Otosclerosis

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Introduction

- **Otosclerosis**
  - Primary metabolic bone disease of the otic capsule and ossicles
  - Results in fixation of the ossicles and conductive hearing loss
  - May have sensorineural component if the cochlea is involved
  - Genetically mediated
    - Autosomal dominant with incomplete penetrance (40%) and variable expressivity
History of Otosclerosis and Stapes Surgery

- 1704 – Valsalva first described stapes fixation.
- 1857 – Toynbee linked stapes fixation to hearing loss.
- 1890 – Katz was first to find microscopic evidence of otosclerosis.
- 1893 – Politzer described the clinical entity of “otosclerosis.”
- 1890 – Bacon describes medical therapy for the condition, and supports the common view that “surgery should not be considered for a moment.”
History of Otosclerosis and Stapes Surgery

- Gunnar Holmgren (1923)
  - Father of fenestration surgery
  - Single stage technique

- Sourdille
  - Holmgren’s student
  - 3 stage procedure
  - 64% satisfactory results
History of Otosclerosis and Stapes Surgery

- **Julius Lempert**
  - Popularized the single staged fenestration procedure

- **John House**
  - Further refined the procedure
    - Popularized blue lining the horizontal canal
History of Otosclerosis and Stapes Surgery

- Fenestration procedure for otosclerosis
  - Fenestration in the horizontal canal with a tissue graft covering
  - >2% profound SNHL
  - Rarely complete closure of the ABG
  - May exhibit vestibular disturbances
History of Otosclerosis and Stapes Surgery

- **Samuel Rosen**
  - 1953 – first suggest mobilization of the stapes
    - Immediate improved hearing
    - Re-fixation
History of Otosclerosis and Stapes Surgery

- John Shea
  - 1956 – first to perform stapedectomy
    - Oval window vein graft
    - Nylon prosthesis from incus to oval window
Epidemiology

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

<table>
<thead>
<tr>
<th>Race</th>
<th>Incidence of otosclerosis</th>
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<tbody>
<tr>
<td>Caucasian</td>
<td>10%</td>
</tr>
<tr>
<td>Asian</td>
<td>5%</td>
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<tr>
<td>African American</td>
<td>1%</td>
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<tr>
<td>Native American</td>
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Epidemiology

- **Gender**
  - Histologic otosclerosis – 1:1 ratio
  - Clinical otosclerosis – 2:1 (W:M)
    - Possible progression during pregnancy (10%-17%)
      - Studies which have demonstrated changes during pregnancy are often retrospective or lack audiometric data.
      - Studies comparing multigravid vs. nulligravid women with otosclerosis have failed to show audiometric differences.
    - Bilaterality more common (89% vs. 65%)
Epidemiology

- **Age**
  - 15-45 most common age range of presentation
  - Youngest presentation 7 years
  - Oldest presentation 50s
  - 0.6% of individuals <5 years old have foci of otosclerosis
Pathophysiology

- Osseous dyscrasia
  - Resorption and formation of new bone
  - Limited to the temporal bone and ossicles
  - Inciting event unknown
    - Hereditary, endocrine, metabolic, infectious, vascular, autoimmune, hormonal
Two phases of disease

Active (otospongiosis phase)
- Osteocytes, histiocytes, osteoblasts
- Active resorption of bone
- Dilation of vessels
  - Schwartzze’s sign

Mature (sclerotic phase)
- Deposition of new bone (sclerotic and less dense than normal bone)
Pathology

- Most common sites of involvement
  - Fissula ante fenestrum
  - Round window niche (30%-50% of cases)
  - Anterior wall of the IAC
Non-clinical foci of otosclerosis
Anterior footplate involvement
Annular ligament involvement
- Incudostapedial Joint
- Otosclerotic Bone
- Footplate
- Saccule
Bipolar involvement of the footplate

Cochleariform Process

Otosclerotic Lesion in Footplate of Stapes
Round Window

- Round Window Membrane
- Otosclerosis
- Internal Auditory Canal
- Posterior Ampulla
Labyrinthine Otosclerosis

1912 – Siebenmann described labyrinthine otosclerosis

- Suggested otosclerosis may cause SNHL via
  - Toxic metabolites
  - Decreased blood supply
  - Direct extension
  - Disruption of membranes
Hyalinization of the spiral ligament
Erosion into inner ear
Organ of Corti

- Otosclerotic Bone
- Degenerated Organ of Corti
- Spiral Prominence
- Severe Atrophy of Spiral Ligament
Cochlear Otosclerosis

- Audiometric studies
  - Some studies have shown that in cases of unilateral otosclerosis ~ 60% may have decreased sensory thresholds even after stapes surgery

- Histologic studies
  - Cases of documented otosclerosis and a large sensory loss have shown large foci of otosclerosis in the otic capsule.
  - Many cases of large otic capsule foci without sensory loss or of sensory loss without foci have also been described.

