

Observations on the Pathological Mechanism of Conductive Deafness in Certain Cases of Neuroma of the VIII Nerve

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UNTIL recently it has been our impression that the tuning-fork tests in cases of unilateral deafness due to VIII nerve neuroma conform to the classical pattern of so-called "nerve deafness". That is to say, the sound of the fork with Weber's test is typically referred to the normal ear, Rinne's test is positive, and perception with the absolute bone conduction test is reduced. Should the Rinne be negative it is a false negative; that is to say, the apparently good perception by bone conduction with the affected ear is due to the transfer of the vibrations to the opposite cochlea, through which it is in fact perceived. This well-known fallacy is recognizable and can be excluded by abolishing the perception by the normal ear of this bone-conducted sound by means of an air-conducted masking sound.

In the course of a previous communication made to this Section (*Proc. R. Soc. Med.*, 1949, **42**, 527), we said that there were some exceptions to this rule, in which a true negative Rinne appeared to be present in the ear affected by the neurofibroma and it is this point to which we return to-day.

So far as we know, all writers who have published their observations upon the tuning fork tests in the deafness of VIII nerve neuroma are in agreement that they conform to the classical pattern of nerve deafness and they have little more to say on the subject. All observers except one, T. H. Just, who in an excellent paper published in 1930 (*Proc. R. Soc. Med.*, **23**, 722) insisted that in certain cases of VIII nerve neuroma the negative Rinne is no artefact, but a true and, as he described it, "disturbing phenomenon".

In our own experience at Queen Square we have encountered a number of cases of this kind. In 2 of these death followed surgical intervention carried out soon after our examination and we were able, through the good offices of Dr. Greenfield, to carry out a histological examination of the temporal bones.

It is the purpose of this communication to describe our findings, and explain how we think they account for this interesting, and as Just described it "disturbing" anomaly—the negative Rinne—in the tuning-fork test results.

The first of our subjects was a man of 57, and the second a woman of 47. They were under the care of our colleagues at Queen Square, Drs. Walshe and Carmichael, to whom we are greatly indebted for permission to investigate the cases and to make use of the clinical notes. The findings in both were characteristic of an VIII nerve neuroma.

CASE I.—A. E., male, aged 57, under the care of Dr. F. M. R. Walshe.

Complaint: Pain behind the left ear and deafness, left ear, four years. Numbness left side of face and unsteadiness in walking, seven months. He had recently experienced difficulty in winking his left eye, and in tasting things on the left side of his tongue.

The pain behind the left ear had been intermittent, never very severe, and never lasted for more than a few hours at a time. The deafness had never been accompanied by tinnitus and in the patient's opinion was quite slight. The difficulty in walking was constant but not severe. He tended to run into things on his left side and was unable to turn round quickly without losing balance. He had never actually fallen or needed support, and there had never been any giddiness.

Neurological findings.—General condition: alert and co-operative.

Visual fields full, fundi normal. Pupils normal. External ocular movements full. Corneal reflexes: right brisk, left absent.

Diminished sensation to cotton-wool, pin-prick and cold over the whole distribution of the left V nerve, including the tongue and hard palate. Some weakness of the left temporal muscle. Slight weakness of the left side of the face of the lower motor neurone type, with impairment of taste over the anterior two-thirds of the tongue on the left. IX, X, XI and XII cranial nerves normal.

Motor system: Normal except for very slight inco-ordination with the heel/knee test on left side.

Neurological diagnosis (Dr. F. M. R. Walshe).—Left VIII nerve neuroma.

X-rays: Normal skull. No evidence of enlargement of the internal auditory meatuses. Ventriculography: Slight, symmetrical hydrocephalus. The aqueduct was lengthened and bowed slightly backwards. The fourth ventricle was displaced to the left, and the anterior border rotated to the right. *Conclusion.*—A left cerebello-pontine angle expanding process (Dr. J. W. D. Bull).

C.S.F. lumbar puncture: Pressure 160 mm. Cells: White nil. Red 5 per c.mm. Protein 260 mg. per 100 c.c.

Otolaryngological examination.—Hypo-æsthesia to pin-prick on left side of soft palate, left side of tongue and left inferior turbinate. Tympanic membranes normal. Other findings normal.

