

Chapter 170: Otosclerosis

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Diagnosis and Nonsurgical Management

Otosclerosis is a bone disease unique to the otic capsule. One or more foci of histopathologically distinct, abnormal bone in the labyrinth may be asymptomatic or may cause progressive conductive hearing loss, mixed hearing loss, or, occasionally, sensorineural hearing loss. Otosclerosis is an important cause of auditory and, to a lesser extent, vestibular symptoms, and both can be treated effectively in most cases.

Epidemiological considerations

The prevalence and incidence of otosclerosis varies, depending on whether clinical criteria or autopsy findings and other descriptive features are used to determine its presence. Race also is a factor. In Caucasians, the disease is found in 7.3% of male and 10.3% of female temporal bones. The stapes appears ankylosed in 12.3% of those with histologic otosclerosis. This means that for every person with a hearing loss caused by otosclerosis, approximately nine have asymptomatic histologic disease. In Blacks, otosclerosis is found in only 1% of temporal bones. The disease is rare to non-existent in American Indians (Altmann et al, 1967; Friedmann, 1974; Guild, 1944; Levin et al, 1988; Nager, 1969; Schuknecht, 1966; Tato, 1967). Joseph and Frazer (1964) found that in Hawaii, otosclerosis was only about half as common in Japanese as in Caucasians.

A study of clinical otosclerosis among Chinese patients in Taiwan (Huang and Lee, 1988) showed that about 1.1% of those seen with hearing loss had the disease. This figure was believed to closely approximate that found among the Japanese and to be much lower than the figures found in comparable studies in Caucasians.

The female to male ratio of patients with otosclerosis seen in clinical practice is about 2:1 (Ginsberg et al, 1978; Larsson, 1960; Robinson, 1983).

Hearing loss is bilateral in more than 90% of patients seeking therapy (Cawthorne, 1955; Ginsberg et al, 1978). This percentage may be falsely inflated because patients with unilateral loss may never seek medical attention. Robinson (1983) has reported laterality data in 551 patients in whom onset of otosclerosis occurred before the age of 18 years. Those who had surgery before 18 years (N = 31) had unilateral disease in about 30% of the cases and in 9 of these 11 cases the patients developed clinical otosclerosis in the second ear after an average of 5.8 years. Only 8% of the 520 patients who underwent surgery in adulthood had unilateral disease at the time of their surgery.

Age also influences the incidence of otosclerosis. Histologically, otosclerosis is rarely seen in patients under 5 years and is most often seen between ages 30 and 49. Clinically, hearing loss usually becomes apparent between adolescence and early middle age (Beales, 1987). Patients usually delay seeking assistance for an average of 2 to 3 years (Robinson, 1983).

Table 170-1 summarizes the prevalence and incidence of clinical otosclerosis cited in various studies throughout the world.

Genetics

Toynbee first recognized the hereditary nature of otosclerosis in 1861; however, only since 1960 have studies determined the genetic mechanism involved. Previous studies were hampered by inadequate numbers of patients, insufficient investigation of family trees, and difficulties in diagnosing otosclerosis in family members. Recent studies of family members have included searches for family trees and thorough evaluation of family members (Causse and Causse, 1984; Larsson, 1960; Morrison and Bunday, 1970).

Approximately 70% of otosclerosis cases have a hereditary basis. This number is higher than the 49% to 58% incidence of positive family history reported in many series (Schuknecht, 1974). Patients apparently may forget about or not be aware of deafness in their own families. In hereditary cases, inheritance is autosomal dominant, with a penetrance of 25% to 40%. No association has been found with chromosome markers or with ABO or HLA (human lymphocyte) antigens. The remaining 30% of cases occur sporadically. The mechanism is speculative, although autosomal recessive inheritance, spontaneous mutation, and nongenetic causes are possible.

Reports in the literature often suggest that the rate of progression and the severity of disease is relatively consistent within a given family (Morrison and Bunday, 1970).

Otosclerosis has been suggested to be a localized form of osteogenesis imperfecta; however, the histologic and gross appearance of otosclerosis is distinct from that of osteogenesis imperfecta. Both lesions can be identified in about 6% of patients with osteogenesis imperfecta (Armstrong, 1984; Garretsen and Cremers, 1991; Hall and Ogilvie, 1961). Extrinsic and intrinsic mechanical stresses, vasomotor imbalance, ground substance abnormalities, and infection have been proposed (Friedmann, 1974).

Definitive studies on the genetics of otosclerosis would require histologic examination of many temporal bones in affected families, and this probably will never be done. For genetic counseling purposes, if both parents are otosclerotic, half the children will have the disease; if one parent has it, then one fourth will (Morrison and Bunday, 1970).

Histopathology

The histologic appearance of otosclerosis in the temporal bone closely resembles that of new bone formation anywhere in the body. Genetic predisposition and the occurrence in endochondral bone make otosclerosis unique.

Otosclerosis begins as discrete foci of abnormal bone that enlarge and may coalesce. The otosclerotic foci demonstrate two phases: an early spongiotic phase and a later, sclerotic phase. The early phase results in highly vascular, less dense, loose spongy bone that replaces the normal bone. At first, the perivascular spaces within the focus enlarge. Bone is resorbed around the vessels and is replaced by cellular, fibrous connective tissue containing numerous osteoclast giant cells and many vascular channels. Within the perivascular connective tissue,

reticular cells and fibroblasts become osteoblasts, which lay down immature bone with a woven, disordered pattern of collagen fibers. This new bone contains much ground substance and few collagen fibers. It stains blue with hematoxylin and eosin, thus forming the "blue mantles" of Manasse. The spongy pattern results from areas of new bone surrounding large areas of highly vascular connective tissue.

During the late, sclerotic phase, the new bone is resorbed and replaced with osseous tissue containing many collagen fibers and little ground substance. The osseous tissue stains red with hematoxylin and eosin. Lamellar bone is laid down to form trabeculae along the margins of the mature osseous tissue. The large areas of vascular, cellular connective tissue are replaced with new bone, and the lesion becomes quiescent.

Within a single focus, small areas of spongiosis and sclerosis are scattered in an irregular, mosaic pattern. A focus may be termed *active* or *inactive*, depending on the extent of areas of resorption versus areas of bone formation. Active lesions have a spongy structure and contain immature osseous tissue. They are soft and crumbly, bleed easily, and contain a highly cellular, fibrous connective tissue with many osteoclasts and dilated vascular channels. An inactive, or sclerotic, lesion contains solid, lamellar, osseous tissue that feels hard on palpation and is poorly vascularized. Fibrous tissue is limited and infrequently has small vessels. Generally, if multiple foci occur within a temporal bone, all have a similar level of activity. In bilateral disease, however, the opposite ear often has a different level of activity (Friedmann, 1974; Nager, 1969).

Location

Foci of otosclerosis may be found anywhere in the temporal bone. Distribution of the lesions is characteristic. Of affected bones, 80% have foci anterior to the oval window; these foci spread along the inferior rim more readily than along the superior rim. The round window is involved in 31% of affected temporal bones. Other sites include the stapes footplate in 12% and the anteroinferior cochlea in 14%. Otosclerosis is rarely seen outside the otic capsule. The fissula ante fenestram has been proposed as the starting focus for otosclerosis because of possible rests of embryonic tissue along its course. However, otosclerotic foci often occur away from the oval window, and several temporal bones have been described with foci near but distinctly separate from the fissula (Guild, 1944).

Stapes fixation

Oval window involvement may ankylose the stapes. The focus of otosclerosis first involves cartilage at the oval window's margin and causes thickening of the annular ligament. Otosclerosis then appears on the footplate. The annular ligament between the two foci then ossifies and fixes the footplate. Some lesions, however, may grow around the oval window and limit stapes motion but never cause ankylosis. Why some large lesions stop short of ankylosis and some small lesions grow onto the stapes is not known (Nager, 1969).

Blood supply

Otosclerosis alters the vascular pattern within the temporal bone. Within the otospongiotic focus, existing vessels increase in size as osteoclasts resorb the surrounding bone. Many new vascular channels develop in the perivascular connective tissue. As the focus grows, the nearby mucoperiosteum reacts with dilatation of blood vessels, giving the mucosa a reddish pink hue. This finding is a correlate of the active or otospongiotic phase, which some otologists consider a contraindication for surgery and an indication for sodium fluoride treatment.

Otosclerotic bone may form new connections between the usually separate blood supply of the bony otic capsule and the membranous labyrinth. These new channels form shunts that can dramatically increase blood flow and dilate labyrinthine vessels; such shunts occur in 47% of involved bones. Abnormal blood flow may contribute to sensorineural hearing loss (Ruedi, 1969).