- Biochemical studies
  - Some authors have noted increased levels of perilymph protein during stapedotomy in patients with radiographic evidence of otic capsule foci and sensory hearing loss.

- Conclusion
  - Many experts believe that extensive involvement of the cochlea will produce sensorineural hearing deficits, although it is not known how this occurs or why it only occurs in a subset of patients with cochlear foci.
Diagnosis of Otosclerosis
History

- Most common presentation
  - Women age 20 - 30
  - Conductive or Mixed hearing loss
    - Slowly progressive,
    - Bilateral (80%)
    - Asymmetric
  - Tinnitus (75%)
History

- Age of onset of hearing loss
- Progression
- Laterality
- Associated symptoms
  - Dizziness
  - Otalgia
  - Otorrhea
  - Tinnitus
History

- **Family history**
  - 2/3 have a significant family history
  - Particularly helpful in patients with severe or profound mixed hearing loss

- **Prior otologic surgery**

- **History of ear infections**

- **Vestibular symptoms**
  - 25%
  - Most commonly dysequilibrium
  - Occasionally attacks of vertigo with rotatory nystagmus
Physical Exam

- **Otomicroscopy**
  - Most helpful in ruling out other disorders
    - Middle ear effusions
    - Tympanosclerotic
    - Tympanic membrane perforations
    - Cholesteatoma or retraction pockets
    - Superior semicircular canal dehiscence
  - **Schwartz’s sign**
    - Red hue in oval window niche area
    - 10% of cases

- **Pneumatic otoscopy**
  - Distinguish from malleus fixation
Physical Exam

- **Tuning forks**
  - Hearing loss progresses from low frequencies to high frequencies
  - 256, 512, and 1024 Hz TF should be used
    - **Rinne**
      - 256 Hz – negative test indicates at least a 20 dB ABG
      - 512 Hz – negative test indicates at least a 25 dB ABG
Differential Diagnosis

- Ossicular discontinuity
- Congenital stapes fixation
- Malleus head fixation
- Paget’s disease
- Osteogenesis imperfecta
- Superior semicircular canal dehiscence
Audiometry

- Tympanometry
- Impedance testing
  - Acoustic reflexes
- Pure tones
Tympanometry

- Jerger (1970) – classification of tympanograms
  - Type A
    - Type A
    - Type As
    - Type Ad
  - Type B
  - Type C
Acoustic Reflexes

- Result from a change in the middle ear compliance in response to a sound stimulus

- Change in compliance
  - Stapedius muscle contraction
  - Stiffening of the ossicular chain
  - Reduces the sound transmission to the vestibule
Acoustic Reflexes

- Otosclerosis has a predictable pattern of abnormal reflexes over time
  - Reduced reflex amplitude
  - Elevation of ipsilateral thresholds
  - Elevation of contralateral thresholds
  - Absence of reflexes
Pure Tone Audiometry

- Most useful audiometric test for otosclerosis
  - Characterizes the severity of disease
  - Frequency specific

- Carhart’s notch
  - Hallmark audiologic sign of otosclerosis
  - Decrease in bone conduction thresholds
    - 5 dB at 500 Hz
    - 10 dB at 1000 Hz
    - 15 dB at 2000 Hz
    - 5 dB at 4000 Hz
Pure Tone Audiometry

- Low frequencies affected first
  - Below 1000 Hz

- Rising air line
  - “Stiffness tilt”
  - Secondary to stapes fixation

- With disease progression
  - Air line flattens
Pure Tone Audiometry

- Carhart’s notch
  - Proposed theory
    - Stapes fixation disrupts the normal ossicular resonance (2000 Hz)
    - Normal compressional mode of bone conduction is disturbed because of relative perilymph immobility
  - Mechanical artifact
  - Reverses with stapes mobilization
Pure Tone Audiometry

- Committee on Hearing and Balance
  - Set standards for reporting results in cases of otosclerosis procedures.
    - Operative hearing results should be reported using post-operative data, specifically, the post-operative air-bone gap.
    - This prevents exaggeration of surgical results and “overclosure.”
  - Adopted by the AAOHNS in 1994
  - Important in reviewing literature regarding surgical outcomes
    - Studies prior to this time often use pre-op bone lines and post-op air conduction measurements which may exaggerate results.
    - This convention is not uniform in all parts of the world, so the methods is very important in determining the consistency of data.
Imaging

