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Otosclerosis – A Disease of Bone Remodeling

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Collaborative Multi-Institutional Otolaryngology Residency Education Program

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Learning objectives

- Understand the pathophysiology of Otosclerosis
- Differential Diagnosis
- Treatment Options
- Thoughts for the future

References
Cummings – chapter 156
The human temporal bone consortium
Surgery Of The Ear And Temporal Bone by Nadol Joseph B Jr, chapter 22
Journal articles (cited en route)
UTMB website

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Introduction

- The normal human otic capsule remodeling rate is extremely slow (<2%, compared with 10% of other bones).
- In otosclerosis normal inhibition of bone remodeling is lost resulting in foci of bone remodeling.
- When remodeled bone bridges the stapedio-vestibular joint, it fixates the joint and impedes sound transmission manifested as conductive hearing loss.
- Sensorineural hearing loss can occur when bone remodeling extends to the cochlea.

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Historical Review

1735-Valsalva- found a stapes fixed to an oval window niche during autopsy of a deaf patient

1857- Toynbee- stated after 1659 ear dissections that "osseous ankylosis of the stapes to the fenestra ovalis was one of the common causes of deafness."

1873- Schwartze- described the **reddish discoloration** of the tympanic membrane, later found to be increased vascularity of the cochlear promontory during the active stage of otosclerosis. This later became known as **Schwartze's sign**, and occurs in 10% of otosclerosis ears.

1881- von Troltsch- used the term "sclerosis" for stapes fixation. He thought that sclerosis of the tympanic cavity mucosa caused stapes fixation.

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Historical Review

1890- Katz- first to find **microscopic evidence of otosclerosis resulting in stapes fixation**.

1893- Politzer- studied the temporal bones of 16 patients whom he had followed while alive with stapes fixation, and **determined that stapes fixation was a primary disease of the otic capsule**, rather than a result of repeated inflammation or infection.

1908- Bezold - described the typical history and physical findings, as well as audiometric findings of otosclerosis.

1912- Siebenmann- found that the **diseased bone of otosclerosis is more porous and less dense** than the normal otic capsule, and suggested that the disease be more accurately termed "otospongiosis." He also postulated that this disease may cause sensorineural hearing loss.

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Gross Pathology

- **Active foci of disease are covered with a thickened vascular mucosa resulting in** "Schwartze's sign".
- **Otosclerotic foci**, specifically if mature, have a white color rather than the yellow hue of the temporal bone.

Location of disease:
- The usual site of onset, or site of predilection is just anterior to the **oval window niche: known as the fissula ante fenestrum** (**80-90%** of temporal bones with evidence of disease).
- Round window niche (30-50%).
  Complete obliteration of the niche is quite rare(1-3%).

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Microscopic Pathology

- Immature/active foci: **a porous spongy structure of irregular bony lamellae** separated by wide vascular spaces containing an abundance of histiocytes, osteoblasts, and some osteoclasts. These lesions have an affinity for hematoxylin, which makes **the bone appear darker**.
- Mature foci: relatively acellular, with narrower vascular spaces and smaller blood vessels.
- Various levels of activity may be seen among foci in the same ear.
- Otosclerosis has been shown by Schuknecht to **conform to the original endochondral bone** which it replaces. For example, the fallopian canal wall is frequently involved, but the canal lumen is never compromised, and facial weakness is not seen.

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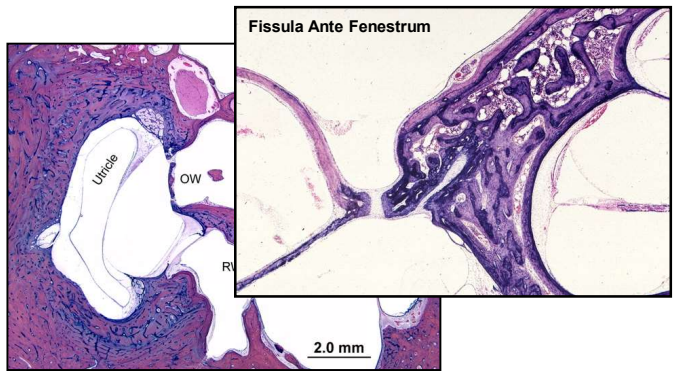
Normal temporal bone histology



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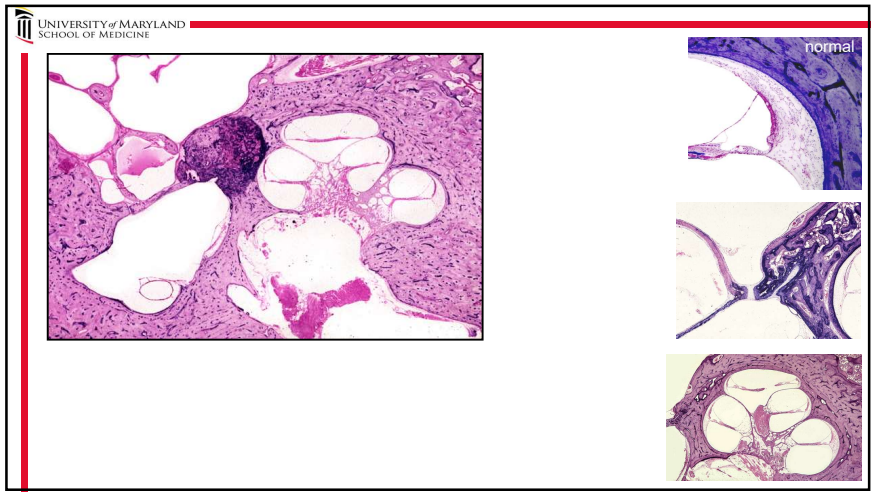
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normal

