MENIERE DISEASE

DEFINITION

- Meniere's disease is a disorder characterized by spontaneous attacks of vertigo, with associated fluctuating sensorineural hearing loss, tinnitus, and aural fullness (TVSA) due to abnormal ion and fluid homeostasis in inner ear. It is usually unilateral.

AETIOPATHOGENESIS

- Idiopathic
- Age – any but usually 20 to 40 yrs of age.

PROPOSED THEORIES:

- Blockade of Endolymphatic duct (Predisposing factors lead to increased production and decreased absorption of Endolymph resulting in endolymphatic hypertension leading to gross enlargement of membranous labyrinth, so called Endolymphatic hydrops (can only be diagnosed by postmortem histopathological examination of temporal bone))
- Immunological Mechanism
- Genetic Predisposition (AD inheritance)
- Viral Predisposition not proved
- Vascular Etiology (Migraine Association) (VIG viral vascular, immunological, genetic)

RUPTURE THEORY ➔ Periodic ruptures of the membranous labyrinth, which result in leakage of the potassium-rich endolymph into the perilymph, are thought to be responsible for producing Meniere’s attacks.

1. Initial Irritative Phase
2. Subsequent Paretic Phase
3. Recovery Phase

- Initial excitation of the hair cells as a result of the increased potassium concentration around their basal surfaces is thought to result in the initial irritative phase of an attack. The subsequent paretic phase is thought to result from a blockade of neurotransmitter release.

- Recovery phase – Healing of the rupture then allows restitution of the normal chemical composition of the endolymph and perilymph, resulting in termination of the attack and improvement in vestibular and auditory function.

- The gradual deterioration in inner ear function probably results from the effects of repeated ionic insults leading to degeneration of Hair cells.

MENIERE SYNDROME ➔

- Many different inner ear or temporal bone diseases, such as
  1. Syphilis
  2. Mumps
  3. Cogan's syndrome
  4. Trauma
5. CSOM, can, after many years, produce the clinical picture of Meniere's disease so called Meniere's syndrome.

- Menieres syndrome in children usually associated with congenital malformations of inner ear.
- Incidence \( \Rightarrow 10-150 \) per 1,00,000 person
- The patient describes attacks of spontaneous vertigo beginning many years after the onset of their unilateral hearing loss.
- Sometimes the attacks of vertigo coincide with the onset of fluctuating hearing loss in the other, previously normal, ear; this is known as contralateral delayed endolymphatic hydrops, which is thought to be due to the development of autoimmunity to inner ear antigens.

**CLINICAL FEATURES:**

- Spontaneous recurrent Vertigo with nausea and vomiting.
- **LOW FREQUENCY** hearing losses.
- Tinnitus and Aural Fullness.
- Vertigo – from min to hours
- Tinnitus and SNHL for days
- A typical attack has three phases \( \Rightarrow \) IRRITATIVE, PARETIC and RECOVERY.
- Irritative phase \( \Rightarrow \) in which the nystagmus, usually horizontal or horizontal-torsional, beats towards the affected ear.(lasts less than an Hr)
- Paretic phase \( \Rightarrow \) nystagmus beats away from the affected ear.(lasts several hours sometimes even one or two days)
- Recovery phase \( \Rightarrow \) nystagmus again beats towards the affected side (lasts same like second phase)
- Some patients, particularly those in the later stages of the disease, develop drop attacks.
- These attacks, also called **TUMARKIN OR OTOLITHIC CRISIS**, are thought to occur as a result of acute otolithic dysfunction.
- The patient simply drops to the ground without warning and can sustain a fracture or other serious injury.
- There is no associated vertigo or loss of consciousness.
- **Positive Henneberts sign** (false positive fistula sign) can be seen in Menieres disease

**DIFFERENTIAL DIAGNOSIS:**

- Vestibular schwannoma
- Multiple sclerosis
- TIA
- Migraine
- Diabetes and thyroid diseases can mimic similar symptoms.

**DIAGNOSIS**

**AAOO Criteria for Meniere’s Disease:**

- **POSSIBLE MENIERE’S DISEASE:**
  - Spontaneous Episodic vertigo without documented Hearing loss
  - SNHL, fluctuating or fixed with dysequilibrium but with no definitive episodes
Other Causes Excluded

- **PROBABLE MENIERE'S DISEASE**:
  - One Definitive episode of Vertigo
  - Audiometrically documented Hearing loss at least one occasion.
  - Tinnitus or aural fullness present in affected ear
  - Other causes ruled out

- **DEFINITIVE MENIERE'S DISEASE**:
  - Two or more definitive spontaneous episodes of Vertigo 20 minutes or longer.
  - Audiometrically documented hearing loss on at least one occasion
  - Tinnitus or aural fullness in affected ear

- In the early stages of Meniere's disease, vestibular and auditory function tests are often completely normal between attacks.

- Audiology → **Low frequency losses**
- Vestibular function test → Rotating chair test, Electronystagmography
- Electrocochleography
- Laboratory test for Syphilis, Mumps etc
- Tests for Inner ear antigens
- **Test for endolymphatic hydrops** – GUS i.e. glycerine, urea and sorbitol stress test
- VEMP → Vestibular Evoked Myogenic Potential.

