Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 3: Middle Ear and Mastoid

Chapter 38: Management of Otosclerosis

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Otosclerosis is a primary disease of the labyrinthine capsule transmitted, in most cases, by an autosomal dominant gene. It represents a constant process of bone resorption and redeposition, and manifests progressive stable conductive, or mixed hearing loss. Otosclerosis occurs clinically to varying degrees in 0.5 to 1.0 per cent of the population, is usually bilateral, and becomes clinically apparent in the teens, second, third, and fourth decade. Women appear to be slightly more susceptible than men, and pregnancy appears to have an adverse effect on the process.

In 1873, Schwartze described the finding of a pink blush behind the tympanic membrane secondary to hyperemia of the promontory mucous membrane in active otosclerosis. This sign now bears his name. Von Troltsch in 1881 coined the term "otosclerosis" from the sclerosing changes observed in the tympanic mucosa. Politzer in 1893 was the first to describe otosclerosis correctly as a primary disease of the labyrinthine capsule, rather than an entity that occurred subsequent to chronic middle ear catarrh, as had been formerly believed.

Conductive hearing loss subsequent to otosclerotic involvement of the stapes footplate is well known and easily appreciated. Less well understood is the sensorineural hearing loss that may occur. Siebenmann in 1912, and later Wittmaack, in 1919, speculated that otosclerosis might cause sensorineural hearing loss as the result of the accumulation of toxic or inflammatory materials in the membranous labyrinth. Although much histologic and biochemical research and much speculation has followed these initial observations, many questions regarding otosclerosis and sensorineural hearing loss remain unresolved. The clinical diagnosis of otosclerosis, as clarified by Bezold in 1908, required a careful history, physical examination, tuning fork evaluation, and audiometric testing. Radiographic examinations are available as adjuncts when needed. Therapeutic modalities include surgery, medicine, and amplification, alone or in combination.

Surgery

Surgery directed at correcting the auditory deficit incurred with fixation of the stapes footplate applies not only to patients with otosclerosis but also to patients with osteogenesis imperfecta, congenital footplate fixation, or fixation due to tympanosclerosis. It must be noted, however, that in patients with congenital footplate fixation, often the annular ligament is less well delineated and the cochlear aqueduct will be widely patent in a higher incidence of patients,

resulting in perilymph "gusher".

Fenestration

Inner ear fenestration to bypass the fixed stapedial footplate was first reported by Passow in 1897. He attempted to create a new site for energy transfer by drilling a hole in the middle ear promontory. Bárány in 1911 observed improved hearing in a patient with otosclerosis and chronic otitis media following inadvertent fenestration of the horizontal semicircular canal during radical mastoidectomy. Subsequent attempts to reproduce this result consistently were made by Bárány and Jenkins in 1913. It was Holmgren who, in 1917, pioneered a long series of fenestration operations for otosclerosis and proved that the inner ear could be opened safely with sterile technique. Holmgren also deserves credit for introducing the operating microscope into otosclerosis surgery. However, his attempts to maintain a permanent fistula were almost always unsuccessful, and it remained for Sourdille, in 1937, using a complex multistage tympanolabyrinthopexy, to achieve consistent, reproducible, and durable results. Julius Lempert in 1938 developed the one-stage operation for otosclerosis, simplifying Sourdille's procedure and making fenestration practical. He trained other otologists in the techniques of this one-stage operation, enabling a host of otosclerotic patients to obtain improved hearing. Although the fenestration operation is rarely performed today for otosclerosis, Lempert's techniques and meticulous attention to detail helped lead to the development of much current otologic thought.

Lempert's operation consists of bypassing the ossicular chain by fenestrating the lateral semicircular canal. This procedure should not be done in the presence of infection and is not helpful on patients who have a significant sensorineural hearing loss with otosclerosis. It is not indicated for routine treatment of otosclerosis today, but in proper hands may rarely be indicated when routine stapedectomy cannot be done for stapes fixation (eg, congenital absence of the oval window or a facial nerve that completely covers the oval window).

With the use of endaural incisions and exposure a tympanomeatal flap is elevated, with special care, from 6 o'clock inferiorly to 9 o'clock (left ear) or 3 o'clock (right ear) anteriorly. A mastoidectomy is performed for exposure and the "bridge" and anterior and posterior buttresses are removed. The incus is identified and saved for future homograft work. The head and neck of the malleus are amputated. The operative field, especially in the region of the dome of the lateral semicircular canal, should be dry and without bone dust. In the cupula technique, two blue lines are outlined and the cap of bone over the fistula is removed in one piece. The tympanomeatal flap is then placed into position and invaginated gently into the fenestra. The ear is packed with nonadherent surgical rayon in rosebud fashion. The packing is removed after 10 days. The patient generally has some vestibular upset secondary to serous labyrinthitis which subsides after a few days.

