SEVERE OTOSCLEROSIS

By RORY WILLIS (Melbourne)

WITH the introduction of the operation of Stapedectomy, the ear surgeon is given a wonderful opportunity of converting almost at one stroke a deaf patient into a normal hearing person. We now take this minor miracle as an everyday occurrence, yet there can be few situations elsewhere in medicine or surgery that can compare with it.

The ordinary otosclerotic patient presents no problems in either diagnosis or indications for surgery. Most cases have a gradually increasing pure conductive deafness until some 50 decibels are lost, and then some of them will show a very slow drop in the cochlear function beginning at the high tones. We will advise surgery to cure the deafness provided the bone threshold is not below 30 decibels in the speech frequencies, and we will confidently expect to close the air/bone gap and the patient will no longer have to use his hearing aid.

The abnormal otosclerotic patient on the other hand may only show 30 to 40 decibels of conductive loss at any stage, but rapidly and apparently simultaneously shows a severe fall in the cochlear function. I am sure that this form of the disease is often misdiagnosed as something other than Otosclerosis, and I admit that the diagnosis is impossible to make with certainty before operation in many of them.

The type of case I want to discuss here is the one that presents with hearing as in Fig. 1.

The air conduction threshold is at the very bottom of the graph, only the deep tones being heard at all, and a hearing aid is virtually useless. Bone conduction may be appreciated at the limit of the audiometer in the low frequencies. Often the only clue to the otosclerotic nature of the deafness is the negative Rinne, but since it is only for the low tones, one cannot be sure that it is true hearing. It may be simply feeling.

Fig. 2 shows what the hearing was like 10 years before the test in Fig. 1 was taken, and Fig. 3 shows what it was like 6 years earlier still.

Studying these three graphs makes the diagnosis of the otosclerotic nature of the patient's deafness quite easy, but one may have only one test to go on, as in Fig. 1. Naturally a very careful history is taken, and evidence of family history, the progressive nature of the deafness, and the presence of paracusis in the early stages, all make the diagnosis of otosclerosis easier. A very useful point is that patients with conductive

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deafness are good hearing aid users, and will cling to their aids even when very little sound is perceived through them. This is in distinct contrast to the patient with pure inner-ear deafness.

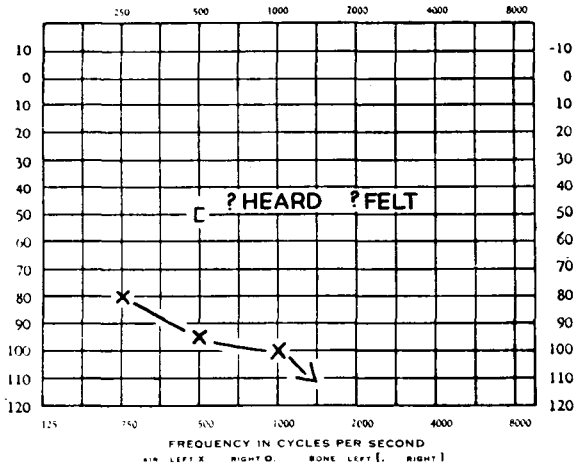


FIG. 1.
Severe Otosclerosis: Mr. A. aged 39 in 1960.

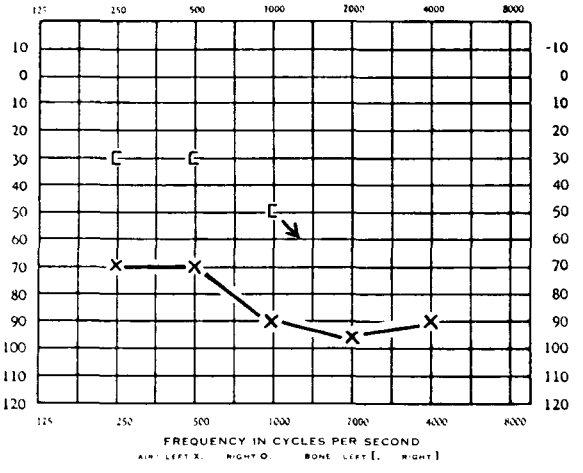


FIG. 2.
Severe Otosclerosis: Mr. A. aged 29 in 1950.

