

**TITLE: Otosclerosis**

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**RESIDENT PHYSICIAN: Alan L. Cowan, MD**

**FACULTY PHYSICIAN: Tomoko Makishima, MD, PhD**

**SERIES EDITORS: Francis B. Quinn, Jr., MD and Matthew W. Ryan, MD**

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## **Introduction**

Otosclerosis is a primary metabolic bone disease of the otic capsule and ossicles that causes fixation of the ossicles with resultant conductive or mixed hearing loss. It is genetically-mediated via autosomal dominant transmission with incomplete penetrance (approximately 40%) and variable expressivity.

Stapes fixation was first described by Valsalva in 1704 during an autopsy on a deaf patient. Joseph Toynbee linked this finding to hearing loss in 1857, and went on to catalog four pathologic variations of stapes fixation in 1868. In 1890 Katz was the first to find microscopic evidence of otosclerosis resulting in stapes fixation. In 1893, Politzer described otosclerosis as a clinical entity and further characterized its pathology in a cadaver. The earliest reports of improved hearing in patients with otosclerosis were anecdotal cases of patients who experienced hearing improvement after head trauma. Toynbee noted that several of his patients noted dramatic improvement after a sudden loud sound or 'traveling in a carriage over a hard road.' These actions presumably freed the fixation of the stapes. Kessel, in 1878 reported on another case. He examined the temporal bones of a patient who had improved hearing after falling off a wagon and found that he had a fracture through the horizontal canal. Politzer in 1894 reported otosclerosis as a "Primary Disease of the Bony Labyrinthine Capsule," in which neoplastic bone "gradually takes the place of normal bone, and ... produces ankylosis of the stapes." Bacon, in 1898, affirmed Politzer's findings, and recommended "administration of large doses of iodide of potash, which should be kept up for a year or more," as treatment. At this time, surgical treatments were considered unthinkable, and in Bacon's Manual of Otology in 1898 he reports that operations on the stapes "should not be considered for a moment." However, Holmgren (1923) had noted the findings of Kessel and Toynbee, and began treating a number of patients with otosclerosis by creating a horizontal canal fenestration and covering it with mucoperiosteum. He is now well known as the father of fenestration surgery. Holmgren's student, Sourdille, later reported a 64% satisfactory result in his three staged operation: (1) mastoidectomy, (2) external ear canal skin flap and (3) horizontal canal fenestration covered by the skin flap. Single stage horizontal canal fenestration was popularized by Lempert in the 1940s

and further refined thereafter by others including John House who described the double blue line technique to avoid drilling directly over the fenestra.

Despite the wide acceptance of the fenestration technique, it was rarely successful in completely closing the air-bone-gap (ABG) and had a > 2% risk of profound sensori-neural hearing loss (SNHL). Therefore, alternate methods were explored. In the 1950s, with initial criticism, Samuel Rosen proposed mobilization of the stapes. This had the advantage of achieving immediate hearing improvement and complete closure of the ABG in most instances. It was soon realized that re-fixation of the stapes was the norm rather than the exception in these cases. In 1956 John Shea performed the first stapedectomy. He covered the oval window with a vein graft from the hand and used an artificial nylon stapes from the incus to the oval window.

Many of the techniques developed by the above mentioned pioneers of stapes surgery still persist today. Variations in technique and style such as grafting material and prosthesis choice, performing stapedectomy versus stapedotomy, and use of cold, sharp instrumentation versus the laser or microdrill have arisen with similar success rates but the basic principle of re-establishing a mobile connection between the tympanic membrane and vestibule has remained the goal of surgery for patients with otosclerosis.

## Epidemiology

The overall prevalence of histologic otosclerosis is about 10%. Approximately 10% of these are affected clinically making the overall prevalence of noticeable hearing loss secondary to otosclerosis approximately 1% in the population. This disease is more common in Caucasians than in other races. 10-20% of Caucasians have histologic otosclerosis while only 1% of African Americans are affected. Overall, clinical disease is present in 1% of Caucasians, 0.5% of Asians, 0.1% of African Americans and 0% of Native Americans.

<u>Race</u>	<u>% incidence of microscopic otosclerosis</u>
Caucasian	10%
Asian	5%
African American	1%
Native American	0%

Women more commonly seek medical attention for hearing loss secondary to otosclerosis, however, studies looking at histologic prevalence of otosclerosis show no difference in men versus women. Approximately twice as many women present with clinical otosclerosis than men. This may be explained by the fact that pregnancy seems to accelerate the progression of otosclerosis. Clinical onset during pregnancy has been reported to be between 10% and 17%. In addition, bilaterality seems to be more common in women than men (89% and 65% respectively).

A review of the literature regarding pregnancy and otosclerosis, however, reveals that current evidence is lacking. The original article which supported treatment of otosclerosis (genetic hearing loss) by cessation of pregnancy and sterilization, was published out of Germany in 1939. This, however, may have been a politically motivated attempt to cleanse a genetic disease. In 1951, abortion and sterilization were still offered as options for patients with progressive hearing loss. In 1983, another article supported the link between progression of hearing loss and pregnancy, however, this was a retrospective review of patient opinions regarding their hearing, without any audiometric data. A more recent article in 2005 used audiometric results to compare a large group of women who had undergone otosclerosis surgery. They compared the women who had never been pregnant to those with children, and found the only difference to be a slightly lower air conduction threshold in women with children. This data, although not age matched, questions the association of hearing loss progression in pregnancy, and would argue against severe measures such as termination of pregnancy or sterilization.

The incidence of otosclerosis increases with age. The most common age group presenting with hearing loss from otosclerosis is 15-45 years, however it has been reported to manifest as early as 7 years and as late as the mid 50s. Microscopic foci of otosclerosis have been found in 0.6% of autopsies of individuals less than 5 years of age. Children presenting with otosclerosis most often have a positive family history and progressive hearing loss.

