Chapter 14: Otosclerosis

Philip H. Beales

The bone of the otic or labyrinthe capsule may be affected by many conditions of diverse aetiology and the majority of these are generalized diseases affecting the skeletal system as well as the temporal bone. Otosclerosis is confined to the otic capsule and, together with a few other conditions, gives rise to deafness and vestibular symptoms.

Definition

Otosclerosis is a hereditary localized disease of the bone derived from the otic capsule. Mature lamellar bone is removed by osteoclasts and replaced by woven bone of greater thickness, cellularity and vascularity.

The otosclerotic focus may be asymptomatic or, if in the area of the stapes footplate, may give rise to ankylosis and conductive deafness. Other parts of the labyrinthine capsule may be involved resulting in sensorineural deafness and vestibular abnormalities. The focus produces an enzyme which may give rise to sensorineural deafness and vertigo, whether or not the stapes footplate is affected. A combination of effects may thus be produced by the otosclerotic lesion sometimes referred to as 'histological', 'stapedial', 'cochlear' and 'combined otosclerosis'. The commonest manifestation seen clinically is of the combined variety where there is both a conductive and sensorineural hearing loss.

Many European otologists use the term 'otospongiosis' when referring to the active vascular focus, but in North America and in the UK the term 'otosclerosis' is used and this refers to the final inactive stage of the lesion where the bone is sclerotic or hardened; neither of these terms is strictly accurate.

History

The first description of ankylosis of the stapes is attributed to Antonio Valsalva who, in 1741, carried out a post-mortem examination on the body of a patient who was believed to be deaf. In 1861, Joseph Toynbee, noted ankylosis of the stapes footplate in 39 out of a total of 1959 temporal bone dissections.

In 1894, Adam Politzer introduced the term 'otosclerosis' and gave the first final account of the histopathology of this condition. In this disease, fixation of the stapes footplate by bony deposits occurs, leading to progressive ankylosis of the footplate in the oval window niche and a progressive conductive deafness. The term 'otosclerosis' refers to the final inactive stage of the process, while the essential pathological lesion is, in fact, a replacement of lamellar bone.

Siebenmann introduced the term 'otospongiosis' in 1912, which referred to the active and vascular stage of the process and this term is widely used in Europe. It is more accurate as it indicates that an active lesion may be present. The belief that the process is inactive, or soon becomes so, has delayed understanding that sensorineural deafness if often an integral part of the disease. Politzer's work was of fundamental importance as he demonstrated, for
the first time, that the stapedial ankylosis was not secondary to 'chronic middle ear catarrh', which was previously believed to be an inflammatory condition, but was the result of a primary disease of the labyrinthine capsule. It was not until nearly 50 years later, when the fenestration operation allowed direct inspection of the oval window in the living patient, that the concept of a chronic catarrhal condition causing secondary fixation of the stapes footplate, was finally abandoned.

**Aetiology**

Otosclerosis is a disorder affecting the growth of collagen and it is only seen in the human species. Despite intensive research, the cause of the development of the disease process remains obscure. The characteristic lesion is a deposit of new bone with a different fibrillar and cellular pattern which is laid down at certain sites in the temporal bone. Known sites of predilection are the oval and round windows, and in these areas cartilaginous rests are found. Otosclerotic foci have been observed in other areas, remove from these special regions and, in these cases, symptoms of conductive deafness do not occur. This condition has been termed 'non-clinical', or 'histological', otosclerosis and is more common than clinical otosclerosis (Guild, 1944).

Many theories of the aetiology of otosclerosis have been proposed and these include metabolic disorders, vascular disease, infection, trauma, and anatomical and histological anomalies of the temporal bone. Although histological studies of focal changes in otosclerosis have shown features in common with bone dystrophies, such as Paget's disease and osteogenesis imperfect, studies of the mineral content of the temporal bone and ossicles have shown variable results. Jense, Neilsen and Elbrond (1979) reported a study of the mineral content of skeletal bone in patients with and without otosclerosis and showed no difference between the two groups. This work supports the assumption that otosclerosis is a localized disease of the labyrinthine capsule.

In 1944, Guild examined 1161 unselected autopsy specimens. Microscopic sites of otosclerotic bone were found in 6.1% of white males, 10.3% of white females, 1.0% of Negro males and 0.5% of Negro females. Bony ankylosis of the stapes was seen in six white and two Negro subjects. There is thus a very marked difference between clinical and non-clinical otosclerosis.

**Incidence**

Guild (1944) first pointed out the importance of making a distinction between clinical and non-clinical, or histological otosclerosis; the latter is about 10 times more common than the former. The results of other workers (Weber, 1935; Engstrom, 1940; Soifer, Weaver and Holdsworth, 1970) supported those of Guild giving an incidence of histological otosclerosis of up to 10% in white adults but much less in the African races. Friedmann (1974) has estimated that 2% of all white persons suffer from deafness caused by otosclerosis. Morrison (1967) studied the incidence of otosclerosis in a population of nearly one-quarter of a million persons in Outer London and showed the incidence to be approximately 2% in patients between the ages of 30 and 59 years of age (Table 14.1). Shambaugh (1949) in North America estimated the frequency of otosclerosis to be at least 0.5%.
Table 14.1 Incidence of otosclerosis 30 years of age and above

<table>
<thead>
<tr>
<th>Age group</th>
<th>Incidence per 1000 population</th>
<th>Age group</th>
<th>Incidence per 1000 population</th>
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<tr>
<td>30-34</td>
<td>2.15</td>
<td>60-64</td>
<td>1.38</td>
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<tr>
<td>35-39</td>
<td>1.94</td>
<td>65-69</td>
<td>0.99</td>
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<td>40-44</td>
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<td>0.53</td>
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<tr>
<td>55-59</td>
<td>1.29</td>
<td>30 and over</td>
<td>1.54</td>
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The variation in the figures produced by different authorities makes it obvious that the true incidence of the disease is not known. In recent years, it has become quite clear that the number of patients seen in clinics and operated on by stapedectomy has fallen dramatically and this is unlikely to be the result of patients failing to attend for treatment as the results of surgery are good and widely known to be highly successful. The most likely explanation is that the original pool of patients has been reduced by energetic otologists, keen to operate on those patients who came to them with a conductive deafness caused by otosclerosis. Hammond (1976) has shown the decline in the number of patients treated by operation at St Thomas’ Hospital, London, between the years 1962 and 1971. Beales (1979) conducted a survey of the number of stapedectomy operations performed in 10 major centres in the UK between 1957 and 1977, in a sample of over four million.

**Racial incidence**

Otosclerosis is most frequently encountered in the Caucasian races and is a common cause of deafness in Europe, the Balkans, the Middle East, and the subcontinent of India, together with the Caucasian peoples of North and South America, Australia, New Zealand, South Africa and elsewhere. It is rarely found in Mongoloid or Negroid man, although it is encountered in the Negro population of America and the West Indies, presumably as a consequence of hybridization. In the latter group, the disease is about 10 times less frequent than among Caucasians (Morrison, 1967).

The disease also occurs, less frequently, in the Negrito populations of Malaya, New Guinea and the Philippines, and in the Japanese, a mixed race of Mongul, Ainu and Negrito blood (Joseph and Fraser, 1964).

**Sex incidence**

In clinical practice, otosclerosis is seen more often in women than men, and a sex ratio of 2:1 has been noted by many authorities. However, the incidence is likely to be the same in both sexes, although hormonal influences may cause the disease to advance more rapidly in the female; it also seems that they are more likely to seek medical advice. Asymmetrical deafness caused by otosclerosis is more common in the male and those working in a noisy environment are less likely to be aware of this because of the phenomenon of paracusis.
Genetic factors

Although Toynbee (1861) noticed the familial pattern of deafness and ear disease, it was not until nearly a century later that a comprehensive investigation into the genetic of otosclerosis was conducted by Larsson (1960, 1962). Larsson carried out a field investigation into 357 otosclerotic families between 1956 and 1958. His analysis pointed to a simple autosomal dominant inheritance with incomplete ‘penetrance’ or manifestation.

Morrison conducted a most detailed and comprehensive survey between 1961 and 1964. In this, the clinical diagnosis was based on a conductive deafness confirmed by audiometry, in the presence of a normal tympanic membrane. In 19 patients, otosclerosis was diagnosed on clinical grounds alone, and in the other 131 it was confirmed by stapedectomy, with histological proof in 109 operations (the specimens being inadequate in the remainder). He confirmed Larsson's result that the disease has an autosomal dominant inheritance. He also found that it occurred in about 40% of individuals carrying abnormal genes; 30% of cases were isolated and this was though to result from failure of manifestations in other family members; some sporadic cases were the consequence of new mutations. Chromosome studies and analyses in otosclerosis and in osteogenesis imperfecta demonstrated normal caryotypes. Morrison concluded that otosclerosis, like osteogenesis imperfecta, belonged to a group of hereditary disorders of collagen, with a similar mode of inheritance, incomplete manifestation, varying degrees of expressivity, and possibly an abnormal enzyme system.

Age of onset of hearing impairment

The disease generally starts in the ’teens and is uncommon before the age of 5 years. The rate of progression varies and in the majority of patients clinical evidence of stapes fixation is not seen. Soifer, Weaver and Holdsworth (1970) showed histological evidence of otosclerosis in 8.3% of 601 temporal bones, but the incidence of stapedial fixation amounted to only 0.09%.

The rate of progression is variable and there are periods of extension alternating with quiescent phases in some patients, while in others the progress of the disease continues relentlessly and, in a small proportion, may be rapid. During periods of endocrine activity, such as puberty, pregnancy and the menopause, the disease may progress; in the majority of patients, signs of hearing loss are complained of between the ages of 20 and 30 years, but sometimes not until 50 years or even later.

The effect of prolonged exposure to industrial noise on patients with otosclerosis

Alberti, Hyde and Symons (1980) reported on the results of an investigation of 135 cases of otosclerosis among men referred for compensation for industrial deafness, all of whom had been exposed to high level industrial noise for a prolonged period of time. When compared to cases without otosclerosis, following similar exposure, both groups showed the typical audiogram of high frequency sensorineural hearing loss associated with acoustic trauma. Most otologists who are experienced in examining patients with industrial deafness will have noticed that the bone conduction audiograms of patients with a marked conductive loss probably caused by otosclerosis, show a high frequency sensorineural deafness which is
typical of noise-induced hearing loss. The explanation of this phenomenon is that it is likely that the person was exposed to damaging levels and gave rise to the sensorineural deafness before the conductive loss developed. The conductive loss developed later and so the audiometric pattern is the combined result of the otosclerosis and the noise deafness present when the person was younger. As otosclerosis may also give rise to sensorineural deafness it can be difficult to arrive at a definite conclusion from the medicolegal point of view.

It is conceivable that a marked conductive deafness does not protect the individual from the damaging effects of a high level of industrial noise, although it is also possible that otosclerosis may make the cochlea of the affected person more sensitive to the damaging effects of noise in the early stages of the disease with minimal hearing loss.

**Otosclerosis and pregnancy**

In 1967, Shambaugh studied 475 mothers who had received surgical treatment by the fenestration operation. He found that 50% had not observed any noticeable effect on their hearing from any of the pregnancies but, in 42%, there was an associated increase in hearing loss. He estimated that the risk of increased hearing loss from any one pregnancy in a woman with stapedial otosclerosis to be about one in 24. Elbrond and Jensen (1979) studied the influence of pregnancy on the hearing threshold, before and after stapedectomy and found that the operation afforded some protection from further hearing loss.

Gristwood and Venables (1975) studied 479 women who were deaf from otosclerosis. They found that in bilateral cases, pregnancy aggravated the deafness and this incidence ranged from 33% after one, to 63% after six, pregnancies. In unilateral cases, it was found that the pregnancy-related deterioration of hearing was much less common.

**Site of otosclerosis in the temporal bone**

Although any part of the bony labyrinth may be affected by otosclerosis, the most common site is between the anterior part of the stapedial footplate, the cochleariform process and the bulge of the promontory.

The focus extends, infiltrates and fixes the footplate in the oval window niche and eventually firm bony ankylosis results. It is more common for the new bone to affect the anterior part of the footplate leaving the centre free. About 85% of lesions are situated in the oval window area.