To try to overcome the question of whether a patient is truly hearing or only feeling vibrations through the bone has been one of the major tasks in this field. So-called "vestibular hearing", in which the balance

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end-organs in the utricle and saccule are known to appreciate low tone vibration transmitted through the temporal bone is a confusing state. Bocca and Perani, 1960, quote cases to support the theory that such a function does exist for frequencies up to 500 cycles per second, and show that an apparent air-bone gap of 30 decibels may be found. However, above 500 cycles the air/bone gap disappears and we have the features of a pure inner-ear deafness. They have no operative observations to exclude middle-ear disease.

I believe that this condition of "vestibular hearing" does exist and

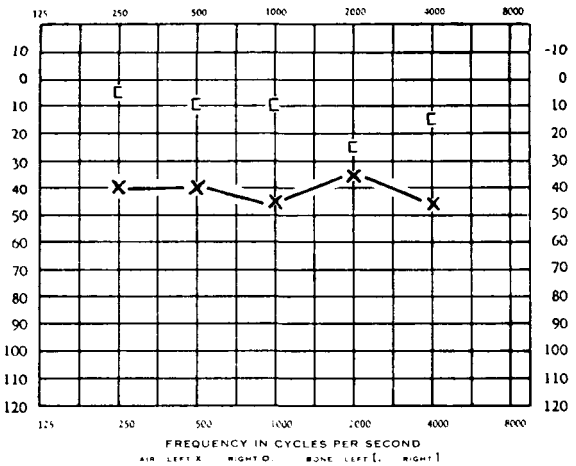


FIG. 3.
Severe Otosclerosis: Mr. A. aged 23 in 1945.

I think that they can be fairly easily excluded because the air/bone gap is small, and because, being truly inner-ear deafness they do not show the very rapid deterioration seen in the severe otosclerotic cases. An occlusion test is also useful to distinguish a nerve deafness from one with a middle-ear element. In a pure nerve deafness, if a 250 fork is sounded on the mastoid until the patient can no longer appreciate it, and then with the fork still in place the ear under test is occluded with the finger tip, in effect producing a negative Rinne, the fork will again be heard for a short period. On the other hand if there is a middle-ear deafness due to any cause, the patient already has an in-built occlusion and you cannot prolong the fork by blocking the ear. Many variations of this test are possible using the fork or audiometer bone conduction unit on the vertex of the skull.

Stimulated by our demands for more accurate diagnosis before undertaking surgery, several new approaches to bone conduction audiometry

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have been instituted and are currently being investigated at the Commonwealth Acoustic Laboratory in Melbourne. These are as follows: (a) To determine whether the patient can discriminate between frequencies when applied through the bone using a sweep frequency oscillator, and (b) to determine whether the patient can appreciate the beats or pulses of overtones or harmonics, using two sources of sound from bone oscillators on the mastoid and on the vertex or forehead. Both are sounded simultaneously say at 500 cycles, and then one is thrown out of phase to produce beats. A modification is to use one at 500 cycles and the other an octave higher at 1,000 cycles, and again beats are produced when one is thrown out of phase. We feel that if a patient can describe these accurately then he really has a functioning cochlea, and is not merely feeling vibrations.

To go a step forward in the discussion of what causes the nerve deafness in otosclerosis, Sercer and Krmpotic, 1958, have demonstrated the presence of foci of otosclerosis in the internal auditory meatus occluding the small foraminae from which emerges the cochlea and vestibular branches of the VIIIth nerve. They maintain that interruption of the fibres at this point explains both the vertigo that accompanies this disease so severely at times, and also the bizarre patterns of nerve loss. As will be immediately appreciated, this also means that severe otosclerotic overall hearing loss does not necessarily mean a severe middle-ear lesion. This is well borne out in my own cases reported elsewhere (Willis, 1963).

