

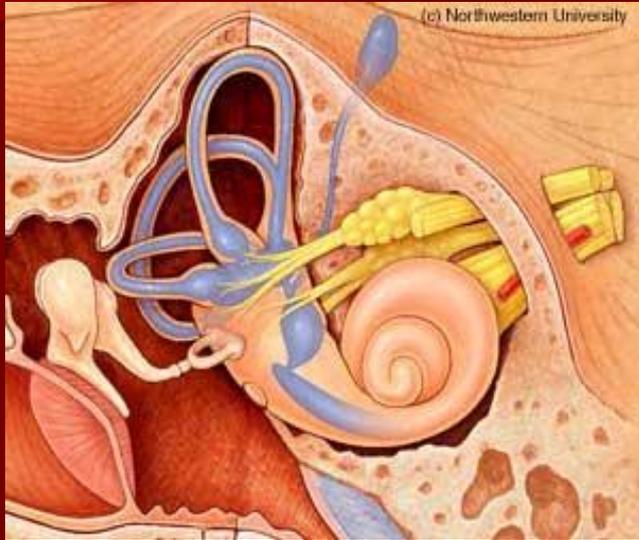
Meniere's Disease

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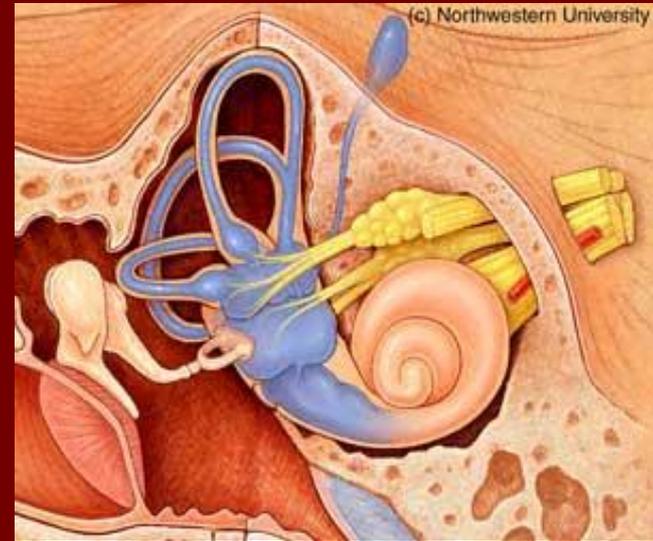
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❖ What is Meniere's Disease?

- In 1861 Prosper Meniere described a syndrome characterized by deafness, tinnitus, and episodic vertigo. He linked this condition to a disorder of the inner ear.
- In 1938 Hallpike and Cairns described the underlying pathology of Meniere's disease as being endolymphatic hydrops but the precise etiology still remains elusive.



Normal membranous labyrinth



Dilated membranous labyrinth
in Meniere's disease (Hydrops)

