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OTOSCLEROSIS

By TERENCE CAWTHORNE (London)

(THE DALBY MEMORIAL LECTURE*)

WHEN Dr. Albert Gray gave this lecture in 1933, he spoke about Otosclerosis.

In recent years there have been such remarkable changes in our knowledge of this disorder, that Otosclerosis will again be presented as the subject of this Dalby Memorial lecture.

Introduction

Although the term "otospongiosis" gives a better description of the disease process, and is preferred in France, the original term otosclerosis, first proposed by Adam Politzer in 1889, continues to be the one generally used to describe the primary disorder of the bony capsule of the labyrinth which, by hindering the entry of sound into the labyrinth, causes conductive deafness; and for which no cause can be found on clinical examination of the auditory apparatus.

Politzer's description of otosclerosis is as fresh and as true today as it was some 65 years ago, and nearly every time that I operate on a case of otosclerosis I am reminded of his observation that "in the majority of the specimens the mucous membrane of the tympanic cavity does not present the least change from the normal".

We still do not know why this disease of bone should happen, nor why it attacks only the bony capsule of the labyrinth. Much has been written about otosclerosis, some of it fact but much of it fancy; and I, too, shall have to mix a little fancy with the facts.

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After enquiring into the incidence of otosclerosis, the clinical features as seen in 2,000 cases will be described. Then the changes in the bony capsule of the labyrinth will be considered and an explanation for their development will be offered. The mechanical effect these bony changes have upon the transmission and reception of sound will be described and finally the way in which this effect can be overcome surgically will be discussed.

Incidence

An analysis of 2,000 unselected cases of deafness in adults attending a Hearing Aid Distribution Centre for the purpose of being fitted with an aid was recently made by one of my associates, Mr. R. M. Harvey,* and the results were as follows:

TABLE I.

ANALYSIS OF 2,000 UNSELECTED CASES OF DEAFNESS ATTENDING A HEARING AID CENTRE

Conductive Deafness	1,125
Perceptive Deafness	711
Mixed Deafness	164

Before considering these cases in any detail it will be as well to define the terms used, as there are still some differences of opinion as to what the term "mixed deafness" means.

Included under the heading of Conductive Deafness are all cases in which the primary cause of the deafness was conductive, even though in the more advanced and long-standing cases there are signs which have generally in the past been attributed to changes in the perceptive mechanism. Such cases are not included under Mixed Deafness because I believe that these changes can be brought about by obstruction to one or both labyrinthine windows alone, provided it is sufficiently severe. Guild (1944) noted that in otosclerosis the nerve of hearing did not appear to be unduly liable to degenerative or other visible changes as the result of otosclerosis and yet nearly half the cases of otosclerosis in this group are severe, which is to say that they might be regarded by many observers as having "mixed deafness". In the present classification the term Mixed Deafness has been reserved for those patients in whom there are definitely two or more causes of deafness present.

TABLE II.

CONDUCTIVE DEAFNESS

Bilateral Chronic Suppurative Otitis Media		251
Otosclerosis		866
Moderate	458	
Severe	408	
Bilateral Otitis Externa		2
Paget's Disease of Bone		6
		1125

The group of 408 cases of severe otosclerosis includes many who might be regarded as mixed deafness, but all of these started as a progressive

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conductive deafness without visible sign of any cause, and only in the later stages developed signs of cochlear changes; signs which we now believe can be given in the presence of a normally acting cochlea, by a sealing-off of the labyrinthine windows:

TABLE III.

MIXED DEAFNESS

Otosclerosis and C.S.O.M.	36
Otosclerosis and Perceptive Deafness	41
C.S.O.M. and Perceptive Deafness	87

In some cases of otosclerosis there was evidence from the speech audiogram that was suggestive of a defective cochlea and so these were included under the heading of Mixed Deafness. In a few others the effects of acoustic trauma or of Ménière's Disease was added to the otosclerotic deafness.

Thus we find that from among 2,000 adults attending a centre for the purpose of being tried out with a hearing aid, no less than 944 or 47 per cent. had otosclerosis.

I will not add figures from my own practice, as they will almost certainly be biased in favour of otosclerosis. The numbers just given do not have any such bias as they do not include any patient seeking operative treatment, and so they can be regarded as a representative cross-section of the adult deaf who wish to try a hearing aid.

As regards the incidence of otosclerosis in the general population, there are the findings of Guild in his study of 1,100 pairs of temporal bones from patients who had died in the Johns Hopkins Hospital, Baltimore. In one-half per cent. of the subjects he found otosclerotic bone with involvement of the stapedio-vestibular articulation and deafness during life. In as many as 5 per cent. there was histological evidence of otosclerotic bone, but as the movement of the stapes was in no way impeded, there had been nothing to suggest the presence of otosclerosis during life.

He found that foci of otosclerotic bone were many times more common in temporal bones of white subjects than those of negro subjects; and as his material was almost equally divided between whites and negroes, it is probable that the incidence among white subjects is higher than the percentage quoted. Indeed Engstrom (1939) and others have found the incidence of foci of otosclerotic bone in their series of temporal bones to be 10 per cent.

In England and Wales, Wilkins (1949) of the Social Survey estimated that 4 per cent. of the adult population was sufficiently hard of hearing not to be able to hear more than tête-à-tête conversation.