Cochlear function.—Whisper (normal 30 ft.): Left, at meatus. Right > 20 ft. Weber (512 c.p.s.) referred to left. Rinne, Left: Negative with masking of right ear. (The masking procedure was carried out by means of a speaking tube through which an assistant applied to the right ear a continuous sh-sh-ing sound.)

Pure tone audiometry (air conduction only).—With the left ear there was an approximately uniform loss (50 db.) for all tones up to 2,048 c.p.s. Above this frequency the loss increased and 8,192 cycles could not be heard at the maximum output of the audiometer (90 db.). There was also a slight loss of hearing in the right ear (Fig. 1A).

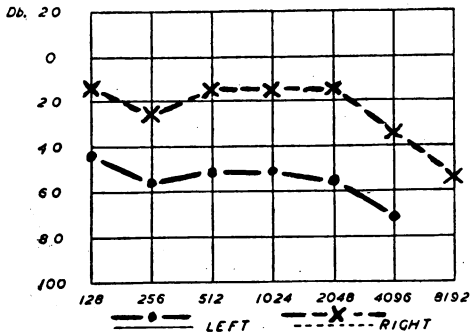


FIG. 1A.—Case I.

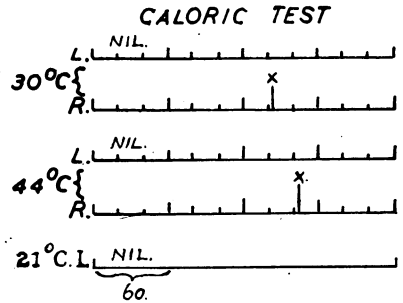


FIG. 1B.—Case I.

Vestibular function.—Posture and gait: A marked tendency to drift to the left when standing or walking with the eyes closed.

Spontaneous nystagmus: With the head erect, first degree vestibular nystagmus was present both to the left and to the right, that to the left being of greater amplitude than that to the right.

Positional nystagmus: In the supine position with the head back and to the left second-degree positional nystagmus of the central type was present to the right. Optokinetic nystagmus: This was markedly diminished to the left, particularly at higher speeds of the drum.

Caloric responses: Complete abolition of the left responses with some directional preponderance to the right of the right responses (Fig. 1B).

Summary of otological findings.—*Cochlear function:* Partial deafness of the left ear; conductive type. *Vestibular function:* Spontaneous nystagmus. Positional nystagmus of the central type and abnormality of optokinetic nystagmus. Complete loss of the left caloric responses.

Otological diagnosis.—Left VIII nerve neuroma.

Operation.—The left cerebello-pontine angle was explored on 28.10.46 by Mr. Wylie McKissock. The left cerebellar hemisphere was lifted and retracted medially to reveal a small typical acoustic neuroma lying in the cerebello-pontine angle. A complete extracapsular removal was carried out.

Following the operation the patient recovered partial consciousness but became drowsy two hours later and died in coma on the following day.

Post-mortem.—The tumour cavity lay between the upper and lower halves of the cerebellum and indented the left side of the pons in its lower half. The tumour had been completely removed. Histological examination showed it to be a neurofibroma of rather unusual histology. Examination of the pons showed almost complete hæmorrhagic destruction of the tegmentum passing inwards as far as the mid-line against the ependymal lining and extending backwards along a line running from the central fovea of the ventricle to the outermost part of the ventral surface.

The temporal bones were removed and on superficial examination appeared normal apart from a moderate degree of enlargement of the left internal auditory meatus.

CASE II.—G. B., female, aged 47. Under the care of Dr. E. A. Carmichael.

Complaint.—Deafness in the right ear three years. Suboccipital headaches six months. Attacks of vomiting with nausea three months.

The deafness was of gradual onset. Recently it had become progressively worse and had been accompanied by "a noise like a railway engine" all over the head. For a short time before admission she had been rather unsteady with a tendency to deviate to the right. No history of giddiness, although objects sometimes tended to "sway up and down".

Neurological findings.—Alert and well orientated in space and time. No impairment of memory. Visual fields full. Mr. Williamson-Noble reports: "Slight blurring of both discs but within physiological limits." Pupils normal. External ocular movements full. Corneal reflexes: Left present, right absent. Loss of sensation to pin-prick and cotton-wool over the facial distribution of all three divisions of the right V nerve but no motor weakness. Slight weakness of the right side of the face of the lower motor neurone type, and slight fibrillation of the right side of the tongue with deviation to the right. Motor system: Upper limbs—some loss of tone and inco-ordination of the right hand and right leg. Loss of sensation to pin-prick over the second cervical segment. Reflexes: Rt. K.J. > left K.J.

