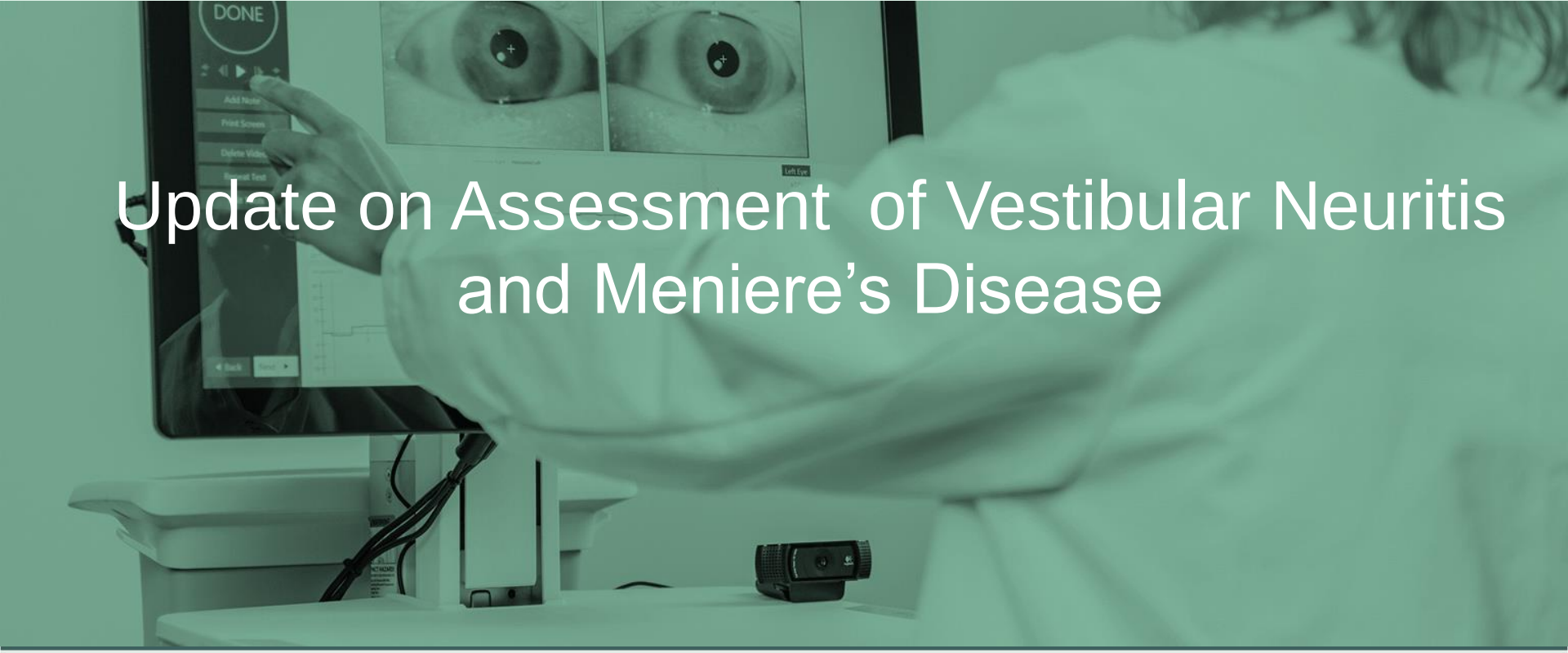

Update on Assessment of Vestibular Neuritis and Meniere's Disease

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Disclosure: Consultant to Interacoustics and Bertec Corp.

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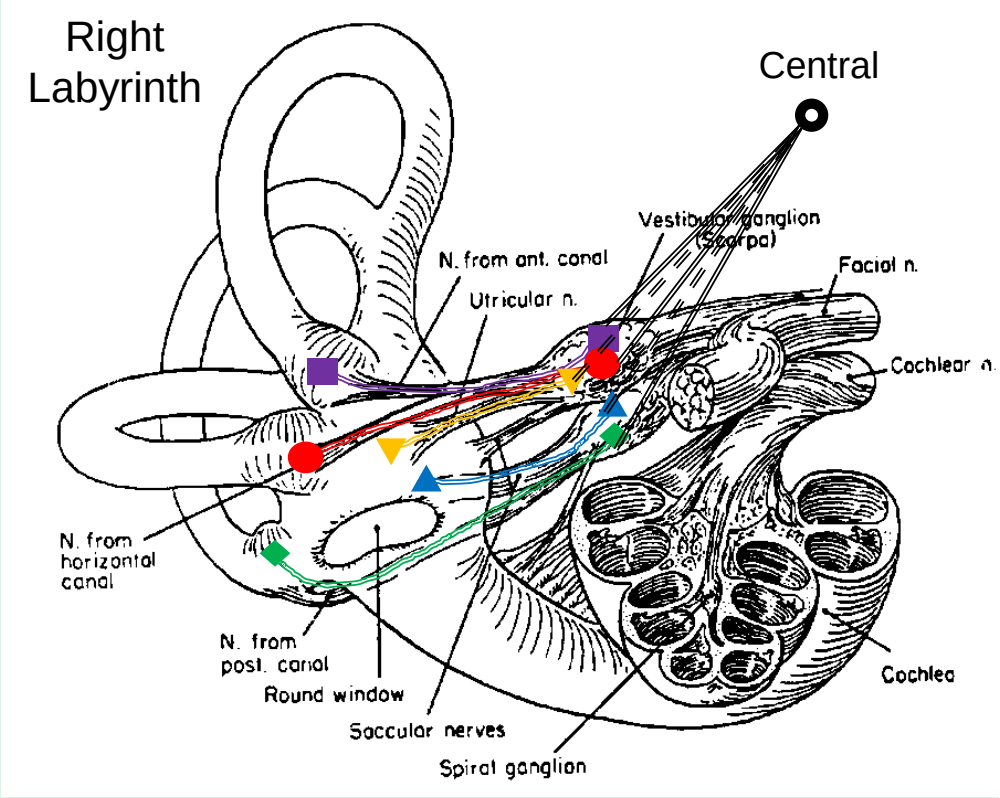
Vestibular Pathways in Different Vestibular Tests