Stapes Mobilization

The first stapes mobilization was employed by Kessel in 1878. Boucheron mobilized 60 stapes in 1888, and in 1890 Minot mobilized 200 stapes and described the operation in detail.

In 1900 Politzer and Siebenmann condemned stapes mobilization, which discredited the procedure for decades. Rosen (1953) deservedly gained recognition for rediscovering mobilization during a fenestration operation and for introducing the mobilization operation for otosclerosis in modern times. He thereby redirected otologic thinking toward a direct approach to the ankylosed stapes.

Stapes mobilization is rarely used at present to treat otosclerosis, but these techniques are useful to determine stapes mobility. There are situations, as for example in tympanoplasty in which the stapes is found to be fixed by tympanosclerosis, in which mobilization is indicated.

Stapedectomy in a Previously Fenestrated Ear

This technique was originated by Schuknecht (1960) and popularized by Sheehy (1962). Stapedectomy can be considered for patients in whom a significant conductive loss remains with good cochlear reserve. A tight tympanic membrane and a good middle ear space are desirable. A large tympanomeatal flap is elevated in such cases, with care being taken not to traumatize the previous fenestration site, which may or may not be healed over by solid bone. A stapedectomy is done and a measured wire-connective tissue prosthesis (5.5 to 6.5 mm) is connected between the malleus and oval window, after which the flap is replaced and held in place by packing. In our opinion it is dangerous to use a wire-piston type of prosthesis for this operation.

Stapedectomy

Stapedectomy is the current treatment of choice for the conductive component of otosclerosis. Dr. John Shea in 1956 revived the stapedectomy operation for otosclerosis and replaced the stapes bone with a polyethylene tube prosthesis and vein graft. The procedure of extracting the stapes for otosclerosis was first performed, however, by Jack of Boston in 1892 but was beset with obvious difficulties because he lacked proper magnification and antibiotic coverage. After, Shea, Schuknecht (1960) introduced the fat or connective tissue-wire prosthesis, further modified by House (1962) with the wire resting on a piece of gelatin sponge. Partial stapedectomy or anterior crurotomy was described by Hough (1960). Various modifications of the surgical techniques and prostheses used continue to date.

Preoperative Considerations

Patient Selection

Critical in achieving consistent satisfactory results in stapes surgery is proper patient selection. One must recognize that those who benefit from stapedectomy may also benefit from amplification and it is for the patient, with the physician's guidance, to determine which modality will best serve the particular condition. Stapedectomy is an elective procedure with minimal but real risks, and no patient should be persuaded toward surgery against his/her will. The patient should demonstrate a socially unacceptable hearing threshold (30 dB or worse), a negative Rinne at two of three speech frequencies, and good discrimination. The objectives of surgery are to improve the patient's ability to communicate, which may be accomplished by improving the

hearing to socially acceptable levels more suitable for successful amplification. Also, as Schuknecht pointed out (1960), it is important for patients to appreciate ambient sounds at about a 40 dB threshold (doorbell, thunder, telephone, dog bark) when not wearing amplification (eg, during the night).

Many patients have a severe mixed hearing loss and even complete deafness due to otosclerosis. If they have a conductive component and any residual speech discrimination, stapedectomy can be done. Improving hearing in these patients can be among the most gratifying results. The patient may become a better candidate for a hearing aid, whereas he or she could not function well with a hearing aid prior to stapes surgery; or, more dramatically, the patient can develop functional hearing with amplification postoperatively, compared with complete deafness preoperatively. Tuning fork testing and live-voice (shouting) testing while masking the opposite ear are helpful in selecting these patients. Audiologic studies are important, but if the patient has a conductive component on top of profound deafness, limits of the audiometer might not permit identification of this loss or measurement of discrimination, which can be measured best by an assessment by live-voice shouting of spondee words.

Another consideration in patient selection is age. Young children are not good risks for stapedectomy because of the appreciable incidence of recurrent eustachian tube dysfunction, predisposing them to changes in the position of their prosthesis and recurrent acute otitis media. Under the age of 20 years there appears to be a higher incidence of oval window reclosure following stapedectomy due to the activity of the otosclerotic focus, and therefore delay in surgical intervention may be wise. The elderly do quite well with stapedectomy and, unless there is a specific medical or otologic contraindication, these patients are satisfactory candidates. Anesthetic risk is minimal since most procedures are conducted under local anesthesia.

