**15th Annual Advances in Neurology**

*Houston Methodist Neurological Institute*

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# *Advances in Vertigo and Neuro-Otology*

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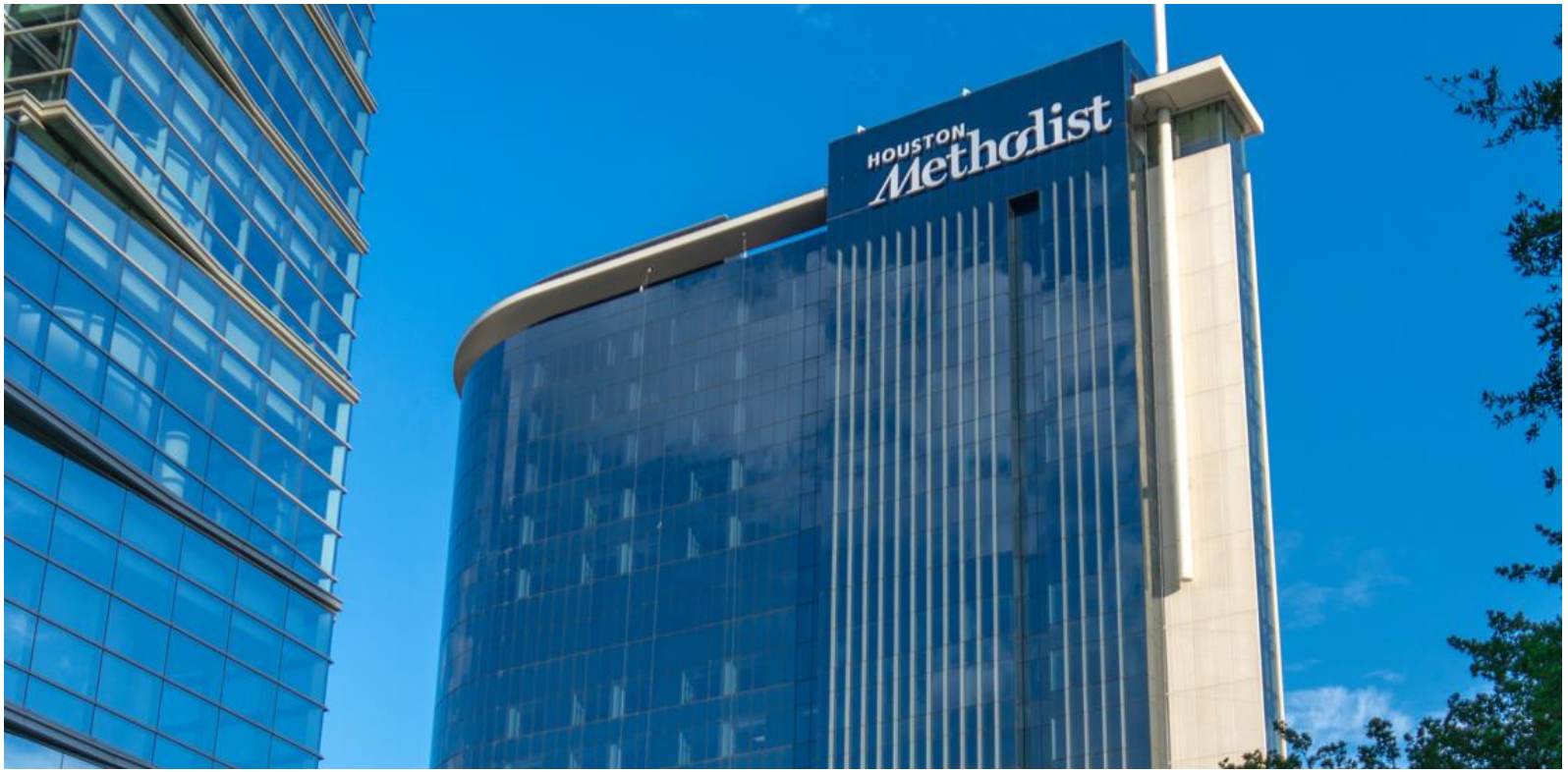
Phoenix, Arizona




**Dr. Fife has no personal of financial conflicts of interest to disclose**

# Overview

1. What is Neuro-Otology?
2. Why is dizziness/vertigo a problem?
3. Ways to Approach to Patients with Dizziness
4. Review of Common Causes of Dizziness
    a. Benign Paroxysmal Positional Vertigo (BPPV)
    b. Acute Vestibular Syndrome (AVS): Stroke vs Vestibular Neuritis
    c. Persistent Postural Perceptual Dizziness (PPPD)
    d. Vestibular Migraine
    e. Meniere's Disease
    f. Superior Canal Dehiscence Syndrome
5. Conclusions

# Neuro-Otology, Otology, Vestibular Neurology, Neurotology

Otology

- Specialty since 1820 NY Eye & Ear
- 1927 MEEI
- LA Foundation for Otology 1959
- House Ear Institute 1981

Neurotology (includes skull base surgery)

- Fellowships since 1960
- An ACGME Otolarygnological Subspeciality Otolaryngology RRC
- Requirements, approved 1994 (40 sites)

Neuro-Otology, neurology trained (Robert W. Baloh, Davis S. Zee)

- Otoneurology, term preferred by some
- Vestibular Neurology with twists
- Currently not ACGME or UCNS

# Why is dizziness/vertigo a problem?

**IT AFFECTS MANY OF US**: Significant dizziness 17-30%, significant vertigo 3-10% lifetime prevalence (1). Females affected by dizziness 2:1, mostly at ages 40-60 years (2)

**REDUCES OUR ABILITIES IN WORK AND LIFE:** Adverse effect on work productivity (2):

- 70% had reduced workloads
- 63% lost work days
- 6% quit or lost their job due to vertigo or dizziness

# Why is the cause dizziness challenging to diagnose?

- Not all dizziness is Vestibular in origin: A surgical ENT neurotology (ENT) clinic found > half of females 60-69 had non-peripheral vestibular causes of dizziness (3).
- About 1 in 6 patients present with 2 or more causes of dizziness at the same time; the most common combinations were VM + BPPV and VM + Meniere's (3)

1.Murdin L, Shilder A. Otol Neurotol 2015;36:387-92
2.Benecke H, et al. Front Neurol 2013;4:136
3.Muelleman T, et al. Otol Neurotol 2017;38:870-5

# Costs and Obstacles to care

Delay in diagnosis is common and leads to unnecessary costs and limitations in function (Fife 2005, von Brevern 2007, Polensek & Tusa, 2008, Wang 2014)

In ED's and other settings, patients with suspected BPPV are often not examined for the diagnosis and ever more rarely treated. This seems due to lack of training or unfamiliarity of technique (Bashir 2015, Kerber 2013, Kerber 2015)

# ICVD Definitions

**Vertigo** = Sensation of self-motion when no self-motion is occurring or the sensation of distorted self-motion during an otherwise normal head movement.

- Spontaneous or Triggered

**Dizziness** = Sensation of disturbed or altered spatial orientation without the feeling of false motion

- Spontaneous or Triggered

**Postural symptoms** = Balance-related symptoms that occur when upright. For example, feeling unsteady or as if one could fall when standing or walking.

- Unsteady or Pulsion or Imminent fall sensation or Due to Vestibular symptoms*

*"vestibular symptoms" refers to sensations of motion, tilting, swaying, etc. but is not meant to imply that all conditions causing these symptom have a basis in vestibular pathways or mechanisms

Bisdorff A, et al. J Vestib Res 2009;19:1-13

# Approach of Dizzy Patients

**Pattern Recognition** – Use history and examination and use data to identify the pattern or overlapping patterns that fit a known cause(s)

**ICVD Scheme** – Hx and exam to define Acute Vestibular Syndrome, Episodic Vestibular Syndrome (episodic or triggered), Chronic vestibular syndrome

**TiTrATE and ATTEST -** TiTrATE is an acronym for **Ti**ming, **Tr**iggers, **A**ssociated symptoms and **T**argeted **E**xamination (1) ATTEST is an acronym for **A**ssociated symptoms, **T**iming, **T**riggers, **E**xamination **S**igns and **T**esting (2)


1. Newman Toker and Edlow. Neurol Clin 2015;33:577-99
2. Edlow JA, et al. J Emerg Med 2018;54:469-83.

# History

Onset, Periodicity and Duration

Description – categorize as vertigo, pre-syncope, imbalance without vertigo, non-specific dizziness

Triggers – Are the obvious reproducible events/activities that evoke symptoms? Also ask about things that aggravate or mitigate symptom severity.

Associated Features – hearing loss? Focal neurological symptoms? Migraine-like headaches? Autophony? Oscillopsia or visual symptoms?

Impact on Quality of Life – How much is this impacting function? How concerned is the patient? Do they have a specific goal for the visit? Just want reassurance they are okay or are they desperate for a treatment?

Prior treatments tried – this can include attempts at physical therapy, medication trials. How did they respond. What was done in physical therapy? What dosage of medications and did they give it an adequate trial time?

# Examination

Observation – demeanor and affect? Did the patient come alone or with others? Assistive device? For how long and why are they using it?

Neurological - Horner's? Hemisensory loss? Unilateral facial weakness? Slurred speech? Limb clumsiness? Dysconjugate gaze? Abnormal reflexes? Orthostasis on vital signs? Gait is normal or abnormal? Able to tandem walk? Gait speed, ignition, floor clearance? Romberg?