The second most common site for the focus is in the region of the round window, and here, evidence of otosclerosis has been found in up to 50% of cases. In the majority of cases (70-80%) both temporal bones are affected, and a characteristic feature of the disease is the striking similarity of the localization and extent of the lesion in each ear and the similar development of the process.

Other areas of predilection are the round window, the anterior wall of the internal auditory meatus, and within the stapedial footplate.
Unilateral otosclerosis

Although both temporal bones are usually affected, unilateral otosclerosis has been described by various authorities as occurring in 10-15% of patients with this disease (Nager, 1939; Cawthorne, 1955).

Histopathology of otosclerosis

In the otosclerotic focus in the temporal bone, there are two types of bone - woven and lamellar - and two types of cell - osteoblasts and osteoclasts. The first bone to appear during the course of embryonic development, and all early membrane bone, is called 'woven bone' because it contains an interlaced tangle of calcifiable fibres. Lamellar bone, on the other hand, consists of fibres in bundles which lie parallel to one another in the intercellular substance and it is formed, like woven bone, by osteoblasts. The mode of action of osteoclasts is not understood; they produce proteolytic enzymes in tissue culture, may actively phagocytose particles of bone and are constantly seen where bone resorption is taking place (Friedmann, 1974).

Stages of the otosclerotic lesion

Although it is usual, on light microscopy, to describe several stages seen, there is no orderly progression from one to another and it is a feature of the otosclerotic lesion that one focus may contain areas at different stages of activity. The stages may be described as active, intermediate, and inactive (final stage). The term otosclerosis refers to the final stage consisting of highly mineralized bone with a mosaic appearance. The osteoclasts have disappeared but osteocytes and/or, osteoblasts may still be seen. The vascular spaces are narrowed, or obliterated, by new bone formation and the lamellar bone which is formed is thicker and more cellular when compared to normal bone.

The term 'otospongiosis' refers to the active phase of the disease and the characteristic feature is the presence of vascular spaces containing some fibrous tissue, and osteoclasts and osteoblasts, forming loosely knit bone. The new bone is called 'woven' because the collagen fibres run in an irregular criss-cross pattern through the otosclerotic focus. A clearly defined boundary between the normal and abnormal bone is a feature, or there may be finger-like processes, which are resorption spaces produced by osteoclasts dissolving some of the perivascular bone. The spaces become filled with osteoclastic bone which stains with haemotoxylin and these areas are sometimes referred to as 'blue mantles'. These are also seen in chronic mastoiditis and other bone diseases (Friedmann, 1974), but Weber (1935 believed that the 'blue bone' was abnormal and otosclerotic foci might be formed through their fusion.

As has been stated previously, there is no orderly development from the active to the inactive stage and all stages may be seen together in one focus which may become quiescent or reactivated at any time.
Clinical features

Deafness

The typical features of otosclerotic deafness are a bilateral, gradually increasing hearing loss, most frequently occurring between the third and fifth decade, the presence of paracusis and tinnitus.

The deafness is often unnoticed by the patient, until the loss reaches 25-30 dB, when difficulty in understanding speech becomes apparent. The patient may remark that the hearing is better in the presence of background noise. The phenomenon of paracusis Willissi is frequently present if there is a predominantly conductive deafness without a sensorineural loss. One explanation of this is that, in general, people with normal hearing raise their voices above the noise level, so that they can remove the masking effects of the noise, and this level of speech sound is above the threshold of the patient with conductive deafness. Although paracusis is seen in other forms of conductive deafness it is most often seen in stapedial fixation caused by otosclerosis.

The patient with otosclerotic deafness has a characteristically quiet voice, which is of good tone, and the change in the speech pattern may be detected by close relatives who often notice the hearing loss before the patient becomes aware of it. The deafness is generally progressive, occurring in a direct linear form; alternatively there may be a plateau-like period, or both features may occur together.

The hearing loss may be almost equal in each ear but often one ear shows a greater loss, and this ratio is usually maintained. Unilateral otosclerosis occurs in approximately 15% of patients. The deafness can remain confined to one ear, or the second ear may become affected later. Many patients with a pure conductive loss will, in later years, develop a sensorineural deafness that is greater than that to be expected as the result of ageing.

Tinnitus

Tinnitus is a common symptom and occasionally the presenting feature. It is sometimes seen in patients without cochlear degeneration when it is the result of an abnormal degree of vascularity of the otosclerotic bone; more often, tinnitus is an indication of sensorineural degeneration.

The tinnitus may be unilateral or bilateral, and of a roaring, hissing or pulsatile character. Fluctuation of the tinnitus is not uncommon and this can be related to metabolic and endocrine disturbances, pregnancy or menstruation. It is more common in the early stages of the disease and it may disappear as the lesion matures and the spongy vascular bone is replaced by the hard sclerotic bone.

Vertigo

Attacks of vertigo, usually of a transient nature, are not uncommon, and they are probably the result of the action of toxic enzymes, which are liberated by the lesion, on the vestibular labyrinth (Causse et al, 1977). A variety of vestibular lesions have been described
in otosclerosis and the most common is true benign positional nystagmus (Colman, 1979). If vertigo is a prominent symptom, the coexistence of Ménière's disease must be considered, as both disorders are common and will be seen from time to time in the same patient.

When taking a history, former episodes of ear disease, head injury, exposure to noise, administration of ototoxic drugs, bone or joint disease must be enquired into, as these factors may be of importance in diagnosis and management.

Morrison (1979) has called attention to the importance of a detailed family history in the assessment of the prognosis when other members of the family are affected.

**Diagnosis**

**Examination**

The examination of the tympanic membranes will include inspection at rest, testing with a Siegle's speculum for mobility and examination after inflation of the eustachian tubes by a Valsalva manoeuvre. The tympanic membranes in otosclerotic patients are sometimes described as being in 'mint condition' but they may be atrophic, thickened, rigid or mobile, and the response to tubal inflation may vary. The 'flamingo blush', or Schwartz sign, is uncommon. It is a result of vascular bone on the promontory, or prominent blood vessels in the submucosal layer of the mucous membrane of the promontory. When seen, it indicates active disease which can progress rapidly.

In every case, examination of the nose, nasopharynx and nasal accessory sinuses is necessary to exclude infection, which may need treatment.

**Clinical assessment of hearing loss**

A rapid estimation of the hearing loss is made in the clinic by simple speech tests, using conversational and whispered voice, the effect of a hearing aid and tuning fork tests. Patients with otosclerosis do not usually show recruitment and are able to hear amplified sounds clearly, unless there is a marked sensorineural deafness.

The tuning fork tests, which will include the Rinne, Weber and Schwabach tests, must be carried out before the more complex tests of auditory function are performed. They are of particular value in two instances:

1. When there is a predominantly unilateral otosclerosis, with poor bone conduction simulating sensorineural deafness. Here the Weber test will show lateralization to the deaf side.

2. When there is a severe combined (stapedial and cochlear) otosclerosis where the bone conduction cannot be recorded as there is more than 60 dB hearing loss (which is beyond the limit for the majority of clinical audiometers). If the Rinne test is negative with 256 fork and the patient's voice has reasonable quality, good results can still nevertheless be obtained from operation (Morrison, 1979).
Audiometry

Bone

Audiometric tests are the most important of the testing methods used in otosclerosis. While the air conduction curve gives an indication of the hearing threshold, and its configuration may give a clue to the diagnosis of otosclerosis in the early stage (Carhart, 1964), bone conduction audiometry is of special value in diagnosis and in the selection of patients for surgical treatment. There are certain difficulties in pure tone audiometry and it is important to realize these problems or false information may result.

Carhart, in 1950, pointed out that bone conduction audiometry revealed distinctive curves in patients with stapes fixation caused by any lesion, congenital or acquired, that interferes with the mobility of the stapes. Anything which reduces the inertia of the movement of the stapes footplate, for example an ossicular discontinuity, can also produce this effect. This in turn depends on the fact that one element of bone conduction is the inertia caused by the weight of the ossicular chain, whereby the stapes footplate vibrates out of phase with the skull as a whole, as the latter is set into vibration by the tuning fork or bone conductor. Thus when the footplate is fixed, it is no longer free to vibrate and so the inertial component of bone conduction is lost. If the main mass of the ossicular chain, the malleus and incus, is disconnected from the stapes it also loses most of the inertial component of bone conduction (it can also occur in chronic otitis media).

This effect is most noticeable in otosclerosis, and reductions in sensitivity described by Carhart are: 5-10 dB at 500 Hz, 10-20 dB at 1 kHz, 15-30 dB at 2 kHz and 5-20 dB at 4 kHz. The maximum reduction in sensitivity is most commonly at 2 kHz, although this is not always so. This shift in sensitivity is known as the 'Carhart's notch'.

The Carhart notch effect may disappear after stapedectomy and this phenomenon is sometimes called 'over-closure of the air-bone gap'. It means that after surgery, only the air conduction thresholds have been measured and then compared with the preoperative bone conduction thresholds; it is important therefore to re-test the bone conduction after surgery.

The otologist can, by correcting the bone conduction audiogram for the Carhart effect, determine with accuracy the degree of sensorineural reserve that a patient with otosclerosis possesses. There is, however, a variation in the mechanical shift in bone conduction response which differs from one patient to another. The use of average figures for correction may be inaccurate and so it is important to have some knowledge of the possible variations.

Variability of the Carhart notch

There are two ways in which an estimate can be made as to whether the notching in the bone conduction audiogram is unusual:

(1) if the bone conduction curve shows an irregularity, or peculiarity, which the air conduction curve duplicates, this should be attributed to sensorineural impairment
(2) if the air conduction curve does not show the peculiarity, and the audiogram shows an unusual modification of the bone conduction response.

Gibb and Mal (1973) carried out an investigation to see if the Carhart notch is abolished after successful stapedectomy surgery; they found that in the majority of cases bone conduction thresholds were improved after operation, especially at 2 and 4 kHz. Their paper made it clear that a correction figure for the Carhart notch was important and failure to make an adjustment might result in borderline cases, suitable for operation, being excluded. They also found that the bone conduction shift, in their series, was much less than Carhart's original estimate.

**Limitations of bone conduction audiometry**

Even when bone conduction audiometry is carefully performed it possesses three defects which can cause serious error in threshold measurement: false lateralization; hyperdistractibility; and shadow response.

**False lateralization**

This error is common when bone conduction audiometry is performed without masking; a 'Weber-type' effect is produced and the bone conduction threshold on the worse side appears to be the better ear (the patient is hearing the sound in the other ear).

**Hyperdistractibility**

In some cases, the masking noise interferes with the response of the ear under test; such patients are hard to test accurately.

**Shadow response**

In some patients, the hearing losses are so great that efforts to mask the contralateral ear are ineffective. This situation arises because effective masking is reduced by the amount of the air conduction loss in the masked ear at that frequency. In the majority of cases good bone conduction audiometry is valid, but incorrect results will be obtained in some patients unless the difficulties which have been outlined are appreciated.

**Objective audiometry**

The application of the concepts of acoustic impedance to clinical audiology dates from 1946, when Metz published his monograph.

The investigations carried out are: tympanometry, acoustic impedance and the measurement of the acoustic reflex threshold.

**Tympanometry in otosclerosis**

If the compliance is greater than 0.6 cm$^3$, it is probable that the footplate of the stapes will be relatively thin and, if the compliance is less than 0.2 cm$^3$, there is a likelihood that
the footplate may be thick or obliterated; if the loss is symmetrical, the information may be helpful in selecting the more suitable ear for surgery (Morrison, 1979).

Browning, Swan and Gatehouse (1985) doubted the value of tympanometry in the diagnosis of otosclerosis and found that measuring compliance is of little help because the results overlap those of the normal range. They believed that testing for the absence of the acoustic reflex is helpful in confirming the presence of a conductive defect when the results of pure tone audiometry are equivocal.

In general, it is true to state that impedance audiometry is valuable in differential diagnosis, when taken in conjunction with other tests.

**Speech audiometry**

As pure tone audiometry does not predict the ability of the deaf person to hear speech sounds clearly, such tests must be included among the other tests of auditory function.

These tests are described in detail in Volume 2. The investigations usually ordered by the clinician are: the speech reception test, the speech discrimination test, and the speech audiogram.