In 1961, House (1961), in a very stimulating article discussed an approach to the internal auditory canal by a Temporal route which takes the surgeon on to the top of the petrous temporal. With a dental burr and under magnification, the greater superficial petrosal nerve is followed back to is origin from the geniculate ganglion of the VIIth nerve, and finally the internal auditory canal is opened. He mentions that this approach has been used to "decompress" the VIIIth nerve, and one can see the most interesting possibilities in it for the future surgery of severe otosclerosis.

We are beginning to appreciate one other cause for the loss of inner-ear function in otosclerosis. The cochlea is a sense organ that functions in a fluid medium, and as sound waves enter it this fluid medium is disturbed, and it could well be that some of the nutrition of the hair cells of Corti's Organ is dependent upon the movement of the fluid. If sound waves do not disturb it periodically then neural degeneration can occur, e.g. Carhardt's notch. This is of course all theoretical, but from a practical angle we have all seen in our stapedectomies the final hearing come up well above the pre-operative bone conduction threshold. Was our initial testing at fault? Was it being masked by the patient's own tinnitus, or did it in fact improve after the operation? In other words did the mobilization of the inner-ear fluid media improve the nutrition of the hair cells and hence the function of the inner ear? Whichever way it

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happens, it happens often enough to be significant, and one begins to hope that it will occur in all cases with a lowered bone conduction threshold.

It is immediately seen why the Stapedectomy operation becomes attractive in the management of severe otosclerosis, because it is often possible to get hearing not only up to the bone conduction but even above it. It is not possible to give these patients hearing without a hearing aid, but it must be realized that these are relatively young patients who have had the depressing experience of seeing themselves steadily drop out of the world of sound, getting stronger and stronger hearing aids, until finally they wear an aid only to advertise the fact that they are deaf, not for any sound it may bring them. Any noise, even the banging of a door, that you can restore to them will take them out of their silent world.

In the past 2 years, 28 patients with very severe otosclerosis have been operated upon by stapedectomy following the Shea technic of vein graft and polyethylene strut. Despite what I have already said about the difficulty of making the pre-operative diagnosis, there has been only one case where a mistake was made and a normal middle ear found at operation. All the others had well marked otosclerotic foci. It appears that males are more prone to this severe form of the disease, there being more males that females in this series, quite the reverse of the normal sex ratio for otosclerosis. All but one of the males had gross disease of the promontory and the footplate so that the oval window niche was completely obliterated, while the females often had a very thin footplate beneath the overhanging promontory lesion. This was a distinct trap and led to the only "dead labyrinth" of the series; the promontory lesion requiring extensive drilling to clear it, finally revealing a very tenuous footplate which became mobile at a touch and dropped away into the labyrinth. No attempt was made to retrieve it, the vein graft being placed over the top. Hearing came up for 3 weeks and then steadily dropped away over the next 6 weeks until no response to bone conduction forks was obtained.

Of the 28 proven otosclerotic cases, a significant hearing gain was obtained in 21, and all but one of these patients are using suitable hearing aids again, and all are adamant that the surgical attempt was well worth while. Three cases will be described in more detail to demonstrate the operative findings and the hearing result obtained.

Case No. I, Mrs. E. K., aged 59. History of progressive deafness since mid-twenties, with tinnitus and paracusis noted in the early stages. Rinne definitely negative. Pre-operative neither air nor bone readings were recordable.

Operation was carried out on October 4th, 1961, using endotracheal anæsthesia as is my usual practice. The incus was normal and mobile. The oval window niche was shallow, the stapes footplate being thin and

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blue. Otosclerotic fixation was of the annular variety and of minimal extent. Removal was easy with hooks, and an excellent vein graft and polyethylene strut inserted. Note that this very severe deafness had minimal disease inside; there is no way of predicting what you will find.

Post-operative course was uneventful and 5 weeks later audiogram showed air conduction threshold to be about 80 decibels average. This has been maintained over the 12 months and she is well adjusted to using her hearing aid again.

