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"Nerve" Deafness: Its Clinical Criteria, Old and New

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ACCORDING to the classical precepts which have long governed the functional tests of hearing, the tuning fork tests of Schwabach, Rinne and Weber make it possible to effect a reasonably clear-cut distinction between nerve or perceptive deafness, as it is often termed, and deafness due to disease of the sound-conducting mechanism of the middle ear.

All otologists are trained in the use of these tests for this differential diagnosis, and few would question their value, but to this must be added that one limitation in particular has long been apparent, namely, that while the tests, which may be called the old criteria of nerve deafness, make it possible to diagnose its existence they do not make it possible to go further and say what, and in what way, particular components of the perceptive mechanism have been affected.

During the last ten years, this situation has shown some change for the better. In the first place more information is available concerning the pathological anatomy of the inner ear and VIIIth nerve. In addition, new knowledge of cochlear function has led to the development of new test procedures. These may be called the *new* criteria of nerve deafness and, as will be seen, they give good promise of enabling us to achieve what the old criteria do not, namely, to add to the diagnosis of nerve deafness some insight into the nature and localization within the perceptive mechanism of the pathological process concerned.

The new criteria which form the subject of this communication are, firstly, the Loudness Recruitment Test, and, secondly, the intelligibility test for amplified speech. These tests have been systematically applied in two groups of subjects: firstly Ménière's disease of the labyrinth, and secondly, VIIIth nerve neurofibroma; and the results obtained will be described and discussed. These two groups have been selected since they exhibit in their pathological anatomy that very difference to which we look to provide a test of our diagnostic criteria. In one group, Ménière's disease, there is good reason to believe that the disturbance of cochlear function is due to an affection of Corti's organ. In neurofibroma, on the other hand, it is known that there occurs a degeneration of the cochlear nerve fibres.

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The pathology of VIIIth nerve neurofibroma has been well described by Cushing (1917), Henschen (1915) and others, and particular mention must be made of the observations of Hardy and Crowe (1936) upon the early stages of its development. The tumours fill the internal auditory meatus, and bring about a gradual pressure destruction of the fibres of the VIIIth nerve with resulting deafness and loss of the vestibular responses. A point of particular interest is the degeneration of the spiral ganglion within the cochlea, although Corti's organ is as a rule well preserved, a point which has also been established with particular clarity in the case of experimental section of the VIIIth nerve in animals. These studies are of particular importance since the vastly superior methods of fixation which are possible in animals establish beyond any doubt the anatomical integrity of the hair cells of Corti's organ which follows a lesion of the VIIIth nerve.

It can therefore be said in summary that neurofibroma of the VIIIth nerve leads to diffuse degeneration of the fibres of the cochlear nerve with preservation of Corti's organ. This statement must be amplified in one important respect. If the neurofibroma or operation of VIIIth nerve section interferes with the blood supply of the cochlea, then Corti's organ *is* affected, and various degrees of necrosis are to be seen in it and in other cochlear structures.

In the case of Ménière's disease, as the term is now used, there is found a very different state of affairs. The primary change within the labyrinth is a distension of the endolymph system. This has been found in all of the five cases which we have had the opportunity of examining, and it has been found, too, in at least a dozen cases examined by other observers. Many of these have described Corti's organ as being normal, but in three of our own cases very striking changes have been present.

Figs. 1 and 2 show Corti's organ in two of these cases. In Fig. 1 the cells have undergone a striking compression and Corti's tunnel is occupied by a structureless coagulum. In Fig. 2 the same striking compression of the cell mass is to be seen. In addition the normal staining differentiation between cytoplasm and nuclei has been obliterated.

No doubt can be entertained concerning the significance of this change in Corti's organ in these two and in one other of our cases, and their importance would appear to be very great. For a full discussion of these changes and their relationship to the deafness of Ménière's disease, reference should be made to a previous communication (Dix *et al.*, 1948). ℓv

It is important to add that in none of our cases have we observed any changes in the peripheral cochlear nerve fibres, or in the cells of the spiral ganglion.

In summary it can be said that while in many cases of Ménière's disease the organ of Corti may appear normal, nevertheless, in such cases there are likely to be present certain submicroscopic residua of the gross changes revealed in three of our cases, and it is to these submicroscopic changes that must be attributed the deafness which in Ménière's disease persists between the attacks. In short, therefore, we are able to say that in VIIIth nerve neurofibroma the anatomical structures affected are the cochlear nerve fibres, the hair cells being preserved, while in Ménière's disease there occurs a primary affection of the hair cells with preservation of the cochlear nerve fibres.

The results of the classical tuning fork tests, the old criteria, reveal little to

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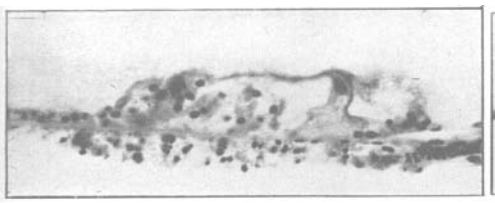


Fig. 1.

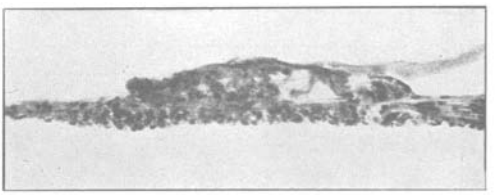


Fig 2.

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distinguish these two groups. In unilateral Ménière's disease Weber's test is typically referred to the normal hearing ear. Rinne's test may give a false negative result if carried out without masking of the normal ear. With masking, however, such a negative result is nearly always reversed. For the measurement of perception by bone conduction we find it best to use the so-called Absolute Bone Conduction Test, with occlusion of the meatus both of the subject and of the examiner. With this test reduced perception is the rule in Ménière's disease, although here again it is necessary to stress the importance of masking the normal ear.

In VIIIth nerve neurofibroma, these three tests give substantially the same results. With Weber's test, perception is referred to the normal ear, Rinne's test is positive in the affected ear with reduction of perception with the Absolute Bone Conduction Test. There are some exceptions to this rule in which a true negative Rinne appears to be present in the affected ear, and we

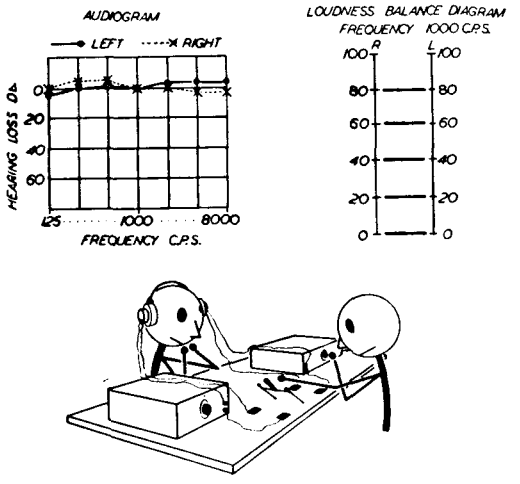


FIG. 3.

hope in a later communication to provide more detailed information upon this apparent anomaly.

