MÉNIÈRE'S DISEASE*

By C. S. HALLPIKE, F.R.C.P., F.R.C.S.

*Aural Physician and Director, Otological Research Unit, Medical Research Council, National Hospital,
Queen Square, London*

The story of scientific enquiry into the problems of human vertigo began, as we know, one hundred years ago with the work of a citizen of Paris, Prosper Ménière. It is, therefore, fitting that I should begin with a few remarks on the history of his work and its remarkably true relationship to modern views.

Ménière's first great achievement was his recognition that paroxysmal vertigo could be caused by a labyrinthine lesion. This may seem a commonplace notion now, but it is necessary to bear in mind that in 1861, when Ménière published his best known paper, the very possibility that a lesion of the internal ear could cause such severe symptoms as vertigo and vomiting was still a moot point. Flourens' experimental work on pigeons, in which he demonstrated for the first time that gross disorders of equilibrium could be produced by injury of the semi-circular canals, had only been published some 30 years earlier, and its significance as regards the problems of human disease had not yet been appreciated. It was Ménière's great merit that he knew of Flourens' work, understood its meaning and applied it to human problems.

Ménière's second great achievement was made possible by his remarkable powers of describing and analysing the symptoms and signs of disease. These enabled him to identify, with an accuracy which has never been seriously questioned, the particular disease of the labyrinth which has since come to bear his name. Of this the essential feature, as we now know, is a distension without evidence of infection or trauma of the endolymphatic vesicle, a finding remarkable in itself and since reproduced with remarkable uniformity in a considerable number of further histological studies of temporal bones carried out, some by myself and also by a number of others.

The present position is, therefore, this: That we know the symptomatology and natural history of Ménière's disease. For this information we need not go far beyond Ménière's own writing.

We know, also, the morbid anatomy of the labyrinth which is associated with the disease, though to be sure the exact mechanism of the endolymphatic distension still escapes us.

But we have also at our disposal what Ménière had not, namely, a good deal of new knowledge derived from special tests of cochlear and vestibular function, which enables us to identify the disease and to assess its severity with greater accuracy than Ménière could command.

I propose to begin my remarks with a brief outline, in essentials a recapitulation of Ménière's own description, of the symptoms and natural history of his disease.

These I shall then endeavour to correlate with the morbid anatomical findings in the labyrinth which have been fully established by recent pathological studies. Thereafter I shall discuss in some detail the results of certain of the tests of cochlear and vestibular function and will again correlate these with the morbid anatomical findings.

Finally, I shall discuss certain problems of differential diagnosis.

We will begin with the clinical features of Ménière's disease. Its onset is usually in the middle period of life, 40 to 60 years, and the sexes are equally affected. Its course is essentially benign in so far as it does not directly shorten the life span and it exhibits no significant association with any other form of organic disease, including allergy, outside the eighth nerve system.

Partial deafness of one ear, accompanied by tinnitus, provides the usual clinical background of established Ménière's disease, upon which are superimposed its better known dramatic features, paroxysmal attacks of vertigo nearly always with nausea, vomiting and prostration. In a minority of cases the vertigo comes on very rapidly, within a matter of seconds, and without any warning. More often, however, the vertigo is of slower onset or may be heralded by some change in the character or intensity of the tinnitus.

The duration of the attacks seldom exceeds five or six hours. They generally end with a state of extreme prostration and are typically followed by

*Lecture given at Cours International de Labyrinthologie Clinique, Paris, January 15-21, 1955.

a long, deep sleep from which the patient awakens without the vertigo, which troubles him no more until the next attack.

In other patients, however, the severe attacks may be interspersed with longer periods of minor unsteadiness which are noticeably aggravated by head movements. In such patients the cochlear symptoms, deafness and tinnitus, are usually overshadowed by the vertigo and occupy a secondary place in the patient's impressions.

One other cochlear symptom must not be forgotten: intolerance of loud sounds with distortion of voice sounds and music. This is common and highly characteristic of Ménière's disease.

We come next to the highly characteristic

FIG. 1.

morbid anatomical changes in the labyrinth which were first described by Cairns and myself in 1938.