X-rays (including Towne's projection): Normal skull. Ventriculography: The fourth ventricle was dilated and displaced upwards and to the left, the displacement being most marked at its lower end. The appearances were those of a space-occupying lesion in the posterior fossa on the right side.

Neurological diagnosis.—? Right VIII nerve neuroma. C.S.F.: Pressure 80 mm. clear and colourless. Cells: 1 lymphocyte per c.mm. Protein 300 mg. per 100 c.c.

Otolaryngological examination.—Tongue, palate, pharynx and larynx normal. Nose and nasopharynx normal. Tympanic membranes: Left—imperforate, marked atrophic changes. Right—imperforate, slight atrophic changes.

Cochlear function: Whisper (normal 30 ft.). Left: > 20 ft. Right: At the meatus. Weber (512 c.p.s.) referred to left. Rinne: Left positive. Right negative, with masking of the left ear. (Masking as in Case I.) Absolute bone conduction: Right slightly reduced.

Pure tone audiometry (air conduction only). Left normal. Right: There was a severe loss chiefly affecting hearing for the highest and lowest frequencies (Fig. 2A).

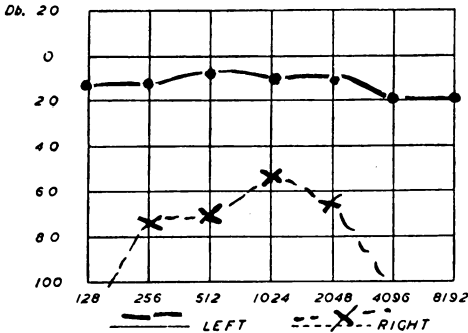


FIG. 2A.—Case II.

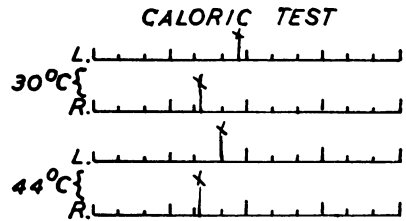


FIG. 2B.—Case II.

Vestibular function: Spontaneous nystagmus: With the head erect, first-degree vestibular nystagmus was present both to left and right, that to the right being slower than that to the left. Positional nystagmus: In the supine position, second degree positional nystagmus to the left was present with the head back and to the right. Optokinetic nystagmus: The response to the left was abolished. Caloric responses: A well-marked diminution of the right responses to both cold and hot stimulation. No directional preponderance was present (Fig. 2B).

Summary of otological findings.—**Cochlear function:** Partial deafness of the right ear. The deafness was, in some respects, characteristic of a conductive lesion.

Vestibular function: Spontaneous nystagmus, positional nystagmus and abnormality of optokinetic nystagmus. Reduction of the right caloric responses.

Otological diagnosis.—Right VIII nerve neuroma.

Operation.—The right cerebello-pontine angle was explored on 7.3.46 by Mr. Wylie McKissock. On retraction of the right cerebellar hemisphere a typical and fairly soft acoustic neuroma was exposed. The surface was coagulated and a considerable amount of tissue removed with the aid of a scoop and suction. Extracapsular removal was then completed.

The patient died on the following day. At post-mortem, hæmorrhagic changes were found in the right side of the pons with flattening of the right middle cerebellar peduncle. A small nodule of tumour remained in the right internal auditory meatus which was not enlarged. Histological examination of the tumour removed revealed a typical acoustic neurofibroma. The temporal bones were removed and on superficial examination appeared normal.

PATHOLOGICAL EXAMINATION OF TEMPORAL BONES

The temporal bones of both subjects were fixed in 10% formalin and embedded in celloidin. Serial sections were then cut in the usual way.