- Caloric (irrigated lateral canal)
- Rotation (both ears)
- Lateral HIT (ipsilateral lateral canal)
- Downward HIT (contralateral anterior canal)
- Upward HIT (ipsilateral posterior canal)
- cVEMP (ipsilateral neck muscle)
- oVEMP (contralateral eye muscle)

- Spontaneous/position (peripheral and central)

Vestibular Neuritis – *Characteristics*

- Sudden onset, protracted episode of incapacitating vertigo, nausea, vomiting that lasts for ~3 days, followed by ~3 weeks of moderate symptoms, followed by another ~3 months of mild symptoms
- No associated hearing loss (distinguishes it from labyrinthitis)
- Usually caused by a viral infection (sometimes epidemic)
  - Some infections cause degeneration of the vestibular nerve (permanent loss)
  - Some infections cause inflammation of the vestibular nerve (function is usually restored once the inflammation subsides)
- Vestibular neuritis can involve:
  - The superior branch of vestibular nerve only (most common) – Patient is susceptible to developing secondary BPPV
  - The inferior branch of vestibular nerve only (rare)
  - Both branches of the vestibular nerve
    - Vestibular findings are similar to labyrinthitis patients

Vestibular Neuritis – *Involving Superior Branch Only*

- VNG/ENG test findings
  - Immediately after the onset of the lesion, the patient will exhibit horizontal and torsional nystagmus both beating away from the side of lesion and up-beating vertical nystagmus

  - Within a day or two, vertical and torsional nystagmus will decrease to the point that they may not be detectable thus making this type of lesion indistinguishable from neuritis involving both branches of vestibular nerve

  - Horizontal nystagmus will decline more slowly and may never disappear entirely
  - Caloric testing will show unilateral weakness on the side of neuritis and will persist as long as the lesion is present regardless of the compensation level
    - Loss of afferent nerve fibers should be large enough to result in an abnormal unilateral weakness finding
    - In case of recovery, such as resolution of nerve inflammation, unilateral caloric weakness will return to within normal limits

Vestibular Neuritis – *Involving Superior Branch Only*

- vHIT findings
  - vHIT results will show catch-up saccades and reduced VOR gain for lateral and anterior canal tests on the side of the neuritis
  - vHIT results will remain abnormal as long as the lesion persists
- VEMP findings
  - Normal cVEMP but oVEMP should be abnormal for the side of lesion
- SVV findings
  - SVV will show significant deviation toward the side of lesion but will improve with compensation

Vestibular Neuritis – *Involving Both Branches*

- Vestibular test findings
  - Immediately after the onset of the lesion, the patient will exhibit horizontal and torsional nystagmus both beating away from the side of lesion but unlike neuritis that involves the superior branch only, there will be no vertical nystagmus
    - Because of the quick resolution of torsional and vertical nystagmus, two types of vestibular neuritis are generally indistinguishable based on the direction of spontaneous nystagmus
  - Caloric and rotation chair findings are identical to those in patients with neuritis involving the superior branch of vestibular nerve only
  - vHIT results will show catch-up saccades and reduced VOR gain for lateral, anterior, and posterior canal tests on the side of the neuritis
  - Both cVEMP and oVEMP should be abnormal for the side of lesion
  - SVV findings should be similar to those in patients with neuritis involving the superior branch of vestibular nerve only

Vestibular Neuritis – *Involving Inferior Branch Only*

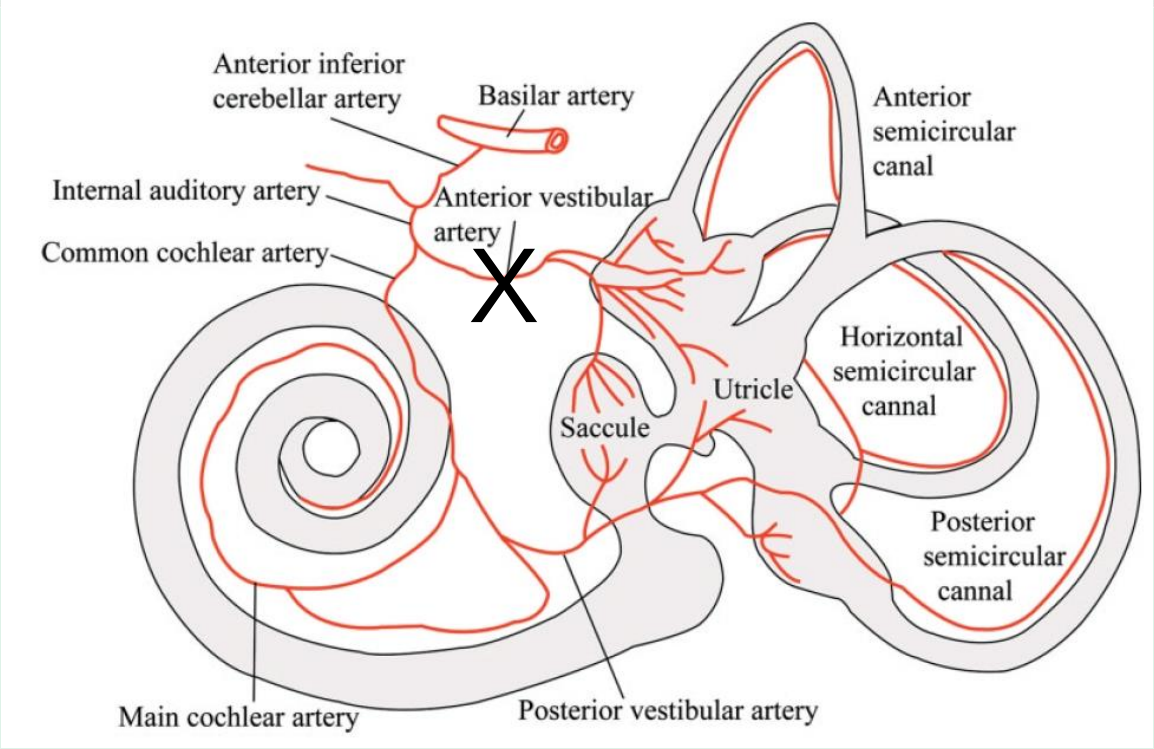
- Vestibular test findings
  - Immediately after the onset of the lesion, the patient will exhibit torsional nystagmus beating away from the side of lesion and down-beating vertical nystagmus
  - Within a day or two, vertical and torsional nystagmus will decrease to the point that they may not be detectable
  - Caloric testing will be within normal limits
    - This type of neuritis could not be detected by traditional vestibular tests!
  - vHIT results will show catch-up saccades and reduced VOR gain for the posterior canal test on the side of the neuritis
  - Normal oVEMP but cVEMP should be abnormal for the side of lesion
  - SVV results may be normal because SVV is primarily considered a test of utricles but more studies are needed

