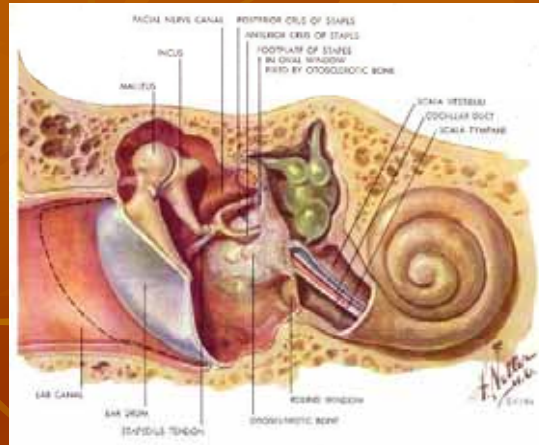


Otosclerosis



Mokhtar Bassiouni

Definition

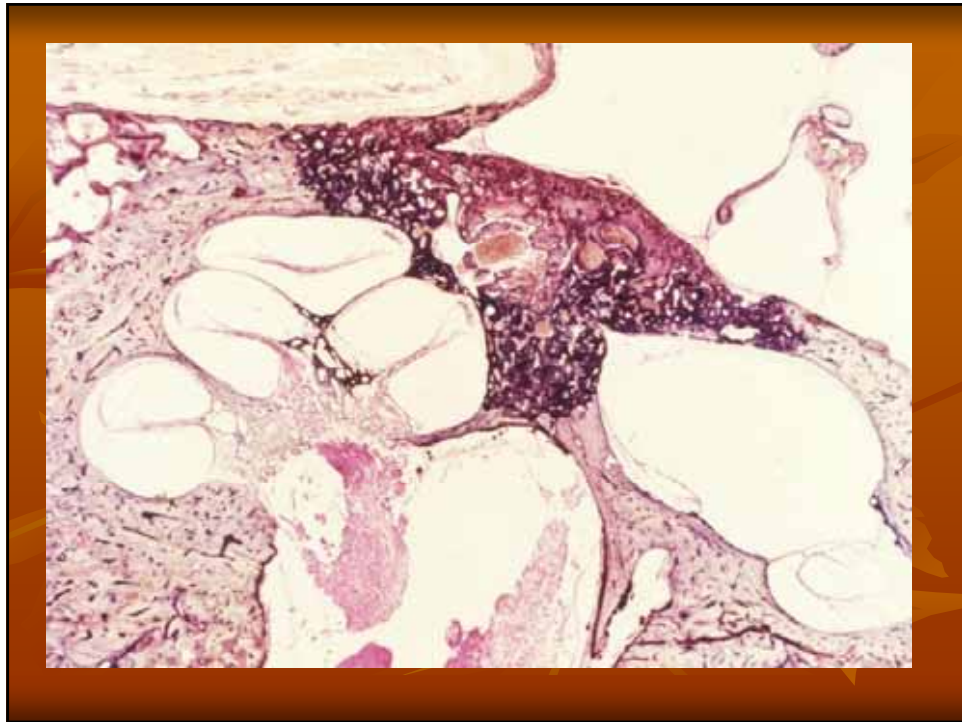
- Genetically-mediated primary metabolic bone disease that affects the human otic capsule, characterized by formation of new spongy bone formation.
- Mode of inheritance
 - Autosomal dominant with incomplete penetrance (40%) and variable expressivity