For the purpose of the present paper, however, it can be taken that the classical tuning fork tests in VIIIth nerve neurofibroma, as in Ménière's disease, yield the conventional picture of so-called nerve or perceptive deafness. In other words, although different anatomical components of the perceptive mechanism are affected in these two groups of subjects, the diagnostic recognition of this difference is not possible by means of these old criteria.

Consideration may next be given to the new criteria and, in particular, the manner in which they succeed where the old criteria fail.

The Loudness Recruitment Test.—This has been described by its originator, Fowler of New York (1936), by Steinberg and Gardner (1937) and others. We have also described it briefly in our previous communication (Dix *et al.*, 1948). It is still not as well known as it deserves to be, and for this reason a further brief outline of the test procedure and its rationale may not be out of place.

The application of the test procedure is shown in Fig. 3. The subject wears

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a pair of telephone receivers, each supplied by a separate pure-tone audiometer, or preferably by a single audiometer with arrangements for independent adjustment of the intensity in the two receivers. The frequency of the sound stimulus is the same in each receiver, and the tester switches it alternately from left to right. The purpose of the test is to ascertain and mark upon the ladder diagram two series of intensity levels, one for the right ear and one for the left, each intensity level for the right ear being connected across the diagram with the level for the left ear found by experiment to give a sensation of equal loudness.

The figures given on the diagram represent decibels above the normal threshold. The subject is a normal one, and equal intensities above threshold give equal loudness sensations in the two ears. The rungs of the ladder, therefore, lie horizontally from bottom to top.

In Fig. 4 are shown side by side with those of a normal subject the audio-

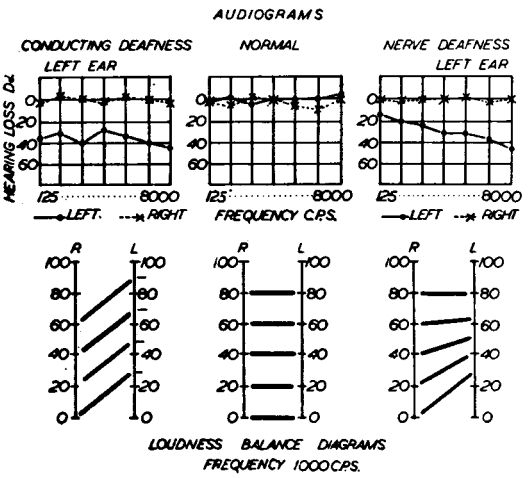


FIG. 4.

grams and loudness recruitment diagrams of two typical cases of unilateral deafness. On the left the deafness is due to a lesion of the conducting mechanism of the middle ear, and on the right to Ménière's disease affecting the left labyrinth. In each case the test frequency selected is 1,000 cycles, at which point the audiogram shows a threshold shift for the affected ear of 30 db. The result obtained in the case of conductive deafness indicates that the loss of sensitivity of the affected ear, 30 db. at threshold, remains constant at 30 db. throughout the entire intensity range. This finding is an unvarying one in conductive deafness, and is explained on the straightforward assumption that the obstruction caused by middle-ear disease to the sound waves on their way to the inner ear introduces an attenuation factor—in this case 30 db.—which is constant at all intensities.

A very different result is obtained in the case of nerve deafness. The audiogram is substantially the same with a threshold shift at 1,000 c/s of

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30 db. The balancing points at threshold are identical with a 30 db. displacement upwards for the L. ear. On ascending the intensity scale, however, it is found that the sensitivity loss or deafness of the left ear, 30 db. at threshold, becomes progressively less until at 80 db. equal intensities at the two ears evoke equal loudness responses.

In other words, the deafness of the affected ear present at threshold disappears at higher intensities, and this, in its simplest terms, constitutes the phenomenon of loudness recruitment.

There is general agreement by all who have since investigated the loudness recruitment phenomenon that it is absent in deafness due to uncomplicated middle-ear disease, so-called conductive deafness. On the other hand, it has frequently been demonstrated in a wide variety of disorders of the internal ear and cochlear nerve, including Ménière's disease, which are collectively described as " nerve deafness " and it has come, therefore, to be regarded in a somewhat

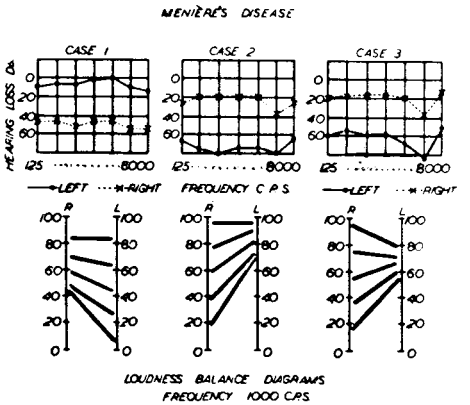


FIG. 5.

uncertain manner as a valuable indication of " nerve deafness ", using the term in the same wide sense connoted by the old tuning-fork tests.

The results of the Loudness Recruitment Test in Ménière's disease and VIIIth nerve neurofibroma can be stated very simply.

In Fig. 5 are shown the audiograms of three cases of Ménière's disease, with their loudness recruitment diagrams. It will be seen that in every case the sensitivity loss at threshold of the affected ear is completely eliminated at high intensities. In the third case well-marked over-recruitment is seen ; that is to say at high intensities the loudness function of the affected ear exceeds that of the normal ear.

In Fig. 6 are shown the audiograms in three cases of VIIIth nerve neurofibroma with their loudness recruitment diagrams. It will be seen that in all three the sensitivity loss of the affected ear at threshold is maintained at all intensity levels and this is the result generally found in degeneration of the cochlear nerve due to VIIIth nerve neurofibroma. In a few cases, partial recruitment is found to occur and to this we shall refer again. In only one case have we encountered complete recruitment.

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Summarizing these findings, it can be said that in Ménière's disease with disorder of the hair cells loudness recruitment is always present. It is generally complete, and over-recruitment is not very uncommon. In VIIIth nerve neurofibroma, however, loudness recruitment, hitherto described as being uniquely distinctive of nerve deafness, is characteristically not present at all. Instead, there occurs a type of response which is much more in accordance with that found in middle-ear deafness.