In the course of this procedure it is normally possible to see a great deal of the finer labyrinthine structures through the clear medium of the celloidin. With the affected temporal

bones of these two subjects, however, the labyrinthine structures were obscured by a fine "coagulum" or gel formation which was evenly distributed throughout and completely filled all the perilymph and endolymph spaces. It was much less dense than that found in labyrinthitis and much more even in its texture and distribution than the exudation which commonly occurs in this condition and indeed in any pathological condition of the labyrinth that we have previously examined. The nature of the abnormality is illustrated in Figs. 3 and 4. These are photographs of two celloidin blocks taken during the course of section cutting. Fig. 3 shows a normal temporal bone in which the normal cavities and membranes of the labyrinth can be clearly seen. Fig. 4 shows the affected temporal bone of a patient with an VIII nerve neuroma in which the naked-eye appearances closely resembled those seen in the 2 cases which form the subject of this paper. The obscuration of the membranes and labyrinth spaces is very evident.

When the sections had been cut and stained in the usual way, with hæmatoxylin and eosin, we were further interested to observe that no sign of the "coagulum" so evident during the process of section cutting could be observed. The histological features of the two cases are shown in Figs. 5 and 6. In both, the external auditory meatuses and the middle ears are normal. The internal auditory meatuses are distorted and occupied by the remains of the neuromata. In the cochleæ, higher magnifications reveal degenerative changes in the nerve fibres and cells of the spiral ganglia. In neither of the sections shown, however, is it possible to appreciate that all the labyrinthine cavities are completely occupied by the homogeneous "coagulum" to which reference has been made. The appearance of the sections was next studied without staining and with dark ground illumination. Under these conditions the presence of the "coagulum" could be readily seen. Figs. 7 and 8 are photomicrographs of unstained sections serially consecutive with those shown in Figs. 5 and 6. They are photographed at the same magnification and with dark-ground illumination. In both, the fine "coagulum" can be seen uniformly filling every part of the labyrinthine spaces. The abnormality of this appearance is illustrated by comparison with Fig. 9 which shows the appearance of a normal labyrinth prepared and photographed under identical condition.

DISCUSSION

It is well known that the labyrinthine spaces are the site of a variety of exudates and transudates, resulting not only from VIII nerve tumours but from many other pathological conditions. Nearly always, however, the distribution and texture of the resulting "coagula", if this term be permissible, are much more irregular than that revealed in the present 2 cases. In addition, it is usually demonstrable quite readily with hæmatoxylin stain. It is justifiable, therefore, to regard the present findings as quite unusual and our next task is to consider how they could be applied to the explanation of the equally unusual but clinically important finding of deafness of the so-called conductive type.

The physical problems concerned with hearing by air and bone conduction, though complex, still seem capable of explanation, in their essentials, in terms which can have changed very little since the days of Bezold. It is known, for instance, that in hearing by air conduction by far the greater proportion of the sound energy reaches the basilar membrane by way of the conducting mechanism which includes not only the tympanic membrane and ossicles but also the fluid contents of the labyrinth. In the case of hearing by bone conduction, while a proportion of the sound energy still reaches the basilar membrane by way of the conducting mechanism, other and more direct pathways through the bony cochlear walls are available.

It follows, therefore, that lesions which disturb the conducting mechanism will have their chief effect on hearing by air conduction; hearing by bone conduction will be affected to a smaller extent. In this way we have the poor hearing by air conduction and the relatively good hearing by bone conduction which are typical of conductive deafness and underlie its characteristic tuning-fork reactions, in particular the negative Rinne. The mechanical characteristics of the middle-ear mechanism are, of course, bound up with its function of overcoming, by virtue of its impedance-matching properties, the severe attenuation which normally occurs when airborne sound enters a fluid medium. The efficiency of this function, however, is necessarily dependent upon normal physical conditions of the labyrinthine fluid, and we think it likely in this connexion that the abnormal histological appearances of this fluid described in our 2 cases connote the existence during life, if not of an actual coagulum, at any rate of some considerable change of its density. The suggestion is therefore made that this density change brings about in turn a mechanical change in this, the terminal component of the conductive mechanism, which disturbs its performance as a whole in a manner which is essentially comparable to that brought about by better-known lesions of its other components, as, for example, middle-ear disease or otosclerosis. As



FIG. 3.



FIG. 4.