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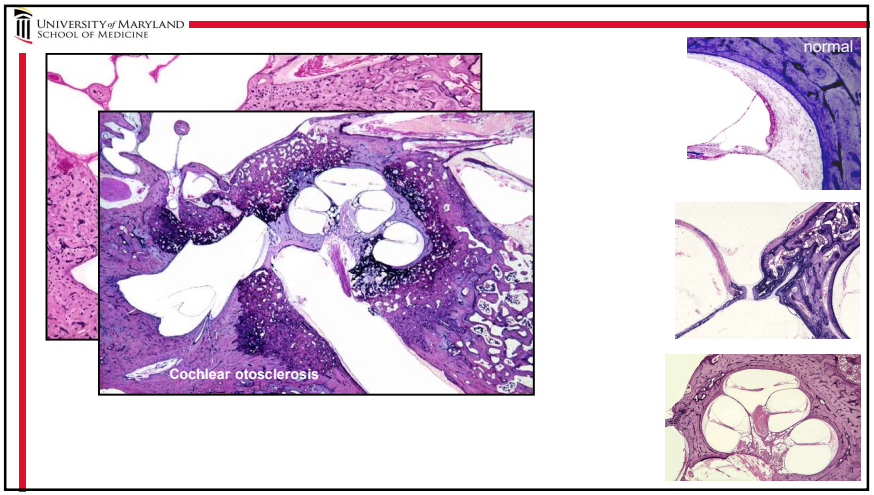
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normal
Cochlear otosclerosis

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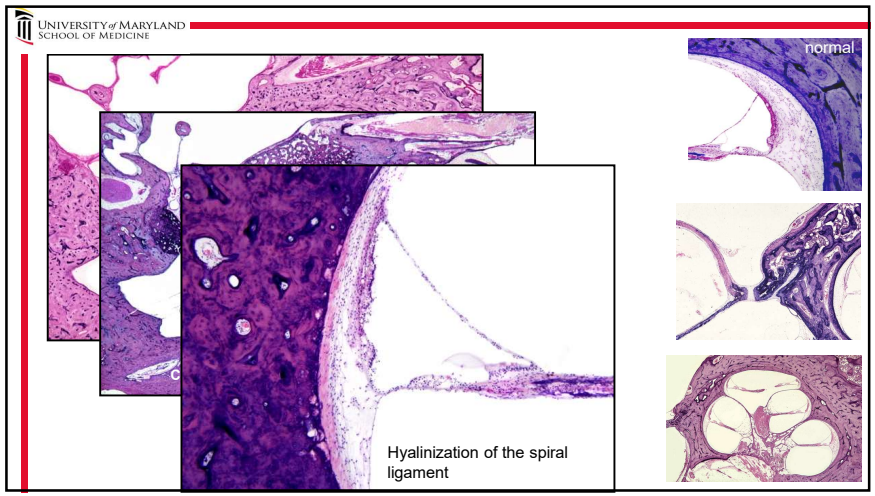
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normal
Hyalinization of the spiral ligament

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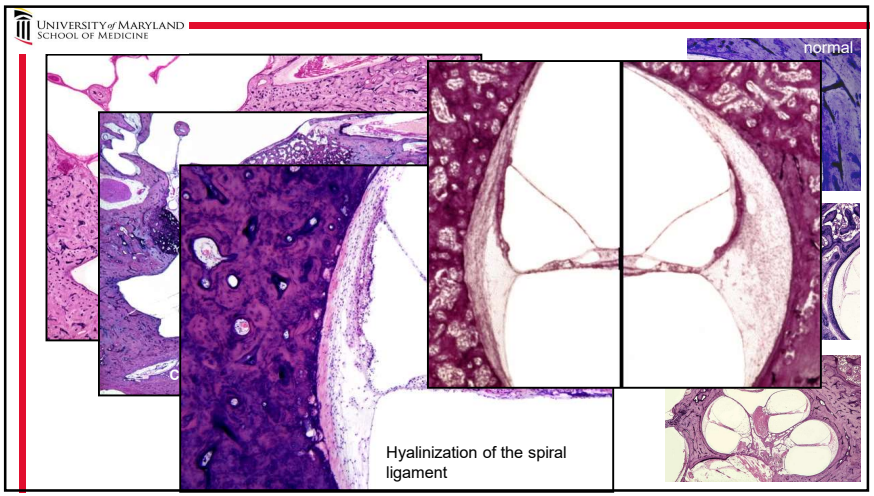
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normal
Hyalinization of the spiral ligament

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Epidemiology (10%m:1%c)

- **In Caucasians: otosclerotic lesions can be found in 7.3% and 10.3% of temporal bones** of men and women respectively. The stapes will be fixed in only 12.3% of those with histopathologic evidence of otosclerosis.

  → **the clinical prevalence is 0.3-0.5%.** Clinical otosclerosis is 1.4-2 higher in women than in men (sex difference under debate).

- Clinical otosclerosis is rare in blacks, Asians, and Native Americans.

- **Otosclerosis eventually involves both ears in 85-90% of patients** (but clinically bilateral in women>men).

Adv Otorhinolaryngol. *2007;65:6-16*

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Age

- The incidence of clinical otosclerosis increases with age. **Microscopic** evidence of otospongiosis:
  - 0.6% of autopsies of individuals less than 5 years old.
  - By late middle age, 10% in white men and nearly 20% in white women.

