INTRODUCTION

Otosclerosis is a bone disease only seen in the otic capsule. It causes hearing loss which may be conductive, mixed, or sensorineural hearing loss. Toynbee first described the condition in 1860 as causing a hearing loss by fixation of the stapes. In 1894, Politzer referred to the fixation of the stapes as otosclerosis. Siebenmann revealed on microscopic examination that the lesion seemed to begin as spongification of the bone and termed the process otospongiosis.

Otosclerosis clinically presents with progressive hearing loss. If the otospongiotic change primarily involves the stapes, then the hearing loss is conductive. The fissula ante fenestram is the most common area for stapedial fixation. The process may progress to involve the entire footplate or may continue anteriorly toward the cochlea, causing a sensorineural hearing loss.

Otosclerosis is an autosomal-dominant hereditary disease. It has variable penetrance and expression. Women are effected by otosclerosis 2:1. Hearing loss usually begins in the late teens or early twenties, but may occur later. The prevalence of otosclerosis varies with race and its expression. The disease is found between 7-10% of cadaveric temporal bones in Caucasians. Clinical otosclerosis is rare in blacks, Asians, and Native Americans.

HISTOPATHOLOGY

Early lesions tend to begin near the fissula ante fenestram. They begin as connective tissue replacing bone, which in turn is remodeled by osteocytes resulting in disorganized bone with enlarged marrow spaces. When this process involves the mobility of the stapes, a conductive hearing loss results. Occasionally, the lesion can involve only the cochlea, causing an isolated sensorineural hearing loss.
EVALUATION

History

Patient history is one of the most important aspects of the evaluation of the otosclerosis patient. Hearing loss usually gradual in onset and slowly progressive over several years. Approximately 70% of otosclerosis cases are bilateral. Typically, hearing loss becomes apparent in the late teens or twenties, but may not become apparent to the patient until age 30 or 40 years. As is typical with conductive hearing loss, patients will report difficulty hearing conversation while chewing and may hear better in noisy rooms because people speak louder in noisy surroundings. Unilateral hearing loss is less noticeable to the patient and results in difficulty with direction of sound and in noisy rooms. Family history is usually positive for hearing loss often with surgical correction.

Physical Examination

Physical examination includes the following:

1. Otoscopy (often with the operating microscope)- look for Schwartze sign which is a red blush over the promontory or the area anterior to the oval window

2. Pneumo-otoscopy- evaluates for middle ear effusion or small perforation

3. Tuning fork exam- may confirm or dispute the finding of a conductive hearing loss on audiometry

Audiometry

The standard audiometric evaluation includes air conduction, bone conduction, and speech audiometry. Additionally, the immittance audiometry battery consists of tympanometry, static compliance, and acoustic reflex testing. The middle ear pressure is not affected by otosclerosis, and, therefore, the tympanogram is normal with a distinct peak that occurs with the normal range. The peak may be lower (As) than normal in the presence of a healthy appearing tympanic membrane, thus alerting the examiner to the possible diagnosis. Acoustic reflexes are a sensitive measure of the movement of the stapes, which in the presence of advanced otosclerosis, the reflex will be absent.

SURGERY

As with all surgery, surgical options for treatment of otosclerosis involves risk. Total sensorineural hearing loss occurs in about 0.2% of cases, but the patient is told that there is a less than 2% chance of further hearing loss and a less than 1% chance of losing all hearing in the operated ear. Dizziness may occur postoperatively also. Dizziness is usually transient and brief. However, it may persist for a short period of time and rarely could be permanent. The possibility of facial palsy should be mentioned as well.

Surgical techniques include stapedotomy or stapedectomy performed with either a laser or microdrill. Please refer to otology texts for further description of surgical techniques.
Multiple problems can be encountered during stapes procedures. These are listed below:

- **Exposed, Overhanging Facial Nerve**

  An exposed facial nerve occurs in approximately 9% of stapes procedures. It may block access to the footplate, making the completion of the procedure impossible. Gentle retraction of the nerve superiorly with a small suction while the drill or laser is used to create the fenestra is possible. If the prosthesis is touching the facial nerve it generally does not create a problem for postoperative hearing or facial function.

- **Fixed Malleus**

  A fixed malleus is a rare problem, but it should be checked for in every procedure. When this occurs, the sound conduction can be reestablished with an incus replacement prosthesis or a total ossicular replacement prosthesis and tragal cartilage.

- **Solid or Obliterated Footplate**

  A solid or obliterated footplate can be managed with a microdrill to create a fenestra. It was a greater problem in the past when a total stapedectomy was performed.

- **Floating Footplate**

  A floating footplate rarely occurs when using the laser or microdrill, especially when the crura are left in place until after the prosthesis is placed. Management includes carefully drilling a small hole in the promontory at the inferior edge of the footplate followed by using a small hook to gently elevate the footplate out of the oval window.