This is a little higher than the 3 per cent. prevalence rate of deafness quoted by Stocks (1949) in a Study of Sickness in the population of England and Wales in 1944-47 which was also based on Social Survey findings.

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If these figures of the prevalence of deafness are considered in the light of our knowledge of Guild's work and of the findings at a hearing aid clinic, then we are forced to conclude that otosclerosis is the commonest cause of deafness in adult life, and that its incidence in the adult population in general cannot be less than ½ per cent.; indeed it may be considerably higher.

I have been able to trace adequate records of 2,000 cases of otosclerosis which have been seen in my clinics and practice during the past few years. Of these, I have personally examined 1,860, the remainder being patients seen in clinics where I work.

Sex Incidence

TABLE IV.

Male	649 (32·5%)
Female	1,351 (67·5%)

Otosclerosis is found more than twice as frequently in women as in men, and even if those cases in which otosclerosis is initiated or made worse by pregnancy are excluded (264) the proportion of females is still 62 per cent.

Age at Onset

TABLE V.

Age Group in Years	Number of Cases
0–10	37
11–20	545
21–30	806
31–40	431
41–50	58
51–60	19
61+	3
No Record	101

The age at onset of deafness due to otosclerosis is not always easy to determine, because the onset is usually gradual and one ear may be affected before the other.

In this series, however, every effort has been made to find out the time at which any difficulty in hearing with either ear was first noticed. The figures in the above table bear out the general impression that the onset of otosclerosis is during the active and growing period of adolescence and early adult life. They reveal that in 70 per cent. the deafness was first noticed between the ages of 11 and 30.

Effect of Pregnancy

TABLE VI.

Deafness initiated or made worse by pregnancy	264
Deafness not affected by pregnancy	155

Deafness due to otosclerosis is sometimes initiated or made worse by pregnancy. In this series 63 per cent. of those who bore children found

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that their hearing deteriorated during the later months of one or more of their pregnancies. In some the hearing was affected after each child, while in others it was affected after one child only, there being no regular pattern of behaviour other than the fact that there is a two-thirds chance that the hearing will deteriorate during the last two months of any pregnancy; and this deterioration may continue during the puerperium.

Paracusis

TABLE VII.

Paracusis present	1,560
Paracusis formerly	35
Paracusis not noted	307
No Record	98

The ability to hear speech more easily in noisy surroundings is, of course, a characteristic feature of conductive deafness, and it is commonly encountered in otosclerosis. In this series it was noted in 84 per cent. Doubt has been expressed about a diagnosis of otosclerosis in the absence of paracusis, but its presence, though usual, is not essential for the diagnosis.

Tinnitus

TABLE VIII.

Tinnitus present	1,570
Tinnitus not present	289
No Record	141

Tinnitus is a usual accompaniment of otosclerosis, though as in paracusis there were 16 per cent. who denied ever noticing tinnitus. It was interesting to find that a majority of those with tinnitus did not find it to be unduly disturbing; and the usual reply from patients to the question of whether the noises were troublesome, was that they were used to them. In a former review of 1,100 cases of otosclerosis, all of which are included in this series, 79 per cent. did not find the noises troublesome. Among those who found the noises to be disturbing was a high proportion of patients with a nervous temperament, Cawthorne (1951).

Family History

TABLE IX.

Positive Family History		1,053
	Paternal	464
	Maternal	448
	Sibling	141
No Family History		886
No Record		61

In 54 per cent. of this series enquiry disclosed that one or more members of the family suffered from a similar kind of deafness, there being a slight preponderance in favour of the father or one of his relatives. In 13·4 per cent. a sibling only was affected. I have found that in the

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more severe form of otosclerosis associated with a pink-tinged tympanic membrane there is often a strong family history of otosclerosis.

I have records of some 30 patients, none of whom have been included in this series, who gave a history of familial deafness, but who on testing appeared to have perceptive deafness. In some instances it has been possible to examine and test other deafened members of the family as they have always responded to the tests as though they were perceptively deaf.

Such cases may have given rise to a diagnosis of cochlear otosclerosis, and I am indebted to my friend Dr. George Shambaugh, Jnr., who kindly drew my attention to an article by his father, Shambaugh (1926), who mentioned such cases. It may well be that these patients have an unusual form of otosclerosis, possibly affecting the cochlea only, but as they do not show any sign of conductive impairment I feel that they should be regarded as having a familial form of perceptive deafness until we have histological evidence to the contrary.

Vertigo

TABLE X.

Vertigo noted	462
Vertigo not noted	1,432
No record	106

A history of vertigo, usually slight and momentary when stooping, was given in 24 per cent. of the cases in this series. As most of the subjects were under 40 I would say that this is in excess of what might be found in the normal population. It was not a constant feature, and it could not be induced at will. I believe that this increased liability to momentary dizziness on bending may be due to the stiffness of the stapes, preventing normal adjustment of the intralabyrinthine fluids to alteration of position. Occasionally in otosclerosis a patient will volunteer the curious information that he notices dizziness when eating. Here again this could be explained by intratympanic pressure changes caused by contraction of the tensor tympani being transmitted in an irregular manner to the labyrinthine fluids.

Other Ear Disease

TABLE XI.

