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THE SURGERY OF OTOSCLEROSIS*

By TERENCE CAWTHORNE (London)

THE restoration, by surgical means, of the sense of hearing impaired by otosclerosis is the greatest otological event of our times. Naturally it has attracted much attention, and much is being written about it. Most of the contributions deal with the technique of the fenestration operation and with personal experiences and results. Some of these articles will be referred to in the present review, which is intended as a sequel to one which I gave some time ago (Cawthorne, 1947).

Principles

Many thousands of years ago, in the late Stone Age or Neolithic Age, our ancestors in these islands, and in certain parts of Continental Europe, were in the habit of making holes in living skulls. The reasons for this practice are a matter for conjecture, but Dr. T. Wilson Parry (1927), who has made an extensive study of prehistoric trephining, is convinced that besides being a ritual operation it was also undertaken for the relief of certain symptoms, among which he mentions giddiness and noises in the ears. If, then, trephining, which it would seem was fairly widely practised in prehistoric times, was done in the hope of letting unwanted noises escape from out of the head, it is not unreasonable to suppose that the same procedure may also have been employed in cases of deafness in the hope of letting very much wanted sounds into the head.

Although there are a few differences in matters of detail, such as technical improvements and in the size and situation of the hole, the underlying principle remains the same to-day: namely, that by making a hole in the skull, sound is enabled to reach the organ of hearing. There are, in addition, two further principles that must be constantly borne in mind in connection with this form of surgery. There must be sufficient

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function in the imprisoned organ of hearing to appreciate the sounds that fall upon it through the agency of the newly-made window; and it follows that the organ of hearing must be strong enough to stand up to the altered physical circumstances brought about by the opening of the bony labyrinth. A further consideration is that the opening made must be so maintained that the transmission of airborne sound waves to the labyrinthine fluids continues unimpaired.

With these principles clearly in mind, namely the functional competence of the organ of hearing and the making and maintaining of the window in the bony labyrinthine capsule, let us examine the present position as regards otosclerosis and its surgical treatment.

Pathology

Nager (1928) and the Shambaughs *et al.* (1942) found that otosclerosis was the commonest cause of deafness in early adult life, and now that there is a cure for this form of deafness more and more otologists are coming to the same conclusion. From the observations of Guild (1944) on the incidence of foci of otosclerosis in human temporal bones, it would seem that the incidence of deafness due to otosclerosis is not less than 0·75 per cent. of the general population. This makes it comparable in frequency with another well-known constitutional disease, namely diabetes. Thus, whilst it is not in any way dangerous to life as are diabetes and many other less commonly encountered constitutional diseases about which we learn so much as students, it is certainly a serious social handicap that deserves the attention of every student of health and disease, particularly now that so many who suffer from it can be restored to a normal working and social life. Otosclerosis is neither a rare, nor is it any longer an obscure, disorder; and though its cause is not yet fully known, its effects are well understood, and they can at last be overcome in many cases.

In otosclerosis the organ of hearing and its surrounding fluids are to a varying extent immobilized because of the tendency for otosclerotic bone to seal off the natural openings through which sound waves are transmitted to the interior of the bony labyrinth from the outside world.

The site of election for the appearance and development of otosclerotic bone is an area just in front of the oval window at the junction of the cochlea and vestibule that has been termed by Bast (1942) " the fissula ante fenestram ". It is nearly always from this site that the footplate of the stapes is slowly fixed in the oval window by spreading otosclerotic bone. Nylén (1949) found this area to be implicated in 90 per cent. of the temporal bones in Nager's collection that he examined, though in more than half other sites were also affected.

In early and limited cases only the oval window may be implicated, and I believe that it is in just such limited cases that the most favourable results are likely to follow surgical treatment.

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The spongy otosclerotic bone may spread, however, down to the round window as well, or inwards to the basal turn of the cochlea and even to the vestibular labyrinth. Nylén (1949) has described its extension over the surface of the bony labyrinth in 10 per cent. of the cases he examined. It is, I believe, in these extensive cases that surgery is often helpless ; and one important aspect of this work is to try and assess from the clinical examination the extent to which the otosclerotic process has invested or invaded the bony labyrinth.

Macroscopic evidence of the disease at operation is limited, and, in my experience, usually takes the form of small, sharply defined bony protuberances, pale yellow or reddish in colour and about the size of a split lentil. These have been noted on the promontory, sometimes spreading on to the adjacent facial nerve and sometimes on to the lateral canal. Only very rarely have I seen a diffuse growth of abnormal bone over the presenting surface of the labyrinth. In by far the majority of cases submitted to operation the appearance has been normal, confirming Politzer's (1892) observation, and I should like to quote his exact words : " that in the majority of specimens, the mucous membrane of the tympanic cavity did not present the least change from normal ".

Development of Surgery

The idea of breaking into the bony box of the labyrinth was initiated by Kessel (1876) who removed the stapes. This and other attacks on the anterior bony labyrinth were abandoned before the end of the nineteenth century, because of the frequency with which infection supervened with disastrous results to function and sometimes even to life. Bárány (1924) suggested the posterior approach through the vestibular labyrinth. It was, however, G. J. Jenkins (1913) who led the way into the bony lateral semicircular canal. In this connection it is interesting to note that he had already used this approach for the relief of vertigo, though he contented himself by merely opening the bony canal, and it was for this operation that the term " fenestration " was first used. The approach was post-auricular and the bony external canal was opened and the posterior wound closed.