Eye movements – saccade velocity, accuracy?

Nystagmus (describe, direction of the fast phase)

- Spontaneous
- Gaze-evoked
- Positional

Exam of VOR function

- HIT, Dix Hallpike, Dynamic visual acuity, head shaking nystagmus, Fukuda stepping test, Past-pointing

# Lab Testing

CBC, CMP, AED levels, Vit B12, HbA1c, Thyroid studies, ANA screen, ESR, FTA-Abs, anti-GAD65 antibodies, thiamine levels

# Cardiac/hemodynamic tests

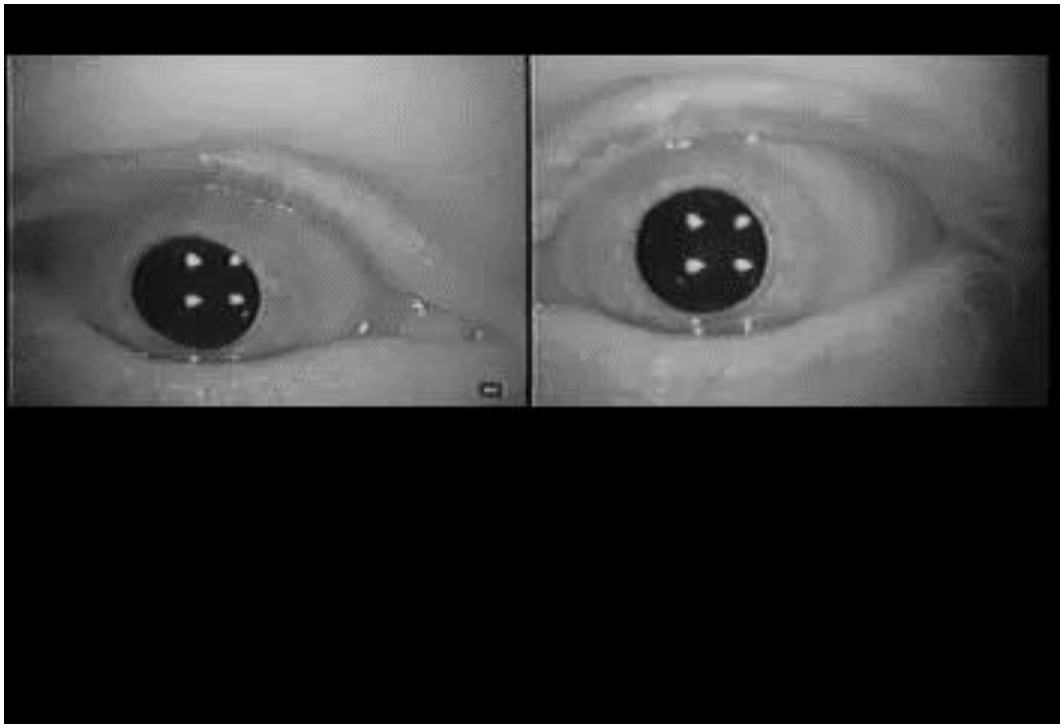
- Cardiology consult
- ECG, Holter, Event monitor
- Tilt table

# Audiovestibular testing

- Hearing test
- Videonystagmography (VNG)
- Vestibular evoked myogenic potential (VEMP)
- Video Head Impulse Test (vHIT)
- Posturography
- Rotational chair testing

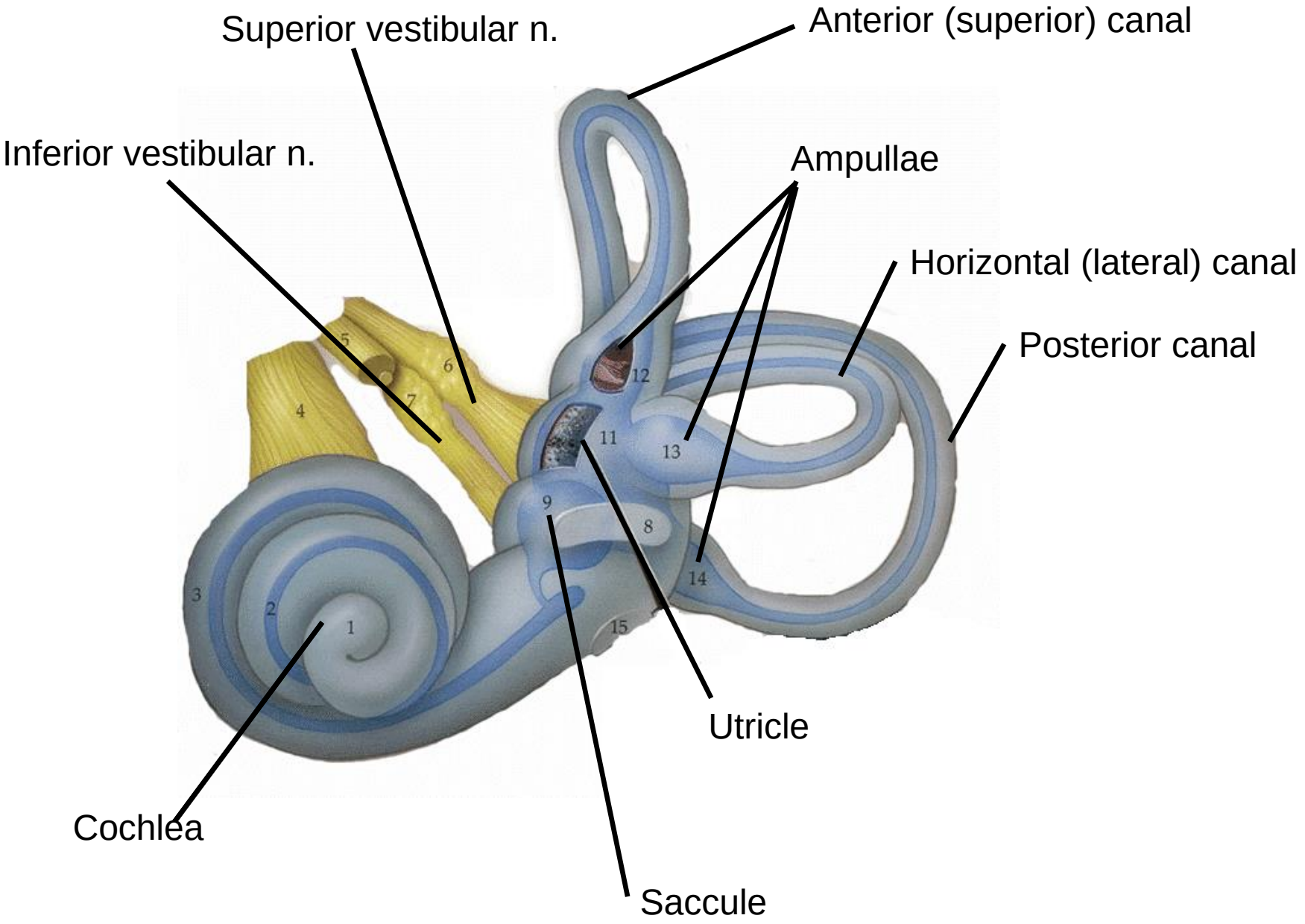
Case:  68 year old woman is seen due to 3 weeks of intermittent spinning vertigo evoked by tilting her head back, turning in bed and sometimes bending over.

