Meniere's Disease

Meniere's Disease, Synonyms: Idiopathic Endolymphatic Hydrops, Labyrinthine Hydrops

Meniere's disease is an idiopathic disorder affecting the membranous labyrinth of the inner ear. It was first described by a French physician, Prosper Meniere. The condition is characterized by an abnormal accumulation of fluid (endolymph) within the labyrinth, leading to increased hydraulic pressure in the endolymphatic system.

This pressure causes a tetrad of symptoms: (i) recurrent, spontaneous episodic vertigo, (ii) fluctuating or progressive sensorineural hearing loss, (iii) tinnitus, typically a low-pitched roaring sound, and (iv) a sensation of fullness or heaviness in the ear. Meniere's disease is often preceded by aural fullness and is frequently accompanied by nausea and vomiting.

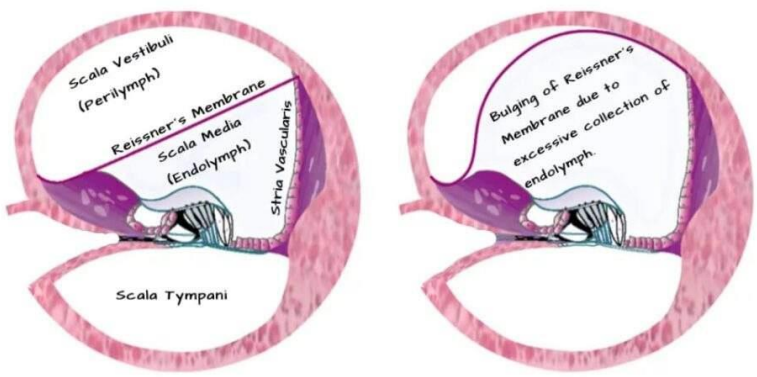
Meniere's Disease vs. Meniere's Syndrome

Meniere's disease is idiopathic in nature, meaning its exact cause is unknown. In contrast, Meniere's syndrome occurs secondary to other underlying conditions, such as autoimmune dysfunction, endocrine disorders, syphilis, trauma, electrolyte imbalances, and parasitic infections. These conditions interfere with the normal production or resorption of endolymph. This distinction is analogous to the differentiation between Bell's palsy and facial paralysis due to a known cause. Just as Bell's palsy is not diagnosed if the cause of facial paralysis is known, Meniere's disease is not diagnosed if the cause of vertigo is known.

Aetiology. The aetiology of Meniere's disease remains idiopathic. However, Meniere's syndrome can arise secondary to various disorders, including:

1. **Autoimmune Disorders:** Meniere's disease has been observed in some patients with lupus and rheumatoid arthritis.
2. **Genetic Factors:** Although most cases are sporadic, 5–15% of cases have a familial occurrence with an autosomal dominant pattern of inheritance.
3. **Viral Infections:** The benefit observed from acyclovir therapy in some patients suggests that the Herpes simplex virus might play a role.
4. **Allergic Reactions:** Certain food triggers may exacerbate symptoms, and the presence of IgG and IgA in cells of the endolymphatic sac supports this hypothesis.
5. **Sympathetic Overactivity:** This can lead to spasm of the internal auditory artery, resulting in deafness and vertigo.
6. **Hypothyroidism:** Approximately 4% of Meniere's disease cases are due to hypothyroidism, which may require thyroid replacement therapy.
7. **Endolymphatic Hydrops:** This can occur secondary to otic capsule diseases such as congenital or acquired syphilis, otosclerosis, Paget's disease, and post-stapedectomy cases.
8. **Exclusion of Other Conditions:** It is important to rule out transient ischemic attacks, endolymphatic sac tumours, vestibular schwannoma, vestibular migraine, vestibular paroxysmia, and recurrent unilateral vestibulopathy.

Pathophysiology. The underlying pathology in Meniere's disease involves the excessive accumulation of endolymph within the membranous labyrinth, leading to distention and rupture of the membranous labyrinth. This results in a mixing of endolymph and perilymph, which disrupts the ionic composition and affects the function of the inner ear. The mechanisms contributing to this excessive fluid accumulation include:



1. **Increased Production of Endolymph:** Endolymph is produced by the stria vascularis, and its overproduction can lead to hydrops.
2. **Reduced Absorption of Endolymph:** Normally, endolymph is absorbed in the endolymphatic sac. Any disruption in this process can contribute to fluid build-up.
3. **Combination of Both:** In some cases, both increased production and reduced absorption may occur, exacerbating the condition.