**Radiological examination** *(see Chapter 2)*

**Plain X-ray examination**

Although otosclerotic involvement of the temporal bone cannot be demonstrated on plain X-ray films, standard views of the mastoid and Stenvers' view of the internal auditory meatus should be carried out for the purpose of differential diagnosis.

Plain X-rays may reveal a sclerotic mastoid, indicating the possibility of past infection, cholesteatoma, an unsuspected acoustic neuroma, or a skull lesion such as Paget's disease. A meningioma may also be detected. These conditions can give rise to a conductive hearing loss, which is not caused by otosclerosis and without X-ray examination they may not be diagnosed.

**Linear tomography**

Linear tomography is available in most radiological departments and is adequate for many purposes, but it does not show sufficient detail of the otic capsule.

**Polytomography**

Polytomography can show details of the auditory ossicles, the oval window and the capsule.

Three variations of capsular otosclerosis can be recognized:

(1) limited changes in the basal coil
(2) diffuse involvement of the cochlear capsule
(3) widespread labyrinthine otosclerosis.

Areas of radiolucency, indicating demineralization of the otosclerotic bone, may be
seen side by side with sclerotic changes. Naunton and Valvassori (1969) have been able to
correrate the bone conduction levels and the evidence of capsular otosclerosis in 74.3% of
patients with combined stapedial and cochlear otosclerosis; Gungovich and Rosenfeld (1974)
have confirmed their work.

Recalcification of a focus may result in a normal radiographic appearance
(Shambaugh, 1971) and this explains the not uncommon finding of normal X-rays in patients
with combined stapedial and cochlear otosclerosis. Positive evidence of radiological changes
is helpful, but their absence does not exclude otosclerosis.

**Differential diagnosis**

The diagnosis of otosclerosis is usually straightforward and is made on the history of
a bilateral hearing loss, an intact mobile tympanic membrane and evidence of conductive
deafness. Unilateral otosclerosis is seen in about 15% of patients and when it does occur, the
possibility that the conductive hearing loss is the result of another cause must be considered.
Otosclerosis can give rise to sensorineural deafness and this must be considered in the
differential diagnosis.

**Middle ear lesions**

**Secretory otitis media**

This is a common condition giving rise to a conductive hearing loss that may persist
for many years. Tinnitus and vertigo are absent and careful examination of the tympanic
membrane shows loss of translucency, and fluid may, or may not, be seen. The tympanogram
is usually typical in this condition and a plain X-ray of the mastoids will show haziness or
increased density. In adults secretory otitis media is often unilateral and may be associated
with a lesion in the postnasal space.

**Middle ear fibrosis (chronic adhesive process)**

In this condition the tympanic membrane may appear normal, but it is usually
retracted, or thickened, and shows lack of mobility. A tympanogram and mastoid X-rays will
establish the diagnosis.

**Tympanosclerosis**

Although tympanosclerosis can occur in the middle ear, with minor changes in the
tympanic membrane, this is not usual and the drum is generally opaque with white patches.
The condition is common and it may result in a severe conductive deafness if the deposits
immobilize the stapes, or the malleus and incus. Tympanometry will show reduction of
mobility of the tympanic membrane and an X-ray may show lack of translucency of the
mastoid air cells.
**Fibro-osseous footplate fixation**

In this condition, the history of previous otitis media is helpful and the tympanic membrane will usually show evidence of past inflammation, but as otosclerosis is not uncommon in conjunction with past otitis media, it is not always possible to diagnose this condition without tympanotomy.

Mobilization, or stapedectomy, may give good results provided there is no associated fixation of the malleus and incus.

**Congenital footplate fixation**

It is important to recognize this condition as stapedectomy carries a risk of perilymph flooding and a sensorineural hearing loss. The deafness is not progressive but the audiogram with a Carhart notch may be identical. If the condition is bilateral, the speech will be affected, but the unilateral cases are usually diagnosed in late childhood or early adult life.

Congenital fixation of the malleus and incus occurs, generally associated with a developmental aplasia of the ear with meatal atresia, but it is occasionally seen without meatal atresia. The condition is rare.

**Ossicular discontinuity**

Traumatic dislocation of the incus is seen after injuries. It is not uncommon as a result of road traffic accidents where the injuries are severe with loss of consciousness, and bleeding from the ear and not infrequently facial palsy. Less commonly, traumatic dislocation of the incus also occurs after minor head injuries. A blow on the ear, or an unskillful attempt to remove a foreign body when it is pushed through the tympanic membrane, may also lead to dislocation (see also Chapter 7).

Tympanometry is of importance in the diagnosis of these lesions and it will show an absent stapedial reflex, an abnormal compliance and low impedance system.

Polytomography may demonstrate ossicular chain abnormalities, but radiography cannot be relied upon if there is only a very small gap, even though the hearing loss may be profound.

Tympanotomy will often be required to make the exact diagnosis, but it is essential for the otologist to recognize the presence of these lesions, as in most cases stapedectomy is not indicated and the deafness will be treated by some form of ossiculoplasty, or occasionally the use of tissue glue. It should also be remembered that injury can also produce ossicular fixation.
Malleus and incus lesions

The fixed malleus - incus syndrome

This lesion was described by Goodhill in 1960, and Morrison (1979) found it in 2% of tympanotomies.

In this condition, there is stiffness, or fixation, of the malleus, incus, or both, but the stapes is not immediately involved. This lesion may be missed if the surgeon who has made a diagnosis of otosclerosis proceeds to remove the stapes without a preliminary testing of the mobility of the malleus and incus. Following the stapedectomy operation, there will be no improvement in hearing and this is attributed to a failure of the stapedectomy.

Impedance audiometry will help to distinguish this lesion and if the mobility of the ossicular chain is tested at every operation, a wrong surgical procedure will be avoided.

Osteoarthritis of the ossicular chain

The entire ossicular chain may be involved by an osteoarthritic condition and when this occurs it may be difficult to distinguish from otosclerosis. It is more commonly unilateral and tympanometry will show a negative stapedial reflex with high total impedance.

Congenital cholesteatoma

A primary cholesteatoma may occur behind an intact tympanic membrane, producing a conductive deafness. Careful examination of the drum with magnification will usually reveal this condition before operation. Tympanotomy and removal of the cholesteatoma with, if necessary, ossicular chain reconstruction will be required.

Fluid in the middle ear: cerebrospinal fluid or perilymph

The most common cause of fluid in the middle ear is secretory otitis media and this has already been described.

A patient at operation for otosclerosis may show the presence of clear fluid in the middle ear and this fills up again after aspiration. The fluid may be cerebrospinal fluid, the result of a fracture of the tegmen tympani caused by a previous head injury, or it could be perilymph which is leaking from the scala vestibuli when there has been a fracture or dislocation of the stapes. Rupture of the round window membrane usually heals spontaneously but this does not always occur and perilymph can leak from an unsuspected fistula.

Degenerative footplate arthritis: crural atrophy

In this condition, which was described by Goodhill in 1979, there is a circumferential ossification of the annular ligament of the stapes, or crural atrophy resulting from latent chronic otitis media. Crural atrophy may be distinguished before operation by tympanometry, which will show the presence of a stapedial reflex and abnormal tympanic membrane.
compliance. Degenerative footplate arthritis cannot be distinguished before operation, but treatment by stapedectomy will give good results.

**Persistent stapedial artery**

A persistent stapedial artery is occasionally seen at operation and stapedectomy may be possible if the whole footplate is not covered. Rarely, a large artery may cover the footplate and fix the stapes, giving rise to a conductive deafness without footplate fixation. Operation is contraindicated in the presence of this condition.

**Paget's disease (osteitis deformans)**

Woodhouse (1973) estimated that about 750,000 people have this disease in the UK and that the temporal bone is involved in about 50% of those with clinical evidence of the disease. Paget's disease rarely gives rise to conductive deafness but, if it occurs, it usually starts after the age of 45 years. In the early stages, there is a conductive type of loss associated with a high tone sensorineural loss (see Chapter 15).

The cause of the conductive deafness is ossicular fixation, mainly of the malleus head. As there is no histological evidence of stapedial fixation, the operation of stapedectomy is not logical, although ossicular mobilization may cause temporary improvement in hearing for a few years before the sensorineural deafness neutralizes the hearing gain.

**Osteogenesis imperfecta**  
*(fragilitas osseum, van der Hoeve's and de Kleyn's syndromes)*

This is a rare disease belonging to a group of hereditary disorders of collagen. The otological features of osteogenesis imperfecta are a progressive conductive and sensorineural deafness, the absence of a Schwartz sign and the absence of vertigo. Tympanometry shows absence of the acoustic reflex with a very high compliance value.

The deafness of osteogenesis imperfecta may be treated by stapedectomy but operation must be delayed until all spontaneous fractures have ceased. A high incidence of floating footplate has been found at operation and increased likelihood of sensorineural deafness occurs after surgery for this condition.

**Sensorineural deafness**

Although pure cochlear otosclerosis is now recognized as a clinical entity by an increasing number of otologists, there are many who remain sceptical about it and the incidence of this manifestation is unknown, and diagnosis is difficult (Schuknecht, 1983).

Progressive sensorineural hearing loss may be the result of one of the rare classified dominant inner ear diseases such as Alport's syndrome, or Norrie's disease, and deafness in retinitis pigmentosa may present in adolescence or early adult life, but in a large number, no definitive diagnosis can be made.
In the present state of our knowledge, it is not possible to exclude otosclerosis as a cause of the sensorineural deafness in some of the patients in this unclassified group.

Otosclerosis can lead to a very severe hearing loss and it must be differentiated from late syphilis of the temporal bone, Ménière's disease and unrecognized ototoxicity.

**Treatment of otosclerosis**

The majority of patients with otosclerotic deafness can be helped by surgical or nonsurgical methods and with the improvements in the design and construction of hearing aids, there are few that cannot be given some help.

**Medical treatment**

**The place of fluoride treatment**

In 1964, Shambaugh and Scott suggested that sodium fluoride, in moderate doses, might promote recalcification and reduce bone remodelling in an actively expanding otosclerotic lesion. In the human subject, fluoride is most effective on the active focus and less so on the mature lesion.

Otosclerotic foci show a tendency to be more active in young persons and less active in older people, although all stages can be found at any age. It should be appreciated that a mature focus can become active again and this may be the result of hormonal activity such as pregnancy, puberty or the menopause. The natural tendency for the active lesion in otosclerosis to become recalcified is inconstant and may be feeble. Shambaugh believed that sodium fluoride, in moderate doses, assists this natural tendency of the focus to become recalcified and inactive, and the evidence in favour of this is given as: fading of the injection of the mucous membrane over an active focus (Schwartze sign); stabilization of the progressive sensorineural deafness which is so often found in otosclerosis; reduction of tinnitus; improvement of mild vestibular symptoms; and X-ray demonstration of recalcification of the focus. Bretlau et al (1985) reported on an experimental and clinical evaluation of sodium fluoride treatment. The results showed that using the calcium/phosphorus ratio as an indication for bone maturity, sodium fluoride could stabilize otospongiotic lesions in retaining calcium relative to phosphorus. The results supported the view that sodium fluoride can change otospongiotic, active lesions to more dense, inactive otosclerotic lesions.

Fluoride is a trace element found in widely varying concentrations of ground water, that is between 0.1 and 16 parts per million. Some local authorities add fluoride to the drinking water to bring the concentration to one part per million and this has proved to be very beneficial in the prevention of dental caries in school children. Bernstein, Sadowsky and Hagstead (1966) studied the incidence of osteoporosis in rural communities in North Dakota, where the farming population remained in the area for a lifetime, and it was found that where there was an abnormally low fluoride content in the drinking water, osteoporosis was four times more common than in areas with a high content. A similar study of otosclerosis by Daniel (1969) showed that stapedial fixation was four times as high in the low area compared with that in the high fluoride area.
Gristwood (1966a), in Australia, reported on the unusually high incidence of the truly obliterated footplate, and it has been the experience of most Australian otologists, that the incidence of the thick and obliterated footplate is in the region of 30%. Gristwood and Venables (1975) have pointed out that the surface water in the most densely populates areas of South Australia is deficient in fluoride ions and it was not until 1971 that fluoridation of metropolitan water supplies was commenced.