My friend and colleague, Mr. W. Wilson, who was closely associated with Jenkins when he was continuing these operations after the first World War, says that the improvement in hearing that resulted from these operations lasted only a few days.

I have also tried this operation and have even preserved the mucosa from the external canal in order to roll it back over the newly-made fenestra or, alternatively, used amniotic membrane as a covering for the fenestra ; but with all these " closed operations " I never got an improvement in hearing that lasted longer than the patient's stay in hospital.

Mr. Wilson also says that Jenkins had the idea of approaching the

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superior canal in the same way that Holmgren did in 1917. In the face of repeated disappointments Jenkins did not persevere with his surgical attempts to cure deafness; and certainly when I first started to work with him in 1925 he was no longer doing these operations. I only wish that he could still be here with us to know that the trail that he blazed has now become the main road to hearing. Further developments of the Jenkins external canal approach were due to the energy, ingenuity and perseverance of Holmgren. He improved the technique, he employed magnifying glasses and the microscope; and he was rewarded in some cases by success (Holmgren, 1938).

Inspired by Holmgren's work, Sourdille (1932) developed his tympanopexy operation. This was the first open operation in which airborne sound waves fell directly upon the skin-covered window in the lateral semicircular canal. The operation was difficult, and it had to be done in two or even more stages, each separated by some months.

From this, Lempert (1938) forged ahead with his one-stage endaural operation that forms the basis of the technique generally employed throughout the world to-day.

This gradual building up of the surgical attack upon deafness in which one worker after another carries the flag farther on may be likened to a form of surgical relay race. Using this analogy, Bárány and Jenkins each ran a dashing 100 yards; Holmgren took over the flag for a gruelling 880; Sourdille spurted ahead in a brilliant 440, and Lempert flashed past the winning-post in a breath-taking 220 to win the race.

But they haven't stopped running yet! Like all good athletes they are not satisfied, and those of them who are still with us continue to strive for better results and to encourage and train others to help in the race.

I cannot leave this aspect of the subject without referring to the debt of gratitude the whole world owes to Lempert, not only for his personal achievement in establishing the fenestration operation, but also for the service he has done otologists all over the world in training them in this work. One of his pupils, Shambaugh, has earned his place in the team and his contributions have played a significant part in improving the results (Shambaugh, 1942, 1946, 1949).

With this short review of the development of the fenestration operation to its present state, it will now be convenient to consider the operation itself as now practised, and what I have to say about this will be based on personal experience in nearly 500 operations for the relief of deafness due to otosclerosis.

Technique

The operation in general use and which I employ is based on Lempert's nov-ovalis approach, and consists in making a window in the anterior part of the bony external semicircular canal and adjacent ampulla. This

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opening is covered by a flap derived from the skin covering the anterior wall, roof and posterior wall of the bony external meatus, continuity with the intact tympanic membrane being preserved.

Its proper performance makes considerable demands on the surgeon and calls for practice, patience and self-control. The surgeon must ensure that he is provided with good illumination and adequate magnification, particularly for the most important part of the operation, namely the making of the fenestra. This must be made on top of a mound, so that when the tympano-meatal flap is placed over the fenestra it will invest it closely and seal it off like a rubber patch sealing off a puncture in a pneumatic tyre. Great care must be taken to avoid injury to the membranous labyrinth, or anything that may encourage the formation of excessive fibrous tissue or bone formation over or within the fenestra. Hæmostasis, a fenestra with clean margins, a perilymph space free of blood and of bone débris, an intact membranous canal and ampulla, and a thin intact tympano-meatal flap from which all flakes of bone have been removed, are essential for success. Lempert and his former pupil, Shambaugh, have furthered the progress of the operation by drawing attention to these and other factors. From my own experience I would like to emphasize the value of adequate magnification whilst actually making the fenestra. I have regularly used a microscope giving ten diameters of magnification for the finer parts of all temporal bone operations since 1938, and Shambaugh (1942) emphasized its value in the fenestration operation. The binocular dissecting microscope, which is freely manœuvrable and which has a brightly illuminated field is the surest way of achieving a clean fenestra, a clean flap, an undamaged membranous labyrinth and a perilymph space free from bone débris and blood.

Only when this stage of the operation is properly performed can a good and lasting improvement in hearing be expected. The following pictures of the fenestra magnified ten times are from a 16 mm. colour film taken by Mr. E. Gwynne-Evans and Mr. E. Mackie. The clean-cut fenestra and the membranous labyrinth are clearly seen.

Therefore, the closest attention and much practice have to be devoted to the making of the fenestra, and the wise surgeon will ensure that he is neither tired out by the preceding steps of the operation, cramped for space by inadequate exposure, handicapped by poor illumination nor, finally, embarrassed by lack of experience in working in a magnified field.

Other aspects of the operation have been fully dealt with elsewhere and, provided that the exposure is adequate, both at the time of operation and for the post-operative inspection and treatment of the cavity, and that important neighbouring structures are not imperilled, the details of the soft tissue and bony approach can be left to the individual surgeon.

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My own preference is for the endaural approach, though, as I have seen when visiting Mr. Simson Hall in Edinburgh, the post-aural approach gives an excellent view and excellent results (Simson Hall, 1948). I have still, however, to be convinced of the value of the antauricular approach advocated by Popper (1947).

