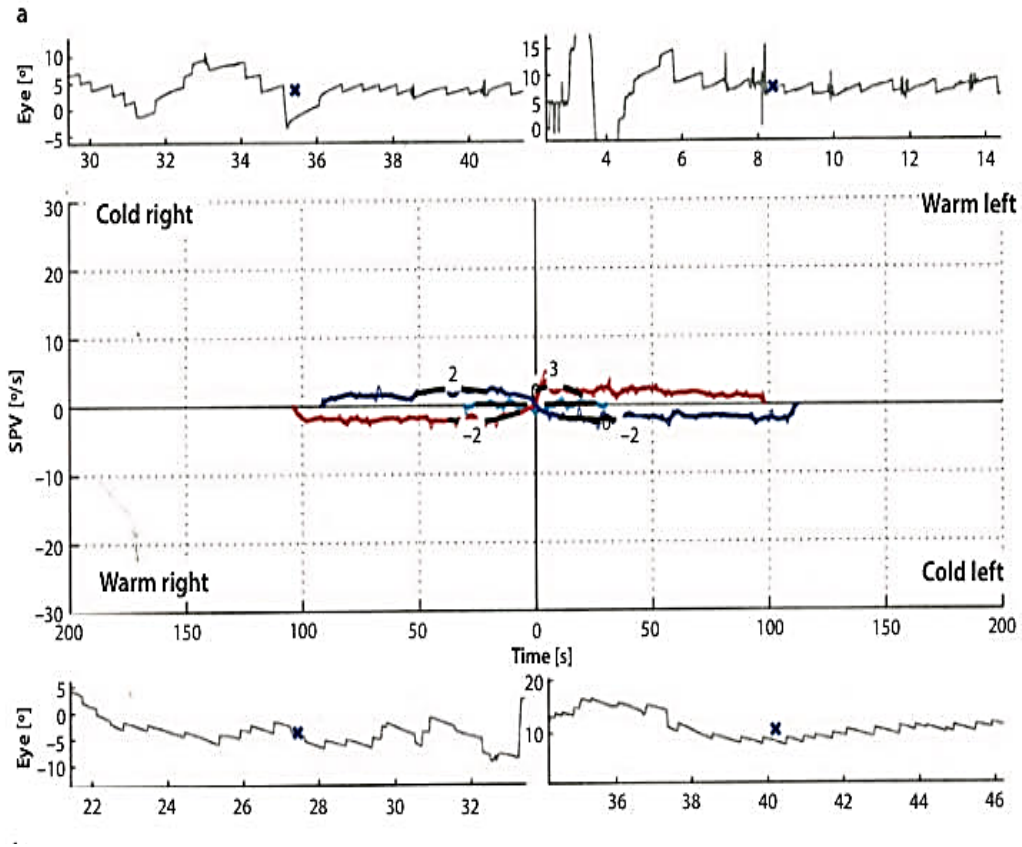


Fig. 7.5 Video head impulse test. Bilaterally significantly reduced VOR gain (left: 0.17; right: 0.13; normal 0.8) indicating bilateral vestibulopathy with a VOR deficit in the high-frequency range

Caloric test



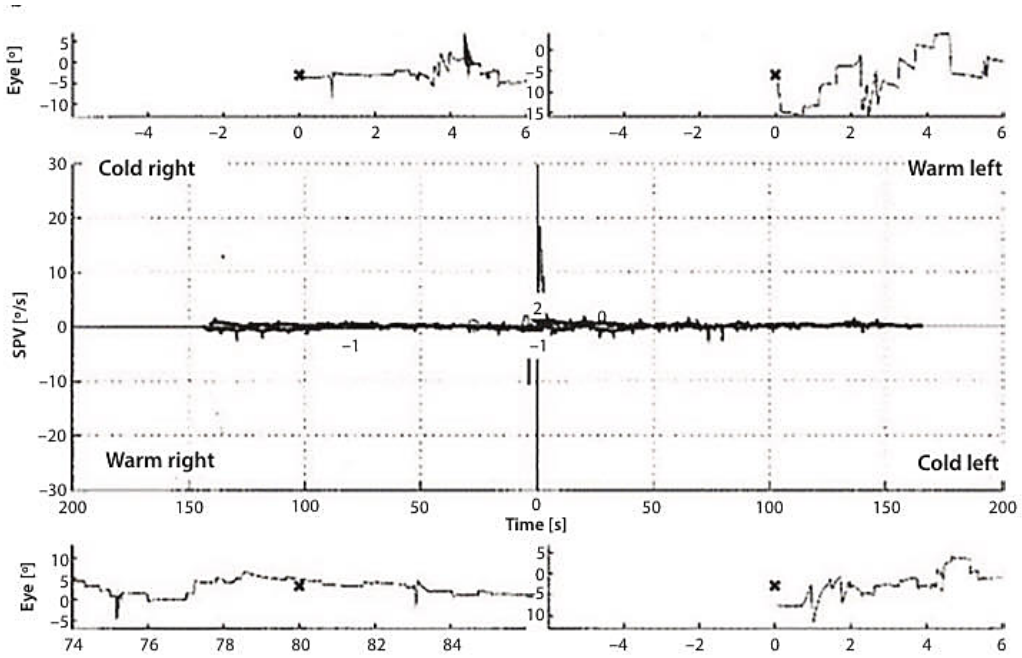


Fig. 7.6 a, b Caloric testing with warm and cold water in two patients with bilateral vestibulopathy. **a** Bilaterally reduced caloric response (sum of the maximum peak slow phase velocity on each side <6°/s). **b** Bilaterally no caloric response. Both patients have a VOR deficit in the low-frequency range

Diagnosis of BVP

- VOR gain of <0.6 (150–300°/s)
- Bilaterally reduced or absent caloric response max. peak SPV < 6°/s
- Reduced horizontal angular VOR gain≤0.1
- the function of the horizontal semicircular canal is always reduced

Complementary Laboratory

- Cervical and Ocular Vestibular-Evoked Myogenic Potentials (c/oVEMP)