Evidence of other ear disease	160
No evidence of other ear disease	1,813
No Record	27

There was evidence of other ear disease in only 8 per cent. of the cases. Only those patients with active ear disease in one ear, while the other normal looking ear exhibited all the features of otosclerosis, were included in this series, and some of the patients excluded from this series had otosclerosis as well as another bilateral ear disease.

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Otosclerosis

One feature in the history of patients with otosclerosis, which does not agree with some of the older views on the disease, is how remarkably healthy these patients seem to be. It is unusual to learn that they have had more than usual childish ailments, and the occasional head cold and, of course, more often than not the ubiquitous catarrh, particularly if they are smokers. In former days gout, syphilis and scrofula were faithfully trotted out as ætiological whipping boys whenever a disease of unknown ætiology such as otosclerosis was discussed. The modern equivalents are I suppose, avitaminosis, hormonal imbalance and of course stress, but I have seen and heard nothing to suggest that such factors are to blame any more than the earlier ones were.

Finally in the history there has been nothing to suggest neglect or lack of treatment, often the reverse, but despite all forms of attention to the eustachian tube, the ear drum and the upper respiratory tract, the deafness has pursued its course uninfluenced by any of these measures.

Cerumen

TABLE XII.

Evidence of cerumen	1,466
No evidence of cerumen	191
No Record	343

It has often been said that sufferers from otosclerosis do not form cerumen so readily as normal hearing persons, and also that the external meatus is relatively insensitive. These phenomena have been attributed to the otosclerotic process which in some way has led to defective circulation and atrophy in the skin lining the external meatus.

I do not think that in otosclerosis there is any deficiency in the cerumen secreting glands, nor do I believe that the external meatus is less sensitive than in normal persons. In 88 per cent. of this series there was evidence of active ceruminous glands, either in the shape of cerumen or a history of having had the cerumen removed.

It is quite true that one does not have to spend much time in removing cerumen from the ears of patients with otosclerosis, nor do such patients often complain of discomfort, but these phenomena can be explained on the grounds that the first thing that happens in deafness due to otosclerosis is that the patient has his ears syringed for wax by his family doctor. This and other manipulations are carried out, perhaps repeatedly, before the patient finally appears before the otologist. By this time not only will most if not all the cerumen have been removed, but also the patient will have become accustomed to having the ear examined and will not find it so unpleasant. Touch corpuscles that are rarely stimulated take a little time to get used to it, as all can remember from the days when as children we had our finger nails cut and for a few minutes afterwards the freshly exposed skin was very sensitive to touch.

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Tympanic Membrane

TABLE XIII.

Normal (including 208 pink-tinged)	1,684
Abnormal	310
No Record	6

Included under the heading of Abnormal Tympanic Membrane are any deviation from the normal lustre and light reflex of the drumhead as the result of external otitis, scars, calcareous patches or other opacities, any interference with the normal mobility of the drum, and even any ballooning of the posterior part of the drum as the result of excessive auto-inflation.

Some of these have already been included under other ear disease, but many slight abnormalities of the drumhead have been noted without any other history or sign of disease except otosclerosis. It is sometimes thought that a small scar on the drum may play some part in the deafness but, as Parnell (1951) showed in an audiometric survey of student hearing this need not be so as he had found several subjects with normal hearing despite a scar in the ear drum.

No case of adhesive otitis has been consciously included in this series. There were, however, two patients thought to have otosclerosis but who at operation were found to have adhesive otitis. In each the tympanic membranes had a uniform milky appearance. In all the other cases in this series submitted to operation, and this is about 1,400, no significant change in the tympanic structures was seen other than an occasional deposit of otosclerotic bone or excessive vascularity of the mucous membrane. The pink-tinge imparted to the tympanic membrane from the inner tympanic wall was present in just over 10 per cent. of the cases. In some it was a faint flush just behind the handle of the malleus, whilst in others the whole membrane appeared aglow, and could have been mistaken for an acute myringitis, if it had not been for the normal sheen and light reflex on the outer surface of the drum. At operation this appearance is found to be due to dilated vessels in the mucous membrane covering the promontory.

In some cases, usually with advanced deafness, a whitish mass could be seen through the tympanic membrane behind the handle of the malleus, and it may be that this appearance has given rise to the observation that the ear drum is thin and atrophic. I believe that in such cases the drum is normal both in texture and in appearance and that what is seen is some abnormality on the inner tympanic wall, possibly some enlargement by otosclerotic bone of the lip of the round window niche. I say this because for the past year I have been noting the appearance of the round window niche at operation and in some it is undoubtedly thickened and can give the picture of a whitish mass on the inner tympanic wall.

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Distribution of Deafness

TABLE XIV.

Unilateral	71
More than 20 dbs difference between ears	165
Symmetrical	1,747
No Record	17

The deafness is usually symmetrical, there being less than 20 decibels difference between the threshold of hearing for pure tones in the two ears in no less than 88 per cent. of this series. Unilateral otosclerosis was diagnosed in 3·5 per cent. in this series and that this can occur is known from the pathological studies of Nager, Guild, Siebenmann and others. Hoople in a recent paper found the incidence of unilateral otosclerosis as high as 10 per cent. and this may be nearer the true incidence of unilateral disease, for in this country I am quite sure that a considerable proportion of persons with unilateral deafness never consult an otologist.

