Otosclerosis
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Definition
A localized hereditary disorder affecting endochondral bone of the otic capsule that is characterized by disordered resorption and deposition of bone

Types
- Clinical involves stapes or stapedovestibular joint
  - Anterior focus
  - Posterior focus
  - Circumferential
  - Biscuit type
  - Obliterative
- Histologic does not involve stapes or stapedovestibular joint, asymptomatic
- Cochlear Pure SNHL

Histopathology
- Earliest Histologic alteration – blue mantle
- Blue Mantle – areas of otic capsule that stain more basophilic than normal
- Resorption of endochondral bone with enlargement of perivascular spaces
- Followed by deposition of immature bone (leads to immature spongiotic bone)
- In time, active resorption and remodeling occur continuously within an otosclerotic focus with production of more mature (lamellar) bone (leads to inactive mature focus)
- Process of bone resorption mediated by osteoclasts
- Process of new bone deposition by osteoblasts
- Proliferation of new blood vessels in otosclerotic focus
- Absence of acute inflammatory cells
- An otosclerotic focus is usually little larger than the bone it replaces, hence leads to thickening of bone

Distribution of Focus
1. Area anterior to oval window (fissula ante fenestrum)(80-95%)
2. Round window niche
3. Apical medial wall of cochlear labyrinth
4. Stapes footplate
5. Post. to oval window
6. Walls of internal auditory canal
7. Around vestibular and cochlear aqueducts
8. Around semicircular canals
9. Around malleus and incus
Etiology

- Genetic predisposition
  o Most common in white (i.e. Caucasians – 10%), less in Asian (5%), least in black (i.e. Africans – 1%)
  o F:M = 2:1
  o Autosomal dominant transmission with incomplete penetrance (25 – 40 %) – COL1A1 gene
  o 50% patients of osteogenesis imperfecta have otosclerosis
- Measles
  o Measles RNA found in stapes footplate with otosclerosis
  o Elevated levels of anti measles antibody reported in these patients
- Autoimmune disease
  o Humoral autoimmunity to type II collagen
- Biochemical changes
  o Changes in mucopolysaccharide composition and concentration
- Stress
- Pregnancy
  o Condition worsens after pregnancy (10 – 17 %)

Evaluation

History

- Hearing loss is of gradual onset and progresses slowly over several years
- Approximately 70% of otosclerosis cases are bilateral
- Become apparent in the late teens or twenties
- Hearing loss may not become apparent to the patient until age 30 or 40 years.
- Because most patients with otosclerosis have a conductive hearing loss, the patients will report difficulty hearing conversation while chewing
- May hear better in noisy rooms because of paracusis willi
- With unilateral hearing loss, the patients tend to have difficulty with direction of sound and in noisy rooms.
- Tinnitus – in cochlear Otosclerosis and active focus
- Vertigo - uncommon but maybe present
- There is usually a positive family history of hearing loss and often of surgical correction of hearing loss.
- The patient usually has a negative history for infections or trauma as a possible cause of the conductive hearing loss
- Speech is monotonous well-modulated soft speech

Examination

- Otoscopy
  o Pneumo-otoscopy is important to rule out middle ear serous fluid that could be the cause of a conductive hearing loss.
  o There may be a red blush over the promontory or the area anterior to the oval window
called the **Schwartzze sign** or flamingo flush

- **Tuning forks tests**
  - Rinnes Negative (initially for 256, later for 512 & 1024 as well)
  - Weber Lateralized to the ear with greater conductive loss
  - ABC may be reduced in cochlear Otosclerosis
  - Bing negative
  - Gelles negative (important in Otosclerosis, see dhingra pg 24)

- **PTA**
  - Loss of air conduction more for lower frequencies
  - Dip in bone conduction curve – max at 2000 Hz – Carhart’s Notch
  - Mixed hearing loss or SNHL might be present

- **Tympanometry**
  - Tympanogram is “A” or “As”

- **Acoustic reflexes**
  - Are sensitive measures of the movement of the stapes.
  - In the presence of otosclerosis, the reflex will be absent.
  - With early stapes fixation, the reflex may be abnormal in that there is a negative on-off effect or diphasic reflex
  - With a more advanced disease, the reflex is absent when the probe is in the involved ear.
  - As the disease and hearing loss worsen, the contralateral reflex will be affected as a result of the degree of hearing loss in the otosclerotic ear.