The bony cortex of the mastoid can be reduced to bone dust by a dental drill or removed by sharp gouges. The drill has the advantage of generally causing less bleeding than the gouge and, in practised hands, can be very accurate and safe. On the other hand, the reduction of the mastoid cortex to fine bone dust by a drill is a messy, laborious and time-consuming procedure. Having tried both, I prefer a little blood at this stage to a lot of bone dust; though, of course, both can be, and are, satisfactorily removed by flushing the cavity with warm saline. In this connection I should like to emphasize the value of irrigation of the operative field with saline at 100° F. In the earlier stages intermittent irrigation is sufficient, but for making the fenestra continuous irrigation as suggested by Shambaugh is most helpful. It is, however, of some importance to ensure that the irrigating fluid is constantly at 100° F., for the exposure of the unprotected vestibular labyrinth to gross thermal changes is obviously undesirable, and it may augment the post-operative vestibular disturbance.

Sufficient bone must be removed to allow full freedom of movement under magnification in the region of the fenestra. Aubry (1946), Hicguet and Van Eyck (1947), Popper (1947) and, lately, Shambaugh (1949) advocate a very limited removal of bone and they try to avoid opening the mastoid or even the antrum. I have not as yet any experience of this limited approach, though if it will hasten post-operative healing without increasing the risk of undesirable complication from infection it will be an advantage. The actual making of the fenestra is, of course, the most important part of the operation. All experienced workers emphasize the value of a fenestra with clean edges, and a perilymph space free from bone débris. Lempert (1948) in his bone-dust-free technique, thins down the bone over the external canal and ampulla until the endosteal layer of bone is reached. Then he outlines the fenestra with a fine knife, so that the " cupola " comes away intact like the top of an egg. Another way of uncapping the fenestra that I have found to be very effective, as it leaves a beautifully clean edge to the fenestra, is to remove the thinned-down " cupola " bone with a 2 mm. gouge. Care must, however, be taken to thin down the bone equally over the area where the margins of the fenestra will be.

Skin grafting the raw areas of the bony cavity at the time of operation will help to shorten the healing time, but unless the graft is very thin indeed it often does not take, particularly if the cavity becomes infected. I think that it is nearly always well worth while covering every bit of raw

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surface with skin graft in the hope of getting primary healing. This can be achieved in about a quarter of the cases.

For the first few post-operative days, the tympano-meatal flap and cavity need to be subjected to continuous, gentle and non-adherent pressure. This can be achieved by lining the cavity with strips of oiled silk—Lister's Green Protective—and then packing it with small pieces of marine sponge. Shambaugh, from whom I got the idea of using sponge, employs two relatively large pieces soaked in sulphadiazine cream, but I believe that smaller pieces give a more evenly distributed pressure, and I am not as yet convinced that soaking the sponges in any local antiseptic is of value, except as a deodorant.

Post-operative Course

With regard to the post-operative period, some vestibular disturbance is the rule, due not so much to infection as has been suggested by some observers, but to the altered physical conditions within the labyrinth. This can best be overcome by the head and balancing exercises developed for the management of vestibular injuries (Cawthorne and Cooksey, 1946). Campbell (1949) has recently reported on the value of Dramamine in combating the post-operative vestibular disturbance. Most patients are out of bed on the fourth post-operative day, the first dressing being done on the eighth day. Very few need to stay in hospital more than twelve days after operation.

Some degree of post-operative infection of the cavity is common, and though continued discharge may be a nuisance it rarely affects the hearing result, and seems to be due to infection of soft tissue rather than bone.

The results are largely governed by the care with which cases are selected and the skill with which the operation is carried out.

Before, however, considering the selection of cases, I should like to deal with operative results.

Results

It is mainly by watching his results that the surgeon can measure his progress and be stimulated to seek for further improvement. Furthermore, by reference to his own results he is able to make the fairest estimate of the amount of improvement in hearing that fenestration at his hands will offer.

Many standards have been suggested for reporting results and the American Otological Society has a sub-committee to enquire into standards for reporting fenestration results, and they have made provisional suggestions.

With regard to my own results, I would prefer to give the figures in terms of decibel improvement or loss for pure tones within the critical

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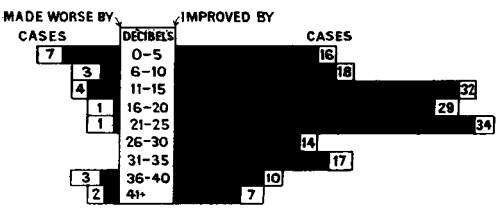
range of speech frequencies (512, 1,024 and 2,048). I have grouped the cases in 5 decibel steps and have included only those patients who have been followed up for eighteen months or more after operation, and I have chosen this time because so far I have not had any patient whose hearing deteriorated after eighteen months. I think that the ideal way of testing the hearing capacity after operation is by means of speech, and Walsh and Silverman (1948) and their co-workers have already published some interesting observations along these lines.

The results now to be considered are those from cases on whom I operated by the technique described, between July, 1946, and April, 1949. Only those of whom I have no record for at least eighteen months after the operation have been omitted.

The following table shows the results in 251 cases in terms of the standards already described :

TABLE I.

HEARING FOR PURE TONES WITHIN THE CRITICAL RANGE OF SPEECH FREQUENCIES (512, 1,024 AND 2,048 C.P.S.) EIGHTEEN MONTHS OR MORE AFTER FENESTRATION IN 251 CASES OF OTOSCLEROSIS



The selection of suitable candidates for surgery from among those deafened patients who hopefully present themselves is no easy task, nor is it a matter about which hard-and-fast rules can as yet be laid down. Some appear to be more suitable than others, whilst some are obviously quite unsuitable.