Vestibular Neuritis – *Findings in vHIT and VEMP*

Neuritis type	Lateral vHIT	Vertical vHIT (RALP/LARP)	cVEMP	oVEMP
Superior vestibular nerve only	Abnormal for impulses toward side of lesion	Abnormal for downward impulses with the head turned away from side of lesion	Within normal limits	Abnormal recordings from the eye muscle away from side of lesion
Inferior Vestibular Nerve	Within normal limits	Abnormal for upward impulses with the head turned toward side of lesion	Abnormal recordings from the neck muscle toward side of lesion	Within normal limits
Total Unilateral Vestibular Loss	Abnormal for impulses toward the side of lesion	Abnormal for downward impulses with the head turned away from side of lesion and for upward impulses with the head turned toward side of lesion	Abnormal recordings from the neck muscle on the side of lesion	Abnormal recordings from the eye muscle away from side of lesion
???	Abnormal for impulses toward side of lesion	Abnormal for downward impulses with the head turned away from side of lesion	Within normal limits	Within normal limits

- Does it matter to know the type of vestibular neuritis?

Vestibular Neuritis or Vascular Origin?



From Kim and Lee, 2009

- Ischemic events lasting over 30 minutes can permanently damage hair cells in different vestibular structures without leaving any trace in the imaging studies
  - Anterior vestibular artery syndrome – damage to the anterior and lateral canals without any damage to the vestibular nerve

Vestibular Neuritis – *Management*

- Within the first few days following the onset of symptoms, the patient can be given a short course of corticosteroids
  - May expedite the recovery in cases of inflammation of vestibular nerve
- In case of severe symptoms, the patient can be given vestibular suppressants but it should be discontinued after a few days so that the vestibular compensation process is not impeded
- The patient should be educated about the course of the disease (~3 days of severe symptoms followed by ~3 weeks of moderate symptoms followed by ~3 months of mild symptoms)
- Vestibular exercises can be given to expedite recovery but in most cases, it is not mandatory at early stages
- The patient should also be informed about the possibility of developing BPPV because the posterior canal and inferior portion of the vestibular nerve are intact

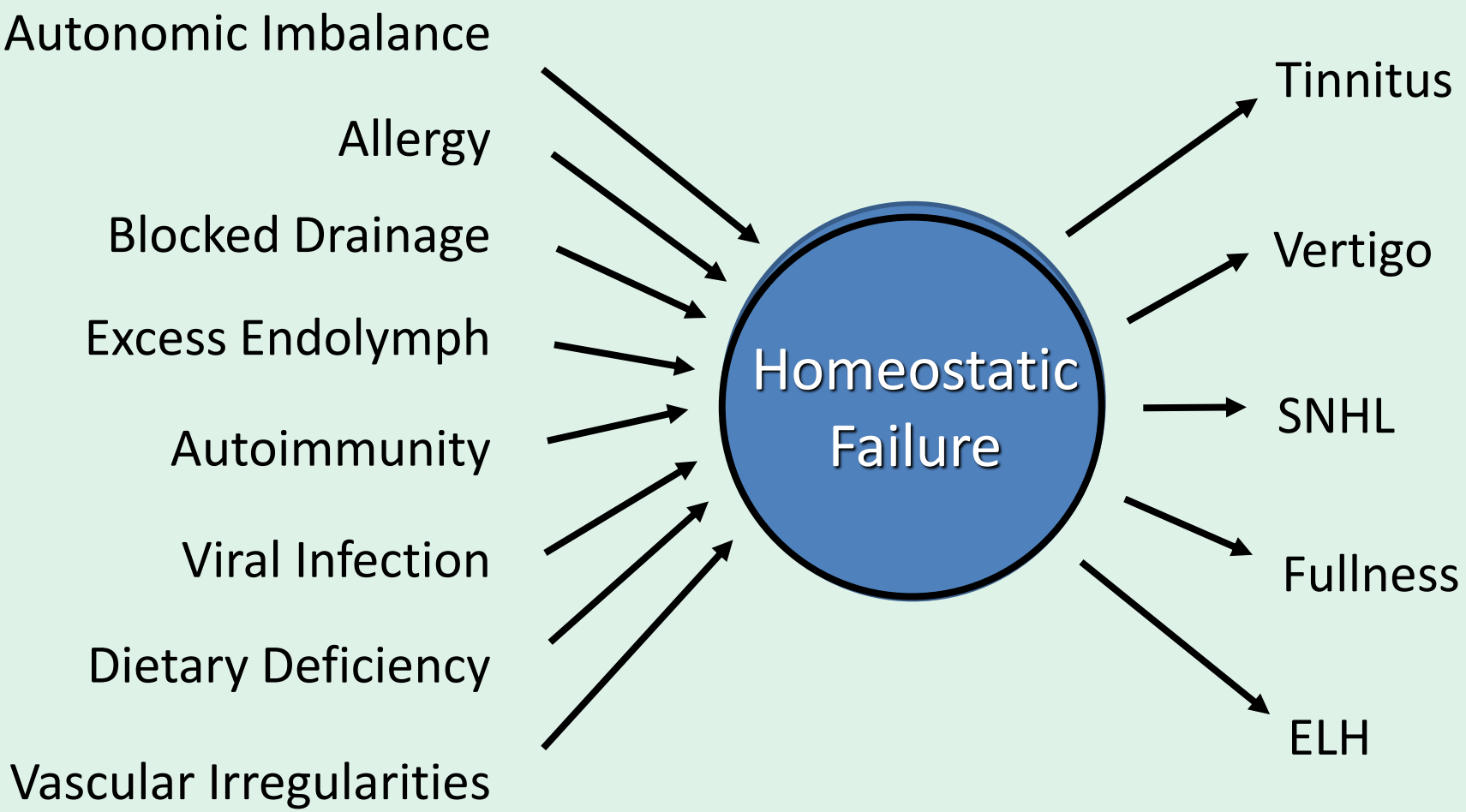
Meniere's Disease (MD)