EXAM:  normal neurological exam but Dix-Hallpike to the left shows…



Courtesy of
J. White, MD

# Benign Paroxysmal Positional Vertigo (BPPV)


Superior vestibular n.
Anterior (superior) canal
Inferior vestibular n.
Ampullae
Horizontal (lateral) canal
Posterior canal
Utricle
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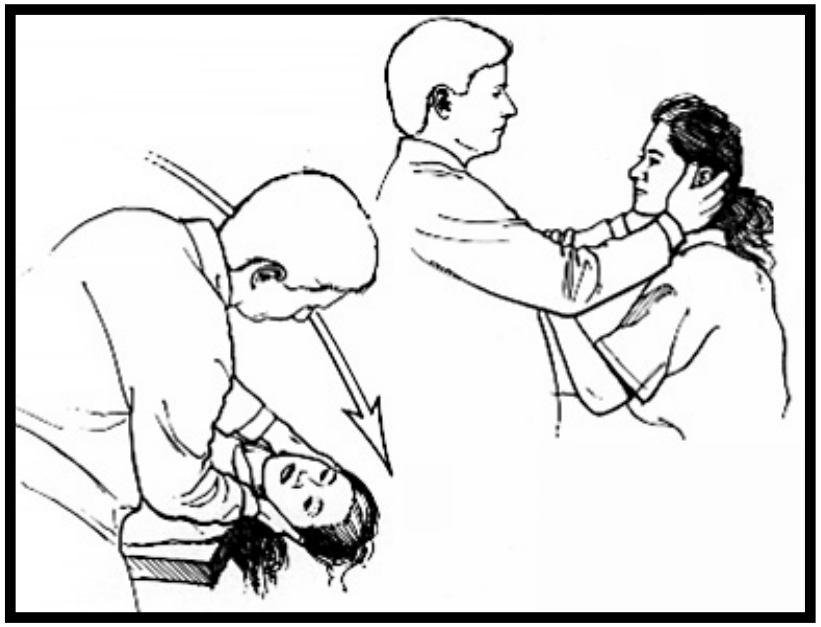
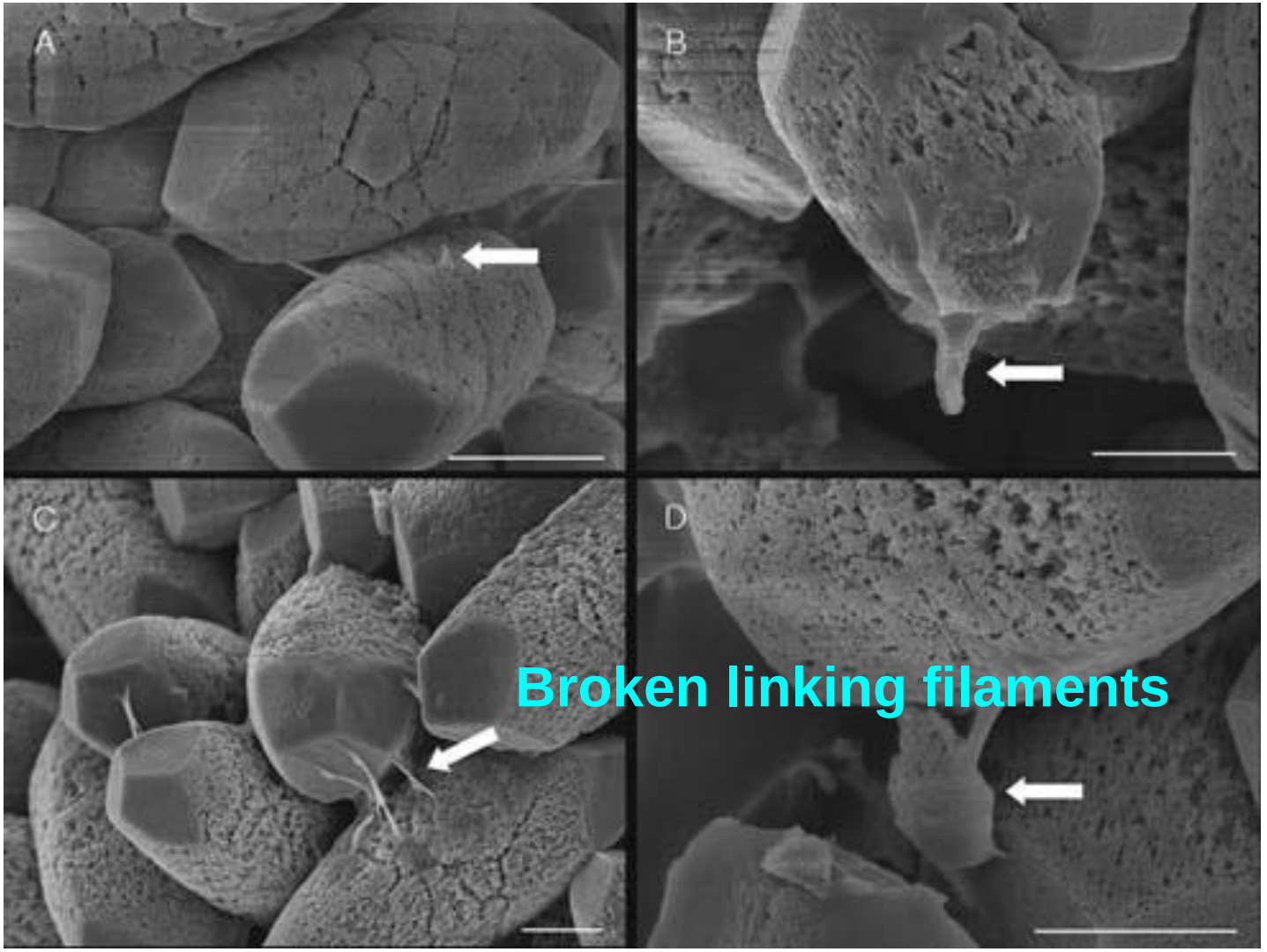
# Dix-Hallpike maneuver




A
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Broken linking filaments

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Otolaryngology–Head and Neck Surg

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Otolaryngology–
Head and Neck Surgery
Vol. 156(3S) S1–S47
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d Neck

Cochrane Library

Cochrane Database of Systematic Reviews

# The Epley (canalith repositioning) manoeuvre for benign paroxysmal positional vertigo (Review)

Hilton MP, Pinder DK

Ri

G.S. Gronseth, MD

Address correspondence and reprint requests to the American Academy of Neurology, 1080 Montreal Ave., St. Paul, MN 55116

BPPV an
tatory response is
otoliths (calcium carbonate
current of endolymph within the affe
cular canal. The most common form of BPPV
curs when otoliths from the macula of the utricle
fall into the lumen of the posterior semicircular

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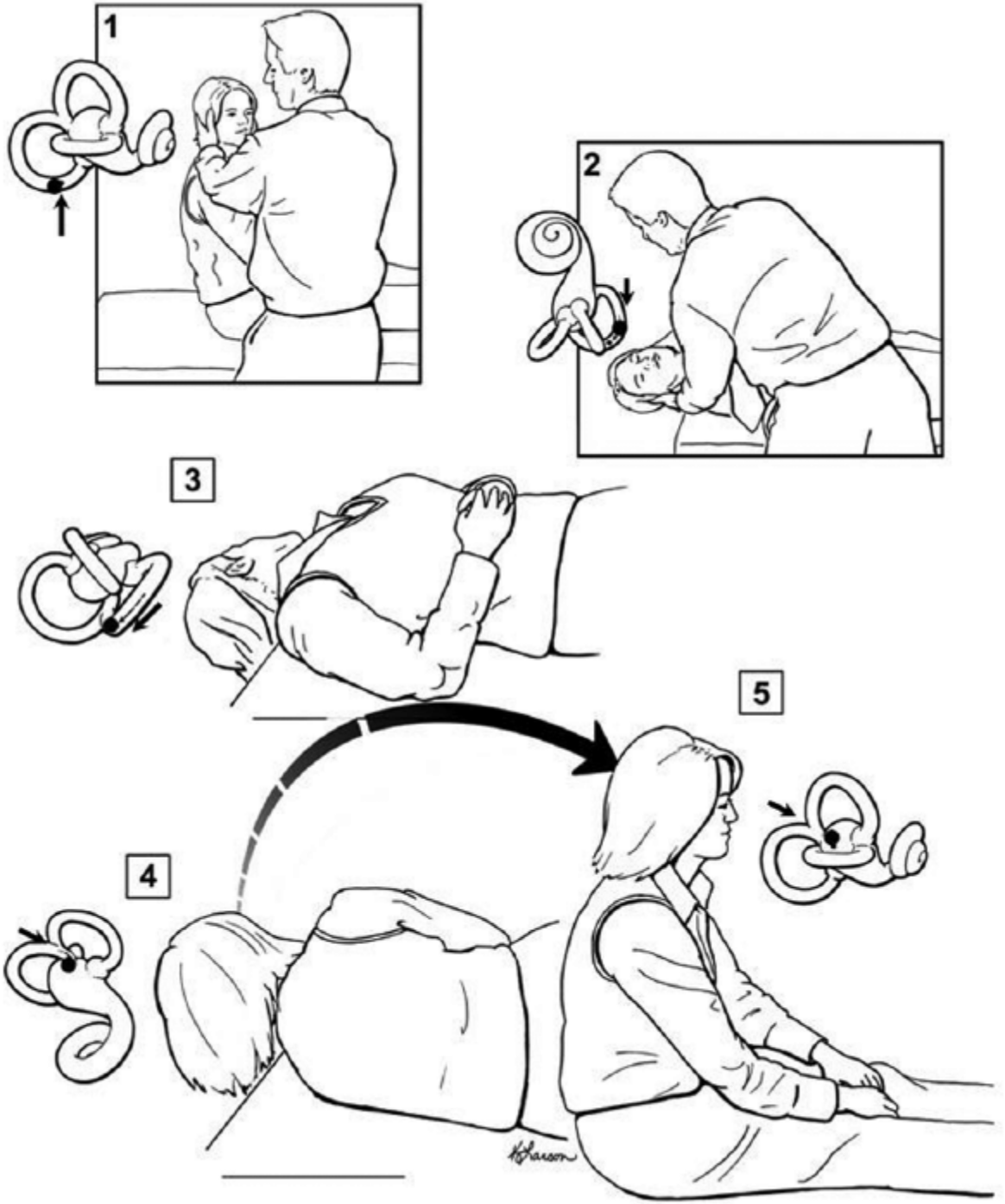
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## Clinical History

59-year-old man had the onset of vertigo at 7 am. He felt some dizziness turning in bed and when he got out of bed he staggered and had to hold on to walk to the bathroom. The dizziness continued to worsen over the subsequent 45 minutes. He says it was spinning with nausea, vomiting and dry heaves. Upon arrival to the ED the spinning continues as does nausea and worsens with any kind of movement of the head about 2 hours after its onset. He has never had any vertigo like this before. Exam was notable for moderate unsteadiness walking and left-beating nystagmus in left gaze not present with gaze to the right.