Epidemiology

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

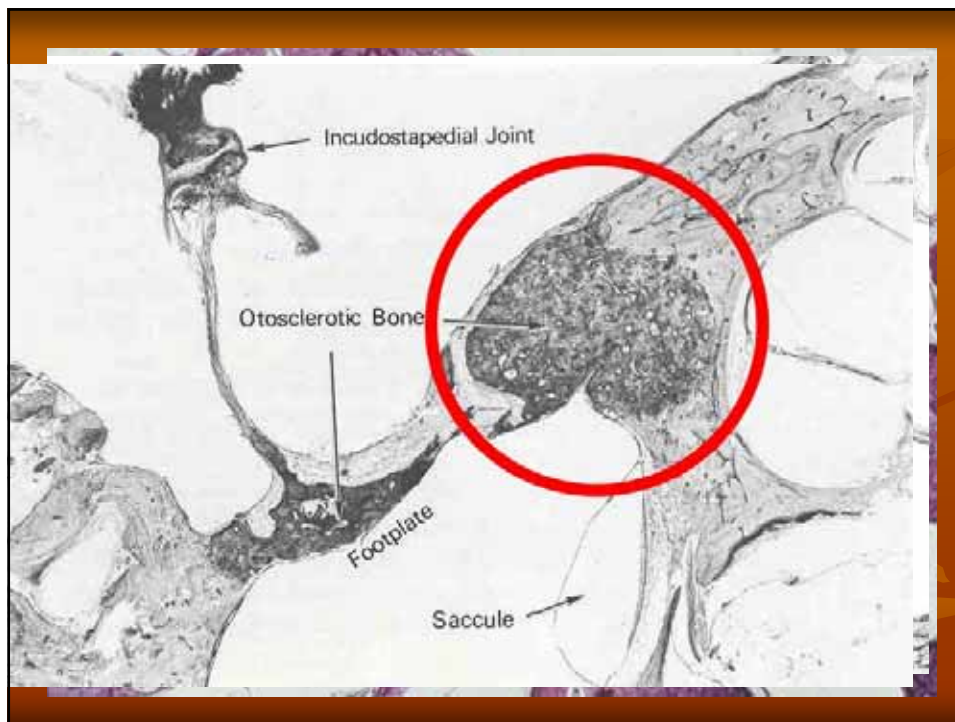
History

- **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%*

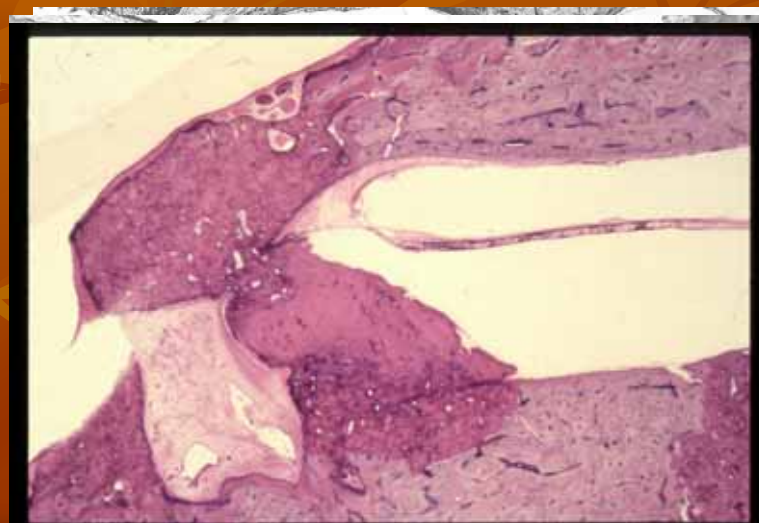


Pathology



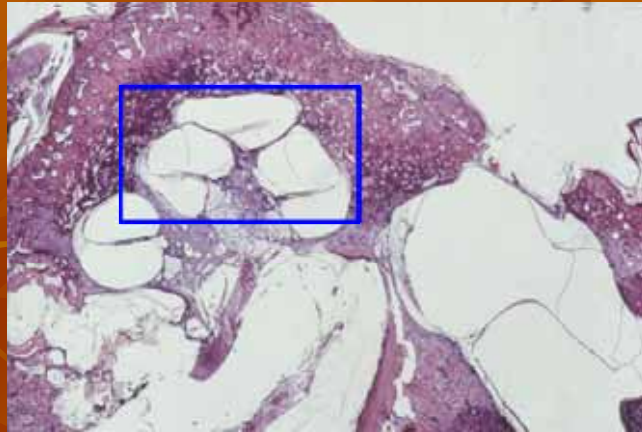


Pathology



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.