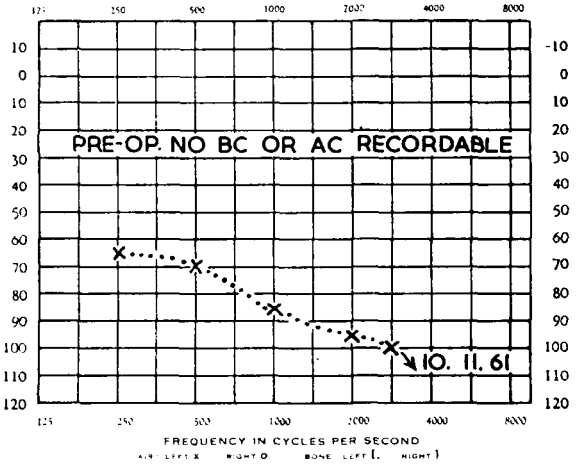


FIG. 4.
Case No. I; Mrs. E.K. showing post-operative hearing level.

Case No. II. Mr. C. L. aged 59. Again a history of progressive deafness, but unable to use his aid in the past few years. Pre-operative audiogram as in Fig. 5 shows one island of air hearing at 100 decibels at 500 cycles. Although Rinne was negative, bone threshold was beyond the limits of the audiometer.

Operation was carried out on August 9th, 1961. Gross otosclerotic disease of the oval window niche was found, heaped up about the stapes crura and obscuring all outlines. Very extensive drilling finally cleared about half the area of the oval window. A good vein graft and a specially cut long and slender strut was fitted. Convalescence was uneventful, and 8 weeks after the operation his audiogram showed air threshold at about 60 decibels. He claimed it was truly remarkable and that he could even hear a little without his aid. He is now maintaining this level and has purchased a small ear-level aid, and is looking forward to the day when I will operate on his other ear.

Case No. III. Mrs. M. McG., aged 36. History similar to the patients

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above. Pre-operative audiogram could record no air threshold, but the patient appreciated bone vibration at 50 decibels at 500 cycles. This was done before my ideas on testing were formulated and in retrospect

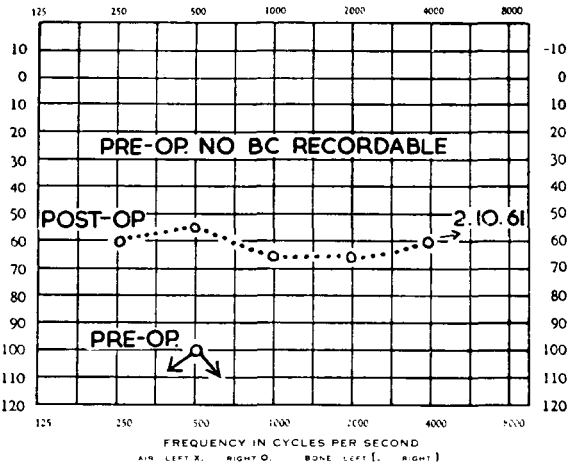


FIG. 5

Case No. II; Mr. C.L. showing pre- and post-operative hearing levels.

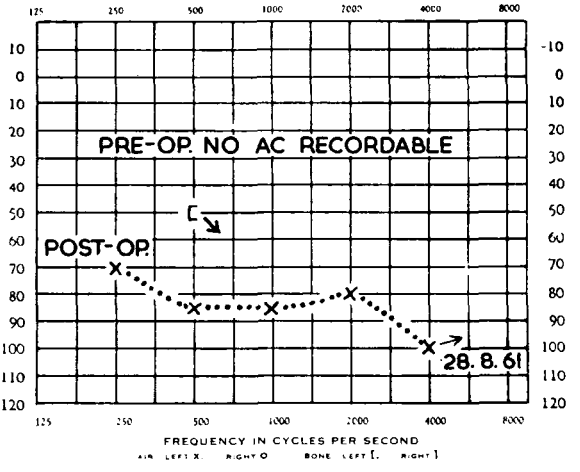


FIG. 6.

Case No. III; Mrs. McG. showing post-operative hearing level.

I am sure that this is not an auditory threshold, but rather a tactile one.