Clinical presentation

History

Hearing loss is the major symptom in otosclerosis. The loss is usually gradually progressive (Larsson, 1960). The age of onset in Cawthorne's patients is shown in Table 170-2. The initial conductive loss usually progresses over months to years, frequently with periods of stabilization. This pattern is consistent with the histologic findings. Otosclerosis may be present for years before stapes fixation begins. Fixation may progress to become complete, resulting in a maximum (50 to 60 dB0 air-bone gap (Cawthorne, 1955; Glasscock and Shambaugh, 1990; Schuknecht, 1974).

Although it has been suggested that pregnancy may hasten the hearing loss, objective data are difficult to obtain (Glasscock and Shambaugh, 1990; Larsson, 1960; Schuknecht, 1974). Cawthorne (1955) found that 63% of his white female otosclerotic patients who had children noted that their hearing had deteriorated during the later months of one or more of their pregnancies. Gristwood and Venables (1983) found that subjective deterioration of hearing in pregnancy ranged between about 33% after one pregnancy and about 63% after six pregnancies.

Paracusis of Willis is found in 20% to 78% of patients. The patient experiences better understanding of speech in a noisy environment. This phenomenon is characteristic of all patients with conductive hearing loss; it occurs because people speak louder in noisy surroundings (Cawthorne, 1955; Larsson, 1960). Tinnitus occurs in the majority of patients with otosclerosis, with or without accompanying sensorineural hearing loss. The tinnitus may be unilateral in bilateral disease. This tinnitus is usually described as a "ringing", and it is not usually classified as bothersome.

Vestibular complaints are more common in otosclerotic persons than in the general population, and they occur more often than most believe. Cody and Baker (1978) carefully studied 500 patients with otosclerosis treated nonsurgically. The most common vestibular symptoms were recurrent attacks of vertigo (26%) and postural imbalance (22%). Virolainen

(1972) questioned 60 nonsurgically treated patients who subsequently had otosclerosis confirmed by surgical exploration. Thirty-four (56.7%) patients attested to having brief periods of vertigo, usually not more than 60 seconds and commonly induced by rapid head movement. Many also experienced unsteadiness when walking in the dark or with their eyes closed. Other observers have not noted as many vestibular complaints; for instance, Bretlau et al (1985) noted vertigo in only 17% of 95 patients. Ginsberg et al (1978) found about 18% of 2405 patients indicated the presence of vertigo prior to surgery, and in most cases it was self-limiting.

The clinical presentation of dizziness in otosclerotic patients falls into three basic patterns: (1) Ménière's disease, (2) otosclerotic inner ear syndrome (OIES), and (3) postural imbalance and vertigo.

Ménière's disease is uncommon in otosclerotic patients, occurring in approximately 4%. The clinical diagnosis is made using the usual criteria for Ménière's disease (see Chapter 181) (McCabe, 1966). Recruitment is often present, as are electronystagmographic (ENG) abnormalities. The clinical picture resembles the added symptoms of Ménière's disease and otosclerosis.

OIES (3% of McCabe's series) occurs even less frequently than Ménière's disease. Characteristic clinical features described by McCabe (1966) include (1) episodic vertigo lasting 20 minutes to 76 hours, which may be a vague floating or semifainting sensation; (2) absence of nystagmus during a spell; (3) normal caloric response, which often reproduces a spell's symptoms; (4) no positional nystagmus; and (5) normal neurological examination. Audiologic features include (1) conductive, nonfluctuating hearing loss, which is frequently unilateral; (2) normal discrimination; and (3) absence of recruitment.

Causes of dizziness and vertigo in otosclerotic patients are not completely understood. Two temporal bones have demonstrated endolymphatic hydrops occurring concomitantly with multiple foci of otosclerosis (Black et al, 1969).

Physical examination

The tympanic membrane is usually normal or has mild abnormalities from other disease processes, with no apparent cause for a conductive loss. Pneumatic otoscopy should be used to rule out an effusion and to demonstrate malleus mobility. The long process of the malleus should be observed with insufflation on all ears to gain a visual appreciation of normal mobility. With malleus fixation, the long process of the malleus is immobile.

Schwartz's sign is hyperemia of the promontory mucosa from increased vascularity, which is visible through the tympanic membrane. Some surgeons prefer to delay surgery until this finding resolves. The patient's ability to turn his or her head should be noted. Patients with significant limitations from arthritis or those with a short, thick neck may have difficulty being positioned on the operating table, thus hampering surgical exposure. Such patients may require general anesthesia for their own and the surgeon's comfort. The size of the external auditory canal determines maneuverability of instruments. Rarely, if the canal is unduly small, a separate canalplasty or a short Lempert No 2 endaural incision may be helpful.

Preoperative testing

Audiometry

A progressive, predominantly conductive hearing loss is the major audiometric sign in otosclerosis. The audiometric tests vary according to the degree of the stapedial ankylosis and the degree of sensorineural (cochlear) involvement. A pure conductive loss is apparent in the early stages and typically is first apparent in the low frequencies (showing an ascending audiometric curve for air-conduction). As the loss progresses, the conductive loss becomes more evident in the higher frequencies and the air-conduction curve usually becomes relatively flatter. Also as the loss progresses, a mild to moderate sensorineural loss may become evident in the bone conduction curve. The degree of this sensorineural component is greater than that expected as the result of aging. The maximum conductive component of the loss seldom exceeds 50 dB to 60 dB. Fig. 170-1 shows examples of various stages of the loss.

In advanced, long-standing disease the sensorineural component may be significant and may dominate the conductive component (Fig. 170-2).

Patients with bilateral loss usually show significantly greater loss in one year in the initial stage of the disease. This asymmetry can predominate for many years, but typically the better ear eventually reaches the degree of hearing loss shown for the originally poorer ear.

Standard audiometric tests are the most critical diagnostic methods used to evaluate the patient. The basic evaluations should include measurements of pure-tone thresholds for air-conduction (250 Hz to 8000 Hz), bone conduction (250 Hz to 4000 Hz), the speech reception threshold, and a measurement of the maximum speech recognition (discrimination) obtained at suprathreshold levels (see Chapter 148).

The air-conduction thresholds indicate the degree of loss and determine if the loss is great enough to warrant surgery. The bone conduction threshold levels indicate the sensorineural function and the optimum hearing levels (improvement) that can be achieved by surgery.

Speech recognition is normal in pure conductive loss in otosclerosis. When a significant sensorineural component exists or when the loss is mixed but primarily sensorineural, speech recognition scores are usually better than expected with comparable losses from other causes.

In 1948, Juers in Chicago and Woods in Dublin coincidentally published papers remarking on the improvement of bone-conduction following successful fenestration surgery. They hypothesized that otosclerosis at the footplate of the stapes somehow falsely decreased audiometric bone conduction responses so as to make the sensorineural levels appear to have greater losses than were truly present.

Carhart and his doctoral students at Northwestern University studied these changes and developed correction factors for bone conduction to estimate the true sensorineural "reserve" (Carhart, 1950; Carhart, 1962; McConnell and Carhart, 1952). Their studies indicated that the preoperative bone conduction curve revealed a distinctive notch with maximum reduction at

2000 Hz. The bone-conduction thresholds after surgery showed average improvement of 0 dB at 250 Hz, 5 dB at 500 Hz, 10 dB at 1000 Hz, 15 dB at 2000 Hz, and 10 dB at 4000 Hz. This shift has since been shown as the "Carhart notch". The shift is felt to be of a mechanical nature caused by impedance changes in the middle ear transformer resulting from stiffness, friction, and mass effects of the otosclerotic lesion.

Subsequent studies of the bone-conduction levels in the stapes mobilization, the stapedectomy and the stapedotomy procedures also have found improvement levels postoperatively. However, the average changes have not been as great as that shown in the "Carhart notch" following fenestration and often have shown greater improvement at frequencies other than 2000 Hz (Boyer and Kos, 1961; Cole, 1966; Gatehouse and Browning, 1982; Gibb and Mal, 1973; Ginsberg et al, 1978; Guilford and Haug, 1958; Harell and Goldwag, 1976; Miller et al, 1961; Odess and Roach, 1962; Rosen et al, 1959; Sooy et al, 1966a, b).

The question arises as to whether or not a surgeon should consider the "Carhart notch" in selecting cases for surgery. It is a particularly critical question in cases such as that shown in Fig. 170-3. If a correction is chosen to account for the notch, the optimum postoperative expectation is 20 dB hearing level for the speech frequency pure tone average (PTA). This is a 25 dB improvement from the surgery. If a correction factor is not appropriate, the PTA following surgery is a 15 dB average improvement. A correction factor would bring the patient within normal limits, but if the correction factor is not appropriate, the patient remains with a mild but possibly significant loss.