Imaging

- contrast MRI CCT

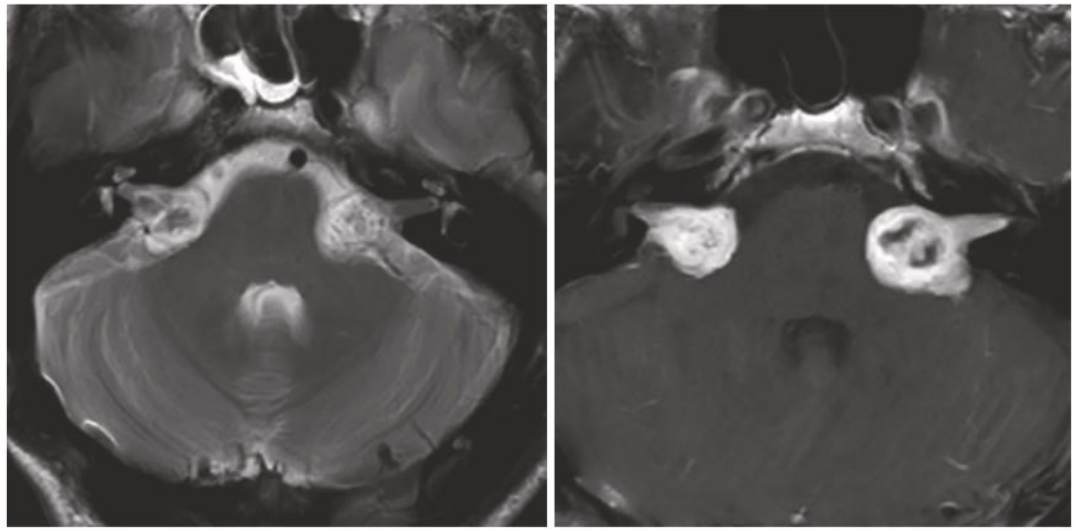
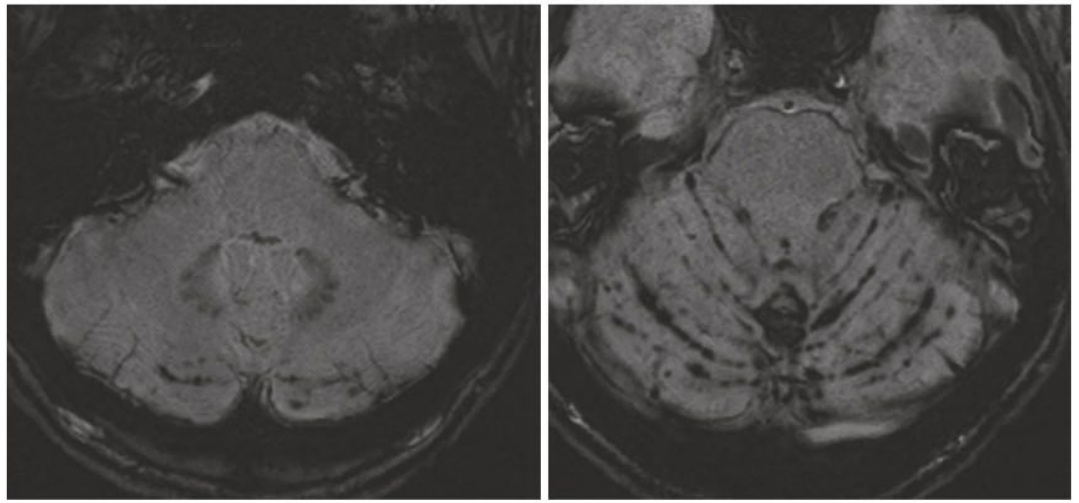


Fig. 7.7 Bilateral vestibular schwannoma with significant contrast enhancement on the right



Summary

- Testing and Clinically initially using video-HIT. If the VOR gain in the video-HIT is <0.6, combination with an appropriate patient history – be made.
- If the VOR gain bilaterally is >0.6, caloric testing is required.
- all patients with BVP, at least one contrast-enhanced MRI (bilateral shcwannoma)

Pathophysiology

the deficits of the vestibuloocular and vestibulo-spinal reflexes

- unsteadiness of posture and gait
- oscillopsia and blurred vision 40–70%
- deficits of spatial memory, navigation and other cognitive functions and morph ological changes : hippocampus , parahippocampus