- **Perilymph Gusher**

  A perilymph "gusher" is the profuse flow of cerebrospinal fluid (CSF) immediately on opening the vestibule. It is rare with a reported incidence of 0.03% and is most often associated with congenital footplate fixation in the pediatric population. The etiology of this CSF leak is thought to be due to either a widened cochlear aqueduct or a defect in the fundus of the internal auditory canal. Management involves placement of a tissue graft over the oval window and completion of the procedure, if possible, rather than packing the ear and terminating the surgery. Placement of a lumbar drain can also be used to reduce CSF pressure.

- **Tympanic Membrane Perforation**

  A tympanic membrane perforation may occur during elevation of the tympanomeatal flap. Perforation does not preclude completion of the operation. Repair involves myringoplasty with either synthetic material or autologous tissue.

- **Chorda Tympani Nerve Damage**

  Damage to the chorda tympani nerve may occur in up to 30% of cases secondary to stretching and mobilization of the nerve during removal of the posterosuperior bony canal wall.
Sequela include complaints of temporary dry mouth, tongue soreness, or a metallic taste that usually subsides in 3 to 4 months. Symptoms are less severe with complete sectioning of the nerve rather than stretching or partial tearing.

**Intraoperative Vertigo**

When the prosthesis is too long, vertigo may result. Vertigo also may be induced when checking the mobility of the prosthesis. In this situation, a shorter prosthesis generally corrects this problem. If the vertigo does not resolve, the prosthesis can be removed and replaced with a 0.25-mm shorter piston.

**POSTOPERATIVE COMPLICATIONS**

**Sensorineural Hearing Loss**

The most devastating complication of stapes surgery is sensorineural hearing loss which occurs in less than 1% of cases. Sensorineural hearing loss may be mild or isolated to high frequencies. When sensorineural hearing loss is suspected, prednisone is started immediately and tapered.

Serous labyrinthitis is common after stapedectomy due to inner ear inflammation. Patients may exhibit mild unsteadiness, positional vertigo, and/or a slight decrease in high-frequency hearing. The above symptoms typically resolve within several days to weeks, correlating with the resolution of the serous labyrinthitis.

Postoperative reparative granuloma is rare and has previously been recognized as a cause of sensorineural hearing loss after stapedectomy. Patients present with initial improvement in hearing followed by a gradual or sudden deterioration 1 to 6 weeks postoperatively. Vertigo can also occur and clinical examination often reveals a reddish discoloration in the posterosuperior quadrant of the tympanic membrane. Treatment consists of prompt recognition and removal of the granuloma from around the oval window.

**Vertigo**

Mild vertigo or dizziness is fairly common and occurs in about 5% of cases. It is rarely prolonged or severe and usually lasts for a few hours, subsiding rapidly. Management is usually not necessary or may be supportive only.

**Facial Paralysis**

Rarely, delayed onset of facial palsy occurs postoperatively usually occurring in the 5 day post-operative setting. It typically lasts for a few weeks. It is usually incomplete and responds quickly and completely to prednisone.

**Tinnitus**

Preexisting tinnitus will usually resolve in patients after stapes surgery. However, some patients will complain of new-onset tinnitus. As discussed previously, tinnitus is possibly related
to serous labyrinthitis and may improve as the ear continues to heal. When tinnitus persists, patients are treated with reassurance and routine tinnitus measures.

**Taste Disturbance**

Taste disturbance occurs in approximately 9% of patients. Stretching of the chorda tympani is usually the cause of this rather than nerve sectioning. Therefore, if the nerve has been stretched or otherwise injured, it is preferable to section it. Usually taste disturbance resolves in 3 to 4 months.

**Tympanic Membrane Perforation**

A small marginal perforation may be repaired by freshening the edges and applying a paper patch. If the perforation has not healed, the process is repeated. If this still fails, a myringoplasty can be performed.

**Perilymph Fistula**

Perilymph fistula is a rare complication after stapes surgery with an incidence ranging from 3 to 10%. With the use of a small fenestra technique, fistula is rarely seen as a cause of failure in stapes surgery. The patient presents with a mixed conductive sensorineural hearing loss with some vague unsteadiness and, rarely, vertigo. In order to repair this, the prosthesis is carefully removed, a tissue seal is placed over the open oval window, and the prosthesis is replaced. A laser is helpful to remove granulation and scar tissue.

**SPECIAL CONSIDERATIONS**

**Ménière's Disease**

Endolymphatic hydrops may occur due to otosclerosis or as two separate disease entities. Stapedectomy in the presence of uncontrolled Ménière's disease can potentially result in a dead ear and should be avoided. A review of patients with otosclerosis and Ménière's disease at the House Ear Clinic revealed that stapedectomy does not increase the risk of sensorineural hearing loss for patients with bone conduction thresholds of 35 dB or better at 500 Hz and no high-tone hearing loss. Stapedectomy is contraindicated for patients with levels of 45 dB or greater at 500 Hz and with high-tone loss. In this later group of patients, postmortem histopathologic analysis revealed contact of the saccular membrane or Reissner's membrane with the stapes footplate, which increased the risk of significant postoperative sensorineural hearing loss.
Bibliography