➤ Possible Causes

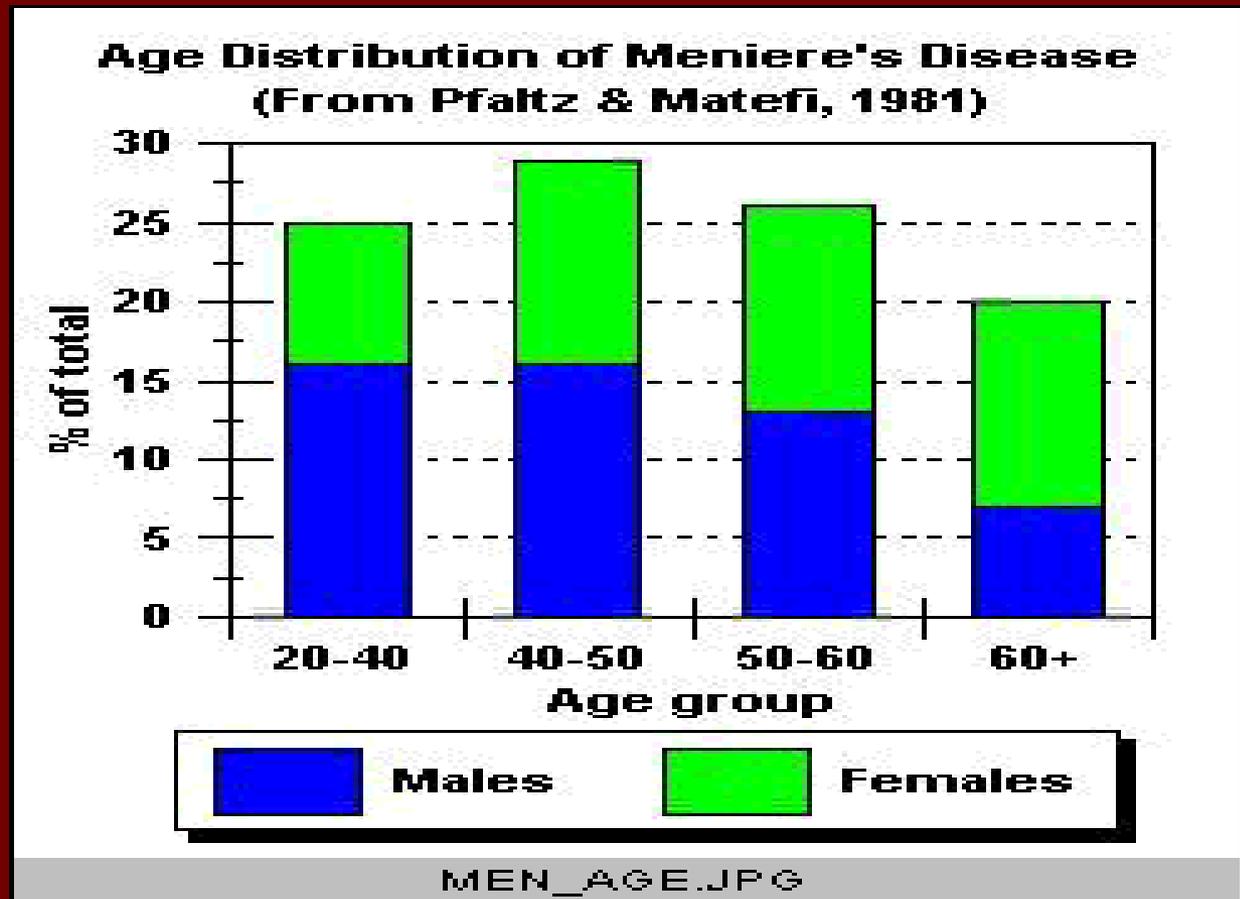
- Anatomical-abnormalities
- Genetic-autosomal dominant
- Immunological-immune complex deposition
- Viral-serum IgE to herpes simplex virus types I and II, Epstein-Barr virus and CMV
- Vascular-associated with migraines
- Metabolic-potassium intoxication

Age Distribution and Incidence

In the US: 50% of patients have a positive family history.
The estimated prevalence is 150 cases per 100,000 population

40's and 50's

Women > Men



Symptoms

- Periodic episodes of rotatory vertigo or dizziness
- Fluctuating, progressive, low-frequency hearing loss
- Tinnitus
- Fullness/pressure

Diagnosis

- The diagnosis of Meniere disease is made based on a careful history and physical exam.
- If the work-up is normal and the classic symptoms continue, the diagnosis of Meniere disease is made.

History

- Most important part of the diagnosis
- Pattern of symptoms
- Association between hearing loss, tinnitus, and vertigo

Physical Examination

- Examination results vary, depending upon the phase of disease. During remission, physical examination findings may be completely normal, particularly if the patient is symptom free.
- During an acute attack, the patient has severe vertigo.
- Patients are sometimes diaphoretic and pale.
- Vital signs may show elevated blood pressure, pulse, and respiration.
- Spontaneous nystagmus directed toward affected ear is typical during an acute attack.

Physical Examination (con't)

- The Romberg test generally shows significant instability and worsening when the eyes are closed.
- The Weber tuning fork test usually lateralizes away from the affected ear.
- The Rinne test usually indicates that air conduction remains better than bone conduction.
- Complete neurologic evaluation is important. New-onset vertigo might be an early sign of stroke, migraine, or brainstem compression that may require emergent evaluation and care.

Lab studies

- No lab studies are specific for Meniere disease.
- A CBC, urinalysis, chemistry panel, and alcohol and drug screening may be helpful if other causes are considered.
- If an infectious cause is suspected, consider blood cultures, urine culture, and a cerebral spinal fluid (CSF) examination.

Imaging Studies

- Magnetic resonance imaging
 - Brain scan should be done to rule out abnormal anatomy or mass lesions. Specifically, acoustic neuromas or other cerebellopontine angle lesions are sought. Other lesions, such as multiple sclerosis or Arnold-Chiari malformations, also can be ruled out.
 - Note that mass lesions rarely are found but are important to exclude.
- CT scans reveal dehiscent superior semicircular canals and/or widened cochlear and vestibular aqueducts

Other tests

- **Audiometry** is particularly helpful to document present hearing acuity and to detect future change.
 - The patient may not notice a loss at specific frequencies. Low-frequency or mixed low- and high-frequency insufficiency may be observed.
 - Typically, the lower frequencies are affected more severely. This is due to preferential sensitivity of the apex to the hydrops.
 - Multiple hearing tests, which document fluctuating hearing loss, are helpful in diagnosing Ménière.