Occupation and lifestyle are important considerations in patient selections; different individuals reach differently to sensory deprivation or maintaining and wearing an appliance. Sudden changes in barometric pressure appear to pose increased risk to the stapedectomized ear as evidenced by a higher incidence of postoperative fistula formation in patients whose occupations or hobbies require repeated exposure to large and rapid pressure changes. These apparent clinical findings are not confirmed in stapedectomized monkeys exposed to experimental changes in barometric pressure. Persistent dysequilibrium which will occasionally occur postoperatively might prove incapacitating and, therefore, a relative contraindication for surgery to one whose occupation requires delicate balance. Prolonged noise exposure may carry some risk of further impairment in patients who have had a stapedectomy and suffered subsequent high-frequency sensorineural hearing loss. Presumably this risk is lessened if the patient's procedure was an anterior crurotomy in which the stapedial tendon was left intact.

Severe episodic vertigo is usually not caused by otosclerosis and may suggest underlying endolymphatic hydrops. Surgery on such an ear may cause a higher incidence of postoperative sensorineural hearing loss if care is not taken in patient selection. Surgery is absolutely contraindicated in the better hearing ear (and only hearing ear) or when active local inflammation or tympanic membrane perforation is present. When obstructing exostoses of the external auditory

canal are encountered, a two-stage procedure may be advisable. Surgery is not contraindicated in a previously operated ear assuming sufficient cochlear reserve and conductive component to work with. It must be noted, however, that endolymphatic hydrops has been identified in patients with otosclerosis and is felt by some to be secondary to the otosclerotic disease process itself (Liston et al, 1984). Also, it has been found that stapedectomy in patients with conductive hearing loss and dysequilibrium has on occasion not only benefited the hearing loss situation but the dysequilibrium as well (Paparella and Chasin, 1966).

A further consideration in patient selection centers around binaural hearing. The question of surgery for unilateral otosclerosis must be individualized, weighing the handicap of unilateral hearing loss carefully. Certainly, no surgery should be contemplated if the contralateral ear is normal and the otosclerotic ear poses no possibility of achieving a functional hearing, with or without amplification. Similarly, stapedectomy on the second ear requires individualized consideration. Important factors include the ease and success of the first operation plus the need for, and reality of, achieving binaural hearing. Some surgeons believe that the observed high frequency hearing loss following stapedectomy dictates against surgery on the second ear. A minimum of 12 months should elapse between operations, as this is the time span during which sudden postoperative hearing loss is most likely to occur.

Patient Counseling

An important aspect in discussing available therapeutic modalities is proper patient counseling. The pros and cons of each possible modality (surgery, medicine, amplification) should be carefully discussed in detail. This aspect is not only essential for good patient care but is a medicolegal necessity. The patient should be advised of the chief risk of stapedectomy - namely, *postoperative cochlear deafness.* The otolaryngologist should not cite the risk statistics (1 to 3 per cent) of the expert otologists in the United States, but should mention the risk in his own hands. Indeed, if the occasional microscopic surgeon finds that his complication rate is too high, he should desist in performing such surgery or better prepare himself so that the benefits will greatly outweigh the risks. It should be recognized that postoperative dysequilibrium may occasionally persist, causing variable handicap to the patient according to the particular life style. Disturbances in taste may follow manipulation or severance of the chorda tympani nerve, and residual tympanic membrane perforation may also occur. The risk of facial nerve paralysis in this procedure is negligible but does deserve mention, especially since one third to one half of facial nerves are dehiscent in the oval window area and may be damaged if alcohol is used in the preparation solution or from surgical trauma.

Immediate Preoperative Care

Preoperatively it has been the common practice to prescribe antimicrobials, although the value of this has not been proven. Sedation should be prescribed approximately 12 hours prior to surgery and again on the morning of surgery. Excess hair is removed with scissors and razor the morning of surgery and the ear is prepared and draped at the time of surgery.

Procedure

The procedure of stapedectomy can be performed under general anesthesia or using local anesthesia with sedation. Proponents of the latter believe that the patient's ability to report intraoperative vertigo may help the surgeon avoid injury to the saccule. Through a speculum placed in the external auditory meatus, the vascular strip is carefully injected with a solution containing a vasoconstrictor (eg, 1 per cent lidocaine with 1:100.000 epinephrine). A tympanomeatal flap is then developed by making a vertical incision from the 12 o'clock position and by a curvilinear incision from the 6 o'clock position. These incisions join approximately 5 to 6 mm from the annulus. A flap of this size should be adequate to cover any resultant defect but small enough to allow adequate exposure. The tympanomeatal flap is elevated, taking care to avoid tunneling or tearing of the annulus. This can be best accomplished with good hemostasis and good visualization. The chorda tympani nerve is identified and preserved if possible (or sacrificed if necessary, usually without any serious postoperative sequelae) to gain exposure to the oval window region. A sharp stapes curet is used to remove bony annulus overlying the oval window. Adequate exposure exists when the tip of the pyramidal process and the horizontal portion of the facial canal over the oval window are seen. The stapedial tendon is divided sharply and, if the footplate is thin, a small control opening is made before separating the incudostapedial joint with a small knife. The stapes superstructure is removed, and the footplate carefully extracted in piecemeal fashion with the use of fine stapes hooks and hoes. It is important not to traumatize the inner ear or remove perilymph from the vestibule. After the entire footplate has been meticulously and carefully removed and bleeding has been controlled, the oval window is sealed and a prosthesis is inserted between the incus and the seal.