**Action of fluoride**

Fluoride reduces osteoclastic bone resorption and increases osteoblastic bone formation. The work of Causse and Chevance (1973) suggested, in addition, that in otosclerosis fluorides have an antienzymatic action on proteolytic enzymes which are cytotoxic to the cochlea and produce sensorineural deafness. In a series of over 4000 patients treated with fluorides, in Chicago, USA and Béziers, France, very few have experienced improvement of the sensorineural element of their deafness, but it has become stabilized in over 80%.

**Indications for sodium fluoride therapy**

Sodium fluoride therapy is indicated in the following groups of patients (Shambaugh and Scott, 1964):

1. patients with surgically confirmed otosclerosis who show progressive sensorineural deafness disproportionate to age

2. patients with pure sensorineural deafness whose family history, age of onset, audiometric pattern and good auditory discrimination indicate the possibility of cochlear otosclerosis

3. patients with radiological demonstration by polytomography of spongiotic changes in the cochlear capsule

4. patients with a positive Schwartz sign.

**Preoperative treatment**

When the patient has an otosclerotic focus which shows activity, as evidenced by a positive Schwartz sign, progressive sensorineural hearing loss, and radiological evidence of a demineralized focus in the cochlear capsule, both Shambaugh and Causse believed that a substantial reduction in vascularity and remodelling of the focus will result from fluoride treatment.

**Postoperative treatment**

If patients are found to have an active focus at operation, fluoride therapy is prescribed for 2 years or longer (Cause and Causse, 1979).
Contraindications to sodium fluoride therapy

Sodium fluoride therapy is contraindicated in the following groups of patients:

1. patients with chronic nephritis with nitrogen retention
2. patients with chronic rheumatoid arthritis
3. patients who are pregnant or lactating
4. in children before full skeletal growth has been completed
5. patients who show an allergy, as demonstrated by an itching rash
6. patients with skeletal fluorosis. This is a rare condition seen in certain areas of India.

Dosage and administration of sodium fluoride

When there is evidence of an active lesion, a daily dose of sodium fluoride of 50 mg is given for 2 years and this can be increased to 75 mg daily in very active cases with a positive Schwartze sign. When there is evidence of stabilization of hearing, fading of the Schwartze sign, and radiological signs of recalcification of the focus, a daily maintenance dose of 25 mg is given for the rest of the patient's life. In the UK, enteric coated capsules of 20 mg are available and may be supplemented with calcium and vitamin D (BPC).

Adverse effects of sodium fluoride therapy

Gastric disturbance is the most common side-effect which is largely prevented by taking enteric coated capsules of sodium fluoride after meals. Patients with a peptic ulcer may complain of a flare up of their symptoms and the treatment must be stopped. An increase of joint symptoms may occur in those with chronic arthritis. A return to the previous state is rapid after cessation of treatment.

There is the remove possibility of skeletal fluorosis being produced and a skeletal survey should be made at the beginning of treatment, and repeated at intervals.

There is still a widespread prejudice and almost an emotional dislike of fluoride therapy by many members of the medical profession, which is not justified and is the result of ignorance about the facts of this form of treatment. At the present time, fluoride therapy is the only known method of promoting recalcification and inactivation of an actively expanding focus of otosclerosis. There is also evidence that sensorineural deafness may be stabilized or even improved in patients who receive fluoride medication.

Hearing aids

The modern transistorized hearing aid with an air conduction receiver gives good results in the great majority of patients with otosclerotic deafness and a bone conduction aid
is rarely prescribed, but may be of value in those with bilateral fenestration cavities. Auditory training and rehabilitation are helpful and those with poor discrimination and severe hearing loss should be advised to have instruction in lip reading.

Many patients prefer natural hearing to the use of a hearing aid and there is evidence that stapedectomy may reduce the rate of cochlear dysfunction which affects all patients with otosclerosis and although surgery is the best method of treatment for otosclerosis if it is successful, there is a high price to pay in the event of failure.

**Surgical treatment**

**Historical**

It is interesting to remember that although the first attempt at mobilization of the stapes was carried out over 100 years ago, it was not until 1958 that John Shea, in Memphis, described the operation of stapedectomy.

The first operation of stapedectomy was carried out by Jack of Boston, Massachusetts, in 1891. He undertook an operation for the removal of the drumhead, malleus and incus, as advocated by Kessel in 1878 for cases of chronic sepsis. During the operation, he found the stapes to be carious and removed this also. In the morning after the operation, the patient informed him that she heard sounds 'never heard before'.

It was unfortunate that, in other hands, stapedectomy proved to be dangerous and was strongly condemned by leading authorities of the time, including Politzer, who have his enormous authority to cautioning against such a procedure and it was not until 1958 that John Shea had the courage to try again and established the modern operation of stapedectomy on a proper footing.

The next stage in the surgical treatment of otosclerosis was concerned with the establishment of an indirect method that allowed the inner ear fluids to move again, under the influence of sound stimuli. The fenestration operation started around 1914 by Jenkins, was developed by Holmgren (1923), Barany (1924) and Sourdille (1930) and later by Lempert (1938); it lasted until 1952 when the operation of mobilization of the stapes was described by Rosen in New York. Early results were good but refixation was common and stapedectomy replaced the mobilization operation. In 1958, Shea introduced the modern operation of stapedectomy which is the basis of all the operations which have been developed since that time. Shea's contribution to the surgery of otosclerosis was monumental and he is rightly regarded as the originator of modern surgery for otosclerosis.

**Indications for surgery**

The majority of patients with a conductive deafness caused by otosclerosis can be treated by stapedectomy and, in general, a patient who will benefit from an operation will also hear satisfactorily with a hearing aid.

The average patient with otosclerosis and a bone conduction level of 0-25 db in the speech range, and an air conduction of 45-65 db, is a suitable candidate for surgery (Goodhill,
The air bone gap should be at least 15 dB and there should be a speech discrimination score of 60% or more for a good hearing improvement.

In the era of fenestration surgery, patients with very severe hearing losses were not suitable for operative treatment, but with the advent of stapedectomy this is no longer the case and patients with hearing losses in the 90-100 dB range and no measurable cochlear reserve on speech discrimination, may still be suitable for operative treatment to enable them to use a hearing aid which was previously of no help. This last group of patients, although small, is the only one where operative treatment is essential as there is no alternative method available. 'The ultimate aim is restoration of available cochlear function, even though this may not carry with it the possibility of unaided hearing' (Goodhill, 1979).

Contraindications to surgery

Morrison (1979) listed 16 contraindications to operation in otosclerosis:

1. The presence of general medical disease when the patient is unfit for surgery, or where the expectation of life is limited.

2. Old age; in those over the age of 70 years there is a 40% chance of discrimination becoming worse, and the risk of fistula formation is greater in the older age group. Unless there is some special reason for operation, a hearing aid should be advised.

3. Most surgeons would not advise operation in children, but Robinson (1983) and von Haacke (1985) have reported good results in young people between the ages of 16 and 21 years.

4. In conductive losses from other causes, the stapes should not be touched. This applies particularly to stapes fixation caused by tympanosclerosis as stapedectomy carries with it a high incidence of sensorineural loss.

5. If other conditions are present, such as otitis externa, or a perforation, stapedectomy is contraindicated until they have been successfully treated.

6. If there is early fixation with a small degree of hearing loss, operation is probably not necessary, although some surgeons might consider stapes mobilization in such cases.

7. In unilateral otosclerosis, surgery may not be necessary, but many patients find loss of binaural hearing a great handicap, and in these cases operation is justified.

8. If the patient has only one hearing ear, operation is not justified unless a hearing aid does not give relief.

9. In stapedial and cochlear otosclerosis, with a poor air-bone gap, operation is not advised if a hearing aid can be used.
(10) The presence of vertigo and clinical evidence of labyrinthine hydrops is a contraindication to operation as there is an increased risk of a 'dead ear' from damage to a distended saccule during operation.

(11) Morrison is of the opinion that revision stapedectomy is dangerous because fine adhesions may exist between the footplate area and the saccule or cochlear duct. If the small fenestra operation is carried out, this criticism does not apply and good results are possible in the hands of the expert.

(12) Second ear stapedectomy is still controversial because of the risk of immediate and delayed sensorineural hearing loss which, in rare cases, can be bilateral. Vestibular damage can occur with permanent loss of coordination. The advantages of bilateral stapedectomy, if it is successful, are the restoration of binaural hearing and the ability to localize the direction from which sound is coming. The majority of surgeons in North America who are specialist 'otosclerosis' surgeons carry out bilateral operations if the criteria are right, and in specialist clinics in Europe this is also the case (Causse and Causse, 1980). Many surgeons, and especially those who do not specialize in the surgical treatment of otosclerosis, feel that the patient should be allowed the safeguard of being able to wear a hearing aid, if necessary in the second ear and, in the UK, bilateral stapedectomy is becoming less common than in the past.

(13) In the young adult with a rapidly spreading stapedial and cochlear otosclerosis and a positive Schwartz sign, surgery should be delayed until the activity is controlled by fluorides.

(14) Stapedectomy is contraindicated in pregnancy, an operation should be delayed for 12 months after parturition.

(15) Stapedectomy may be inadvisable on those whose occupations demand considerable physical strain, in those engaged in sport and in airmen, especially those flying small unpressurized aircraft, as there is an increased risk of perilymph fistula.

(16) If there is evidence of poor eustachian tube function in one ear, detected by tympanometry, and there is bilateral otosclerosis, it is advisable to operate on the ear with normal atmospheric middle ear pressure rather than the poorer hearing ear.

Preoperative counselling of the patient

As there is an alternative method of treatment, a hearing aid, which in most cases is satisfactory, it is essential to explain to the patient the advantages and possible disadvantages of surgery. In a suitable case there is at least an 85% chance of obtaining a good hearing improvement. About 10% gain only slight improvement of hearing and the remaining 5% may expect some degree of sensorineural loss after operation, which may become total in perhaps 2% (Shea, 19850. It should be explained to the patient that the operated ear may fail after an initially good result, and this can occur many years later. Slight vertigo, for a few weeks after operation is common, and a transient weakness of the facial muscles can occur. If bilateral stapedectomy is carried out the possibility of alteration of taste from chorda tympani damage must be mentioned. The patient must be warned against violent nose blowing at all times, as
this can lead to a fistula. Flying is contraindicated for 2 weeks after surgery and strenuous exercise, or the lifting of heavy weights, must also be avoided for a similar period.

The surgeon who does not tell the patient of the possible risks of operative treatment is, today, likely to become involved in medicolegal problems.

**Surgical technique**

The technique of operation is fully described elsewhere (Schuknecht, 1971; Beales, 19810 and the figure shows the various types of operations performed today.

The surgeon will use the technique which he has found to be the most satisfactory. A prosthesis is most commonly used for the reconstruction of the ossicular chain and that made from Teflon is the most widely employed. Some surgeons use a stainless steel piston, while others, a wire prosthesis - the 'Schuknecht method'. A few surgeons avoid the use of a prosthesis by using the crura of the stapes which remain after the footplate has been removed or fenestrated. This latter method is not always technically possible and long-term results are not as satisfactory as those obtained by the use of a prosthesis.

In recent years, it has been shown that the risks of damage to the internal ear at operation and the long-term risk of perilymph fistula is less if a small fenestra is made in the posterior part of the footplate (Marquet, Creten and van Camp, 1972; Marquet, 1983, 1985).

**Small fenestra stapedectomy**

Marquet claimed that this method has the following advantages:

(1) The fine instruments used with this technique prevent the risk of tearing of the mucous membrane of the structures of the vestibule.

(2) The calibrated hole in the centre of the footplate, avoids rupture of the annular ligament, and so the vestibular endothelium is not disturbed and the contents of the vestibule are undamaged.

(3) The prosthesis does not penetrate more than 0.1 mm into the vestibule and this avoids the risk of tearing, or irritation, of the underlying membranous structures. The growth of the vestibular endothelium beneath the lower end of the prosthesis is rapid and is guided by the meniscus of the perilymph. The inner ear is sealed rapidly from the middle ear as an extremely small opening has been made which is closed by the end of the piston.

(4) The small curvature of the meniscus of perilymph prevents the entry of waste particles of bone, which in any case are very small.

