1. (Dara) Review the histopathology of this disease.

The characteristic lesion of otosclerosis is a pleomorphic replacement of normal bone with spongiotic or sclerotic bone. The histiologic disease progresses in stages. Early lesions appear adjacent to the fissula ante fenestram as sheets of connective tissue replacing bone. Bone is absorbed by osteoclastic activity and new bone is deposited by osteocytes. The osteocytes are found at the advancing edge of the lesion, which extends into the otic capsule in fingerlike projections. The lesions contain vascular spaces in the center. The result is disorganized bone rich in osteocytes with enlarged marrow spaces rich with blood vessels and connective tissue ("spongiotic bone"). Healthy bone has few viable osteocytes and chondrocytes and is relatively avascular. Formation of dense sclerotic bone in areas of previous resorption signifies the late phase of otosclerosis. Lesions that have ceased all activity appear as sclerotic lesions.

The extent and location of lesions vary. Some are relatively small and do not involve the stapes. As the disease advances, the lesions spread across the stapedial annular ligament, causing stapedial fixation. If the lesion progresses in the opposite direction to the cochlea, it results in SNHL. It may spread in both directions, resulting in a mixed hearing loss. In 80-90% of patients, lesions are limited to the anterior oval window and affect its pathology by calcification of the annular ligament or by involving the stapes, resulting in the characteristic CHL. In 8% of patients, the process involves the cochlea and parts of the labyrinth (labyrinthine otosclerosis), resulting in SNHL. Approximately 2% of patients display both labyrinthine and ossicular chain involvement.

2. (Dara) Discuss the etiology of otosclerosis.

Otosclerosis is an autosomal dominant hereditary disease with variable penetrance and expression. 2/3 of those affected are women. Hearing loss usually begins in the late teens or early 20s and may be accelerated by pregnancy. Evidence has recently mounted that the measles virus may play a role in gene activation of otosclerosis. This hypothesis is supported by a declining incidence of otosclerosis since measles vaccinations became widespread.

3. (Deya) What are the “Blue Mantles of Manasse”?

4. (Deya) What is “Schwartz’s sign”?

Otosclerosis is a pleomorphic bone remodeling process. In the normal otic capsule, the calcification process is completed by one year of age and bone remodeling is rarely seen after this process. The otic capsule contains small regions of immature cartilaginous tissue called globuli interossei which may be the loci of the earliest lesions of otosclerosis.

The earliest phase of otosclerosis (spongiotic phase) is characterized by resorption of bone around blood vessels, with an increase in space around the vascular channels, which is replaced by cellular, fibrous connective tissue. Vascular spaces become wider and if the active focus reaches the periosteal surface of the promontory, they may cause a red-pink glow that can be seen through the TM on otoscopy, known as the Schwartz’s sign. This represents vascular shunts between the otosclerotic foci and the submucosal vessels of the promontory. Within the connective tissue laid around vascular channels, reticular cells and fibroblasts become osteoblasts, which lay down immature bone with a woven, disordered pattern of collagen fibers. This basophilic bone regions stain blue with H&E, thus forming “blue mantles of Manasse,” characterized by plexus-like projections.

During the late, or sclerotic phase, the new bone is resorbed and replaced with osseous tissue containing many collagen fibers and little ground substance. The osseous tissue stains red with H&E. The large areas of vascular, cellular connective tissue are replaced with new bone, and the lesion becomes quiescent. Within each temporal bone containing otosclerosis, lesions can be found in each of these phases although the overall histologic status seems to be fairly uniform.

5. (Amy) Discuss the epidemiology of otosclerosis.

Otosclerosis can be considered a hereditary disorder; the genes implicated are autosomal-dominant with highly variable penetrance and expression. Two thirds of affected individuals are women. Clinical otosclerosis is most common in whites (1%), and it is rare in blacks (0.1%), Asians (0.5%), and Native Americans (0.5%). Histologic otosclerosis is found in temporal bones of 7.3% of white women, 10.3% of white men, and approximately 1% of blacks. When one ear is affected, the contralateral ear is usually affected (80-90%), irrespective of race or age.

6. (Amy) What is the clinical presentation of otosclerosis?

Patients most often present with progressive hearing loss. Noticeable hearing loss usually begins in late teens or early twenties, but it can also present later (30's-early 40's). Hearing loss may be accelerated by pregnancy. If the process involves the stapes, the hearing loss is conductive. The most commonly affected area is the anterior crura. Otosclerosis may progress to involve entire stapes footplate or continue anteriorly toward the cochlea, causing a sensorineural hearing loss. Patients may also complain of tinnitus that progresses with the hearing loss. Occasionally, mild to severe vertigo is also present.