A variety of prostheses can be used, some in conjunction with the oval window graft. The use of a sharp or beveled polyethylene prosthesis has resulted in a high incidence of postoperative fistula, and therefore its use has almost entirely been abandoned. Similarly, clinical and experimental observations have identified a higher incidence of postoperative fistula, vertigo, and temporary and permanent sensorineural hearing loss with the use of a preformed gelatin sponge-wire prosthesis. Connective tissue oval window grafts appear to give the best long-term hearing results clinically and cause the least amount of vestibular inflammation experimentally. The wire prosthesis, when used, must be crimped tightly around the incus; one can check gently for a round window reflex, which is sometimes seen by palpating the incus. The absence of the reflex is not important in all cases.

The anterior crurotomy technique can be used successfully if otosclerosis is confined to the anterior margin of the oval window. In this case the anterior crus is carefully dissected away and a crack is made across the midportion of the footplate. The anterior portion of the footplate is removed, and a small connective tissue plug is placed to seal the small oval window defect. This technique has the advantage of preserving the incudo-stapedial joint and the stapedial tendon which attaches to the neck of the stapes; this may be especially beneficial to those patients required to work in noisy environments.

In some patients a very thick footplate with an indistinct window annulus is found as a result of what is called obliterative otosclerosis. If this thick bone is mature and not too vascular, the footplate can be thinned with a fine microstapes drill and a small 1-mm opening made through the footplate to allow insertion of a piston prosthesis. The stainless steel piston or Teflon wire piston prosthesis can be used.

After stapedectomy, the tympanomeatal flap is replaced in its normal position and, in many patients, restoration of hearing is immediately apparent. In some, hearing will not be immediately improved because of serous labyrinthitis. This may continue for several weeks or rarely as long as a month following surgery.

One dressing technique consists of using a strip or two of surgical silk with superimposed cotton balls soaked in an antibiotic-steroid solution. Another method is merely to fill the external auditory canal with Gelfoam. The dressing is removed after 3 to 5 days, and a postoperative audiogram is obtained 1 month after surgery. There may be a mild amount of postoperative vestibular difficulty lasting 1 or 2 days.

Complicating Factors

The stapes surgeon should be aware of variations in anatomy that may occur in the course of the operation. Rarely, on turning the tympanomeatal flap, a high and dehiscent jugular bulb is encountered. Bleeding is usually overcome with pressure, while persistent oozing may require rescheduling. A persistent stapedial artery is rarely seen. It courses between the crura of the stapes, embryologically from the stump of the second aortic arch (stapedohyoid artery), and traverses the stapes through the obturator foramen. Injury to this vessel is met with profuse hemorrhage and its presence usually dictates termination of the procedure. The procedure should also be terminated if total obliteration of the round window by an otosclerotic focus is encountered. Attempts at stapedectomy in this rare situation are fraught with difficulty, complications, and a high incidence of poor results. In such cases, a small fenestra stapedectomy may be performed (see the following section).

Upon intraoperative palpation of the ossicular chain, the surgeon may discover a heretofore unrecognized fixed malleus. This may occur alone or in conjunction with stapedial footplate fixation, and may be congenital or secondary to trauma or infection. Mobilization of the malleus by forceful manipulation is frequently followed by refixation. Suggested treatment depends upon the degree and type of fixation. Occasionally, for minor epitympanic adhesions, lysis and mobilization will suffice while the thick anterior fixation will require (1) removal of the incus; (2) removal of the stapes, as would be done in a stapedectomy; (3) covering of the oval window with a tissue graft; (4) removal of the head of the malleus and severance of the anterior ligament of the malleus; and (5) placement of a malleus-to-oval window prosthesis or interposition of a total ossicular Teflon and Plasti-Pore prosthesis between the drumhead and the oval window. If a total ossicular replacement prosthesis is used, fascia, perichondrium, or preferably, a small block of cartilage should be placed between the prosthesis and the drumhead. Other forms of incudomallear fixation may occur that require specialized approaches.