There has been much debate over the relationship of fluoride and incidence of otosclerosis. Many feel that a lack of environmental fluoride may result in clinical disease. Others feel that treatment with fluoride may stop progression of hearing loss. However, Hough noted two patient populations with different levels of fluoride but no relationship between hearing loss. He also noted extremely high levels of fluoride in areas of India which also are known to have a high incidence of otosclerosis. He concluded that there was no evidence for causation.

## **Pathophysiology**

Otosclerosis is an osseous dyscrasia, limited to the temporal bone, and characterized by resorption and formation of new bone in the area of the ossicles and otic capsule. The inciting event that initiates the onset this disease is unknown. Many theories have been proposed such as hereditary, endocrine, metabolic, infectious, vascular, and most recently, autoimmune, however, none have been proven. Hormonal factors have been suggested to play a role in otosclerosis based on the observation that pregnancy sometimes accelerates the progression of the disease. Siebenmann first described the microscopic appearance of an otosclerotic focus as spongy in appearance. He noted that the disease was usually quiescent and limited to the anterior aspect of the stapedial foot plate. Less frequently the focus encompasses the annular ligament of the stapes resulting in decreased mobilization and subsequent conductive hearing loss. In 1912 Siebenmann came up with the term labyrinthine otosclerosis referring to foci of otosclerosis involving the otic capsule of the cochlea and labyrinth. He proposed that labyrinthine otosclerosis could result in SNHL. To this day, this is a debated topic. Proposed theories of how SNHL may result from otosclerosis include liberation of toxic metabolites into the inner ear, decreased blood supply, and direct extension of the otosclerotic focus into the inner ear resulting

in changes in electrolyte concentrations and alterations of the biomechanics of the basilar membrane.

The most common site of involvement in otosclerosis is the anterior oval window near the fissula ante fenestrum. When both the anterior and posterior ends of the footplate are involved it is termed “bipolar” involvement or fixation (if the footplate is immobile). If only the footplate is involved, it is sometimes referred to as a “biscuit footplate”. When the entire footplate and annular ligament are involved it is known as an obliterated footplate or obliterative otosclerosis. The round window is involved in approximately 30% to 50% of cases but the niche is rarely completely obliterated.

Histologically, otosclerosis has two main forms: an early or spongiotic phase (otospongiosis) and a late or sclerotic phase. The early phase is characterized by multiple active cell groups including osteocytes, osteoblasts, and histiocytes. It develops a spongy appearance because of vascular dilation secondary to osteocyte resorption of bone surrounding blood vessels. This can be seen grossly as red hue behind the tympanic membrane termed “Schwartz's sign” (described by Schwartze in 1873). Microscopically, a focus of active otosclerosis reveals finger projections of disorganized bone, rich in osteocytes particularly at the leading edge. In the center of the focus, multinucleated osteocytes are often present. In the sclerotic phase, dense sclerotic bone forms in the areas of previous resorption. Both the sclerotic and spongiotic as well as intermediate phases may be present at the same time. Otosclerotic foci always begin in endochondral bone but may progress to involve endosteal and periosteal layers and even enter into the membranous labyrinth.

## **Diagnosis**

### **History**

The most common clinical presentation of otosclerosis is a patient in their 20s or 30s (more commonly women) with slowly progressive, bilateral (80%), asymmetric, conductive hearing loss and tinnitus (75%). As in any patient with an otologic complaint, a complete history should be ascertained including the age of onset of hearing loss, progression, laterality, and associated symptoms including vertigo or dizziness, otalgia, otorrhea, and tinnitus. A history of prior otologic surgeries should be elicited as well. History of significant ear infections (particularly chronic otitis media) makes the diagnosis of otosclerosis less likely.

Approximately 25% of patients with otosclerosis present with some vestibular complaints. This has been termed by some as otosclerotic inner ear syndrome. True attacks of vertigo may be present, however, mild dysequilibrium is the more common. When vertigo is present, it is usually paroxysmal and may be difficult to distinguish from Meniere's syndrome. If there is any question, further vestibular testing should be performed to differentiate the two. Some experts are proponents of performing electrocochleography on any patient suspected of having otosclerosis who have vestibular complaints. Stapedectomy performed in patients with Meniere's disease carries with a much higher incidence of postoperative profound SNHL, and most surgeons feel that any vestibular complaints are a contraindication to the procedure.

Patients will often have low-volume speech. Because of the conductive nature of their hearing loss, they perceive their voice as louder than it actually is. As can occur in other forms of conductive hearing loss, patients with otosclerosis will sometimes report improved hearing in the presence of background noise (opposite of those with high frequency SNHL). This is known as the paracusis of Willis. It occurs because the CHL reduces the volume of the background noise, and improves the ability to hear someone who is speaking directly to them with a higher volume to overcome the background noise.

Two-thirds of patients will report a family history of hearing loss. This is particularly important to ascertain as it may help to make the diagnosis in less clear cases.

The differential diagnosis for otosclerosis includes any number of middle ear disorders resulting in conductive or mixed hearing loss. Disorders such as ossicular discontinuity, congenital stapes fixation, malleus head fixation, superior semicircular canal dehiscence, Paget's disease (osteitis deformans), and Osteogenesis imperfecta (van der Hoeve syndrome) present with similar otologic findings as otosclerosis. Sometimes definitive diagnosis is delayed until surgical exploration of the middle ear. Below are some clinical findings for each of these diagnoses that differentiate them from one another:

**Ossicular discontinuity**

- conductive loss of 60 db usually without sensorineural component
- flaccid tympanic membrane on pneumatic otoscopy
- type Ad tympanogram

**Congenital stapes fixation**

- Family history less likely (10%)
- usually detected in the first decade of life
- 25% incidence of other congenital anomalies (3% for juvenile otosclerosis)
- non-progressive CHL

**Malleus head fixation**

- when congenital, associated with other stigmata (aural atresia)
- presence of tympanosclerosis
- pneumatic otoscopy
- almost always associated with type As tympanogram (only in advanced otosclerosis)

