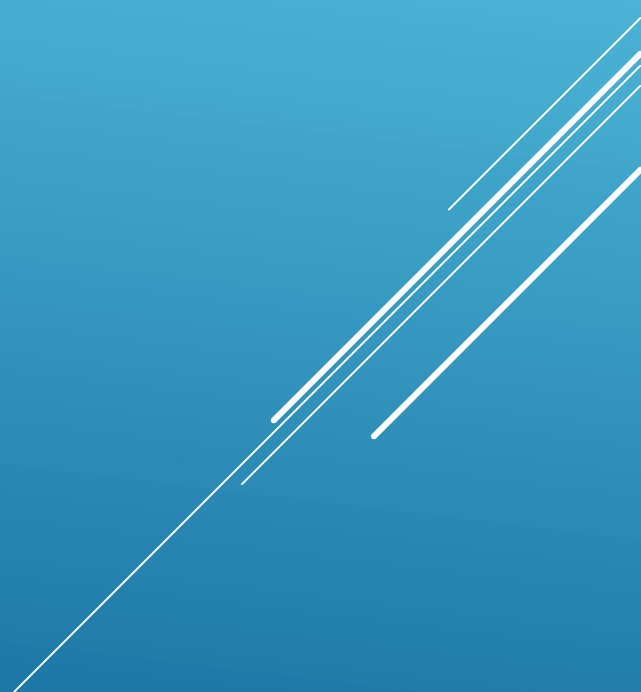
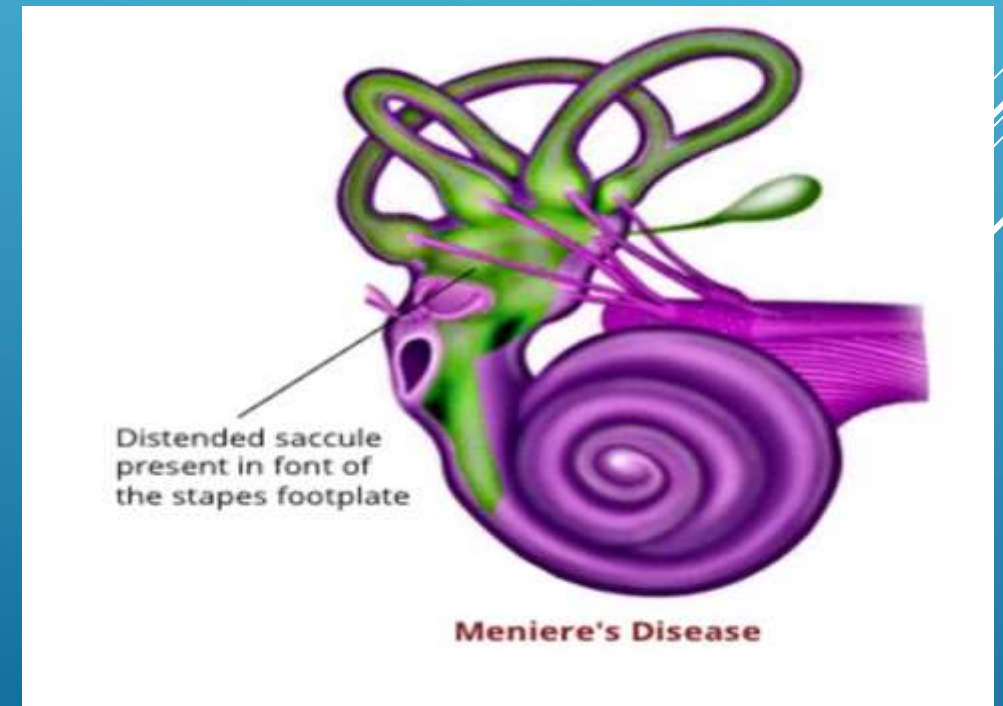
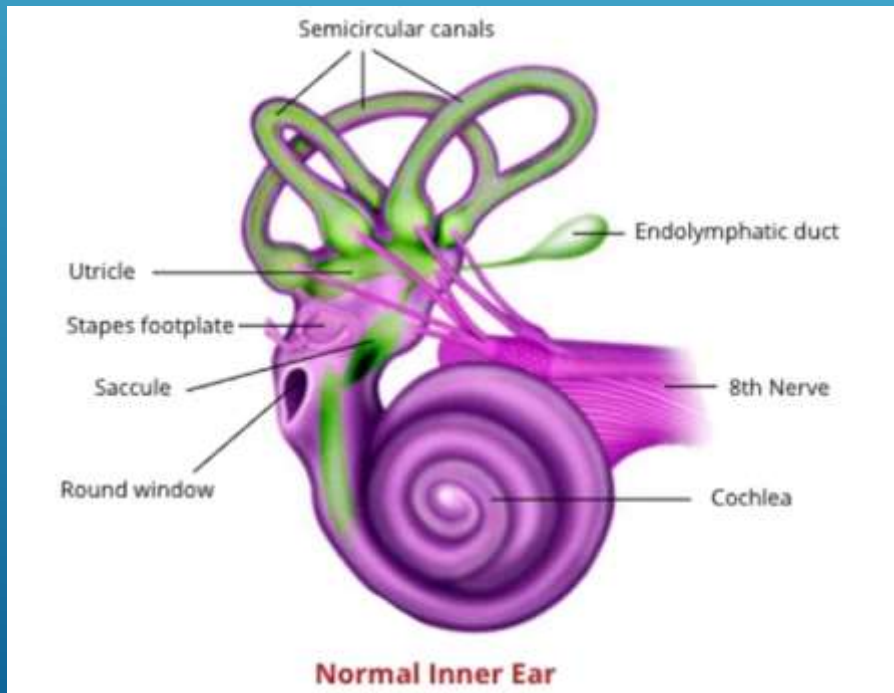


HEARING AND VESTIBULAR TESTING IN MENIÈRE'S DISEASE

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Inner ear spaces of the anterior and posterior labyrinth communicate in between and endolymphatic hydrops present in the Menière's disease usually affects both auditory and vestibular sensorial structures located in the two parts of the inner ear.