On physical, tuning fork exams are helpful but may be confusing in patients with mixed hearing loss. The remainder of the physical exam is often normal. The Schwartze sign is found in approximately 10% of cases and is characteristic of otosclerosis; it occurs when increased vascularity of the promontory during the active phase of otosclerosis is visualized as a pinkish hue behind the TM.
Jan 3: Otosclerosis (updated 09/06)

7. (Kathy) You have given a clinic patient the diagnosis of otosclerosis. She is not interested in surgery. What other options does she have?
   - Observation - In cases of mild CHL that does not bother patient, follow with yearly CAE.
   - Medication -
     - Sodium fluoride therapy:
       - Reduces osteoclastic resorption and increases osteoblastic bone formation. Together, these actions may promote recalcification and reduce bony remodeling in actively expanding osteolytic lesions.
       - Inhibits proteolytic enzymes that are cytotoxic to cochlea and may lead to SNHL.
       - Start at 50mg daily for active disease, may taper to 25mg daily with stabilization of symptoms (hearing stable, reduced tinnitus, reduced dizziness, fading of Schwartz sign, and radiologic signs of recalcification).
     - Bisphosphonates (alendronate, risedronate, zolendronate):
       - Incorporated into bone, where they inhibit osteoclastic remodeling.
       - Currently widely used for osteoporosis, but holds some promise in controlling otosclerosis.
   - Amplification -
     - Most patients have normal cochlear function with excellent speech discrimination, and are therefore good hearing aid candidates.
     - When compared to successful surgery, disadvantages include poorer sound quality, cosmesis, cost of maintenance, occlusion effect, comfort, and being able to hear only when aid is in use.

8. (Rosow) Discuss stapedectomy vs. stapedotomy

The key difference between the two procedures involves removal of all or part of the stapes footplate (stapedectomy) vs. creation of a small hole in the footplate where the prosthesis will articulate (stapedotomy). In stapedectomy, a tissue graft is always necessary to prevent perilymphatic fistula, whereas it may not be necessary in a stapedotomy. In a stapedectomy, a control hole is made in the midportion of the footplate with a straight pick prior to disarticulation of the incudostapedial joint. The stapes superstructure may be downfractured by exerting delicate force in a superior-to-inferior plane just medial to the capitulum of the stapes. The control hole is widened and the footplate bisected with a right-angle pick. The posterior one third of the footplate may be removed to perform a partial stapedectomy. A total stapedectomy involves removal of the entire footplate with a right-angle pick. A small fenestra stapedectomy involves widening the control hole to 0.6-0.8 mm. Whichever stapedectomy technique is used, the oval window opening is now covered with a tissue autograft composed of either fascia, vein, or perichondrium. A prosthesis may now be placed between the oval window and the incus. In a stapedotomy, the posterior crus of the stapes is divided with either the laser or crurotomy scissors. The stapes superstructure is downfractured and removed. A hole is made in the footplate using either the laser or a microdrill. A tissue graft is placed over the oval window opening when a bucket handle prosthesis is used, or the oval window may remain uncovered when other types of prostheses are used. A blood seal can be created by abrading the mucosa of the superior promontory and allowing blood to ooze onto the prosthesis-oval window interface. This may prevent perilymphatic fistulization.
9. (Rosow) What are the contraindications to surgery for otosclerosis?

**Indications:**
- Unilateral or bilateral otosclerosis in which the average air-bone gap of four frequencies (500, 1000, 2000, 4000 Hz) is 40 dB or more
- Otosclerosis with severely declining bone conduction and an air-bone gap < 40 dB
- Fixation of the footplate in tympanosclerosis with an intact or repaired TM
- Congenital anomalies with fixation of the footplate

**Relative contraindications:**
- Air-bone gap less than 40 dB with normal bone conduction
- Pediatric patient
- Significant medical comorbidities
- Profound post-op vertigo from contralateral ear

**Temporarily absolute:**
- Otitis externa
- Perforated TM (for large perforations that do not heal spontaneously, tympanoplasty should be done prior to and separate from stapes surgery)

**Absolute:**
- Only hearing ear
- Significant complication in contralateral ear (arterial bleed, profound post-op sensorineural hearing loss)
- Cochlear otosclerosis
- Coexistent Meniere’s disease (significantly increases the possibility of residual hearing loss in the operated ear)
- Documented dilation of the vestibule or the vestibular aqueduct on CT or MRI (also negatively affects hearing outcome)

All patients with otosclerosis should be offered the option of wearing a hearing aid prior to proceeding with stapes surgery.