In a proportion of cases, however, an incomplete degree of recruitment is found to be present. This we think likely to be due to some degeneration of the hair cells, secondary to involvement of the cochlear blood supply by tumour pressure.

It will therefore be seen that the first of the new criteria, the Loudness Recruitment Test generally makes possible a clear distinction between nerve deafness due to disease of the end-organ and nerve deafness due to disease of the cochlear nerve fibres.

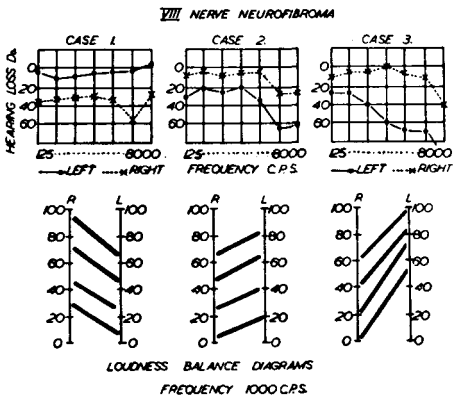


FIG. 6.

Consideration may next be given to the second of the new criteria.

The Intelligibility Test for amplified speech.—Otologists have for many years been much interested in the way in which deaf patients vary in their ability to hear speech and to understand it. In general they have come to recognize that in nerve deafness, as assessed by the old criteria, inability to understand speech tends to be out of proportion to the threshold sensitivity loss or deafness. This means, in effect, that while such patients hear the sounds of speech, their capacity to understand them is disappointingly low. With conductive deafness however, this disproportion between hearing and understanding is not present. If, in fact, the sounds of speech can be made to reach the cochlea at all, they seem in general to be well understood.

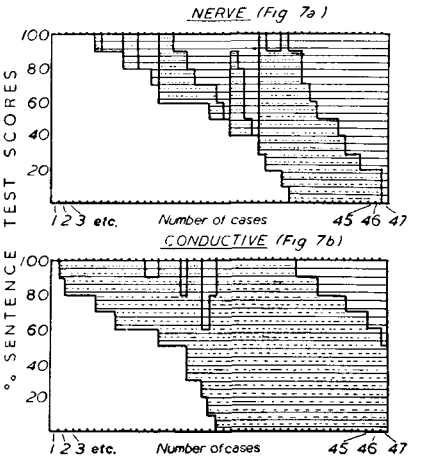
Otologists have also come to recognize that in the former group, restoration of intelligibility, as, for example, by means of hearing aids, tends to be inadequate. In the latter group, however, hearing aids are generally very successful in restoring intelligibility.

The validity of these two important beliefs is illustrated by the results of

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two investigations we were able to carry out at Queen Square in 1945 in the course of work carried out on behalf of the Electro-Acoustics Committee of the Medical Research Council.

From our clinical material 47 cases were selected in which the tuning-fork tests yielded an unambiguous picture of nerve or perceptive deafness, and another 47 in which these tests were equally clear in indicating conductive or middle-ear deafness. Otoscopic and other investigations were in agreement with the results of the tuning-fork tests. A direct investigation was then carried out of the subject's capacity to understand speech by means of the Fry-Kerridge sentence tests spoken in an ordinary conversational voice at 3 ft. both with and without hearing aids. In all cases lip-reading was excluded. Disability or deafness for tête-à-tête conversation was rated at nil when 100 per cent. of the sentences were understood. When none of the sentences was understood, the disability was rated at 100 per cent. Further tests were then



FIGS. 7A and 7B.

carried out with hearing aids, and the degree of improvement ascertained. The results are shown graphically in Fig. 7.

In Fig. 7A each case is represented by one of the small sections along the base-line. There are 47 in all. The sentence percentage scores are ranged along the vertical axis. The clear area represents hearing, i.e. percentage of sentences understood, while the hatched area represents deafness or percentage of sentences not understood. The hatched area which is stippled represents the amount of deafness compensated by means of a hearing aid. It will be noted for example that the first five cases understood 100 per cent. of the sentences and had, therefore, no disability.

At the other end of the scale, Case 47 has a disability of 100 per cent. unaided, and regained none of this with a hearing aid. The impression gained from the figure is of a rather large clear area indicating an initial deafness which is not very severe. Secondly, of a rather small stippled area, indicating a rather small amount of improvement with hearing aids.

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In Fig. 7B, however, a different state of affairs is observed, namely, a much smaller clear area and a much larger stippled area. These results are expressed quantitatively in Fig 8 which shows that in the case of conductive deafness the unaided hearing capacity, " intact hearing ", is small and that practically all of the hearing loss is regained by means of hearing aids, leaving only a small fraction which may be termed residual hearing loss.

With nerve deafness, however, the unaided hearing capacity is very much larger, but comparatively little of the hearing loss is regained and the residual hearing loss is quite considerable.

It may, therefore, be said of these findings that they illustrate very well the truth of the generally accepted belief that deafness lends itself readily to alleviation by means of hearing aids when it is due to disease of the middle ear, but much less readily when it is due to disease of the nervous mechanism of the internal ear or cochlear nerve.

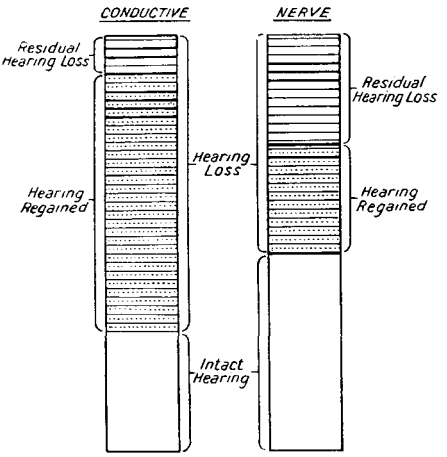


FIG. 8.

A further point which calls for consideration is the belief generally held by otologists that the pure-tone audiogram and the capacity to understand speech tend to be quite differently related in nerve deafness and in conductive deafness, and in this respect also a comparative study was made of the same two groups of subjects. For the evaluation of the pure-tone audiometric losses, we have made use of a development of the Sabine Percentage Hearing Loss Chart recommended by the American Medical Association (1942). This is shown in Fig. 9. The values inscribed in each column are cumulative from above downwards. Thus, the value for the percentage of total hearing loss contributed by the hearing loss at any one of the four frequencies, 512, 1024, 2048 or 4096 cycles/sec. can be read directly from the chart, the figure in question being given in the square immediately above the level at which the hearing loss curve crosses the column concerned. Addition of the four cumulative values then gives the total percentage loss.

In the case of an audiometric loss represented by the line A———A′ the

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total percentage loss would be given by addition of the figures immediately above the line : 10·4, 20·5, 25·1 and 15.