An overhanging or dehiscent facial nerve may be of concern, as it frequently impedes access to the oval window and makes footplate removal and prosthesis placement difficult. If the operating surgeon believes that the footplate can be removed safely and a prosthesis placed without resulting in pressure on the facial nerve, this finding is not a contraindication to proceeding.

Occasionally, a profuse perilymph leak will be encountered upon making a control hole in the stapes footplate. An abnormally patent cochlear aqueduct is felt to be responsible for this happening, which occurs more commonly on the left than on the right and is more frequent in patients with congenitally fixed footplates than in those with otosclerosis. With the patient's head elevated, connective tissue seal of the oval window and wire prosthesis will often be successful in stopping the leak. In stubborn cases, a Teflon cannula may be inserted in the lumbar subarachnoid space, draining about 100 mL of fluid. This cannula should be maintained in place for 3 to 4 days to remove 100 to 150 mL of cerebrospinal fluid daily to reduce its volume and, therefore, decrease pressure.

The floating footplate is another problem that may be encountered in stapes surgery. This usually occurs in patients who have had previous stapes mobilization, those where fixation is minimal, or those in whom a biscuit type of dense otosclerosis footplate exists. A floating footplate can usually be avoided if a control hole is made in the footplate prior to removing the stapes suprastructure. If, in spite of preventative measures, this condition should occur, it frequently can be remedied by making a small (0.5 mm) notch in the promontory at the inferior margin of the oval window. Through this notch a small right angle pick is introduced to remove the free footplate. Should the footplate, either in part or in its entirety, become severely depressed into the vestibule, it should be left and the oval window sealed with a wire-tissue prosthesis. Many methods have been described to retrieve the submerged footplate fragment, most of which result in moderate to severe sensorineural hearing loss. Postoperative dysequilibrium is to be expected, and may persist for a considerable time.

Fracture of the long process of the incus may be found incidentally during stapedectomy or may be the result of overzealous curetting or crimping. In any event, if the prosthesis cannot be satisfactorily placed on the remaining long process, the incus should be removed and an incus replacement prosthesis used.

The sensation of vertigo during stapedectomy has a variety of causes, should alert the surgeon to impending complications, and is a major reason for performing stapedectomy under local anesthesia. It may be simply the result of a cold caloric effect on the inner ear but, more likely, suggests violation of the vestibule (especially the utricle) by mechanical or chemical (anesthetic) irritation.

Upon return of the tympanic membrane to its anatomic location, any small tears (which should be meticulously avoided) are bridged with medially placed connective tissue or Gelfilm and the edges approximated. A perforation that persists into the extended postoperative period may further require myringoplasty.

Postoperative Considerations

Care

Postoperatively the patient should be placed at bed rest with the head of the bed elevated to promote physiologic eustachian tube function. Preoperative antibiotics should be continued for 5 to 7 days postoperatively, or at least until the pack is removed, and elective systemic corticosteroids are avoided. Subsequent activity is restricted only insofar as limiting straining for a period of up to 12 weeks and instructing the patient not to fly and to keep water out of the operated ear for at least 2 weeks.

Results

In approximately 90 per cent of patients early closure of the air-bone gap by decreasing the air threshold to within 10 dB of the preoperative bone can be expected. This percentage decreases slightly with time. Ten per cent of patients will experience initial postoperative sensorineural hearing loss, but in only 3 per cent of cases should this sensorineural hearing loss persist. Recent studies have identified a slowly progressive high frequency sensorineural hearing loss in stapedectomy patients. Whether this observation is the result of cochlear otosclerosis or the long-term effects of the surgical procedure itself has not been determined.

Complications

Postoperative complications may appear immediately or may be delayed for several weeks or years. Mild vertigo during the immediate postoperative period is not uncommon, and usually represents a serous labyrinthitis. Vertigo may be associated with irritative nystagmus and nausea. These symptoms and findings should gradually improve over 24 to 48 hours. Should they persist for several days the anti-inflammatory effect of corticosteroids may be helpful. Permanent vestibular alteration as measured by electronystagmography has been reported. Acute otitis is unlikely during the immediate postoperative course when prophylactic antibiotics are employed. If encountered, however, this process is potentially dangerous and should be treated aggressively with appropriate antibiotics. Labyrinthitis and meningitis are dread complications which may occur in the early or late postoperative period. The oval window, with its weak or incomplete neopartition, serves as a potential path for infectious diseases of the middle ear to extend to the labyrinth and, subsequently, the meninges. The sharp, beveled polyethylene prosthesis placed on compressed absorbable gelatin sponge appears to predispose to this complication, as does a preoperative history of eustachian tube dysfunction and recurrent acute otitis media. Patients should be instructed to report to their physician for urgent treatment if symptoms of acute otitis media occur. Treatment then consists of administration of parenteral antibiotics.