**Paget's disease**

- diffuse involvement of the bony skeleton
- elevated alkaline phosphatase
- CT - diffuse, bilateral, petrous bone involvement with extensive de-mineralization
- More commonly crowds the ossicles in the epitympanum, partially fixing the ossicular chain

**Osteogenesis imperfecta**

- presence of blue sclera
- h/o of multiple bone fractures
- CT - more common involves the otic capsule and to a greater extent

**Superior semicircular canal dehiscence**

- conductive hearing loss similar to otosclerosis

- mobile ossicles and tympanic membrane on pneumatic otoscopy
- vertigo or eye movements with loud noise, pneumatic otoscopy, or valsalva
- CT findings of superior canal dehiscence

## **Physical Exam**

Otomicroscopy is essential in making the diagnosis of otosclerosis, particularly with respect to ruling out other causes of conductive hearing loss. Middle ear effusions can mimic the symptoms of otosclerosis but is readily seen on examination of the TM. Tympanosclerosis usually occurs in a patient with a history chronic otitis media. It alone can cause significant CHL either through fixation of the malleus in which case the focus is often seen just anterior to the neck of the malleus, or when it surrounds the stapes and oval window area. Pneumatic otoscopy can be helpful in making the diagnosis of malleus fixation. Cholesteatoma, chronic adhesive otitis media, and tympanic membrane perforation are usually easily ruled out by the physical examination.

Recent literature also describes a condition of superior semicircular canal dehiscence which may mimic the audiometric findings of otosclerosis. While these patients may present with an air-bone gap and a normal appearing middle ear on exam, they have several findings which may elicit the correct diagnosis. The patients often complain of hyperacusis to their pulse or eye movements. Application of pressure or loud sound to the ear canal may evoke a vertical torsional nystagmus. The tympanic membrane is mobile on pneumatic otoscopy. In addition, audiometric examination may reveal bone conduction levels at less than 0 dB NHL, intact reflexes, and normal speech discrimination.

The TM appears normal in the majority of patients with otosclerosis. In 10% of patients, a red hue may be seen in the area of the promontory and oval window niche (Schwartz sign). This occurs in patients with active disease where increased vascularity is present in these areas. 256, 512, and 1,024-Hz tuning forks (TF) should be used in all patients. The Rinne test is particularly useful. Early in the disease, low frequency CHL will predominate resulting in a negative Rinne test with the 256-Hz TF only. As progression occurs, the 512 and then the 1,024-Hz TF will become negative.

## **Audiometry**

The audiogram, particularly with respect to pure tones, tympanometry, and acoustic reflexes, is the most important objective test in diagnosing and planning treatment for patients with otosclerosis.

Tympanometry is a graphic representation of change in acceptance of sound energy through the middle ear as a function of air pressure applied into a sealed external auditory canal. Jerger (1970) classified the tympanogram into type A, B, and C. Type A tympanograms are further subdivided into type A, As, and Ad. All type A tympanograms have a clearly defined peak of function in the range of  $\pm 100$  daPa. The location the peak of the tympanogram along the x-axis is a representation of the air pressure in the middle ear space. The height of the peak is determined by the peak compliance of the tympanic membrane and middle ear. Because middle ear air pressure is not affected by otosclerosis, the peak of the tympanogram is always in

the normal range along the x-axis. Compliance of the system, however, is reduced as otosclerosis progresses resulting in depression of the height of the peak. A low peak in the normal middle ear pressure range is called a type As (s-stiffness curve) tympanogram and is characteristic of advanced otosclerosis but more commonly, malleus fixation.

Acoustic reflexes result from a change in compliance of the middle ear/TM system in response to a sound stimulus. The change in compliance results from contraction of the stapedius muscle in response to a loud sound which stabilizes the stapes reducing the transmission of sound energy in to the vestibule. Progressive stapes fixation results in predictable pattern of acoustic reflex abnormalities. The first sign of early otosclerosis (even before any conductive hearing loss is detected) is a diphasic reflex pattern (ie increase compliance at the onset and cessation of the sound stimulus). Why this happens is unclear but it has been postulated that it results from inherent elasticity in the otosclerotic anterior footplate and crura allowing the non-affected posterior footplate to move with stapedius contraction and relaxation. As stapes fixation progresses, the acoustic reflex amplitudes are reduced, followed by elevation of ipsilateral, then contralateral thresholds, and finally, disappearance of the reflexes altogether.

Pure tone audiometry has advantages over tympanometry and acoustic reflex testing in that it better characterizes the severity of disease and is frequency specific. The first affect of early otosclerosis on pure tones is a decrease in air conduction in the low frequency, especially below 1000 Hz. The rising air line seen in these patients is referred to as a “stiffness tilt” and is called such because it is caused by decreased compliance secondary to stapes fixation. As the disease progresses, the air line flattens. This occurs because the otosclerotic focus has a mass affect on the entire system in addition to decreasing its compliance. Further progression of otosclerosis to involve the cochlea may result in increased bone conduction thresholds. Usually high frequencies are affected first because the disease focus is usually adjacent to the basal turn of the cochlea. More isolated cochlear otosclerosis may sometimes result in a mixed hearing loss with a “cookie-bite” pattern with both air and bone lines.

The *Carhart notch* is the hallmark audiologic sign of otosclerosis. It is characterized by a decreased in the bone conduction thresholds of approximately 5 dB at 500 Hz, 10 dB at 1000 Hz, 15 dB at 2000 Hz, and 5 dB at 4000 Hz. It results from a mechanical artifact and is not a true representation of the cochlear reserve. It is postulated that this phenomenon occurs because stapes fixation disrupts the normal ossicular resonance and that the normal compressional mode of bone conduction is disturbed because of relative perilymph immobility caused by stapes fixation. These theories are supported by the fact that *Carhart's notch* disappears after stapedectomy.