10. (Josh) You have signed up your first patient for a stapedotomy-drill vs. laser? What types of lasers would you consider?

Current methods of performing small fenestra stapedotomy involve various techniques including the use of lasers, microdrills, or fine picks. Evaluation of postoperative PTA, frequencies, and SNHL shows no significant difference between the two methods. Limited opening of the vestibule may carry lower risk of damage to the inner ear and resultant SNHL and/or vertigo.

**Microdrills**
- Increased usefulness when one is faced with a thick footplate

**Lasers**
- May reduce the mechanical trauma to the stapes, causing decreased labyrinthine irritation and possibly better results
- Two types of lasers are being used to create the fenestra in the footplate of the stapes

  - **Visible argon OR Potassium-titanyl-phosphate (KTP-532)**
    - Both similar frequency
    - Carried through fiberoptic cable
    - Preferentially absorbed by red pigment, such as hemoglobin
    - Handheld probe has greater divergence than the microscope mounted micromanipulator, and therefore has less potential to damage the contents of the vestibule
    - Some believe should not be used for revision stapes surgery because poorly absorbed by collagen and passes readily through perilymph

  - **Infrared Carbon Dioxide (CO₂)**
    - Carefully aligned series of mirrors and lenses
    - More efficiently absorbed by collagen and bone than KTP
    - Some believe more effective in primary and revision stapes surgery
11. (Tali) What is Carhart’s notch and why does it disappear after successful surgery?

Carhart’s notch: An audiometric finding characteristic of otosclerosis is a decrease in bone conduction threshold with a peak at 2,000 Hz. Although the notch occurs at 2,000 Hz, a reduction in bone conduction sensitivity is seen from 500 to 4,000 Hz which is, on average, 5 dB at 500 Hz, 10 dB at 1000 Hz, 15 dB at 2000 Hz, and 5 dB at 4,000 Hz. Carhart attributed this phenomenon to “mechanical factors associated with stapedial fixation.” The Carhart notch is not a true indication of “cochlear reserve” and this apparent bone conduction loss may be corrected by surgical intervention. There are a variety of theories but generally accepted to be result of stapes fixation from otosclerosis which causes disruption of the around normal ossicular chain resonance (at 2000 hz). This eliminates the middle ear component of sound amplification. Post-operatively, once the stapes is mobilized, it reverses. Other theories for this audiometric finding include mechanical artifact and perilymph immobility.

12. (Scott) What problems might you encounter intra-op and how would you deal with them?

Intraoperative complications of stapes surgery include:

- TM perforation: 1.9% incidence, repair if needed via underlay technique.
- Chorda injury: metallic taste, 30% of cases, usually due to stretch on nerve and resolves 3-4 months
- Bleeding: mucosal trauma (which can cause oval window fibrosis w/reclosure), persistent stapedial artery (may bipolar artery or work around)
- Malleus fixation: 1-10% cases, usually occurs later in life, air-bone gap usually 10-15 dB; can either break ossified ligaments (high rate of recurrence) or perform incus bypass procedure.
- Incus: necrosis due to COM or previously placed prosthesis with erosion. Intraop subluxation in 0.2% cases. Can perform incus bypass procedure.
- Tympanosclerosis: can be otosclerosis with mild CHL, hx of OM and myringosclerosis. If progresses to involve footplate then can perform stapes surgery but results not as good as for otosclerosis.
- Solid/obliterated footplate: 7-11%, solid more common. Most thin out footplate with drill, but partial or complete reclosure of oval window after stapedectomy may occur = early failure. May use CT to follow progression post-op.
- Floating stapes footplate: can occur when stapes in mobilized, may be pushed into vestibule if not careful. Control holes made to prevent further mobilization and use of hooks/needles to get under footplate and remove safely.
- CSF gusher: CSF gush upon entering vestibule; Two sources: widened cochlear aqueduct or defect in fundus of IAC. Two clues of possible CSF gusher – avascular congenital middle ear and abnormally anterior insertion of posterior crus of footplate. More common in footplate fixation and on L sided stapes surgery. Immediate HOB elevation, small hole stapedotomy and fascial plug prior to prosthesis insertion to prevent further leak. Occasionally needed lumbar drain.
- Prothesis failure: 30-82% failures due to displaced prosthesis and depends on prosthesis used. Short prosthesis 8-9% failures.

Other important complications (post-op)

- SNHL: 0.2-10%
- Perilymphatic fistula: drop/fluctuation in hearing, tinnitus/vertigo.
- Facial nerve injury: rare