MD REMAINS A CLINICAL DIAGNOSIS BASED ON PATIENT-REPORTED SYMPTOMATOLOGY AND AUDIOMETRIC DATA

: Clinicians should obtain an audiogram when assessing a patient for the diagnosis of Me´nie`re's disease. Strong recommendation

Clinicians should diagnose definite or probable Me´nie`re's disease in patients presenting with 2 or more episodes of vertigo lasting 20 minutes to 12 hours (definite) or up to 24 hours (probable) and fluctuating or nonfluctuating sensorineural hearing loss, tinnitus, or pressure in the affected ear, when these symptoms are not better accounted for by another disorder.

AUDIOMETRY SHOULD INCLUDE PURE TON AIR CONDUCTION THRESHOLD ANY CONDUCTIVE COMPONENT OF THE HEARING LOSS (BONE CONDUCTION THRESHOLDS ESPECIALLY IN ATTACK, TYMPANOMETRY, ACOUSTIC REFLEX AND INCLUDING A MEASURE OF SPEECH RECOGNITION IN EACH EAR.

Diagnosis	Criteria
Certain	Definite Ménière's disease confirmed by histopathology
Definite Ménière's disease	≥ Two definitive spontaneous episodes of vertigo lasting 20 minutes to 12 hours + Audiometrically documented low- to medium-frequency sensorineural hearing loss in the affected ear on at least one occasion before, during or after one of the episodes of vertigo + Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear
Probable Ménière's disease	≥ Two episodes of vertigo or dizziness, each lasting 20 minutes to 24 hours + Fluctuating aural symptoms (hearing, tinnitus or fullness) in the reported ear

PURE TONE AUDIOMETRY

In early stages, lower frequencies SNHL is recorded and the curve is of rising type. When higher frequencies are involved, curve becomes tent or falling type later it becomes flat

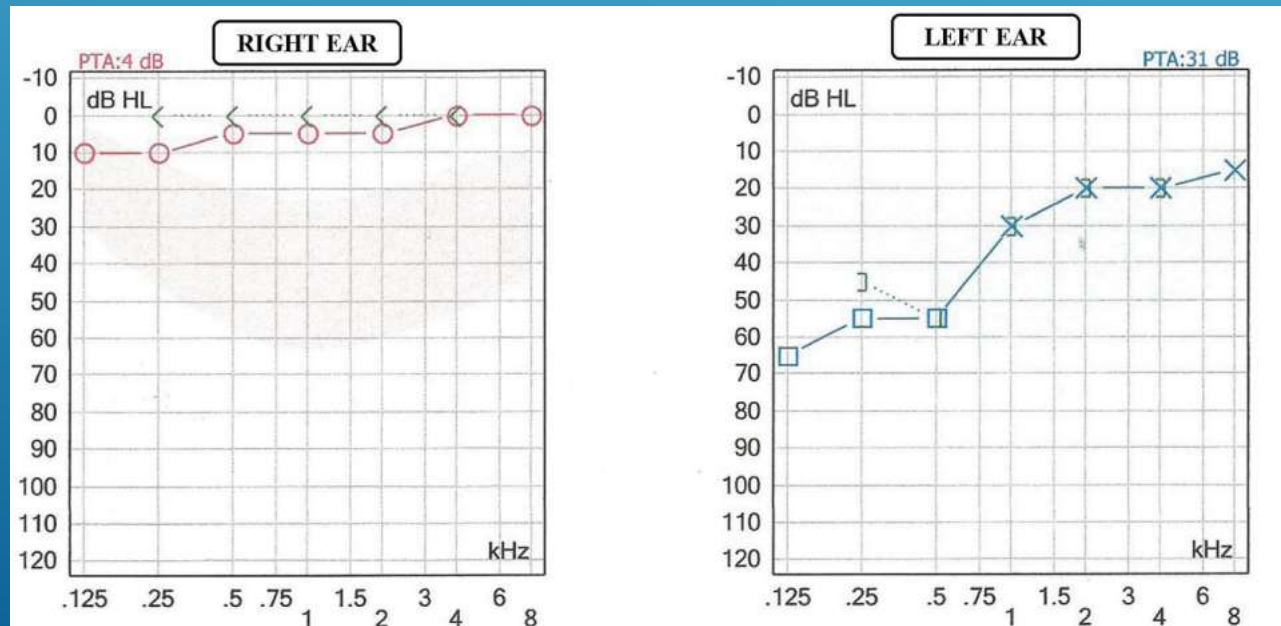
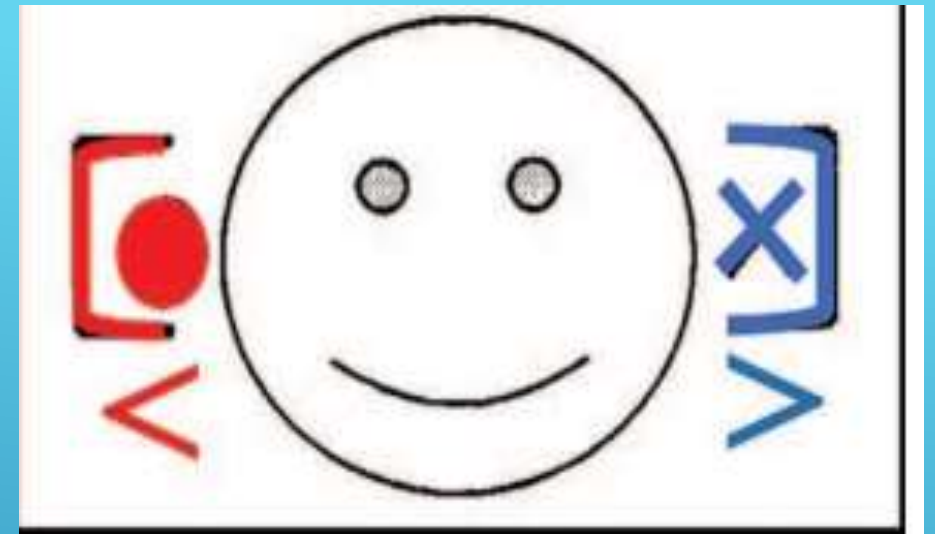


Figure 3. Low frequency sensorineural hearing loss in left ear.