- **Clinical onset is usually between 15 and 35 years of age**, but the disease may manifest itself as early as age 6 or 7, and as late as the mid 50's. Higher activity is more prevalent in those less than 50 years old, and very low activity is common after 70 years of age.

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Causes / Pathophysiology

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Genetics - Inheritance

- A complex disease with some mongenic forms. In these cases, the disease is often autosomal dominant with variable penetrance (25-40%). Can be sporadic (40-50%).

- Difficulty in cloning genes underlying otosclerosis as only 10% of otosclerosis becomes clinically apparent. True inheritance may not be known until epidemiological studies incorporate microscopic evaluation.

Otolaryngol Clin North Am. 2018 Apr;51(2):305-318

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Table 1
Genetic loci contributing to familial cases of otosclerosis with mendelian segregation identified using family linkage studies

Locus	Position	Family Countries of Origin
OTSC1	15q25–26	South India, Tunisia
OTSC2	7q34–36	Belgium, England
OTSC3	6p21.3–22.3	Cyprus, Tunisia
OTSC4	16q21–23.2	Israel
OTSC5	3q22–24	Netherlands
OTSC6[a]	—	—
OTSC7	6q13–16.1	Greece, Netherlands
OTSC8	9p13.1–q21.11	Tunisia
OTSC9[a]	—	—
OTSC10	1q41–44	Netherlands

[a] Loci names OTSC6 and OTSC9 have been reserved by the Human Genome Organisation Gene Nomenclature Committee but have yet to be published.
Data from Refs.[11,38,76]

Otolaryngol Clin North Am. 2018 Apr;51(2):305-318

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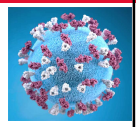
The Measles Virus Infection Hypothesis



- 1986 – McKenna et al: implicate persistent measles infection – identified filamentous structures similar to the virus in the bone.
- Shown high affinity to the otic capsule.
- Uses CD46 and CD150 cell surface receptors expressed in human and primates to enter cells.
- Lower incidence since the Measles virus vaccine and in the 1970's.

- Low rate of otosclerosis in countries that are endemic to Measles such as African countries.
- Numerous studies, including recently published reports, could not detect the virus RNA or CD46 expression in over 95% of stapes extracted from patients with otosclerosis.

Otolaryngol Head Neck Surg. 2018 Jan;158(1):158-162
Otolaryngol Clin North Am. 2018 Apr;51(2):305-318

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The Sodium Fluoride Deficiency Hypothesis

- Historical association between drinking water fluoridation and the prevalence of otosclerosis.
- Possible prevention of hearing deterioration by sodium fluoride oral supplementation.
- Suggested mechanism: inhibition of proteolytic enzymes to stabilize and decrease bone turnover.

- Inconsistency across studies as shown by a systematic review of the literature.

Otology & Neurotology. 35(6):1052–1057
Otolaryngol Clin North Am. 2018 Apr;51(2):305-318

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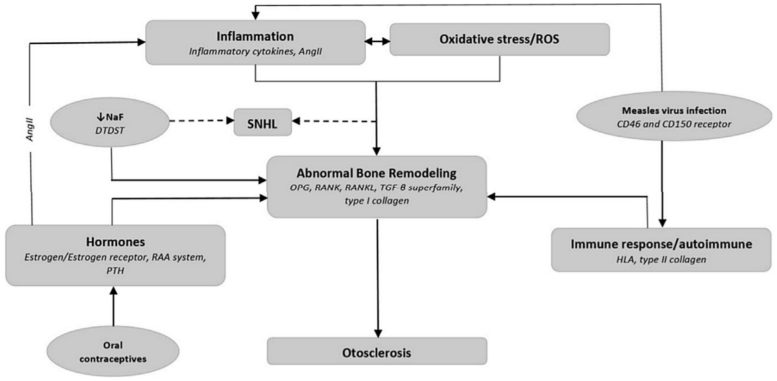

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Clinical Evaluation

- History
- Symptoms
- Physical examination
- Audiologic evaluation

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History

- **Gradual onset conductive/mixed hearing loss, slowly progressive.**
  - **70% bilateral, apparent late teens/twenties.**
  - **HL may not be apparent until age 30 or 40.**
- **Negative history for infections/trauma.**
- **Positive family history, often of surgical correction of HL.**

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Symptoms

- Hearing loss.
- "Paracusis of Willis" (false paracusis) – patients report difficulty hearing better in noisy rooms. Characteristic of conductive HL – people speak louder in noisy environments (e.g., in a bus or at Starbucks).
- Difficulty with direction of sounds and in noisy rooms (sign of asymmetric hearing loss).

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Physical Exam

- **Otoscopy**:
  - Normal TM. 10% of patients have "**Schwartze sign**" which is a pinkish hue behind the TM over the promontory or the area anterior to the oval window.
  - Normal pneumo-otoscopy.

- **Tuning forks**:
  - Weber – would lateralize to the ear with a greater conductive component with as little as 5 dB CHL.
  - **Rinne – 512-Hz reversal**: conductive loss of at least 15-20 dB. 512-Hz and 1024-Hz reversal: loss of at least 30 dB.

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Audiometry

- **Audiogram** – air conduction, bone conduction and speech audiometry. Typically a worsening conductive HL that progresses from low to high frequencies. Most patients also have a sensorineural loss that is disproportionate to age. Carhart's notch is a 10-20 dB drop in the bone conduction at 2000Hz induced by stapes fixation.

  Speech discrimination usually is better than normally expected, except when associated with severe SNHL.

- **Immittance** – compliance should be normal.