Etiology

Table 7.1 Causes and underlying disease leading to bilateral vestibulopathy

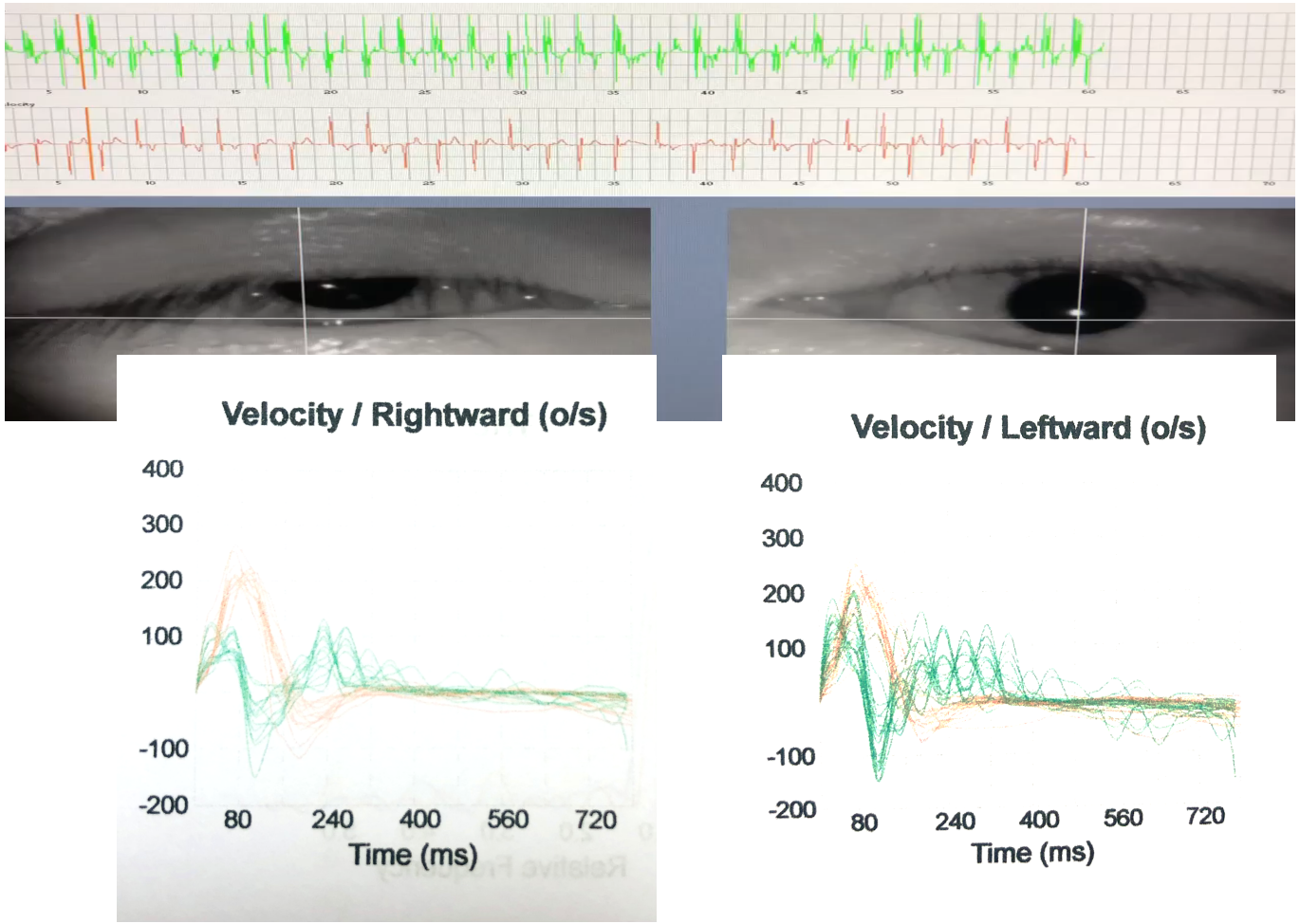
Relatively frequent	Underlying cause
Idiopathic (>30–50%)	
Ototoxicity	Gentamicin and other antibiotics
	Anticancer chemotherapy
	Loop diuretics Amiodarone
	Aspirin
Bilateral Menière's disease	Endolymphatic hydrops
Meningitis or labyrinthitis	e.g., Streptococci, Neisseria meningitis, Mycobacterium tuberculosis
Combination with cerebellar disorders	Cerebellar ataxia and/or cerebellar ocular motor disorders, in particular downbeat nystagmus (frequent) CANVAS (cerebellar ataxia, neuropathy, and vestibular areflexia syndrome) Spinocerebellar ataxias Multiple system atrophy

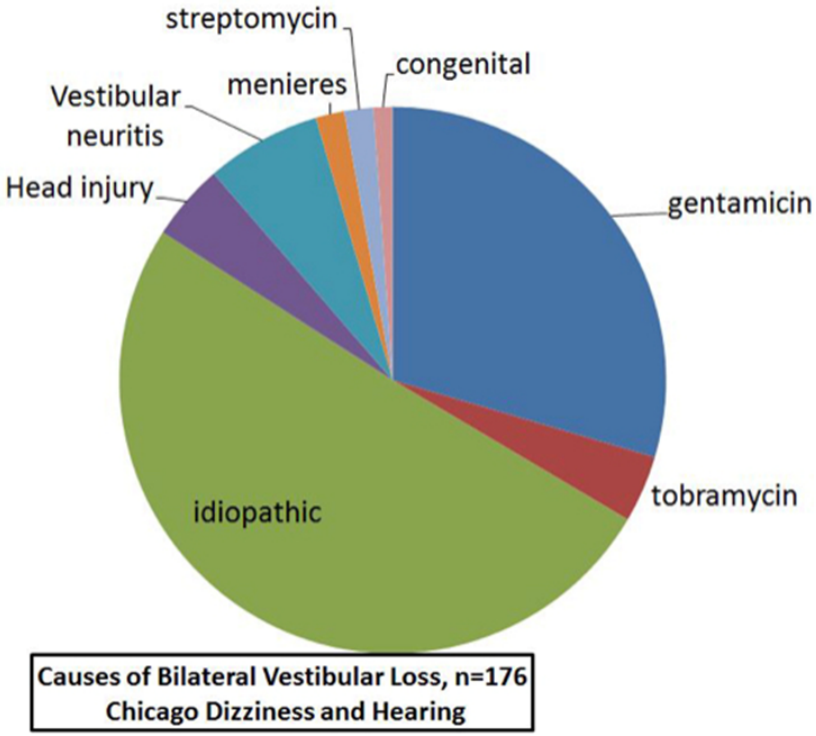
Etiology

Table 7.1 (continued)

Relatively frequent	Underlying cause
Tumors	Neurofibromatosis type 2 (Bilateral vestibular schwannomas)
	Lymphomas
	Meningeosis carcinomatosa
	Infiltration of the skull base
Autoimmune disorders	Cogan's syndrome
	Neurosarcoidosis
	Behçet's disease
	Cerebral vasculitis
	Systemic lupus erythematosus
	Polychondritis
	Rheumatoid arthritis
	Polyarteritis nodosa
	Wegener's granulomatosis/ANCA-associated small vessel associated vasculitis and others
	Giant cell arteritis/large vascular vasculitis
	Antiphospholipid syndrome
	Hereditary sensory and autonomic neuropathy
Vitamin deficiencies	B1, B6, B12 deficiency
Bilateral sequential vestibular neuritis	Herpes simplex virus type I, varizella zoster
Other causes	
Congenital malformation	Usher's syndrome and other rare hereditary conditions
Other causes	Bilateral labyrinthine concussion Bilateral petrous bone fracture
	Paget's disease
	Macroglobulinemia
	Vertebrobasilar dolichoectasia Superficial siderosis