STAGING OF MÉNIÈRE'S DISEASE

Pure tone average in dB in previous 6 months

1	≤25
2	26–40
3	41–70
4	>70

COCHLEAR CONDUCTIVE HEARING LOSS



Figure 4. Conductive (a) or mixed (b) hearing loss due to cochlear conductive hearing loss.

It is not unusual to have a conductive component of the hearing loss in Menière's disease **acute phase**—disturbances in endolymph metabolism lead to pressure variations at the round and oval window with secondary increases of impedances.

High impedances diminish air transmission of the sounds, (. In these cases, middle ear test (tympanometry and acoustically evoked stapedius reflex) shows no impairment of the middle ear as cause of the conductive component of the hearing loss.

cochlear sensorineural hearing loss (SNHL) accompanied by **recruitment**, a phenomenon of increased loudness perception—above an increase threshold, higher intensity sounds are as loud to the hearing impaired



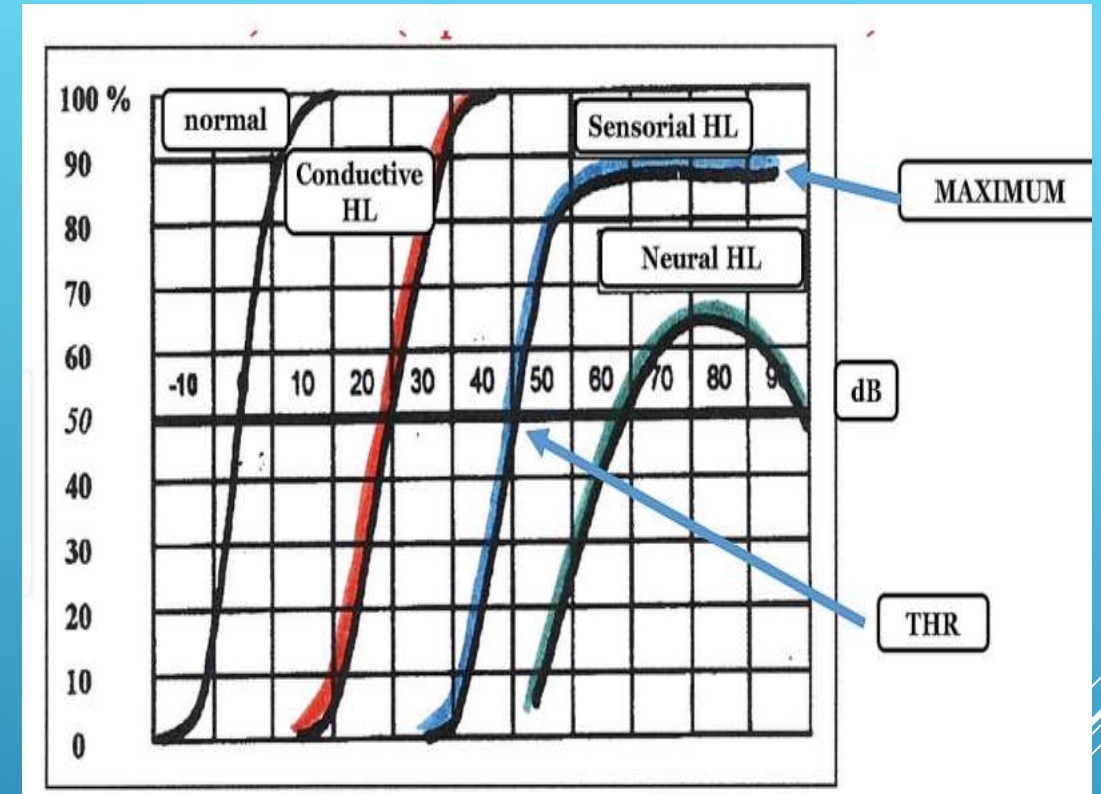
SPEECH AUDIOMETRY

speech audiometry. It is a more complex test, since evaluates the entire auditory pathway as hearing is a cortical process. Speech audiometry is also a subjective audiological test where the tested person has to repeat the heard stimuli—numbers, monosyllabic, disyllabic words, or sentences.

Speech Audiometry: Speech Discrimination score is 55–85% and discrimination ability is impaired during and immediately following an attack.

- Word recognition (discrimination) has been reported in some studies as poor as 32%.
- However, the degree of hearing loss seldom exceeds a 70 dB average

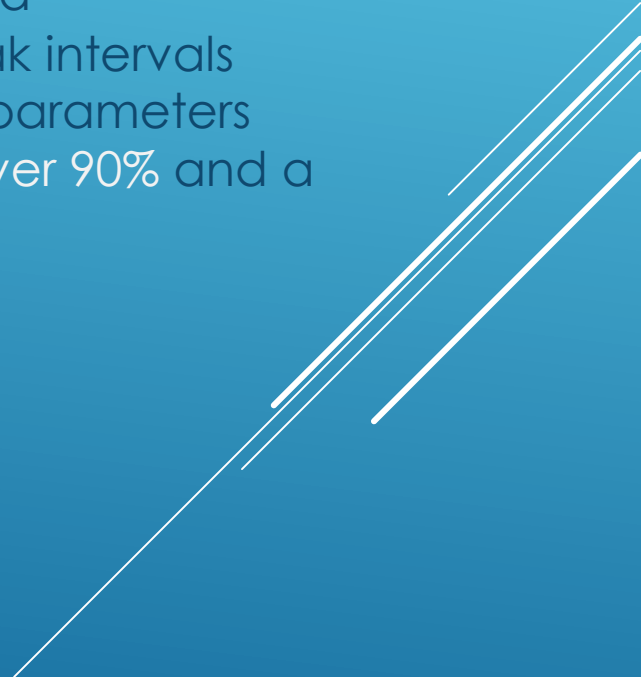
If in cochlear sensorineural hearing loss of other etiology there is a good correlation (± 7 dB) between pure tone and speech audiometry THR, in prolonged Menière's disease, some differences may appear. Another parameter used in interpretation of the speech audiometry is the maxim of intelligibility/discrimination. It represents the highest percentage of correct repeated stimuli the patient obtains. For normal hearing or conductive hearing loss persons, 100% intelligibility is reached. Sensorineural hearing loss induced distortions in audition which can limit the maximum of discrimination. Speech audiometry can draw attention on the estimated site of hearing loss, cochlear, or retrocochlear: in cochlear lesions, once the maximum score of discrimination is reached, it remains constant as higher intensities are tested. In retrocochlear sensorineural hearing loss an odd phenomenon occurs—as intensity increases, the patient understands less word (roll-over phenomenon).