This method of arriving at a percentage evaluation of hearing loss from a pure-tone audiogram has considerable advantages over any earlier method of this kind. It is, however, still open to the objection that it deals only with threshold intensities and its results, therefore, are not likely always to be related to the behaviour of the ear at intensities well above threshold. Direct measurement of speech intelligibility at different intensity levels is, therefore, a method which we have come to find indispensable.

The method we have employed was devised in connection with the work of the Electro-Acoustics Committee of the Medical Research Council (1947).

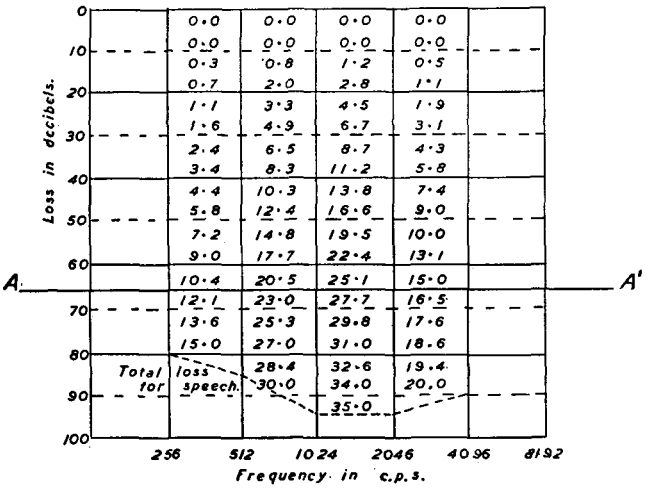


FIG. 9.

Pure-tone audiometer chart for evaluating percentage hearing loss (as approved by the Council on Physical Therapy, American Medical Association).

The patient is seated with the ear under test at a fixed distance from a loud speaker. Recorded lists of words are then reproduced on the speaker by means of an amplifier system. The method used for recording the results is shown in Fig. 10. The amplification can be adjusted within the range of 100 db. and an intensity level at the ear of the listener corresponding to that of the conversational voice at 3 ft. is obtained with an amplifier setting at 50 db. Other settings specified on the chart correspond to the loud conversational voice at 3 ft. This is obtained at 64 db. and the so-called church level at 36 db. The response curves of a normal subject are shown on the right of the chart. At a setting of 30 db. (well below church level) the score would be 90 per cent. At 17 db. the score would be 40 per cent. of words corresponding with the 90 per cent point of the sentence intelligibility curve. This 40 per cent. score

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for words is a very critical level since understanding of sentences falls off precipitously at any point below it, as shown by the curve. Thus, if it is desired to know the hearing efficiency of any subject at any of these three levels, church, ordinary conversation or loud conversation, it is required only to know whether at these levels the subject's word score is above or below the critical 40 per cent. level.

The curves A, B and C are obtained from three different subjects with varying degrees of deafness.

Subject A.—Score with ordinary conversation, nil. Score with loud conversation below the critical level.

Subject B.—Score with loud conversation well above the critical level ; and with ordinary conversation just below the critical level.

Subject C.—Score for ordinary conversation well above the critical level ; at church level, just below the critical level.

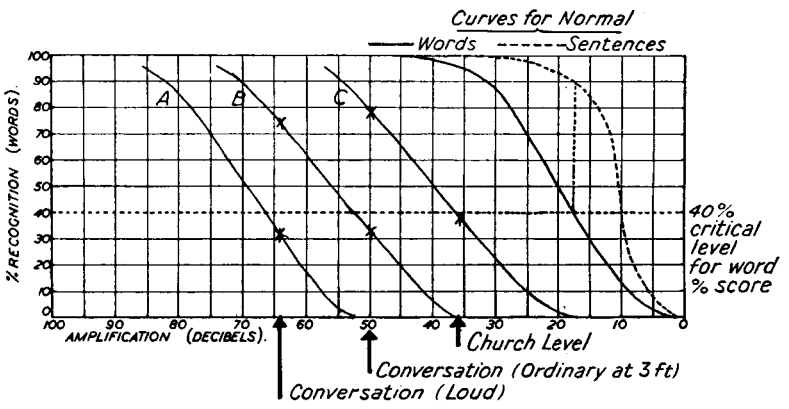


FIG. 10.

In all cases the loss for speech is concisely expressed by the displacement of the curve to the left of the normal, as measured along the critical 40 per cent. level.

In Case A, this displacement is 50 db. In Case B, 35 db. and in Case C 18 db.

The results of the comparative study in our two groups of subjects of speech audiometry carried out in the manner described and of pure-tone audiometry evaluated in the manner of Sabine, is shown in Fig. 11 in the form of a scattergram. As already stated, the otological findings in one group including the tuning-fork tests were characteristic of middle-ear deafness, and in the other, of " nerve " or perceptive deafness.

In Fig. 11 the percentage losses as derived by the Sabine method from the pure-tone audiogram are plotted along the vertical axis, while speech deafness in decibels is plotted along the horizontal axis.

The distribution of the two groups of symbols reveals a systematic differentiation. Each of the vertical columns bounded by successive ordinates can be regarded as a particular grade of speech deafness. In each such

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column the symbols are widely distributed, dots lying well above the crosses. In other words, a particular grade of speech deafness is compatible with a degree of pure-tone deafness which tends to be higher if it is due to a middle-ear lesion than to a nerve lesion.

Conversely, each horizontal band of the chart may be regarded as a particular grade of pure-tone deafness. In each such band, the dots or middle-ear symbols are well to the right of the crosses or nerve symbols. In other words, in any one grade of pure-tone deafness the deafness for speech tends to be less if it is due to a middle-ear lesion than to a nerve lesion. These results confirm and systematize the generally-held belief that without knowledge of the pathological basis of the deafness, i.e. whether it is of the " conductive " or

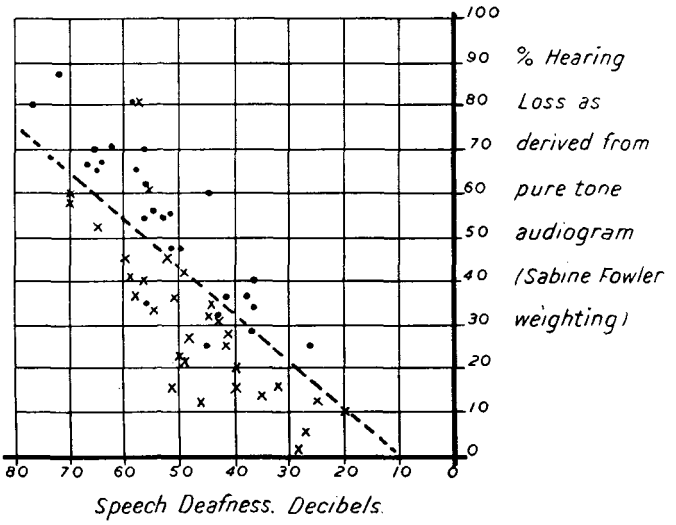


FIG. 11.