# Acute Vestibular Syndrome

**Acute Vestibular Syndrome** = acute vertigo, n/v worse with head motion, associated acutely with nystagmus, lasting 24 hours to several weeks (Hotson 1998) and caused by 2 main conditions:

**(1) Stroke of the brainstem or cerebellum (5%)**

**(2) Acute Unilateral Peripheral Vestibulopathy (95%)** = acute unilateral loss of peripheral vestibular function by any cause.

**a. Vestibular neuritis (VN)** = inflammation of the vestibular nerve, by viral infection, reactivation of herpes family virus or by non-viral immune mechanisms. *Other terms:* Vestibular neuronitis, neurolabyrinthitis, labyrinthitis, acute vestibular paralysis, epidemic vertigo.

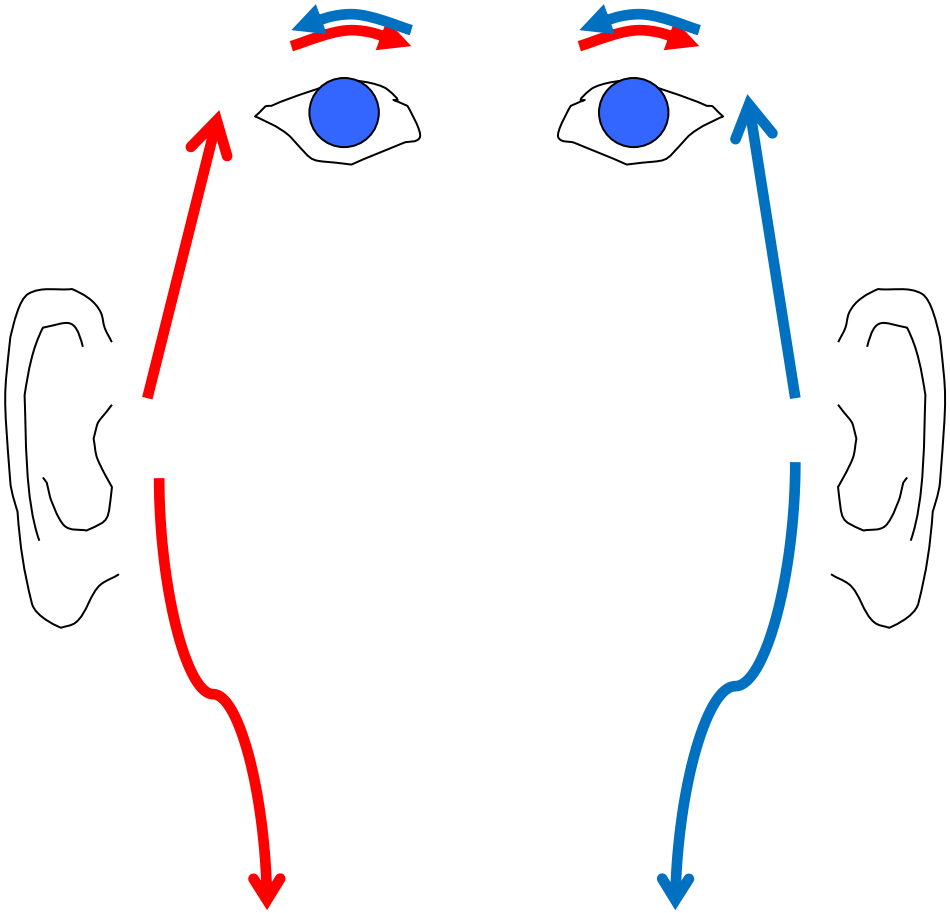
**b. Meniere's disease**

**c. Other causes of acute unilateral vestibular loss**

# HINTS Exam

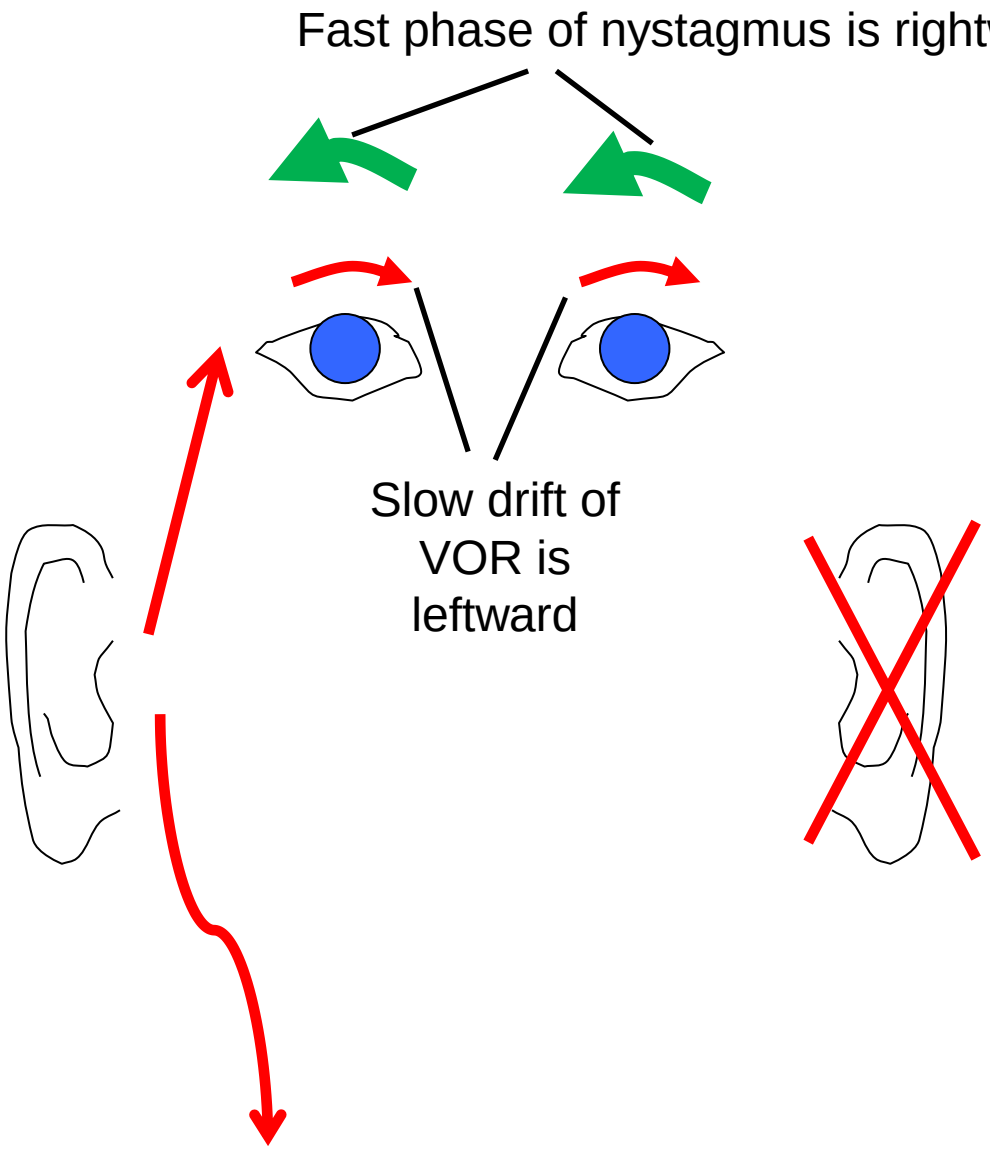
| Hints exam test | Peripheral vertigo | Central vertigo |
|---|---|---|
| Horizontal head Impulse test (h-**HI**T) | Positive with catch up saccade | Normal with no catch up saccade |
| Nystagmus (**N**) | Horizontal only in one direction | Spontaneous vertical horizontal gaze evoked |
| Test of Skew (**TS**) | No skew (negative) | Skew present |