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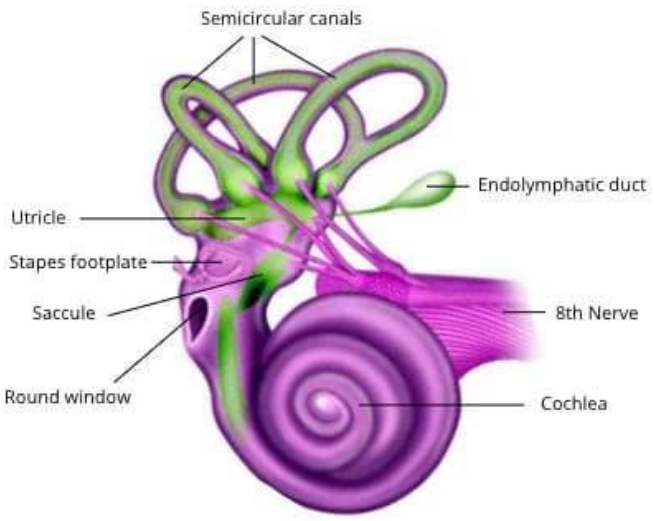
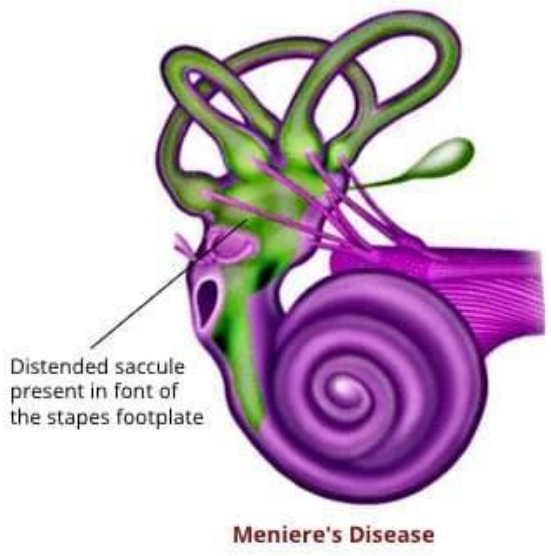
Flowchart showing Pathophysiology of Meniere's disease

Raised endolymphatic pressure.
↓
Dilated cochlear duct may fills the scala vestibuli
↓
Bulged Reissner's membrane herniates through helicotrema into the apical part of scala tympani
↓
Rupture of the membrane that separates perilymph (low potassium extracellular fluid) from endolymph (potassium-rich intracellular fluid)
↓
Mixing of fluids
↓
Sudden change in the rate of vestibular nerve firing
↓
Acute vestibular imbalance producing sudden hearing loss & vertigo.
↓
When perilymphatic compartment is restored to normal, symptoms subside.
↓
Aided by the collapse, the rupture heals, and the process is repeated.



Normal Inner Ear



Meniere's Disease

Clinical Features. The clinical presentation of Meniere's disease is characterized by a combination of vertigo, hearing loss, tinnitus, and aural fullness.

1. **Vertigo:** This is the hallmark symptom of Meniere's disease, characterized by sudden, episodic attacks of vertigo that may last from a few minutes to several hours. These episodes are often accompanied by nausea, vomiting, ataxia, nystagmus, abdominal cramps, diarrhea, cold sweats, pallor, and bradycardia. Importantly, the patient remains conscious during these attacks. Some patients may experience warning signs such as aural fullness, changes in tinnitus, or ear discomfort prior to an attack. During remission periods, patients may report hearing loss, a sense of imbalance, or be asymptomatic. For diagnosis, at least two definitive episodes of vertigo lasting for at least 20 minutes are required.

Tullio phenomenon: Attacks of vertigo on hearing loud noise probably due to the distended saccule lying against the stapes footplate. However, this phenomenon is also noticed when there are three functioning windows in the ear, e.g., a fenestration of horizontal canal in the presence of mobile stapes.

Table : Grading of severity of Vertigo

0	No vertigo	–
1	Mild attack	Brief vertigo < 20 minutes and/or vague sense of disequilibrium lasting < 2 hours.
2	Moderate attack	Vertigo for 20–60 minutes and/or disequilibrium lasting > 2 hours hampering daily activity.
3	Severe attack	Vertigo lasting >1hour, associated nausea and vomiting, with or without disequilibrium lasting > 2 hours hampering daily activity.
4	Extreme attack	Vertigo > 1 hour with nausea, vomiting and severe disequilibrium requiring bed rest throughout day.