Due to this change in bone conduction levels following stapes surgery, the Committee on Hearing and Equilibrium created guidelines for reporting audiometric results in stapes surgery. Since the bone conduction can be improved along with the air conduction, the committee recommended basing the results on postoperative bone measurements, specifically postoperative air-bone gaps. This prevents ‘overclosure,’ where the postoperative air line is compared to the preoperative bone line, which may result in a air-bone gap closure of more than 100%, even when a postoperative gap is present. While this was adopted by the AAOHNS in 1994, not all

areas of the world use these standards, so results reported in the literature should be carefully examined for their ABG closure criteria.

The vast majority of cases of otosclerosis are associated with conductive or mixed hearing loss. Occasionally, a case of pure sensorineural otosclerosis will be described, but its existence is controversial. The first case of pure sensorineural otosclerosis was described by Shambaugh Sr. in 1903. In 1967, Shambaugh Jr. developed seven criteria for diagnosing pure sensorineural otosclerosis.

1. A positive Schwartz's sign
2. Family history of surgically confirmed stapedial otosclerosis
3. Symmetric progressive sensorineural loss, with stapes fixation in one ear
4. Unusually good speech discrimination for a pure sensorineural loss
5. Onset of hearing loss at usual age for stapes fixation and progressing without other known etiology
6. CT scan in patient with one or more of above criteria that shows cochlear capsule demineralization (a negative scan does not preclude diagnosis because mature disease resemble normal bone on CT)
7. On-Off effect of impedance tympanometry

Since that time, many investigators have studied the relationship between otosclerotic foci within the otic capsule and the existence of sensorineural hearing loss. A number of purely audiometric studies without histology have been conducted; several show no evidence of sensory loss, while others have shown marked differences in sensory levels between ears in unilateral otosclerosis. Histologic studies with clinical correlation have shown that some cases of sensorineural loss have associated otosclerotic foci in the endosteal bone; however, cases of endosteal involvement without sensory loss and cases of sensory loss without endosteal involvement have also been frequently documented. At the present time, no firm evidence to support or refute the relationship of sensorineural hearing loss and cochlear endosteal involvement exists.

## **Imaging**

It is controversial whether imaging studies are needed, beyond the physical exam, tuning forks, and audiogram, in diagnosing and managing patients with otosclerosis. Those in favor of computed tomography (CT) scanning of the temporal bone in these patients cite a number of advantages. CT can characterize the extent of the otosclerotic focus at the oval window and can be used when the clinical diagnosis is in doubt (patients with profound mixed hearing loss). It is also indicated by some when patients have significant mixed hearing loss to determine if capsular involvement is present. An enlarged cochlear aqueduct may be seen which increases the index of suspicion for a potential perilymph gusher during footplate fenestration or removal.

CT scanning is also useful in evaluating the post-stapedectomy patient who initially had a good hearing result but was subsequently lost. The 20 degree coronal oblique projection is the best view to evaluate for re-obliteration of the oval window or for determining the position of the prosthesis. Necrosis of the long process of the incus can also be detected. If the patient is experiencing attacks of vertigo, the piston may be seen displaced medially into the vestibule. In

addition, patients with complaints of hyperacusis and a Tullio phenomenon or Hennebert sign may warrant a CT evaluation for superior semicircular canal dehiscence. CT scanning of the temporal bone should be taken in the axial and coronal oblique planes. The axial projection is obtained by having the patient lay supine with the canthomeatal line perpendicular to the table top. 1 mm cuts are then taken from the top of the superior semi-circular canal to the inferior aspect of the cochlea. The coronal oblique view should be taken with the patient supine with the head overextended so that the X-ray beam passes perpendicular the Frankfurt's line. The patients face is rotated 20 degrees towards the side under examination.

Foci of early or active otosclerosis or otospongiosis are poorly calcified and may actually make the oval window appear larger than normal. As re-ossification occurs the oval window appears smaller or obliterated by bone. In cochlear otosclerosis, active foci give the appearance of a ring around the cochlea. This is termed "double ring effect" or the "halo" sign. In order for a mature cochlear or labyrinthine focus of otosclerosis to be seen on CT, the focus must be larger than 1mm, its density must be different for that of the normal otic capsule, and it must appose the periosteal or endosteal surfaces of the capsule.

Paget's disease is in the differential diagnosis for otosclerosis. Although other physical findings will likely be present to distinguish them from one another, it is worth mentioning the unique CT findings of Paget's disease in the temporal bone. This disease is characterized by initial bone resorption followed by diffuse bony hypertrophy. Most commonly the petrous bone is affected first in which the internal auditory canal is usually the first structure involved followed by the cochlea and vestibular system. On CT there is a typical washed-out appearance of both temporal bones due to severe demineralization. As remodeling of bone occurs the entire temporal bone becomes thickened. The stapes is often involved and less commonly the incus and malleus.

CT findings in osteogenesis imperfecta often reveal an abnormally thin calvarium. Active otosclerotic foci in the temporal bone usually involve the entire otic capsule.

## **Management**

### **Natural History**

Roughly 90% of cases of otosclerosis are never clinically apparent. 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 presentation of patients with clinically evident disease is 20 years. Over the next two to four decades, the conductive hearing loss progressive and may become mixed. Rarely is pure SNHL a result of otosclerosis. 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 mild imbalance. The foci usually mature by age 50 to 70, leaving patients with as much as a 50 dB CHL, and varying degrees of SNHL.

## **Patient selection and preoperative counseling**

Factors that contribute to determining the best method of treatment for a patient with otosclerosis include the results of tuning fork testing and audiometry, skill of the surgeon, medical condition of the patient and patient wishes. Studies have shown a higher satisfaction rate in patients undergoing successful surgical treatment for otosclerosis than those who use hearing aids, however, each patient should be treated on an individual basis. Once the diagnosis is made, options for treatment including hearing aids and surgery along with the advantages and disadvantages of each should be discussed extensively with the patient. A repeat clinic visit one week later is often beneficial, giving the patient time to think about the options and avoid rushing into something they may regret. The best surgical candidate is a patient in good health with a socially unacceptable ABG, a negative Rinne test, excellent discrimination, and the desire for surgery after an appropriate period of time for deliberation.