(5) The piston is firmly held in position at both ends, no pendulum movement can take place and the tip remains in the centre of the footplate perpendicular to it.
Vein graft-Teflon piston interposition operation

This method has been used in the Causse clinic in Béziers, France, since 1962 and over 20,000 operations have been performed by this technique which is a combination of the vein graft stapedectomy and Teflon piston technique (Causse, 1964; Causse and Causse, 1984). The advantages claimed for this modification of the Shea Teflon piston operation are:

1. Immediate closure of the oval window fenestra with no risk of the Teflon piston being manipulated in the open window, or dropped into it, when attempts are made to place it in position.

2. The tip of the piston is protected by soft tissue and this is an additional safeguard for the underlying membranous structures in the vestibule.

3. The strong seal over the oval window fenestra leads to a reduced tendency for perilymph fistula.

4. A further advantage is that an immediate seal of the window is obtained making it possible to monitor the progress of the patient more accurately than with other methods which produce a fistula and rely on spontaneously healing for it to close. Postoperative audiograms in the latter case do not give useful information for some time, and so immediate postoperative monitoring of the patient is of little value.

Large fenestra stapedectomy: wire and fat graft

The Schuknecht operation is described in detail in the monograph *Stapedectomy* (Schuknecht, 1971). This has proved to be a satisfactory procedure and it is used by many surgeons today (McGee, 1969).

The advantages of the large fenestra operation are that it is relatively easy to carry out in most cases, an immediate seal of the oval window is obtained by the fat graft, and the use of a stainless steel wire prosthesis makes accurate placement of the graft possible.

The disadvantages of the procedure are that removal of the whole or greater part of the footplate is a more traumatic procedure, and that there is an increased risk of damage to the underlying membranous structures in the vestibule; the incus is left bare with a diminished blood supply and a greater risk of necrosis and if the wire is crimped too tightly, the terminal portion of the long process may be damaged.

Schuknecht has shown, by post-mortem studies, that after some operations the wire wanders to the edge of the oval window and conductive loss develops. The rare complication of a granuloma of the oval window can occur after this procedure.

Use of the argon laser in stapedectomy

The potential value of the laser in middle ear surgery is considerable, since there is elimination of mechanical trauma, tremendous accuracy and reduction of bleeding. The CO₂
laser, used in other branches of otolaryngology, cannot be used for operations on the middle ear as it gives rise to inner ear damage.

Perkins (1980) using a laser focal spot size of 50-100 microns, makes multiple small holes in a rosetta fashion around the perimeter of the desired aperture and the central bone is removed with special superfine 45° pick. An aperture slightly larger than 0.6 mm is made in the footplate to accommodate the wire piston assembly, covered with a thin graft of autologous vein. If no vein is used, there is less difficulty in placing the prosthesis into the fenestra. McGee (1983) and Portmann (1983) have reported their results with the argon laser in stapedectomy.

This new method makes it possible to create a small fenestra without significantly fracturing the footplate and it should eliminate the problem of the floating footplate.

Problems found at operation

Although many stapedectomy operations are straightforward and present little technical difficulty, anatomical and pathological variations can be encountered which may present great difficulty, and while the experienced surgeon may be able to overcome them with a successful result, the less experienced may be wiser to abandon the operation.

The most common abnormalities are discussed below.

Abnormalities of the facial nerve

Dehiscences of the facial nerve are not uncommon and in about 0.5% of middle ears there is a sizeable dehiscence, so that the nerve bulges down and obscures the arch and footplate. In some cases, it is possible to displace the nerve upwards and complete the operation, but if footplate surgery is likely to be blind, it is safer to abandon the operation. Very rarely, the facial nerve takes an anomalous course, either splitting to surround the stapes or coursing inferior to the oval window (Hoogland, 1977).

Persistent stapedial artery

A persistent stapedial artery of sufficient size to prevent the completion of the operation is very rare and is found in 0.2% of operations. A small vestigial vessel is not uncommon and must not be damaged as it can cause troublesome bleeding.

Perilymph flooding

This is a rare complication. Causse and Causse (1980) have only encountered it as occurring six times in more than 20,000 operations, an incidence of 0.0287%. It is seen in an ear with an abnormally patent cochlear aqueduct and polytomography before operation may show an abnormally-shaped vestibule. The complication is more common in ears with congenital fixation of the footplate. If a small safety hole is made in the footplate before the crura are detached, the condition will be detected and it may then be possible to close it by a connective tissue graft placed between the footplate and the crura.
In other cases, it may be possible to seal the 'perilymph gusher' with a soft tissue graft and the prosthesis put into position in an attempt to hold the graft in contact with the oval window. The flow of cerebrospinal fluid may last for several days and a severe sensorineural hearing loss is likely to occur with this complication.

**Floating footplate**

This is a potentially serious complication and may result in a 'dead ear' if attempts are made to extract the footplate, which in some cases becomes hinged on itself, or even disappears from view. A preliminary drill hole in the footplate, before attempts to remove the crural arch are made, is a wise precaution and will often prevent the complication. If the footplate is visible, it may be possible to remove it by manipulation and extraction with a fine hook, or a small drill hole may be made at the margin of the oval window and fine hook used to remove it. If it cannot be removed without excessive manipulation it should be left in place, a soft tissue graft placed over the oval window and the operation abandoned. Some authorities apply a prosthesis from the incus to the floating footplate, if it remains slightly hinged inwards, and good results have been reported from this procedure.

**Depressed footplate, submerged footplate**

This is more common than 'floating footplate'. The footplate may be totally submerged and this is caused by trying to remove a floating footplate, or the posterior part may become submerged when the fixation of the footplate is confined to the anterior part.

No attempt should be made to retrieve the submerged footplate by instruments as this may cause severe cochlear damage. Roche, Wayoff and Moeller (1971) described a method of dealing with this complication, which is not damaging to the contents of the vestibule. Drops of blood are poured laterally into the vestibule and when it is full of blood and a clot has formed, which takes 10-12 minutes, the clot is removed by a lateral application of the sucker bringing the footplate to the surface.

**Presence of blood in the vestibule**

Excessive bleeding during operation is a hazard that many otologists have to face in the UK if they use general anaesthesia as too many anaesthetists do not think that 'hypotensive' anaesthesia is justified in stapedectomy. The benefits of a dry field cannot be overemphasized and if the surgeon cannot persuade his anaesthetist to provide this he would be advised to use local anaesthesia. There is still some disagreement about the possible serious effects of leaking blood into the vestibule. Linthicum and Sheehy (1969) could find no evidence of any detectable ill-effects from this complication, while Smyth and Hassard (1978) believed that labyrinthine trauma from this cause cannot be dismissed. Preoperative fluoride therapy may help in reducing the activity and thus the vascularity of otosclerotic bone.

**Tympanic membrane tear**

If a small tear of the drum is found at the end of the operation and it is only a slit, it is covered with gelatin sponge. A larger tear is closed by rotating the flap and covering it
with gelatin sponge. Healing almost always occurs. If inadequate flaps have been made and this is associated with excessive bone removed, a defect may be formed at the end of the operation and this should be covered with temporalis fascia placed beneath the edge of the drum and on the adjoining meatus.

**Obliterative otosclerosis**

This condition is less common than it used to be. It requires special treatment which is discussed in a later section.

**Damage to the chorda tympani nerve**

During a stapedectomy operation, it is frequently necessary to displace the chorda tympani nerve to gain adequate exposure, and there is controversy concerning the advisability of stretching the nerve or cutting it. It should be appreciated that if the nerve is cut, it will produce permanent loss of sensation of taste in the anterior two-thirds of the tongue on the same side and no re-innervation of the taste buds can take place, either from the chorda tympani nerve of the opposite side or the posterior third of the tongue supplied by the glossopharyngeal nerve. Permanent loss of chorda tympani innervation results in atrophy of the taste receptors in the anterior two-thirds of the tongue; the dorsum of the tongue becomes smooth and pale. The patient may not complain of the loss of taste sensation, if one nerve is cut, as the tongue retains sensation in 66% of its surface (Diamond and Frew, 1979).

If the nerve is stretched during operation, a persistent abnormal sensation in the tongue may occur, described by the patient as salty, or metallic, and this is caused by paraesthesia of this sensory nerve. Cutting the nerve may also produce these unpleasant sensations of taste. If the nerve is displaced but not divided the taste disturbance is less (Bull, 1965; Wiberg, 1971).

Bilateral loss of the chorda tympani nerve produces marked symptoms in the majority of patients and, in addition to the loss of taste, there is loss of the secretomotor supply to the submandibular and sublingual salivary glands which produces an uncomfortable dry mouth. If at all possible, the surgeon must preserve the chorda tympani nerve, and this is essential in bilateral operations.

**Obliterative otosclerosis**

Special considerations of technique must be applied to the massive otosclerotic focus filling in and obliterating the oval window as the early attempts to drill out the oval window were accompanied by a high incidence of sensorineural deafness and reclosure of the oval window by regrowth of otosclerotic bone (House, 1962; Schuknecht, 1963; Shea and Sanabira, 1963).

Gristwood (1966b) reported an unusually high incidence of the truly obliterated footplate seen in 350 consecutive operations for otosclerosis. It has been the experience of Gristwood and other Australian otologists that the incidence of the thick footplate in Southern Australia is not known, but it is thought that the deficiency of fluoride ions in the drinking water and the late fluoridation of water supplies, which was not started until 1971, in
metropolitan areas, may be a factor. The most common finding was that about 50% of those with a thick and obliterated footplate developed their hearing loss during childhood or adolescence, and 80% of this group had noticed some hearing impairment before the age of 25 years. It was also found that the obliterated case could not be identified before operation by audiometry and that the condition was rarely unilateral.

Gristwood described three varieties of obliterative oval window otosclerosis.

**Truly obliterated footplate**

In this condition, the stapes footplate is replaced by a massive otosclerotic focus that fills in the oval window niche. The rim of the footplate cannot be identified and the crura of the stapes may be buried.

**Solid partly-obliterated footplate**

The footplate was found to be diffusely and greatly thickened, but a rim of delineation, often spurious, could be seen over a small segment of its circumference, the remainder of the margin being obliterated.

**Solid spuriously delineated footplate**

This is a rare type in which a complete gutter of delineation surrounds an apparently thick solid footplate. The spurious nature of the delineation is only revealed when attempts to remove either half of the footplate fail because of the obliteration deeper in.

**solid delineated footplate**

This is the thick biscuit or rice grain footplate which, although diffusely thickened, retains a delineated rim and an intact annular ligament. This type of footplate may be firmly wedged in the oval window and it can be pushed into the vestibule during operation giving rise to a 'floating footplate'.

**Narrowed oval window niche**

Otosclerotic foci around the oval window may lead to a marked narrowing of the niche which produces a slit-like effect at the oval window. Attempts to remove the footplate or make a fenestra within it should not be made until the overhanging bone has been removed and the footplate can be seen.

**Sensorineural deafness in otosclerosis**

Sensorineural deafness is frequently associated with the conductive hearing loss of otosclerosis, but there is still argument about the exact mechanism by which it occurs. There is controversy about the concept of 'cochlear otosclerosis' which is a sensorineural hearing loss caused by otosclerosis of the labyrinth in the absence of stapes fixation. Shambaugh (1965), Derlacki and Valvassori (1965) and Balle and Linthicum (1985) have produced strong arguments for supporting the theory of cochlear otosclerosis while, on the other hand, Gross
(1969), and Schuknecht and Kirschner (1974), and Schuknecht (1983) have failed to show otosclerotic foci of significant size or incidence in the temporal bones of patients with pure sensorineural deafness of unknown cause.

The possible causes of the cochlear degeneration seen in otosclerosis are:

1. bony invasion of the scala tympani of the cochlea (Politzer, 1894)

2. circulatory changes in the cochlea as a result of abnormal bony foci (Mayer, 1911; Ruedi, 1965)

3. damage to the cochlea by toxic metabolites from abnormal bone (Siebenmann, 1912; Witmaack, 1919; Chevance et al, 1970).

**Bony invasion of the scala tympani of the cochlea**

In the very early descriptions of the disease by Politzer (1894), Habermann (1904) and Siebenmann (1912), sporadic cases are mentioned showing bone formation in the scala tympani which were thought to be caused by otosclerosis. In 1921, Lange found the scala tympani to be partially filled with newly formed bone tissue into which the otosclerotic process of the labyrinthine wall had penetrated.