Studies of postoperative improvement for stapedectomy and stapedotomy often cite a few cases of improvement beyond the Carhart notch. The critical question of whether to use the correction should not be concerned with these few cases but should be answered by looking at the postoperative distribution of bone-conduction responses for all cases. A correction would be appropriate if most cases had an average PTA overclosure of 10 dB. It would not be appropriate if a relative few maximum overclosure cases raises the average to the Carhart notch figures, while most cases do not achieve such overclosure. Shambaugh and Adamson (1964) found 20% to 25% of stapedectomies exceeded the Carhart correction. Moncur and Goodhill (1963) and Sooy et al (1966a) reported that the majority of their cases did not have overclosure reaching 10 dB PTA suggested by the Carhart notch.

The data suggest that the Carhart notch is a valuable finding that suggests a diagnosis of otosclerosis in a patient with conductive loss and a compatible history. The data further suggest that the Carhart effect (overclosure) usually should *not* be considered in estimating optimum postoperative levels because of individual variation of responses (especially in currently used surgical procedures for otosclerosis).

Standard audiometry, especially in cases of bilateral loss, requires a skilled audiometrist and equipment that can evaluate profound losses with high-intensity maximum limits for air-conduction, bone-conduction, and speech audiometry. Masking should be done with narrow-band masking for pure tone tests and speech (pink) noise for speech testing. Overmasking may be a problem in some cases but intense levels of masking should be tried, if necessary, because of broad variability in individual overmasking possibility. Masking for speech reception thresholds and word recognition is required if the obtained threshold is about

40 dB or greater relative to the bone-conduction levels of the non-test ear and if the presentation of the words for speech recognition reach this same level.

The possibility of tactile stimulation must be evaluated in bone-conduction testing. The bone conduction stimulus can be felt at 25 dB or greater for 250 Hz, at 35 dB or greater at 500 Hz, and at 75 dB or greater at 1000 Hz. The maximum limits of bone-conductors at higher frequencies are not known to provide tactile stimulation. Individuals' ability to feel rather than hear such signals varies greatly, and the levels cited above are lower than average ability. When measurements are difficult to interpret in such cases, the questions can often be resolved by simply asking the patient whether the signal was "heard" or "felt".

Anderson and Barr (1967) have reported a series of congenital loss cases with "pseudo-mixed" progressive deafness that may be confused with footplate fixation and otosclerosis. The confusion concerns the falsely enhanced conductive involvement reflected in audiometry for 250 to 1000 Hz in these cases. Exploratory surgery indicated normal mobility of the ossicular chain, yet the audiometry indicated average air-bone gaps of 35 dB for 250 Hz, 35 dB for 500 Hz, and 20 dB for 1000 Hz. The false enhancement for bone conduction was thought to be caused by malformations (enlarged ossicles). The false bone-conduction responses were at levels too weak to cause tactile stimulation. Acoustic reflexes were present. The authors supported their causal hypothesis with previous studies showing similar results when ossicles were experimentally weighted in awake human subjects.

Tuning fork tests are mandatory to establish that the hearing loss has conductive component and to confirm the audiometric findings. A discrepancy should prompt reevaluation for a collapsing canal, a functional loss, or a primarily sensorineural loss that was not recognized by audiometry. It should be noted that an audiometric Weber test may be superior to a tuning fork Weber test because the maximum intensity from a bone oscillator is far more intense than that from a tuning fork (especially for high frequencies).

Acoustic immittance testing

Acoustic immittance testing includes tympanometry and acoustic reflex measurements (see Chapter 148). Tympanometry is not a good measurement to detect stapes fixation. The peak of the tympanogram usually is evident at a normal pressure level. Although the height of the peak for otosclerotics has often been described as abnormally low, actually it is often borderline normal or within the normal range (Browning et al, 1985; Ivey, 1985; Jacobson and Mahoney, 1977). A high peak of compliance is not compatible with a diagnosis of otosclerosis.

Two distinct patterns of abnormal acoustic reflexes are seen in otosclerosis. If the stapes is firmly fixed, no reflexes can be elicited from the affected ear. If footplate mobility has decreased but is not fixed, three abnormalities may be found separately or in combination. A *diphasic response* is seen at the onset and again at the end of the stimulus, the magnitude of the reflex may be reduced, and the polarity of the reflex may be reversed from normal findings.

The diphasic response, so-called on-off effect, is virtually pathognomonic for otosclerosis but usually is seen only with early disease. If symptoms are present less than 5 years, nearly all patients will show the on-off effect; if present more than 10 years, there is usually no reflex. In patients with unilateral disease, half the normal hearing ears will show the on-off effect. The mechanism producing the abnormal reflex is not well understood (Bel et al, 1976; Forquer and Shehy, 1981b; Mangham et al, 1983; Rane et al, 1978; Terkildsen et al, 1973). Fig. 170-4 illustrates these reflex findings.

Some clinicians feel it is useful to run stapedial reflex tests on patients with a small or even with no air-bone gap. Patients with a small or no air-bone gap, sensorineural loss with good discrimination, and a positive family history may be suspected of having otosclerosis and should be evaluated with acoustic reflexes. A positive on-off effect is found in 30% of such patients and, when present, supports the diagnosis (Causse and Causse, 1984; Forquer and Sheehy, 1981a).

Electronystagmography

Vestibular symptoms appear to be more common in non-surgically treated patients with otosclerosis than in the general population, but vestibular testing also often demonstrates ENG abnormalities. Cody and Baker (1978) tested 83% of 230 such patients who had vestibular symptoms in addition to hearing loss. Almost 60% had an abnormal thermal caloric test, usually "unilateral depression in labyrinthine function, occasional bilateral depression in function, and rarely unilateral absence in function".

Virolainen (1972) compared ENG tracings during bithermal caloric testing, and acceleration and deceleration thresholds from the rotating chair test in 60 nonsurgically treated otosclerotic patients and 20 controls. In order of frequency, the vestibular abnormalities in the otosclerotic patients were caloric hyperexcitability with a heightened threshold of angular acceleration and deceleration, directional preponderance, and positional nystagmus. Caloric hypoexcitability was evident in 42% to 57% of the patients (depending on which test parameter was measured), directional preponderance in 37% to 53%, and positional nystagmus in 33%. Differences from the control series were all statistically significant. The abnormalities appeared more often with greater sensorineural hearing loss but were not necessarily seen in the ear with the poorer hearing.

Although not necessary in all patients, preoperative ENG is desirable in those with vestibular symptoms and in those who have undergone previous stapes surgery particularly if they had significant postoperative vertigo. In a patient with bilateral otosclerosis and vestibular symptoms but without previous surgery, the ear with abnormal vestibular function should probably be chosen (if surgery is recommended and the hearing is similar on the two sides), since if further vestibular injury should occur, good compensation is likely because the other side is still normal. If the normal ear is damaged from stapes surgery, chronic imbalance is more likely, since both vestibular end organs are damaged. If a patient who has had previous surgery has abnormal ENG findings on the surgically treated ear, surgery in the second ear may increase the risk of chronic disequilibrium.

Radiography

Tomographic findings in otosclerosis correlate with the phase of the disease. Active, spongiotic disease results in decreased density of involved areas. This ranges from a small dehiscence in the normal, crisp outline of the capsule to entire loss of anatomic details. The inactive, sclerotic phase demonstrates areas of sclerosis mixed with areas of demineralization. These changes are not specific for otosclerosis and may be found in osteogenesis imperfecta, syphilis, fibrous dysplasia, neurofibromatosis, and some normal patients. Otosclerosis, however, is the most common cause of these otic capsule changes (Derlacki and Valvassori, 1965; Valvassori, 1969).

Clinical usefulness of tomography is controversial because of difficulty in interpreting findings. Extensive changes and normal findings are readily recognized; however, findings are equivocal in many cases. At best, tomography can make the diagnosis in only about two thirds of temporal bones with histologically confirmed otosclerosis. One millimeter cuts are necessary for an optimal study (Balle and Linthicum, 1984; Cody and Baker, 1978).

Computed tomography, using densitometry for diagnosis of cochlear otosclerosis appears to be more exact than using polytomography (Valvassori, 1984; Valvassori and Dobben, 1985).

Differential diagnosis

A broad differential diagnosis must be considered for acquired conductive hearing loss in patients with an apparently normal tympanic membrane. A proper diagnosis is essential to avoid unnecessary surgery and to anticipate operative difficulties.

Incus or malleus fixation

Ossicular fixation to the tegmen resulting from healing after trauma or infection comprises about 1% of the cases of conductive hearing loss in patients who have normal tympanic membranes and a progressive conductive type hearing loss. Suspensory ligaments may become ossified and fix the malleus-incus complex. Superior and anterior ligaments are most often involved. The diagnosis is suspected in less than 50% of cases preoperatively (Singleton, 1976). Careful assessment of malleus mobility with pneumatic otoscopy is helpful in making the diagnosis preoperatively. Audiometry is not helpful in the distinction from otosclerosis.