Key : Conductive deafness ●. Nerve deafness ×.

" nerve " type, pure-tone audiometric findings cannot reliably be related to the hearing capacity for speech. They exemplify, too, the general truth of the belief that in nerve deafness, as assessed by the old criteria, loss of intelligibility for speech tends to be disproportionately larger than the pure-tone audiometric loss, and tends also to be disproportionately larger and more difficult to compensate by means of hearing aids. This does not, however, apply to all of our cases of " nerve " deafness as will readily be seen from the situation of some of the x symbols in the scattergram, and a closer analysis of the anatomical basis of our cases of " nerve " deafness has, therefore, been attempted by means of systematic application of the speech audiometric tests in two groups of subjects in whom the deafness was due respectively to Ménière's disease and VIIIth nerve neurofibroma.

In Fig. 12 is shown, for comparison, the unaided and aided speech audiogram of two cases of middle-ear deafness.

In Case A the speech deafness as measured at the critical level is 61 db. It

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will also be noted that the unaided curve lies approximately parallel to the normal curve, a point of considerable importance.

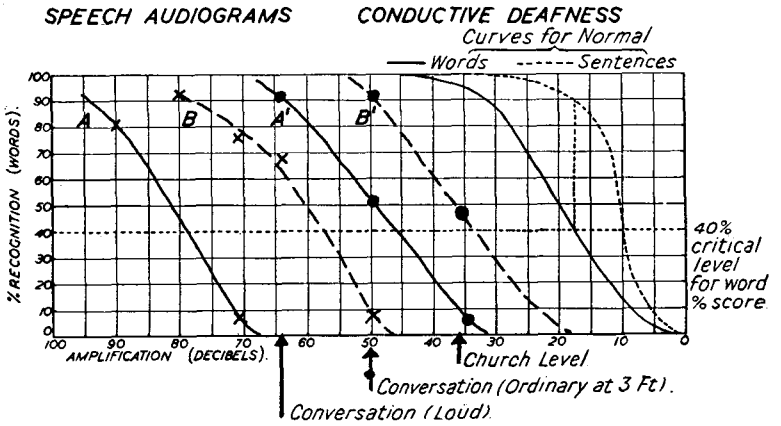


FIG. 12.

Speech audiograms. Conductive deafness.

Speech deafness (Decibels)	Unaided	Aided
Case A	61	27
Case B	40	16

The aided curve shows very substantial displacement to the right, towards the normal unaided curve, the speech deafness, in fact, being reduced to 27 db.

With Case B a similar result is obtained. The unaided curve is of normal contour. It is displaced some 40 db. to the left of the normal curve. With an aid the speech deafness is reduced to 16 db. As would be expected, the aided curves reproduce in both cases the form of the unaided curves.

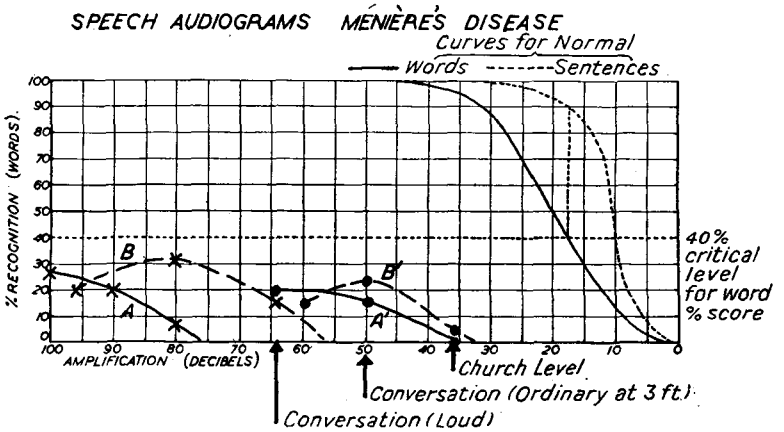


FIG. 13.

Speech audiograms. Ménière's disease.

In Fig. 13 are shown the very different results obtained in two cases of Ménière's disease. The unaided curves do not lie parallel to the normal curve.

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Both show a downward slope towards the left, indicating a disproportionate failure of intelligibility with amplification. With hearing aids, although the unaided curves are displaced to the right, their form is unchanged and the improvement in intelligibility is disappointingly small. Thus in neither case is there obtained a word percentage score for ordinary conversation which is above the critical 40 per cent. level. Both of these cases were typical of Ménière's disease in showing complete loudness recruitment and *it can thus be said of Ménière's disease that while recruitment of loudness is the rule, recruitment of intelligibility, which might be expected, may be conspicuously absent. In fact, the opposite would appear to occur, namely, that as loudness increases intelligibility falls off.*

We come, finally, to speech audiometry in cases of nerve deafness due to VIIIth nerve neurofibroma. The results of speech audiometry in two cases of nerve deafness due to VIIIth nerve neurofibroma are shown in Fig. 14.

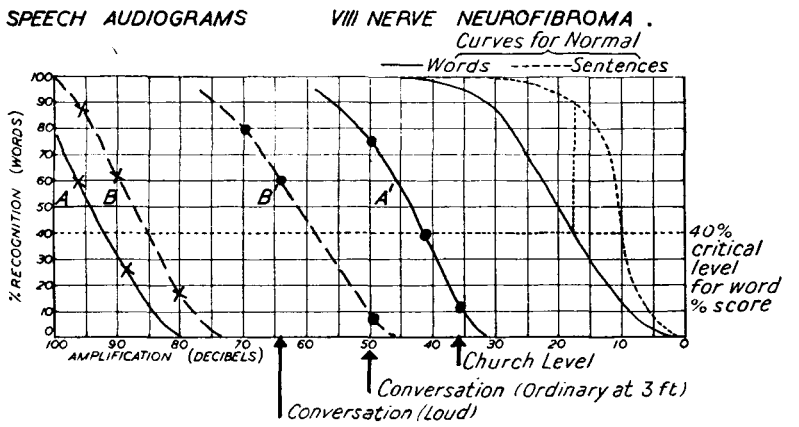


FIG. 14.

Speech audiograms. Eighth nerve neurofibroma.