- Diagnostic criteria for Definite MD (Lopez-Escamez, et al 2015)
  1. Two or more spontaneous episodes of vertigo, lasting 20 mins to 12 hours
  2. Audiometrically documented low- to medium-frequency SNHL in one ear, defining the affected ear on at least one occasion before, during or after one of the episodes of vertigo
     a. Unilateral: BC 30 dB poorer than better ear at 2 contiguous frequencies below 2k Hz
     b. Bilateral LF SNHL: absolute BC thresholds 35 dB or poorer at 2 contiguous frequencies below 2k Hz
     c. Delayed MD: SNHL may antedate the onset of vertigo episodes by several weeks, months or years
  3. Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear
     a. Change of hearing should be noticed within 24 hours of vertigo episode
  4. Not better accounted for by another vestibular diagnosis

Meniere's Disease (MD)

- Diagnostic criteria for Probable MD (Lopez-Escamez, et al 2015)
  1. Two or more episodes of vertigo or dizziness, each lasting 20 mins to 24 hours
  2. Fluctuating aural symptoms (hearing, tinnitus or fullness) in the reported ear
     a. Fluctuating symptoms must be reported during the vertigo episode
     b. Increased tinnitus intensity or aural fullness in the affected ear is usually associated with episodes of vertigo in the first years
  3. Not better accounted for by another vestibular diagnosis

Pathophysiology of Meniere's Disease



Meniere's Disease – *Vestibular Test Findings*

- Spontaneous nystagmus
  - Immediately after the onset of the attack, the patient will exhibit horizontal spontaneous nystagmus beating toward the side of lesion indicating excitation of the involved ear
  - After a few minutes, spontaneous nystagmus will beat away from the side of lesion indicating inhibition of the involved ear
  - Spontaneous nystagmus will decrease over several days or a few weeks
  - After about 5 days, some patients will exhibit nystagmus that again beats toward the side of lesion most likely due to the return of function to the involved labyrinth during the cerebellar clamping stage of vestibular compensation (inhibition of the responses from the intact side)
    - This is called recovery nystagmus and lasts no more than a few days

Meniere's Disease – *Vestibular Test Findings*

- Caloric testing
  - Shortly after the onset of the attack, the patient will exhibit unilateral caloric weakness in the involved ear
  - In the early stages of the Meniere's disease, caloric responses return within normal limits after several days
  - In the latter stages, caloric weakness will persist indicating permanent loss of vestibular function

Meniere's Disease – *Vestibular Test Findings*

- VEMP
  - VEMP findings vary depending on the elapsed time since the onset of the last attack and the disease stage
  - ~50% of patients with Meniere's disease have asymmetric cVEMP responses, indicating differences in the saccular function (returns to normal in ~50%)
  - Recent reports show different frequency tuning in cVEMP thresholds for 250 Hz – 1 kHz stimuli in patients with endolymphatic hydrops (Rauch et al 2004)
    - Some patients show the same pattern in the unaffected ear, perhaps indicating delayed endolymphatic hydrops
  - 40% of Meniere's patients show improvement in cVEMP amplitude following the administration of furosemide (Seo et al, 2003)
  - VEMPs can be used to monitor gentamycin therapy