Gentamicin toxicity

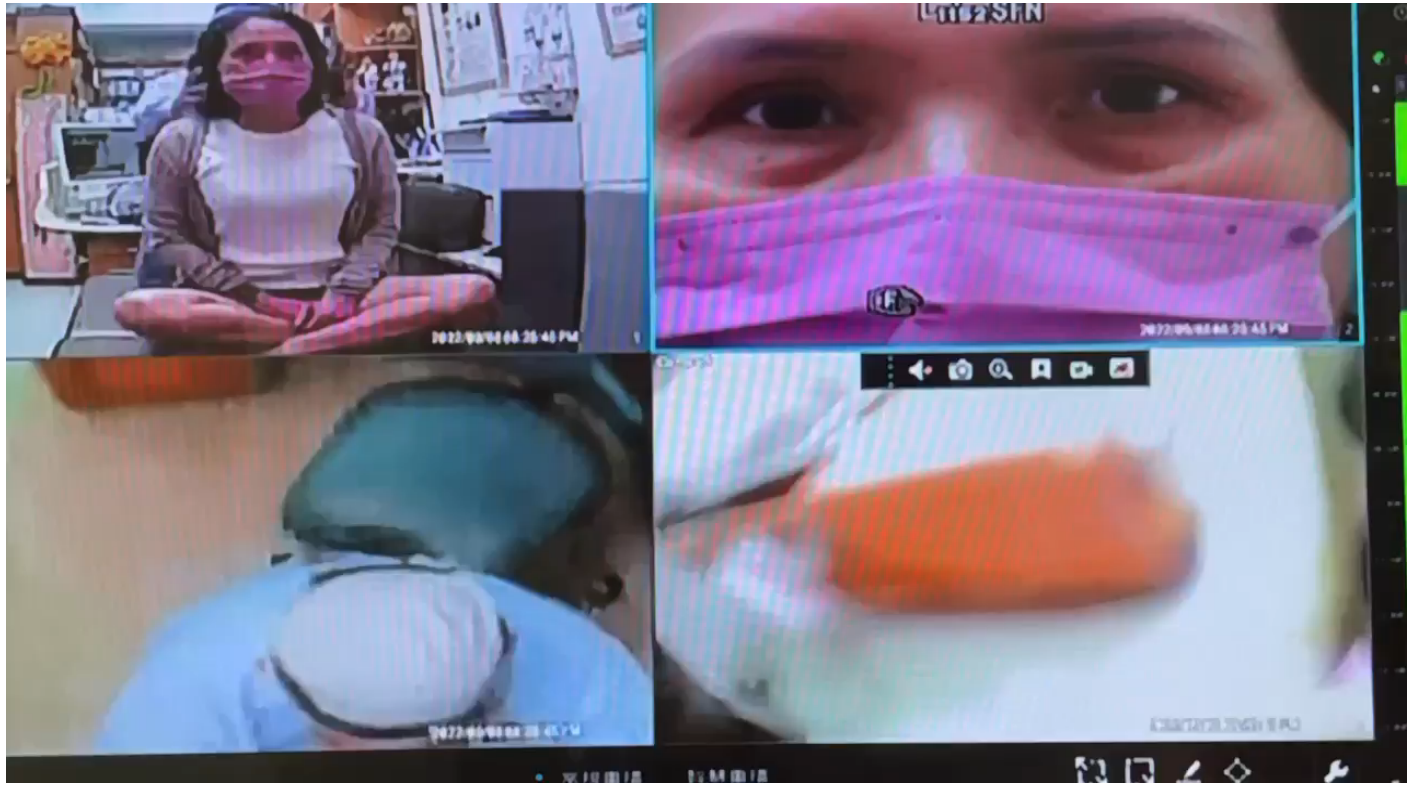




Causes of Bilateral Vestibulopathy
Timothy C.Hina MD March 10 2021

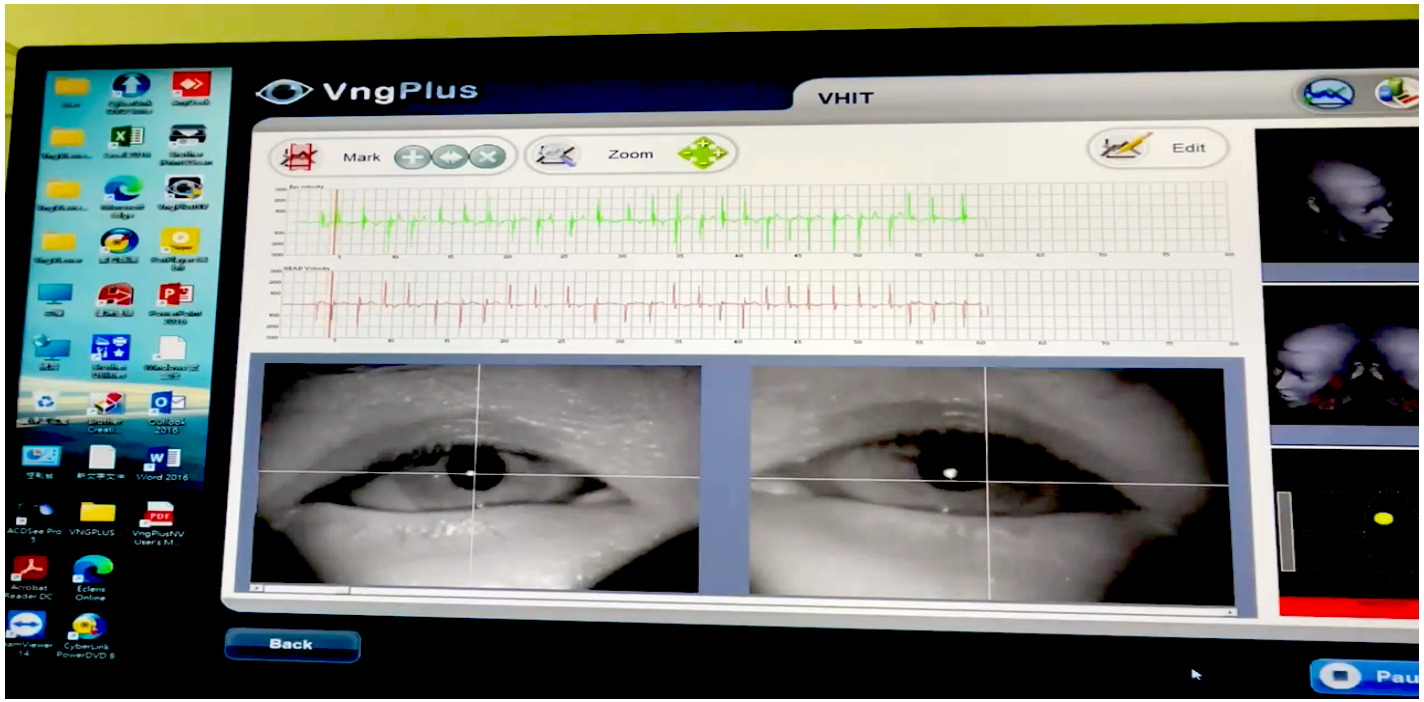
Lung cancer，

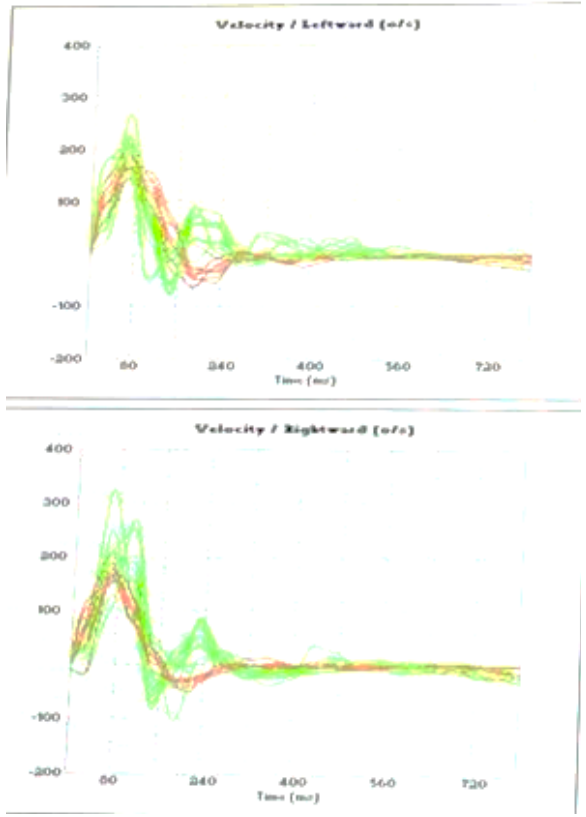
adenocarcinoma， metasatic brain tumor



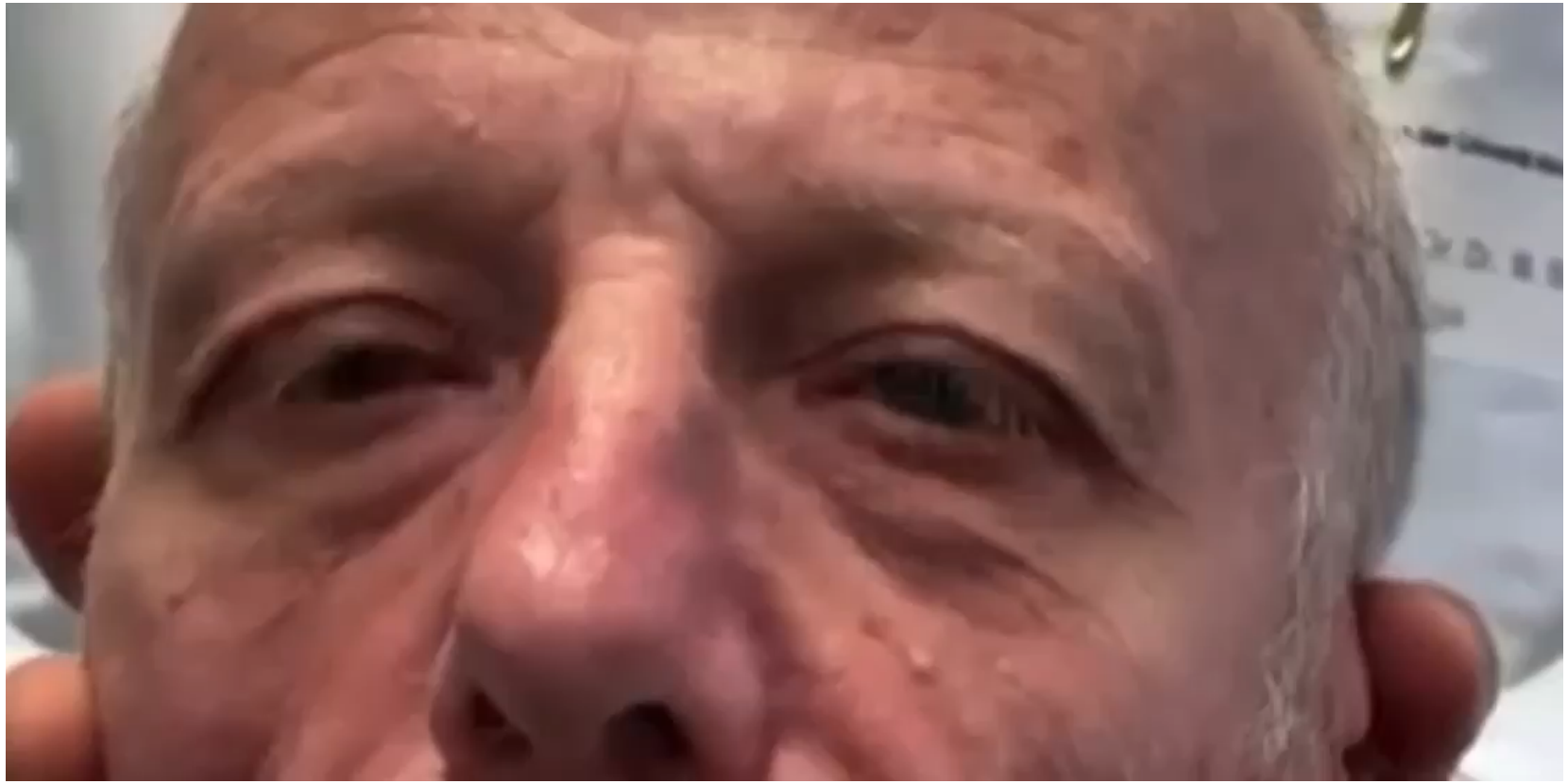
Lung cancer，

adenocarcinoma， metasatic brain tumor



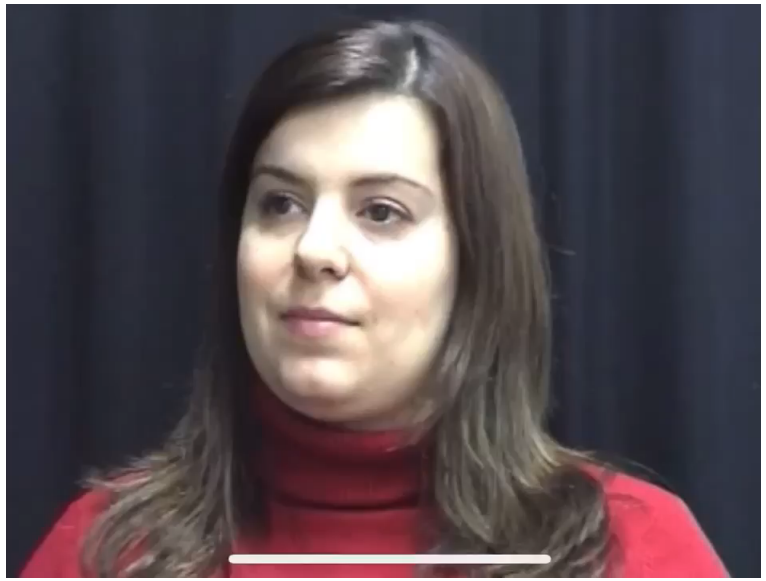


“Cerebellar Ataxia with Neuropathy and Vestibular Areflexia Syndrome” (CANVAS)



Impaired walking in cerebellar ataxia, neuropathy and vestibular areflexia (CANVAS)

Cogan- Sydrome



patient history

- acute subacute spinning /non-spinning vertigo,
- reduced hearing, tinnitus,