In Fig. 1 is shown a transverse section of the unaffected temporal bone of a typical case of Ménière's disease. In the vestibular area can be seen the macula of the saccule with the thin saccular wall. Outside this, and underlying the stapes footplate, is the large perilymph space of

FIG. 2.

the vestibule. In the cochlea are seen the two perilymph scalae and between them the scala media filled with endolymph and containing the sensory cells of Corti's organ. It is separated from the scala vestibuli by the thin membrane of Reissner.

In Fig. 2 is shown, for comparison, a section of the opposite affected temporal bone. In the vestibule the saccule is distended, the membrane being pushed back everywhere upon the bony vestibular wall with obliteration of the perilymph space which lies deep to the stapes footplate. In the cochlea the outstanding abnormality is the apparent disappearance of Reissner's membrane. In fact this appearance is brought about by a gross

FIG. 3.

distension of the scala media with displacement of the membrane on to the bony wall of the scala vestibuli.

In Fig. 3 is shown the affected cochlea at a higher magnification. No abnormality can be seen in the fibres of the cochlear nerve as they lie in the spiral osseous lamina, while the cells of the spiral ganglion are also normal in number and structure.

We come finally to the cells of Corti's organ. These are notoriously difficult to display in histological preparations of the human temporal bone. Nevertheless, it is possible even at this magnification to detect certain obvious pathological changes, and these are better shown in Fig. 4. Above we see Corti's organ of the affected ear with its hair cells and Corti's rods enclosing a well-formed tunnel. Below, on the affected side, the cell mass of Corti's organ is greatly compressed and its outline irregular, while Corti's tunnel is occupied by a structureless coagulum.

Changes closely resembling these have been found both by myself and by others in the temporal bones of a considerable number of subjects

FIG. 4.

with Ménière's disease, and it is now safe to describe them as constituting its essential morbid anatomy.

As to the cause of the endolymphatic distension, little even now can be said. Some evidence, mainly histological, has been advanced for a primary failure of the absorptive mechanism of the endolymphatic sac. The evidence, however, is far from strong and the alternative possibility of some abnormality, quantitative or qualitative, of the endolymph itself as secreted by the stria vascularis, remains to be investigated.

Although in this sense the morbid anatomical findings give us no full explanation of the disease, they do enable us to say this; that the essential events which underlie the characteristic paroxysms consist of recurrent bouts of endolymphatic distension. As a result of the accompanying physicochemical changes in the endolymph the sensory cells of hearing and equilibrium undergo acute structural changes with resulting derangement of hearing and equilibrium.

These changes are incompletely reversible between the attacks, and this we expect to find

reflected in the results of functional tests applied during these periods. We can, in fact, return to the subject of these tests enlivened by the assurance that we can interpret their results in terms of known structural changes in particular anatomical elements, namely the sensory end-organs of hearing and equilibrium.

We come now to the tests themselves. Of the vestibular tests we have come to rely chiefly upon the caloric tests practised according to a technique first described in 1942 in association with my colleague, the late Dr. Gerald Fitzgerald. The subject of these tests is still, we find, looked upon as a matter of some difficulty, and I propose, therefore, to begin with a brief description of the way in which we perform them and interpret their results.

FIG. 5.

The tests are carried out with the patient lying supine, as shown in Fig. 5, the head being raised 30 degrees from the horizontal, thus bringing the external canal into its position of maximum sensitivity. The cold and hot stimuli, water at 30° and 44° C., are equidistant from body temperature, and are applied for 40 seconds. Their effects are substantially limited to the external canal and are calculated to bring about equal and opposite deflections of the cupula.

The subject is asked to fix his gaze upon a small object in front of him, usually a mark upon the ceiling. The observations of the resulting nystagmus are, therefore, of second degree nystagmus. The subject's eyes are well illuminated, and the observer inspects them from a distance of about 30 cm. The observational conditions are thus very good.

What we measure is the time interval between

the application of the stimulus and the end of the response. In this, the latent period of the response is included, and we make no attempt to measure it separately. The results are graphically recorded in the manner shown in Fig. 6.