Speech deafness (Decibels)	Unaided	Aided
Case A	75	25
Case B	67	41

In both cases, the unaided and aided curves lie parallel to the normal as shown in Fig. 12. With all four curves intelligibility continues to improve with amplification up to high levels and in both cases it is possible to bring the percentage score for ordinary conversation well above the critical 40 per cent. level.

It will be seen, therefore, that as with the first of our new criteria, so with the second. Quite different results are obtained in end-organ lesions and in nerve-fibre lesions. Both, therefore, succeed where the old criteria fail, in making clear the important distinction between these two conditions.

The principles which underlie the use of these new criteria for so useful an analysis of the anatomical basis of nerve deafness are still far from being well understood. In certain particulars, too, the facts themselves cannot yet be regarded as well established. This applies in particular to the reactions to

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amplified speech which are encountered in Ménière's disease. Speech audiometric curves, having the form displayed in Fig. 13, though highly characteristic of this disorder, are by no means always present. Indeed, some subjects of Ménière's disease, particularly in its quiescent phases, derive considerable benefit from hearing aids, although here, again, they practically all show a marked unwillingness to use these instruments, a clinical fact which is perhaps more distinctive of the condition than the results of audiometric or indeed any tests.

It is necessary finally to emphasize the remarkable way in which, both with the Loudness Recruitment Tests and with speech audiometry, the results obtained in nerve-fibre lesions approximate to those obtained in lesions of the conducting mechanism of the middle ear.

Some explanation for this resemblance has been offered in our previous discussion of the loudness recruitment phenomenon (Dix *et al.*, 1948) and it is hoped that further light will be thrown upon it by the results of investigations at present in progress.

Clinically, the resemblance is of great practical importance, and helps to explain the serious diagnostic error which is still encountered of mistaking an early VIIIth nerve neurofibroma for early unilateral otosclerosis.

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The Causes of Perceptive Deafness

By R. R. SIMPSON, F.R.C.S.Ed.

I WANT to approach this subject, in the first place, from the clinical aspect. For this purpose I have taken 100 cases of perceptive deafness seen by me during the last year at the Hull Royal Infirmary. All the cases showed the accepted clinical findings, including audiograms both for air and bone conduction. The causes of the deafness were, in my view, as follows:

Cause		No. of cases
(1) Results of chronic otitis media	.. .. ..	16
(2) Traumatic :	(*a*) Gunfire, bomb-blast, explosions	12
	(*b*) Occupational	11
	(*c*) Concussion	5
	(*d*) Fracture of the skull	2

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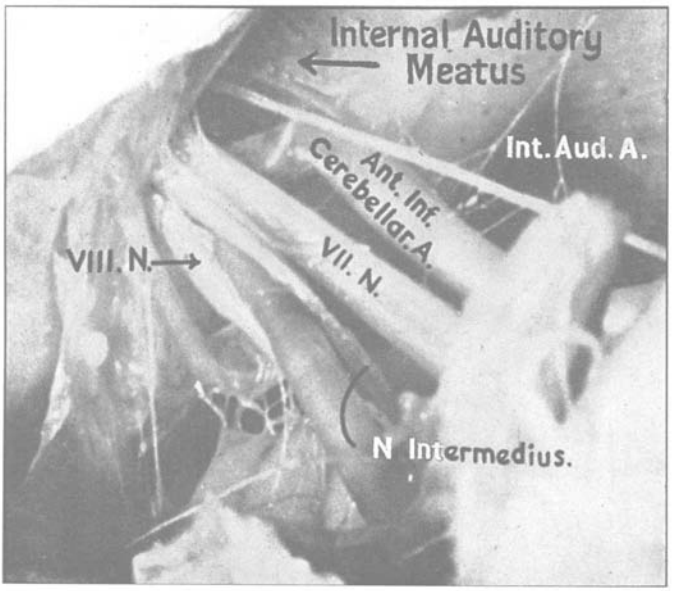


FIG. 1.

After S. Sunderland, *Brain*, 1945, **68**, 25. "The arterial relations of the internal auditory meatus."

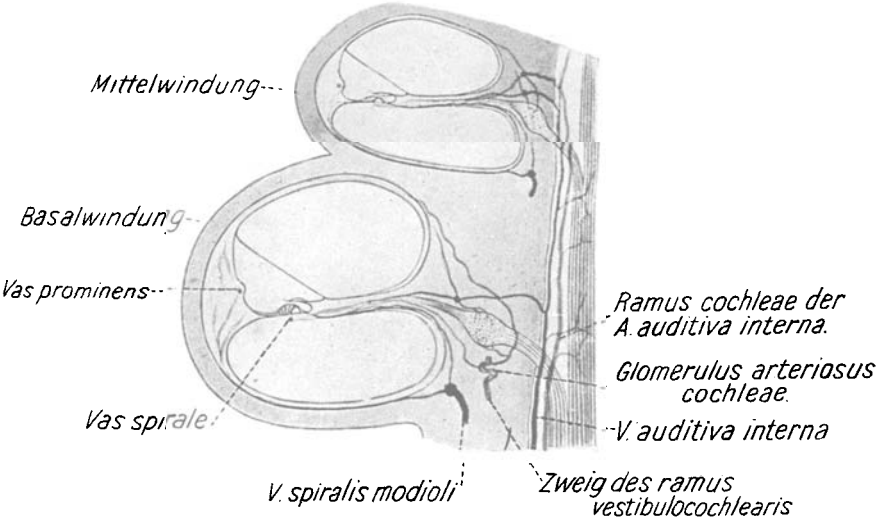


FIG. 2.

Diagram of the distribution of blood vessels in the cochlea. (From Abderhalden.)

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(3)	Ménière's disease	10
(4)	Syphilis	9
(5)	Arteriosclerosis	15
(6)	Influenza	4
(7)	Measles (without otitis media)	2
(8)	Paget's disease	2
(9)	Diphtheria	1
(10)	Diabetes	1
(11)	Anæmia (hypochromic microcytic)	1
(12)	Mumps	1
(13)	Scarlet fever (without otitis media)	1
(14)	Cerebral thrombosis	1
(15)	Thyrotoxicosis	1
(16)	Lead poisoning	1
(17)	Undetermined	4
	Total	100

It is not a comprehensive list. I have excluded, for example, all congenital causes. It is not to enumerate the detailed causes that I show this list. It is rather to consider the *primary* causes of perceptive deafness as illustrated by such a list. Such a list can be divided into three main primary causes: (*a*) neuritic; (*b*) traumatic; (*c*) vascular.

(1) Cases where the nerve elements of the Perceptive Mechanism are primarily involved, e.g.