- Computed tomography (CT) of the temporal bone

  - Proponents of CT for evaluation of otosclerosis
    - Pre-op
      - Characterize the extent of otosclerosis
      - Severe or profound mixed hearing loss
      - Evaluate for enlarged cochlear aqueduct
    - Post-op
      - Recurrent CHL
        - Re-obliteration vs. prosthesis dislocation
        - Vertigo
“Halo sign”
Paget’s disease
Osteogenesis Imperfecta
Management Options

- Medical
- Amplification
- Surgery
- Combinations
Patient Selection

Factors

- Result of tuning fork tests and audiometry
- Skill of the surgeon
- Facilities
- Medical condition of the patient
- Patient wishes
Surgery

- Best surgical candidate
  - Previously un-operated ear
  - Good health
  - Unacceptable ABG
    - 25 to 40 dB
    - Negative Rinne test
  - Excellent discrimination
  - Desire for surgery
Surgery

- Other factors
  - Age of the patient
    - Elderly
      - Poorer results in the high frequencies
    - Congenital stapes fixation (44% success rate)
    - Juvenile otosclerosis (82% success rate)
  - Occupation
    - Diver
    - Pilot
    - Airline steward/stewardess
Surgery

- Other factors
  - Vestibular symptoms
    - Meniere's disease
  - Concomitant otologic disease
    - Cholesteatoma
    - Tympanic membrane perforation
Surgical Steps

- Subtleties of technique and style
  - Local vs. general anesthesia
  - Stapedectomy vs. partial stapedectomy vs. stapedotomy
  - Laser vs. drill vs. cold instrumentation
  - Oval window seals
  - Prosthesis
Canal Injection

- 2-3 cc of 1% lidocaine with 1:50,000 or 1:100,000 epinephrine
- 4 quadrants
- Bony cartilaginous junction
Raise Tympanomeatal Flap

- 6 and 12 o’clock positions
- 6-8 mm lateral to the annulus
- Take into account curettage of the scutum
Separation of chorda tympani nerve from malleus

- Separate the chorda from the medial surface of the malleus to gain slack
- Avoid stretching the nerve
- Cut the nerve rather than stretch it
Curettage of Scutum

- Curettage a trough lateral to the scutum, thinning it
- Then remove the scutum (incus to the round window)
Curettage of Scutum

- **Exposure**
  - **Vertical:**
    - Facial nerve to round window
  - **Horizontal:**
    - Pyramidal process to malleus

- **Preservation of bone over incus**
Middle ear examination

- Mobility of ossicles
  - Confirm stapes fixation
  - Evaluate for malleus or incus fixation

- Abnormal anatomy
  - Dehiscent facial nerve
  - Overhanging facial nerve
  - Deep narrow oval window niche
  - Ossicular abnormalities
Measurement for prosthesis

- Measurement
  - Lateral aspect of the long process of the incus to the footplate
Total Stapedectomy

**Uses**
- Extensive fixation of the footplate
- Floating footplate

**Disadvantages**
- Increased post-op vestibular symptoms
- More technically difficult
- Increased potential for prosthesis migration
Stapedotomy/Small Fenestra

- Originally for obliterated or solid footplates
  - Europe
  - 1970-80

- First laser stapedotomy performed by Perkins in 1978
  - Less trauma to the vestibule
  - Less incidence of prosthesis migration
  - Less fixation of prosthesis by scar tissue
Drill Fenestration

- 0.7mm diamond burr
  - Motion of the burr removes bone dust
  - Avoids smoke production
  - Avoids surrounding heat production
Laser Fenestration

- **Laser**
  - Avoids manipulation of the footplate
  - Argon and Potassium titanyl phosphate (KTP/532)
    - Wave length 500 nm
    - Visible light
    - Absorbed by hemoglobin
    - Surgical and aiming beam
  - Carbon dioxide (CO2)
    - 10,000 nm
    - Not in visible light range
    - Surgical beam only
      - Requires separate laser for an aiming beam (red helium-neon)
    - Ill defined fuzzy beam
Oval window seal

- Tragal perichondrium
- Vein (hand or wrist)
- Temporalis fascia
- Blood
- Fat
- Gelfoam (now discouraged)
Reconstructing the annular ligament
Placement of the Prosthesis

- Prosthesis is chosen and length picked
- Some prefer bucket handle to incorporate the lenticular process of the incus
Stapedectomy vs. Stapedotomy