Other factors that should be considered when counseling patients about surgery are the age of the patient, occupation, and history of prior stapes surgery. Meyer (1999) showed that older patients undergoing stapes surgery had poorer results in the high frequency range. Children with congenital stapes fixation have a much lower success rate of ABG closure (44%) as compared to children with juvenile otosclerosis (82%). Younger patients are more likely to develop re-ossification of the stapes footplate over their lifetime. They can undergo revision surgery which carries with a substantially reduced success rate and higher rate of profound SNHL. In the past patients who were exposed to frequent barometric changes (divers, pilots, airline stewardess) were counseled on the increased risk for postoperative fistula and prosthesis dislocation. This was true when the use of gelfoam, wire prostheses and total stapedectomy were more common. Now, with the increased practice of the small fenestra technique, use of newer pistons, and autologous tissue for oval window seals, displacement of the prosthesis and perilymph fistula is no more common in these patients than in the general population. It is certainly not unreasonable for these theoretical risks to be discussed with the patient. Patients whose hobbies or profession requires excellent balance are questionable candidates for stapes surgery.

Other otologic disease processes should be considered as well. Most authors discourage performing stapes surgery in patients with Meniere's disease, especially when it is active. Some feel that any vestibular symptom is a contraindication to surgery. This is because patients with endolymphatic hydrops have a much greater risk of SNHL after stapes surgery. It has been proposed that this is due to intra-operative injury to the saccule which is dilated and abuts the oval window. Further testing in these patients with electronystagmography and electrocochleography should be considered. Tympanic membrane perforations should be repaired prior to stapedectomy. Active otitis media or the presence of cholesteatoma is a contraindication to stapedectomy.

Patients with downslowing sensorineural loss as part of a mixed loss usually have poor speech discrimination, which does not improve after surgery and may even worsen. In addition, these patients are considered to have a "compromised cochlea" with sensorineural loss often worsening after any degree of intra-operative trauma.

## Surgery

When surgery is decided upon, the worst hearing ear should be approached first. The basic order of steps in stapes surgery for otosclerosis have remained relatively constant over the past 30 years. What have changed are subtleties in technique and style in performing each of these basic steps. This starts with the anesthetic choice. This is determined by the surgeon's preference, patient wishes, and medical condition of the patient. Those who use local anesthesia with conscious sedation tend to be experienced otologic surgeons in a non-teaching atmosphere. The advantages cited are that hearing restoration can be immediately evaluated and that the patient can report symptoms of vertigo. In addition, local anesthesia is useful in patients with pre-existing medical conditions that preclude them from general anesthesia. General anesthesia is useful in teaching institutions where inexperienced surgeons take longer to perform the operation. In these cases, patient comfort is improved. Hearing results are similar for both methods.

The patient is seen initially in holding and the correct ear to be operated on is confirmed with the patient and by the audiogram and CT (if available). The ear is then marked externally to ensure that there is no confusion with the rest of the OR staff. The CT scan and audiogram are then placed in a visible location in the OR for ease in intra-operative reviewing. Once anesthesia has been attained, the patient's ear is prepped, usually with Beta-dine (if there is no history of allergy to iodine), and the ear is draped in a sterile fashion. The external auditory canal is injected with 1% lidocaine with epinephrine in four quadrants at the bony cartilaginous junction.

**Raising the tympanomeatal flap:** The tympanomeatal incision is carried out using a variety of cutting devices (roller knife, round knife, or various straight blades). The canal skin is incised at the six and twelve o'clock positions and are carried out approximately 6-8 mm lateral to the annulus. A lateral curved cut parallel to the annulus connects these incisions. It is important to make the tympanomeatal incision lateral enough to the annulus to allow enough skin to drape back over the scutum which will be partially removed. If the flap is too big, however, it will become difficult to retract it out of the way of the operating field. The flap is elevated with a round knife anteriorly to the annulus. Cotton soaked with adrenaline can be helpful in with elevation and controlling bleeding. The annulus is then removed from its sulcus with a Rosen needle. The chorda tympani nerve is identified and is separated from the TM. The nerve should be preserved if possible. Releasing its attachments to the medial surface of the malleus can increase the slack on the nerve and allow it to be move out of the way without stretching it. In some cases it is in the way of the operating field, despite maneuvers to increase its mobility. In these cases, it should be cut rather than stretched out of the way. Patients develop more severe and prolonged dysgeusia from stretching the nerve than from cutting it. In addition, there is a risk of retrograde inflammation to the facial nerve with possible delayed facial nerve paralysis.

**Exposure:** Removal of a portion of the scutum is almost always necessary for adequate visualization of the stapes footplate, oval window niche, and surrounding structures. A curette is used to remove bone just lateral to the edge of the scutum first. This thins the scutum and allows for easy removal. Attempts to remove the edge first may result in slipping of the instrument and injury to surrounding structures. It is important to remove enough bone posteriorly to allow for visualization of the pyramidal eminence and superiorly to allow visualization of the inferior

border of the facial nerve. A small amount of bone should be left over the incus superiorly to prevent retraction of the tympanic membrane onto the lateral surface of the ossicles.

**Middle ear examination:** All three ossicles should be palpated for fixation prior to and after division of the incudostapedial joint. If malleus or incus fixation is encountered, these must be addressed as well to attain optimal hearing results. Occasionally a very small focus of active otosclerosis is found, usually at the anterior pole of the stapes footplate. Gentle palpation of the footplate will sometimes mobilize it. The patient can then be followed and treated with sodium fluoride to convert the focus into the sclerotic phase. In general, re-fixation usually occurs, making this technique less appealing. The tympanic segment of the facial nerve should be examined and overhang into the oval window niche as well as any bony dehiscence should be noted. Dehiscence occurs in 50% of all ears and usually occurs on the medial and inferior aspect of the nerve. The distance between the footplate and the medial surface of the lower aspect of the long process of the incus is then measured (usually 4.5 mm). About 0.25 to 0.50mm is added to the measured distance to accommodate entrance of the prosthesis slightly in to the vestibule. Measurement before disarticulation of the incudostapedial joint may be more accurate, since movement of the incus is possible with joint disarticulation. Some surgeons still prefer to perform the measurement after disarticulation.