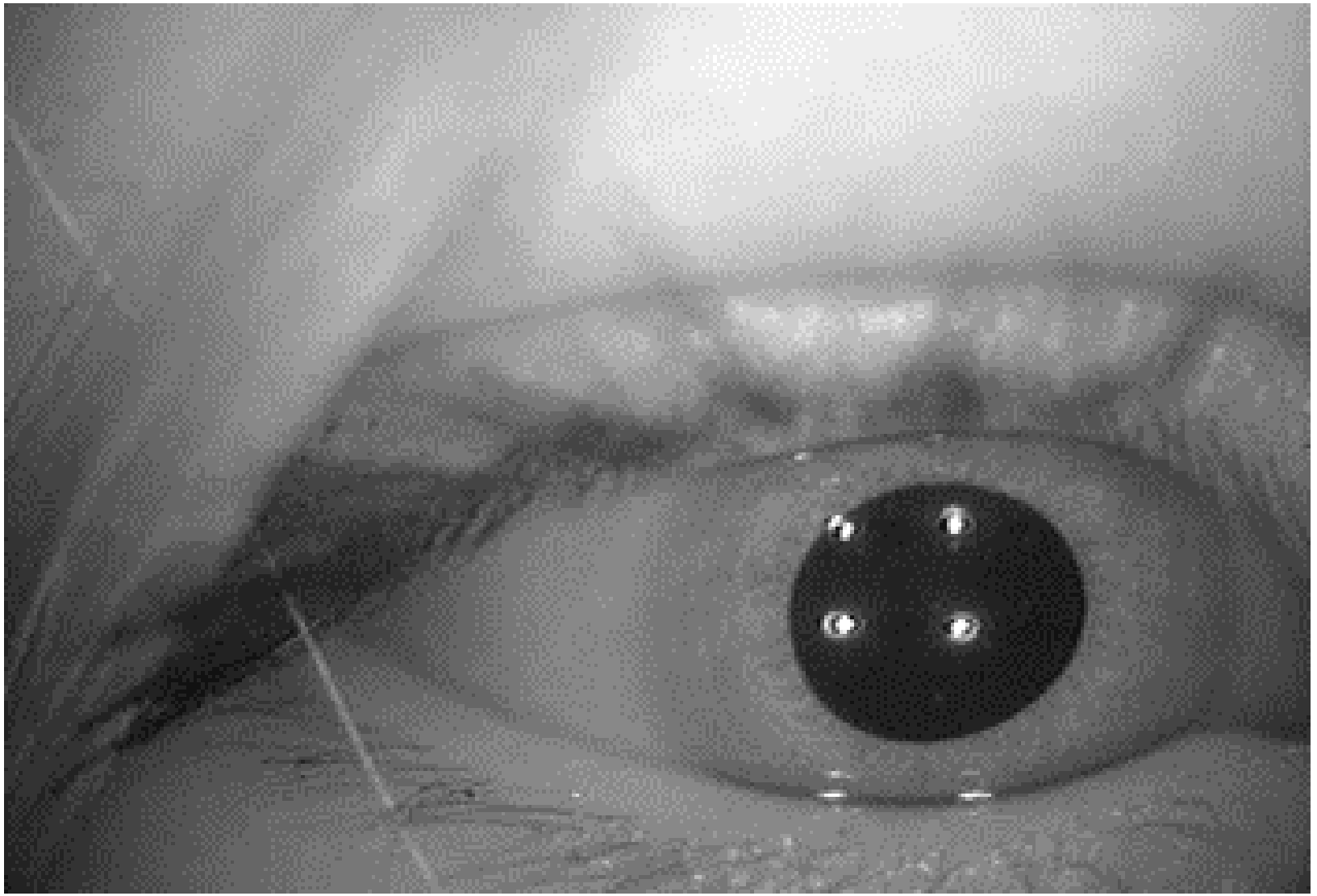
Head impulse test:  It shows that the response is normal with quick turn to the patient's right, abnormal with quick turn to the left.  This indicates impaired function of the left horizontal semicircular canal and normal function on the right. (courtesy of RW Baloh)

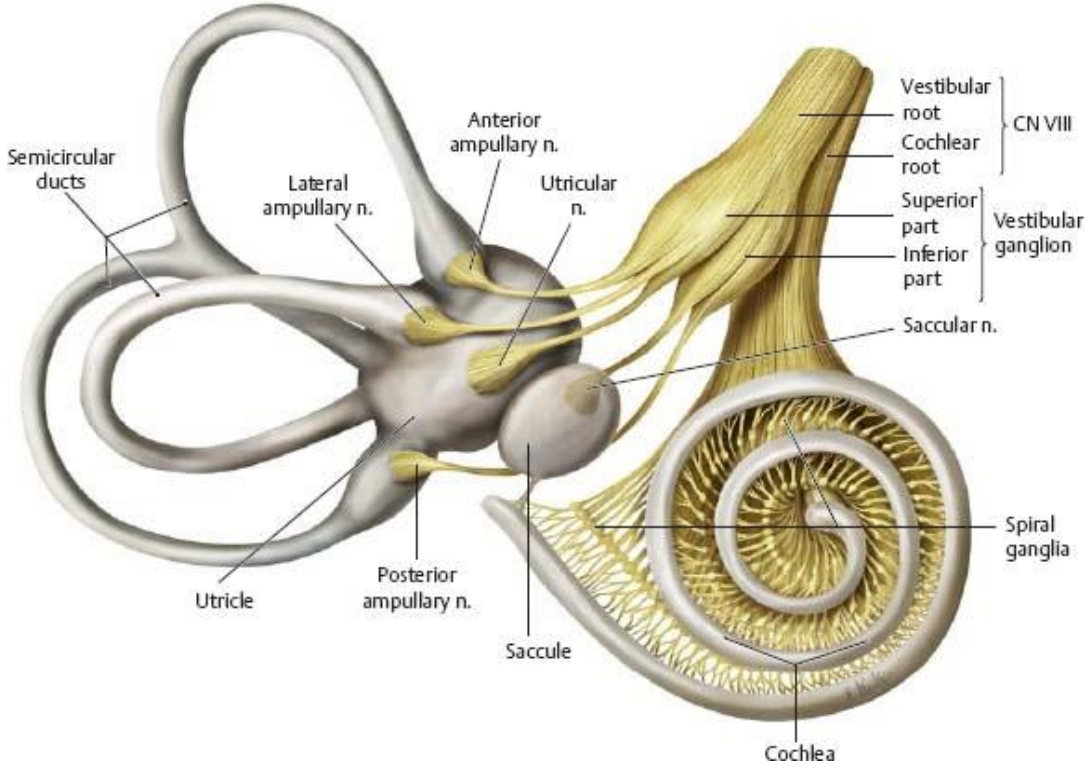


Increases muscle tone to anti-gravity muscles

Increases muscle tone to anti-gravity muscles


Fast phase of nystagmus is rightward
Slow drift of VOR is leftward
Increases muscle tone to anti-gravity muscles






**Types of vestibular neuritis**

- Superior vestibular nerve supplies: anterior canal, horizontal canal, utricle
- Inferior vestibular nerve supplies: posterior canal, saccule
- Complete vestibular neuritis = both superior and inferior nerve distribution affected.

# Vestibular Neuritis

## Prognosis

1. 22-50% of VN patients had normalization of caloric vestibular function by 6 weeks (Kim HA 2008, Buki 2016)

2. Recurrence rate is 2% (Neuhauser 2016)

3. 30-40% of VN patients have persistent dizziness (Neuhauser 2016). Possibly from anxiety, vestibular migraine, slow compensation

# Management of
# VN vs Stroke presenting with isolated vertigo

**Vestibular Neuritis**

- Benign prognosis, recovery usually in weeks to months
- Administration of corticosteroids, weak evidence supporting
- Administration of anti-herpetic, only if Ramsay-Hunt Syndrome
- Speed and possibly extent of improvement enhanced by VRT
- About 1/3 recover from the vestibular loss within 3 months, the remainder adapt well.

**Stroke with isolated vertigo**

- Possible harbinger of larger stroke to follow
- tPA or TNK (Tenecteplase) may be indicated
- Recovery is generally good if truly isoled vertigo presentation

Case: 38 year old woman has had 3-4 years of periodic dizziness worse in the past 6 months

- Dizziness characterized as either rocking/floating; not spinning
- Dizziness lasts 30 min to all day, varies in severity day to day
- Dizziness is present 30/30 days per month
- Under substantial work and personal/family stress
- Dizziness worsened by visual commotion, in crowds where people are moving about, with multi-tasking
- She is severely bothered by the dizziness and has sought multiple medical evaluations

EXAM: Normal neurological exam. No nystagmus. Gait and balance normal.
Brain MRI, CMP, CBC, B12, TSH, Audiogram, VNG, Ophthalmology, Neurology, Medical, Endocinology, Cardiology, ENT evaluations all normal.

# Persistent Postural Perceptual Dizziness (PPPD)

# Diagnostic Criteria for Persistent Postural Perceptual Dizziness

(paraphrased) (Staab et al. 2017)

A. Dizziness or unsteadiness or non-spinning vertigo on most days for $\geq$ 3 months (may be intermittent, wax and wane)
B. Persistent dizziness occurs without provocation but worsened by upright posture, head motion, exposure to moving visual stimuli or complex visual patterns
C. Symptoms associated with onset after a vestibular syndrome, acute or prolonged stress, medical illness
D. Symptoms cause significant distress and impairment (usually out of proportion to exam findings)
E. Not better accounted for by another disease

# Treating Persistent Postural Perceptual Dizziness

| Drug Class | Example, Dose Range | Adverse effects |
|---|---|---|
| Tricyclics | Nortriptyline 25-100 mg qd<br>Imipramine 25-75 mg qd | Wt gain, dry mouth, sedation, perspiration |
| SNRIs | Venlafaxine 25-150 mg qd<br>Duloxetine 30-90 mg qd | Insomnia, nausea, perspiration |
| SSRIs | Sertraline, citalopram, escitalopram, fluoxetine, paroxetine | Jittery feelings, weight gain, reduced libido |
| BZDP | Clonazepam 0.25-0.5 mg qAM | Sedation; dependency |

Case: 38 year old woman has had 3-4 years of periodic dizziness worse in the past 6 months

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Brain MRI, CMP, CBC, B12, TSH, Audiogram, VNG, Ophthalmology, Neurology, Medical, Endocinology, Cardiology, ENT evaluations all normal.