Nager and Fraser (1938), in their paper on bone formation in the scala tympani in otosclerosis, stated that the main change occurs in the labyrinthine capsule with the inner ear showing only minor alterations. After the examination of a large number of temporal bones, they found that in rare cases there was extensive bone formation in the scala tympani, and in the more advanced cases it was almost filled with new bone. They believed that the cause of this in the scala tympani was the result of the otosclerotic focus in the wall of the labyrinth producing a certain alteration, or irritation, of the endosteal layer and perilymphatic spaces leading to circumscribed fibrosis and bone production. This type of bone formation in the inner ear is found only in otosclerosis, but it is a rare and uncommon cause of the inner ear deafness which is so common in this condition.

**Circulatory changes in the cochlea as a result of abnormal bony foci**

Ruedi (1965) re-examined Otto Mayer's theory that venous obstruction caused by invasion of the root of the spiral lamina in the basal turn of the cochlea by otosclerotic bone, gave rise to incompetence of the venous drainage of the anterior and middle spiral veins, leading to neuroepithelial degeneration of the inner ear. Ruedi described how the actively growing otosclerotic focus advanced, giving rise to thrombosis in the vessel adjacent to it. The vessels became walled in, so that a sharp demarcation was apparent between the new vascular channels of the otosclerotic focus and the old vessels of the otic capsule. It was noticed that each focus developed its own self-contained, largely autonomous vascular system, and it was found that a connection between a wide capillary of the old otic capsule and the vascular space of an active otosclerotic focus could develop. When the otosclerotic lesion had penetrated the region of the promontory to appear under the mucous membrane, shunts were often seen between the blood vessels of the otosclerotic deposit and those of the mucosa.
These vascular shunts are well known clinically as the 'flamingo blush' first described by Schwartze, and when seen on examination, are an indication of active otosclerosis.

Ruedi also demonstrated vascular shunts between otosclerotic blood vessels and spiral capillaries, which caused marked congestion in the region of the modiolus and he was of the opinion that the formation of new lamellar bone within the inner ear was caused by this stasis, as opposed to Nager's theory that it was the stimulation of the osteoblastic activity of the endosteal capsule.

Ruedi's final investigation, after establishing the presence of the shunts, was to determine whether the atrophy of the labyrinth could be the result of the disturbances in the inner ear brought about by the otosclerosis. He found, in seven out of 10 temporal bones examined, that the organ of Corti was disintegrated or missing altogether, with a degeneration of the corresponding nerve fibres and ganglion cells. In the majority (six out of seven bones), he detected a shunt in the region of the spiral capillary and the inferior spiral vein. In one of these there was no sign of a venous shunt but an obliterated artery, thought to be the vestibulocochlear, and thrombosis of this accounted for the disintegration of the neuroepithelium and atrophy of the stria vascularis, within the basal turn, seen in the specimen.

Although Ruedi believed that a vascular aetiology accounted for all the inner ear changes seen in otosclerosis, it is only in advanced disease that such abnormalities are seen. The sensorineural hearing loss so common in otosclerosis cannot be explained, in all cases, as being the result of venous congestion alone and his theory is only applicable in some cases.

It is necessary to examine the other theory, of a humoral factor, to explain more satisfactorily the phenomenon of sensorineural degeneration in otosclerosis.

**Damage to the cochlea by toxic metabolites from the abnormal bone**

In 1912, Siebenmann postulated that the abnormal bone of a focus of otosclerosis, which was invading the labyrinth, poured out inflammatory products into the fluid and these contained toxic metabolites which caused the labyrinthine lesions. Witmaak (1919) also assumed that degeneration of the labyrinth was caused by the diffusion into the labyrinthine fluids of an acid liberated by otosclerotic lesions dissolving the bone.

The actively growing deposit of otosclerosis, as it erodes the endosteum, comes into close relationship with the perilymph of the basal scala tympani. Such active foci have a rich blood supply and extensive marrow tissue; the spaces of the latter may communicate directly with the scala tympani, so that the perilymph flows, not only over the otosclerotic bone, but also into the spaces, allowing mixing of their contents.

It is surprising that, until recently, so little attention has been paid to the humoral theory, for there is such a close relationship between the actively growing deposit of otosclerosis and the perilymphatic spaces. In 1958, Harrison and Naftalin formulated a theory of the active circulation of the labyrinthine fluids which suggested that the inner ear damage in otosclerosis is humoral in origin. They believed that perilymph is formed by ultrafiltration from blood vessels in the perilymphatic space. The perilymph, as a result of the hydrostatic
pressure in the general circulatory system, passes across Reissner's membrane and the basilar membrane as a plasma transudate and reaches the scala media. The plasma transudate is then converted into endolymph by a specific process of the stria vascularis, which replaces sodium from the plasma transudate with potassium, by a low energy exchange mechanism analogous to the resorption process of the renal tubules.

If metabolites and other breakdown products of the otosclerotic process were to contaminate the perilymph they could in fact pass across Reissner's membrane and the basilar membrane, with the perilymph, and on reaching the scala media cause damage to the organ of Corti. A simpler explanation of the route of entry of toxic products is through the canaliculi.

In 1970, a research project was undertaken by a biochemist, two histopathologists, an enzymologist and an otologist from three different centres (Paris, Coppenhagen and Béziers), which has led to further development of the theory that the inner ear damage in otosclerosis may be humoral in origin (Chevance et al, 1970).

They found that osteoclasts are rarely, and only exceptionally, found in the extension zone of the focus, or in the marrow spaces that constitute the active or otospongiotic focus. This confirms the observations of Ogilvie and Hall (1953) who had noted that in the diffuse form of otosclerosis 'the osteoclasts were remarkable for their scarcity, small size with degenerate cytoplasm and nuclei, and loss of direct application to the bone...'. It has been generally believed that these cells are responsible for the entire bony resorption which takes place in the lytic phase of the otosclerotic lesion.

Chevance et al have been able to demonstrate that apart from fibroblasts, fibrocytes, osteoblasts, and osteocytes there is, in addition, a special type of cell, containing lysosomes which are dense vesicular bodies in the cytoplasm. The frequency of this cell, its location and morphology indicated that, in their opinion, it was a histiocyte taking an active part in bone resorption.

These cells were most often found in the 'front' of the otosclerotic process and the bone surrounding them showed the presence of lysis. It is well known that lysosomes contain a number of hydrolases with a very high enzymatic content and the activity of acid phosphatases is generally considered to be the best index of lysosomal content. It was found that the histiocytes exhibited strong acid phosphatase activity and these workers are of the opinion that the histiocytes play the decisive role in the process of otosclerotic resorption.

In addition to the demonstration of the presence of histiocytes in the active focus, otosclerotic microfoci have been found beyond the advancing edge of the lesion, and in these the lytic and rebuilding phases were found to be proceeding simultaneously.

The perilymph of patients subjected to stapedectomy operations, with the presence of otosclerosis confirmed by biopsy, has been examined: six enzymes have been identified: phosphatasic acid, collagenase, alpha-chymotrypsin, lactic dehydrogenase, ribonuclease and trypsin. These enzymes control the evolution of the otosclerotic microfoci and they also pass into the labyrinthine fluids through the cochlear barrier, previously thought to be impassable, entering through the canaliculi.
The actual passage of these enzymes has been demonstrated in a series of perilymph specimens, first studied by Adams' method (Adams and Tuqan, 1961), later by qualitative study and finally by quantitative methods using a microelectrophoretic technique (Uriel and Avrameas, 1964; Uriel, 1971).

Perilymph specimens were studied by multiple statistical analysis and it was found that:

1) proteases or hydrolases enter the labyrinthine fluids in about 75% of cases of otosclerosis and this corresponds to the 75% incidence of cochlear degeneration which is seen in patients with clinical stapedial otosclerosis

2) statistical correlation have been arrived at by standard tests, and binary and threefold correlations have been established between the proteolytic activity of the perilymph and the progressive sensorineural hearing loss which is seen in 75% of the cases with stapedial fixation from otosclerosis.

A correlation was also seen between the proteolytic activity of the perilymph and impairment of the posterior labyrinth, shown by the torsion swing test and electronystagmography, in patients without clinical symptoms of vertigo.

The enzymatic concept of otosclerosis

The experimental findings of Chevance, Causse and their co-workers have led them to postulate the theory of the enzymatic concept of otosclerosis.

The process begins in one or more of the numerous cartilaginous rests scattered through the enchondral layer of the otic capsule. Hydrolytic enzymes and proteases, causing cellular destruction, spread from the original focus to the different parts of the cochlea. If the focus is close to the stapediovestibular joint the process of bone rebuilding may produce a fixation of the stapes footplate and a conductive hearing loss. If the proteases and proteolytic enzymes reach the inner ear a sensorineural hearing loss occurs, and if the enzymes reach the posterior labyrinth vertigo may be caused. If the focus is situated far from these sites, the disease may never be detected clinically.

It is well recognized that conductive deafness in otosclerosis may exist without substantial hearing loss, but a sensorineural hearing loss that is disproportionate to the age of the patient is commonly associated with the conductive deafness. The sensorineural hearing loss may precede fixation, although it is usually associated with it. It is also recognized that a gradually developing sensorineural deafness occurs in many patients who have had a successful stapedectomy operation, and this also is often disproportionate to the patient's age.

If otosclerosis of the cochlear capsule can cause sensorineural hearing loss when there is stapedial otosclerosis, it can cause sensorineural hearing loss when there is no stapedial fixation and when this occurs, the condition is known as cochlear otosclerosis.
The enzymatic concept of otosclerosis, elaborated by Chevance and Causse and their colleagues, is of considerable interest as it explains many facets of the disease which still remain obscure, and it explains the long, slow and variable progress of the disease.

These workers believe that otosclerosis is a local disease in which there is an upset of the equilibrium between enzymes and antienzymes in the microfoci of otosclerosis and this gives rise to variable clinical results; for example if the focus is stapedial a conductive loss occurs, but if the focus is outside the region of the footplate a sensorineural deafness may result, and if the focus is in the region of the vestibule, vertigo may result. This theory gives strong support to those who believe that medical treatment, by antienzymes or enzyme inhibitors, is important in the treatment of this disease.

The belief that sensorineural hearing loss can occur in a pure form, as the result of otosclerosis without a conductive hearing loss, has been strongly criticized (Schuknecht and Kirschner, 1974; Schuknecht, 1983). The criticism is based entirely on histological examination of temporal bones, is dogmatic, and does little to help in the explanation of the obscure aspects of this disease.

Audiometric studies were carried out by Glorig and Gallo (1962) who compared bone conduction levels of patients with otosclerosis with air conduction levels in the general population. The assumption was made that, because the hearing losses found in the general population are largely sensorineural, the comparisons would be valid. Their results indicated that otosclerosis does not increase the sensorineural hearing loss above that to be expected in the general population and audiometric patterns for higher frequencies, in those with otosclerosis, resemble those found in general populations. In patients over the age of 60 years, it was found that the sensorineural hearing loss in high frequencies was greater in patients with otosclerosis than in the general population. This finding contradicts their conclusions.

Although otologists are in agreement that gross lesions cause sensorineural deafness in otosclerosis by direct invasion of the scala tympani of the cochlea, and gross lesions interfere with the circulation of the stria vascularis and this may also give rise to sensorineural deafness, there is still controversy concerning the humoral theory.

Causse and Chevance have developed the humoral theory, first postulated by Siebenmann in 1912, and their theory of enzymatic concept of otosclerosis is the only one that attempts to explain many of the enigmas of this disease.

**Diagnosis of sensorineural deafness in otosclerosis**

In 1978, Shambaugh pointed out that, before Lempert's fenestration operation for otosclerosis came into general use, few clinicians in North America were able to diagnose stapedial ankylosis caused by otosclerosis. As recently as 1931-1932, not one patient coming to the Massachusetts Eye and Ear Infirmary during those 2 years was diagnosed as having otosclerosis. He believes that the situation concerning pure cochlear otosclerosis is similar today in that this diagnosis is denied in some large clinics and is made with hesitation in others.
Shambaugh and Holdermann, in 1926, gave three criteria necessary for the probable diagnosis of cochlear otosclerosis and these were:

(1) an insidious onset beginning in early adult life
(2) the absence of any reason for the nerve loss
(3) conductive deafness in other members of the immediate family.