Transtympanic electrocochleography (ECOG)

- Transtympanic electrocochleography (ECOG) specifically detects distortion of the neural membranes of the inner ear.
- This is presumably due to perilymph pressure fluctuations and can show evidence of cochlear involvement.
- ECOG measures the ratio of the summing potential (probably from the movement of the basilar membrane) and the nerve action potential in response to auditory stimuli. Hydrops is suggested when this ratio is greater than 35%.
- This is most accurate when Ménière is active.

Electronystagmography (ENG)

- Electronystagmography (ENG) is a test of the inner ear function (particularly the semicircular canals).
- It tests central and peripheral function and can help localize the site of lesion.
- Typically, Meniere disease causes a reduced vestibular response in the affected ear, although response may be increased secondary to an irritative lesion.
- The direction of the spontaneous nystagmus during or after an attack of Ménière is not a reliable indicator of the site of the lesion. An irritative phase may occur during the attack (fast phases directed toward involved ear) followed by a parietic phase (fast phases directed toward opposite ear).

Differential Diagnosis

- The differential diagnosis is broad and includes:
- perilymph fistula,
- recurrent labyrinthitis,
- otosclerosis,
- migraine ,
- congenital ear malformations of many kinds,
- viral meningitis, viral encephalitis,
- neurosyphilis,
- stroke,
- tumors,
- trauma,
- autoimmune disorders,
- MS, etc.

Treatment

- Medical therapy is both symptomatic (ie, acute attacks) and prophylactic.
- If Ménière is due to a secondary cause (ie, Ménière syndrome), primary first-line management is the diagnosis and treatment of the primary disease (eg, thyroid disease).
- Vestibulosuppressants (eg, meclizine) decrease symptoms, but generally only mask the vertigo by decreasing the brain's response to vestibular input.

Treatment Cont'd

- Diuretics or diuretic-like medications (eg, hydrochlorothiazide) actually decrease the fluid pressure load in the inner ear. These medications help prevent attacks but do not help once an acute attack has started.

Treatment Cont'd

- Anti-inflammatory properties of steroids are helpful in endolymphatic hydrops. This is probably due to reduced endolymphatic pressure. Steroids actually can reverse vertigo, tinnitus, and hearing loss.

Treatment cont'd

- Aminoglycosides are a class of antibiotics that were discovered serendipitously to be preferentially toxic to the vestibular end organ.
 - Destruction of the vestibular end organ renders the brain insensitive to the fluctuations in the inner ear pressure during an acute Ménière attack.
 - If given systemically, aminoglycosides affect both ears.
 - Although these drugs can be used to treat extremely severe bilateral Ménière disease, they leave the patient with little or no balance function. The resulting Dandy syndrome, a complete loss of inner ear function, can be debilitating.

Treatment cont'd

- During the quiescent phase, medical treatment of Ménière disease is tailored to each patient. Lifestyle and dietary changes are usually the first step. Avoiding trigger substances (eg, caffeine) alone may be sufficient. Smoking cessation also is recommended.

Treatment cont'd

- In an acutely vertiginous patient, management is directed toward vertigo control.
 - Intravenous (IV) or intramuscular (IM) diazepam provides excellent vestibular suppression and antinausea effects.
 - Steroids can be given for anti-inflammatory effects in the inner ear.
 - IV fluid support can help prevent dehydration and replaces electrolytes.

Treatment Cont'd

■ Surgical Care:

- Surgical therapy for Ménière disease is reserved for medical treatment failures and is otherwise controversial.
- Surgical procedures are divided into 2 major classifications as follows:
 - Destructive surgical procedures
 - Nondestructive surgical procedures

surgical procedures Cont'd

- Destructive surgical procedures
 - Rationale to control vertigo: Endolymphatic hydrops causes fluid pressure accumulation within the inner ear, which causes temporary malfunction and misfiring of the vestibular nerve. These abnormal signals cause vertigo. Destruction of the inner ear and/or the vestibular nerve prevents these abnormal signals. As long as the opposite inner ear and vestibular apparatus function normally, the brain eventually will compensate for the loss of one labyrinth.