Reparative granuloma occurs in about 1.5 per cent of stapedectomy patients and usually becomes manifest 1 to 6 weeks postoperatively with the sudden or gradual onset of sensorineural hearing loss, vertigo, tinnitus, and sound distortion. Pain and fever are noticeably absent. Otoscopic examination reveals a red-grey discoloration in the posterior superior quadrant of the

tympanic membrane. Audiologic evaluation indicates a decrease in discrimination scores which are out of proportion to that anticipated by the mixed hearing loss alone. Etiology is uncertain although infection, foreign body, hypersensitivity reaction, or simply an exaggerated response to the trauma or surgery have all been implicated.

The outcome of reparative granuloma management depends upon early diagnosis and surgical intervention. Exploratory tympanotomy discloses are red-gray, fleshy, vascular mass about the prosthesis. Histologic examination reveals acute and chronic inflammatory cells, neovascularization, fibroblastic proliferation, and foreign body giant cell formation. Occasionally foreign bodies (starch granules and bone dust) may be identified. The mass should be removed in its entirety and an oval window seal, different from that used in the original surgery, placed.

Prompt and proper therapy will result in satisfactory hearing gain in most patients, although some will continue to have high frequency sensorineural hearing loss and decreased discrimination.

Oval window fistula, although an infrequent postoperative complication, may occur subsequent to oval window surgery. The usual presentation includes fluctuating or progressive sensorineural hearing loss and tinnitus. Vertigo may also occur. Occasionally an inner ear conductive hearing loss may be the result of an oval window fistula theoretically due to loss of impedance match associated with perilymph spillage. The diagnosis is made primarily on history and audiologic evaluation. The hearing loss, as with reparative granuloma, is usually associated with a disproportionate decrease in discrimination. Fistula testing, electronystagmography, and myringotomy may prove of little diagnostic help. Oval window fistula is related to the length, type, and position of the prosthesis. Of all oval window grafts gelatin foam appears most susceptible to fistula formation. The cochlear aqueduct, although closed in the large majority of patients, may be open, allowing a potentially large pressure gradient to be transmitted from the cerebrospinal fluid to the oval window area. This, coupled with the negative pressure that may occur in the middle ear, may also play a role in the formation of oval window fistula. If the diagnosis is suspect, early exploration is indicated. Occasionally the fistula may be repaired by local débridement and connective tissue interposition, but many cases will require removal of the prosthesis and regrafting. Approximately two thirds of patients will receive significant benefit from surgical intervention, whereas the remainder will continue to suffer some hearing loss and/or dysequilibrium.

Other causes of delayed hearing loss, such as allergy or autoimmune reactions within the inner ear, are also possible. The exact nature of delayed cochlear losses following stapedectomy is not thoroughly understood. For example, experimental stapedectomy in animals or prospective oval window grafting experiments in cats have resulted in a high incidence of lymphoid follicles in the inner ear, characteristic of autoimmune disease.

When long-term follow-up is carried out, it has been found that persistent or progressive conductive hearing loss greater than 10 dB may occur in up to 25 per cent of patients who have undergone stapedectomy. This conductive hearing loss may be secondary to serous otitis media,

loosening of the prosthesis, resorption of the long process of the incus, adhesions from the promontory to the prosthesis or incus, and an eccentrically placed prosthesis. Most of these mechanical malfunctions, if causing significant conductive hearing loss, can be corrected surgically at re-exploration. It must be recognized, however, that reopening the vestibule, if necessary, is associated with a greater risk of sensorineural hearing loss than that reported for the original operation.

Stapedotomy (Small Fenestra Stapedectomy)

Stapedotomy, or the surgical development of a small fenestra placed centrally in the stapedial footplate, is an alternative to total footplate removal (Bailey et al, 1983). This approach to the surgical correction of the conductive hearing loss that accompanies stapes footplate fixation was initially employed for those cases in which the stapes footplate was excessively thickened and the stapedial annulus obliterated. Smythe and Hazard (1978), however, reported successful long-term surgical results using this technique in patients without obliterative otosclerosis. The results were not only comparable in the low-frequencies with those achieved by total footplate removal, but they were also superior in frequencies of 2000 Hz and above. The surgical approach for this operation is identical to that described for stapedectomy except that, instead of removing the total stapes footplate, a small (approximately 0.8 mm in diameter) fenestra is centrally placed in the footplate by using picks, a hand drill, or an argon laser. A piston prosthesis is then inserted through the fenestra to a depth of about 0.25 mm in the vestibule. This prosthesis is then crimped to the long process of the incus. A tissue seal is made around the piston with fascia or perichondrium to help avoid postoperative fistula.