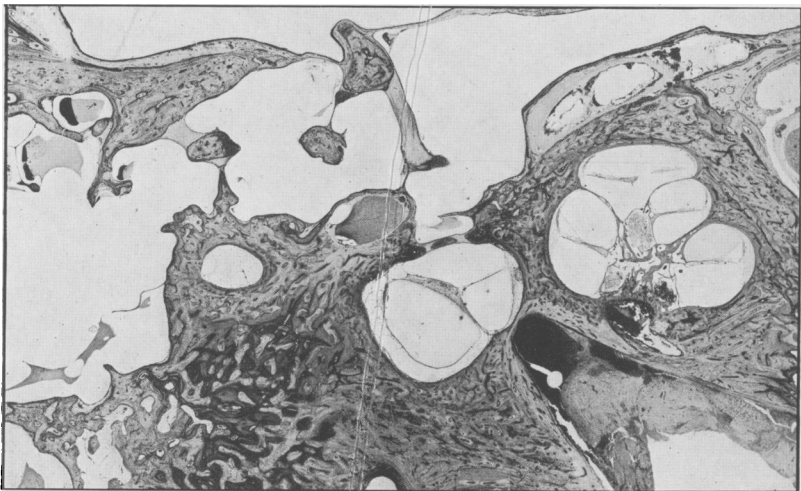


FIG. 5.



FIG. 6.

M. R. DIX and C. S. HALLPIKE: *Observations on the Pathological Mechanism of Conductive Deafness in Certain Cases of Neuroma of the VIII Nerve.*

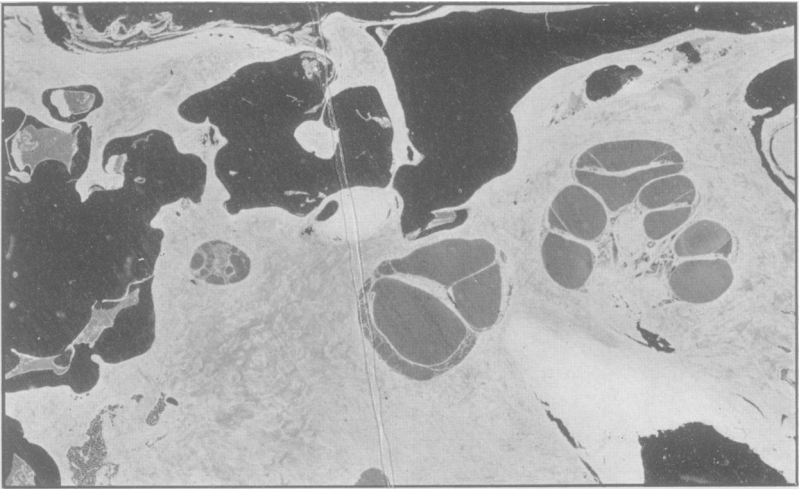


FIG. 7.

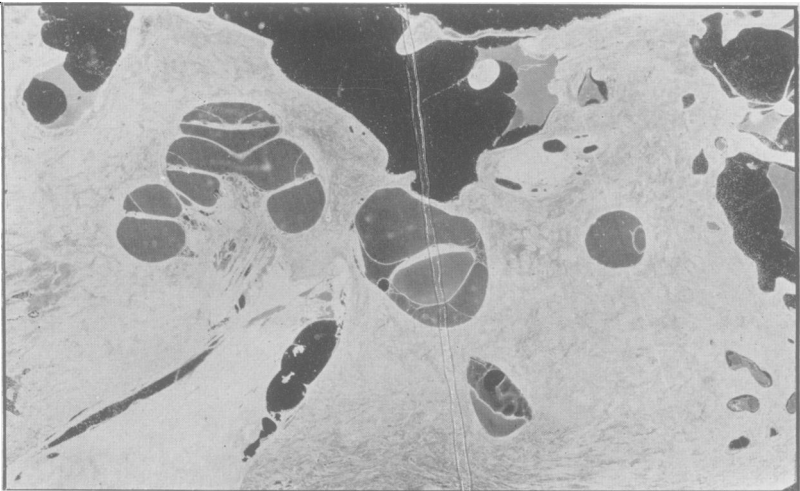


FIG. 8.