Operation was carried out on February 22nd, 1961. The incus was normal. There was gross disease on the promontory forcing the stapes

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crura up out of position. The bone lesion was cleared by drilling to reveal a fragile and almost membranous stapes footplate. This was removed without difficulty and the vein graft and strut were well placed.

One becomes rather used to patients saying how their personality alters after a successful Shea operation for otosclerosis, but this particular patient was really quite extraordinary in her descriptions. Audiogram taken 6 months after the operation shows an air threshold of about 85 decibels and this has been maintained to the time of writing some 20 months after the operation. She is using her aid well and is able to enjoy

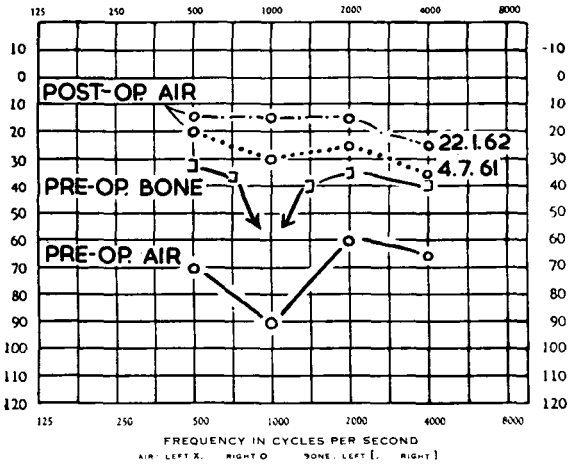


FIG. 7.

Mrs. J. W. Illustrating post-operative air conduction level well above pre-operative bone conduction level.

shopping, dancing, T.V. and picture theatres. As in most of these patients, the hearing aid is so much a part of them that they feel they have normal hearing despite the audiogram showing 85 decibel loss.

Before proceeding to sum up this subject, I would like to illustrate the way in which the post-operative air threshold can come up well above the pre-operative bone threshold. Fig. 7 shows such a case, Mrs. J. W., aged 41 years. Pre-operative bone tracing showed an average level of 40 decibels with a severe dip at 1,000 cycles which went down below the capacity of the audiometer. Right stapedectomy was carried out on May 26th, 1961, and minimal annulus disease was found.

Within 6 weeks of the operation her hearing was up above 30 decibels, and by 8 months after the operation it was at 15 decibels. This is the sort of case that would never have been tackled in the Fenestration era; nor would I have attempted Stapes Mobilization with such a low bone threshold. Now she has normal hearing.

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Summary

A series of cases is presented of extreme otosclerosis deafness to show that careful operation is justified.

McHugh (1961) discussing the brain-injured child, makes some very good comments upon the great disrupting effect of sensory isolation—"It has been demonstrated that the lack of sound and other sensory stimulation has a profound effect upon human behaviour, and the modern surgical techniques that permit the dramatic restoration of severe conductive deafness to normal hearing levels have also been found to produce far greater effects on overall brain activity than were ever previously suspected."

We regularly see this personality change in the normal successful stapedectomy patient, but we see it also in these very severely deaf subjects who have known the most depressing experience of their hearing steadily failing as they change from stronger to stronger hearing aids, until finally no electronic device can help them. Now we are able to bring back some sound, even if it is only the banging of a door.

Finally we must remember that the surgical restoration of a little bit of hearing is only the start of the patient's rehabilitation. Some of the hearing levels obtained are very low indeed, and although all of these patients are hearing aid users of long standing they should still be taught to use the new hearing correctly. This programme of rehabilitation extends over about 6 months and begins with the use of the hearing aid on the operated ear for only 20 to 30 minutes twice a day to begin with, and then only in controlled conditions. I ask them to tune in a radio to music, and then to adjust both the radio volume and their aid volume so that the sound is comfortable. This caution is dictated by the fact that we do not know just how these subnormal cochleae are going to tolerate loud sound. The training period is gradually extended, and sessions with recorded Phonetically Balanced (P.B.) word lists are included. It is extraordinary how their "score" mounts as the weeks go by. These patients are all wonderful lip-readers, and training with T.V. programmes allows both the aural and visual senses to work together.

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