Meniere's Disease – Frequency Tuning in VEMPs

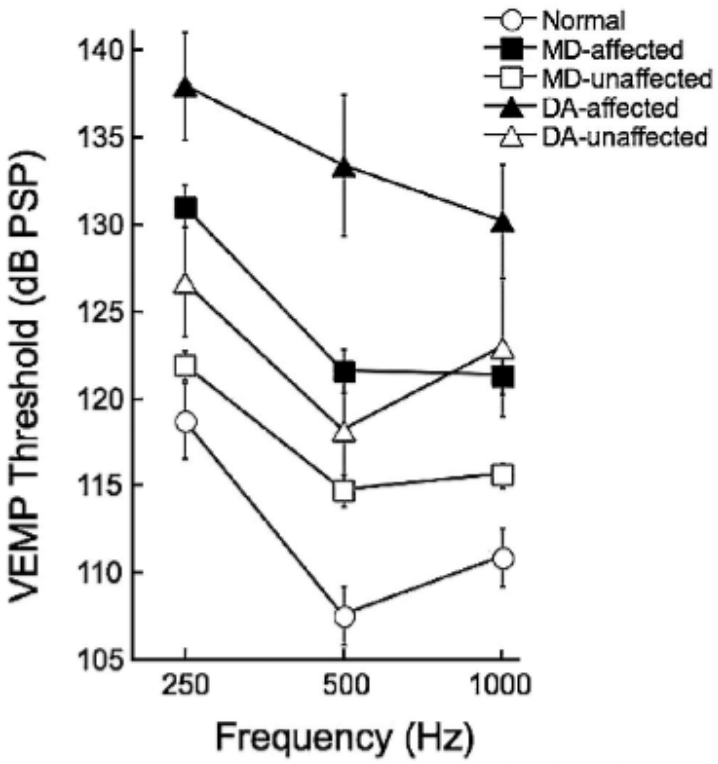
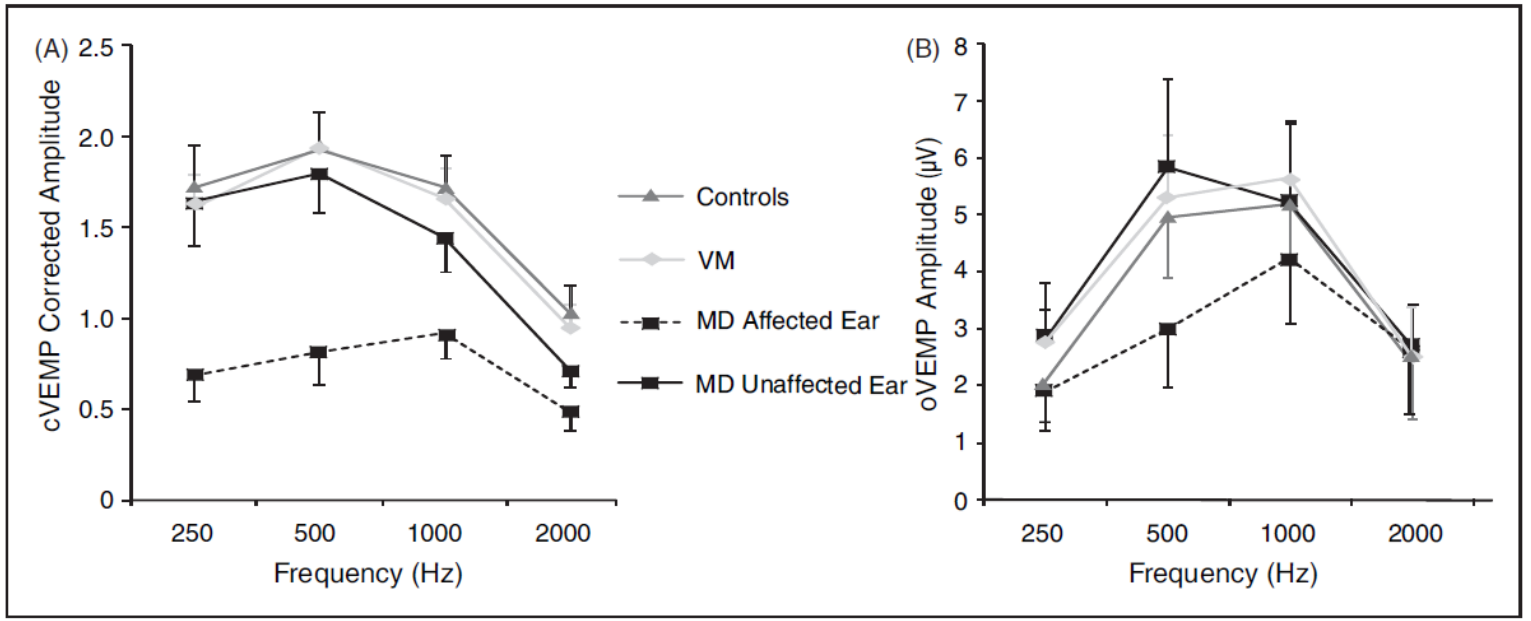


Fig. 2. Mean (± standard error of mean) vestibular evoked myogenic potential threshold versus stimulus frequency for each type of ear. MD – Ménière's disease; DA – drop attacks.