- eye pain with red eyes.

if not immediately treated , BVP and bilateral hearing loss

MRI typically shows bleedings and contrast enhancement in the labyrinth and/or the cochlea.

Differential Diagnosis

- Cerebellar ataxias without BVP.
- Cerebellar dizziness
- Downbeat nystagmus syndrome
- Extrapyramidal diseases, such as Parkin-son’s disease, multiple system atrophy, or tauopathies,
- Functional dizziness.
- Intoxication, e.g., benzodiazepine or chronic alcohol abuse.
- Normal pressure hydrocephalus or subcortical vascular encephalopathy
- Orthostatic dizziness
- Persisting unilateral peripheral vestibular deficit.
- Reduced visual acuity, also due to nystagmus
- Syndrome of the third mobile windows

Differential Diagnosis

- Combined Central and Peripheral Vestibular Deficits
- acute lesions of the cerebellar flocculus
- Wernicke encephalopathy,
- Gaucher’s disease
- Infarctions in the AICA territory

Course of the Disease

- 80% of the patients, there is no significant improvement of vestibular function

Treatment

- Therapeutic Principles, Aims,and Pragmatic Therapy

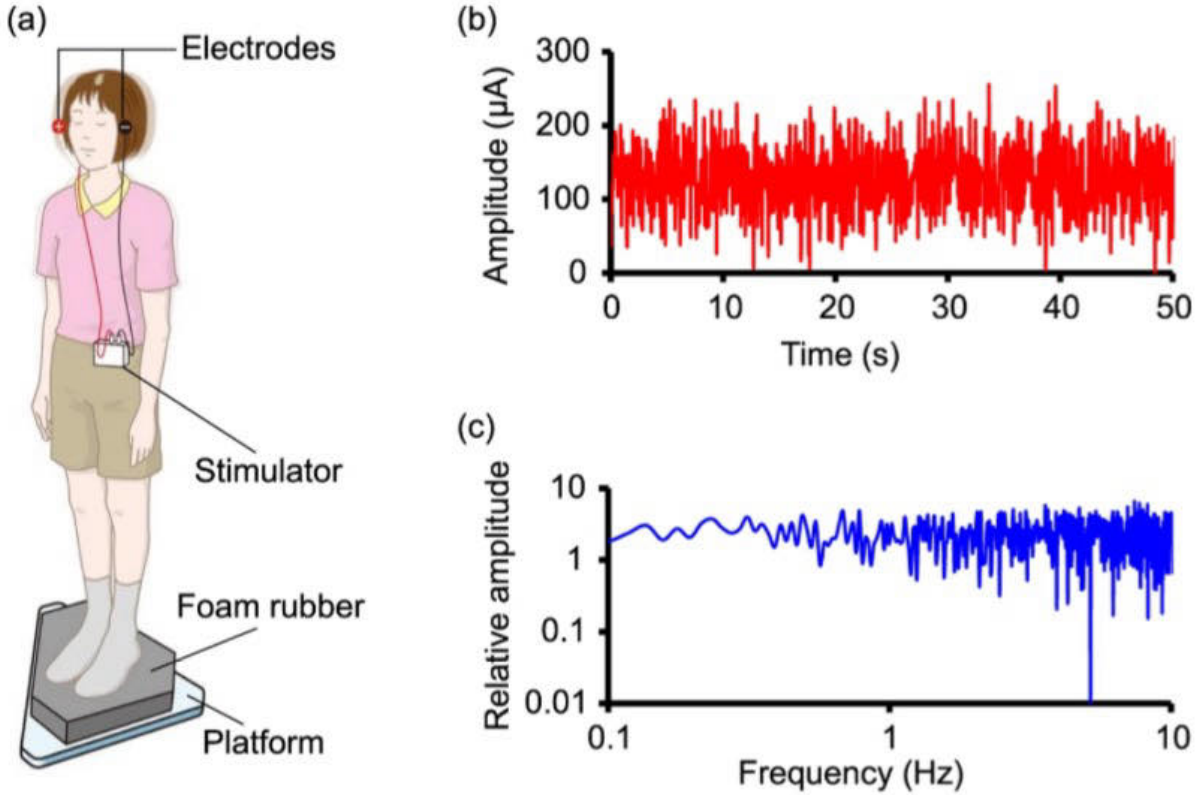
1. Explanation : S/S , benigh course, vestibular exercise (6-12 weeks)
2. Primary prophylaxis : ototoxic labyrinthine drugs (antibiotics,diuretics)
3. Therapy of the underlying disease : postmingitis , autoimmune inner ears