**Harvesting a graft:** Multiple grafts have been used as an oval window seal including vein (usually from the hand), temporalis fascia, tragal perichondrium, ear lobule fat, and periosteum. Gelfoam has been abandoned due to its higher incidence of post-operative reparative granuloma formation and SNHL. The graft is harvested prior to footplate fenestration or removal so that it is immediately available for use to prevent excessive perilymph leakage. This is particularly important if a perilymph gusher is encountered for immediate patching.

**Stapedectomy/Stapedotomy:** Decision to perform total or partial stapedectomy versus stapedotomy depends on the extent of stapes fixation and other characteristics of the foot plate, and surgeon preference. Hough 1960 suggested performing a partial anterior stapedectomy for small anterior foci of otosclerosis. He performed this technique by fracturing the middle of the footplate and anterior crus and extracting the anterior half of the stapes. He did this without cutting the stapedius tendon. This has a potential advantage in patients who work around loud noises.

Total stapedectomy has been recommended for extensive fixation of the footplate and by some authors for a floating footplate. The incudostapedial joint is first divided with a round or joint knife prior to division of the stapedius tendon. Pressure is applied at the joint, parallel and opposite that of the tendon. The tendon supports the footplate preventing inadvertent avulsion and inner ear trauma. Howard House recommends placing a small hole with a pick into the center of the footplate (if it is thin) prior to division of the joint. This allows a place for a hook to be used to extract the footplate if it becomes mobile during manipulation of the joint. Incus and malleus mobility are then checked. The stapedius tendon is divided with microscissors or a laser. A curved pick is then used to down fracture the superstructure towards the promontory and is then extracted. The footplate is then removed with right angle hooks. Autologous tissue graft is placed over the oval window and an appropriate sized piston or prosthesis is placed from the long process of the incus to the graft.

Stapedotomy or the small fenestra technique was originally used in patients with obliterated or solid footplates. It was initially performed in Europe and began to gain acceptance in the United States in the late 1970s and 80s. Because of the early success of this technique, many began using stapedotomy for non-obliterative cases. The advantage of stapedotomy include less risk of trauma to the vestibule and less incidence of migration of the prosthesis and fixation by scar tissue as is seen in stapedectomy techniques. Initially sharp instrumentation was used to make the fenestration which required a great degree of technical proficiency. In 1978, the first laser stapedotomy was performed by Perkins and has since become a widely accepted method for fenestrating the footplate.

The three most commonly used lasers for stapedotomy are the potassium titanyl phosphate (KTP/532), argon, and carbon dioxide lasers. The KTP/532 and argon lasers have similar biosurgical effects. They both are visible beams of light and therefore, do not require a separate laser to act as an aiming beam. This allows for far greater target accuracy because they act as their own aiming beam. The CO<sub>2</sub> laser is an invisible surgical beam and must be used with a separate aiming beam, usually a red helium-neon beam which produces an ill-defined fuzzy border. The CO<sub>2</sub> beam must be perfectly lined up with the aiming beam or the target will be off center. Both the KTP/532 and the argon lasers are absorbed by hemoglobin making them better coagulators than the CO<sub>2</sub> laser. The CO<sub>2</sub> laser is less expensive and requires less maintenance. When using the laser, it is important to evaluate all equipment in the pre-operative period to ensure proper function. The risk of using the any laser in the oval window area is overheating of surrounding tissue including the facial nerve. Causse recommends allowing 10 to 15 seconds between pulses to allow for cooling. In addition a suction must be used to evacuate the smoke and inadvertent suctioning of perilymph may occur.

In addition to the laser, a small microdrill can be used to create a fenestration in the footplate. Most commonly a 0.7mm diamond burr is used. Some surgeons use the drill after using the laser to complete the fenestration which allows the turning of the drill bit to remove bone dust and fragments (Causse et al., 1993). The fenestration site is usually aimed at the center of the footplate, however, Causse recommends placing the fenestration more posteriorly. He argues that because the stapes actually produces a vestibular fluid wave by rocking posteriorly, placing the piston into a more posterior fenestration is more physiologic.

**Sealing the oval window and placement of the piston:** As mentioned previously, multiple substances can be used with equal efficacy in sealing the oval window. Causse is a proponent of vein grafts. He makes the argument that it is important to reconstruct the annular ligament. The function of the annular ligament is to provide both resistance (dissipation force) to protect the inner ear from acoustic trauma and elasticity for acoustic transfer (disrupted by otosclerosis). Because of the inherent elastic properties of vein, it is better suited to recreate this physiologic function. Vein, fat, and perichondrial grafts require additional incisions unlike temporalis fascia or blood.

Once the oval window has been sealed the appropriately sized prosthesis is placed into the fenestra and around the incus. Multiple stapes prostheses have been developed over the past 30 years. The sharp or beveled polyethylene strut developed by Shea is associated with higher incidence of postoperative fistula, so it is rarely used today. Similarly, the wire prosthesis attached to compressed gelfoam has been associated with higher incidence of postoperative

fistula, due to reparative granuloma formation. Most stapedectomies today are repaired with a wire prosthesis attached to a connective tissue graft that covers the oval window. Stapedotomies are usually repaired with a piston type prosthesis of 0.6 mm or 0.8 mm diameter, which again rests on a connective tissue graft. The prosthesis usually attaches to the long process of the incus, either with a wire that is crimped into place, or a bucket handle that fits under the lenticular process of the incus. After placement of the prosthesis, the malleus is palpated to insure appropriate movement of the repaired ossicular chain. One consideration when choosing the diameter of the prosthesis to use is how wide the oval window niche is. A deep narrow niche may require a thinner prosthesis such as a wire. If the patient is awake, hearing can be tested by whispering into the patient's ear. The tympanomeatal flap is then redraped and the ear canal is packed with Gelfoam.