Stapedotomy may be performed by using a laser. The CO_2 laser provides precise surgical control without causing bleeding and, when used properly, produces insignificant thermal changes within the vestibule. Long-term effects on hearing ability with the use of laser stapedotomy are at least equal to those following mechanical stapedotomy, and there are few complications.

A Flexible Approach to Otosclerosis Surgery

Surgical approaches in treating otosclerosis need not be stereotyped. It is possible to use a variety of techniques, depending on the pathologic conditions and anatomic characteristics encountered during stapes surgery. The major objective is to improve hearing significantly, with or without a hearing aid, and to avoid complications. As discussed in Chapter 18 of this volume, in most cases it is safe and cost-effective for patients to have same-day surgical procedures, avoiding an overnight hospital stay.

The multiple approach as used by one author (M.M.P.) is described as follows.

Mobilization of Stapes. Occasionally, stapes mobilization is still a good procedure. If the pathologic conditions are excessive, as in obliterative otosclerosis or extensive otospongiosis in a young adult, and the risk of cochlear damage is of greater concern, in some cases stapes mobilization can result in improved and sustained hearing. Stapes mobilization may be enhanced

by fracturing or removing part of the footplate in certain rare cases. Hearing improvement postoperatively may be sustained or, if recurrent fixation and conductive hearing loss occurs, stapedectomy can be considered at a later date with better planning and perhaps under safer conditions. For example, if the patient has spongiotic bone, one would use sodium fluoride and calcium gluconate in an attempt to convert spongiotic to sclerotic bone to enhance the safety of a possible subsequent stapedectomy procedure.

Anterior Crurotomy. If the pathologic condition is isolated only to the fistula ante fenestram and involves only the margin of the anterior oval window, an anterior crurotomy can be used quite successfully. The method is to score the footplate across the middle and to remove the anterior half of the footplate along with the anterior crus, leaving the incudostapedial joint intact along with its attached stapedial tendon. If the oval window and the posterior crus are otherwise normal, this patient will achieve a long-lasting, good result. This procedure is less traumatic, when done carefully, insofar as possible labyrinthine damage is concerned, and has the beneficial adjunct of preserving the incudostapedial joint and the stapedial tendon.

Stapedotomy. A piston is used whenever the oval window is narrowed or small due to anatomic or pathologic configurations. This might be a bulging facial nerve, either dehiscent or in its fallopian canal, or a bulge from the promontory. The oval window varies in size and may be small in a given patient or may be small because of dense otosclerosis, which will allow removal of a central, thinner portion of the footplate. In such an instance, a piston is utilized, usually, a 4.25-mm piston. A measurement is taken, and if needed a 4.5-mm piston also can be used. Tissue grafts are used to seal; however, a very satisfactory graft consists of using available blood clot mixed with Gelfoam, which provides an acceptable seal and yet does not encourage collagen or bony fixation in the region of the piston postoperatively.

Obliterative Otosclerosis. For cases with obliterative otosclerosis, the entire thick footplate is carefully drilled, the oval window is opened, and a piston is used as above.

Complete Stapedectomy and Wire/Connective Tissue Prosthesis. In patients who have complete stapedectomy, a very satisfactory method is to custom-make a prosthesis of wire/connective tissue or wire/fat. The usual length from the top to the bottom of the prosthesis is 4.5 mm. This method requires that the surgeon has practiced the making of this prosthesis in the temporal bone laboratory. Stainless steel wire (0.005 inch) is used.

Stapedectomy/Sacculotomy for Patients with Otosclerosis and Ménière's Disease. We have successfully used this procedure in selected cases with patients with symptoms of vertigo, Ménière's disease, and otosclerosis. To date, we have had no complications in this subset of patients and the majority have had a very satisfactory result in terms of improvement of hearing and treatment of vertigo. If vertigo persists, endolymphatic sac enhancement can be considered, subsequently (Paparella et al, 1984).

Missing Incus. We have treated patients who have had a previous fenestration procedure or, for some reason or another, have an absent incus. We have also had cases in which patients

have otosclerosis - that is, fixation of the stapes - along with other diseases such as chronic otitis media. Once the infection is totally under control after tympanoplasty and/or mastoidectomy, after a year a stapedectomy can safely be done. This may include a malleus-to-oval window prosthesis; if there is no infection, this can result in a very satisfactory hearing result, if cochlear reserve and a mesotympanic space are reasonably intact.