Influenza	4
Measles	1
Diphtheria	1
Diabetes	1
Mumps	1
Scarlet fever	1
Lead poisoning	1
	10 cases

In none of these cases was there any evidence of a previous otitis media. The membranes appeared normal and intact. In the history of the case there appeared to me to be a direct connection between the disease mentioned and the incidence of the deafness. It appeared that these diseases exerted a direct toxic effect on the nerve cells of the cochlea or the nerve itself.

Eggston and Wolff[2] suggest that in the cases of perceptive deafness due to drugs, the lesion is an allergic reaction in the cochlea itself.

(2) *Traumatic causes.*—These cases are divided into four groups, roughly on the degree of trauma applied. In the case of the fracture of the skull and concussion (seven cases) the degree of trauma is the most severe. Next come the cases caused by explosions, bomb-blast and gunfire (twelve cases). And the least in degree of trauma are those due to various occupations (eleven cases).

In connection with the cases of occupational causes and the gunfire and explosion causes, it is interesting to refer to recent views on the theories of

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hearing. Reboul[7] considered the hydrodynamics of the propagation of a fluid wave within small conical cavities. He computed the movements of these waves within such tiny elongated cavities as the scala vestibuli and scala tympani. He maintains that the pressure waves and waves of elongation set up by each tone is a function of the vibratory frequency of that tone. Such waves will impinge upon the reception endings of the organ of Corti at specific points. The point of impingement in the cochlea of a given wave can be calculated mathematically. This theory makes it unnecessary to assume that thousands of little microphones are lined up in the cochlea waiting for the tones to locate them. On the contrary, fluid vibrations at a given frequency will always produce sufficient pressure at a mathematically predictable point to set into action a relatively simple perceptive mechanism. Much more proof of these propositions is, of course, needed. But there is some experimental support for it, e.g. Larsell's[5] histological studies of the developing cochlea of the pouched opossum embryo, from which movement reflexes and oscillographic responses were secured only as various areas reached functional activity. In discussing these findings, one of the joint authors of the paper, McCrady, comes to the same conclusions as Reboul. McCrady[5] puts it this way: "The maximum efficiency of pressure exerted by the energy of sound wave travelling through a liquid in a tube, depends upon the relation of wavelength to the circumference of the tube."

He found that by mathematical calculation he could expect a certain area of the cochlea to be receptive to the precise sounds which, from other experimental evidence, such as Hallpike's, we believe are received there. This theory may well explain the mechanism by which sound waves reach the appropriate area of the cochlear mechanism and set off the stimulus which is interpreted in the cortex by us as the sound corresponding to that stimulus. But it does not explain why sounds of varying pitch but of great intensity, or of varying pitch but of sustained and lesser intensity, i.e. the conditions producing occupational deafness, etc., all tend to produce the lesion in the cochlea which we have come to refer to as "the 4096 dip". On the basis of this theory we should expect to find the lesion in occupational deafness and gunfire, etc., at the point in the cochlea where we believe the pitch involved is received by the cochlea. It seems to me that there must be some other explanation of the "4096 dip" commonly found in traumatic cases. Here I would point out that this "dip" is not confined to traumatic cases. It is also found in many cases of chronic otitis media of long standing, where there has been no associated trauma. Is there an explanation common to both causes, i.e. other than trauma?

(3) *Vascular causes.*—First let me mention the cases in this group which I think you will accept without question. They are:

	No. of cases
Ménière's disease	10
Arteriosclerosis	16
Anæmia	1
Cerebral thrombosis	1
Thyrotoxicosis	1
	28

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The next group, sixteen cases, are those which are attributed to the results of chronic otitis media. In support of my contention that these are vascular in origin I would quote Eggston and Wolff[2]:

"No inner ear appears microscopically perfectly normal in the presence of a well-established otitis media. Dean and Bunch on the basis of audiometer records gave clinical evidence of this fact . . . Clinically the hearing and vestibular functions return to normal when the otitis has been cured, as a general rule."

It is my belief that the sixteen cases here mentioned were, for various reasons, caused by vascular changes in the cochlea as a result of vascular changes in the middle ear from old chronic otitis media. If we accept these as being of vascular origin, then we have a total, so far, of forty-four.

There are still nine cases of syphilis to classify. We assume that some of the cases will show cochlear changes due to an endarteritis obliterans and some will show trophic changes in the nerve. If, for the purpose of this discussion, we say that four out of the nine cases show endarteritis obliterans, we have now a total of forty-eight cases out of 100 in which the vascular changes are the primary cause of the perceptive deafness.

I want to acknowledge here my indebtedness to Professor Harris[4] of the Anatomy School, University of Cambridge. It was he who drew my attention to some of the anomalies in the accepted ideas of the vascular supply of the cochlea.

Sunderland[8] has drawn attention to the fact that the exact size of the internal auditory artery, in relation to the other structures in the internal auditory meatus, is rarely appreciated. The reason being that, in the dissection of this area, the brain is lifted up and the attachments cut in order to expose the internal meatus. If, however, the brain is gently retracted, Fig. 1 is the picture that is seen. Note, first, the unexpected length of the artery, and secondly, its small calibre when compared to the nerves in the meatus. Sunderland found that in 17 per cent. of specimens the internal auditory artery took origin independently from the basilar artery and entered the meatus as a separate arterial channel anterior to the VIIth and VIIIth nerves. In the remaining 83 per cent. the artery took origin from a variable point on the anterior inferior cerebellar artery. The evidence favours, therefore, a predominant origin of the internal auditory artery from the anterior inferior artery. When the anterior inferior cerebellar artery passes between the nerves entering the internal auditory meatus, it does so, in most instances, between the main motor division and the portio intermedia.

Sunderland adds that the knowledge of such a relationship is of considerable functional importance in surgical explorations of the cerebello-pontine angle and in connection with certain disturbances of nerve conduction which may possibly arise as a direct consequence of mechanical pressure exerted on the nerves by atheromatous, enlarging, abnormally pulsating or tortuous vessels.

In the internal auditory meatus, this artery gives off a cochleo-vestibular branch, which divides into a trunk supplying the proximal two-thirds of the basal coil of the cochlea, and a posterior vestibular artery. The continuation of the internal auditory artery divides into two trunks, one entering the modiolus and supplying all of the cochlea excepting the proximal two-thirds of the basal

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coil, and the other the anterior vestibular artery. These branches are all terminal or " end-arteries " constituting the sole blood supply for their respective areas of distribution.