## **Special Considerations and complications in stapes surgery**

### ***Overhanging and dehiscent facial nerve***

Occasionally the facial nerve is dehiscent in the fallopian canal and protrudes over the oval window niche such that it obstructs the view of the stapes footplate. If the surgeon is not familiar with management of this problem the surgery should be aborted. Facial nerve displacement can be used for conventional manual stapedectomy or in laser stapedotomy. Perkins (2001) describes the maneuver for use with the laser. The nerve is first displaced to allow for examination of the footplate. This is done by compressing it superiorly for short periods of time (5 seconds) using No. 24 suction. 10 to 15 seconds is allowed to lapse before performing the maneuver again. This should be repeated until adequate assessment of the footplate is complete. Then, while the nerve is retracted and the footplate visualized, the laser is pulsed and the usual rosette pattern is created. A wire type prosthesis is then used. It can be bent to accommodate the facial nerve, however hearing results will not be as good. An addition 0.50 to 0.75mm should be added to the selected length of the piston to allow for curve around the nerve.

### ***Floating footplate***

A floating footplate occurs usually during the course of a stapedectomy when the footplate dislodges from the surrounding oval window niche. It may be encountered incidentally but is more commonly due to iatrogenic causes such as palpation of the footplate with a pick or the when burring a fenestra, or during division of the incudostapedial joint. The first issue in management of the floating footplate is prevention. Use of the laser to perform stapedotomies may prevent this complication by reducing direct manipulation. H. House recommends drilling a safety hole into the footplate at the beginning of the procedure to help in the removal of the footplate should it become mobile.

There are several ways of managing a floating footplate once it has occurred. If the surgeon is inexperienced, termination of the procedure and referral to someone more familiar with this complication is not unreasonable. One way to manage this problem is to perform a total stapedectomy. Proponents of this technique do so to prevent a sinking footplate. Attempts at removing the footplate can, however, result in fragments of the stapes falling into the vestibule. These fragments should not be removed. Howard House (2001) recommends drilling down the posteroinferior promontory to allow access of a right angle cooked underneath the

footplate without touching it. It is then elevated toward the facial nerve and is removed. Other surgeons advocate performing a fenestra with the laser. In these situations hearing results are adequate (97% ABG closure) if the footplate is thin or blue (diffuse footplate). If the footplate is very thick or white (“biscuit footplate”), hearing results are much poorer (52% ABG closure) and re-fixation occurs in 30%. Perkins recommends that if the laser is not available, the surgery should be postponed for 4 months followed by laser stapedotomy.

### ***Diffuse obliterative otosclerosis***

When the footplate, annular ligament and edges of the oval window niche are all involved with otosclerosis it is termed “obliterative otosclerosis”. These cases are managed by thinning the bone with a small cutting burr. First the anteroposterior edges are thinned until they are blue. Then the opening is enlarged sufficiently to cover the area with a graft and to insert a prosthesis. Long-term hearing results for obliterative disease are poorer, with re-ossification and obliteration occasionally occurring.

### ***Perilymph gusher***

This complication is associated with a patent cochlear aqueduct and results in excessive perilymph leakage after fenestration of the footplate. It occurs more common on the left side and in patients with congenital footplate fixation. A perilymph gusher increases the risk of post-operative SNHL. Another benefit of the control hole placed initially during the surgery is that it can lead to early identification of a gusher. Management includes roughing up the surface of the footplate followed by rapid placement of an oval window seal. Completion of stapes reconstruction with a prosthesis can still proceed, however stapedectomy is contraindicated. Postoperatively the patient should be treated as if they had a cerebrospinal fluid leak or perilymph fistula with the head of the bed elevated, bed rest, stool softeners, avoidance of Valsalva maneuvers, occasionally a lumbar drain.

### ***Round window closure***

Unlike the oval window, hearing loss is not affected by closure of the round window unless it becomes complete. Upwards of 50% of patients with otosclerosis have round window involvement, however <1% have complete closure affecting hearing. In these patients, surgery should be avoided because they have a higher risk of post-operative SNHL.

### ***Sensori-neural hearing loss***

Permanent sensori-neural hearing loss is the most dreaded complication of stapes surgery. The incidence of this complication ranges between 0.6% and 3% in large series. The incidence depends on multiple factors including extent of disease (particularly the presence of cochlear involvement), experience of the surgeon, techniques employed, age of the patient, and history of prior surgery. Most authors report that in experienced hands, the risk should be  $\leq 1\%$  for non-revision cases.

SNHL after stapes surgery may be either temporary or permanent. Most patients develop some degree of serous labyrinthitis in the first couple of postoperative days which is manifested clinically by gait unsteadiness, vertigo with head movement, and a slight decrease in hearing for frequencies above 2 kHz, along with reduced speech discrimination. By definition, all symptoms

including hearing loss resolve. Reparative granuloma may cause temporary SNHL if it is addressed within the first two weeks of development.

Permanent SNHL may result when serous labyrinthitis becomes suppurative. More commonly it is due to direct trauma during surgery particularly if extensive drilling is carried out. This is confounded in patients with extensive cochlear otosclerosis with endosteal involvement. In these cases the cochlear is fragile and prone to basilar membrane breaks. Other potential causes of permanent SNHL are large or sudden drops in perilymph pressure (perilymph gusher, or extensive suctioning of perilymph), impaired cochlear blood flow following neck torsion during surgery, and medial displacement of the prosthesis.

### ***Facial nerve paralysis***

The risk of facial nerve paralysis is always present which any manipulation of the middle ear structures. Despite the close proximity of the nerve in stapes surgery, the risk of injury and paralysis is low (<1/1000 cases). In primary stapes surgery it may be a result of a couple of factors. Of course direct trauma with cold instrumentation or with the laser may occur. If the laser is pulsed too frequently, the nerve may become heated and swell. As mentioned previously, stretching of the chorda tympani nerve may result in retrograde inflammation of the facial nerve. The majority of cases of facial nerve paralysis are incomplete and temporary. A steroid taper is usually given and resolution of the paralysis or paresis can be expected in several weeks

### ***Vestibular symptoms***

The purpose of performing stapes surgery under local anesthesia besides immediate hearing evaluation is to avoid injury to the sacculle. There are multiple other sources of vestibular stimulants during these surgeries, however. These include irrigation around the lateral semicircular canal, palpation of the stapes, and local anesthetic. Some authors feel that these confounding factors make intra-operative monitoring of vestibular symptoms less reliable.