- **Acoustic reflex** –
  - Early stapes fixation: reflex normal.
  - Advanced disease – **reflex is absent**.
  - With progression of disease – the contralateral reflex is affected.

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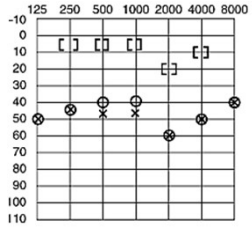
Carhart's notch

- Classically described as elevation of the bone conduction thresholds of approximately 5, 10 and 15 dB at 500, 1000 and 2000 Hz respectively. This is thought to result from the disruption of normal ossicular resonance, which, in humans, is approximately 2000 Hz.



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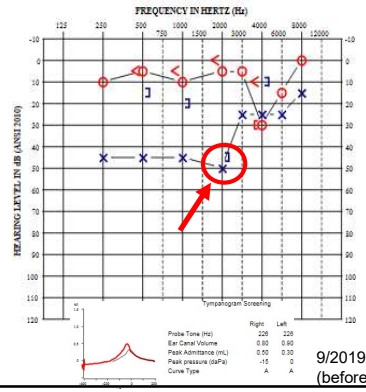
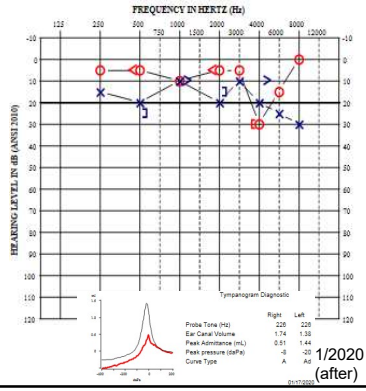
Carhart's notch



9/2019 (before)



1/2020 (after)

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Imaging of Otosclerosis - HRCT

- Not part of the routine workup.
- Can be helpful in confirming or excluding other pathologies.
- Sensitivity ~75-85%. 80% of positive cases are bilateral. – some studies suggest 100% sensitivity.
- Active (spongiform) lesions can be detected even <1mm. Inactive and sub-millimeter foci are difficult to detect.
- A correlation exists between the size of the otosclerotic focus and extent of conductive hearing loss, but not between the cochlear involvement and extent of sensorineural hearing loss.

Ann Otol Rhinol Laryngol. 2019 Nov;128(11):1054-1060
Ann Otol Rhinol Laryngol. 2005 Sep;*114*(9):*709-16*
Otolaryngol Head Neck Surg. 2006 Apr; *134(4):685-92*

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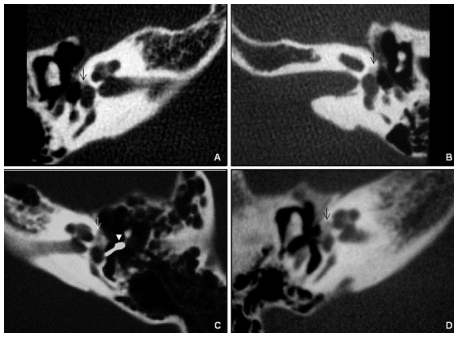
Fenestral Otosclerosis: Foci of hypodensity anterior to the oval window (FAOW)



Computed tomography, axial cuts,
A and B: FAOW with small size, **without involvement of the cochlear endosteum** (*arrows*);

C and D: FAOW probably involving the cochlear endosteum (*arrows*).
Note the presence of the stapedectomy prostheses (*arrowhead*).

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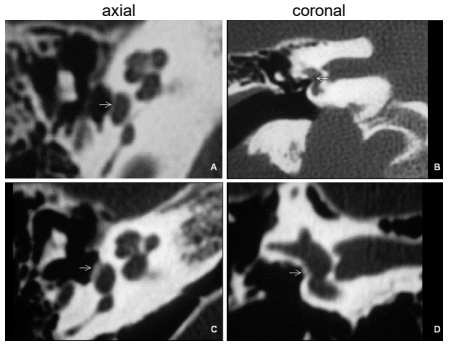
Fenestral Otosclerosis: Thickening of the stapedial footplate



Normal stapedial footplate

Thickened stapedial footplate

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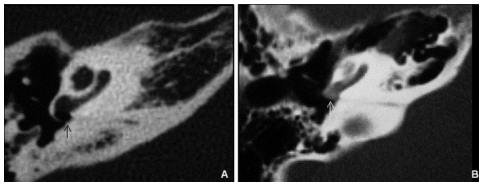
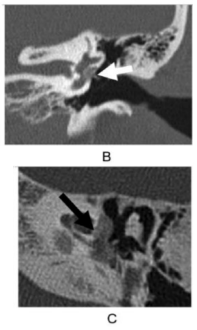
Fenestral Otosclerosis: Obliterative foci in the round window





Computed tomography, axial cut, in the level of the round window.
A: Normal round window niche (*black arrow*).
B: Note the presence of obliterative focus in the round window (*white arrow*).

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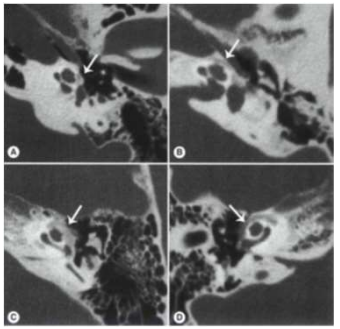
Retrofenestral/pericochlear otosclerosis: Pericochlea lucency

Limited to one turn

Entire capsule



More than one turn but not the entire capsule

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CT-Evaluation Checklist recommended by Dr. Redleaf and colleagues:

1. Lucency at the fissula ante-fenestrum
2. New bone formation around the round and oval window
3. Round window obliteration in a coronal view
4. Radiolucency surrounding the cochlea, semicircular canals and IAC
5. Rule out superior semicircular canal dehiscence

Sensitivity of HRCT can be up to 100% and imaging prior to surgery aids in counseling patients, preparing surgically, and excluding other pathologies

Rofo. 2020 Mar 26. doi: 10.1055/a-1131-7980
Ann Otol Rhinol Laryngol. 2019 Nov;128(11):1054-1060

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Differential Diagnosis

- **Ossicular discontinuity**.