The results shown are characteristic of an average normal subject. The two continuous lines represent time, three minutes sub-divided into intervals of ten seconds. The interrupted lines represent the response durations. Above are the two cold responses for the left and right ears, and below the two hot responses. It is convenient for reference to number these responses 1 to 4 from above downwards. Responses 1 and 4 consist of nystagmus to the right. Responses 2 and 3 consist of nystagmus to the left. The direction of the nystagmus is specified in accordance with convention in terms of its rapid component.

The commonest and most obvious type of abnormality is shown in Fig. 7. We have termed it a canal paresis. One or other of the two external canals exhibits a loss of sensitivity. Above is seen a left canal paresis. Responses 1 and 3 are reduced with respect to responses 2 and 4 which are normal. Below, a right canal paresis. Responses 2 and 4 are reduced.

The degree of paresis varies. The responses, as shown, may be moderately reduced. It may be severely reduced or totally extinguished. The

question which now arises is: Is it really necessary to use both cold and hot stimuli? Does not the information derived from the hot responses merely reduplicate that derived from the cold responses?

This would certainly be the case if the only abnormality which occurred were a loss of sensitivity of one or other external canal. In point of fact it is not the only abnormality, and this brings us to the interesting and important phenomenon for which, since 1942, we have used the term 'directional preponderance.' The nature of this is shown in Fig. 8.

Above is shown the normal caloric pattern. Below is a typical directional preponderance to the left. Reactions 1 and 4, which consist of nystagmus to the right, are inhibited. Reactions 2 and 3, which consist of nystagmus to the left, are increased.

Below is seen an example of directional preponderance to the right. In this, reactions 2 and 3, which consist of nystagmus to the left, are inhibited. It is easy to understand that this type of abnormality could not possibly be demonstrated by means of cold stimuli alone. Thus in this case responses 1 and 2 would indicate a loss of sensitivity of the *right* labyrinth. Responses 3 and 4, however, would indicate a loss of sensitivity of the *left* labyrinth.

It is only when all four responses are known that the directional nature of the derangement is made apparent.

It is necessary at this point to ask and answer a practical question. What is the clinical importance of directional preponderance which makes it worth while demonstrating? The answer is found in the fact, established by observation, that in a significant proportion of cases of unilateral vestibular

disease, affecting either the labyrinth or the eighth nerve, it occurs as the sole abnormality of the caloric responses. Thus we have found it in some 20 per cent. of our series, extending to several hundreds of cases of unilateral Ménière's disease. The direction of the preponderance is always to the side opposite to the lesion; its presence can be accepted as evidence of that lesion, and this is the fact which gives it its clinical importance. What now of its cause? This will best be understood if we consider the neurological mechanisms to which it is usual to attribute certain well-known disturbances which follow experimental destruction of one labyrinth. These it is customary to explain in the following way:

Each labyrinth, in addition to responding in a phasic manner to angular accelerations by means of its semicircular canals, is also the source of tonic impulses which exercise an asymmetrical influence upon the limbs and eyes. Thus the right labyrinth alone causes a rotation of the head to the right, together with a sustained nystagmus with its rapid component to the right; effects which in the intact animal are cancelled by the opposing influence of the left labyrinth.

If now the right labyrinth be destroyed, then the unopposed tonic impulses from the left labyrinth will cause rotation of the head to the left with nystagmus to the left. As we know, this nystagmus, severe at first, gradually disappears in the course of a few weeks, its abolition being brought about by a process of central compensation. Thus the right vestibular nuclei, being deprived of incoming tonic impulses, set up a rhythm of their own and in this way the output of the two sets of vestibular nuclei are once more brought into balance and the nystagmus ceases.

Let us now take the case in which the right labyrinth, instead of being completely destroyed, is, as in Ménière's disease, the seat of a partial lesion which has brought about no more than a partial loss of its tonic discharge. In this case we assume that the resulting minor imbalance of the tonic discharges would be insufficient to cause any spontaneous nystagmus to the left. Nevertheless, its tendency to occur will still be present and will manifest itself as a directional preponderance to the left of the caloric responses.