Ossicular fixation and conductive hearing loss can also result from tympanosclerosis. Most affected individuals have a long history of middle ear infection and some evidence of the same process affecting the tympanic membrane (myringosclerosis).

Stapes fixation

Stapes fixation from other bone diseases, such as Paget's disease or osteogenesis imperfecta, may be found in rare instances, although the surgical techniques used are the same as for otosclerosis.

Abnormal perilymph pressure

Certain individuals have an abnormal communication between cerebrospinal fluid and perilymph because of abnormalities in the cochlear aqueduct or internal auditory canal fundus. This transmits excessive pressure to the undersurface of the stapes footplate and ossicular chain. A mixed hearing loss is seen because of the extra pressure on cochlear structures and the footplate (Cremers et al, 1983).

If stapedectomy is performed, a perilymph gusher will result, with a high probability of total sensorineural hearing loss in the involved ear and an increased risk of meningitis.

The critical finding is that normal acoustic reflexes may be present, instead of the diphasic or absent reflexes seen in otosclerosis. If computed tomography is performed, the Mondini deformity is often found. Intraoperatively, mobility of the ossicular chain may be reduced, but the ossicles are normal and no otosclerosis is seen (Cremers et al, 1983). Perilymph gushers are said to occur in 1:500 to 1:800 stapes procedures (Farrior and Endicott, 1971; Glasscock, 1973).

Middle ear effusion

An effusion is rarely missed preoperatively; the physical findings together with a tympanogram confirm the diagnosis. When effusion coexists with otosclerosis, most surgeons consider it a contraindication to surgery if the effusion is more than an occasional occurrence. Retraction of the tympanic membrane results in an abnormal medial displacement of the stapes prosthesis, stressing the oval window seal and risking a tear. The risk of bacterial infection is also higher in ears with an effusion. A ventilating tube is undesirable, but occasionally necessary, in an ear that has had a stapedectomy.

Ossicular discontinuity

Ossicular discontinuity may result from erosion of the lenticular process of the incus or from trauma. Hearing loss may be maximal (50 to 60 dB) and acoustic reflexes absent if separation is suggestive. In contrast to otosclerosis, both air and bone conduction tracings are usually flat across the audiometric frequencies; no Carhart notch exists. The other distinguishing feature is the tympanogram showing a deep notch which reflects increased compliance. If a fibrous remnant remains between the incus and stapes, the air-bone gap may be only 30 to 40 dB and acoustic reflexes may be present, although abnormal.

Inner ear conductive hearing loss

Inner ear conductive hearing loss occurs in 1:700 patients with otosclerosis (Shea, 1984). It is characterized by a valid conductive loss and normal impedance curves and stapedia reflexes. The cause is obscure.

Congenital cholesteatoma

A conductive hearing loss in a child with a normal appearing tympanic membrane and no middle ear effusion can occasionally be caused by a congenital cholesteatoma. Usually the hearing loss is stable and the family history is negative. Careful microscopic examination is important to rule out both effusion and cholesteatoma. A high-resolution, thin-section CT scan usually demonstrates the soft-tissue attic mass.

Collapsing ear canal

Spurious conductive hearing loss from a collapsing external auditory canal can be misdiagnosed as otosclerosis. This can occur when the weight and pressure of an earphone causes the opening of the canal to close or almost occlude. Canal inserts to prevent the collapse during testing eliminate the air-bone gap. Confirmation of audiometric air-bone gaps with tuning fork tests is necessary to prevent this error.

Congenital footplate fixation

Congenital footplate fixation presents with a nonprogressive conductive hearing loss present (but not necessarily known) since birth. It may be unilateral or bilateral. It results from failure of the footplate of the stapes to separate from the otic capsule. This lack of separation may be partial or complete and consequently the associated conductive hearing loss may be maximal (50-60 dB) or somewhat less. Surgery is essentially the same as for otosclerosis, but other associated congenital abnormalities (involving the other ossicles or facial nerve) or a potential perilymph gusher may exist (Teunissen et al, 1990). These ears must be approached with caution and only by experienced surgeons.

Management

Vestibular symptoms. When Ménière's disease coexists with otosclerosis, two approaches have been used to eliminate the vestibular symptoms; medical treatment directed at Ménière's disease (Cole and Funkhouser, 1972) and medical treatment directed at the otosclerotic bone lesions (Cody and Baker, 1978). Unfortunately, neither study was controlled.

Hearing loss. After a diagnosis of otosclerosis is made on clinical grounds, four basic management options are available for the hearing loss: observation, amplification, NaF therapy, and surgery; they can be used singly or in combination. Only after carefully evaluating the symptoms and findings in relationship to the whole patient can the surgeon recommend the best course of action.

Observation

Many unilateral losses and some bilateral conductive losses in the 30 to 40 dB range do not seem to hamper job performance, school performance, or social communication. These patients may prefer to be observed. Follow-up with audiograms every 6 to 12 months is helpful to watch for progression. Personal vanity, occupation, age, and risk of the procedure are important considerations.

However, for some patients the same 30 to 40 dB loss may provide a very significant problem. For example, a driver of a vehicle with a normal left ear and such a loss in the right ear may have great communication problems if required to carry on conversation with a rider on the right side. Salespersons often require better hearing to adequately perform their duties. Each patient has his own individual needs and motivation. Patients with special needs should be encouraged to consider surgery or amplification even if they have unilateral loss.

Amplification

Speech discrimination usually is excellent in patients with a pure conductive loss and usually is well preserved in patients with mixed losses. Low-gain hearing aids in patients with relatively mild or moderate losses can be very helpful. Most patients with otosclerosis can wear a hearing aid successfully and obtain good hearing improvement. Many are satisfied using the aid only when they feel it is necessary (Smyth, 1982). Amplification is especially useful in those who cannot undergo surgery, those who prefer not to accept the risks of the procedure, those with great fears or phobias about surgery, or those who prefer an aid after being presented with options of an aid or surgery.

Sodium fluoride

Few controversies in medicine have been as polarized as the use of NaF in otosclerosis. For a discussion of the proponent's data and the use of NaF, the reader is directed to Shambaugh, 1990. For a discussion of the opposing points, the reader is directed to Kerr and Hoffman, 1989.

Much of the controversy surrounding the use of NaF in otosclerosis has been resolved. The following points can be accepted.

1. A progressive sensorineural component to the mixed hearing loss may occur in otosclerosis.
2. A pure sensorineural hearing loss associated with otosclerosis not involving the stapedial footplate and apparently caused by this otosclerosis has been documented in a few cases (Balle and Linthicum, 1984).
3. Oral NaF (with calcium and vitamin D) induces metabolic changes in otosclerotic bone (Bretlau et al, 1985; Linthicum et al, 1973).
4. The combination NaF, calcium, and vitamin D appears to retard the development of sensorineural hearing loss in otosclerosis in selected cases (Bretlau et al, 1985; Linthicum and Forquer, 1985).

Indications. The indications for NaF, calcium, and vitamin D therapy are said to be (1) vestibular symptoms, (2) progressive mixed hearing loss, (3) preoperative stabilization, and (4) sensorineural hearing loss (cochlear otosclerosis).

Treatment of the vestibular symptoms is mentioned earlier. For a progressive mixed hearing loss, Linthicum and Forquer (1985) recommend instituting treatment if there is a decrease in the pure-tone sensorineural average of 2 dB or more per year or a 5 dB worsening in any one of the thresholds at 500, 1000, or 2000 Hz. They also recommend rechecking the hearing every 6 months and increasing the dosage if progressive deterioration occurs. Careful reassessment is made after 2 years of treatment; if the condition is stable or improved at that time, treatment may be stopped or may be continued for life. Continued periodic assessment is important (Linthicum and Forquer, 1985).

Dosage and administration. NaF is given most easily as Florical (8.5 mg NaF and 364 mg calcium carbonate per capsule), which is available without a prescription and can be ordered through pharmacies and health food stores.

The usual dosage is 40 to 50 mg NaF per day, usually two tablets with meals three times a day, plus 500 mg of vitamin D daily. If calcium and vitamin D are not given with the fluoride, secondary hyperparathyroidism may result.

Precautions and side effects. Side effects of NaF are usually not serious but can be troublesome. In one study, 18% of 343 patients experienced side effects and 8% stopped taking the medication for this reason (Cody and Baker, 1978).