The series under consideration does not include any patient who :

1. Was not thought to be suffering from otosclerosis.
2. Did not have an intact and healthy tympanic membrane.
3. Could not tolerate and understand speech with the help of a high quality amplifier.

Each patient selected for operation has, at the time of the first interview, been put into one of three groups : Suitable, Borderline, and Unsuitable, and the distribution in this series has been as follows :

TABLE II.

Suitable	131
Borderline	105
Unsuitable	15

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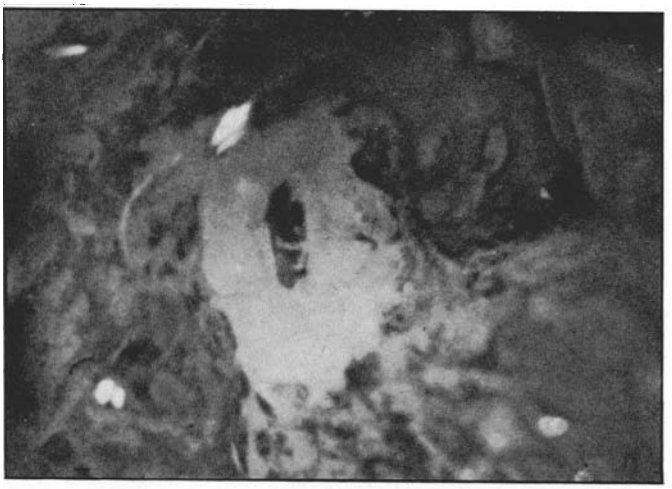


FIG. 1.

Magnified view ($\times 6$) of a newly created fenestra. The top of the fenestra has been cut off and turned over, where it can be seen on the left of the fenestra. The membranous canal with some shreds of endosteal débris and bone will have to be cleared away.

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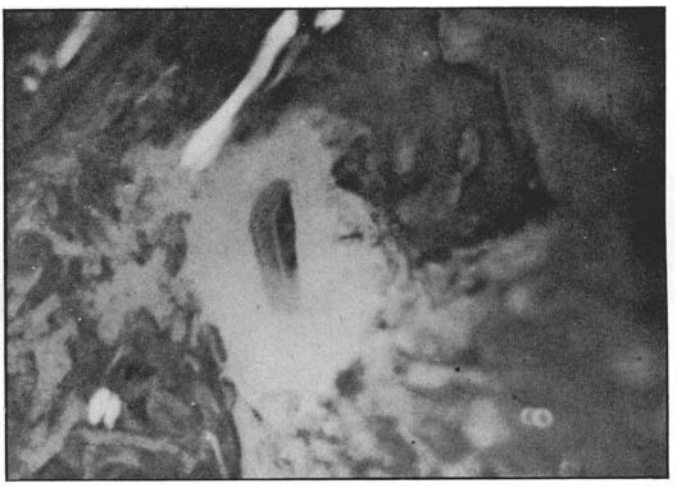


FIG. 2.

The same fenestra as in Fig. 1, after all the shreds of bone have been cleared away.

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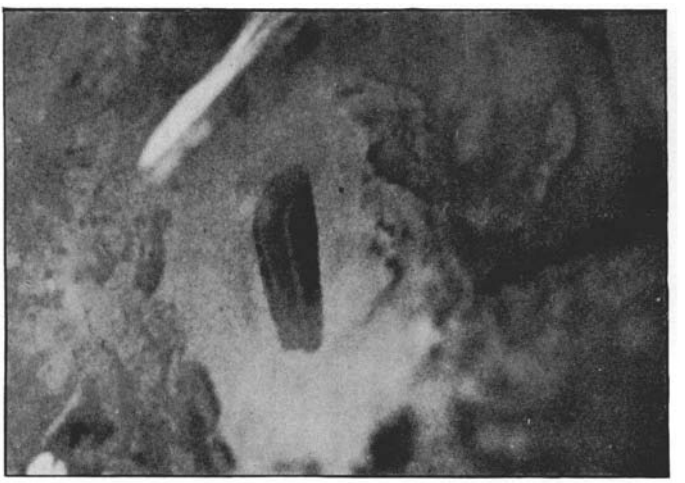


FIG. 3.

The completed fenestra. The membranous labyrinth can be clearly seen and in the upper part of the fenestra the membranous canal can be seen bending into the ampulla (×10).

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I started to do this at the beginning of this series, and recently Shambaugh (1949) has described a similar scheme.

I do not think that I can possibly attempt to lay down any definite rules about classifying cases except to say that there is no one definite feature to go by. The most intriguing cases are the failures in the suitable group. Some may be due to an unsuspected operative or post-operative hazard, particularly to bleeding; but I think that others are due to an inability of the organ of hearing to stand up to the strain: an inability which often does not, in the present state of our knowledge, give any pre-operative clue. One has to assess the patient as a whole, taking into consideration the history, the appearance of the tympanic membranes, tuning fork tests, pure tone audiogram, the response to amplification and the tone of the voice.