# Diagnostic Criteria for Vestibular Migraine

(paraphrased) (Lempert et al. 2012)

A. $\geq$ 5 episodes of moderate to severe vestibular symptoms (spontaneous vertigo, illusions of motion, visual vertigo, motion sensitivity, rocking)
B. Current or previous migraine by ICHD criteria
C. $\geq$ 50% of episodes have (i) unilateral pulsating headache or (ii) photophonia or phonophobia or (iii) visual aura
D. Not better accounted for by another disease

# Treating vestibular migraine

| Drug Class | Example, Dose Range | Adverse effects |
|---|---|---|
| Verapamil | Verapamil 120-360 mg qd | Constipation, GERD |
| Tricyclics | Nortriptyline 25-100 mg qd<br>Imipramine 25-75 mg qd | Wt gain, dry mouth, sedation, perspiration |
| SNRIs | Venlafaxine 25-150 mg qd<br>Duloxetine 30-90 mg qd | Insomnia, nausea, perspiration |
| Antiepileptic | Topiramate 25-100 mg qd | Cognitive SFx, sedation, dysgeusia |
| | Valproate 125-500 mg bid | Wt gain, hair loss, tremor |
| Beta blockers | Propranolol 80-180 mg qd | Fatigue, low BP/HR, palpitations |
| Clonazepam* | .25 -1 mg bid regularly, prn | Sedation; dependency |

Case: 37 year old woman has had 5 severe vertigo attacks over the past 2 years.

- The vertigo attacks: severe, random, last 2-6 hours
- Spells are sometimes preceded by a change in the pitch of tinnitus in the left ear and muffled hearing that can take a day to resolve.
- She has no symptoms once she has recovered but has had some reduction in hearing on the left side.

EXAM is normal except for reduced finger rub hearing on the left.

# Diagnostic criteria for Menière's disease

(Lopez-Escamez JA, et al. 2015)

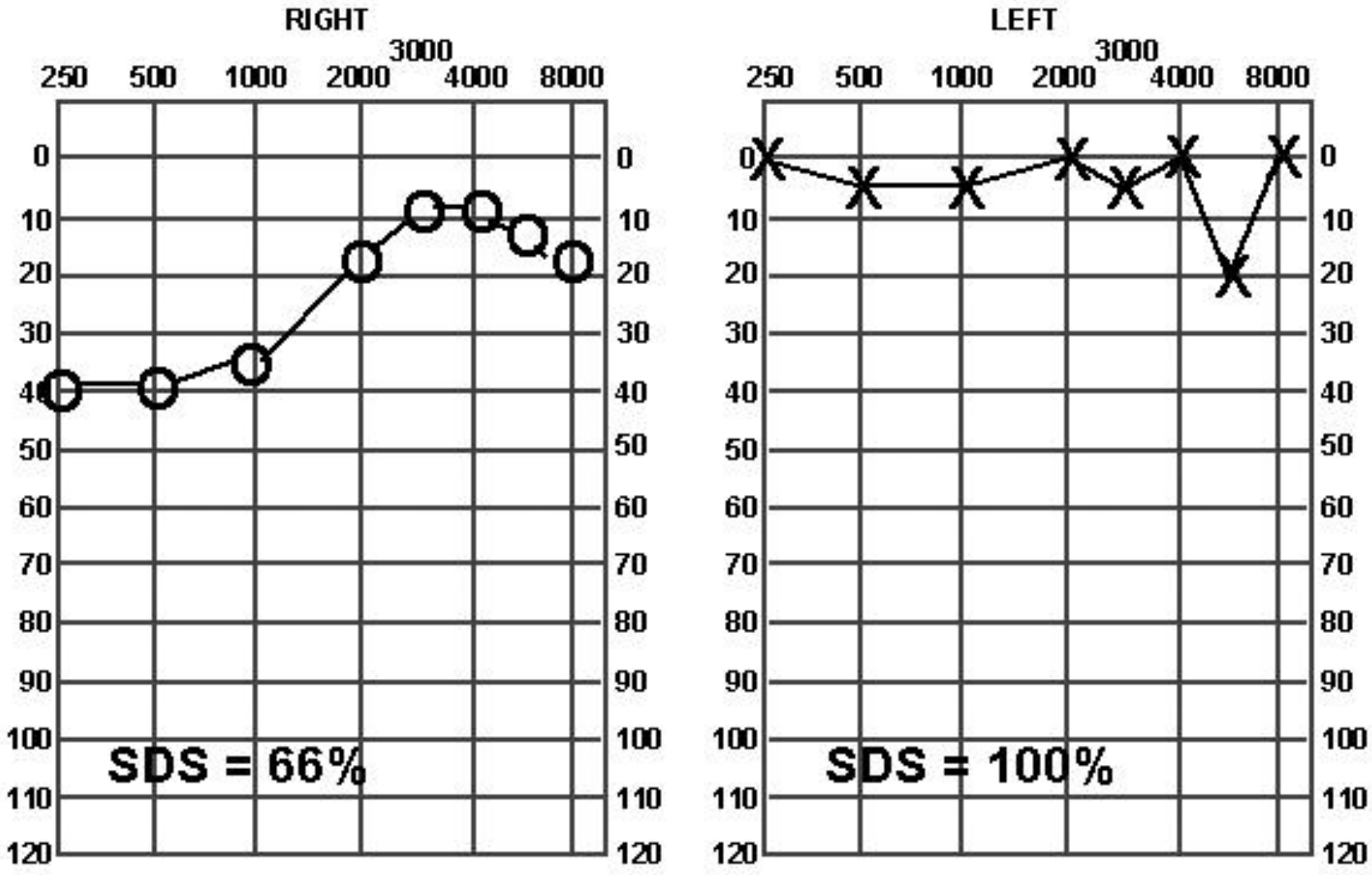
A. $\geq$ 2 spontaneous episodes of vertigo, 20 min to 12 hours

B. Low to medium frequency sensorineural hearing loss in on ear on an audiogram that the patient as identified to be the "affected ear" during at least one episode

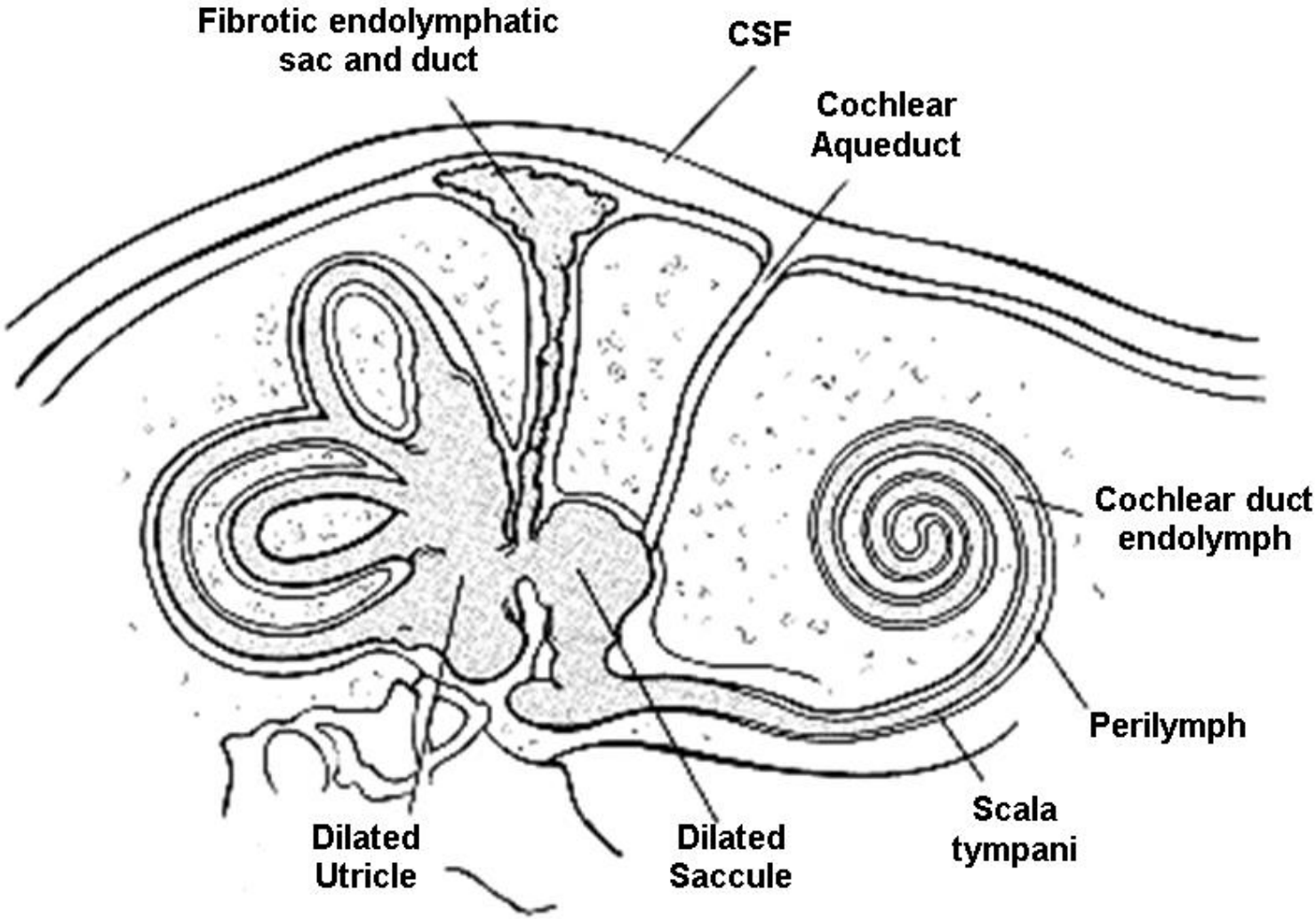
C. Fluctuating hearing, tinnitus, fullness in the affected ear