Shambaugh (1966) gave six reasons for suspecting that otosclerosis may be the cause in cases of pure sensorineural deafness:

(1) a positive Schwartze sign, in one or both ears
(2) a family history of surgically confirmed stapedial otosclerosis
(3) the presence of symmetrical sensorineural hearing loss in both ears, one of which has stapes fixation
(4) a flat, rising, or a 'cookie-bite' audiometric air conduction curve with unusually good speech discrimination for someone with a pure sensorineural loss
(5) pure sensorineural hearing loss beginning insidiously in early, or middle, adult life and progressing with no apparent cause
(6) the demonstration of stapes fixation in a patient with previous pure sensorineural deafness of no apparent cause.

Causse, Shambaugh and Chevance (1977) have described three categories of diagnostic criteria for making a diagnosis of cochlear otosclerosis.

Criteria of presumption

There is a slowly progressive hearing loss in childhood of a sensorineural type which becomes worse at puberty, or at period of endocrine activity, in a family with a history of a progressive sensorineural hearing loss.

If a sensorineural hearing loss is aggravated in a woman by pregnancy, the menopause, or by treatment with oestrogens.

If there is good discrimination for speech in a patient with sensorineural deafness using a hearing aid and especially if there is good discrimination in noisy surroundings.

Criteria of probability

If there is a positive Schwartz sign in one or both ears, if the audiogram shows a sensorineural loss with a 'cookie-bite' curve and if there are positive polytomographic findings in the cochlear capsule.
Criteria of certainty

If, in a case of progressive sensorineural deafness, there develops an 'on-off' impedance effect or diphasic impedance change indicating impending fixation of the stapes and if, in a case of slowly progressive pure sensorineural deafness, the beginning of an air bone gap develops.

Kelemen and Linthicum (1969) found that severe sensorineural hearing loss was usually associated with extensive invasion of the cochlear capsule by otospongiotic bone, but mild degrees of sensorineural hearing loss were associated with lesser foci which did not always reach the cochlear endosteum.

Radiological demonstration of cochlear otosclerosis

Conventional radiography is of little value in the diagnosis of otosclerosis and linear tomography does not give adequate information about the very small structures in the temporal bone. Multidirectional hypocycloidal polytomography is of considerable value and Derlacki and Valvassori (19650 have developed this technique.

It is necessary for the lesion to be greater than 1 mm for it to be visible and the density of the focus must be different from that of the normal capsule for it to be detected. The normal capsule of the labyrinth is the densest bone and cannot become more sclerotic, but it can become thicker when mature otosclerotic bone increases the thickness of the capsule, which then appears roughened, or scalloped on its edges caused by the irregular outline of the new bone. Derlacki and Valvassori have shown that capsular changes can be demonstrated in 65% of patients with confirmed stapedial otosclerosis and in 30% of patients with clinical findings suggestive of cochlear otosclerosis.

Applebaum and Shambaugh (1978) stated that: 'Caution must be exercised in the interpretation of subtle polytomographic changes in the cochlear capsule and restraint used in the X-ray diagnosis of pure cochlear otosclerosis until there is evidence of correlation with pathological material'.

High resolution computerized tomography

This method has proved to be valuable in assessing the pathology and extent of chronic suppurative otitis media, and precise information concerning the normal anatomy of the ossicles, facial nerve, tegmen and semicircular canals. It is of value in the differential diagnosis of a conductive deafness as it will reveal, for example an interrupted ossicular chain from a lesion of the incus. With improvements in technique, this method may prove to be of value in the study of labyrinthine otosclerosis (de Groot, 1985).

The perilymph fistula after stapedectomy

This is a serious complication as it is potentially dangerous from the risk of meningitis and it gives rise to hearing loss which will progress if the fistula does not close, either by spontaneous healing, or as the result of a revision operation.
The signs and symptoms of perilymph fistula were first described by Lewis (1961) and by Farrior (1962), and the complication, at one time thought to be unusual, is now accepted as being the most common single complication of stapedectomy.

Fistulae occur after all operations on the stapes footplate and Table 14.2 shows the incidence with various types of operation from a survey of a considerable number of operations which were carried out by Harrison, Shambaugh and Derlacki in 1970.

**Table 14.2. Symptoms and findings**

<table>
<thead>
<tr>
<th>Primary fistulae</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Dysequilibrium</td>
<td>77%</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>71%</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>45%</td>
</tr>
<tr>
<td>Fluctuating hearing</td>
<td>42%</td>
</tr>
<tr>
<td>Secondary fistulae</td>
<td></td>
</tr>
<tr>
<td>Dysequilibrium</td>
<td>61%</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>78%</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>44%</td>
</tr>
<tr>
<td>Fluctuating hearing</td>
<td>83%</td>
</tr>
<tr>
<td>Discrimination loss</td>
<td>72%</td>
</tr>
</tbody>
</table>

(Reproduced from Moon (1970) by kind permission of the Editor of *The Laryngoscope*)

**Diagnosis of perilymph fistula**

The symptoms of a perilymph fistula are a fluctuating hearing loss, tinnitus, a feeling of fullness in the ear and vertigo. The symptoms are those of labyrinthe hydrops, which this condition can simulate.

The fistula may be primary, dating from the time of operation when there is failure of the seal of the oval window, or it may be secondary, when it can appear many months or even years after the original operation.

**Primary perilymph fistula**

When the opening, created by the surgeon in the oval window region, fails to heal after the operation, a disturbance of equilibrium persists in the days and weeks after operation, until vestibular paralysis and compensation occurs. In other patients, there may be brief periods of vertigo continuing over a long period.

**Secondary perilymph fistula**

The characteristic symptom of secondary perilymph fistula is a change of hearing coming on after an interval, which may be months or years after a successful operation; associated with this are feelings of fullness, tinnitus and dysequilibrium. There may be
considerable variation in the symptoms but a conductive deafness may be the early sign of a fistula and this may precede a serious irreversible labyrinthine lesion (Goodhill, 1979).

The symptoms and findings in a review of 49 cases of perilymph fistula are given in Table 14.3 (Moon, 1970). Sometimes the patient may give a history of symptoms developing after an incidence such as severe nose blowing, strenuous exercise or flying in an unpressurized aircraft, but often no precipitating factor may present and the patient may not seek advice until there is a considerable degree of sensorineural deafness.

Table 14.3. Hearing loss

<table>
<thead>
<tr>
<th></th>
<th>Primary fistula</th>
<th>Secondary fistula</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensorineural</td>
<td>16 (52%)</td>
<td>7 (39%)</td>
</tr>
<tr>
<td>Mixed</td>
<td>13 (42%)</td>
<td>5 (28%)</td>
</tr>
<tr>
<td>Conduction</td>
<td>2 (6%)</td>
<td>3 (17%)</td>
</tr>
<tr>
<td>None</td>
<td>0</td>
<td>3 (17%)</td>
</tr>
<tr>
<td></td>
<td><strong>31</strong></td>
<td><strong>18</strong></td>
</tr>
</tbody>
</table>

(Reproduced from Moon (1970) by kind permission of the editor of *The Laryngoscope*)

**Clinical findings in perilymph fistula**

The clinical examination of the ear is usually normal, rarely the tympanic membrane may be retracted or there is evidence of fluid in the middle ear.

**Audiometric tests**

Hearing tests carried out soon after the onset of a fistula will show findings similar to those seen in labyrinthine hydrops, that is a pure tone sensorineural hearing loss, in the low frequencies initially, followed by a flat loss which fluctuates. There may be recruitment and short increment sensitivity index (SISI) scores are often above 75%. In the early stages, discrimination scores fluctuate with the pure tone threshold, but later they may be disproportionately lower than expected when compared with the stapedius reflex threshold. In some cases, and particularly when vertigo is the main complaint with little or no depression of pure tone levels, there may be a markedly diminished speech discrimination score. It has already been pointed out that a variable conductive loss may occur without a sensorineural component, and this is characteristic of the 'slipped strut', an early technique now seldom performed.

**Vestibular tests**

**Hallpike caloric test**

The characteristic finding is canal paresis, or hypoactive response, and this may be present at an early stage of the condition. It must be appreciated that after stapedectomy there
is a high incidence of diminished caloric response and so the significance of the caloric test is difficult to evaluate and of little value in the diagnosis of a fistula.

**Electronystagmography**

Electronystagmography may reveal a directional fixed positional nystagmus but this cannot be relied upon as an indication of a fistula.

**Fistula test**

Fistula test with the pneumatic otoscope may be helpful but have been found to be negative in one-third of cases. The negative pressure fistula test with electronystagmography produces nystagmus with the quick phase away from the affected ear. Fistula test with electronystagmography and the impedance bridge have superseded the older methods as they give a higher degree of accuracy.

**Diagnosis of perilymph fistula using electronystagmography and impedance bridge**

If tympanometry is carried out after normal stapedectomy with the opening into the oval window sealed, the change of pressure will usually produce nystagmus which can be detected on the electronystagmographic tracing. The changes of pressure produced by tympanometry, if a fistula is present, do not lead to corresponding pressure changes in the vestibule of the inner ear as the perilymph leaks through the fistula and so there is no nystagmus.

Daspit, Churchill and Linthicum (1980) have found the electronystagmographic fistula test to be the most valuable and positive of all the tests. They used the Madsen impedance bridge; the canal (probe) ear pressure was increased to + 200 mm H₂O, held at this level for 15 seconds and then rapidly decreased to - 400 mm H₂O. An electronystagmographic recording was made throughout this procedure, which was repeated two or three times, with a visual inspection for rotatory nystagmus; the patient was also asked if there was any feeling of giddiness, or any condition similar to the symptoms that had been experienced. The fistula test was considered to be positive if nystagmus was elicited, especially in the absence of spontaneous nystagmus.

When a positive result was obtained, a similar procedure was performed on the other ear together with a routine test of positional nystagmus which, if present, could influence the test results. All recordings were carried out with eyelids closed.

**Safety of the test**

The pressures used in impedance measurements were compared with those used in pneumatic otoscopy where a positive pressure as high as + 300 mm H₂O and as low as - 600 mm H₂O could be obtained. These levels exceeded any that were used with the impedance bridge and it was considered that the test was a safe procedure.
Results of electronystagmographic fistula test

Daspit, Churchill and Linthicum (1980) reported that the electronystagmographic fistula test is of considerable value, with a degree of accuracy greater than the fistula test with pneumatic otoscopy. Caloric tests, tragal compression and a Valsalva manoeuvre were found to be of little value in the diagnosis of perilymph fistula.

Aetiology of perilymph fistula

Primary fistula

The surgeon creates a fistula at every stapedectomy operation and relies on the natural process of healing, or in some techniques a graft of soft tissue, to seal the opening which has been made into the vestibule. In most operations, there is enough surgical trauma to the oval window mucoperiosteum to lead to the production of an inflammatory repair envelope around the prosthesis sealing the opening into the oval window. Fistulae are more common with plastic prostheses than with interposition techniques; however, the hearing results using prostheses are better.

There is no doubt that a small fistula remains after many stapedectomy operations with incomplete closure of the air bone gap and the hearing result may be acceptable to the patient. Although perilymph fistulae usually lead to a sensorineural hearing loss this is not always the case and a persisting conductive loss after an operation, which appeared to be satisfactory at the time, should warn the surgeon that there may be a perilymph fistula and a revision operation to close it will be the best line of treatment. It must be appreciated that a small fistula may become larger under the influence of barotrauma; a sensorineural hearing loss will follow and it may then be too late to save the hearing.

Secondary or acquired fistula

Secondary perilymph fistulae are usually the result of barotrauma which breaks the fragile seal and can occur at any time after operation. In most cases, the stapedius tendon has been cut and a sudden change of intratympanic pressure may produce an abnormal movement of the prosthesis, giving rise to a rupture of the oval window seal.

The following case records from the author's practice are examples of fistulae developing after stapedectomy operations.