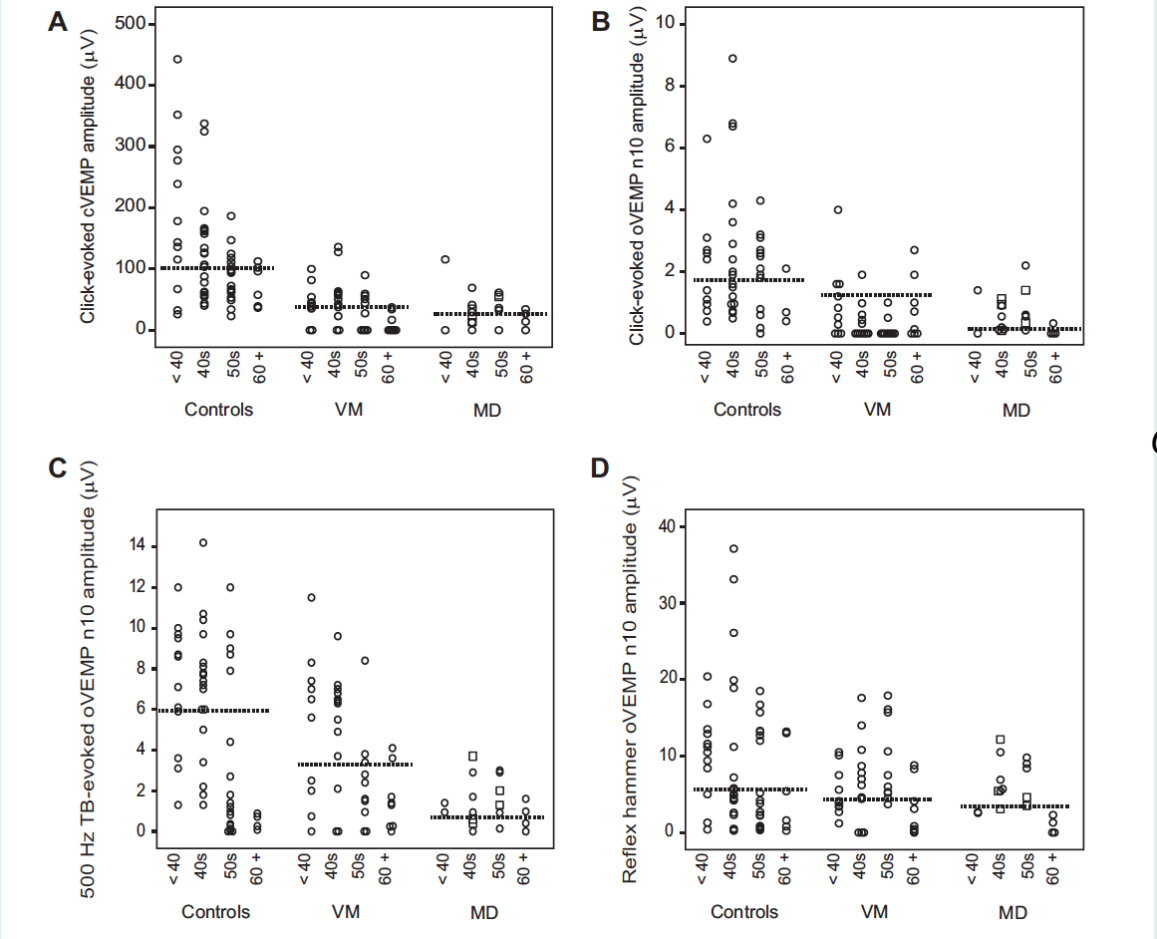
Ferdinand et al., *The Laryngoscope*, 2006.

Meniere's Disease – Frequency Tuning in VEMPs



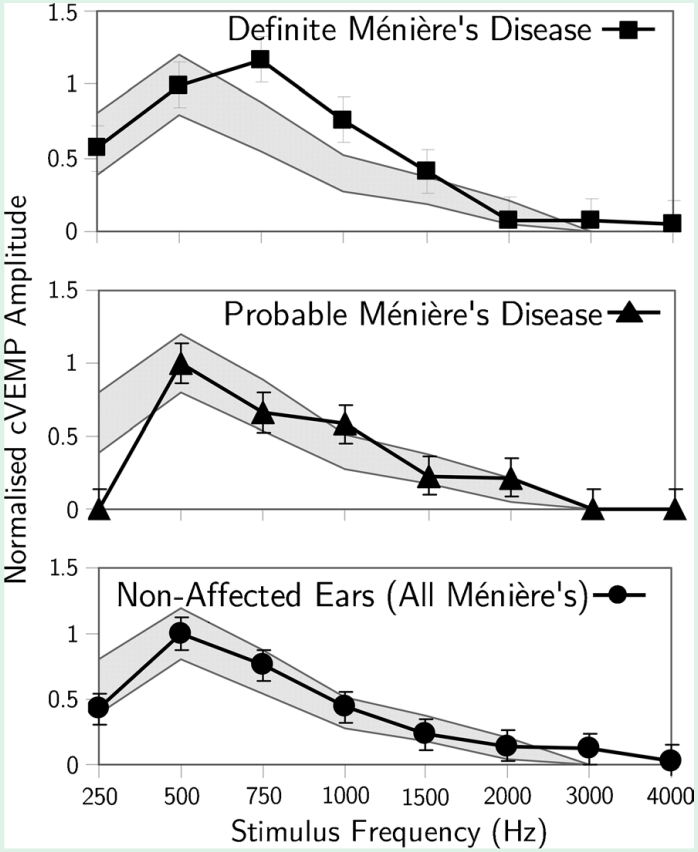
Taylor et al., *Cephalalgia*, 2012

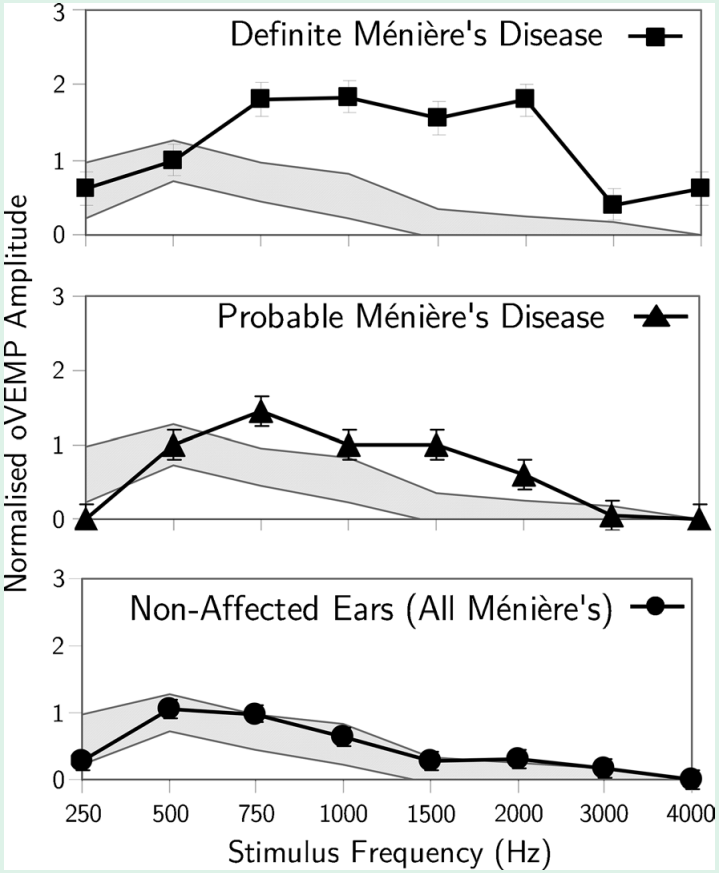
Meniere's Disease – VEMPs



Zuniga et al., *Otolaryngology Head-Neck Surgery*, 2012

Meniere's Disease – *VEMPs*





From Sandhu et al., (2012)