Hearing Tests

I have always regarded tuning-fork tests as being very helpful, not only in deciding upon the type of deafness, but also in trying to gauge the extent of the disease. The results of such tests were given in a former series of 1,100 cases, Cawthorne (1951). A negative Rinne for all forks up to and including 2,048 cps. suggests limited disease without complete fixation at the oval window and with, of course, the activity of the cochlea unimpaired. Routine bone conduction audiometry was not practised in the earlier cases, but it is now always carried out, and it can give valuable information. Shambaugh and Carhart have shown that the bone-air gap offers a useful guide to the prospects of improvement by a fenestration operation. Speech audiometry is an important and informative part of the routine examination which helps to complete the picture.

Finally, comes a live voice test with the patient using a hearing aid, and for this I use a Medresco Mark IIb or III aid. If a patient with otosclerosis can, in a reasonably quiet room, understand conversation easily with the aid, I regard this as further evidence of the integrity of the cochlea.

If a patient can hear sounds with the aid, but is unable to distinguish the meaning of the sounds, then the possibility that the cochlea is affected must be considered. If the patient cannot hear with the air-conduction aid, then a Medresco bone conduction aid is tried. Here again, if speech appreciation is good, an intact cochlea is assumed.

Impairment of hearing by air and bone conduction both for pure tones and speech is of course found in perceptive deafness from a cochlear defect. But I believe that it can also happen in pure conductive deafness with normal cochlear function if the sound is prevented from reaching the cochlea by both labyrinthine windows being sealed off as may happen in widespread otosclerosis.

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Classification of Otosclerosis

Now this leads to the classification of otosclerosis, and here it is helpful to try and think in terms of what is happening to the bony capsule of the labyrinth.

We know that the earliest sign of otosclerosis comes when new spongy bone starts to bridge across the ligament between the margin of the oval window and the stapes footplate. This is nearly always at the anterior end and, at first, may only cause some stiffness of the joint with sight impairment of hearing. If and as the bony bridge extends and becomes wider and firmer, so the stiffness of the stapes hinge-joint increases and the deafness deepens. In other words, the amount of deafness depends upon the stiffness of the stapes. You will note that, so far, I have avoided the word "fixation" of the stapes and I have done this for a very good reason. The term "fixation" as applied to a joint, means immobility; so does ankylosis. But are we sure that the stapedio-vestibular joint is absolutely fixed and immobile in otosclerosis? That it can be I know, because on two recent occasions I have found at operation the stapes more than half buried in otosclerotic bone. But then these two patients were profoundly deaf. I prefer to think of the stapedio-vestibular articulation being stiffened by bony deposits in the joint and in the footplate, so that its free movement is impaired rather than fixation or ankylosis, both of which terms imply immobility.

In the Wernher Research Unit, we are at present engaged in investigating the stiffness of the stapes in otosclerosis. Until we know something more about the state of the stapedio-vestibular articulation, I prefer to avoid such absolute terms as fixation and ankylosis. Instead I use the word stiffness.

The deafness in otosclerosis is more often than not progressive, though as a rule, the rate of progress is often slow. In patients who have had one ear operated upon, it has been interesting to find that in some, the deafness in the unoperated ear has remained stationary over several years. It is well known that pregnancy may reactivate the disease, and it is possible that it may be stimulated to spread after a big operation or accident. Apart from this, I cannot think of any other activator of the process; and though it is said that in women the menopause may have this effect, I have not found this happen often enough to make it significant. Thus when we come to make a diagnosis of otosclerosis I think that it is of help to try and picture what is actually going on in the labyrinthine capsule and to use one or more qualifying terms such as limited, extensive, widespread, active or quiescent, and I hope that the day is not far distant when we can also say whether the oval, the round or even both windows are affected.

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Otosclerosis

Pathology

Otosclerotic bone, which is only found in the bony capsule of the labyrinth, consists of patches of new spongy bone, containing wide Haversian canals, large medullary spaces with abundant blood vessels, the whole of the affected area staining dark blue with hæmatoxylin eosin. It is thought by many that this excess of new bone is there to replace bone previously absorbed, but others believe that the new bone erodes the old by pressure and infiltration. In some areas Manasse has described finger-like processes of bluish staining bone, each surrounding a blood vessel. These blue mantles of Manasse probably represent the earliest stage of the disease, and Nager (1927) believes that this indicates that the activating agent, whatever it may be, arrives in the bony capsule of the labyrinth *viâ* the blood-stream. This otosclerotic bone may appear in any part of the labyrinthine capsule and in a few unfortunate cases may almost envelop the cochlea; but in the large majority of cases the first focus will be found just in front of the anterior margin of the oval window and in many this will be the only focus. This is the site of election for otosclerotic bone.

New otosclerotic bone does not in itself cause deafness, for Guild showed that for every case of histological otosclerosis with deafness due to involvement of the stapedio-vestibular articulation, there were eight others in which the stapes was quite free and in whom there had not been any sign of otosclerosis during life. Once, however, that the otosclerotic bone has bridged across the annular ligament of the stapes, a patch of otosclerosis no larger than a pin's head is sufficient to cause deafness. Bengt Nylèn (1949) in an interesting study of Nager's specimens of otosclerosis found that in 90 per cent. of the specimens the region of the fissula ante fenestram was affected by otosclerotic bone whilst the round window region was affected in 40 per cent.