Thus we see that in the absence of spontaneous nystagmus, and in the absence also of any alterations in sensitivity of the external semi-circular canal, a unilateral loss of labyrinth tonus may still be made evident by means of the cold and hot caloric tests.

Thus in Fig. 8 the upper pattern, preponderance to the left, is indicative of a tonus defect of the right labyrinth. The lower, preponderance to the

right, is indicative of a tonus defect of the left labyrinth.

Whether or not this is the true explanation of the directional preponderance of the caloric responses which we have encountered in unilateral labyrinthine lesions, such as occur in Ménière's disease, cannot yet be stated with certainty. We can only say that the circumstantial evidence in its favour is very strong.

What we can state as matters of fact are: That it does occur in association with such lesions; that it can be accepted as evidence of their presence; that its demonstration requires both cold and hot caloric stimuli and that for this reason their use is essential.

To complete this exposition of the caloric test abnormalities encountered in Ménière's disease, I must explain that while these two primary abnormalities, canal paresis and directional preponderance, usually occur separately, they are not infrequently combined. The resulting abnormalities in the patterns of the caloric responses, though they may be very confusing, are nevertheless readily recognizable as algebraic summations of the primary abnormalities.

The results of the caloric tests obtained in a series of 50 cases of Ménière's disease were published in 1942 by my colleague, Mr. Cawthorne, and myself, and the figures therein given continue to accord very well with our subsequent findings which cover several hundred subjects and are shown in Table 1.

We may now turn to the tests of cochlear function; in particular to two which are especially deserving of our attention. These are the intelligibility test for amplified speech and the loudness recruitment test.

Otologists have been aware for many years that the capacity of deaf patients to derive benefit from hearing aids depends very much upon the pathological basis of their deafness. Knowledge of this subject has been greatly advanced by the numerous and searching enquiries into the problems of hearing aid design which have been carried out in the last ten years by investigators upon both sides of the Atlantic, in particular the Harvard Group in the U.S.A. and the Electro-Acoustics Committee of the Medical Research Council in Britain. In the course of this work great advances have been made in the principles and practice of speech audiometry and as a result this valuable test procedure has now been put upon an accurate quantitative basis.

By its means otologists are now able to answer with rapidity and precision one of the most important practical questions that can arise in connection with any deaf subject. To what extent can his social disability, loss of speech intelligi-

CALORIC TEST RESULTS IN 100 CASES OF MÉNIÈRE'S DISEASE.

DIRECTIONAL PREPONDERANCE	21 CASES
CANAL PARESIS	49 CASES
DIRECTIONAL PREPONDERANCE + CANAL PARESIS	18 CASES
NORMAL	12 CASES

TABLE I.

bility, be corrected by means of amplification. This criterion, applied by means of speech audiometry, makes it possible to divide deaf subjects into two groups.

In the first group the deafness is generally due to conductive lesions and restoration of speech intelligibility by amplification can be satisfactorily accomplished. It would seem that in these subjects, if the sounds of speech can be made to reach the cochlea at all, then they are well understood.

In the second group the deafness is of the so-called perceptive type and restoration of intelligibility by amplification cannot be achieved. The reactions to amplification of four subjects belonging to these two groups is shown in Fig. 9.

FIG. 9.

The tests are carried out with the patient seated in an anechoic soundproof room with the ear under test at a fixed distance from a loudspeaker. Recorded lists of monosyllabic words are reproduced on the speaker by means of an amplifier system.

The amplification can be adjusted over a range of 100 db. and this intensity scale is set along the base line. The percentage of words understood

are recorded upon the ordinates. The amplifier setting of 50 db. gives an intensity at the ear of the subject which corresponds to that of an ordinary conversational voice at a distance of one metre. Other useful amplifier settings specified on the chart are at 64 db., which corresponds roughly to a loud conversational voice at 3 ft., and at 36 db. This we call 'church level' since it corresponds roughly to the speech intensity levels which reach the ears of listeners in churches and other auditoria. The response curves of a normal subject are shown on the left. At an amplification of 30 db., well below church level, the score would be 90 per cent., which corresponds to a sentence intelligibility of 100 per cent. Curves A and B were obtained from two subjects with conductive deafness.