The most common problems were gastrointestinal (14%), including nausea, vomiting, heartburn, diarrhea, and, rarely, loss of appetite. The second most common side effect was musculoskeletal pain (3%), usually joint pain. Rare side effects included ankle edema and light-headedness. One patient developed renal calculi 6 months after therapy began, and another developed them after 14 months. Two patients with moderate renal failure developed spontaneous bilateral hip fractures while on NaF. Biopsies showed severe fluorosis in the first case and osteomalacia and skeletal fluorosis in the other. Gerster et al (1983) consequently recommend reduced dosages in patients with renal insufficiency. Shambaugh and Causse (1974) detected fluorosis radiographically in 0.25% of 4000 patients. With fluorosis, abnormally hard and brittle bony changes develop because of excessive fluoride intake, and the affected individual is at increased risk for fractures. The diagnosis is made by detecting sclerosis and thickened trabeculae radiographically; these changes are readily apparent, especially on pelvic and spinal views. In the Shambaugh and Causse (1974) series, the fluorosis resolved in half the patients within 2 years and caused no sequelae in the others.

Surgical Management

History

Surgery for otosclerosis has a long history (Hillel, 1983). One-stage fenestration of the lateral semicircular canal was the standard procedure for 20 years after Lempert perfected it. Stapes mobilization and later stapedectomy replaced it in the 1950s because the hearing results were better; fenestration left an average residual 20 to 25 dB conductive loss even in expert hands.

Prosper Ménière described the first stapes mobilization as having taken place over 160 years ago. On February 18, 1842 Ménière watched, through a large meatus, while a deaf judge revived the sensitivity of his own ear with a long gold needle. Ménière saw the needle impinge on the umbo of the malleus and noted the concavity of the drum increase at the same instant the patient became aware of increased hearing. Ménière concluded that movement was transmitted to the stapes and inner ear via the ossicular chain.

Kessel did the first surgical stapes mobilization in 1878. By 1888, Boucheron reported 200 cases of mobilization - many with excellent results.

Although he did not do it for otosclerosis, Frederick L. Jack, performed the first stapedectomy June 18, 1892. Blake had planned the project, but having temporarily lost the use of his right hand, he had Jack proceed. The patient was a 12-year-old girl who had experienced aural discharge since childhood scarlet fever. After removing the tympanic membrane, malleus, and incus, Jack noticed the head of the stapes was carious. He passed a knife around it to loosen it and extracted it with a small hook. The morning after surgery, the patient heard sounds, whereas before surgery she had heard very little. Many of Jack's first 16 cases were done for suppurative otitis media! In fact, he noted specifically that in sclerotic cases, part or all of the footplate frequently remained behind and that the results were not as good in cases resulting from suppuration. He concluded that inflammatory reaction was unusual, and that the procedure was followed by no bad results. Blake mentioned one case in which vertigo lasted five months!

In March of 1898 Alderton turned down the posterior-superior quadrant of the tympanic membrane as a flap while his patient was anesthetized with cocaine. Alderton cut the stapedius muscle, disarticulated the incudostapedial joint, and removed the incus. There was no hearing improvement. In the fall of 1898, Alderton again reflected the flap and removed the stapes superstructure. The patient became dizzy but heard better for 24 hours. On January 25, 1899, Alderton again exposed the ear and drilled a hole through the stapedial footplate with a guarded trephine, producing perilymph without producing dizziness. At 6-month follow-up, the patient's hearing was improved.

In 1945 and 1946 Cawthorne performed stapedectomies on 14 patients using a fenestration approach. He placed amniotic membrane over the oval window and pressed a tympanomeatal flap into the oval window - holding it in place with bone wax. In 1950, he did an additional 8 stapedectomies. In these, he removed the malleus to aid in the flap displacement. Five of eight patients improved, but as they did not improve as much as is usual with fenestration, he abandoned the procedure.

In 1950, John Lindsay observed a patient whose husband, in play, had accidentally pushed a bobby pin into her ear - after which she noted that her hearing improved. Lindsay noted a healed scar over the patient's incudostapedial joint. He found that the hearing in her ear had improved 40 dB from the incident, taking it to a level 20 dB better than her opposite, fenestrated ear. Eight years later, her hearing in the "bobby pin" ear was at the same level as that in the fenestrated ear (Lindsay, 1962).

Samuel Rosen, in September of 1951, while using the Lempert approach he had previously used for sectioning the chorda tympani and Jacobson's nerve for Ménière's disease, began palpating the stapes in fenestration candidates. His hypothesis was that patients with only partial fixation of the stapes were poor candidates for fenestration whereas those with complete fixation were good candidates. April 3, 1952, while using this technique on a 43-year-old electrical engineer, he found the stapes to be incompletely ankylosed. Pressure against the neck of the stapes caused free mobility of the stapes with a sudden return of hearing that was maintained.

Knowing of Rosen's success mobilizing the ossicular chain, Sylvester Daly attempted to accomplish the same thing by passing bougies up the eustachian tube. He was able to see the bougie reach the incudostapedial joint at times, and he believed he could massage the stiff ossicular chain. He pointed out that the technique was ineffective in completely fixed stapes.

E. P. Fowler, Jr, in July of 1955 reported his anterior crurotomy. He weakened the footplate posterior to the anterior focus, after which he fractured the anterior crus.

Victor Goodhill recommended using transincudal pressure instead of capitular pressure to avoid joint disarticulation. If the joint were loose or became loose, he glued it with pentocryl before mobilizing the stapes. If he were unable to mobilize the stapes, he applied a microvibrator to the incus, which yielded a hammerlike force.

In July of 1956, Howard House began shattering the stapes footplate and placing a plastic prosthesis between the incus and the shattered footplate.

Harold Schuknecht in 1956 began mobilizing the stapes footplate using a pick at the footplate margin when mobilization at the capitulum failed. Later the same year, he began using Heermann chisels at the edge of the footplate to accomplish mobilization. During the same year, he began mobilizing a portion of the footplate and placing a tantalum wire from the incus to the mobilized portion of footplate.

In December of 1956, Myerson began mobilizing the stapes by applying pressure to the short process of the malleus. Later he touched the short process of the malleus with an electromagnetic microvibrator, which he used to individual pain tolerance. If there were no improvement in 1 month, he reflected the tympanic membrane and used the vibrator on either the incudostapedial joint or on the footplate.

J. Brown Farrior, in September of 1957, did a partial stapedectomy, cut the stapedius tendon, cut the posterior crus of the stapes very low, performed an anterior crurotomy, created a mobilized central island, and repositioned the posterior crus on the island.

Arthur Juers did a similar procedure although he fractured the footplate into multiple fragments and placed the posterior crus over the fragments, packing Gelfoam around the junction. At times he removed the posterior footplate, closed the oval window with perichondrium, and transposed the posterior crus over the perichondrium.

J. V. D. Hough, independently, in June of 1956 performed partial stapedectomy - removing the otosclerotic focus, sealing the window with Gelfoam, and positioning the posterior crus of the stapes over the Gelfoam.

Michel Portmann actually removed the stapes intact and replaced it over a vein. In other cases, he removed the footplate, sealed the oval window with a vein, and repositioned the crura over the vein.

John Shea, Jr, in May of 1956 fenestrated the oval window, covered the opening with subcutaneous tissue, and placed a Teflon stapes between the incus and the oval window. This evolved into the oval window fenestration, vein graft seal, and placement of a polyethylene tube between the incus and the vein (August, 1957).

In 1957, Harold Schuknecht and Claire Kos independently began using tissue plugs. Each used wire to make the prosthesis. For tissue, Schuknecht selected connective tissue and Kos selected vein for the plug to seal the oval window.

Bruce Cornish (1959) of Auckland, New Zealand approached mobilization in a different manner. He inserted an electrode into the stapedius tendon and placed an indifferent electrode on the posterior canal wall. By applying a current, he was able to mobilize the stapes in some patients.

Indications

Unilateral otosclerosis

In adults with unilateral otosclerosis and normal hearing in the opposite ear, optimal results can be obtained only if the hearing in the two ears becomes nearly equal after surgery. The ideal candidate has preoperative bone conduction levels between 0 and 30 dB and a negative Rinne test with a 512 Hz tuning fork. Adequate cochlear reserve should also be present, as indicated by a speech discrimination score of at least 80%.

A speech-in-noise discrimination test has shown (Donaldson and Selters, 1961) that there is no "magic level" above which a patient whose unilateral hearing loss has been surgically corrected will do well and below which he or she will do poorly. Although improvement in a unilateral or second ear (of bilateral ears) is more subtle than in the first ear, in general patients are pleased with improvement in unilateral losses in proportion to the improvement.

Surgery is contraindicated in the patient with unilateral otosclerosis if the otosclerotic ear is the better hearing ear.

Bilateral cases

When patients with bilateral clinical otosclerosis have the preoperative audiometric conditions already mentioned for the ideal unilateral candidate, they also are candidates for surgical correction. When bilateral bone conduction thresholds are poorer than 30 dB, patients may still be good candidates for surgery even if discrimination scores are slightly less than

80%. Postoperative amplification may still be necessary for optimal hearing, even though the hearing in the treated ear improves considerably. The need to use the aid only part-time, the advantage of a longer battery life, and the need for a smaller aid may make the surgery worthwhile.