Now Bast and Anson in studying the development of the bony labyrinth paid particular attention to a small fissure in front of the oval window because they believe it is here that otosclerosis usually originates. Perozzi (1916) named this Cozzoloni's zone after his uncle and chief, but the descriptive term fissula ante fenestram applied by Bast and Anson is the one in general use. They observed this fissure through fœtal life, and though in the fully developed bone it is constantly present, its size and extent are subject to wide variations.

They found this fissure to be occupied by cartilage and fibrous tissue, the cartilage varying somewhat in the regularity of its formation, sometimes being irregular and knarled.

They believe that the available evidence favours the hypothesis that the otosclerotic process starts in this cartilaginous rest in the fissula and that it may well be more likely to occur when the cartilaginous formation is irregular. On the other hand Guild noted that in some bones in his

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collection the fissula ante fenestram was normal despite otosclerotic bone elsewhere in the capsule of the labyrinth. Nevertheless the available evidence suggests that an unstable cartilaginous rest in the fissula is the starting point in most cases of otosclerosis. (Fig. 1.)

Now it is known that this disease often runs in families and just as we inherit many anatomical features, such as the colour of our hair and our eyes and our facial structure, from our ancestors, so may a wide fissula ante fenestram with irregular cartilage formation also be inherited.

Let us now turn to the bony capsule of the labyrinth as a whole. It is unique in that it is the only part of the bony skeleton to be fully developed at birth and normally no further growth of bone takes place

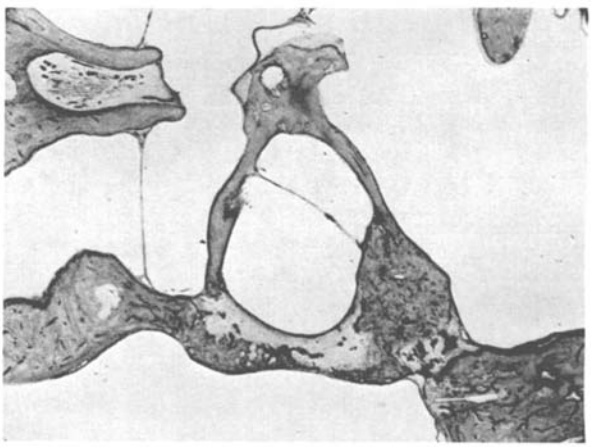


FIG. 1.

× 10. Showing stapes with otosclerotic bone involving the stapedio-vestibular articulation. Though the main focus of otosclerotic bone is at the site of election in front of the oval window, the fissula ante fenestram can be clearly seen and in this section is free from otosclerotic bone. This is from a woman aged 33, who had a fairly severe conductive deafness who committed suicide. Sections prepared at the Ferens Institute.

in the bony labyrinth. The reason for this lies in the thick middle enchondral layer of the bony capsule of the labyrinth which is relatively avascular and not capable of laying down new bone. This is seen in fractures through this enchondral layer which are said never to heal.

Here then is a bone which is the only bone in the body which is fully developed at birth; and which in normal circumstances does not have further bone added to it even during times when new bone is being laid down in cartilaginous epiphyses, or in other circumstances where cartilage is being transformed into bone. Such circumstances are found during adolescence and during the latest months of pregnancy, and there surely must be at these times some form of bone growth promoting substances circulating in the blood stream. That the exciting agent in the production

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of otosclerotic bone is blood borne is suggested by the blue mantles of Manasse, the earliest manifestation of the disease which, as has already been noted, follows the blood vessels. I believe that Siebenmann (1899) was right when he said that otosclerosis is nothing more nor less than an attempt by the body to add bone to the labyrinth, an attempt that though normal in every other bone is abnormal for the labyrinthine capsule.

I would also recall the fact that otosclerosis is usually found in people who are in all other respects remarkably healthy. All this suggests to me that the otosclerotic process is not so much a disease as an exaggeration of the normal.

As regards the prevention of otosclerosis it seems to me that the only



FIG. 2.

× 200. Cupula from external semicircular canal in an advanced and widespread case of otosclerosis showing normal and otosclerotic bone. Sections prepared at the Wernher Research Institute.

way to do this would be to prevent bone growth generally; and if this were possible then those with large fissulae might have to choose between being deaf or being dwarfs. Why in some cases the process is limited to one small area while in others it spreads at times widely over the labyrinthine capsule, and once having started what makes it stop are important questions to which at present we have no answer.

Since the introduction of the fenestration operation it has been possible to study the pathology of otosclerosis in the living. Simson Hall (1954) found evidence of otosclerotic bone in some 14 per cent. of the bone specimens removed from the lateral semicircular canal at operation and I can now say that since we have been examining such bone we have found otosclerosis in a similar proportion. (Fig. 2.)

More recently, however, we have been interested in the round window

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niche and one of our workers in the Wernher Research Institute, Dr. Aronson, is studying the anatomy of this region. We have been carrying out a certain number of operations on patients with very profound deafness due to otosclerosis and we have found that in some the round window niche has been partly blocked by new bone formation, and furthermore chips of bone from this region have been found to contain otosclerotic bone. (Fig. 3.)