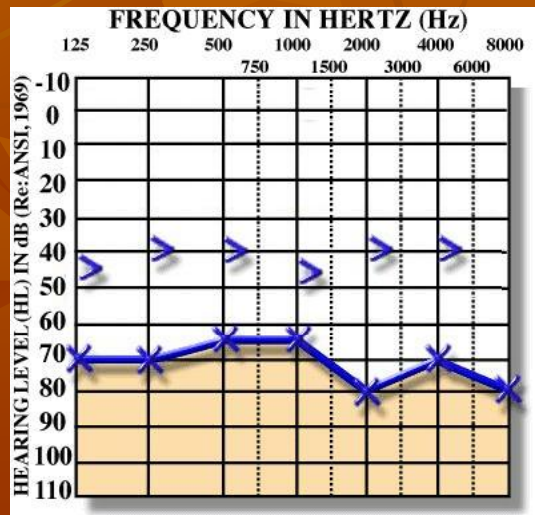
Examination

- Tuning fork tests:
 - Rinne test is -ve
 - Weber test lateralizes to the right
 - Schwabache test is positive
 - Gelle test: +ve



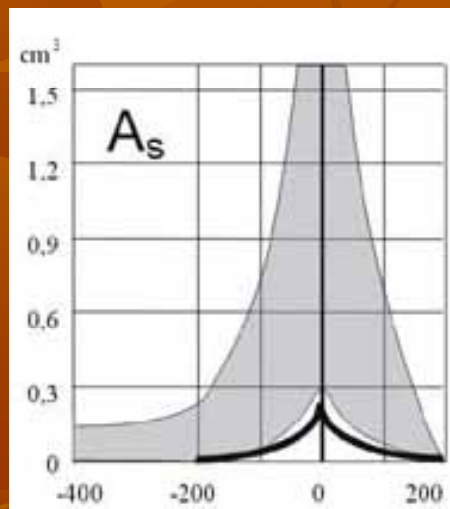
Pure Tone Audiometry

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

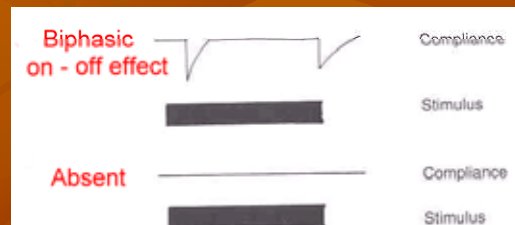


Tympanometry

- 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

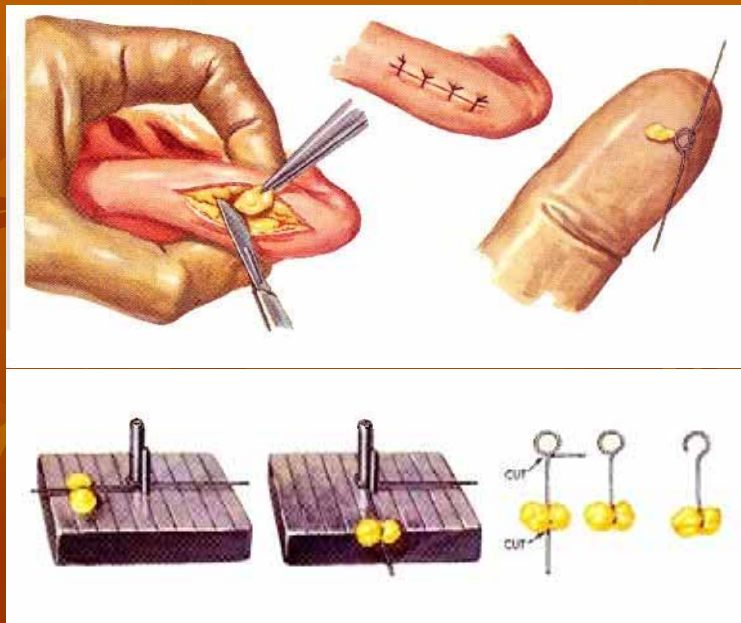
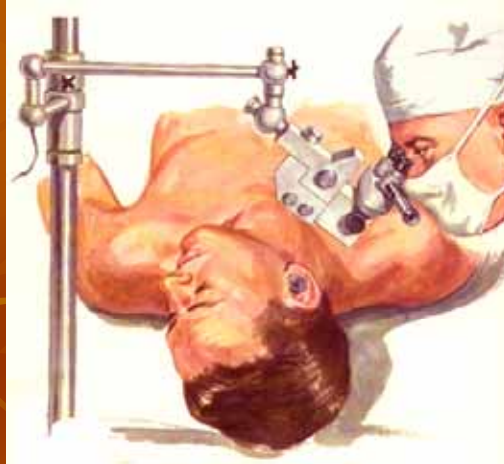
Management