Subject 'A' at church level can only understand an occasional word. With increasing amplification, however, his score steadily increases and reaches 90 per cent. level when the intensity is raised to the level of ordinary conversation at 3 ft. With this degree of deafness the patient would, of course, experience no difficulty with a tête-à-tête conversation. 'B's deafness is more severe. He understands a few words at 50 db. Nevertheless he reaches the score of 90 per cent. when amplification is raised to 80 db. We see that in both these cases it is possible by means of amplification to restore full intelligibility of speech.

In Fig. 9 are also shown for contrast the very different results that are obtained in the type of perceptive deafness which we encountered in Ménière's disease. Both curves C and D begin at points which correspond closely to those of the

two cases of conductive deafness. As the amplification is increased, however, intelligibility, after an initial slight increase, falls off again and at no point is any serviceable restoration of intelligibility achieved. These results are highly characteristic of Ménière's disease, at any rate in its active phases.

We come now to the Loudness Recruitment Test. Most of our work has been carried out with patients suffering from unilateral Ménière's disease and in these we have found it best to use the so-called 'alternate binaural loudness balancing procedure' with which Fowler first demonstrated the phenomenon.

FIG. 10.

The way the test is carried out is shown in Fig. 10. The subject wears a telephone receiver on each ear, each supplied by a separate pure tone generator. The frequency of the stimulus is the same for each ear but the intensity can be varied independently. With the switch the stimulus is applied alternatively to each ear, the intensity being adjusted to give equality of the loudness sensations in the two ears. The two intensity scales are marked in decibels above the normal threshold, one for the right ear and one for the left. The test procedure is designed to identify and connect, as shown, points on the two scales which give equal sensations of loudness. The ladder diagram was derived from a normal subject

and the rungs of the ladder, as would be expected, run horizontally from top to bottom.

On the left is shown the result obtained with a case of unilateral deafness due to middle ear disease. The hearing loss at threshold is 30 db. at 1,000 c.p.s. This is shown by the lowest rung of the ladder. The intensity is then increased by some 20 db. at the normal ear and the matching intensity at the affected ear is found to lie at 50 db. In the same way the sensitivity difference between the two ears, 30 db. at threshold, is maintained all the way up the intensity scale.

On the right, by contrast, is shown the result obtained with a case of unilateral deafness due to Ménière's disease. The hearing loss at threshold is also 30 db. at 1,000 cycles, and this too is shown by the lowest rung of the ladder. As the intensity scale is mounted, however, the difference in sensitivity between the two ears, 30 db. at threshold, diminishes until at 60 db., equal intensities give equal loudness sensation, and this in brief constitutes the Loudness Recruitment phenomenon.

In describing the technical features of this test in such detail I have had a special point in mind. As has been clearly shown by my colleagues, Drs. Dix and Hood, and myself, the Loudness Recruitment phenomenon is characteristic of hair cell disease. It depends, in fact, upon the capacity of diseased hair cells to give responses of normal magnitude to stimuli of high intensity. They cannot, however, sustain such responses and, indeed, if the stimuli are at all prolonged the responses rapidly relapse to sub-normal level. Hence we insist upon using a test procedure which, as in the one that I have described, short interrupted stimuli are used.

In all that I have so far told you of Ménière's disease, I have endeavoured to include none but well attested facts. The information, although not complete, is impressive both in character and amount. It enables us to say of the disease that it has a highly characteristic symptomatology and natural history, matched by an equally characteristic morbid anatomy. Further, the tests of vestibular and cochlear function all give results in a uniform and, we should like to think, distinctive character.

Now, finally, comes the practical question of diagnosis. To what extent do these clear-cut clinical features enable us, in practice, to distinguish it from other organic disorders of the eighth nerve system? What, in fact, are these other disorders and in what way do their clinical features and pathology differ from those of Ménière's disease?