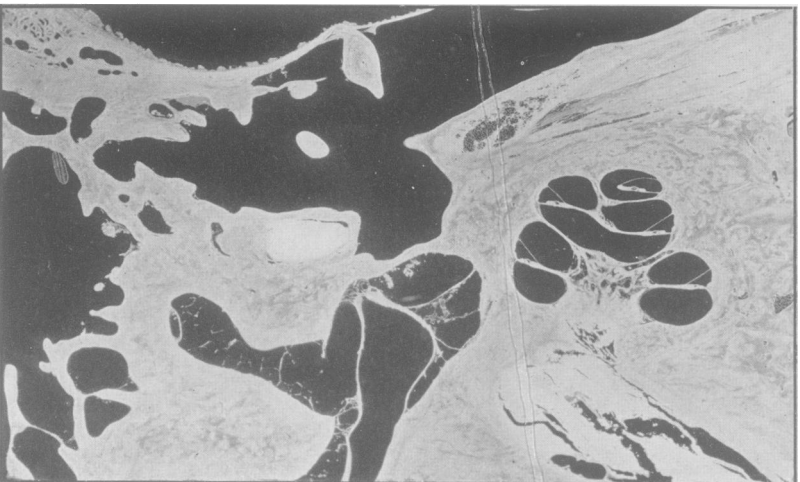


FIG. 9.

M. R. DIX and C. S. HALLPIKE: *Observations on the Pathological Mechanism of Conductive Deafness in Certain Cases of Neuroma of the VIII Nerve.*

a result, the consequent deafness is of the same conductive type. Loss of hearing by air conduction is severe, hearing by bone conduction relatively good and the Rinne test negative.

If this suggestion be acceptable, then it follows that a conductive element would also be likely to occur in the deafness which results from other forms of labyrinthine disease which are characterized by similar changes in the labyrinthine fluid. Of these, neuro-labyrinthitis, in particular that secondary to meningitis, may be taken as a likely example. In the majority of such subjects the deafness, which is due in large part to the destruction of the nervous elements, is extreme both by air and bone conduction, and no qualitative tests are therefore possible. We have, however, encountered some cases of this kind with partial deafness in whom the tuning-fork tests could be carried out with reasonable accuracy, and in a few of these we have been interested to note the presence of a true negative Rinne. A summary of the clinical features of three such cases is appended.

P. I., aged 10.

History.—Meningitis aged 3. Deafness in left ear noticed at age of 8.

Examination.—Tympanic membranes normal. Rinne (512 c.p.s.): Left negative with masking of right; right positive. Absolute bone conduction (left) much reduced with masking of right.

Audiograms.—(Tests of left ear carried out with masking of right ear, *see below*, Figs. 10A, 10B.)

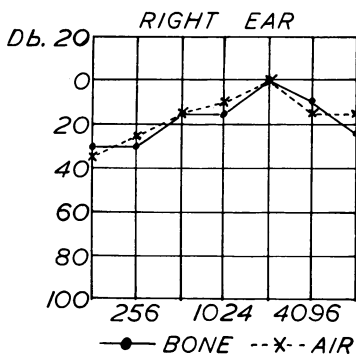


FIG. 10A.

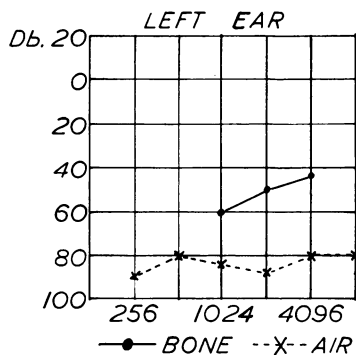


FIG. 10B.

A. P., aged 32.

History.—Deaf since meningitis aged 2.

Examination.—Tympanic membranes normal. Rinne (512 c.p.s.): Left negative; right false negative. Absolute bone conduction (left) much reduced. (*See Fig. 11.*)

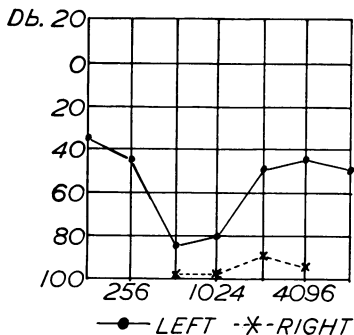


FIG. 11.—Audiogram of Case A. P. (Air conduction.)

R. T., aged 8.

History.—Maternal rubella at second month. Deafness noticed at age of 5.