Case record - secondary perilymph fistula from barotrauma

Female aged 47 years: a stapedectomy-Teflon piston with vein graft had been performed 18 months previously, with a good hearing result. After a severe cold, the patient blew her nose very vigorously and complained of slight vertigo and hearing loss. A revision operation 4 weeks after these symptoms developed revealed a fistula close to the inferior margin of the oval window. The oval window was regrafted with vein, a new Teflon piston placed in position and the hearing was partially preserved.
**Case record - primary and secondary perilymph fistulae**

Female age 49 years: the patient has a stapedectomy with Teflon piston performed by another surgeon one year before she was referred to the author. The operation had produced some hearing gain but incomplete closure of the air-bone gap. Slight vertigo had persisted since the operation and some months later this became worse and the hearing disappeared. When seen she had a severe sensorineural deafness with marked vertigo and loss of vestibular function in the operated ear.

A revision operation showed that the Teflon piston had fallen off the incus and was lying in the vestibule and there were three fistulae visible in the membrane clothing the oval window. As there was no useful hearing and severe vertigo a labyrinthectomy was carried out and the oval window grafted with vein.

This is an example of a primary fistula resulting from a loosely attached Teflon piston and secondary fistulae had also developed. When this piston fell into the vestibule a 'dead ear' resulted and the vestibular system was severely damaged.

This case illustrates the importance of closing a Teflon piston after it has been applied to the incus. If the piston is opened too widely the edge of the 'eye' may be damaged and it may never close properly.

**Case record - fistula 20 years after a successful stapedectomy**

Male aged 59 years: the patient had a fenestration operation on the left ear in 1952 and a right stapedectomy operation, polyethylene strut and vein graft of the oval window in 1959. The operations were performed by the late Sir Terence Cawthorne and both were successful. In 1979, 20 years after the stapedectomy operation, the patient complained of a small hearing loss and slight vertigo when he carried out yoga exercises.

A revision operation on the right ear revealed that the tip of the polyethylene strut had penetrated the vein graft and there was a slight leak of perilymph. Necrosis of the tip of the long process had occurred, but the strut was still attached to the incus by a strand of mucous membrane. The strut was removed and the oval window regrafted with a vein graft. A slight sensorineural hearing loss resulted from the fistula but the speech frequencies were not affected. The patient did not wish for further surgery and obtained good hearing with an aid.

**Prevention of perilymph fistula**

The very thin membrane which will develop to close the oval window if fat or gelfoam is used, can be avoided if the oval window is covered with a vein graft. If the greater part of the footplate is removed, not only is there a greater risk of sensorineural deafness being produced by such a traumatic procedure, but there is a greater risk of a fistula developing later.

The small fenestra stapedectomy with or without a soft tissue graft is an advance on techniques where a large opening is made in the footplate.
The interposition operation, when the posterior crus of the stapes is used to make contact with a soft tissue graft closing the opening in the oval window, is the safest technique for prevention of a fistula.

**Eustachian tube function**

Hemenway, Hildyard and Black (1968) found that perilymph fistula accounted for 30% of poor results in a series of 1788 operations and they found that the most common condition present at the onset of symptoms was inadequate eustachian tube function.

They believe that it is most important to question prospective patients about their occupation and to investigate the eustachian tube function. If a patient is a pilot, if his hobby is mountain climbing, ski-ing, water ski-ing or diving, he is advised to wear a hearing aid.

**Advice given to the patient after operation**

It is important to warn the patient who has had a stapedectomy operation that:

1. nose blowing should be avoided and the mouth should be kept open on coughing and sneezing
2. flying or going over a mountain pass should be avoided for at least 10 days after operation, or when an upper respiratory tract infection develops
3. diving when swimming should be avoided
4. lifting heavy objects should be avoided
5. any hearing loss, vertigo or ear infection must be reported immediately.

When eustachian tube dysfunction develops after operation, Hemenway and his colleagues placed ventilation tubes in the tympanic membrane.

**The treatment of perilymph fistula**

The treatment of perilymph fistula is a tympanotomy at the earliest possible moment, with an attempt to close the fistula.

When the leak is detected the fistulous track is excised and the prosthesis removed with great care. The opening in the vestibule is covered with a soft tissue graft which is held in place by another prosthesis. A disadvantage of a two-stage method of closing a fistula is that the graft may float off with a recurrence of the fistula as there is no prosthesis to hold it in place.

All methods of repairing a fistula that have been described, rely on the application of a living graft to close the defect. It would seem logical to believe that if the original stapedectomy had included a graft to seal the oval window there would be fewer postoperative perilymph fistulae.
Results of treatment of fistula of the oval window

Unless early treatment is instituted, the chances of restoration or improvement of hearing are small and in some cases a troublesome vertigo may remain. It is imperative that the surgeon must be fully aware of this complication and realize that some techniques are safer than others. The use of gelfoam to seal the oval window produces a very thin membrane, and gives the highest incidence of fistula formation. Most surgeons have abandoned this technique today. The true small fenestra technique as practised by Marquet, Creten and van Camp (19720 is a safe method and the small fenestra stapedectomy, with vein graft and Teflon piston, as practised by Causse has also proved to be safe.

If the results of treatment of perilymph fistula are to show improvement, early diagnosis and immediate revision surgery are essential.

Revision operations

Conductive deafness may occur after a successful stapedectomy and when a prosthesis has been used, there are four principle causes: necrosis of the tip of the incus; loose attachment between the incus and the prosthesis; detachment of the prosthesis from the incus; and displacement or dislodgement of the prosthesis from the oval window area.

Recurrence of otosclerosis may spread from the anterior footplate area and recurrence of the lesion can lead to a closure of the oval window area even if the whole footplate has been removed. Recurrence of the bony deposit may occur after a stapedioplasty (Portmann interposition) and the author has noticed this in 8% of operations carried out by this method.

Dawes and Curry (1974) described their experiences of 82 revision operations giving special consideration to 50 operations in 44 patients, with a minimum follow-up period of 12 months; operative and postoperative details were known in over 1000 stapedectomy operations. These authors described the causes of failure in the two groups, of immediate and delayed conductive loss, and the results of revision surgery in both.

Immediate

The causes of early failure were found to be incus necrosis and loose fit of the prosthesis; in some cases, no cause could be found. The results of treatment proved to be worthwhile with a success rate of 50%.

Delayed

When the original Shea operation was being carried out, the causes of late conductive deafness were found to be necrosis of the lenticular process of the incus, displacement of the strut and new bone formation. In the piston cases, six losses were due to detachment from the incus and four of 'short piston'. The importance of not overstretching the 'eye' of the piston and closing it over the long process has been mentioned in an earlier section.
Results of treatment of delayed conductive loss

Dawes and Curry (1974) found that the results of revision surgery, after an initial good result from stapedectomy were good, but if a sensorineural deafness is to be prevented, early revision in the period soon after operation must be avoided.

The development of obliterative otosclerosis, or closure of the round window membrane is, in general, a contraindication to further surgery. The exact procedure to be adopted depends on the findings, but Morrison (1979) has stated that re-opening of the vestibule carries a 50% risk of inner ear damage; he has found that it is worthwhile attempting reconstruction of the sound-conducting mechanism after an assembly failure and the experienced operator can obtain success rate in the region of 50%.

Sensorineural deafness after operation

In the majority of cases, hearing loss after stapedectomy is caused by cochlear damage and a sensorineural deafness results. In some cases, the cause is a failure of the linkage system, producing a conductive deafness which is often associated with a sensorineural loss. Treatment of conductive deafness after operation is discussed in the section on revision operations.

Sensorineural deafness after operation may occur in the immediate postoperative period, in the intermediate period weeks or months after operation, or be delayed for months or even years after surgery.

Immediate sensorineural deafness after operation

The reported incidence of severe sensorineural deafness following stapedectomy varies from 0.5 to 4% and it is important to realize that these are the figures from a series of operations by expert surgeons with a special interest in stapedectomy. The results of the occasional operator are not reported and it is likely that the incidence of cochlear damage is much higher when the surgeon is inexperienced.

Smyth and Hassard (1978) reported an incidence of 'dead ear' of 3.5% (713 operations). Morrison (1962, 1979) has shown that in a series of 1000 operations, there was a hearing loss of 4% in the first 50, 2% in the next 50, no immediate losses in the next 500 and 0.25% in the remaining 400 operations.

Causes of hearing loss after operation

The causes of cochlear loss produced by operation are numerous - they are the direct result of trauma of varying types to the inner ear at the time of, or soon after, operation.

All operations involve the possibility of trauma to the inner ear; some techniques are more traumatic than others and the pathology of the lesion will influence this. In addition, a small group of patients is particularly sensitive to the creation of a window in the footplate of the stapes and so there is always the risk of this disaster after every operation, even by the
most expert surgeon. This risk is small and indeed almost non-existent with some techniques, if correctly managed.

The causes of immediate sensorineural deafness include: acoustic trauma from drilling; excessive movement of the stapes producing an hydraulic effect; rupture of the membranous inner ear; rapid loss of perilymph; footplate fragments or bone dust in the vestibule; and the floating footplate. Attempts at removal of the last of these may result in a 'dead ear' and it is essential that this complication is correctly managed.

**Acoustic trauma**

The modern microdrill is a safe instrument if it is used correctly. The drill must be light, preferably fixed to a small motor which allows slow rotary motion, and the drill ends must not be toothed unless there is an obliterative footplate, and this condition requires a special technique which is discussed elsewhere.

**Excessive movement of the stapes**

If an attempt is made to remove a prematurely mobilized footplate, and this can occur when there is minimal fixation, an hydraulic effect can be produced which is damaging to the membranous structures in the vestibule. It is a wise precaution to make a small opening in the footplate, with a slowly rotating microdrill before attempting to detach the crura from the footplate, and is a precaution that may prevent the hydraulic effect.

**Rupture of the membranous inner ear**

Meticulous technique and the making of a small fenestra should prevent this complication, but if small particles of bone do enter the vestibule, *no* attempt should be made to remove them either by instruments or suction.

**Rapid loss of perilymph**

This may be caused by the use of suction in the oval window which must therefore be avoided. It is occasionally seen in an ear with an abnormally patent cochlear aqueduct, when cerebrospinal fluid will enter the ear. The treatment has been described in the section on problems found at operation.

**Presence of blood in the vestibule**

This condition has already been discussed. It is unlikely that blood in the vestibule will cause any damage to the inner ear. The problem is the difficulty in dealing with the pathology found at operation when the operative field is obscured. In clinics which specialize in the surgery of otosclerosis, a dry field is achieved by suitable anaesthetic techniques.
Treatment of postoperative cochlear loss

If a technique is adopted which does not lead to a fistula after operation for example, the 'vein graft-interposition' operation or the true small fenestra technique, it is possible to obtain complete closure of the air bone gap and avoid postoperative high tone loss.

Causse et al (1970) are of the opinion that careful monitoring of the hearing after operation is of vital importance since it may be possible, if a sensorineural hearing loss is detected within a few hours of its appearance, to reverse the hearing loss by medical treatment.

The monitoring of the patient involves strict audiometric surveillance by means of bone conduction audiometry and Weber tests of the pure tone and speech variety. Speech and pure tone audiometry are carried out later and the final speech and pure tone audiogram is made on the twentieth postoperative day. After discharge the patient is instructed to report immediately to the surgeon, by telephone, if there is any sudden hearing drop, or the onset of tinnitus or vertigo.

The treatment of sensorineural deafness after operation is similar to that given for Ménière's disease and consists of the repeated injections of nicotinic acid intravenously, three to five times per day. Intravenous heparin is given in small doses, as it is believed to act as a vasodilator, an antispasmodic and as an agent that helps to absorb exudates. In large doses, over 150 mg/day, it acts as an anticoagulant. Hydrocortisone is also used, in gradually decreasing doses, as it is believed that it protects cell membranes, and has an antiinflammatory, antiedematous and antihaemorrhagic action. Sodium fluoride is added to this basic treatment for antienzymatic action on the cochlear lesions.

Medical treatment of sensorineural deafness after stapedectomy is carried out by few surgeons. Diagnosis is difficult as there is loss of discrimination for a time after most stapedectomy techniques. This is caused by the fistula created at surgery which usually closes spontaneously. Audiometry after operation often shows an incomplete closure of the airbone gap and a high tone loss in the immediate postoperative period. The audiometric pattern improves during the course of a few weeks after operation but the high tone loss takes longer and may never recover completely.

If medical treatment can lead to recovery of sensorineural hearing loss after stapedectomy, it should be tried more frequently although it will remain difficult to say if a successful outcome is the result of therapy, or due to the remarkable power of the inner ear to recovery after injury.