The subject is a large one and was dealt with in some detail by Dr. Dix and myself in a paper communicated in March 1952 to the Otological

Section of the Royal Society of Medicine of London. In the time at my disposal it will be necessary to confine myself to a brief review of the salient points of that paper.

We stated that by far the greater part of the patients seen by us at Queen Square, with symptoms indicative of organic derangement of the eighth nerve system, could be divided into four distinct clinico-pathological groups:

1. Ménière's disease.
2. Vestibular neuronitis.
3. Positional vertigo of the benign paroxysmal type.
4. Tumours of the cerebello-pontine angle including neurofibromata of the eighth nerve.

1. On the subject of Ménière's disease I need say no more.

2. Vestibular neuronitis is chiefly distinguishable on clinical grounds from Ménière's disease by the conspicuous absence of cochlear signs and symptoms. It affects adults without preference for sex and is relatively more common than Ménière's disease under the age of 30.

Apart from the absence of cochlear signs and symptoms the condition is sometimes, though not always, distinguishable from Ménière's disease by the character of the vertigo. This may consist of sudden and transient seizures accompanied by sensations of blackout. On the other hand there may be no severe paroxysms and the disequilibrium may take the form of feeling 'top heavy' or off balance when walking.

In a fairly high proportion of the subjects the onset of the symptoms is associated with some kind of febrile illness or with some evidence of infection in the nose or throat. Infective ear disease, as in Ménière's disease, does not seem to be a factor.

As stated, tinnitus or deafness are conspicuous by their absence, and the most searching tests of cochlear function yield normal results. Tests of vestibular function, however, tell a different story. In particular the caloric tests constantly show severe and often bilateral derangement, while the galvanic responses, too, are often much reduced. The condition is essentially a benign one and the symptoms usually respond well to treatment of infective foci when these are present. Under these circumstances a restoration of the caloric responses has also been observed. The evidence of vestibular disease, taken in conjunction with the absence of cochlear symptoms or signs, makes it necessary to locate the lesion in the vestibular pathways at some point central to the labyrinth. It was, however, impossible to go further and specify the particular elements of the neurones, nerve cells or nerve fibres, which were involved. When, therefore, it came to naming the condition, we required a term comprehensive enough to encompass this ambiguity. We

chose the name 'vestibular neuronitis' and have since continued to use it.

3. *Positional nystagmus of the benign paroxysmal type.* Since Bárány's original paper published in 1921, otologists and neurologists alike have shared a growing interest in a group of patients in whom nystagmus and vertigo are precipitated by certain critical orientation of the head. As Nylén has shown in his well-known monograph, a particular variety of this condition tends to occur in association with space-occupying lesions in the posterior fossa. In addition, however, positional nystagmus may occur in patients without any evidence whatsoever of any serious intracranial disease and an increasing body of evidence has accumulated to support Bárány's original thesis that in these subjects the lesion is situated in the otolith organs themselves.

In our paper in 1952, Dr. Dix and I analysed our findings in 100 such subjects and came to conclusions which were in striking agreement with those of Bárány. Our findings and conclusions can be stated quite briefly. The condition chiefly affects adults of the age group 30 to 60 years without preference for sex. In none was there any evidence of disease of the central nervous system outside the eighth nerve system.

FIG. 11.

The method of eliciting the nystagmus, and also its highly characteristic features, are depicted in Fig. 11. In this the critical head position is also indicated with the head turned either to the left or, as shown, to the right. In taking up this position the patient is first seated upon the couch with the head turned to one side and the gaze fixed upon the examiner's forehead. The examiner then grasps the patient's head firmly between his hands and briskly pushes the patient back into the critical position. The reaction which results calls for some detailed description.