The following case sheets have been selected as representative of an average case in each group:

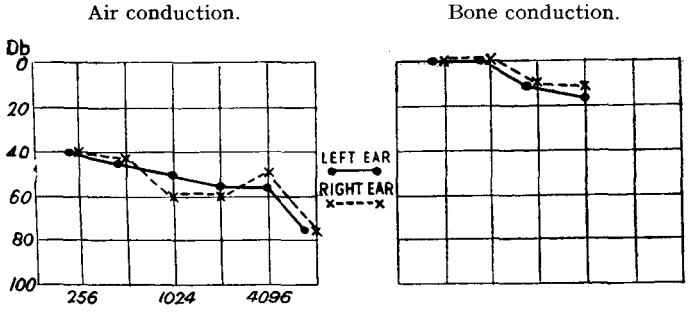
TABLE III.

SUITABLE

Mrs. O. D., age 48. 23.7.48.

HISTORY.—*Deafness.*—Onset, 10 years. Course, Progressive. Affected by, Not by only child. Present Hearing, Raised voice. *Paracusis.*—Noted. *Family History.*—None. *Tinnitus.*—None. *Vertigo.*—None. *Discharge.*—None. *Trauma.*—None. *Drugs.*—Quinine. *Other Illness.*—Malaria. *Previous Investigation.*—Inflation.

EXAMINATION.—*Complexion.*—Dark. *Temperament.*—Equable. *Amplifier.*—Well tolerated. *Voice Tone.*—Soft, normal timbre. *Upper Respiratory Tract*—Clear. *Ears and Hearing* (Left and Right).—External Meatus, Traces of cerumen. Tympanic Membrane, Normal. Conversational Voice, 3 inches. Eustachian Tube, Patent. *Tuning Fork* (Left and Right Rinne).—Absolute Bone Conduction: 512, Normal. Negative. 1024, Normal. Negative. 2048, Slight loss. Negative. *Audiometer.*—Maico D.9.



CONCLUSIONS.—Otosclerosis.

RECOMMENDATIONS.—Suitable. 50 per cent. chance of gaining and maintaining improvement in right ear sufficient to hear speech at conversational level within radius of 8 feet.

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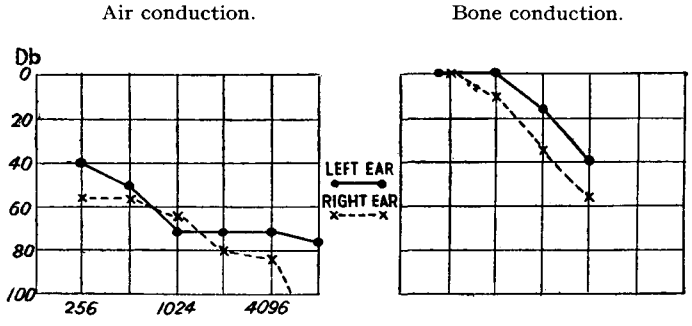
TABLE IV.

BORDERLINE

Mrs. E. D., age 45. 23.7.48.

HISTORY.—*Deafness.*—Onset, 31 years. Course, Slowly progressive. Affected by, Not by only child. Present Hearing, Raised voice. *Paracusis.*—Noted. *Family History.*—Sister, niece. *Tinnitus.*—Moderate, not troublesome. *Vertigo.*—Occasional, momentary. *Discharge.*—None. *Trauma.*—None. *Drugs.*—Alcohol. *Other Illness.*—None. *Previous Investigation.*—Inflation.

EXAMINATION.—*Complexion.*—Dark. *Temperament.*—Equable. *Amplifier.*—Well tolerated (air and bone). *Voice Tone.*—Soft, normal timbre. *Upper Respiratory Tract.*—Clear. *Ears and Hearing* (Left and Right).—External Meatus, Moderate cerumen. Tympanic Membrane, Normal. Conversational Voice, Raised voice. Eustachian Tube, Patent. *Tuning Fork.*—Absolute Bone Conduction (Left Rinne): 512, Normal. Negative. 1024, Slight loss. Negative. 2048, Nil. Absolute Bone Conduction (Right Rinne): 512, Normal. Negative. 1,024, Slight loss. Negative. 2048, Moderate loss. Bone only. *Audiometer.*—Maico D.9.



CONCLUSIONS.—Otosclerosis.

RECOMMENDATIONS.—Borderline. 30 per cent. chance of gaining and maintaining improvement in right ear sufficient to hear speech at conversational level within radius of 3 feet.

TABLE V.

UNSUITABLE

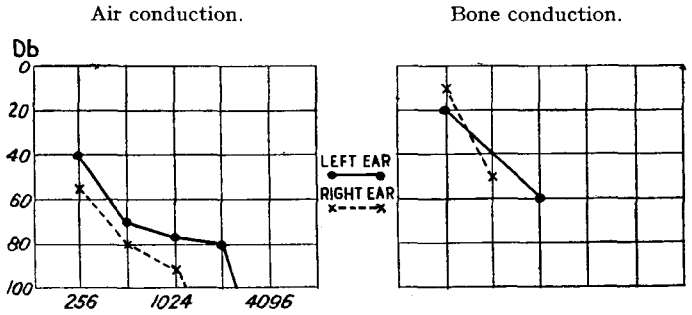
Mr. F. C. H., age 43. 15.9.48.

HISTORY.—*Deafness.*—Onset, 16 years. Course, Slowly progressive. Affected by, Head colds. Present hearing, Raised voice. *Paracusis.*—Noted. *Family History.*—Brother slightly deaf. *Tinnitus.*—None. *Vertigo.*—None. *Discharge.*—None. *Trauma.*—None. *Drugs.*—None. *Other Illness.*—None. *Previous Investigation.*—Septum operation.