2. **Fluctuating Hearing Loss:** Hearing loss may accompany or precede vertigo. Initially, hearing may be normal during remission phases, but repeated episodes can lead to permanent deafness. Hearing loss typically affects low-frequency sounds, and music may seem disharmonious. Patients may also experience recruitment, where sounds become intolerably loud at certain intensities, making them poor candidates for hearing aids. Diplacusis, where a single sound is perceived differently in each ear, is also common.
3. **Tinnitus:** This is usually described as a low-pitched, ocean-like roaring, whistling, or hissing sound. Changes in the intensity and pitch of tinnitus may serve as a warning sign of an impending vertigo attack.
4. **Age and Sex:** Meniere's disease typically affects individuals in their fourth to sixth decades of life, with an equal prevalence in males and females

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5. **Spontaneous Nystagmus:** The nystagmus associated with Meniere's disease progresses through three phases:

- Irritative Phase: Nystagmus beats toward the affected ear in a horizontal or horizontal-torsional direction.
- Paretic Phase: Nystagmus beats away from the affected ear.
- Recovery Phase: Nystagmus beats toward the affected side as peripheral vestibular function recovers.

Clinical Examination

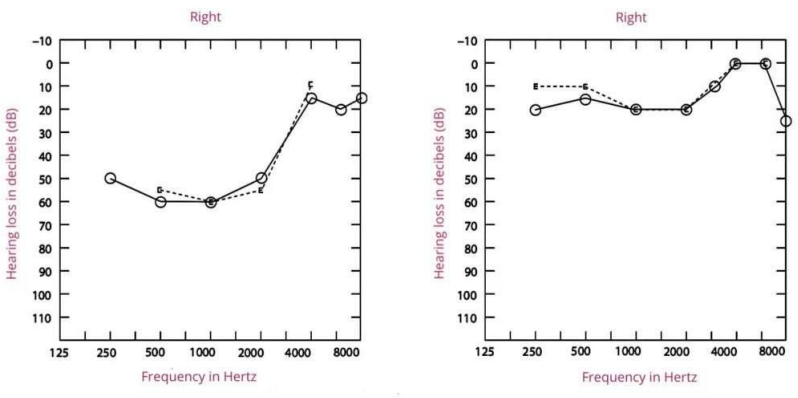
1. Otoscopy: The findings are usually normal, but pneumo-otoscopy of the affected ear may elicit symptoms.
2. Nystagmus: The quick component of nystagmus is directed toward the normal ear.
3. Tuning Fork Tests: These tests often reveal sensorineural hearing loss:

- Weber Test: Sound is lateralized to the better ear.
- Rinne Test: The test is positive, indicating that air conduction is better than bone conduction.
- Absolute Bone Conduction: This is reduced in the affected ear.

4. Neurologic Examination: This is essential to differentiate Meniere's disease from other conditions that can present with vertigo, such as stroke, migraine, or brainstem compression.

Investigations

1. **Pure Tone Audiometry:** In the early stages of Meniere's disease, low-frequency sensorineural hearing loss is recorded, producing a rising curve on the audiogram. As higher frequencies become involved, the curve may become tent-shaped or falling, eventually flattening in advanced stages.



Audiogram in Ménière's disease.
(a) Audiogram showing 60 dB low-frequency sensorineural hearing loss — the rising curve.
(b) Audiogram after 4 months of treatment with a rigorous low-sodium diet shows normal pure-tone thresholds.

2. **Speech Audiometry:** The speech discrimination score ranges from 55% to 85%, with discrimination ability impaired during and immediately after an attack.

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3. **Electrocochleography:** This electrophysiological test measures the ratio of summating potential (SP) to action potential (AP) through either transtympanic or extratympanic methods. In Meniere's disease, the SP/AP ratio is elevated due to basilar membrane distention toward the scala tympani. Normally, the SP/AP ratio is 30%; in Meniere's disease, it is raised.
4. **Caloric Test:** This test often shows a decreased response and canal paresis on the diseased side in 75% of cases. In some instances, directional preponderance to the healthy side or a combination of canal paresis and directional preponderance may be observed.
5. **Glycerol Test:** This test involves the oral administration of glycerol, a dehydrating agent, along with an equal amount of water and lemon juice. This reduces endolymphatic pressure and may lead to improvements in hearing. An improvement of 10 dB in two or more adjacent octaves on an audiogram, a gain of 10% in speech discrimination score within 1–2 hours, and an improvement in tinnitus and aural fullness after glycerol ingestion are considered positive results. The test has both diagnostic and prognostic value and is often combined with electrocochleography.
6. **Posterior Fossa MRI:** This imaging study is essential to rule out acoustic neuromas or other cerebellopontine angle lesions in the absence of contraindications to MRI.
7. **Delayed Gadolinium-Enhanced MRI:** Intravenous gadolinium contrast is administered four hours prior to MRI, or intratympanic gadolinium is given 24 hours prior to MRI. This imaging technique serves as a biomarker to differentiate between the perilymphatic and endolymphatic spaces, as gadolinium loads into the perilymphatic space without entering the endolymph in a healthy inner ear.