First of all there is nearly always a marked latent period. Sometimes this is as long as five or six seconds. Occasionally it is very short and indeed the reaction may seem to come on at once. This, however, is uncommon. The onset of the nystagmus is nearly always preceded by an appearance of

distress. The colour may change; the patients may close their eyes, cry out in alarm and make active efforts to sit up again. At this point it is necessary to reassure the patient and maintain the position of the head. The nystagmus is chiefly rotatory, the direction of the rotation being towards the undermost ear. (Note—In specifying the direction of the rotation, reference is made to the displacement of the 12 o'clock point of the corneal circumference.) In addition to the rotatory element there is generally a horizontal component which is again directed towards the undermost ear. The nystagmus increases in a rapid crescendo in a period which may be as short as two to three seconds or as long as ten seconds. Thereafter it rapidly declines and the patient's distress is relieved. If the patient is then allowed to sit up, a recurrence of the vertigo in a slighter form is generally noted, and if the eyes are examined at this point nystagmus can be seen, the direction of which is, on the whole, reversed. If this is allowed to disappear and the critical supine position is again assumed, the nystagmus again makes its appearance, but generally in slighter form and disappears more rapidly than before. After two or three repetitions of this test it is generally found that the reaction has been eliminated altogether and cannot be elicited except, as Bárány pointed out, after a period of rest.

Now the evidence put forward by Miss Dix and myself led us to conclude that the symptoms were due to an irritative lesion of the otolith apparatus of the labyrinth towards which, when undermost, the nystagmus was directed. Thus in Fig. 11 the subject's right labyrinth would be the affected one. This conclusion was derived from the following chain of evidence:

1. The benign course of the disease and the absence of evidence of any involvement of the central nervous system indicated a labyrinthine lesion due to infection, trauma or some vascular lesion.

2. A labyrinthine lesion was also indicated by the fact that in 55 of our 100 cases substantial evidence of ear disease was present, usually in the form of gross middle ear infection or labyrinthine trauma.

3. In many of these 55 cases with ear disease this disease was unilateral and in practically all of these latter the nystagmus was directed towards the affected ear when this was placed undermost.

4. In one typical case we were able to carry out a histological examination of the affected labyrinth and this revealed chronic changes in the otolith apparatus.

The case was that of a woman of 40 who exhibited before death a characteristic positional nystagmus of the benign paroxysmal type with the right ear undermost. Her death was due to an unrelated disorder. Histological examination of the

FIG. 12.

left temporal bone showed a normal vestibular apparatus. In the right temporal bone, however, gross changes were found in the maculae of the otoliths.

In Fig. 12 is shown the appearance of the normal healthy human utricular macula. The layer of sensory cells is seen evenly arranged with the superimposed otolith membrane. Beneath the layer of sensory cells lies a loose connective tissue meshwork in which run the fibres of the utricular nerve.

FIG. 13.

In Fig. 13 is shown a view at higher magnification of the sensory epithelium and the underlying connective tissue meshwork.

In Figs. 14 and 15 is shown the macula of the utricle in our case of positional nystagmus. It can be seen that in addition to the absence of the otolith membrane and the disorganization of the sensory cells, there is also present a considerable thickening of the sub-epithelial connective tissue network with the presence here and there of irregular cellular infiltrations. At one point there occurs a number of irregular spaces occupied by fluid or cell remnants. The general picture is one

FIG. 14.

FIG. 15.

of chronic tissue changes resulting either from infection or trauma, and it accords very well with our conception of the responsible lesion which we have reconstructed from our clinical evidence. Changes very similar in character but lesser in degree were present in the macula of the saccule.

4. *Tumours of the cerebello-pontine angle.* The best known and commonest of these tumours are the acoustic neurofibromata. From what Crowe and Hardy have told us of their early development they seem to have their starting point in the region of Scarpa's ganglion of the vestibular nerve as it lies in the internal auditory canal.

The course of such a tumour falls typically into two distinct stages, each with its own clinical features.

In the first or otological stage there is a slow and progressive destruction of both cochlear and vestibular nerve fibres. Clinically, the condition is extremely insidious. Thus, the deafness being unilateral, causes little social disability and is not as a rule accompanied by severe tinnitus. For these reasons it seldom attracts serious attention. As to

the vestibular destruction its progress, being slow and steady, causes no paroxysmal disturbance, and with it keep pace the processes of central compensation. As a result, the tumour may progress to complete unilateral destruction of vestibular function without causing any vestibular symptoms apart from slight unsteadiness and momentary vertigo on walking.