Mechanics of the Obstruction in Otosclerosis

Before dealing with these and other cases I would like for a few moments to discuss the mechanical problem presented by otosclerosis and what I have to say about this is inspired by the work of Wever and

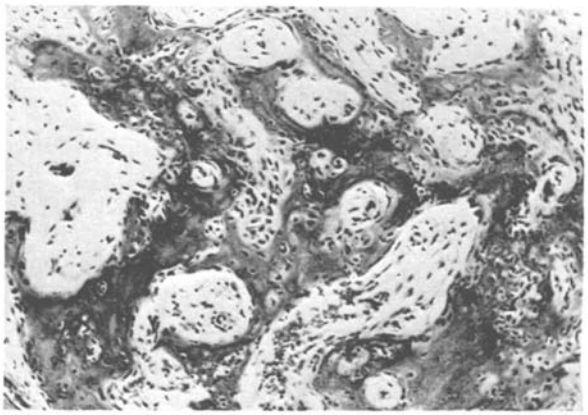


FIG. 3.

× 200. Fragment of bone from round window niche in a case of advanced otosclerosis which at operation appeared to be partly filled with new bone formation.

Lawrence which is fully reported in their recent book *Physiological Acoustics*.

Normally sound waves enter the labyrinth by the oval window from whence they travel in a fluid medium being drawn, so to speak, by the action of the round window into the cochlea and across the basilar membrane thus stimulating the hair cells of the organ of Corti. This action calls for two mobile windows, one on the vestibular and the other on the tympanic side of the basilar membrane. In this way a movement of the stapes footplate in the oval window sets up a movement of the fluid inside the bony labyrinth. Instead of being dispersed all over the labyrinth this is encouraged to enter the scala vestibuli of the cochlea as this is the path of least resistance. Once in the scala vestibuli the wave can travel along the coiled cochlear tube or take a short cut across the membranes which separate it from the scala tympani. It is expended on the round

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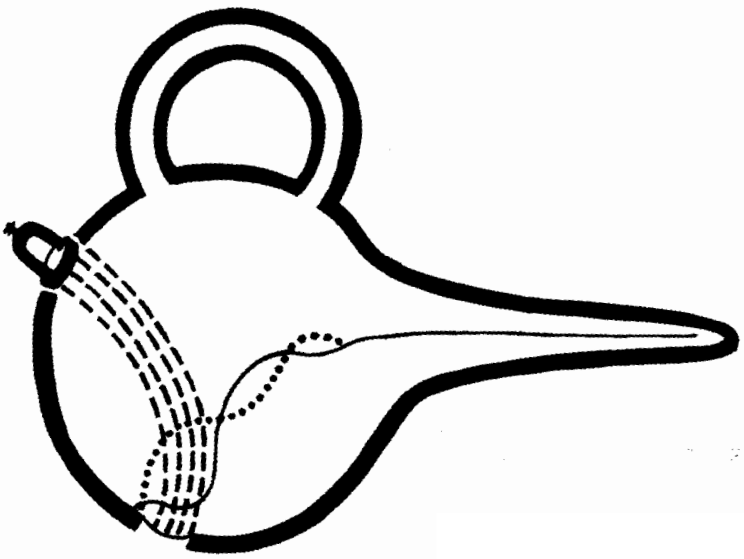


FIG. 4.

Diagrammatic representation of the cochlea, vestibule and lateral canal, showing the stapes, round window and basilar membrane (after Wever). This diagram depicts the normal mechanism whereby the sound wave travels through the labyrinthine fluids so as to effect the optimum movement of the basilar membrane.

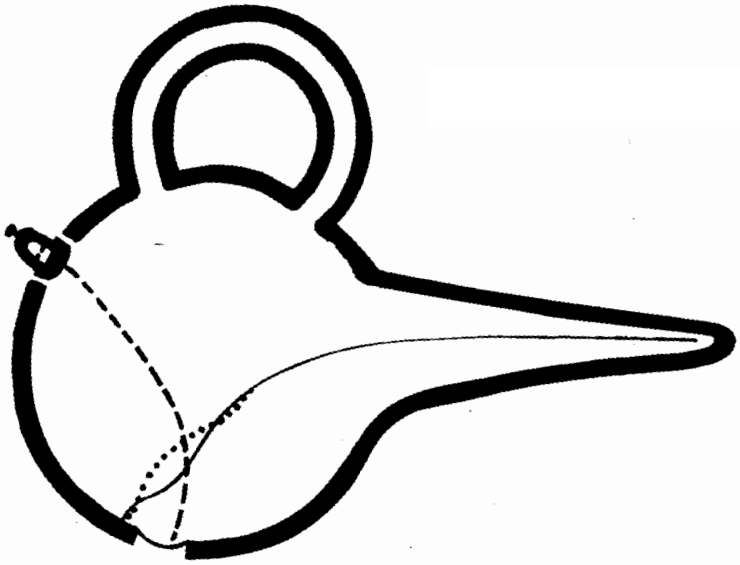


FIG. 5.

Diagrammatic representation of the cochlea, vestibule and lateral canal, showing the stapes, round window and basilar membrane (after Wever). In this diagram the stapes is stiff, though not fixed, as in early otosclerosis. The resistance of the stiffened stapes to sound waves trying to enter the labyrinth results in a smaller fluid wave in the labyrinthine fluids and in consequence a smaller excursion of the basilar membrane.