. BRAINSTEM EVOKED RESPONSE AUDIOMETRY (BERA)/AUDITORY BRAINSTEM RESPONSE (ABR)

ABR—is an **objective electrophysiological** audiological method that allows recording of the electrical activity evoked by neural activity in the auditory pathways, from the cochlea to the brainstem (lateral lemniscus)

In Menière's disease patients, **BERA is mandatory** in order to rule out a retrocochlear etiology of the sensorineural hearing loss. Latencies, interpeak intervals and interaural differences of the latencies and interpeak intervals are the parameters used for this differential diagnosis . In general, ABR exhibits a **sensitivity of over 90%** and a **specificity of approximately 70–90%**.

Several white lines of varying lengths and slopes are positioned in the bottom right corner of the slide, creating a modern, abstract graphic element.

VESTIBULAR EVALUATION

- 1) recommended battery test for positive DIAGNOSIS
- 2) Evaluate degree of vestibular lesion which is present from the beginning of the Menière's d

Both vestibulo-ocular reflex (VOR) and vestibulospinal reflex (VSR) should be evaluated in disease.

Clinicians should not routinely order vestibular function testing or electrocochleography (ECoChG) to establish the diagnosis of Ménière's disease

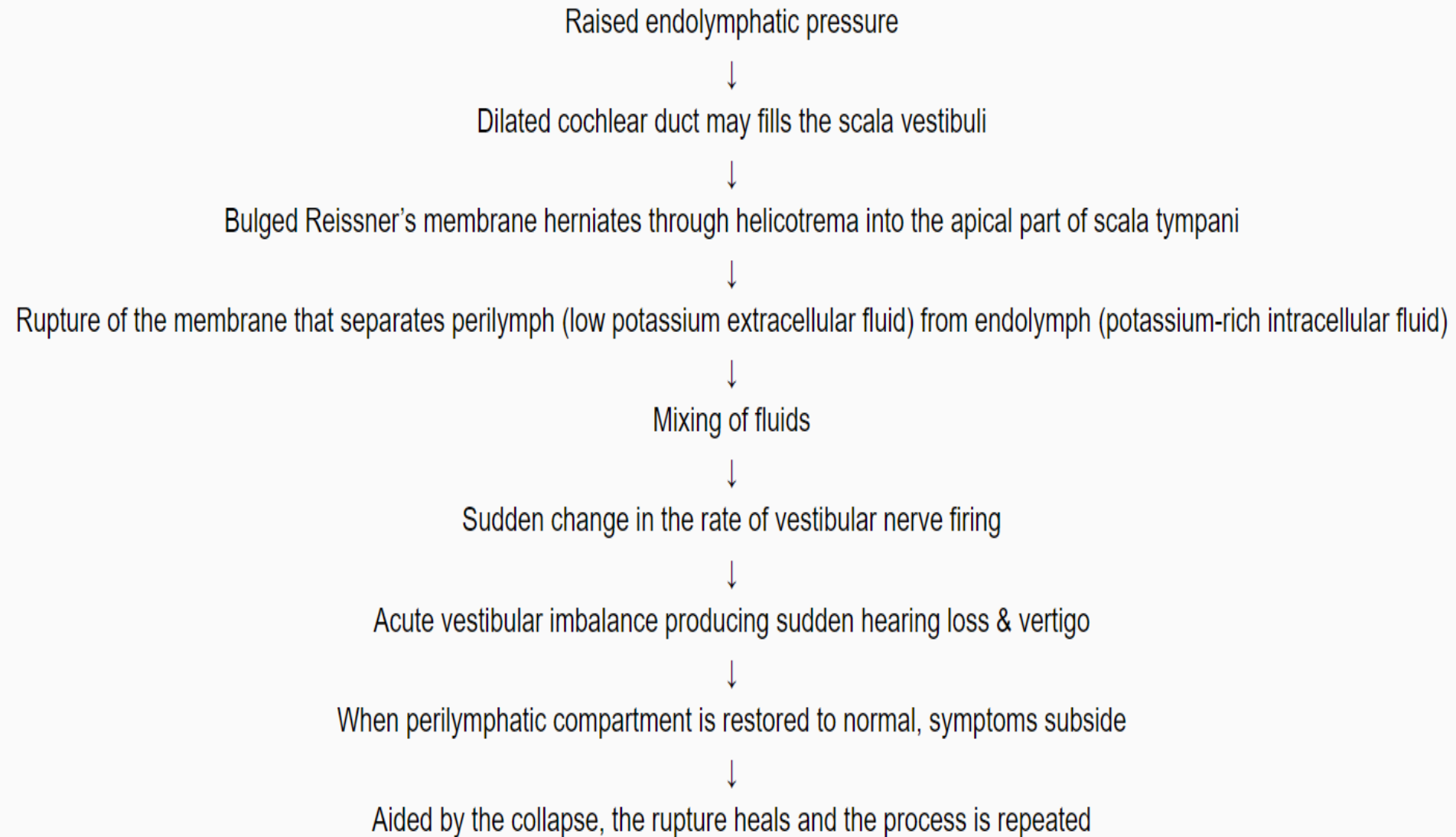
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In Menière's disease, results in ENG/VNG differ depending on the phase (acute/subacute, or chronic) and the duration of the disease.