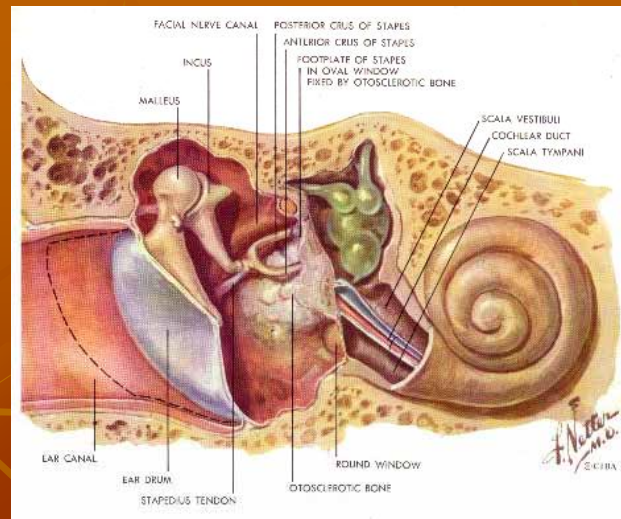
- Surgery
- Amplification
- Medical therapy

Surgery

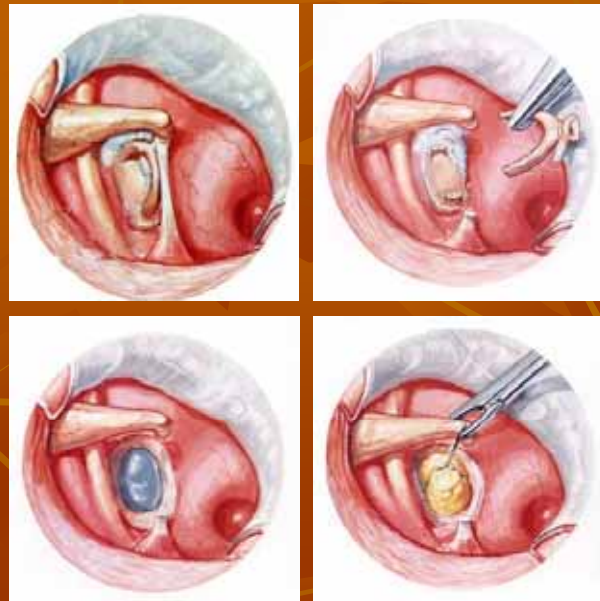
- Stapes surgery
 - Stapedectomy
 - Stapedotomy