May cause a conductive loss of up to 60db, and the tympanic membrane shows increased compliance on tympanometry.

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Differential Diagnosis

- Ossicular discontinuity.
- **Malleus head fixation**.

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Differential Diagnosis

- Ossicular discontinuity.
- Malleus head fixation.
- **Congenital fixation of the stapes**.

Congenital stapes fixation may also occur in conjunction with an abnormally small tympanic membrane, partial meatal atresia, or a shortened manubrium. A high degree of suspicion must be maintained in these patients. The sometimes abnormal course of the facial nerve in these patients makes operative repair a greater challenge.

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Differential Diagnosis

- Ossicular discontinuity.
- Malleus head fixation.
- Congenital fixation of the stapes.
- **Chronic secretory otitis media.**
- **Tympanosclerosis.**

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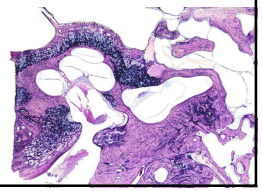
Differential Diagnosis

- Ossicular discontinuity.
- Malleus head fixation.
- Congenital fixation of the stapes.
- Chronic secretory otitis media.
- Tympanosclerosis.
- **Paget's disease (osteitis deformans) of the temporal bone.** Other bone diseases, e,g,osteopetrosis.

Begins in the periosteal bone outside the capsule instead of in the endochondral capsule like otosclerosis. It causes sensorineural loss similar to that sometimes seen in otosclerosis, probably due to hydrolytic enzymes entering the cochlear fluid. Conductive hearing loss is rare in Paget's disease.



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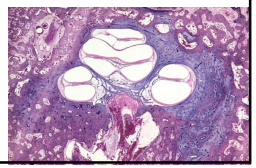
Differential Diagnosis

- Ossicular discontinuity.
- Malleus head fixation.
- Congenital fixation of the stapes.
- Chronic secretory otitis media.
- Tympanosclerosis.
- Paget's disease (osteitis deformans) of the temporal bone. Other bone diseases, e,g,osteopetrosis.
- **Osteogenesis imperfecta** (van der Hoeve-de Kleyn syndrome).

An autosomal dominant condition in which a defect in osteoblast activity results in fragile bones and blue sclera. In addition to multiple long bone fractures, around half of these patients develop stapes fixation. The short term surgical response to stapes surgery in these patients is similar to that seen in otosclerosis, but progressive postoperative sensorineural hearing loss is more common.



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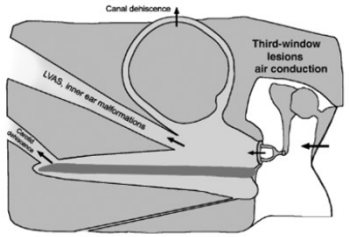
Differential Diagnosis

- Ossicular discontinuity.
- Malleus head fixation.
- Congenital fixation of the stapes.
- Chronic secretory otitis media.
- Tympanosclerosis.
- Paget's disease (osteitis deformans) of the temporal bone. Other bone diseases, e,g,osteopetrosis.
- Osteogenesis imperfecta (van der Hoeve-de Kleyn syndrome).
- **Third Window lesions** (SSCD, large vestibular aqueduct syndrome, dehiscent IAC).

'the great mimicker' – however, in third window pathologies, acoustic reflexes should be intact.



S.N. Merchant and J.J. Rosowski

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Finding	Middle ear pathology	Third window pathology
Air-Bone gap	0-60 dB, any frequencies	0-60 dB greatest in lower frequencies
Bone conduction thresholds	Usually normal or elevated	Can be negative (-5 to -20 or better) for frequencies below 2000Hz
Acoustic Reflexes	Absent	Present
VeMP	Absent	Present, thresholds lower than normal, can have higher amplitude
OAE	Absent	May be normal
Umbo velocity on laser Doppler vibrometry	Stapes fixation –normal Malleus fixation – low Ossicular discontinuity – high	High normal
Sound/pressure induced vertigo	Absent	May be present
Imaging	May show middle ear / otosclerotic anomalies	Third window pathology
Exploratory tympanotomy	Ossicular lesion	Normal ossicular mobility

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Treatment

- Natural history of the disease
- Amplification
- Medical
- Surgical

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Natural history

- **Roughly 90% of cases of otosclerosis are never clinically apparent**. The foci originate in childhood, remain immature for several years but never reach the cochlea or oval window, and then mature into the inactive form before ever causing hearing loss.
- The average **age of onset of subjective hearing loss is 20 years**.
- Typically, the disease progresses in a stepwise fashion, with periods of quiescence alternating with periods of deterioration.
- In addition to worsening hearing, periods of disease progression are characterized by worsening tinnitus, a positive Schwartze's sign, and sometimes a mild imbalance.
- The foci usually mature by age 50 to 70, leaving patients with as much as 50dB of conductive hearing loss, and varying degrees of sensorineural hearing loss.