   (predisolone 80 mg/kg, tapered 3-4 weeks)
4. Physiotherapy:

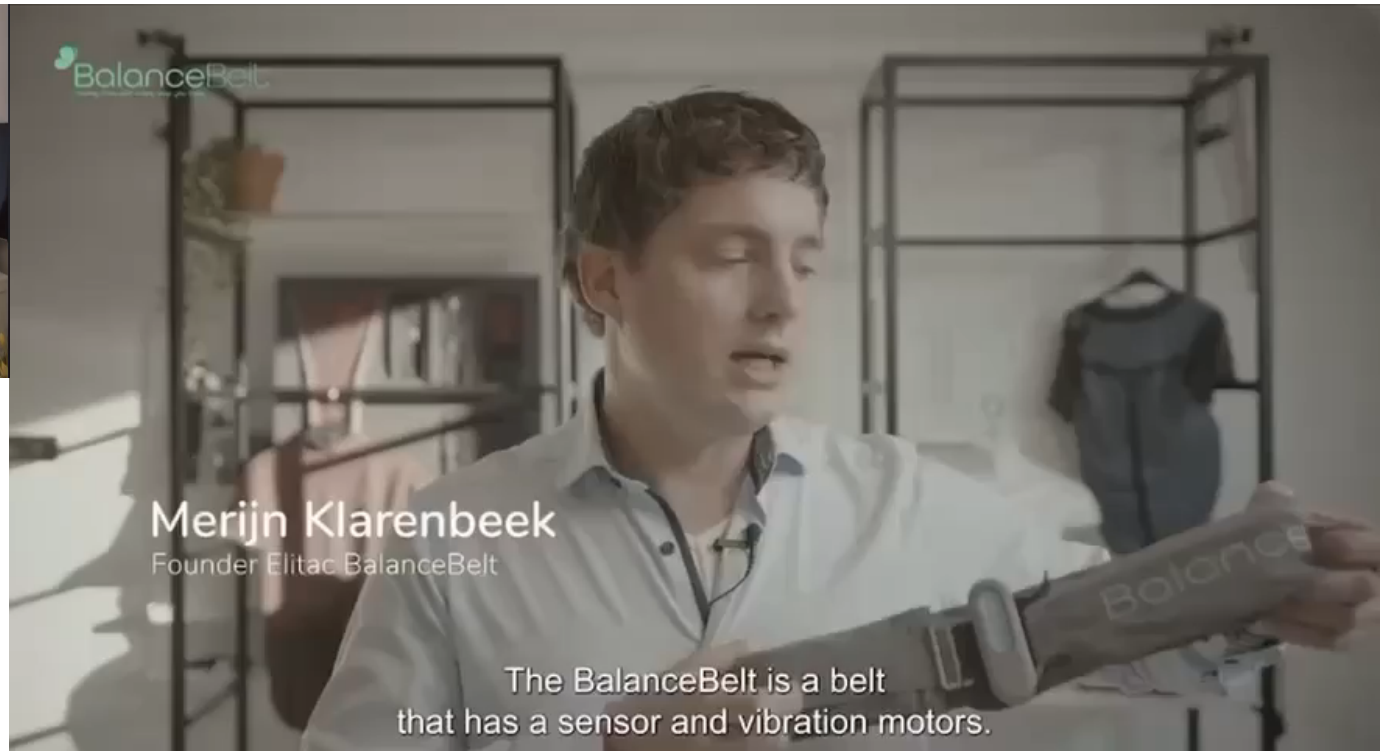
New therapeutic measures, such as

- noisy galvanic stimulation
- the balance belt
- the foreseeable future—the vestibular implant