Table : **Differences between cochlear and retro-cochlear lesion**

	Normal	Cochlear lesion	Retrocochlear lesion
Pure tone audiogram	Within normal limits	SNHL	SNHL
BERA	Normal interval between wave I & V	Normal interval between wave I & V	Wave V delayed or absent
Recruitment	Not present	Present	Not present.
SISI score	0–15%	Over 70%	0–20%
Threshold tone decay test	0–15 dB	Less than 25 dB	Above 25 dB
Speech discrimination score	90–100%	Below 90%	Very poor
Roll over phenomenon	Not present	Not present	Present
Stapedial reflex	Present	Present	Not present
Stapedial reflex decay	Within normal limits	Within normal limits	Abnormal

Meniere's Disease Variants

Meniere's disease can present in various forms, often classified based on the predominant symptoms and the specific areas of the inner ear affected. Understanding these variants is crucial for accurate diagnosis and treatment. Below are the common variants of Meniere's disease:

Tumarkin's Otolithic Crisis

1. Tumarkin's Otolithic Crisis (Utricular Crisis)

- Clinical Presentation: Patients experience sudden episodes of being forcefully pushed to the ground without warning, often described as a "drop attack." These episodes occur without loss of consciousness, vertigo, or hearing loss, distinguishing them from typical Meniere's disease attacks.
- Pathophysiology: The underlying mechanism involves the deformation of the otolithic membrane in the utricle or saccule, caused by fluctuations in endolymphatic pressure. This deformation disrupts the normal functioning of the otolith organs, leading to the sudden loss of postural control.
- Treatment: Intratympanic injection of gentamicin is commonly used. Gentamicin is vestibulotoxic and helps in reducing the frequency of these crises by partially ablating the vestibular function.

2. Cochlear Hydrops

- Clinical Presentation: This variant involves symptoms solely related to cochlear dysfunction, such as hearing loss and tinnitus, without vertigo. The absence of vertigo differentiates cochlear hydrops from classic Meniere's disease.
- Pathophysiology: The condition is caused by a blockage at the level of the ductus reuniens, which connects the cochlear duct and the saccule. This blockage prevents the spread of endolymphatic hydrops from the cochlea to the vestibular system, resulting in isolated cochlear symptoms.

3. Vestibular Hydrops

- Clinical Presentation: Patients experience episodic vertigo without accompanying cochlear symptoms like hearing loss or tinnitus. This variant highlights the exclusive involvement of the vestibular system.
- Pathophysiology: Similar to other forms of hydrops, vestibular hydrops result from endolymphatic pressure changes, but these affect only the vestibular apparatus. The cochlear function remains unaffected, leading to vertigo as the sole symptom.

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4. Lermoyez Syndrome

- Clinical Presentation: Lermoyez syndrome is essentially Meniere's disease with a reversed sequence of symptom onset. Patients first notice a deterioration in hearing, which is later followed by episodes of vertigo. In some cases, vertigo is followed by a temporary improvement in hearing.
- Pathophysiology: The mechanisms are similar to those of classic Meniere's disease, involving fluctuating endolymphatic pressure. However, the order in which symptoms appear is reversed, possibly due to differing rates of fluid buildup and pressure changes within the cochlear and vestibular systems.

5. Delayed Hydrops

- Clinical Presentation: Delayed hydrops may present after a period of normal inner ear function, often following an episode of viral, bacterial, or spirochetal infection. Patients may experience hearing loss, tinnitus, and vertigo, sometimes years after the initial infection.
- Pathophysiology: This variant is thought to result from latent or chronic infections that cause inflammation and subsequent endolymphatic hydrops. The delayed onset of symptoms may be due to a slow, progressive accumulation of endolymph or scarring in the inner ear structures.

DIAGNOSIS OF MENIERE'S DISEASE.

The most recent guidelines for the diagnosis of Ménière's disease were issued in 2015 by the Classification Committee of the Bárány Society, The Japan Society for Equilibrium Research, the European Academy of Otology and Neurotology (EAONO), the Equilibrium Committee of the American Academy of Otolaryngology– Head and Neck Surgery (AAO–HNS) and the Korean Balance Society. It is as follows.