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Nonsurgical management

- **Amplification**- a good option. These patients typically have good speech discrimination and do not have the recruitment problems common to most types of sensorineural hearing loss.
- **Sodium fluoride**- taken orally has been shown to stabilize the hearing loss associated with otosclerosis in 80% of patients. The fluoride ion replaces the usual hydroxyl ion in periosteal bone, forming fluorapatite, instead of the usual hydroxyapatite. In addition, bone resorption is reduced, and calcification of new bone is enhanced. Actively expanding foci of otosclerosis are inactivated, as has been documented by computed tomography.

  Tinnitus and imbalance are reduced, and Schwartze's sign frequently becomes negative.

  Side effects: rash, arthritis, and gastric distress.

  Available over the counter as Florical (sodium fluoride and calcium carbonate), and the usual dose is about 20-40 mg of fluoride a day. If overall stabilization of the disease has occurred, the patients is placed on a life-long maintanence dose of about 25mg of fluoride a day. Stopping the treatment in those patients whose disease process stabilized while on therapy may result in a reactivation of the disease in 2-3 years.
- **Bisphosphanates –** possible utility.

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Surgical treatment

- **1878-1899-** Kessel, Boucheron, Miot, Blake and Jack, Faraci- In all, more than 300 **stapes mobilizations** were performed. The incudostapedial joint was seperated, then the stapes was mobilized by applying pressure in various directions. There were many reported improvements in conductive hearing loss.
- **1900-** International Congress- Siebenmann, Politzer, and other leaders in Otology condemned stapes mobilizations and other surgery for otosclerosis as both dangerous and ineffective.
- **1930s**- Lempert- performed **horizontal canal fenestrations**, with some improvement in hearing. This procedure did not correct all of the conductive hearing loss.
- **1952-** Rosen- While exposing an ossicular chain in preparation for horizontal canal fenestration, a **stapes** became mobilized with dramatic improvement in hearing. He then began attempting **mobilizations** on several patients, evidently unaware of similar treatments used more than 50 years earlier.
- **1954**- Shambaugh - **First used a microscope** for stapes operations.
- **1956-** Shea- With an operating microscope, **he removed the stapes, then reconstructed the ossicular chain** and sealed the oval window. Modifications of his original technique are still used today.

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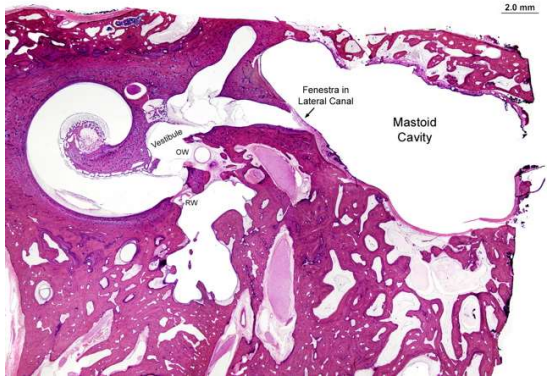
Surgical treatment



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Pre-operative considerations

- Indications
- Contraindications
- Counseling/ informed consent
- Operative note
- Type of anesthesia
- Perioperative antibiotics/steroids

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Indications for surgery

- Air-bone gap ≥25 dB at 250 Hz to 1 kHz.
- Negative Rinne at 512 Hz.
- In bilateral HL – worst hearing ear first. Second ear will follow in 8-12 months.
- Concomitant SNHL is not a contraindication.

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Contraindications for surgery

- **Only hearing ear** – except for patients with profound mixed hearing loss that are candidates for CI.
- Contra-lateral ear with disease that may threaten hearing in the future – relative C/I.
- Perforated TM.
- Infected middle/external ear.
- Patients with vestibular pathology – r/o Meniere's.
- Patients that depend on balance for their work, must be well informed of potential disequilibrium following surgery.

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Pre-operative counseling

- Explanation of treatment alternatives including amplification.
- Realistic expectations.
- Complications: failure to correct CHL, partial/complete HL (1%) – can also be from a sudden SNHL post-op, vestibular disturbance, perforation of TM, facial nerve injury, perilymphatic fistula and disturbance in taste. Complications of usage of Laser in surgery.
- Need to avoid engaging in activities that might subject the ear to high pressures (e.g. parachute jumping, diving, flying).
- Short term and long term failure.

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Operative note

Must be detailed both for future revision surgery and when considering surgery on the contralateral ear:

- Shape and mobility of incus and malleus.
- Presence of otosclerosis.
- Fixation of stapes.
- Patency of round window.
- Location and bone covering of the facial nerve.
- Status of the chorda tympani at the end of the procedure.

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Type of Anesthesia

Local anesthesia	General Anesthesia
•Hearing can be tested •If the patient complains of vertigo during the surgery the surgeon can alter his technique •Avoid postoperative nausea and extreme pressures associated with arousing •Shorter duration of surgery	•Patient comfort •Some patients are not amenable to local anesthesia

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Stapedotomy / Stapedectomy

- Positioning
- Exposure
- Fenestration / Removal of footplate
- Placement of prosthesis

Illustrations Adapted from Surgery Of The Ear And Temporal Bone by Nadol Joseph B Jr, chapter 22

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Stapedotomy

Positioning –

Goal: bring the TM to a horizontal plain.

Method:

• Supine, head hanging and rotated to the opposite shoulder.

• Use self-retaining speculum holder.

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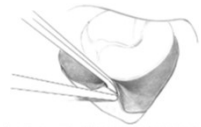
Local anesthesia:
Lidocaine+Epinephrine.