At the beginning of an acute phase, due to the minor ruptures in the Reissner's membrane and an increase of potassium concentration in the endolymph, the vestibular sensorial epithelium in the affected ear is stimulated and the spontaneous nystagmus beats toward the Menière's ear). Soon after, due to constantly increasing of the potassium concentration, the vestibular hair cells are intoxicated and their function decrease. In this stage, spontaneous nystagmus changes its direction toward the healthy ear

Results of vestibular testing and ECoChG often fluctuate throughout the course of MD, and the degree of damage detected correlates poorly with patient-perceived disability

HERE ARE PATIENTS WHO MEET DIAGNOSTIC CRITERIA FOR MD BUT HAVE NORMAL VESTIBULAR TESTING RESULTS. THESE RESULTS DO NOT NECESSARILY RULE OUT MD. THERE ARE INSTANCES IN WHICH VESTIBULAR FUNCTION TESTING AND ECOCHG MAY BE HELPFUL IN EVALUATING AND MANAGING INDIVIDUAL PATIENTS WITH MD

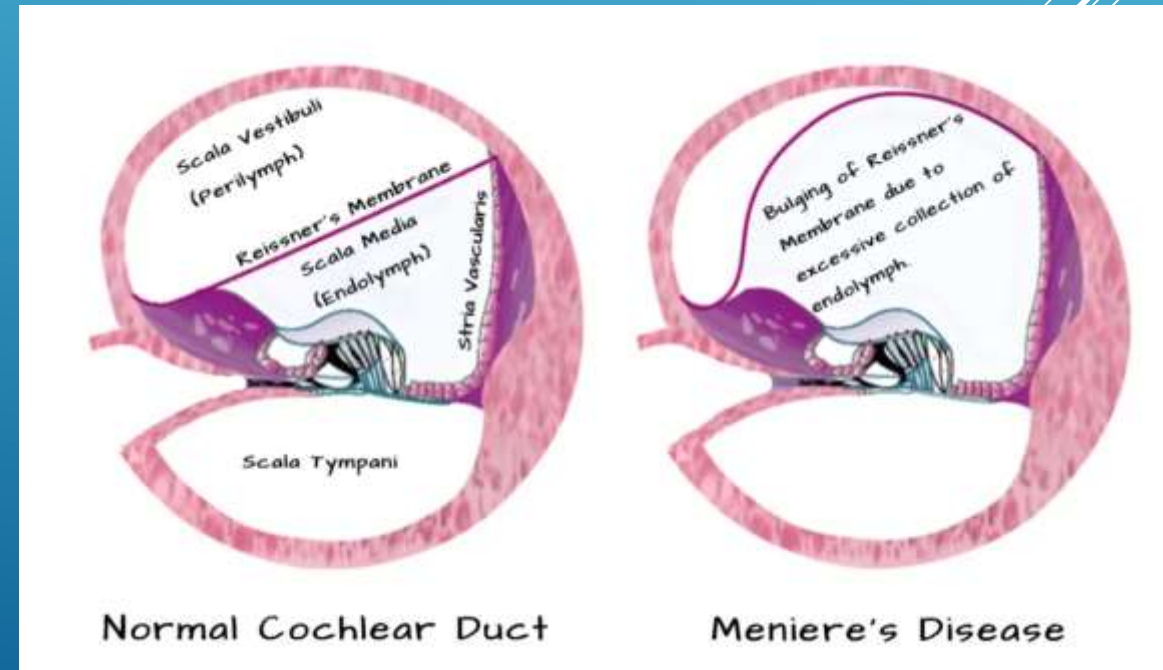


MÉNIÈRE'S DISEASE VARIANTS

- 1.**Tumarkin's otolithic crisis:** The patient gets feeling of being pushed on the ground suddenly without loss of consciousness. There is no vertigo or hearing loss. The underlying mechanism is deformation of the otolithic membrane of the utricle/sacculle as a result of changes in the endolymphatic pressure. T/m : Intratympanic injection of gentamicin
- 2.**Cochlear hydrops:** Symptoms and signs of only cochlear disease are present because there is block at the level of ductus reunions. Vertigo is not seen.
- 3.**Vestibular Hydrops:** Patient gets attacks of episodic vertigo while cochlear functions are normal.
- 4.**Lermoyez Syndrome:** It is Ménière's disease in reverse order. First hearing starts deteriorating followed by episodes of vertigo.
- 5.**Delayed hydrops:** caused by viruses, bacteria or spirochaetes.

ECOG

In endolymphatic hydrops, due to the **increased pressure in scala media, basilar membrane vibrates asymmetrical**. These changes of the traveling wave lead to several dysfunctions: distorted cochlear microphonics, enlargement of the summing potential and broadening of action potential. Magnitude of the AP compared with SP (SP/AP ratio) is increased in endolymphatic hydrops (>30%). The SP/AP amplitude ratio has 50–60% sensitivity in Ménière's disease diagnosis and 95% specificity in Refs.



. ELECTROCOCHLEOGRAPHY

Electrocochleography (ECochG) is **an objective** audiological test that measures the electrical potentials derived from the cochlear hair cells and the auditory nerve. ECochG measures the electrical responses of the cochlea and auditory nerve to acoustic stimulation.

An auditory stimulus is presented to the ear, and electrical responses are recorded, including the cochlear microphonic, the summing potential (SP) generated by cochlear hair cells, and the cochlear nerve action potential (AP), which is equivalent to wave I of the auditory brainstem response.

ECochG has historically been used in assessment of patients with presumed ELH. ELH is believed to generate abnormally large SP amplitudes relative to AP amplitudes by distending the basilar membrane toward the cochlear scala tympani.

1 Patients with a shorter duration of disease may not have developed cochlear changes that result in abnormal ECochG, therefore decreasing the sensitivity to detect pathology