**MANAGEMENT OPTIONS**

**NON INTERVENTIONAL TREATMENT**

**LIFESTYLE MODIFICATIONS**

- Low Caffeine, Alcohol, Nicotine, Salt (CANS) diet, Stress also precipitates Menieres
- Salt restriction to 2 to 3 gms per day

**MEDICAL MANAGEMENT**

- **Acute Attacks** – Vestibular suppressants like BZD’s, antihistaminic, anticholinergics
- Promethazine (antihistaminic (phenargan)) and prochlorperazine (D2 antagonist)
- Beta Histine (H1 agonist and H3 antagonist) → Vasodilator
- Diuretics
- Glucocorticoid treatment for possible immunological etiology.
- Other vasodilators and vitamin regimes

**REHABILITATION**:

- Hearing aids
- Vestibular rehabilitation – for patients who responded to medical or surgical treatment for vertigo but have some remaining disequilibrium.

**INTERVENTIONAL TREATMENT**:

**NON DESTRUCTIVE SURGICAL PROCEDURES**
1. Endolymphatic sac surgery (Enhancement, Shunting or Both)
2. Sacculotomy
   - Have an advantage of low risk of SNHL

**INTRATYMPANIC GENTAMYCIN**

- Delivered by injection or cannula ➔ Vestibulotoxic ➔ penetrates via round window membrane ➔ destroys hair cells ➔ Ablates labyrinthine function.
- Multiple applications of Gentamycin controls vertiginous symptoms but doesn’t affect hydrops and associated fullness, hearing fluctuations and sensory loss.
- Treatment has risk of SNHL which is irreversible.
- Low dose Gentamycin has been tried to affect only vestibular function without affecting cochlear function.
- Gentamycin is Vestibulotoxic and causes damage to dark cells of secretory epithelium thus leading to decreased endolymph production.

**TRANSTYMPANIC GENTAMYCIN**

- Gentamycin (4mg/ml) buffered with NaHCO₃ until pH is 6.4 ➔ T tube grommet in posteroinferior quadrant of TM as round window niche comes there ➔ 1ml administered ➔ 3 injections per day (7am-1pm-7pm) ➔ Patient lies half hour supine with ear facing up.
- Treatment stopped only if PTA/Nystagmus/Vertigo deteriorates.

**INTRATYMMPAINIC GLUCOCORTICOIDS** have been tried

**ENDOLYMPHATIC SHUNT SURGERY**

- External shunts (sac to mastoid or subarachnoid space)
- Internal Shunts (eg Fick’s sacculotomy, Cody Tack operation, Cochleosacculotomy of Schuknecht (LABYRINTHOTOMY), Otic periodic shunt of House and Pulec)

**LABYRINTHOTOMY**

- **TRANS CANAL LABYRINTHOTOMY (LEMPERT’S)**
- **TRANSMASTOID LABYRINTHOTOMY**

**VESTIBULAR NEURECTOMY**

- **TRANSLABRYNTHINE**
- **RETROLABRYNTHINE**
- **RETROSIGMOID**
- **MIDDLE CRANIAL FOSSA**

- Positive pressure pulse generator (Meniett’s) increases the fluid exchange in inner ear.

**ENDOLYMPHATIC SAC SHUNT/DECOMPRESSION**

**INDICATIONS**
Servicable hearing of involved.
Significant chance of developing Menieres in contralateral ear i.e. younger patients.

CONTRAINDICATIONS

- Congenital inner ear dysplasia
- Tertiary syphilis or perilymph fistula.

PROCEDURE:

- Mastoidectomy done → Lateral SCC and fossa incudis identified → sigmoid sinus skeletonised → dura exposed at the part of bone anterior to Sigmoid sinus and posterior to posterior SCC → Endolymphatic Sac identified (thickened whitish appearing area of dura) → lateral wall of SCC incised and sialistic implant introduced.

COMPLICATIONS:

- Vertigo and Disequilibrium
- Hearing loss
- Facial Weakness
- Tinnitus
- CSF leak and infection

COCHLEOSACCULO TOMY or LABYRINTHOTOMY

- TM flap elevated → round window niche exposed → 3mm rt angled pick advanced through round window in direction of oval window → when introduced to full length end of pick is located below footplate of stapes → pick withdrawn and round window perf covered by tissue graft.

LABYRINTHECTOMY

- Surgical removal of bony and membranous labyrinth by removal of all the neuroepithelium from treated side, but causes irreversible hearing loss.

LEMPERT’S TRANS CANAL LABYRINTHECTOMY

- Posterior annulus curetted to expose horizontal segment of facial nerve and entire limits of round window niche → incus removed, stapedius tendon cut and stapes removed intact → Oval window enlarged by removing its inferior margin → 3-4mm rt angled utricle hook is used to locate and remove the utricle → ampullary ends of SCC are probed to ensure destruction of neuroepithelium, posterior ampullary nerve is sectioned → vestibule is filled with absorbable gelatin sponge/fat graft

TRANSMASTOID LABYRINTHECTOMY:

- Canal wall up → Incus removed → SCC skeletonised opened and 3 ampullar and neuroepithelium removed with rt angled hook → vestibule packed with Gentamycin soaked Gelfoam