Otosclerosis

Christopher Muller, M.D.
Faculty Advisor: Arun Gadre, M.D.
The University of Texas Medical Branch
Department of Otolaryngology
Galveston, Texas
Grand Rounds Presentation
June 4, 2003
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”
History of Otosclerosis and Stapes Surgery

- Gunnar Holmgren
  - 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
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>
</tr>
</thead>
<tbody>
<tr>
<td>Caucasian</td>
<td>10%</td>
</tr>
<tr>
<td>Asian</td>
<td>5%</td>
</tr>
<tr>
<td>African American</td>
<td>1%</td>
</tr>
<tr>
<td>Native American</td>
<td>0%</td>
</tr>
</tbody>
</table>
Epidemiology

- Gender
  - Histologic otosclerosis – 1:1 ratio
  - Clinical otosclerosis – 2:1 (W:M)
    - Increase progression during pregnancy (10%-17%)
    - 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
Pathophysiology

- Siebenmann – first to describe the microscopic appearance
  - Spongy
  - Usually limited to the anterior footplate
Pathology

- 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)
“Blue mantles of Manasseh”
Pathology

- Most common sites of involvement
  - Fissula ante fenestrum
  - Round window niche (30%-50% of cases)
  - Anterior wall of the IAC
Fissula ante and post fenestrum
Fissula ante fenestrum
Non-clinical foci of otosclerosis
Annular ligament involvement
Footplate Involvement
Anterior footplate involvement
Bipolar involvement of the footplate
Round Window

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

- 1912 – Siebenmann described labyrinthine otosclerosis
  - Suggested otosclerosis may cause SNHL
    - Toxic metabolites
    - Decreased blood supply
    - Direct extension
Hyalinization of the spiral ligament
Erosion into inner ear
Diagnosis
History

- Most common presentation
  - Women in her 20s or 30s
  - 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

- Vestibular symptoms
  - 25%
  - Most commonly dysequilibrium
  - Occasionally attacks of vertigo with rotatory nystagmus
- Prior otologic surgery
- History of ear infections
History

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

- **Ototoxicscopy**
  - Most helpful in ruling out other disorders
    - Middle ear effusions
    - Tympanosclerosis
    - Tympanic membrane perforations
    - Cholesteatoma or retraction pockets
  - Schwartzte’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
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
  - Diphasic reflex pattern
  - Reduced reflex amplitude
  - Elevation of ipsilateral thresholds
  - Elevation of contralateral thresholds
  - Absence of reflexes
Acoustic Reflexes

1. Normal
   Compliance
   Stimulus

2. Mixed
   Compliance
   Stimulus

3. Diphasic
   Compliance
   Stimulus

4. Fixed
   Compliance
   Stimulus
Pure Tone Audiometry

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

- Low frequencies affected first
  - Below 1000 Hz
- Rising air line
  - “Stiffness tilt”
  - Secondary to stapes fixation
Pure Tone Audiometry

- With disease progression
  - Air line flattens
    - Secondary to mass effect
Pure Tone Audiometry

- 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

- 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
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 enlarge cochlear aqueduct
    - Post-op
      - Recurrent CHL
        - Re-obliteration vs. prosthesis dislocation
        - Vertigo
Imaging

- **CT**
  - **Axial cuts**
    - Patient position – canthomeatal line perpendicular to the table top
    - 1 mm cuts
    - Top of sup. SCC to bottom of the cochlea
  - **Coronal**
    - Patient position – supine w/ head overextended face turned 20 degrees ipsilateral
“Halo sign”
Paget’s disease
Osteogenesis Imperfecta
Natural history of otosclerosis

- 90% of all cases are never clinically apparent
- Foci begins in childhood
- Most commonly becomes symptomatic in the 3rd and 4th decades
- After clinical presentation
  - Conductive hearing loss progressive
  - Periods of quiescence and deterioration
  - Worsening tinnitus
  - Associated SNHL (rarely purely SN)
- Matures by age 50-70 with max. CHL of 50 dB
Management

- Medical – Sodium Fluoride
- Amplification
- Surgery
- Combinations
Patient Selection

- Factors
  - Result of TF tests and audiometry
  - Skill of the surgeon
  - Facilities
  - Medical condition of the patient
  - Patient wishes
Patient Counseling

- Options for treatment
  - Advantages and disadvantages of each
- Repeat clinic visit
Surgery

- Best surgical candidate
  - Previously un-operated ear
  - Good health
  - Unacceptable ABG
    - 25 to 40 dB, bilateral ABG recommended by different authorities
    - 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
Endolymphatic Hydrops
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
Pre-op

- Confirm the correct ear (largest ABG)
  - With the patient
  - Audiogram
  - History and physical exam
- Place CT and audiogram in a visible location in the OR for easy intra-operative evaluation
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)
- Visualize the pyramidal process and facial n.
Curettage of Scutum

- Exposure of pyramidal process and facial n.
- 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
Measurement for prosthesis

- Measurement
  - Lateral aspect of the long process of the incus to the footplate
- Average 4.5 mm
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
- Advantages
  - 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
Fenestration

- Causse et al. (1993)
  - Recommends posteriorly placed fenestration to better recreate the natural physiologic dynamics of the footplate
Pivoting stapes
Energy transmission to the stapes
Posterior Fenestration

- Posteriorly placed fenestration with the laser
- Causse also recommends following the laser with the diamond burr to remove char
Oval window seal

- Tragal perichondrium
- Vein (hand or wrist)
- Temporalis fascia
- Blood
- Fat
Vein graft
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)
Stapedectomy vs. Stapedotomy

- ABG closure at 4 kHz
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
Floating Footplate

- Hearing results
  - Thin or blue footplate – 97% ABG closure (<10dB)
  - White or “biscuit” footplate – 52% ABG closure
**Diffuse Obliterative Otosclerosis**

- Occurs when the footplate, annular ligament, and oval window niche are involved
  - Bone is thinned with a small cutting burr
  - Blue lined at anteroposterior edges first
Perilymphatic Gusher

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

Management

- Ruff 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
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
- Satisfaction rate less than with successful Sx
  - 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
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
- **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
References

- Lempert J. Improvement in hearing in cases of otosclerosis: A new, one stage surgical technique. *Arch Otolaryngol* 1938;28:42-97
- Shambaugh G. Clinical diagnosis of cochlear (labyrinthine) otosclerosis. *Laryngoscope* 1965;75:1558-1562
- Reference