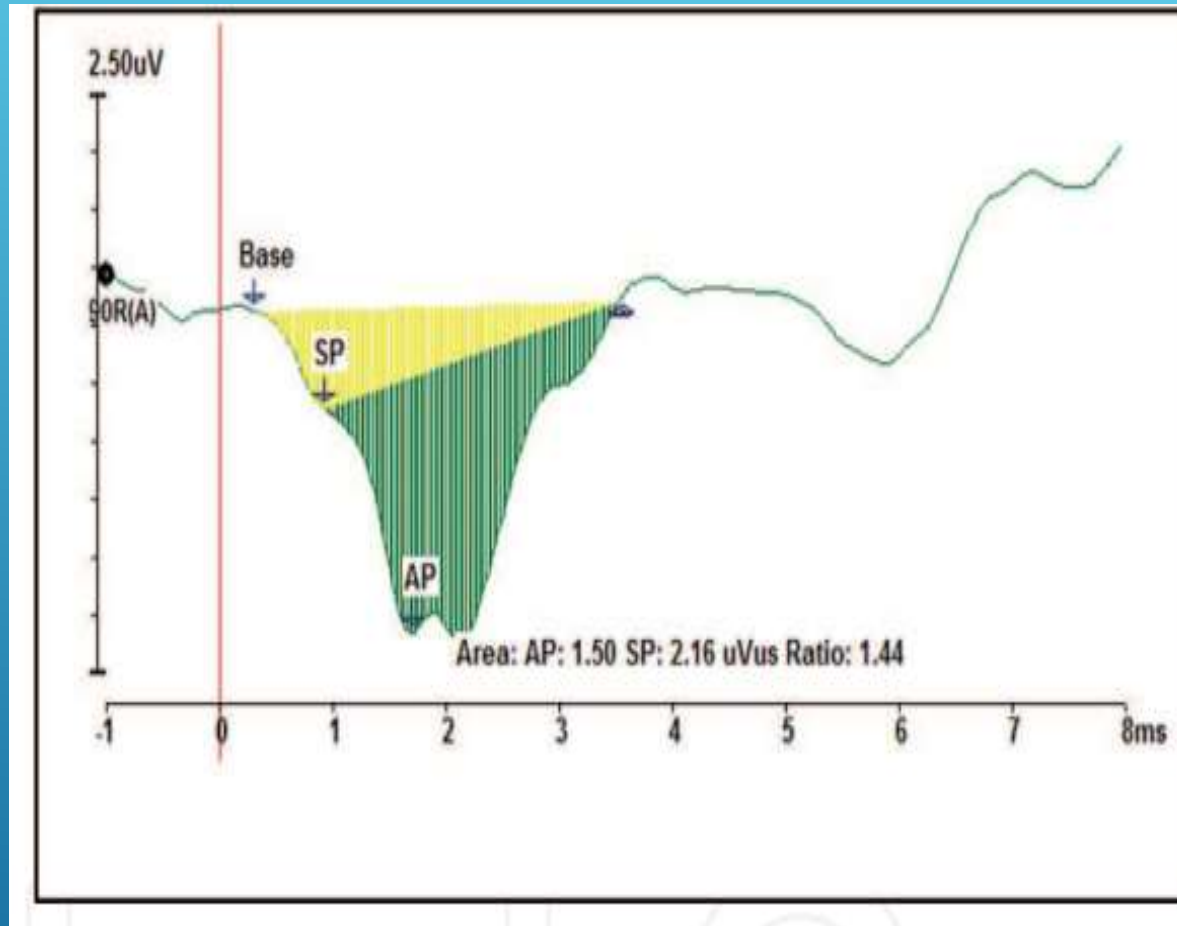
different stimuli and techniques for measuring ECochG responses create variations in measurements

Tone burst stimuli have demonstrated greater sensitivity in detecting cochlear hydrops comparative to click stimuli with transtympanic electrodes.

Other calculations and techniques to measure ECochG with the SP/AP amplitude and **area ratio** have also been suggested to improve the diagnostic accuracy for MD.,

elevation of the SP/AP amplitude and area ratio is not unique to patients with MD and may also be observed in the presence of **a third mobile window** of the inner ear, such as a superior semicircular canal dehiscence.

ECOG has limitation in patient with hearing loss



an area ratio (seems to be a more sensitive parameter for detecting endolymphatic hydrops [An increase of more than 2 of SP/AP area together with the increase of SP/AP amplitude ratio increases sensitivity and specificity in Menière's disease diagnosis to 92 and 83.9%, respectively]).

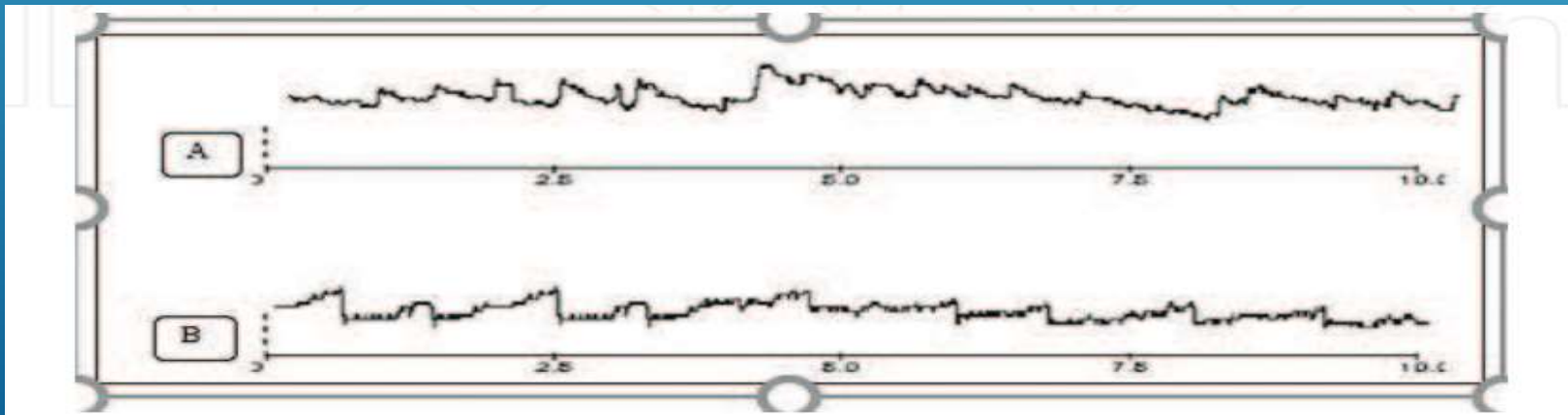
“ VESTIBULO-OCULAR REFLEX/VNG VHIT

Electronystagmography (ENG)/videonystagmography (VNG) 11

Electro- or videonystagmography allows quantification of the **nystagmus**, **as specific sign of vestibule-ocular reflex dysfunction**. ”

Nystagmus, as a conjugate movement of eyes with a slow and a fast phase provoked by vestibular asymmetry, reflects variations of the corneo-retinian potential during eyes movement. The **slow** phase is the effect of vestibular stimulation and its amplitude is proportional to the intensity of vestibular **stimulation**. The **fast phase** is central in origin and reflects only the reflex movement of the eyes to return to their normal position in the orbit. **The fast phase direction gives the nystagmus direction**

- ▶ In Menière's disease patients, results in ENG/VNG differ depending on the phase (acute, subacute, or chronic) and the duration of the disease.
- ▶ At the beginning of an acute phase, due to the minor ruptures in the Reissner's membrane and an increase of potassium concentration in the endolymph, the vestibular sensorial epithelium in the affected ear is stimulated and the spontaneous nystagmus beats toward the Menière's ear
- ▶ Soon after, due to constantly increasing of the potassium concentration, the vestibular hair cells are intoxicated and their function decrease. In this stage, spontaneous nystagmus changes its direction toward the healthy ear.