Destructive surgical procedures Cont'd

- Problems with destructive procedures:
 - Destruction of one inner ear depends on the adequate function of the opposite ear. Unfortunately, Ménière disease can be bilateral (7-50%), in which case this method is contraindicated. Since balance and hearing are closely intertwined within the labyrinth, destruction of the balance portion carries a high risk of hearing loss. Note that destructive procedures are irreversible and reserved for severe cases.

surgical procedures Cont'd

- Nondestructive surgical procedures:
 - These are directed toward improving the state of the inner ear. They are less invasive than destructive procedures and do not preclude the use of other treatment modalities. Discussion here is limited to the 4 most generally accepted management options:
 1. endolymphatic sac decompression or shunt
 2. vestibular nerve section
 3. Labyrinthectomy
 4. transtympanic medication perfusion.

surgical procedures cont'd

- Endolymphatic sac decompression and/or shunt
 - In theory, the endolymphatic sac procedure decreases endolymph pressure accumulation by removing the petrous bone, which encases the endolymph reservoir. This procedure allows the reservoir sac to expand more freely, thus dissipating pressure. A drain or valve from the endolymphatic space to either the mastoid or subarachnoid space can be inserted as another means of further reducing pressure.
 - Success rates (in terms of controlling vertigo and stabilizing hearing acuity) with this procedure are reported at 60-80%.

surgical procedures Cont'd

■ Vestibular nerve section

- For patients with useful hearing in the affected ear, sectioning the diseased vestibular nerve can be the ultimate solution.
- Although the hearing and balance functions are housed in one common chamber within the inner ear, their neural connections to the brain separate into distinct nerve bundles as they course through the internal auditory canal.
- This anatomical separation allows balance function to be isolated and ablated without affecting hearing function.

surgical procedures Cont'd

■ Labyrinthectomy

- This management option for Ménière disease has the advantage of a high cure rate (>95%) and is useful in the patient whose hearing on the diseased side has been destroyed already by Ménière disease.
- Labyrinthectomy involves ablation of the diseased inner ear organs.
- This procedure is less complex than vestibular nerve section because labyrinthectomy does not require entry into the cranial cavity.
- Labyrinthectomy is less invasive than vestibular nerve section.

Labyrinthectomy Cont'd

- This procedure carries less danger of cerebrospinal fluid leak and meningitis since craniotomy is not required.
- Like those who undergo vestibular nerve section, patients require a few days of inpatient care.
- Accommodation to the surgical loss of one vestibular apparatus usually takes weeks or months.
- Vestibular rehabilitation during this time period is also helpful.

surgical procedures Cont'd

- Transtympanic perfusion of medication
 - Medications for Ménière disease are applied through a myringotomy within the middle ear cavity, where they presumably are absorbed through the round window membrane into the inner ear.
 - Transtympanic perfusion is a relatively low-risk, simple procedure that applies a high concentration of medicine with minimal systemic effects.

Treatment cont'd

■ Diet:

- Dietary management is appropriate in patients not severely affected; patients avoid substances that may trigger or exacerbate fluid pressure buildup in the inner ear.
- Similar to managing systemic hypertension, the goal for Ménière disease is to reduce the total body fluid volume. This, in turn, may reduce the inner ear fluid volume.
- Since sodium seems to play a major role in fluid retention within the inner ear, avoiding salt (eg, pizza, preserved foods, smoked fish) is paramount.

Diet Cont'd

- Consult with a nutritionist to establish a rigid salt-restricted diet (1.5 g sodium per day).
- Avoiding other trigger substances (eg, caffeine, nicotine, alcohol, high-carbohydrate substances, high-cholesterol/triglyceride foods) also can help.
- Note that many preserved and smoked foods contain sodium nitrite, which can contribute to high sodium content.

Treatment Cont'd

■ Activity:

- Endolymphatic hydrops does not preclude regular activity. Exercise is recommended in moderation.
- Because of the unpredictable nature of the disease, balance-intensive, dangerous tasks (eg, especially climbing ladders) should be avoided.

Prognosis

- Prognosis is variable, since the disease pattern of exacerbation and remission makes evaluation of treatment and prognosis difficult to predict.
 - In general, Ménière symptoms tend to stabilize spontaneously with time. With regard to vertigo, about half of patients stabilize over several years.
 - Patients tend to "burn out" over time and with residual poor balance and hearing.

Prognosis Cont'd

- Ménière disease can be classified into several stages of progression. Early stages involve cochlear hydrops, which proceeds to affect the vestibular system.
 - Ménière disease is most bothersome during these early stages.
 - As patients progress to later stages, the hydrops fills the vestibule so completely that no further room is available for pressure fluctuation and the vertigo spells disappear.
 - The acute attacks are replaced by constant imbalance and progressive hearing loss.