- **Speech Audiometry**
  - Speech discrimination score \(\rightarrow\) high

- **HRCT**
  - Detection of hypodense area around oval window and cochlea
  - Gray scale has to be calculated as visually grey areas with differing density not discernible
  - Sensitivity (based on research by Miura et al) up to 94%
  - CT Grading of otosclerosis (by Symons & Fanning)
Shambaugh’s criteria for diagnosing cochlear otosclerosis

1. Positive Schwartze’s sign
2. Positive Family history
3. Symmetrical SNHL
4. SNHL cannot be attributed to any other cause
5. Flat, rising or cookie bite audiogram with Good Speech Discrimination Score
6. Fixation of stapes

Presently CT findings are also required.

Differential diagnosis

- Tympanosclerosis
- Ossicular discontinuity
- Congenital stapes fixation
- Malleus head fixation
- Paget’s disease
- Osteogenesis imperfecta
- Third window lesions

Treatment

Medical

- Fluoridation of drinking water
- Oral fluorides
  - 20 – 120 mg /day (Shambaug)
  - Fluoride ions replace hydroxyl ions → fluoroapatite instead of hydroxyl apatite → more stable & resistant to osteoclastic activity

Both are controversial. Researches are on but no evidence at present of benefit in Otosclerosis.

Hearing Aids

- Conventional hearing aids
- BAHA

Roles

- Primary treatment
  - Hearing thresholds comparable to those obtained from surgery
  - Factors considered
    - Attitude to risk for surgery
    - Aesthetic and comfort considerations
    - Hearing specific functional status
    - Quality of life
- In combination with surgery in far advanced Otosclerosis
- During post stapedectomy rehabilitation- improves word recognition score
- As a rescue treatment many years after surgery
Surgery

- Stapedectomy
- Partial Stapedectomy
- Stapedotomy
- Stapes footplate mobilization
- Revision Surgery

Before surgery is considered, the risks, complications, and alternatives are thoroughly discussed with the patient.

Indications for surgery
- AC threshold 30 dB or worse
- AB gap >20 dB
- Speech Discrimination score >60%

Contraindications
- Only hearing ear
- Associated meniere’s disease
- Young children
- Professional athletes, high construction workers, divers, frequent air travelers
- Those who work in noisy surroundings
- Otitis externa
- Positive schwartzze’s sign

Although the risk of sensorineural hearing loss is low, the patient needs to be informed of this possibility.

Total sensorineural hearing loss occurs in about 0.2% of cases, but the patient is told that there is a less than 2% chance of further hearing loss and a less than 1% chance of losing all hearing in the operated ear.

The slight chance that dizziness may occur postoperatively also is discussed.

Dizziness is usually transient and brief but may persist, and in extremely rare cases could be permanent.

The possibility of facial palsy is discussed.

Surgery is usually performed with the patient under local anesthesia.

Local anesthesia is preferred because:
- An awake patient can inform the surgeon if vertigo occurs
- An intra operative assessment of improvement in hearing can be done
- The surgery is usually of short duration with minimal discomfort and does not warrant the risks or costs of general anesthesia.

Steps of Stapedectomy
- TM flap raised
- Care is taken during elevation to identify the chorda tympani nerve so that it is not stretched or injured.
- The superior elevation is carried forward to the malleus.
- Care is taken not to dislocate the incus.
- Usually the stapes is not entirely visualized because of the bony postsuperior external auditory canal overhang.
- This overhang is removed with a curette, or some prefer a microdrill.
- The exposure is adequate when the facial nerve can be visualized superiorly and the pyramidal process posteriorly.
- Once adequate exposure is established, the malleus, incus, and stapes are palpated to ascertain mobility.
- The distance from the incus to the stapes footplate is measured.
- The usual distance from the lateral surface of the incus to the foot-plate is 4.5 mm.
- Because the piston prosthesis is usually measured from the medial surface of the incus, 0.25 mm is added to allow for this distance and to allow for a small extension into the vestibule.
- Usually 4.5 mm piston used in males and 4.25 in females.
- 0.7-mm diamond microdrill is used to create the fenestra.
- The fenestra is created with a light touch of the drill in post 1/3rd of footplate.
- The 4.25-mm or 4.5mm length, 0.6-mm diameter platinum polytef prosthesis is placed from the incus to the fenestra.
- It is crimped firmly on the incus.
- The incudostapedial joint is separated, and the stapedial tendon is sectioned.
- The superstructure of the stapes is fractured inferiorly with a quick, decisive movement and removed.
- A test of the prosthesis is performed.
- Blood, fat or a connective tissue graft is then placed around the footplate perforation in an attempt to seal it, if a vein graft has not been placed earlier over the stapedotomy.
- TM flap is replaced.

External ear canal is packed.