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window at the end of the scala tympani and in this way the reciprocal action of these two windows encourages the to and fro movement of fluid in the cochlea that is essential for stimulation of the hair cells of the Organ of Corti (Fig. 4). If one window does not transmit sound waves freely then the balance is upset and some degree of deafness results. I favour the view that the degree of the deafness depends upon the extent of the impairment of mobility at the window. If the impairment is slight then the deafness will be slight. If the mobility is greatly reduced so will the hearing be greatly reduced. If there is no movement at all then it may be that there is little or no hearing at all (Fig. 5). When one window is blocked hearing can be restored if a new window on the same side of the basilar membrane as that which was occluded is provided (Fig. 6). To provide two mobile windows on one side of the basilar membrane would result in all the movement inside the labyrinth taking place on one side only of the basilar membrane. This would cause little effective movement of the basilar membrane and little or no improvement in hearing. What is much worse two mobile windows on the vestibular side of the basilar membrane might provide a vestibular stimulus (Fig. 7). Furthermore things must be so arranged that the pressure from incoming sound waves falls on and presses in one window, the other being shielded from the incoming wave and so free to move outwards. Otherwise if the incoming sound wave is allowed to fall equally on each window so that both are pressed inwards at the same time then very little of this energy will be able to get inside as the outgoing movement of each window will cancel the other out.

Early attempts at providing a new window failed because these simple physical principles were not appreciated. Recent work by Skoog (1951) and his associates has emphasized the importance of this "Differential Sound Pressure" between the two labyrinthine windows.

The Fenestration Operation

The fenestration operation is now firmly established as the only way in which natural hearing can be restored to a patient whose deafness is caused by otosclerotic bone interfering with the movement of the stapes. Jenkins (1913) was the first to report the results of making an opening into the bony lateral semicircular canal and when discussing Jenkins's paper Bárány (1913) described how in 1910 he had tried an opening into the posterior canal. Holmgren (1939) tried the superior canal but later changed to the lateral canal, and his pupil Sourdille (1938) first conceived the idea of a procedure which left the fenestra in direct communication with the air, so that air borne sound waves could act directly upon it. This procedure called for a series of operations spread over several months.

In 1938 Lempert introduced his one stage fenestration operation which was further modified in 1945 so that the fenestra lay over the ampulla of the lateral semicircular canal, the incus and head of the malleus

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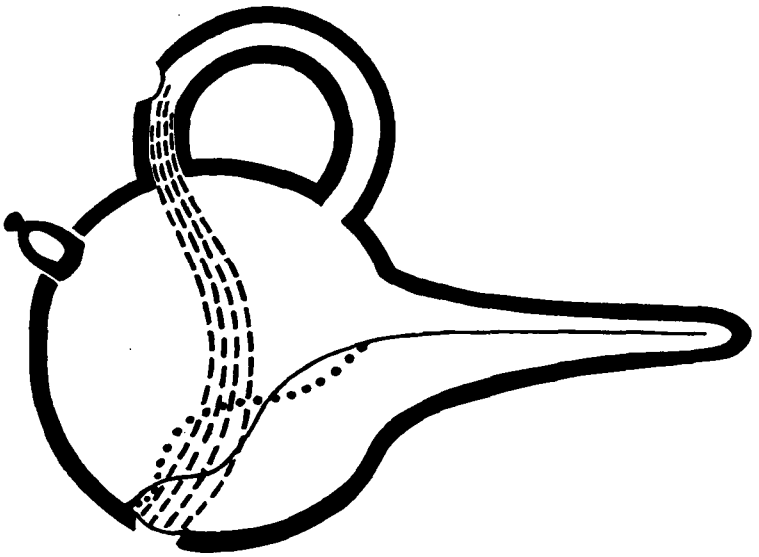


FIG. 6.
Diagrammatic representation of the cochlea vestibule and lateral canal, showing the stapes, round window and basilar membrane (after Wever). In this figure the stapes is stiffened by otosclerotic bone, and a fenestra has been made in the lateral semicircular canal, through which sound waves are able to enter the labyrinth on the vestibular side of the basilar membrane and are conducted across the membrane to the round window on its tympanic side. The success of the fenestration operation depends upon a mobile window on each side of the basilar membrane.

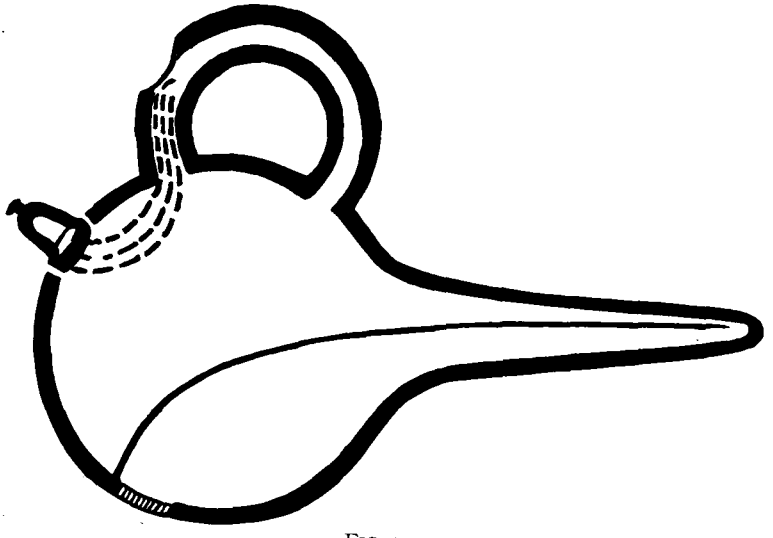


FIG. 7.
Diagrammatic representation of the cochlea, vestibule and lateral canal, showing the stapes, round window and basilar membrane (after Wever). In this figure the round window is covered over by otosclerotic bone but the stapes is free to move. Therefore, a fenestra in the lateral semicircular canal results in a movement of the labyrinthine fluids on the vestibular side of the basilar membrane only. This does not improve the hearing and may result in a vestibular response to a sound stimulus or the Tullio phenomenon.