When selecting which ear (in bilateral otosclerosis) on which to operate, the cardinal rule is to operate on the poorer hearing ear. To decide which is the poorer hearing ear, the otologist will need to consider pure-tone averages, speech discrimination, the effects of amplification, and the patient's comment as to which ear hears better. If one ear has had previous surgery, the results of this procedure need to be factored into the decision as to whether a revision operation ought to be carried out on the previously operated ear or whether surgery should be performed on the unoperated ear. At all times it must be borne in mind that in rare instances, after the operation, the unoperated ear will become the only hearing ear and the patient will need to depend on it for the rest of his or her life.

A few patients with long-standing otosclerosis have audiograms that show no measurable bone or air conduction thresholds although they have a history typical of otosclerosis. Usually such patients maintain easily understandable speech and have successfully used amplification in the past. They usually have both stapedial and cochlear otosclerosis. A 70 to 75 dB bone conduction threshold level in such a patient will not be measurable, since most audiometers have bone conduction output no greater than 60 to 65 dB. A superimposed 40 to 50 dB air-bone gap makes the air conduction threshold levels barely measurable, if measurable at all. Preoperatively, it is difficult to establish with certainty that the patient is a candidate for successful surgery. These patients may have severe to profound loss such that the speech recognition list cannot be presented at a loud enough level to estimate the maximal score for this aspect of the hearing function. The patient's hearing is beyond the maximal limits of the equipment. It is helpful in such cases to test discrimination with a powerful hearing aid in a free-field test paradigm. This technique gives you the maximal intensity limits of the hearing aid, which may be 130 to 145 dB. Improving the air conduction threshold in the treated ear to 80 dB or better may allow successful use of amplification.

Vestibular symptoms

Most authors agree that stapes surgery is contraindicated in the presence of active Ménière's disease. Whether one can prudently recommend it as a therapeutic alternative in these patients when vestibular symptoms have abated is another question. Black et al (1969) reviewed the case of a man with reduced hearing from progressive otosclerosis and Ménière's disease (both extensive bilateral labyrinthine otosclerosis and endolymphatic hydrops were verified at autopsy) who benefited from stapedectomy with no vestibular symptoms postoperatively. When this case is added to that of Linthicum and Neely (1977) and those of Issa et al (1983), however, two of six surgically treated ears in patients with otosclerosis and Ménière's disease experienced postoperative loss of all labyrinthine and auditory function, despite the preoperative absence of vestibular symptoms for more than 2 years. Only under unusual circumstances ought such risks be accepted.

Smyth et al (1975) detected decreased caloric responses 3 months postoperatively in 30% of 26 ears with normal values preoperatively. In most instances the abnormalities improved and sometimes returned entirely to normal by 1 year postoperatively. None of these patients had symptoms of vestibular dysfunction. Clinical experience has shown that when stapedectomy damages one vestibular end organ, as indicated by symptoms persisting for several weeks postoperatively, an increased likelihood exists for permanent imbalance after stapedectomy on the second ear. ENG is recommended before a second stapedectomy in any patient who exhibits vestibular symptoms longer than 2 weeks after the surgery on the first ear. The results of the vestibular findings should be considered in making the surgical recommendation.

Second ear

The surgeon must deliberate carefully before recommending surgery on the second ear for otosclerosis. The advantages of symmetric, bilateral hearing for determining the direction of the sound source, as well as hearing equally well from each side, are compelling, but other considerations exist. The results of surgery on the second ear are less dramatic than on the first, since often little improvement occurs in overall hearing ability. Simply improving sound perception close to the involved ear and slightly improving sound localization may disappoint some patients. Realistic expectations must be stressed in preoperative counseling (Smyth, 1982). The second ear should be operated only if the functional result in the first ear is good and the hearing is stable; waiting 1 year is appropriate. Surgery on the second ear should not be done if the first ear had a significant delayed sensorineural hearing loss.

Surgery on the second ear is also contraindicated when significant vestibular injury has resulted from the first stapedectomy. Careful questioning about vestibular symptoms after the first surgery is important before considering surgery on the second ear. ENG studies have shown a 30% rate of abnormal vestibular testing in asymptomatic patients who preoperatively had normal vestibular tests (Smyth et al, 1975). Such patients can often compensate well for this unilateral vestibular loss and be entirely asymptomatic, but if surgery similarly damages the second ear, symptoms may result. ENG is appropriate when vestibular symptoms followed the first stapedectomy. Bilateral vestibular injury may result in chronic disequilibrium and imbalance, especially in older adults.

Otosclerosis in children

Surgical findings indicate that otosclerosis in children tends to be more severe than in adults. Whereas "biscuit type" and obliterative otosclerosis make up less than 10% of adult cases, in Cole's series of children with otosclerosis, 42% of his children either had circumferential otosclerosis, biscuit type otosclerosis, or oval window obliterative otosclerosis (Cole, 1982). The incidence of advanced otosclerosis slowly increases with the duration of the hearing loss (Robinson, 1983).

Results of surgery show that stapedectomy in children is as effective and safe as in adults when performed by experienced surgeons (von Haacke, 1985). Closure to within 10 dB in speech frequencies can be expected in more than 80% of cases. Delayed sensorineural loss (greater than 10 dB) is found in 1% to 3%.

Although stapedectomy in children is safe in expert hands, potential benefits must be weighed against risks. Improving the potential development of speech and language is not reason enough to operate, since surgery is never recommended in children under age 5 years and amplification can successfully meet the need for adequate hearing to develop speech and language. Similarly, amplification can adequately manage the communication needs of children with conductive hearing losses. Personal and family counseling may be necessary for acceptance of amplification, but the lack of interest in a hearing aid should not be an indication for surgery in a child except in the most unusual circumstances. The senior author ordinarily waits to operate on children until they are old enough to have the procedure under local anesthesia and can make a mature informed decision regarding the surgery (House et al, 1980).

Clinical otosclerosis in children presents a more difficult problem than when it appears in later life, both because of the more aggressive bony abnormality and the special considerations of the child's activities and risks. Thus it is recommended that surgery for otosclerosis that develops in childhood be performed only by those with extensive surgical experience.

Informed consent

Since a predicable risk of a poor surgical result exists, all patients must realize this and be prepared to accept it. Those who have reservations about accepting a bad result should use a hearing aid until they can accept the appropriate risks of stapedectomy or stapedotomy. As with any surgery, details of the procedure, the expected perioperative course, and the potential complications and ill effects should be completely explained to the patient. It is important that the otosclerosis surgeon keep a running record of his or her results so that expectations related to these results rather than those from the literature are quoted to the patient.

Technique

Instrumentation

There are many well-designed instruments available to help the surgeon perform stapedectomy. Experienced surgeons develop marked preferences for specific instruments, but it is preferable to learn to use a few instruments well than to attempt to have a large array available.

Anesthesia

Stapes procedures can be done safely under local or general anesthesia. Local anesthesia supplemented with intravenous sedation is safe and effective and is recommended for most adults. The senior author prefers to give 2.5 to 5 mg of diazepam IV as soon as the patient, who already has an IV line in place, is on the operating table. Additional diazepam is used as needed during the procedure.

All surgery is done with ECG and blood pressure monitoring. Intermittent verbal contact with the patient by the surgeon or the circulating nurse is important to monitor the level of sedation. An initial injection of 1.8 mL 2% Xylocaine with epinephrine added is injected in the lateral part of the ear canal prior to preparation of the ear canal. It is convenient to use an Astra 9002-00 dental syringe; 1.8 mL cartridge; and a 30 gauge needle. Although other surgeons prefer 1:8000 to 1:100.000 epinephrine, high concentrations can sometimes result in cardiac arrhythmias. The senior author uses 1:50.000 epinephrine and finds that it provides adequate, safe vasoconstriction and more than sufficiently prolonged anesthesia. In patients with elevated blood pressure, it is important to achieve optimal medical control of the pressure prior to surgery.

An additional injection of about 0.5 mL of the same anesthetic agent is made into the medial part of the ear canal after the ear is prepared and the ear canal has been irrigated with Betadine followed by normal saline. In hypertensive patients, the lateral injection is made with 2% Xylocaine without epinephrine. The injection of the flap medially is with 2% Xylocaine with 1:100.000 epinephrine added. This usually results in excellent hemostasis. If it does not do so, the procedure is terminated.

Any local anesthetic reaching the middle ear may be absorbed through the secondary tympanic membrane with resultant postoperative dizziness. Occasional transient facial paralysis lasting from 2.5 to 4 hours may result from injection of the local anesthetic. Local anesthesia has a number of advantages. The effectiveness of ossicular chain reconstruction can be readily determined by voice testing. Vertigo resulting from footplate manipulation or prosthesis placement can warn the surgeon to alter technique. Finally, the patient avoids the cardiac and pulmonary risks of general anesthesia. General anesthesia is preferred by some surgeons, who find it is more comfortable for the patient and surgeon and eliminates their concern about sudden patient movement or nausea and vomiting during the procedure.