Postoperative vestibular symptoms are usually short lived but may persist for several days and less commonly up to months. Small fenestra techniques, particularly with the laser, have been shown to reduce the frequency, severity, and duration of vestibular symptoms after stapes surgery when compared to stapedectomy techniques.

An oval window fistula should be suspected in patients with vertigo and associated fluctuating or progressive SNHL and tinnitus. This complication usually occurs early in the postoperative course. A history of sudden barometric trauma should be elicited from the patient and documented and audiometry should be performed. If there is a high index of suspicion, middle ear exploration should be performed. Findings may include a medially displaced prosthesis with or without perilymph leakage. The oval window should be patched. Progression of the hearing loss should be halted and vestibular symptoms should diminish.

### ***Reparative Granuloma***

Reparative granuloma is a well known cause of SNHL after stapes surgery. It was more commonly reported when gelfoam was used as an oval window sealant. The incidence is estimated to be between 1% and 2% of cases. The usual presentation is patient with an initial

good hearing result who develops a drop in their bone conduction thresholds approximately one to two weeks postop with associated tinnitus and vertigo. On physical exam, a reddish discoloration can be seen through the TM posteriorly. Treatment is exploration and removal of the granuloma which usually surrounds the prosthesis and incus. Most patients can expect near complete resolution of their original postop hearing if the problem is addressed early in its course.

### ***Recurrent conductive hearing loss and revision stapedectomy***

The most common cause of failure of stapes surgery for otosclerosis is displacement or slippage of the prosthesis. Increase ABG may occur immediately secondary to technique or over months to years secondary to gradual thinning of the incus with subsequent slippage of the prosthesis. If the prosthesis is placed too tight around the incus compression necrosis may result. The tip of the prosthesis sometimes becomes adherent to the edge of the niche. Other causes may include stapes re-fixation or continuation of disease progression with re-obliteration of the oval window niche and ankylosis of the malleus or incus.

Any of these causes may be an indication for revision stapes surgery. It is important to counsel patients extensively on the reduced success rate of revision surgery. Glasscock reported on 828 cases of stapes surgery from 1970-1994. Air-bone gap closure to within 5 dB was achieved in 91% of primary operations compared to 66% of revision cases. The best candidates for revision surgery are those patients who had excellent results from their first surgery and have experienced gradual worsening of the ABG over a period of months to years.

### ***Non-surgical***

**Amplification:** A hearing aide is an acceptable option for patients who do not want to undergo surgery for otosclerosis or in those patients who are not fit for surgery. Certainly the option of amplification as an alternative to surgery should be discussed with all patients. The benefit over surgery is the avoidance of the risk of significant SNHL. Patients who undergo successful surgery have a higher satisfaction rate than those wearing hearing aids. This is probably due to a couple of reasons. The first is the canal occlusion effect in which the hearing aide gives the patient the sensation they are hearing in a barrel. The other is the fact that amplification is not used at night.

**Medical:** In 1923, Escot was the first to suggest that calcium fluoride be used in the treatment of otosclerosis. It was later popularized by Shambaugh (1965). The fluoride ion replaces the usual hydroxyl ion in periosteal bone, forming fluorapatite, instead of the usual hydroxyapatite. This results in decreased bone resorption and increased calcification of new bone. Actively expanding foci of otosclerosis are inactivated, as has been documented by computed tomography. In addition, tinnitus and imbalance are reduced, and Schwartze's sign frequently becomes negative. The most frequent side effects are rash, arthritis, and gastric distress. Permanent serious side effects are very rare. It is available over the counter as Florical (sodium fluoride and calcium carbonate), and the usual dose is about 20-120mg of fluoride a day in adults. This dosage is decreased for children and pregnant women. After two years, the efficacy of the treatment can be evaluated. Schwartze's sign, and the degree of tinnitus and imbalance are reassessed, and a CT scan is repeated. If overall stabilization of the disease has occurred, the patient is placed on a life-long maintenance dose of about 25mg of fluoride a day.

The use of sodium fluoride has been heavily debated. Schuknecht found no evidence of histologic improvement with fluoride treatment. Hough concluded that there is no evidence that fluorides help in otosclerosis. Shambaugh, however, supported the use of fluoride, and reported a long term clinical series that “demonstrated its value in arresting previously progressive sensorineural hearing loss.” Causse published a placebo controlled study of fluoride in otosclerotic patients. Although no significant hearing improvement was noted, the number of patients who remained stable with therapy was much higher than placebo. He concluded that “there should no longer be any hesitation in prescribing sodium fluoride ... for patients with otospongiosis who show progressive sensorineural hearing involvement.”

Sodium fluoride treatment should be considered in patients who are not surgical candidates, those who decide against surgery, and those who have SNHL or vestibular symptoms believed to be due to otosclerosis. In addition, patients who are candidates for surgery with an active focus detected by a positive Schwartze’s sign may be given fluoride treatments for 6-12 months prior to surgery to induce the focus to mature and potentially prevent the progression of disease after surgery. If the focus is determined to be active during surgery, postoperative treatment can be initiated. Overall, 50% of patients have stabilization of their disease, 30% improve, and the rest continue to progress. 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.

Bisphosphonates have recently been administered to patients with active otosclerosis and neurotologic complaints such as dizziness or Meniere’s symptoms. These patients have are generally considered poor candidates for surgery. Initial results with Etidronate have been promising, with large numbers either improving or resolving. However, long-term follow-up is lacking. In addition, recent reports of bisphosphonate induced osteonecrosis bring long-term therapy with this medication into question.

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