- ▶ In the next days after the acute spell of the Menière's disease results in rotatory and caloric test varies—either hypofunction in the affected ear. or symmetric functionality of the inner ears.
- ▶ The **absence of a fixed vestibular lesion** is the case in most of patients. In prolonged Menière's disease (long-term/chronic effect) usually patients' express caloric hypofunction of the affected ear (1/2—2/3 of patients) as VOR reflects the decreased input from the damaged

VNG involves recording eye movements during a battery of tests that assess vestibular function.

Caloric testing is one component of VNG and is best used to identify **unilateral peripheral vestibular hypofunction**. The caloric test provides ear-specific information with temperature-driven nonphysiologic **low-frequency** stimulation of the horizontal semicircular canal.

In cross-sectional studies and case series of patients with MD, 65% of patients have unilateral weakness noted on caloric testing. Thus, a substantial proportion of MD patients are expected to have normal results.

Normal caloric testing should not rule MD out



VHIT

The video head impulse test (HIT) evaluates as well semicircular canal function. Integrity of the VOR allows the tested subject to maintain sight fixed during **high-acceleration high-velocity head** .. Rotation is performed in each plane with an excitatory effect on each of the six semicircular canals. A positive HIT stands for complete lesion of the fibers connected with the tested semicircular canal. **In comparison with caloric testing, video HIT is abnormal in much more small numbers of Menière's disease patients, maybe because vestibular lesion is not complete**

vHIT is another vestibular test that uses high-frequency stimulation to assess function of all 6 semicircular canals independently. By using high-speed recordings of eye movements during and after high-velocity head impulses, vHIT yields a measure of vestibulo-ocular reflex gain (eg, ratio of slow-phase compensatory eye velocity to head velocity) as well as the pattern of corrective saccades that result from a canal functional deficit.

Discordant results between vHIT and caloric testing have been observed in multiple studies of patients with MD. In a series of 88 patients with definitive MD based on AAO-HNS 1995 criteria, 67% of patients had abnormal caloric testing, of which 45% had normal vHIT results

IN MD there is selective damage to type II hair cells that affected LOW FREQUENCY during caloric test and type I haircells not involve by menieres and normal VHIT produde

Currently there is insufficient evidence to support use of this pattern of discordant caloric testing and vHIT results as a diagnostic tool for MD

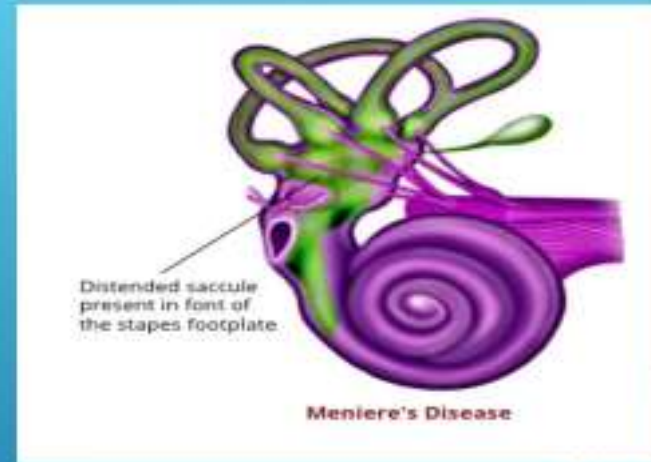
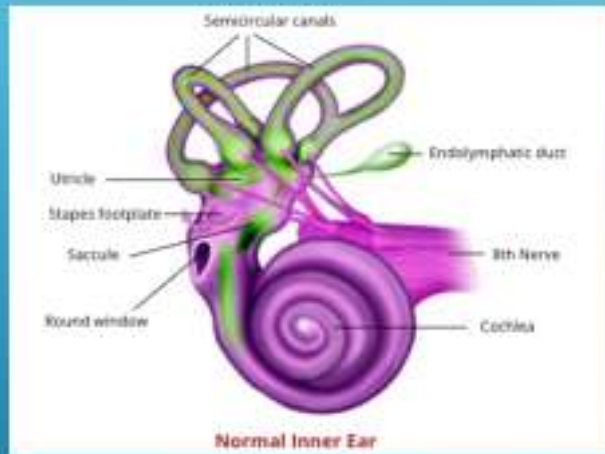
. However, these tests can be useful to identify a unilateral peripheral hypofunction, which may help guide further management, specifically in uncompensated cases
† MD results in selective damage to type II hair cells that affect the low-frequency response of the

BPPV IN MENIERES

- ▶ In between the acute spells, Menière's disease patients can experience positional vertigo, usually due to benign paroxysmal positional vertigo (BPPV).
- ▶ Disturbances in endolymph metabolism affect the function of the stria vascularis with secondary negative effects on the otolithic membrane. Still, BPPV is more frequently associated with vestibular migraine than Menière's disease

caloric asymmetry in MD results from alterations in inner ear fluid dynamics from ELH rather than from actual canal paresis.