Approaches

Wide fenestra stapedectomy. Wide fenestra stapedectomy involves removal of a third or even more of the footplate. The wide fenestra technique has been used since the early days of stapedectomy and has gained wide acceptance as a safe and effective procedure (Robinson, 1981).

After footplate removal, Gelfoam ought not be used as the oval window seal. Although fast and technically easy to use, the wire-Gelfoam prosthesis has a higher incidence of complications than tissue seals. The incidence of granuloma formation, drop in discrimination score, perilymph fistula with or without a high neomembrane, recurrence of the conductive hearing loss from migration of the wire, and long-term drop in sensorineural function are all higher with Gelfoam when compared with tissue seal (Sheehy and Perkins, 1976; Sooy et al, 1973).

Use of a sheet of tissue (fascia, perichondrium, vein) to cover the oval window can almost eliminate the incidence of perilymph fistula. Vein has the advantage of greater elasticity than fascia and perichondrium and lays more smoothly along the niche walls; however, vein is slightly less convenient to harvest since it requires preparation of an additional field.

Small fenestra stapedotomy. Small fenestra stapedotomy was first described in 1972 and has become increasingly popular. Theoretic advantages include less risk of direct damage to the saccule and utricle and prevention of delayed, scar-induced migration of the prosthesis.

The diameter of the piston in the small fenestra technique is important. As the diameter becomes larger, lower frequencies (0.5, 1, 2, kHz) are conducted well, but the high-frequency response (4 kHz) suffers. As the prosthesis diameter becomes smaller, the converse is true. Most authors agree that 0.4 to 0.6 mm prostheses give acceptable high- and low-frequency responses.

The advantage of stapedotomy includes better air-bone closure at 4 kHz. Since much of the information in consonants is found in the second- and third-level formants, better high-frequency response can give better speech discrimination. Also stapedotomy gives better long-term sensorineural function at 4 kHz, a reduced incidence of total sensorineural loss, and fewer postoperative vestibular complaints (Bailey et al, 1983; Fisch, 1982). Unfortunately long-term results are not available. In theory, tissue placed around a piston, rather than between the piston and the inner ear, may not adhere to the piston. Such a circumstance may lead to the formation of a fistula many years after the procedure if there is no seal between the tissue placed around the piston and the piston itself. Only long-term results in significant numbers of patients will answer this question. The major disadvantages are technical. The crura must be fractured without mobilizing the footplate. The fenestra must be centered on the footplate. These may be made mechanically or with a laser. Positioning the prosthesis can be difficult because of the fenestra's small size. The prosthesis length is critical (McGee, 1981).

The method of Causse et al (1985) uses stapedotomy but combines it with vein as a tissue seal. This method ought to be as free from the potential complication of a fistula as wide fenestra stapedotomy with a tissue seal.

Management of advanced disease. In severe and profound hearing loss with advanced disease states, the success rate falls far below that of blue footplate disease, to less than 50%. The chance of significant inner ear loss also rises, with the rate depending on the surgeon's judgment, experience, and skill. These cases are definitely not for the occasional stapes surgeon. Fig. 170-5 illustrates the results of a successful surgery for a patient with advanced disease.

The biscuit footplate in the advanced otosclerotic group is the most amenable to management. In this type the footplate is white and mounded up in the center with the edges of the oval window niche visible. A piston is essential during surgery because recurrent otosclerosis will rapidly engulf a tissue-wire prosthesis in most cases. This is essentially a drill-out operation to convert a white footplate into a blue one, which is then managed as any blue footplate. A slowly rotating diamond burr is used to go over the footplate in circular and anteroposterior strokes and effect a uniform reduction in the footplate's thickness before perforation occurs. Once perforation occurs, drilling must be stopped, since it may result in excessive mobilization of inner ear fluids and significant damage to inner ear function. After the footplate is appropriately thinned, the distance between the long process of the incus and the thinned footplate is carefully measured. A piston 0.25 mm longer than the measured distance is selected and placed after removal of sufficient footplate to accept it.

In obliterative otosclerosis, the edges of the oval window are not visible. In fact, the oval window niche may be filled with otosclerosis enabling the surgeon to see only a mass of bone extending from the facial nerve to the promontory, with only the capitulum and part of the crural arch visible. These are dangerous ears in which to perform a drill-out, since the facial nerve and the inner ear may be injured. Bulging of otosclerotic bone into the vestibule may occur. Less severe cases can be satisfactorily drilled out with a microdrill thinning the entire window area until a uniform bluish color indicates the proper thinness. A circumferential thin blue line is then created, the footplate is transected at its midpoint, fractured, and removed (Sooy et al, 1964).

Otosclerosis obliterating both the oval and round windows is the rarest type of advanced disease. Unfortunately, it is not amenable to current surgical techniques.

Intraoperative complications

Floating footplate. Every surgeon who performs many stapedectomies will occasionally experience a floating footplate. The senior author prefers to make a small "pot hole" in a thin area of the footplate by scratching with a straight pick prior to fracturing the stapes superstructure. Should a floating footplate develop, a fine pick can be used to enlarge the "pot hole", and the footplate can be removed with a fine hook. In addition, the "pot hole" decreases the suction effect of the inner ear if the stapes is extracted in one piece during the attempt to fracture the superstructure. Because of lack of visibility, a "pot hole" cannot always be made. If one has not been made and a floating footplate results, the surgeon has several options. A fine pick or microdrill can be used to make an opening in the edge of the promontory adjacent to the midpoint of the caudal edge of the promontory adjacent to the midpoint of the caudal edge of the footplate. Once this opening is large enough, a fine hook can be used to extract the mobile footplate. Should the footplate remain mobilized, the patient will be able to hear. Should the footplate refix, it can be removed as originally intended. A third approach is to use a laser to make a hole in the footplate and then to extract the footplate or place a piston through the hole as the surgeon prefers.

Submerged footplate. If the footplate or a major piece of it drops into the vestibule, no attempt should be made to chase it. Instruments should never be placed in the vestibule. The surgeon should simply accept that it happened and finish the surgery. Many of these patients have few vestibular symptoms postoperatively. Some will experience postural vertigo for several weeks, but fortunately the vertigo is usually self-limited.

Vertigo. A patient will rarely have vertigo, nausea, and vomiting before any footplate work is done. This may be a caloric effect from the suction or from absorption of local anesthetic through the secondary tympanic membrane. Both problems respond to additional intravenous diazepam.

If vertigo occurs later, it is usually during footplate manipulation or at the time the prosthesis is placed, and it is usually momentary. If it continues, the prosthesis may be too long and should be replaced after careful remeasuring. If a prosthesis cannot be placed without causing vertigo, the ear should be closed. The surgeon should wait for a neomembrane to form in its natural position and attempt to place the prosthesis 1 or 2 months later.

Partially dislocated incus. The incus may be partially dislocated if the surgeon, while removing a hook from the ear catches the long process. This is a very unusual complication for an experienced surgeon. If the incus has been dislocated straight lateralward, it should be carefully returned to its original position where it will usually remain. If the mucosal folds and ligaments holding the incus in place have been too disrupted and the incus does not appear secure when returned to its original position, the surgeon should use a tissue-wire prosthesis rather than the prosthesis originally intended. The tissue bulk should be large enough to prevent it from sliding into the vestibule. Should the post-operative air-bone gap be unsatisfactory, the ear can be reopened and a piece of tragal cartilage inserted between the long process of the incus and the tympanic membrane.

Perforated tympanic membrane. Although it is very unusual, even an experienced otologist could tear the tympanic membrane while elevating a flap or while separating a thin tympanic membrane from an incus to which it is adherent. If this is identified prior to removal of the footplate, the tear should be buttressed with tissue and the procedure terminated. If the tear is not identified until after the footplate is removed, the operation should be complete and the perforation buttressed with some of the same tissue taken to seal the oval window.

Postoperative complications

Otitis media. Acute suppurative otitis media is a rare but potentially life-threatening complication. It usually results in a total hearing loss in the involved ear and may lead to secondary bacterial labyrinthitis and meningitis. Risk factors include recent or synchronous upper respiratory infection, traumatic surgical technique, tympanic membrane perforation, break in sterile technique, and chronic systemic disease with immune compromise (Matz et al, 1968). Use of prophylactic perioperative antibiotics is controversial, and no controlled studies have demonstrated benefit. The senior author prescribes a 5-day course of penicillin or erythromycin postoperatively.