EXAMINATION.—*Complexion.*—Dark. *Temperament.* Equable. *Amplifier.*—Well tolerated. *Voice Tone.*—Early voice changes. *Upper Respiratory Tract.*—Evidence of former nasal operation. *Ears and Hearing* (Left and Right).—External Meatus, Traces of cerumen. Tympanic Membrane, Normal.

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Conversational Voice, Shout. Eustachian Tube, Patent. *Tuning Fork.*—Absolute Bone Conduction (Left Rinne): 512, Moderate loss. Air equals Bone. 1024, Moderate loss. Air equals bone. 2048, Moderate loss. Positive; Absolute Bone Conduction (Right Rinne): 512, Moderate loss. Negative. 1024, Moderate loss. Negative. 2048, Nil.



CONCLUSIONS.—Otosclerosis.

RECOMMENDATIONS.—Unsuitable. 20 per cent. chance of gaining and maintaining improvement in right ear sufficient to hear speech at conversational level within radius of 1 foot.

The following tables show the results in each of the three groups: (?)

TABLE VI.

HEARING FOR PURE TONES WITHIN THE CRITICAL RANGE OF SPEECH FREQUENCIES (512, 1024 AND 2048 C.P.S.) EIGHTEEN MONTHS OR MORE AFTER FENESTRATION FOR OTOSCLEROSIS IN 131 CASES CLASSIFIED AS SUITABLE

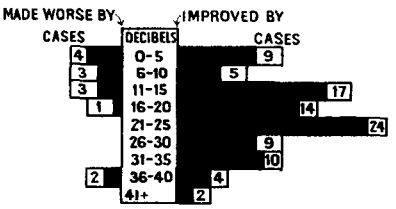
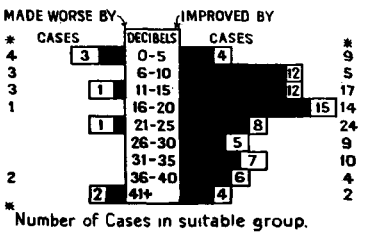


TABLE VII.

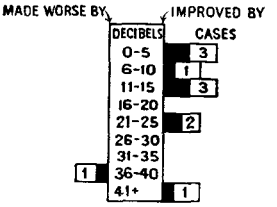
HEARING FOR PURE TONES WITHIN THE CRITICAL RANGE OF SPEECH FREQUENCIES (512, 1024 AND 2048 C.P.S.) EIGHTEEN MONTHS OR MORE AFTER FENESTRATION FOR OTOSCLEROSIS IN 105 CASES CLASSIFIED AS BORDERLINE



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HEARING FOR PURE TONES WITHIN THE CRITICAL RANGE OF SPEECH FREQUENCIES (512, 1024 AND 2048 C.P.S.) EIGHTEEN MONTHS OR MORE AFTER FENESTRATION FOR OTOSCLEROSIS IN 15 CASES CLASSIFIED AS UNSUITABLE

TABLE VIII.



This method of grouping cases does not pretend to be exact, but it does guide the otologist in advising his patient as to the chance of being helped by an operation. Provided the patient is suffering from otosclerosis and is able to hear with an aid, I think that it is unfair to deny him or her the chance of hearing again, though, of course, the chance of success should always be frankly estimated. I always try to discourage cases in the unsuitable group but, as the results show, nearly 25 per cent. of these can have their hearing restored to the practical level.

It will also be interesting to group the results according to the provisional suggestions of the American Otological Society, and these are approximately as follows:

GROUP 1. PRACTICAL HEARING RESTORED

Improved up to or above the 30 decibel level for the three critical speech frequencies (512, 1024 and 2048 d.v.).

GROUP 2. IMPROVED

Improved by at least 15 decibels for the three critical speech frequencies, but not up to the 30 decibel level.

GROUP 3. UNCHANGED

Improved by less than 15 decibels or not made worse by more than 10 decibels for the critical speech frequencies.

GROUP 4. MADE WORSE

Made worse by more than 10 decibels for the critical speech frequencies.

The figure in each case is arrived at by adding together the readings for each of the three frequencies and dividing by three.

TABLE IX.

EFFECT ON HEARING OF FENESTRATION FOR OTOSCLEROSIS EIGHTEEN MONTHS OR MORE AFTER OPERATION IN 251 CASES

		Suitable	*Borderline*	*Unsuitable*
GROUP 1				
(Practical hearing restored)	132	88	41	3
GROUP 2				
(Hearing improved)	51	18	29	4
	64			

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		Suitable	*Borderline*	*Unsuitable*
GROUP 3				
(Hearing unchanged) ..	56	21	30	5
Group 4				
(Hearing made worse) ..	13	5	5	3

Indications for Operation

Now I believe that in its present form the fenestration operation is most likely to be successful when the otosclerotic bone is limited to the region of the oval window and, *per contra*, that extensive spread of otosclerotic bone, either over the surface or into the cavities of the bony labyrinth, renders a case unsuitable. Therefore any clinical indication of such extensive growth should be carefully sought for and, if present, taken into consideration in assessing suitability for operation.

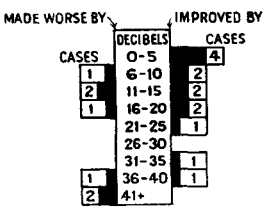
In a previous review of the surgery of otosclerosis (Cawthorne, 1946), I mentioned four signs of poor prognostic significance, and I should now like to add a fifth :

1. Alteration in the TIMBRE of the voice.
2. Intolerance of AMPLIFICATION.
3. Disappearance of PARACUSIS.
4. Reduction of BONE CONDUCTION.
5. Pink-tinged TYMPANIC MEMBRANE.