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were removed, and the fenestra was finally covered by a tympano-meatal flap taken from the deep meatal wall and continuous with the tympanic membrane.

This now-ovalis technique introduced by Lempert has been generally adopted throughout the world with of course minor variations and in skilled hands it results in permanent improvement in hearing in at least three-quarters of the cases chosen for operation. To those whom I have had the pleasure of training in this work I always suggest that if they are unable to get a permanent improvement in hearing in at least 50 per cent. of the cases they operate upon then they should either revise their technique or abandon the operation.

I would like to say something about the possibility of closure of the fenestra, because I think that it is still generally felt that this is often liable to happen. In my first 850 operations it happened in 8 per cent. of the cases. The main problem in fenestration is still the group which despite an adequate operation fails to improve. This has happened in 13 per cent. of my cases and much of my time now is being taken up by enquiring into the cause of these failures. Among these is a smaller group in which the hearing is at once worse after the operation and there is troublesome vertigo though the labyrinth remains active and the fistula sign is brisk. It is possible that these may be cases in which the main block was in the round window, and that by providing two mobile windows on the vestibular side of the basilar membrane the hearing was made worse and post-operative vertigo was unduly troublesome.

Tullio Phenomenon

Tullio described how pigeons with an artificial opening into the bony vestibular labyrinth gave a vestibular response to a sound stimulus.

This Tullio phenomenon has also been noticed in man and in 1949 I described a case of Ménière's Disease in which an opening had been made into the bony lateral semicircular canal without however affecting the membranous labyrinth. After recovery from the operation the patient experienced sudden giddiness when exposed to nearby loud sounds. At examination it was possible to provide eye and body movements by sounding a loud pure tone in the operated ear. This was attributed to the presence of two mobile windows (fenestra and oval window) in the vestibular part of the labyrinth.

I have seen three patients in whom a fenestration had been carried out for perceptive deafness—that is to say they had a freely mobile stapes. In each case there was such troublesome vertigo after the operation that the labyrinth had to be subsequently destroyed. In two of these a Tullio phenomenon was present.

Now why is the Tullio phenomenon not seen more frequently after the fenestration operation. It may of course be said that if the sound

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stimulus be powerful enough there will be some vertigo. But generally speaking there is no dizziness on exposure to sound, and the reason for this is that the stapes is not free to move. If, however, the otosclerotic bone has fixed the round window and left the oval window free, or at any rate comparatively free to move then I have found there is troublesome vertigo and no improvement in hearing. I am at present investigating more closely this small but unfortunate group of cases.

While in the present state of our knowledge it may not be possible always to avoid operating on such cases, it is certain that the fenestration should not be done if there is any reason to suppose that the normal movement of the stapes is not impeded.

Operative Findings

At operation the middle ear and its contents rarely look anything but normal. Occasionally with the help of the microscope it is possible to see whitish opacities in the normally translucent footplate of the stapes, and in about 1 per cent. of cases obvious bone growth can be seen. The stapes does not move readily, but with magnification it is sometimes possible to see slight movements of the footplate in response to gentle pressure on the head of the stapes. It will of course be appreciated that in ordinary circumstances the excursions of the ossicular chain in response to sound stimulation are not very great and indeed would scarcely be visible to the naked eye. Probing the stapes needs to be done gently as the crura very easily fracture away from the footplate and indeed this can be regarded as evidence that normal movement of the footplate is impeded.

I have found otosclerotic bone in the lateral canal with a fixed stapes. I have also found otosclerotic bone in the round window niche with a fixed stapes. I have now found otosclerotic bone both in the lateral canal and in the round window with a fixed stapes. I wonder if I shall ever find otosclerotic bone in the round window with a mobile stapes. If I do then I shall not be surprised if the patient comes within that small group about whom I spoke earlier in which the hearing is worse and the giddiness is troublesome. Where we are to make the new window in such cases remains to be seen, though I have on a recent occasion made a new round window in a patient in whom both windows were entirely covered over with otosclerotic bone which had spread all over the capsule of the labyrinth.

Conclusions

Our knowledge of otosclerosis has been greatly increased since the introduction of the fenestration operation which now enables hearing to be restored to the majority of those selected for operation. By studying the mechanics of the disorder and by improving testing procedures it may

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be possible to restore hearing to some of those who at present cannot be helped by an operation.

Finally may I say how grateful I am to all those of my friends and colleagues who have entrusted their patients to me, to all my friends at King's College Hospital and in the Wernher Research Unit on Deafness who have done so much to make this work possible and in particular to Mr. A. B. Hewlett who drew the diagrams and who has helped me in all stages of this work.

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