Granuloma. Granuloma of the oval window occurs after 0.6% to 3% of stapedectomies. Most develop within 6 weeks and appear with sudden or gradual hearing loss or vertigo. Physical examination shows a red-gray discoloration posterosuperiorly. Audiograms demonstrate sensorineural hearing loss in almost all patients and decreased speech discrimination in most.

Exploration of the ear should be done as soon as possible to prevent progression of a serous labyrinthitis to an irreversible fibrinous phase and permanent hearing loss. At surgery, a red, soft, vascular mass is seen extending from the niche around the prosthesis and long process. The granuloma and prosthesis should be very gently removed and a different graft material placed. Recovery of cochlear function is complete in 20% of patients and partial in 30%. No change occurs in 30% and further loss occurs in 20%. Kaufman and Schuknecht (1967) did not find the granulomas were related to "graft material used, the surgical technique, starch glove powder, or infection".

Perilymph fistula. A fistula should be considered in the differential diagnosis of almost any problem that arises after stapedectomy and is evident in 9% to 16% of ears undergoing revision for any reason. Characteristic symptoms should arouse suspicion. Hearing

loss is present in 63% to 100% of cases. It usually fluctuates and, when present, is mixed or sensorineural in 85% of patients and entirely conductive in 15%. Vertigo occurs in 58% to 70% and tinnitus in 50%. The symptoms may begin any time after stapedectomy; the longest reported interval is 15 years (Althaus and House, 1973; Harrison et al, 1967; Mawson, 1975; Sheehy et al, 1981).

Physical examination may aid in diagnosis if a positive fistula test is present (using a Politzer bag, pneumatic otoscopy, tympanometry, or platform test), but demonstrated fistulas may not evoke one.

Fistulization rate is highly dependent on the type of oval window seal. Use of no seal or Gelfoam gives rates as high as 4% in both stapedectomy and stapedotomy (Sooy et al, 1973). The rate for fat-wire prosthesis is about 1%. The lowest rates are found with a sheet of tissue (vein, fascia, perichondrium) beneath the prosthesis (0% to 0.6%).

At exploration, the area around the prosthesis should be closely observed. Fluid leakage can be elicited by pressure on the jugular vein, Trendelenburg's position, Valsalva's maneuver by the patient, or positive pressure ventilation by the anesthesiologist if general anesthesia is used. If a fistula is identified, the prosthesis should be removed and replaced over a tissue graft, if this can be safely accomplished. If dizziness is encountered when removing the prosthesis or seal, it is best to leave it in place, freshen the surface of the tissue around the fistula, and apply a fresh tissue graft to the area.

Results of fistula repair after stapedectomy are encouraging. Vertigo is much improved or eliminated in 76% to 100% of patients. Hearing is improved or stabilized in 73% to 78%. Patients with dizziness have a worse prognosis for return of an associated hearing loss. Tinnitus persists in 87%. Results tend to be better if the fistula is repaired early (Halvey and Sade, 1983).

Sensorineural hearing loss. Severe cochlear loss can be detected in the early postoperative period. When it is caused by an inadvertent occurrence at surgery, an associated complete loss of vestibular function usually results in a characteristic clinical picture (Chapter 145). Errors in technique that can contribute to these incidents include excessive movement of inner ear fluids during extraction of the stapes, injudicious suctioning, rupture of the membranous labyrinth during attempts to remove pieces of footplate, and acoustic trauma imparted by the surgical drill (Schuknecht, 1962; Sheehy et al, 1981; Smyth, 1982).

If the patient has an uneventful hospital course after stapedectomy and suddenly develops persistent vestibular symptoms and tinnitus, returning to the hospital for emergency tympanostomy is advised. Fistula or granuloma will most likely be found.

Persistence of air-bone gap. Persistence of a negative Rinne's test at 3 to 4 months with no visible explanation indicates the need for an exploratory tympanotomy. The surgeon first palpates the malleus for mobility, since a fixed malleus and otosclerosis may occur in the same ear. (This ought to have been checked for and detected at the initial operation, but the surgeon may have felt the motion was adequate when it was not. The more experienced the surgeon, the more likely he or she will correctly interpret the amount of malleus motion). The surgeon next palpates the incus for mobility. If either the malleus head or the incus body

is fixed, it will fix the other. To find out which is at fault, the surgeon inserts a sickle knife into the incudo-malleal joint, and twists it slightly. It should then be obvious which ossicle is at fault.

The surgeon then palpates the long process of the incus while visualizing the round window niche in search of a light reflex. It is usually necessary to place a drop of Ringer's solution or normal saline in the niche to detect motion when the incus is moved. If the malleus and incus are normal, nothing more can be done and the ear should be closed. If no light reflex occurs on incus palpation, the surgeon attempts to elicit a light reflex by palpating the prosthesis itself. If none is obtained, the area of the neomembrane is palpated. If a light reflex is obtained with this maneuver but not by palpating the prosthesis or incus, the oval window is at fault and should be explored very carefully. The prosthesis is fixed by regrowth of otosclerotic bone, scarring to the edge of the oval window, or hung up on remaining footplate.

Loose wire. The most common symptom of a loose wire is fluctuating hearing. Middle ear air pressure, even in normal ears, is never absolutely constant, and a slight shift will favor or disfavor the contact between the wire and the incus. Since a loose wire tends to increase erosion of the long process of the incus, the situation compounds itself.

Incus necrosis. Incus necrosis results in a maximal conductive hearing loss, with an air-bone gap of 55 to 60 dB. If adhesions between the prosthesis and the incus or tympanic membrane favor sound transmission, however, the loss may be only 20 to 30 dB. In restoring the hearing, the surgeon may need to insert a columella between the neomembrane and the tympanic membrane, or if the angles are favorable, to the malleus. A three-ossicle chain, however, is consistently superior to any other arrangement. Recrimping the wire loop on the stump of the long process is seldom possible because of its taper, but its chances of staying will be improved by drilling a notch (Steffan) in the long process to receive the wire. Another useful procedure is to place a stainless steel bucket prosthesis (Lippy-Robinson) under the long process stump and turn the handle up over the lateral surface of the stump of the incus. The senior author has had the best results by placing a homograft stapes, upside down, between the long process of the incus and the oval window membrane.

Delayed sensorineural hearing loss. Slowly progressive postoperative sensorineural hearing loss frustrates the surgeon and the patient. It may result from the natural history of otosclerosis or the surgical technique. In a large series of patients, the loss averaged 5.5 dB per decade and reached 40 dB by 18.5 years postoperatively.

Surgical technique may affect this loss. Stapedotomy preserves response better at 4 kHz than stapedectomy and results in better discrimination scores. In speech frequencies postoperative stapedectomy patients reach a 40 dB loss after 13 years, whereas stapedotomy patients do not reach this level until 21 years after surgery.

For patients with bilateral disease, when only one ear underwent surgery, the average bone conduction levels are better preserved in the treated than in the untreated ear (Smyth, 1982).

Vestibular dysfunction. Vestibular symptoms in the immediate postoperative period usually persist only for hours or for one to two days. ENG studies (Ali and Groves, 1964) reveal that 80% of patients after stapedectomy have spontaneous nystagmus and 43% have positional nystagmus that resolves during the first week after stapedectomy. Detailed neurologic examinations show abnormal gait (53%), past pointing (7%), and Romberg's sign (16%), all of which resolve in the first week.

Vestibular symptoms that persist for more than a few weeks are unusual and usually are caused by uncompensated vestibular end organ damage. Severe end organ damage may sometimes be well compensated without lasting symptoms or hearing loss. Caloric tests done at 6 to 12 weeks postoperatively reveal paresis or no response in 27% of patients. Most of these returned to normal, 9% eventually progress to a total loss of caloric response but had minimal vestibular symptoms (Ali and Groves, 1964; Stroud, 1963).

Expected results

The surgeon must be cautious when evaluating reports that compare different types of prostheses. Some reports compare preoperative bone levels with postoperative air levels, not taking into account the Carhart effect. The postoperative air level should be compared with the postoperative bone level (Elonka et al, 1982). In general, hearing results and the incidence of complications depend more on the surgeon's skill and experience than the technique and prosthesis used (Sheehy, 1983).

When no preoperative vertigo develops and a blue footplate is found, experienced surgeons should expect a postoperative air-bone gap of 10 dB or less in approximately 90% of cases, despite the technique or prosthesis used. No more than 1% of ears should develop a marked sensorineural loss. Results that are worse than this should prompt a reassessment of technique, but if results are up to these standards, no changes are necessary. The results will be poorer when a biscuit footplate is present and much poorer if attempts are made to correct obliterative otosclerosis.

Revision surgery is less successful than primary stapedectomy. The results will depend on the problem responsible for the initial failure and the expertise with which the problem is handled (Lippy and Schuring, 1983; Sheehy et al, 1981).