Migraine	Ménière's Disease
Tinnitus: high-pitched	Tinnitus: low-pitched, roar
May have ear fullness (ache), phonophobia, and photophobia	Usually ear fullness or hearing loss
True spontaneous vertigo is rare; can occur for minutes	True spontaneous vertigo is common; can occur for hours
Short nap usually helps	Short naps usually do not help
Visual auras are common	Visual auras are uncommon
Motion sickness is common	Motion sickness is uncommon

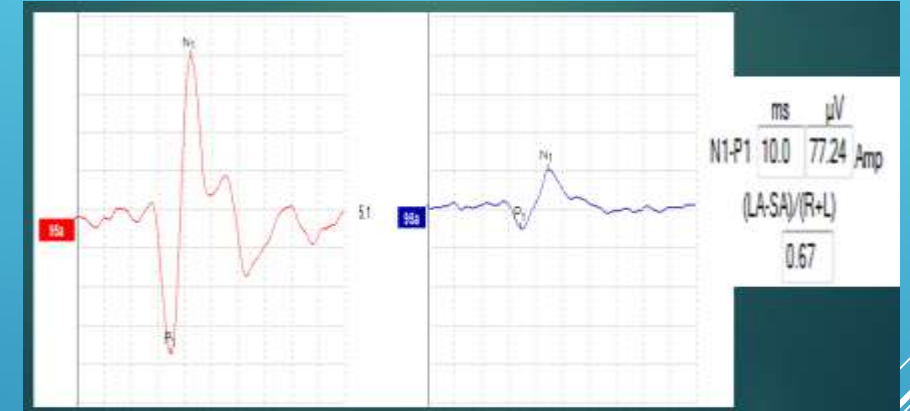


VESTIBULAR EVOKED MIOGENIC POTENTIALS

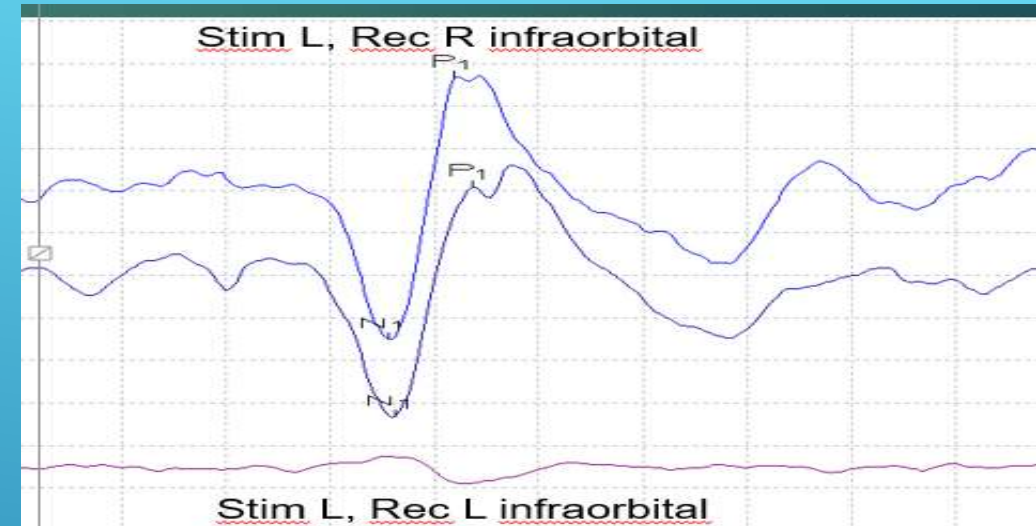
Vestibular evoked myogenic potentials (VEMPs) measure otolithic function.

- ▶ . In response to **loud sound stimulation** (95–97dB nHL), **saccular** vestibular sensorial epithelium generates activity in the inferior vestibular nerve and further in the vestibulospinal and vestibulo-ocular pathway.
- ▶ Action potential transmitted through the vestibulospinal pathway generates muscular responses in the effectors of the vestibulospinal (cervical muscles—cervical vestibular evoked myogenic potential: **cVEMP**) or vestibulo-ocular reflex (extraocular muscles—ocular vestibular evoked myogenic potential: **oVEMP**)

VEMPs may also have a role in prediction of evolving bilateral MD



- ▶ It also has been studied frequency tuning of cVEMP in endolymphatic hydrops and it appears that VEMP is recorded at higher frequencies and across broader frequency ranges than in normal inner ears due to changes in saccular resonance characteristics .
- ▶ These two changes (blunting and frequency shift of cVEMP) are greater as the Menière's disease has a longer evolution and greater severity in Additionally, over 20% of Menière's
- ▶ disease patients have abnormal cVEMP results in the non-affected ear in Ref.
- ▶ recommending VEMP as a predictor test for bilateral Menière's dis



In response to 500 Hz tone burst AC or BC

N1: ~10 msec

P1: ~15 msec

Contralateral response occurs slightly earlier and is larger than ipsilateral response (contralateral pathway is faster)

. Ocular VEMP Ocular VEMP (oVEMP) is a newer variant of VEMP which measures **saccular** function in response to very loud sound stimulation (about 120–130 dB SPL) or **utricle** function in response to vibrations applied to the cochlea.

Electrodes placed **below the orbit** record excitatory response in **the contralateral inferior oblique** muscle when in a flexed state by looking upward in **the** **contralateral inferior oblique** muscle. The first negative (excitatory) component of the oVEMP at a latency of about 10 ms is called n10. This n10 component most likely indicates the myogenic potentials of inferior oblique muscle.

in patients with early Menière's disease tested at attack, the contralateral oVEMP n10 is enhanced compared to measures in the same patients at quiescence. We speculate that this enhancement by Menière's disease attack could be due to mechanical changes in the labyrinth that enhance the sensitive response of utricular receptors to bone conduction vibrator stimulation. It seems that alterations in frequency tuning discussed in cVEMP are also present in sound-evoked oVEMP in Menière's disease patients in Ref.

CONTRALATERAL PATHWAY IS FASTER

patients present with **atypical symptoms** or when there is difficulty determining the affected ear, which may be helpful when considering **ablative interventions**.

, vestibular testing should be performed to assess the integrity of the vestibular system prior to completing an inner ear ablative procedure for MD treatment. **As bilateral peripheral vestibular** hypofunction has a significant impact on QOL and functioning,¹⁸⁷ full assessment of the vestibular function in the contralateral ear is warranted to determine the risks prior to permanent vestibular ablation. Vestibular testing may also be used to assess the effectiveness of ablative treatment.

, if patient symptoms are suggestive of other vestibular disorders, vestibular testing can be helpful to evaluate for these other causes. However, use of vestibular testing is best directed by patient history for appropriate interpretation of testing results and guidance of patient management

some **elderly patients** with long-standing and now recurrent MD may not clinically manifest frank vertigo symptoms but rather present with episodes of vestibular disturbance or “vague” dizziness.