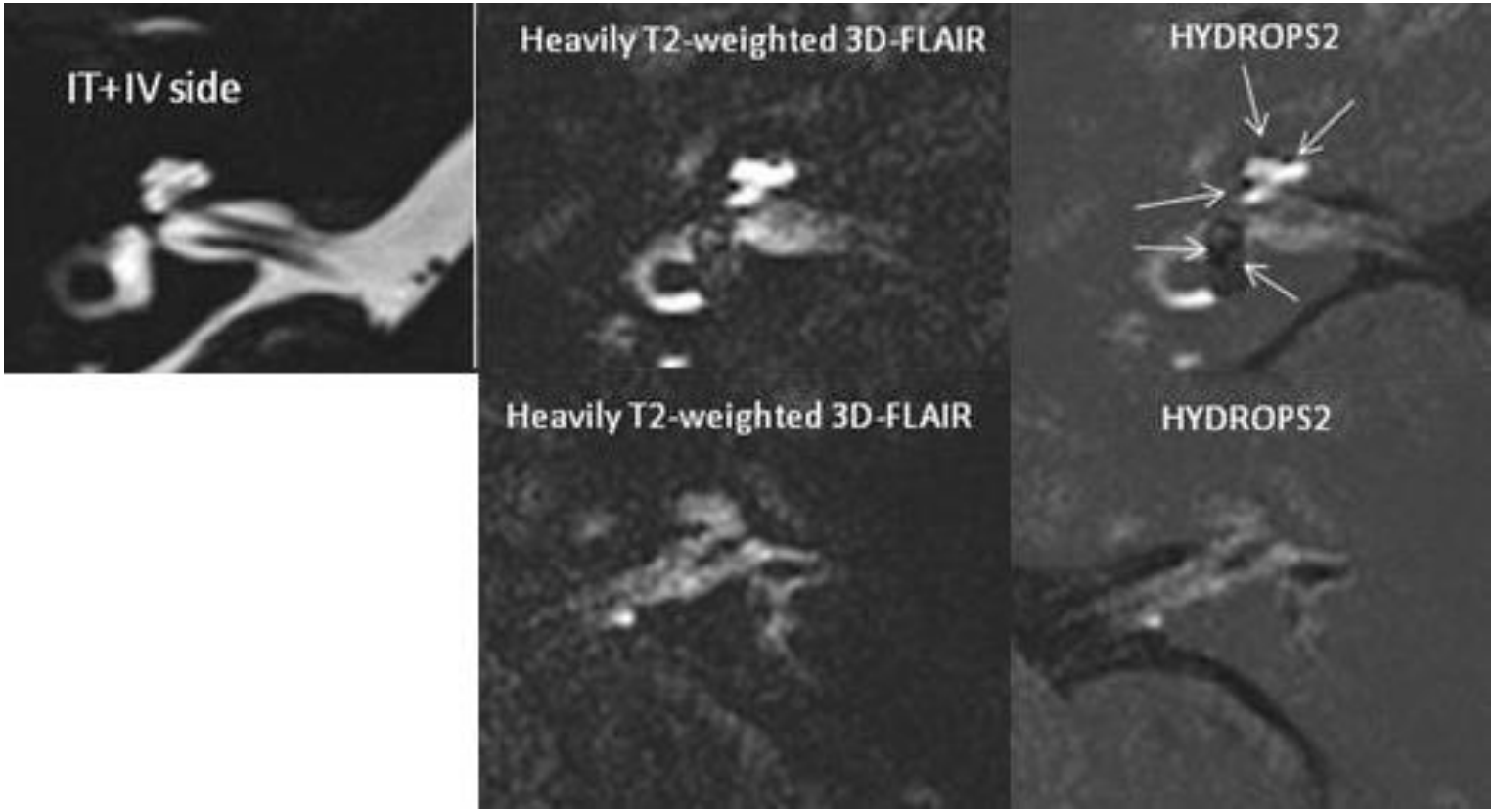
D. Not better accounted for by another vestibular diagnosis

# Right Sided Meniere's Disease



# Pathophysiology of Meniere's disease






Right ear

Left ear

MRI can detect endolymphatic hydrops seen in Meniere's using contasted or non-contrasted MRI:

- Heavily T2 weighted 3D FLAIR
- 3D Real Inversion Recovery after IT, IV Gad

.

(Naganawa & Nakashima 2014)

Case: 45 year old woman has had 6 months of:

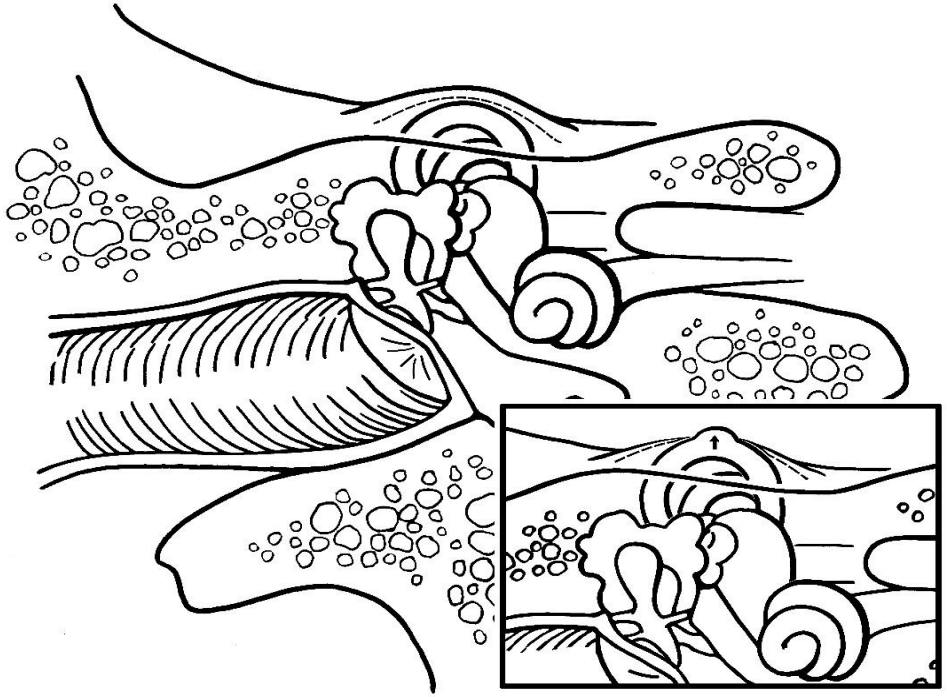
- Intermittent dizziness lasting minutes to an hour
- Dizziness is described as brief spins, rocking, swaying
- Associated brief bouncing vision, nausea, tinnitus
- Reports the dizziness is evoked by certain sound triggers such as low pitched sounds
- She can hear her own heart beat loudly (autophony)

EXAM: Normal neurological exam. No nystagmus.

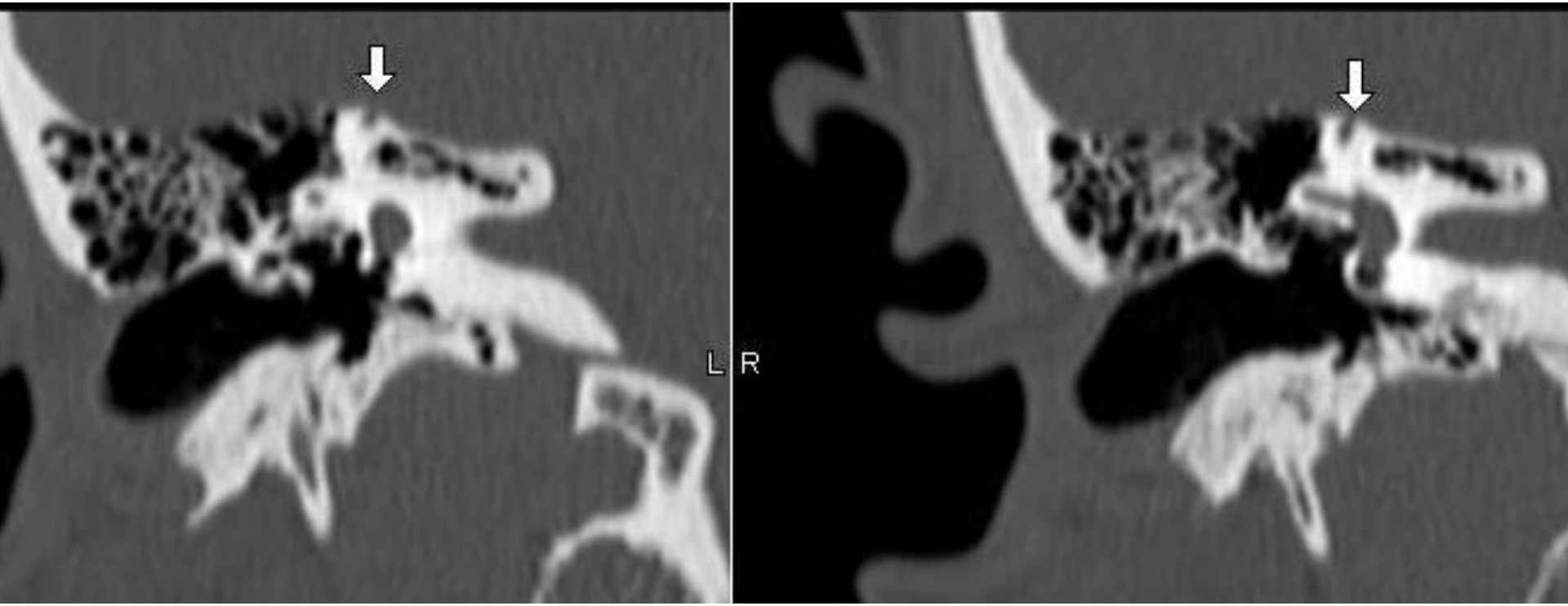
# Superior Canal Dehiscence Syndrome

In normal subjects, bone overlying the SSCC has an average thickness of 0.67 mm $\pm$ 0.38 mm (Carey 2000)