Stapedectomy

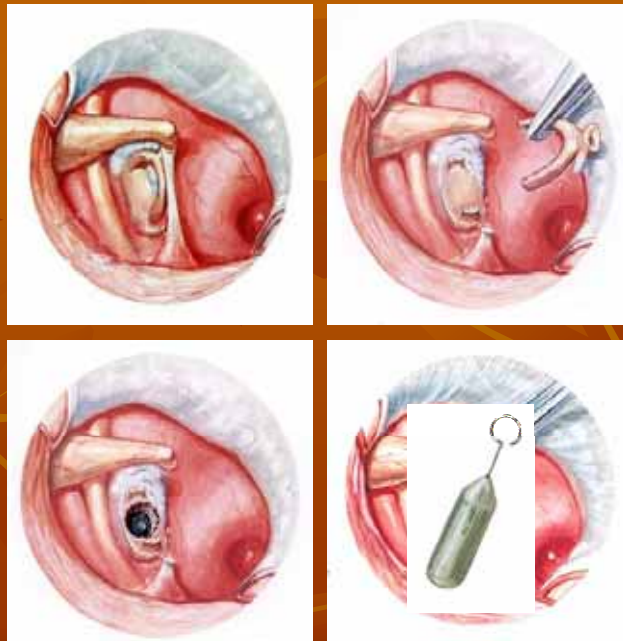
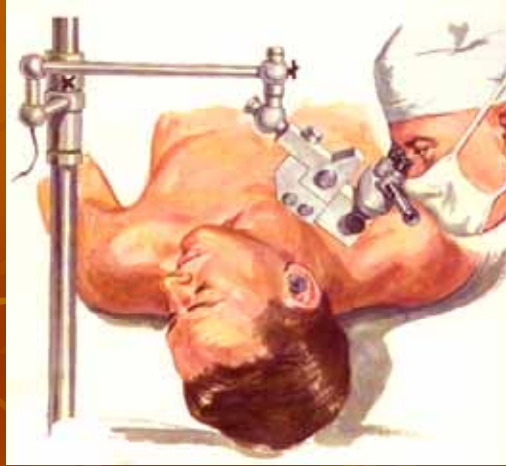


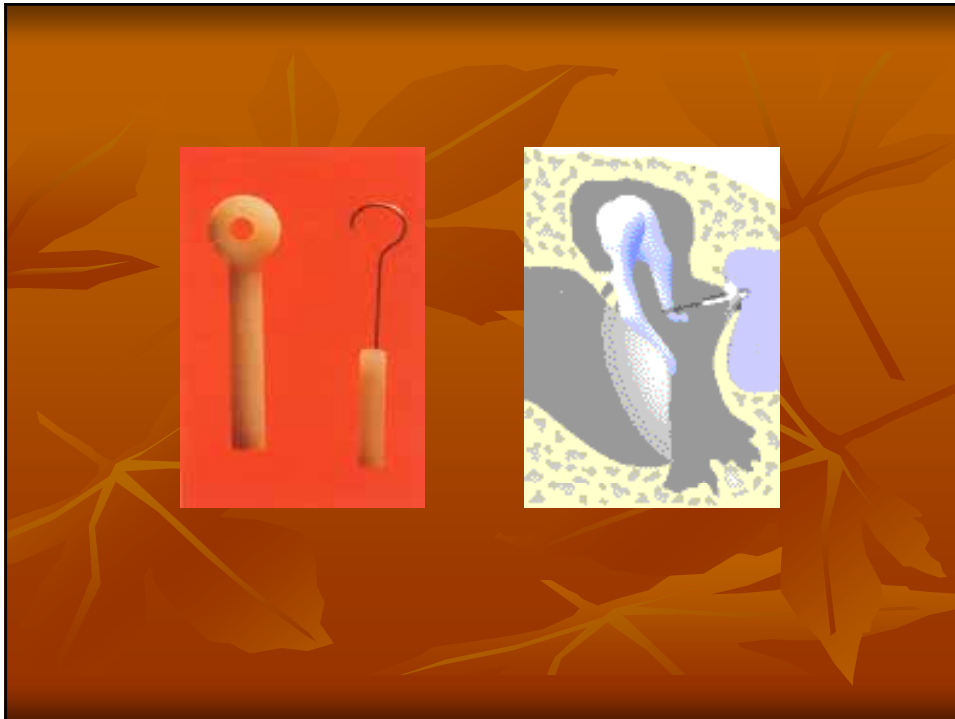


■ Endomeatal incision



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