Exposure:

- Anteriorly based tympanomeatal flap (roller knife).



- Curette the posterior bony annulus to allow visualization of:
  - Facial nerve superiorly
  - Pyramidal eminence posteriorly
  - Entire circumference of the oval window

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Checkpoint !

- Assess:
  - Patency of round window
  - Mobility of incus and malleus
  - Fixation of stapes

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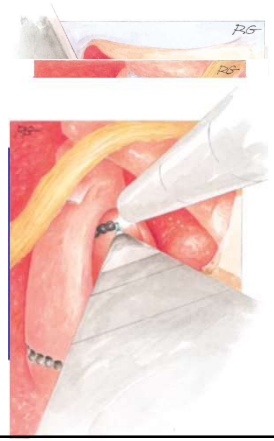
Separate incudostapedial joint (joint knife)

Section stapedial tendon (laser or scissors)

Posterior crurotomy and possibly anterior crurotomy

Fracture the remaining superstructure toward the promontory



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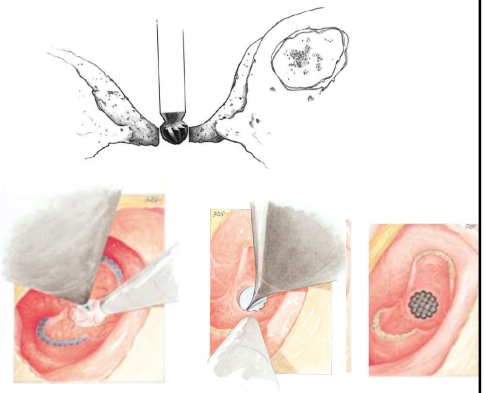
Fenestration

Motor driven short cutting/ diamond bur measuring 0.7mm in diameter

OR

Laser – argon/KTP (visible) or CO2 (invisible)

CO2 advantage – no caloric effect on the perilymph, however, has to be attached to an operating microscope with a separate coaxial aiming beam



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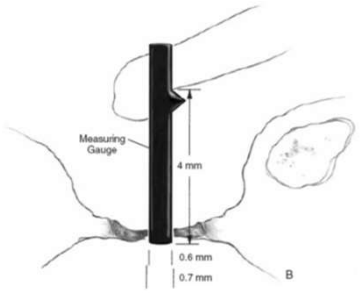
Measurement of the prosthesis



Width of measuring rod is 0.6 mm. 0.25 mm are added on to the length to allow sufficient penetration of the vestibule.

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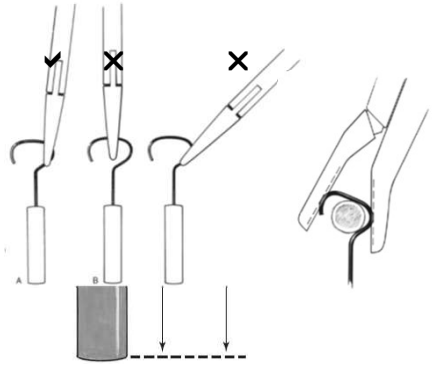
The stapes prosthesis

The Schuknecht Teflon-wire piston: Teflon head and malleable rod from stainless steel. Other pistons are made of platinum.

The piston is grabbed so that the long axis of the alligator and the prosthesis are aligned.

After testing prosthesis and ruling out nausea, the loop is then crimped. Prosthesis is tested. Gelfoam / tissue graft are placed around the piston. The tympanomeatal flap is returned.



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Types of stapes prosthesis

The original prosthesis was carved from teflon and implanted by Dr. Jon J. Shea, 1956

Otol Neurotol. 2008 *Apr;29(3):407-15*

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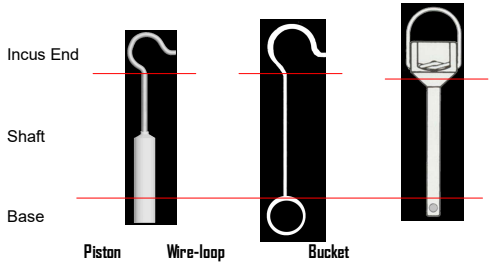
Types of stapes prosthesis



Otol Neurotol. 2008 *Apr;29(3):407-15*

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Types of stapes prosthesis

- Gyrus – SMART incus prosthesis: uses the elastic memory of a nitinol metallic wire that coils around the incus in response to heating.
- All prosthesis currently implanted are safe for MRI scanning.

Otol Neurotol. 2006 Dec;*27(8):1064-9*

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Stapedectomy

Indications:

- Floating footplate
- Comminuted fracture of the footplate
- Unavailability of lasers or micro-drills

Hearing results are similar for stapedectomy and stapedotomy but the occurrence, duration and severity of vestibular symptoms is higher for stapedectomy.

- Different tissue grafts can be used. Historically reconstruction was done with an ear lobule Fat-wire prosthesis.

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Post-Operative care

- Same day surgery.
- Keep ears dry.
- Avoid Valsalva maneuvers.
- Air travel allowed from a few days post-op.
- F/U in one week, audiometry in 6-8 weeks.

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Intra-Operative Complications

- **Tears in the tympanomeatal flap** → repair with underlay tragal perichondrium or fascia graft.
- **Subluxation of the Incus** → continue with surgery/ use a malleus attachment prosthesis.
- **Overhanging Facial nerve** (3% of cases secondary to a dehiscent fallopian canal) → abort surgery if the nerve is covering more than 50% of the stapes footplate.
- **Obliterative otosclerosis of the oval window** (4.7% of cases) → thin obstructing bone, then use a 0.7mm diamond burr to drill fenestration.
- **Otosclerosis involving the round window** → only complete obliteration will cause conductive HL. Complete procedure. If residual HL then no revision. Resolving obliteration is contraindiacted (d/t SNHL).