Examination.—Tympanic membranes normal. Rinne (512 c.p.s.): Left negative; right negative. Absolute bone conduction grossly reduced left and right. (*See Fig. 12.*)

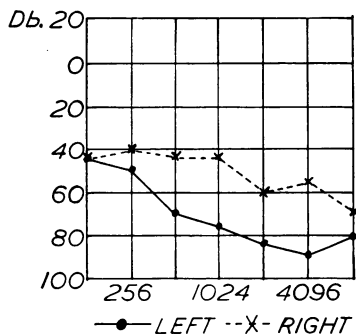


FIG. 12.—Audiogram of Case R. T. (Air conduction.)

A clinical feature of all 3 cases, and one of importance in clarifying the diagnosis, was the reduction of perception with the absolute bone conduction test, a finding to which we attach clinical importance in the diagnosis of disease of the cochlear nervous elements. In 2 of the 3 cases also (P. I. and A. P.) a gross abnormality of the caloric responses was also present. In the third case (R. T.) the caloric tests were not carried out. It seems, therefore

necessary to stress the clinical value in these difficult cases, both of the absolute bone conduction and the caloric tests, since they make so plain that co-existent with the conductive element of the deafness a perceptive element is also present.

In conclusion, it may be said that although the conception of the labyrinthine fluid as constituting an element of the conductive mechanism is not a new one, this would appear to be the first occasion on which it has been possible to give it the support of direct anatomical and clinical observations.

Mr. F. C. Ormerod recalled a discussion on acoustic nerve tumours by this Section in 1923 when the negative results of the Rinne test were mentioned but not discussed at any length. In 1929, T. H. Just definitely described the true negative Rinne and in 1934 Albert Gray described the histology of the temporal bones of two cases of bilateral acoustic nerve tumours (*Proc. R. Soc. Med.*, 27, 1179). Both patients, one under the care of the late Professor de Kleijn and the other under the care of the speaker, showed a negative Rinne test. In both cases Gray demonstrated otosclerotic changes in the neighbourhood of the oval window and he felt that this explained the negative Rinne. He felt also that the occurrence of otosclerotic changes following acoustic tumours confirmed his theory as to the vasomotor origin of otosclerosis. The findings were illustrated in the second volume of Gray's Atlas of Otolaryngology.

Mr. T. A. Clarke asked Mr. Hallpike how the coagulum found in cases of VIII nerve tumour with apparent negative Rinne differed from that found in similar cases with the usual finding of a positive Rinne. It was agreed that perfect masking of the good ear had still to be achieved in testing bone conduction on the diseased side. Could any failure in masking account for the apparent negative response described in the Rinne test in these cases?

Mr. I. Simson Hall said that Mr. Hallpike's findings had interested him enormously. On one occasion during the fenestration operation, in a case of fairly extreme deafness, he opened the labyrinth and found a condition he could not understand. The bone surrounding the canal was rather soft and the outlines were blurred completely. He supposed that the patient must have had a labyrinthitis, but it now occurred to him that Mr. Hallpike's explanation might have been the correct one. The operation was a complete failure.

Mr. Philip Scott said that he was stimulated, on getting notice of this Paper, to refer to some of his notes, and now he was stimulated again to go back and look at his sections. In the sections at which he had had an opportunity of looking he was dissatisfied in one particular case with the negative Rinne. That case did have a noticeable coagulum throughout the section. He now intended to re-examine the section in the light of the observations of Mr. Hallpike and of some others.

Mr. Hallpike, in reply, said that when they began to encounter these cases with evidence of conductive deafness they used to think of the likelihood of an associated otosclerosis, but as they had the opportunity of examining further material they never found otosclerosis, and in the cases shown that day the stapes footplates and middle ears were normal.

Mr. Clarke had asked how this particular coagulum compared with the coagulum which occurred not uncommonly in other conditions of the labyrinth. Generally speaking, such coagulum was much more scattered and less homogeneous than that found in the 2 cases under discussion. As to guaranteeing the efficiency of masking, this it was difficult to do.

He had been very much interested in what Mr. Simson Hall had told them, because in some cases of deafness in children of 5 to 10 years of age the diagnosis of juvenile otosclerosis was sometimes made with some confidence on the basis chiefly of a negative Rinne test. It seemed to him that had caloric and absolute bone conduction tests been done in Mr. Simson Hall's case a different view might have been taken of the diagnosis.