I would suggest that in the first three groups the organ of hearing is definitely at fault. In Group 4, the organ of hearing may be responsible, but I believe that loss of bone conduction can also be caused by widespread otosclerotic bone, particularly if it seals up both labyrinthine windows. In the last group, the tympanic membrane is seen to have a pink tinge due, it is believed, to transmitted colour from the mucosa covering the promontory—the so-called Schwartze sign. This is often associated with other unfavourable signs, particularly loss of bone conduction, and is generally regarded as an unfavourable sign. It was not until I was preparing this paper, however, that it was borne upon me how unfavourable a sign this really is. (vp. 116)×

TABLE X

HEARING FOR PURE TONES WITHIN THE CRITICAL RANGE OF SPEECH FREQUENCIES (512, 1024 AND 2048 C.P.S.) EIGHTEEN MONTHS OR MORE AFTER FENESTRATION FOR OTOSCLEROSIS IN 25 CASES WITH PINK-TINGED TYMPANIC MEMBRANES



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This group can be analysed further as follows, and it will be seen that those with pink-tinged tympanic membranes account for many of the failures, and in the suitable group for all the failures :

TABLE XI

		Suitable	*Borderline*	*Unsuitable*
GROUP 1				
(Hearing restored to practical level)	8	6	2	
GROUP 2				
(Hearing improved)	2		2	
GROUP 3				
(Hearing unchanged) ..	7	1	5	1
GROUP 4				
(Hearing made worse) ..	8	5	2	1

In the present series, 25 cases were noted as having pink-tinged tympanic membranes, and only 10 of these sustained an improvement of more than 10 decibels for the critical speech frequencies, despite the fact that 16 had been assessed before operation as suitable cases. On the other hand 9 were made worse. I have found that such cases have been the cause of much disappointment, and in future I shall put all such cases in the borderline or unsuitable categories ; though by doing so I do want to emphasize that they should not be denied operation provided that they appreciate that the chance of success is slender.

The successful cases are both gratifying and satisfying, but to dwell on them too much is enervating. The failures are disappointing and sometimes distressing, but they can and should be stimulating. The cause of failure in some instances is only too obviously—and I refer to my own cases—faulty technique, in others faulty selection ; but there are some in which the cause of failure is not as yet obvious. But it is only by doing a certain proportion of apparently unsuitable cases that we can advance this work, and I would draw attention to the fact that in 6 out of the 15 apparently unsuitable cases that I operated upon there was an improvement in hearing of more than 20 decibels for the critical speech frequencies. Therefore, although the prospect of improvement is not necessarily bright in cases classified as unsuitable, I do not think that they should be absolutely denied operation if there is evidence of effective cochlear function.

Removal of Stapes

Before concluding, I should like to offer a few observations on two other forms of surgical treatment that I have tried.

I have removed the stapes in 23 cases of otosclerosis, all but four of which would be classified as UNSUITABLE. Cornelli (1949) has also

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recently described his experiences with this operation. One of the SUITABLE cases has been demonstrated, and he has retained his improvement for nearly four years. He has also had a successful fenestration on the other ear. If we can solve the problem of sealing off the oval window and of retaining the tympanic membrane intact and of normal tension, I believe that this might prove to be a most effective and lasting form of treatment; because, after all, it is Nature's window and is unlikely to become sealed over by bone again, as anyone will realize who has seen the oval window under the binocular microscope.

In another small group, all considered most unsuitable by the standards that I have mentioned, I have performed a fenestration operation and in addition removed the stapes. In all the four cases that were so treated, I believed that the round as well as the oval window was impeded by otosclerotic bone. In none, however, was there any improvement in hearing, but I still feel that in such cases, provided that there is sufficient cochlear function to appreciate and tolerate amplification, some form of operative treatment may be found to be of help, and I intend to continue to search for it.

There are two final aspects of this problem that are of considerable physiological interest and which may help us to appreciate further the mechanics of hearing.

Tullio Phenomenon and Fenestration

I have not as yet seen a case of otosclerosis submitted to the fenestration operation in which exposure to loud sounds resulted in a vestibular stimulus. In the Tullio phenomenon, a window made in the lateral canal renders the animal—pigeons, in the cases Tullio described—liable to a vestibular stimulus on exposure to a loud sound. I had a patient suffering from Ménière's disease on whom, in 1938, I made an opening into the bony lateral semicircular canal, without disturbing the membranous labyrinth. This patient, after leaving hospital, complained that exposure to any loud sound caused giddiness, and investigation revealed that exposure to a pure tone of 80 db. within the range 250-4,000 c.p.s. caused a movement of the eyes and deviation of the head and body. This was relieved by a second operation, in which the membranous external semicircular canal was removed, and as a result cochlear function destroyed.

Now why is it that Tullio's phenomenon is not seen after the fenestration operation for the relief of deafness due to otosclerosis? For I have not as yet seen it, nor have I knowledge of any record of it. I have, however, seen it in two patients who had been submitted to a fenestration operation; but in each case there was some doubt as to whether the deafness was due to otosclerosis, and I suspect that in these two cases the

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footplate of the stapes was not impeded. I believe that the Tullio phenomenon is dependent upon unimpeded round and oval windows. For this reason I would hesitate to advise the fenestration operation where there was any reason to suppose that the stapes was mobile and the round window unimpeded.