noisy galvanic stimulation



the balance belt



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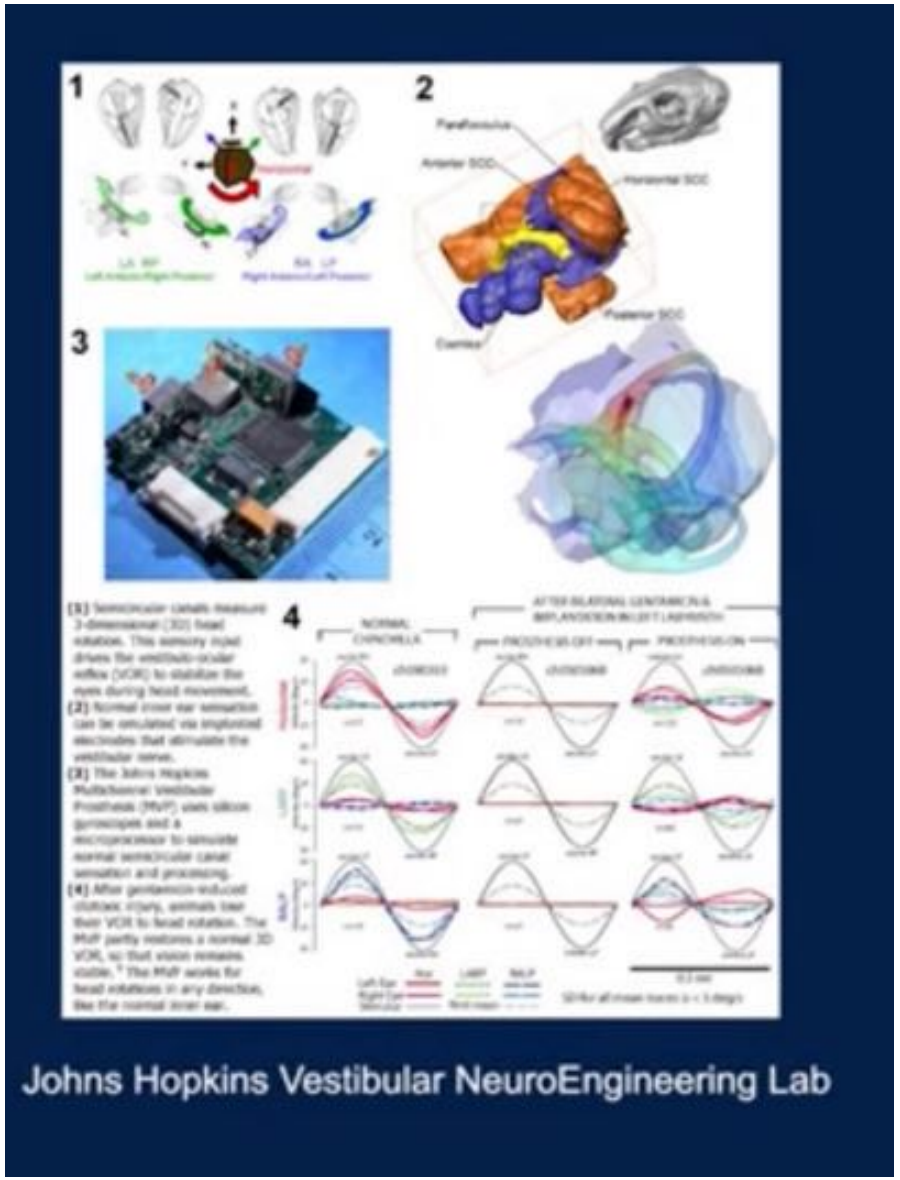
vestibular implant

Johns Hopkins Multichannel Vestibular Prosthesis

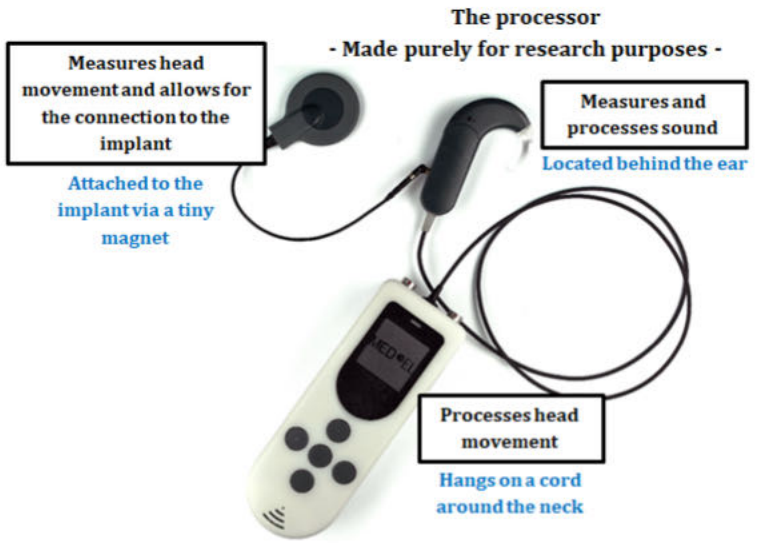


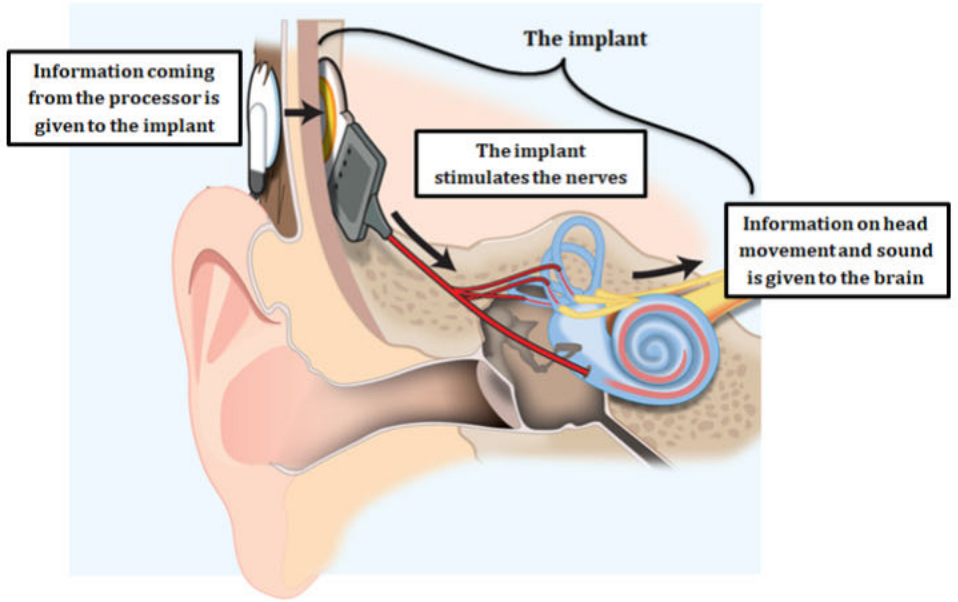
Vestibular implant (VI)

Vestibular Implantation Can Work Even After >20 Years of Severe Bilateral Vestibular Hypofunction

Desi P. Schoo, MD; Andrianna I. Ayiotis, BS; Celia Fernandez Brillet, BS; Margaret R. Chow, PhD; Kelly E. Lane; Bryan K. Ward, MD; John P. Carey, MD; Charles C. Della Santina, MD PhD

Multichannel Vestibular Implant Early [redacted] NCT02725463)





Thanks !!

The road to Barrany Sociey Meeting
Uppsala Swedon 2018