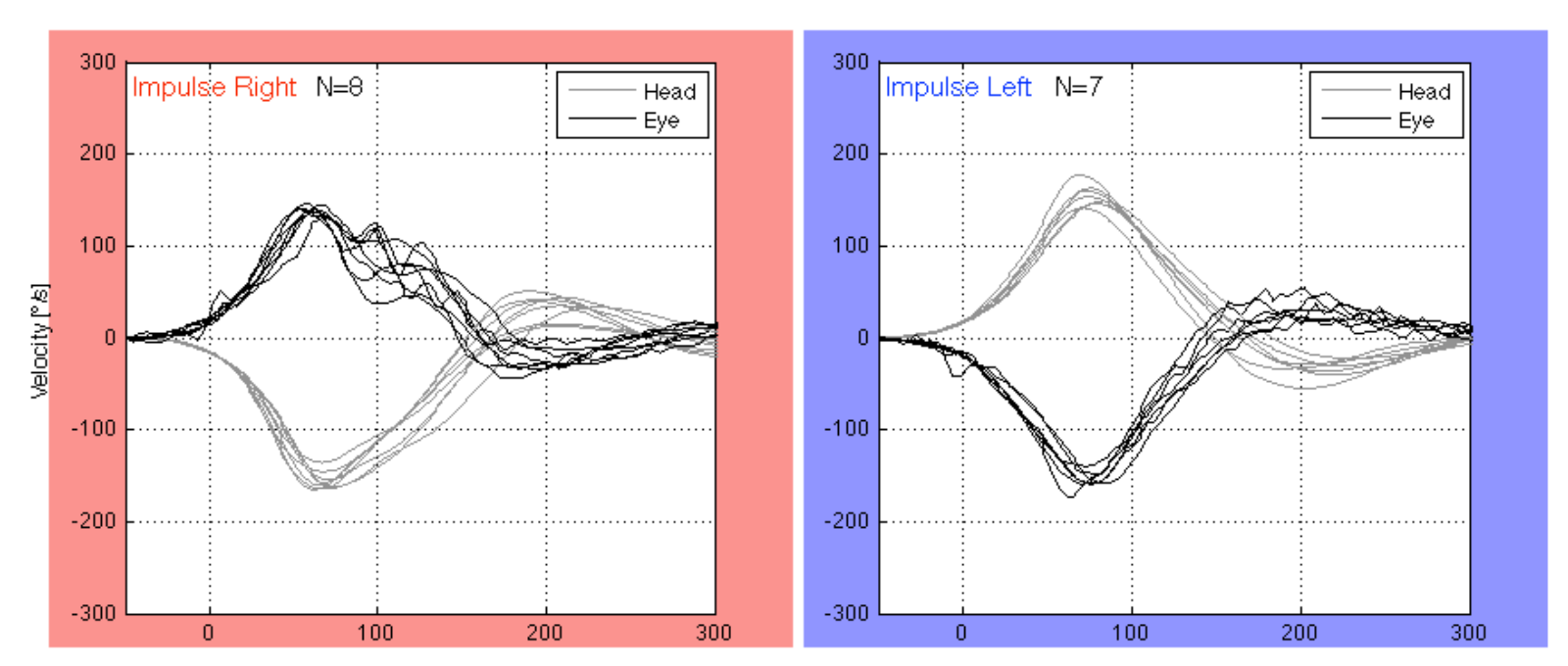
- P1-N1 amplitudes for different frequencies in patients with suspected Meniere's disease

Meniere's Disease – *Vestibular Test Findings*

- vHIT
  - Lateral vHIT and caloric test findings should have a similar time course depending on the elapsed time from the onset of the last attack and the disease stage
  - There has been reports of Meniere's patients with normal vHIT but abnormal calorics
    - Frequency dependency or whirlpool effect?
  - There has been a report about fluctuations of vHIT results in patients with Meniere's that suggests increased VOR gain in lateral HIT during the silent period between Meniere's attacks (Manzari et al, 2010)
    - Needs further validation, probably an artifact
  - vHIT can be used to monitor gentamycin therapy
  - No reports of vertical HIT testing in Meniere's patients yet

Meniere's Disease – *Lateral vHIT*

- Head thrust testing (bedside HIT) – negative bilaterally
- Video head impulse test (vHIT) – normal response from both lateral semicircular canal
  - Lateral semicircular canal gain: Right = 0.87 and Left = 0.92



ANNALS OF THE NEW YORK ACADEMY OF SCIENCES
Issue: *Dizziness and Balance Disorders*