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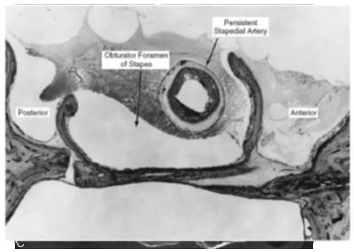
Intra-Operative Complications (2)

- **Persistent Stapedial artery** (1:5000 surgeries) → when persists arises from the internal carotid to replace the middle meningeal artery or branch into arteries that accompany CN V. Do not complete procedure if the artery can not be left intact.

Presentation: a pulsatile middle ear mass or that may appear as an incidental finding during middle ear surgery or in a CT scan.

The CT findings include the absence of the ipsilateral foramen spinosum and a soft-tissue prominence in the region of the tympanic segment of the facial nerve.



AJNR Am J Neuroradiol. 2000 Mar;*21*(*3*):*572-7*.

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Intra-Operative Complications (3)

- **Malleus ankylosis** (0.5% of temporal bones) → suspect when there is reduced movement of malleus in pneumatic otoscopy. Confirm with Doppler vibrometry. Treat with a malleus attachment prosthesis.
- **Perilymph Gusher** → can result from a congenital anomaly such as fixation of the stapes. Pack fenestra with tissue graft or cotton pledget. Place lumbar drain. Complete surgery with a perichondrium or vein tissue graft. Important to prevent SNHL.
- **Floating / depressed footplate** → if depressed will cause vertigo. Mainly – try to avoid by using laser when possible. Alternatively carefully perform a stapedectomy.

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Post-Operative Complications

- **Facial palsy →**
  - Secondary to local anesthesia – resolves within hours.
  - Damaged during surgery/ uncertain etiology – explore and possible graft.
  - Delayed paralysis (0.5%, 5-20 days post-op) – will resolve in 1-2 months. Treat like Bell's palsy.
- **Chorda tympani dysfunction →**
  - Dried nerve – temporary hypogusia/dysgusia.
  - Stretched nerve – chronic symptoms of metallic taste.
  - Cut nerve – temporary symptoms that usually resolve within 3-6 months.
- **Otitis media →** If immediate post-op admit with broad spectrum antibiotics.

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Post-Operative Complications (2)

- **Vertigo →**
  - During surgery – change length of stapes prosthesis.
  - Immediately following – suspect pneumolabyrinth. Should resolve in 24-48 hours. If persists may be associated with SNHL.
  - Delayed – BPPV, PLF.
- **Reparative Granuloma →** onset 5-15 days post-op; symptoms and signs of labyrinthitis: dizziness, tinnitus, HL, nystagmus toward the non-operated ear. Otoscopy: edema, thickening and hyperemia of flap and TM. Audiometry – mixed HL. Tx – steroids and immediate exploration with removal of prosthesis and closure of fenestra with a tissue graft.
- **SNHL →**
  - Early, high frequency – surgical trauma.
  - Delayed – suspect perilymphatic fistula.
  - Up to 1% of stapes surgery patients will suffer permanent SNHL.

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Post-Operative Complications (3)

- **Conductive hearing loss →** air-bone gap closure to within 10 dB occurs in approximately 90% of primary stapedectomies
  - Immediate post-op –
    - Malfunctioning prosthesis
    - Unrecognized malleus fixation
    - Unrecognized round window obliteration
    - Middle ear effusions
    - Unrecognized SSCD
  - Delayed – **recurrent conductive hearing loss occurs in 35% of cases**
    - Erosion of the incus at the site of attachment (64%)
    - Malpositioned prosthesis (41%)
    - Bony regrowth of the oval window (14%)
    - Round window obliteration (23%)
    - Adhesions in the middle ear (14%)

Otol Neurotol. 2001 Mar;22(2):*162-9*.

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Revision stapedectomy

- Indications:
  - An air-bone gap greater than 20 dB over the three-frequency range 0.5 to 2 kHz. Better chance for improvement if the initial hearing post-op was greatly improved.
  - Dizziness, suspected PLF.
- Results:
  - Closure of ABG to < 10 dB is achieved in ~60-80% of patients.
  - SNHL >10 dB can be as high as 7% and profound SNHL is ~1%.
  - A poorer outcome is related to incus necrosis, multiple revisions, and indications for surgery other than conductive hearing loss

Otol Neurotol. 2003 Jul;24(4):560-6
Otolaryngol Head Neck Surg. 2000 Dec;123(6):728-32
Laryngoscope. 1998 Dec;108(12):1794-800

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Other surgical options

- **BAHA – a handful of studies that show good results for selected patients.**

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Far advanced otosclerosis

- **Far advanced otosclerosis** (House and Sheehy 1961): Long term otosclerosis with air conduction > 85 dB and a non-measurable bone conduction.
- **Very far advanced otosclerosis** (Iurato 1992): non-measurable air and bone conduction (blank audiogram).
- Patients with far advanced otosclerosis can benefit from CI. However, several studies suggest that stapedectomy may be attempted before CI as some of the patients will do better with stapedectomy + amplification. Both options should be presented to the patients.

Turk Arch Otorhinolaryngol. 2020 Mar;58(1):35-40
J Laryngol Otol. 2017 Nov;131(11):961-964
Acta Otolaryngol. 2007 Jun;***127***(**6**):***574-8***

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THANK YOU!

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