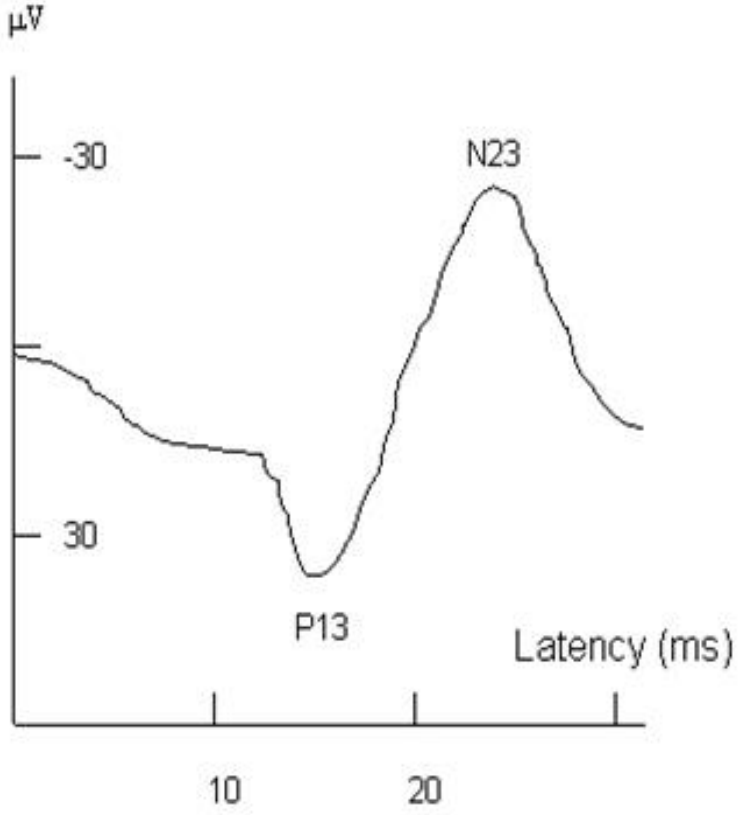
2% of the population has an abnormal thinning of the temporal bone though perhaps only ¼ of those actually have symptoms.  High resolution CT temporal bone should be 0.67 mm or thinner slices if possible.



# Superior Canal Dehiscence

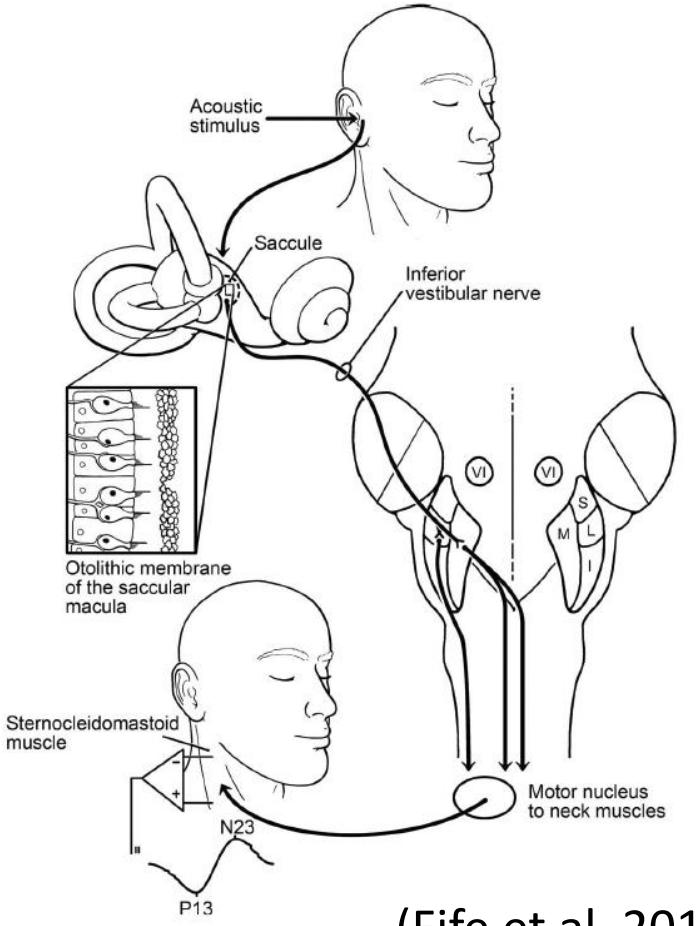


# Vestibular Evoked Myogenic Potentials (VEMP)

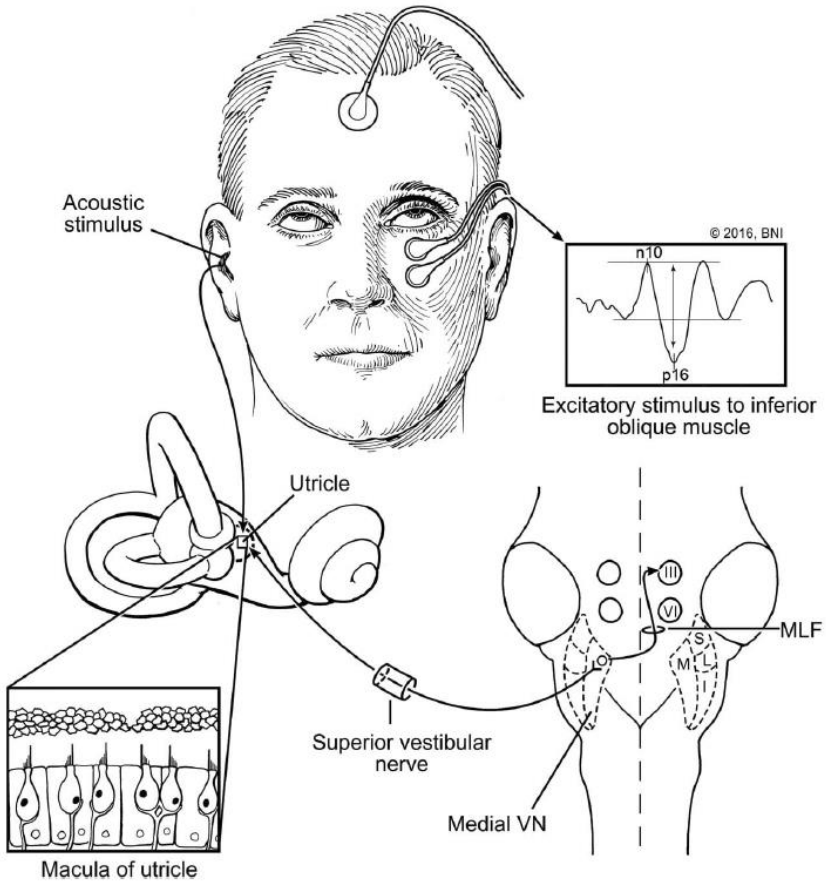



# Vestibular Evoked Myogenic Potentials

Cervical VEMP (cVEMP




Ocular VEMP (oVEMP)




(Fife et al. 2017)

| Description | Onset | Timing | Trigger | Assoc. Feature | Dx |
|---|---|---|---|---|---|
| Spin, whirl, tilt, float | Sudden | 5-60 sec | Tilt head, roll in bed | Occas. nausea | BPPV |
| Spin, whirl, tilt, float | Sudden | Days-wks | None | Worse with head movement; nystagmus; Abn HIT | Vestibular Neuritis |
| Spin, whirl, tilt, float | Sudden | 30 min – 12 hrs | None | Tinnitus, muffled hearing one side | Meniere |
| Near-syncope | Abrupt, usually upright | 2-20 min | Upright position | Pallor, sweating, low BP/HR with spell | Orthostasis |
| Spin, tilt, float, VID, motion sensitive | Varies | Varies | None | Migraine hx; exam normal | Vestibular Migraine |
| Rocking, floating | Often gradual | Often constant | None | Prior vertigo/distress; distressing | PPPD |

# Conclusions

1. Know your patient, what is their goal for the visit, their fear, their hope
2. Take a detailed history, focus on the symptom features
3. Avoid getting lost in the tests and prior opinions
4. On exam, understand basics of nystagmus
5. Know how to do Dix Hallpike and what BPPV nystagmus looks like
6. Learn how to do head impulse testing by practicing on normal patients
7. Try to always assess gait and describe it if abnormal
8. Learn the usual patterns of the various causes of vertigo and dizziness to improve your diagnostic skill set
9. When you find you missed a diagnosis, find out why, learn from it