Post-operative complications
- Conductive hearing loss
  - Displacement of prosthesis out of footplate
  - Bony re-growth over fenestration
  - Necrosis of long process of incus
  - Fibrous adhesions fixing ossicles
- SNHL
  - Surgical trauma, extensive drilling around footplate
  - Floating footplate
  - Perilymph or blood aspiration into oval window
  - Congenital footplate fixation
CSF Gushers
  - Delayed SNHL
    - Barotrauma from air travel
    - Blast injury
    - Reparative granuloma
    - Perilymph fistula
    - Suppurative labyrinthitis
    - Influenza
  - Facial palsy
  - Vertigo
  - Perilymphatic fistula
  - Reparative granuloma
  - Discomfort to loud noise
  - Alteration in loud noise & taste – injury to chorda tympani
  - Cholesteatoma
  - Meningitis

OUTCOME EVALUATION
- Glasgow benefit plot
- Belfast rule of thumb
- Amsterdam hearing evaluation plots
- Sofia profile plot

Other treatment techniques / Recent Advances
- Microdrill / Microperforator / Laser In Stapedotomy
- Malleus handle(malleus grip) stapedectomy
- Smaller diameter fenestration with no use of connective seals
- Heavier piston for low frequency hearing loss
- Lighter piston for high frequency hearing loss
- Preservation of stapedius tendon to improve blood supply to incus
- Cochlear implantation
- Fibreoptic micro-handpiece
- Robot-assisted stapedotomy

REVISION SURGERY
- Poorer hearing outcomes
- Increased risk of complications – permanent SNHL
- Use of Laser advocated in
  - Clearing of adhesions in oval window area
  - To free ankylosed prosthesis from incus
  - Sculpting incus
- Malleostapedotomy
  - Incus erosion
  - Fixation of malleus & incus
SECOND EAR STAPEDECTOMY

- Prognostic factors
- Interval between surgeries 1 yr
- Contraindications for second ear stapedectomy
  - Complications following first surgery
  - Tinnitus and / or persistent vertigo following first surgery

FLOATING FOOTPLATE  
(D’Souza & Glasscock)

- Footplate is not properly fixed and becomes mobile (though still hinged to OW → if this contact is also lost then known as submerged footplate)
  - Incidental finding
  - Most commonly occurs during stapedectomy when fenestra is placed in centre (instead of posterior 1/3rd)
- Prevention
  - Creating a control hole in footplate before removing arch of stapes
  - Laser
- Management
  - Tissue (vein) grafting & prosthesis over graft (Lippy)
    - If footplate blue → success 95%
    - If footplate white or biscuit → success 52%
  - Graft, allowing re-fixation for 4 months → re operate with laser & vein clad prosthesis (Perkins) → if recurrence during revision → abandon
  - Laser fenestration & piston implantation
  - Extraction → create a pothole in promontory near footplate → then lift and remove the footplate (House)

OR
- Abort procedure

Obliterative Otosclerosis  
(D’Souza & Glasscock)

- Total obstruction of oval window & it’s margins
- Evidence of very advanced otosclerosis
- Young patients (10 -15 yrs), when active otosclerosis develops around puberty
- More in males than females (as opposed to otosclerosis)
- Bilateral (Uncommon in unilateral otosclerosis)
- Uncommon in RW but if present is associated with SNHL
- Pathology
  - Same as otosclerosis
  - Early stages in fissula ante fenestrum → disease advances → stapes suprastructure & crura eventually buried in extension of otosclerotic bone → Bone thickness 0.5 to 2 mm
- Incidence 7 to 11 % (Shambaug)
- Types
  - Truly obliterated footplate → Boundaries of footplate can’t be identified
  - Partly obliterated footplate → Rim of delineation can be made out
o Spuriously obliterated footplate → surrounding rim in presence of solid thick footplate
o Biscuit footplate → Well delineated rim & intact annular ligament → prone for floating footplate

- Parameters for diagnosing obliterative footplate before surgery
  1. Hearing impairment in teenage & young adults
  2. Positive Schwartz’s sign
  3. Bilateral rapidly decreasing hearing
  4. “As” type curve on tympanometry
  5. CT scan findings of obilitation of footplate

- Management
  - Sodium fluoride
  - Surgery
    - Small fenestra with long thin piston
    - Large fenestra till blue lined vestibule is left
    - Laser large fenestra

- Results
  1. Chances of SNHL higher
  2. High chances of re closure → new bone is not otosclerotic but reparative → options are revision laser stapedectomy or NaF or Hearing aids → re drilling with instruments has very high chances of SNHL

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