Definition

- Genetically-mediated primary metabolic bone disease that affects the human otic capsule, characterized by formation of new spongy bone formation.
- Mode of inheritance
  - Autosomal dominant with incomplete penetrance (40%) and variable expressivity
Epidemiology

- 10% overall prevalence of histologic otosclerosis
- 1% overall prevalence of clinically significant otosclerosis

History

- **Gender:** Female to Male ratio = 2 : 1
- **Race:** More common in Caucasians, rare in Negroes
- **Age:** 15-45 most common age of presentation
  Rare onset in children and after 55 years.
- **Side:** Otosclerosis is bilateral in 70% of cases.
- **Family history:** positive in more than 50%
Pathology

Two phases of disease

1. Active/immature (otospongiosis phase)
   - Osteolytic resorption of bone, with sheets of vascular connective tissue replacing the bone. Schwartze's sign is positive.

2. Inactive/mature (sclerotic phase)
   - Formation of dense sclerotic bone in areas of previous resorption, signifies the late phase of otosclerosis. The result is disorganized bone, with narrow vasculature and few recognizable haversian systems.

Pleomorphism is largely due to normal coexistence of both stages of otosclerosis in any single temporal bone.
1. Clinical otosclerosis
   - Pathology involving and fixing the footplate of stapes.
   - Clinically represented by conductive hearing loss.

2. Histological otosclerosis
   - Pathology involving areas of the otic capsule away from the footplate of stapes, which remains freely mobile.
   - Only discovered accidentally after death during histological examination of the temporal bone at autopsy.

3. Cochlear otosclerosis
   - Pathology involves a large part of the labyrinthine endosteum.
   - Metabolites diffuse to the inner ear, injuring the hair cells.
   - Clinically represented by SNHL component which adds to the existing HL (resulting in Mixed or rarely Pure SNHL).

Fissula ante fenestram (80%--90%)
- Round window niche (30%--50% of cases)
- Anterior wall of the IAC

Pathology
Pathology
Secondary Endolymphatic Hydrops

Etiology

The etiology of this disease is still unknown and appears to be multifactorial.

- **Hereditary:**
  - +ve family history in 50%.
  - Mode of inheritance:
    - Autosomal dominant with incomplete penetrance (40%) and variable expressivity.

- **Viral etiology:**
  - Measles virus RNA in 80% of otosclerotic stapedes.

- **Hormonal factor:**
  - More common in females.
  - Rapid progression during pregnancy.

- **Other**
  - Autoimmune.
  - Vascular.
  - Metabolic.
Clinical picture

Symptoms

- Hearing loss (Slowly progressive)
- Tinnitus 75%
- Vestibular symptoms (uncommon) 25%
- Paracusis Willisii: The patient hears better in a noisy environment.

*This occurs because people speak louder in noisy surroundings.*
Examination

- Tuning fork tests:
  - Rinne test is –ve
  - Weber test lateralizes to same side
  - Schwabache test is prolonged
  - Gelle test: +ve
Pure Tone Audiometry

- Conductive HL (80%): air-bone gap
- Carhart’s notch
- Mixed HL (15)
- Pure SNHL (5%)

Tympanometry

- Normal type A
- Shallow type As
Acoustic Reflexes

- Reflexes are absent
- Reflexes are biphasic (on-off effect)

Differential Diagnosis

- Ossicular discontinuity
- Congenital stapes fixation
- Malleus head fixation
- Paget’s disease
- Osteogenesis imperfecta
- SNHL in young adults
Management

- Surgery
- Amplification
- Medical therapy

Surgery

- Stapes surgery
  - Stapedectomy
  - Stapedotomy
Stapedectomy
- Endomeatal incision
Stapedotomy
Amplification

- Satisfaction rate is less than with successful surgery
- Indications
  - Poor surgical candidates
  - Only-hearing ear
  - Patients who do not desire surgery.
- Postop:
  - Patients with mixed HL
  - Failed surgery
Medical Treatment

**Sodium Fluoride**

- Causes maturation of active foci of otosclerosis
- It only alters the course of active not the mature form of otosclerosis
- Reduces tinnitus, reverses Schwartz’s sign.
- Indications
  - Active otosclerosis with progressive mixed hearing loss
  - Otosclerosis with + Schwartz’s sign
  - Treat for 6 m. pre-op
  - Postop if active otospongiosis is detected intra-op
THANK YOU
FOR YOUR ATTENTION