- ABG closure < 10dB (PTA)
Special Considerations and Complications in Stapes Surgery
Overhanging Facial Nerve

- Usually dehiscent
- Consider aborting the procedure
- Facial nerve displacement (Perkins, 2001)
  - Facial nerve is compressed superiorly with No. 24 suction (5 second periods)
  - 10-15 sec delay between compressions
  - Perkins describes laser stapedotomy while nerve is compressed
- Wire piston used
  - Add 0.5 to 0.75 mm to accommodate curve around the nerve
Floating Footplate

- Footplate dislodges from the surrounding OW niche
  - Incidental finding
  - More commonly iatrogenic

- Prevention
  - Laser
  - Footplate control hole

- Management
  - Abort
  - H. House favors promontory fenestration and total stapedectomy
  - Perkins favors laser fenestration
Diffuse Obliterative Otosclerosis

- Occurs when the footplate, annular ligament, and oval window niche are involved
- Closure of air-bone gap < 10 dB less common.
- Refixation commonly occurs
Perilymphatic Gusher

- Associated with patent cochlear aqueduct
- More common on the left
- Increased incidence with congenital stapes fixation
- Increases risk of SNHL

Management
  - Rough up the footplate
  - Rapid placement of the OW seal then the prosthesis
  - HOB elevated, stool softeners, bed rest, avoid Valsalva, +/- lumbar drain
Round Window Closure

- 20%-50% of cases
- 1% completely closed
- No effect on hearing unless 100% closed
- Opening has a high rate of SNHL
SNHL

- 1%-3% incidence of profound permanent SNHL
  - Surgeon experience
  - Extent of disease
    - Cochlear
  - Prior stapes surgery

- Temporary
  - Serous labyrinthitis
  - Reparative granuloma

- Permanent
  - Suppurative labyrinthitis
  - Extensive drilling
  - Basilar membrane breaks
  - Vascular compromise
  - Sudden drop in perilymph pressure
Reparative Granuloma

- Granuloma formation around the prosthesis and incus
- 2-3 weeks postop
- Initial good hearing results followed by an increase in the high frequency bone line thresholds
- Associated tinnitus and vertigo
- Exam – reddish discoloration of the posterior TM
- Treatment
  - ME exploration
  - Removal of granuloma
- Prognosis – return of hearing with early excision
- Associated with use of Gelfoam
Vertigo

- Most commonly short lived (2-3 days)
- More prolonged after stapedectomy compared to stapedotomy
  - Due to serous labyrinthitis
- Medialization of the prosthesis into the vestibule
  - With or without perilymphatic fistula
- Reparative granuloma
Recurrent Conductive Hearing Loss

- Slippage or displacement of the prosthesis
  - Most common cause of failure
  - Immediate
    - Technique
    - Trauma
  - Delayed
    - Slippage from incus narrowing or erosion
    - Adherence to edge of OW niche
    - Stapes re-fixation
    - Progression of disease with re-obliteration of OW
    - Malleus or incus ankylosis
Amplification

- Excellent alternative
  - Non-surgical candidates
  - Patients who do not desire surgery

- Patient satisfaction rate lower than that of successful surgery
  - Canal occlusion effect
  - Amplification not used at night
Medical

**Sodium Fluoride**

- 1923 - Escot suggested using calcium fluoride
- 1965 – Shambaugh popularized its use

**Mechanism**

- Fluoride ion replaces hydroxyl group in bone forming fluorapatite
- Resistant to resorption
- Increases calcification of new bone
- Causes maturation of active foci of otosclerosis
Medical

- **Sodium Fluoride**
  - Reduces tinnitus, reverses Schwartze’s sign, resolution of otospongiosis seen on CT
  - OTC – Florical
  - Dose – 20-120mg
  - Indications
    - Non-surgical candidates
    - Patients who do not want surgery
    - Surgical candidates with + Schwartze’s sign
      - Treat for 6 mo pre-op
      - Postop if otospongiosis detected intra-op
Medical

- Sodium fluoride
  - Hearing results
    - 50% stabilize
    - 30% improve
  - Re-evaluate q 2 yrs with CT and for Schwartze’s sign to resolve
  - If fluoride are stopped – expect re-activation within 2-3 years
Medical

- **Bisphosphonates**
  - Class of medications that inhibits bone resorption by inhibiting osteoclastic activity
  - Dosing not standard
  - Often supplement with Vitamin D and Calcium
  - Studies conducted on otosclerosis patients with neurotologic symptoms report the majority of patients with subjective improvement or resolution.
  - Future application of this treatment unclear, especially with new reports of bisphosphonate related osteonecrosis.
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