Effect on Hearing of Damage to Membranous Labyrinth

The other feature that is of particular interest to me is that limited damage to the membranous canal does not, in cases of otosclerosis, necessarily result in a complete loss of hearing. In every case of Ménière's disease in which I have removed all or even part of the membranous external semicircular canal, total and irreversible deafness has resulted. I have, however, accidentally torn the membranous external semicircular canal across in four of the cases in the present series. In none was the hearing improved, whilst in three it was not improved or made worse than by 10 db. either way. In the fourth, it was made much worse, but was not abolished. In each case the membranous canal was torn across at the posterior end, well away from the ampulla. I hope in due course to report the result of deliberately carrying out a similar procedure in cases of Ménière's disease.

Other Complications

There are some whose ear continues to discharge, though this does not necessarily modify a good result ; whilst others have a varying degree of narrowing of the meatus.

As yet, I have not found it necessary to re-operate on a patient for either of these complications.

In two cases there was a facial paresis, one due to pressing in of the thin bony fallopian canal in trying to stop a bleeding point, and one which came on three days after operation for no obvious reason. In both cases recovery was complete within two weeks.

In 37 cases the initial improvement was not maintained, due, I believe, in most instances to a partial or complete closure of the fenestra. On the other hand, 23 cases continued to improve three or more months after the operation, and one only started to improve seventeen months after operation. Finally, in one case the hearing in the operated ear went altogether. Because of the possibility of making the hearing in the operated ear worse, I make it a rule always to operate on the worse hearing ear. If this is successful the other ear can if desired be operated on at a later date—not less than one year after.

Revision

With regard to revision operations, I must confess to being disappointed in the results. In the seven that I have done in the series under

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review, I found bone extending into and as a rule surrounding the membranous canal. In three of these, the membranous canal was torn in endeavouring to free it from the surrounding bone. In none was there any permanent improvement in hearing as a result of the revision operation. I think that the next time I undertake a revision I shall be prepared, if I find the membranous labyrinth entangled in new bone, to make a fresh opening in the bony labyrinth, possibly in the posterior canal.

Conclusion

The surgical treatment of deafness due to otosclerosis is now firmly established. In skilled hands—and only skilled hands should attempt this delicate surgery—more than two-thirds of the patients who submit themselves to operation can expect a worth-while and lasting improvement in hearing. There is still much more work to be done and many problems to be solved. For those who are unsuitable for or unwilling to face surgery, a good wearable valve-amplified hearing aid rarely fails to improve the hearing. We must not lose sight of the fact that surgery is only part of the management of the patient suffering from this form of deafness.

REFERENCES

AUBRY, M., 1946, *Ann. d'Otol.*, Jan.-Feb.

BÁRÁNY, R., 1924, *Acta oto-laryng., Stockh.*, vi, 260.

BAST, —., 1942, *Arch. Otolaryng., Chicago*, xxxvi, 816-26.

CAMPBELL, E. H., and MACFARLANE, D., 1948, *Arch. Otolaryng., Chicago*, xlvii, 590.

CAWTHORNE, T. E., 1947, *Proc. roy. Soc. Med.*, ix, No. 6, 320-6.

———, and COOKSEY, F. S., 1946, *Proc. roy. Soc. Med.*, xxxix, 270.

CORNELLI, G., 1949, *L'Ospedale Maggiore*, a. xxxvii, n. 2.

GUILD, S. R., 1944, *Ann. Otol. Rhin. Laryng.*, liii, 246.

HICGUET, G., and VAN EYCK, M., 1947, *Ann. d'Otol.*, lxiv, 634.

HOLMGREN, G., 1938, *Surgical Therapy for Otosclerosis*, Nelson's Loose Leaf Surgery of the Ear, New York.

JENKINS, G. J., 1913, *Int. Congr. Med.*, xvii, Sect. 16 (Otol.), Pt. 2, 609.

KESSEL, J., 1876, *Arch. Ohrenheilk.*, xi, 199.

LEMPERT, J., 1938, *Arch. Otolaryng., Chicago*, xxviii, 42.

———, 1948, *Arch. Otolaryng., Chicago*, xlvii, 551.

NAGER, F. R., 1928, *J. Laryng. and Otol.*, xliii, 15.

NYLÉN, B., 1949, *Histopathological Investigations on the Localization, Number, Activity and Extent of Otosclerotic Foci*, Upsala.

PARRY, T. WILSON, 1927, *Postgrad. med. J.*, May.

POLITZER, A., 1892, *Anatomical and Histological Dissection of the Human Ear in Normal and Diseased Conditions*, Baillière, Tindall & Cox, p. 287.

POPPER, O., 1947, *J. Laryng. and Otol.*, lxi, 24.

SHAMBAUGH, G. E., 1942, *J. Amer. med. Ass.*, cxix, 243.

———, 1949, *Acta oto-laryng.*, Supplementum lxxix.

———, and JUERS, A. L., 1946, *Arch. Otol., N.Y.*, xliii, 549 (June).

SIMSON HALL, I., and YOUNG, A., 1948, *J. Laryng. and Otol.*, lxii.

SOURDILLE, M., 1932, *Rev. Laryng., Paris*, liii, Suppl.

WALSH, T., and SILVERMAN, R., 1946, *Laryngoscope, St. Louis*, lvi, 536.

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