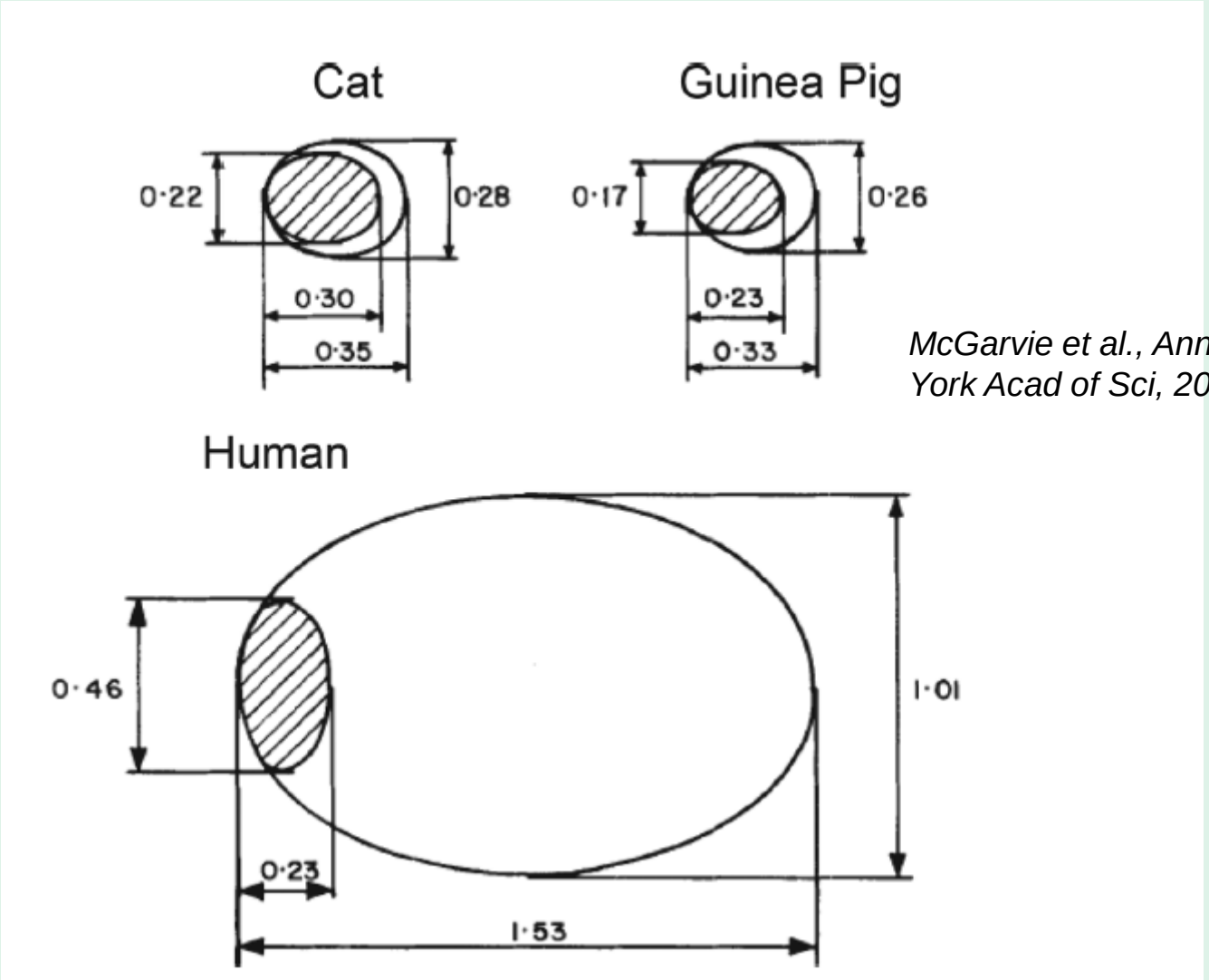
What does the head impulse test versus caloric dissociation reveal about vestibular dysfunction in Ménière's disease?

Leigh A. McGarvie,[1] Ian S. Curthoys,[2] Hamish G. MacDougall,[2] and G. Michael Halmagyi[1]

The Dissociation of Video Head Impulse Test (vHIT) and Bithermal Caloric Test Results Provide Topological Localization of Vestibular System Impairment in Patients With "Definite" Ménière's Disease

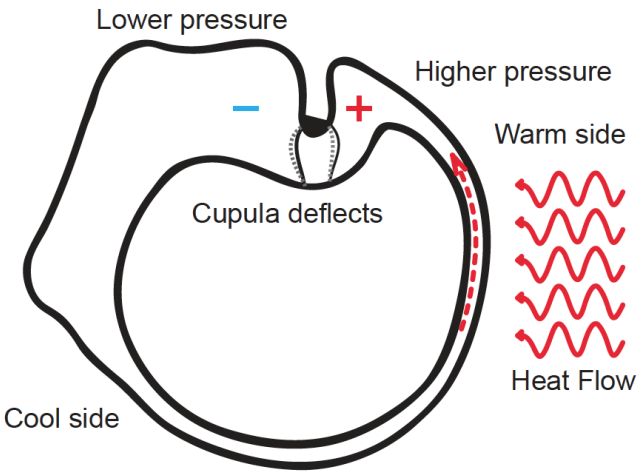
Devin L. McCaslin,[a] Alejandro Rivas,[a] Gary P. Jacobson,[a] and Marc L. Bennett[a]

Meniere's Disease – Dissociation Theory

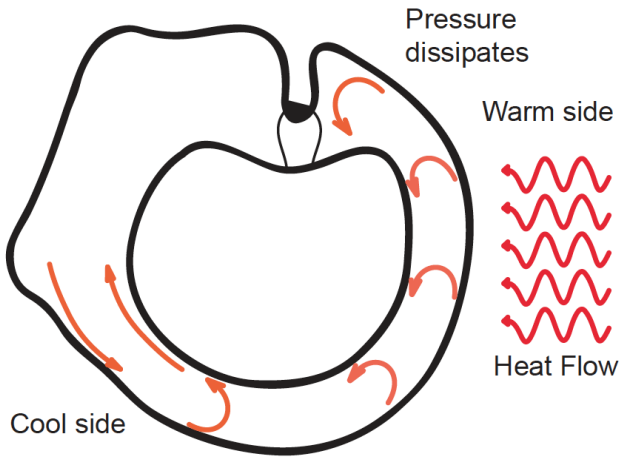


McGarvie et al., Annals of New York Acad of Sci, 2015

Meniere's Disease – Dissociation Theory



Normal Duct: No local flow, hydrostatic pressure drive retained and Cupula bent.



Hydropic Expansion of Duct: Local convective flow dissipates hydrostatic pressure across Cupula.

McGarvie et al., Annals of New York Acad of Sci, 2015

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