Table : Diagnostic criteria for Meniere's disease (other causes excluded)

Diagnosis	Criteria
Certain	Definite Meniere's disease confirmed by histopathology.
Definite Meniere'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 Meniere'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
Possible Meniere's disease	Meniere's type episodic vertigo without documented hearing loss, or sensorineural hearing loss, fluctuating or fixed, with vertigo but without definitive episodes. Other causes excluded

Hearing loss:

The staging system is essential in the diagnosis of certain, definite and probable Meniere's disease. It is based on the average of pure tone thresholds at 0.5, 1, 2 and 3 kHz (rounded to the nearest whole) of the worst audiogram during 6 months before start of treatment.

Table: Staging of ménière's disease	
	Pure tone average in dB in previous 6 months
1	≤25
2	26–40
3	41–70
4	>70

Treatment of Meniere's Disease.

General Advice:

1. Relief of Anxiety: Patients are encouraged to engage in relaxation exercises and yoga to manage anxiety, which can exacerbate symptoms.
2. Smoking Cessation: Nicotine can cause vasospasm, which may worsen Meniere's symptoms. Patients are advised to quit smoking.
3. Dietary Restrictions: A diet that is free of salt and caffeine is recommended to help control fluid retention and reduce symptoms.
4. Activity Modifications: Patients should avoid activities that require precise balance, such as flying, underwater diving, or working at heights.
5. Stress and Hydration Management: Patients are advised to manage stress levels and avoid excessive water intake.

Management of Acute Attacks

1. Reassurance and Rest: During an acute attack, patients should be reassured and advised to rest with their head elevated and minimize head movements.
2. Vestibulosuppressants: Medications such as dimenhydrinate (Dramamine), promethazine theoclate (Avomine), or prochlorperazine (Stemetil) are used to dull the brain's response to inner ear signals. Diazepam (Valium or Calmpose) 5–10 mg intravenously can be administered to suppress the activity of the medial vestibular nucleus.

Management of Chronic Phase

1. Vestibulosuppressants and Anti-nausea Medications: Cinnarizine and prochlorperazine (Stemetil) are commonly prescribed. A regimen of Stemetil 10 mg TDS orally for two months, followed by 5 mg TDS for one month, is typical.
2. Diuretics: If vestibulosuppressants and vasodilators are ineffective, furosemide 40 mg on alternate days with potassium supplements, or thiazide diuretics (hydrochlorothiazide) 12.5 mg daily, may be prescribed to control recurrent episodes.
3. Vasodilators: Betahistine (Vertin) 8–16 mg TDS orally increases labyrinthine blood flow and is commonly used.
4. Microwick: A small wick (1 mm × 9 mm) made of polyvinyl acetate can be used to deliver drugs from the external canal to the inner ear, avoiding repeated intratympanic injections. A tympanostomy tube is inserted into the tympanic membrane, and the wick is passed through it to deliver medications like steroids or gentamicin.
5. Intratympanic Injections: If medical treatments fail, repeated intratympanic injections of dexamethasone are indicated. In cases refractory to dexamethasone, gentamicin injections can be considered. Gentamicin, a vestibulotoxic drug, can control vertigo in 60-80% of patients but may lead to sensorineural hearing loss as a long-term complication.
6. Trans-Tympanic Low-Pressure Therapy: The MENIETT device delivers low-pressure pulses (0-20 cm H_2O) to the middle ear via a grommet, which helps redistribute endolymph and reduce pressure.
7. Allergen Elimination: Identifying and eliminating allergens that trigger attacks may be beneficial.

Surgical Treatment

1. **Conservative Procedures:**

- Endolymphatic Sac Surgery: Decompression or shunt procedures can drain excess endolymph.
- Tenotomy of Tensor Tympani and Stapedial Tendons: This procedure can relieve symptoms by reducing the impact of increased cochlear pressure.
- Cochleosacculotomy: Puncturing the cochlear duct to drain it into the perilymph, which helps alleviate symptoms.
- Ultrasonic Destruction of Vestibular Labyrinth: This preserves cochlear function while targeting the vestibular labyrinth.
- Vestibular Nerve Section: A surgical option to control vertigo while preserving hearing, this involves sectioning the vestibular nerve while sparing the facial and cochlear nerves.

2. **Destructive Procedures:**

- Labyrinthectomy: Complete destruction of the membranous labyrinth to relieve vertigo, typically performed when cochlear function is non-serviceable. Approaches include:
- Transmastoid Route: Through the lateral semicircular canal.
- Transcanal Approach: Through the oval window.
- Laser Destruction of Inner Ear: A less invasive option for destroying the inner ear structures responsible for vertigo.

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