Next, Professor Harris draws attention to the generally accepted or conventional diagrams of the vascular supply of the cochlea in order to contrast them with real specimens. Fig. 2 is taken from Abderhalden. Eggston and Wolff[2], quoting Nabeya and Assia, also accept this arrangement of the vascular supply. Commenting on this, Professor Harris lays down the general principle that whenever an artery goes through a long fibrous canal, it is *not* accompanied by one vein but by a plexus of veins (cf. pampiniform plexus, vertebral venous plexus, etc.). Professor Harris has confirmed this view by examining sections to show the vascular supply in the labyrinth of a ten-and-a-half weeks human embryo. Therefore, in the internal ear one should expect not only arterial phenomena but also venous phenomena, e.g. varicosities, thrombosis, etc.

Concerning the physiology of the vascular supply of the labyrinth, Mygind and Falbe-Hansen[6] maintain that : (*a*) In contrast to the general reaction of the organism, the labyrinth shows vasoconstriction after the injection of histamine, vasodilatation after adrenaline, presumably thus compensating the universal change of blood-pressure. (*b*) The water-binding power of the labyrinthine cells is increased by injections of histamine, lowered by adrenaline.

There is some evidence, therefore, for supposing that the vascular mechanism of the internal ear is designed to counteract sudden changes in the vascular tension of the rest of the body.

I mentioned earlier that Reboul's theory did not explain " the 4096 dip ". It will be seen that a vascular lesion would. If we assume, therefore, that in the traumatic causes where a " 4096 dip " is found, the lesion is vascular, we can add to the vascular causes a further twenty-three cases (twelve gunfire, blast, explosion and eleven occupational). This would bring the total of the vascular causes up to seventy-one out of 100 cases, a figure very much higher than I had previously considered.

The evidence to support this vascular theory, so far as I am aware, is based on clinical grounds and theoretical speculation. I know of no direct experimental evidence to support it.

Asherson[1], in discussing the question, maintains that " any noise above a certain intensity is a stimulus, which reacts through the ear and produces a spasm of an end-artery distributed to that part of the high-tone division of the cochlear nerve which transmits the frequencies 4096 ". But he does not venture an explanation of how this reflex works. Instead he asks : " Is the original arteriole spasm a reflex *viâ* the tympanic membrane and the stapedius muscle ? Is this reflex protective in gearing down the conductivity of the nerve to prevent too loud a sound being transmitted ? "

These and many other questions must be answered by the evidence of future experiments in this almost unexplored field.

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MR. E. D. D. DAVIS asked Mr. Hallpike for his views on the subject of masking, since in his own experience the procedure was often of little clinical value.

MR. TERENCE CAWTHORNE said that with the new concept presented by the authors it was possible clinically by means of these tests to distinguish between a lesion of the end-organ as seen in Ménière's disease and a lesion of the nerve trunk as seen in a neurofibroma. At a neurological hospital such problems were common and they had the experience that the method put forward was of real practical value. He believed that this study would be of great help in differentiating between a lesion of the end-organ and the trunk of the VIIIth nerve.

MR. SIMSON HALL said that he had listened to the paper read by Dr. Hallpike with great pleasure and profit and congratulated him and his colleagues on the very practical approach which they had adopted towards these problems. He had had the advantage of hearing Dr. Hallpike talking on this subject some time ago in the Section of Neurology, and what he had said that morning put in a more practical and everyday fashion what he had advanced on the previous occasion. He was extremely interested in his speech intelligibility differential test. One question arose on this matter. Dr. Hallpike told them that he was using a loudspeaker for testing. He would like his opinion as to whether he preferred to use live speech or reproduced speech. Would he consider these tests of the same value if carried out with a more crude method? In other words, was it essential to have scientifically graded speech? The speech intelligibility curve which he had shown demonstrated in a graphic form a point which was of interest to those who were dealing with the operative side of the correction of deafness.

There was no doubt that direct assessment of the intelligibility of speech was needed in assessing the value of the fenestration operation. They had found in many cases that although they had got a satisfactory rise in the audiometric chart, the raising of the lower tone limit did not give the patient any real satisfaction, and sometimes his condition was worse than before, because owing to nerve loss, the intelligibility suffered very considerably.

Referring to Mr. Simpson's paper, he was extremely interested in the result of vascular change in otitis media. In a paper which he himself had read to the Section a little while ago he had pointed out that in early otitis media the nerve change was obvious and was the last thing to recover. He wondered whether they could link that up with some of the vascular changes of which Mr. Simpson had spoken. The same thing occurred after the fenestration operation. Mr. Simpson had certainly given them some new ideas on that subject.

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AIR COMMODORE E. D. D. DICKSON showed records of cases of perceptive deafness occurring in airmen who had undertaken high altitude diving. None of these experienced pain during the dive. The eustachian tubes were patent and the tympanic membranes were normal in appearance when seen. Audiometric tests showed a perceptive type of deafness and the caloric stimulation of the labyrinth gave a normal response. (These latter findings were checked and corroborated by Hallpike at Queen Square.) The first case dived from 14,000 ft. and the second from 30,000 ft. to 10,000 ft. The third case lost consciousness whilst looping an aircraft and regained it at 3,000 ft. On landing all complained of deafness in one or other ear which has persisted. What was the cause of such deafness? Was it due to pressure changes in the inner ear confined to the cochlea?

MR. H. V. FORSTER said that having listened to the discussion, he had been reminded of an old question put to us with anxiety by our deafened patients. Would the use of an electrically powered aid eventually damage the hearing?

Recent contributions by European workers, for example, de Maré, Huizing and Pothoven, renew interest in this question when applied especially to cases of perception deafness associated with the loudness recruitment phenomenon.

He thought it would be agreed that our answer should be definitely encouraging in cases of obstructive or middle-ear deafness, but when the inner-ear system is at fault, the answer must be guarded and be dependent on the improved design of hearing aids.

Certain developments in the design of these instruments are expressed by such terms as "peak clipping" and "compression amplification", but he, Mr. Forster, would not press Mr. Hallpike and his co-workers for information on this technical problem, which he hoped might be discussed at some future meeting of the Section.

Older people are grateful to the practised public speaker and experienced politician, not only for his clear diction, but also for the well-spaced words and sentences. This latter quality, however, is often impatiently criticized by the younger listener.

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DR. C. S. HALLPIKE in reply to Mr. Davis said that for their own part they paid a great deal of attention to masking, and they found it absolutely essential in many of the cases with which they had to deal. He would not like to say that they knew the best way to do it, for the technical difficulties were quite considerable, but he did think it a very important subject indeed.

In answer to Mr. Simson Hall, he did not think they had any objection to live speech, which had certain advantages, but it was a question of time and trouble. The technical difficulties of reproducing speech were now so small that he would have thought that the use of properly recorded speech for this kind of work might be recommended very heartily.

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