Medical Therapy

Siebenmann in 1889 suggested a correlation between otosclerosis and sensorineural hearing loss; there has been much subsequent speculation as to the etiology and incidence of this hearing loss. Today there is little doubt that otosclerosis can cause sensorineural hearing loss but equally little agreement as to the pathogenesis of the mechanism. Some authors postulate histologic otosclerosis to be a fairly common cause of sensorineural hearing loss in adults, while others believe that the existence of pure sensorineural hearing loss secondary to an otosclerotic lesion has been exaggerated. Young adults or adults presenting with a fairly flat audiometric configuration may have genetically induced delayed sensorineural hearing loss mediated by atrophy of the stria vascularis.

Politzer in 1894 described the actual invasion of the scala tympani by an otosclerotic process. Subsequent histologic findings described include cochlear capsule invasion, interruption of the cochlear endosteum, and mechanical disturbance of the anchorage mechanism for the basilar membrane. The spiral ligament is the most common membranous structure altered in cochlear otosclerosis with atrophy, fibrosis, thickening, and collagenization. These changes, coupled with changes in the stria vascularis, are thought by some to be secondary to vascular shunting between the bony and membranous cochlea, resulting in increased pressure and congestion in the labyrinthine veins and subsequent build-up of toxic metabolites within the corresponding membranous labyrinth.

Those who believe in its existence make the diagnosis of pure cochlear otosclerosis in patients who have insidious, progressive, symmetric, sensorineural hearing loss with no apparent underlying etiology. Frequently there is a family history of otosclerosis and physical examination may or may not reveal Schwartze sign. Diagnostic confirmation relies upon tomographic study, including study by computed tomography, which may reveal radiolucent patchy areas about the cochlear capsule in early otosclerosis, radiolucent areas with adjacent areas of increased density as the process matures, and diffuse sclerosis in late otosclerosis. Caution is advised in the detailed interpretation of studies of the labyrinthine capsule with polytomography, since vagaries of dense and less dense bone occur in normal patients.

Medical treatment is reserved for patients with otosclerosis who have pure sensorineural hearing loss or who, because of age, degree of hearing loss, occupation or social life, lack of discrimination, resistance to surgery, or other reasons are not good surgical candidates. Medical therapy may also be used in conjunction with surgery in those patients with mixed hearing loss. We have employed the use of fluoride therapy most commonly with patients who have achieved a good air-bone gap closure from stapedectomy who, over time, developed a distinctively

sensorineural hearing loss. Some authors recommend prophylactic treatment when a strong family history exists or a Schwartze sign is present in the absence of hearing loss.

Escot (1923) was the first to propose the use of small doses of fluoride (calcium fluoride) for the treatment of progressive deafness associated with otosclerosis. Based on the favorable influence large doses of fluoride have on the osseous lesions of Paget's disease, the histologic similarities between early otosclerosis and Paget's disease, and the histologic similarities between mature otosclerosis and fluoride-induced sclerosis in bone, Escot, and later Shambaugh (1965), anticipated arrest of the otosclerotic process with fluoride therapy.

Fluorine, when ingested as sodium fluoride, is absorbed up to 97 per cent from the gastrointestinal tract; most of it is excreted through the kidneys. Wherever fluorine comes in contact with bone it forms a chemical union with the fluoride ion, replacing the hydroxyl ion in bone crystals. This new fluorapatite complex is much more stable than the hydroxyapatite crystal found in the normal cell. Sodium fluoride also results in a positive calcium balance by accelerating calcification of young bone and retarding bone resorption.

Side effects of sodium fluoride treatment are minor and infrequent. Gastric disturbance is most prevalent and manifests as nausea and vomiting. Decreasing the dosage and prescribing enteric-coated tablets usually alleviates this problem. Occasional patients will complain of joint pain and swelling, muscle pain, and bone pain. These symptoms are reversible but require termination of therapy. Fluid retention, ocular discomfort, and cutaneous eruption are unusual side effects.

The current recommended therapy is in the form of oral sodium fluoride, 30 to 120 mg daily. This may be combined with 400 units of oral vitamin D and 10 mg of oral calcium carbonate daily, since recent evidence suggests an association between otosclerosis and deficiencies in vitamin D and calcium (Brooks, 1985). Evaluation of therapeutic efficacy is based on disappearance when present, of a Schwartze sign, audiometric improvement, and tomographic signs of maturation. Approximately 50 per cent of patients treated will remain unchanged, whereas one-third will improve and the remaining one-sixth will continue to worsen.

Amplification

Amplification is of benefit in patients who have not achieved serviceable hearing from available therapeutic modalities or for those who, for one reason or another, are not considered good candidates for surgical intervention.