In the second, or neurological stage, the tumour expands from the internal auditory meatus and begins to press at first upon the brain stem in the region of the vestibular nuclei, thereby causing trigeminal disturbances and spontaneous nystagmus to the opposite side. Later interference with the cerebellum develops with cerebellar ataxia of the homolateral limbs. It is with regret that we have to admit that in most cases of eighth nerve tumour the diagnosis is not made until they have passed from the otological stage in which their removal is relatively easy, into the neurological stage when the brain stem changes add greatly to the hazards of the operation. The morbid anatomical changes in the labyrinth are well known. The vestibular and cochlear neurones, including the spiral ganglion, degenerate completely, usually with preservation of the cells of Corti's organ. Sometimes, however, the tumour interferes with the blood supply of the labyrinth and changes in the hair cells then do occur. Distension of the Scala media of the kind found in Ménière's disease is not found.

FIG. 16.

In Fig. 16 is shown a transverse section of the temporal bone from an early case of eighth nerve neurofibroma. The tumour has been removed. There is a characteristic funnel enlargement of the internal auditory meatus. Remains of the tumour are still present in the fundus of the meatus at this point. Extensive degeneration of the cochlear neurones, including the cells of the spiral ganglion, have taken place within the labyrinth. The hair

	CHARACTER OF VERTIGO.	COURSE OF VERTIGO	RELATIONSHIP OF VERTIGO TO HEAD POSITION	CALORIC RESPONSES.	DEAFNESS.	TINNITUS.	CHARACTER OF DEAFNESS.	RESPONSE TO AMPLIFICATION.	LOUDNESS RECRUITMENT.	PATHOLOGY.
MENIERE'S DISEASE	PAROXYSMAL. WITH NAUSEA AND VOMITING	ATTACKS OFTEN SEVERE AT ONSET. LONG PERIODS OF FREEDOM COMMON	ABSENT	ABNORMAL RESPONSES OCCUR IN 95%	ALWAYS PRESENT	ALWAYS PRESENT. OFTEN SEVERE	VARIABLE. SOUNDS AND VOICES DISTORTED	POOR	ALWAYS PRESENT. OVER-RECRUITMENT MAY OCCUR	DISTENSION OF ENDOLYMPH SYSTEM. DEGENERATIVE CHANGES PRESENT IN END ORGANS. NERVE FIBRES & GANGLION CELLS NOT AFFECTED
TUMOURS OF CEREBELLO-PONTINE ANGLE	PAROXYSMS RARE. PERSISTENT IMBALANCE USUAL	SLIGHT IN EARLY STAGES. THEREAFTER PROGRESSIVE	RARE	ABNORMAL RESPONSES OCCUR IN 100%	ALWAYS PRESENT	ALWAYS PRESENT. NOT OFTEN SEVERE	PROGRESSIVE	MAY BE GOOD	USUALLY ABSENT OR INCOMPLETE	DEGENERATION OF FIBRES OF VIIIth NERVE & ASSOCIATED GANGLION CELLS. DEGENERATION OF END ORGANS NOT USUAL BUT MAY RESULT FROM DERANGEMENT OF VASCULAR SUPPLY
VESTIBULAR NEURONITIS	BLACK OUTS OR DROP SEIZURES. SOMETIMES PERSISTENT IMBALANCE	ATTACKS FREQUENT DURING ACTIVE STAGES. RECOVERY OR LONG REMISSIONS USUAL	ABSENT	ABNORMAL RESPONSES OCCUR IN 100%. OFTEN BILATERAL	COCHLEAR SYMPTOMS AND SIGNS ABSENT				UNKNOWN. ? TOXIC DEGENERATION OF VESTIBULAR NEURONES	
POSITIONAL NYSTAGMUS OF BENIGN PAROXYSMAL TYPE	PAROXYSMAL	ATTACKS FREQUENT DURING ACTIVE STAGES. RECOVERY OR LONG REMISSIONS USUAL	ALWAYS PRESENT	OFTEN NORMAL	COCHLEAR SYMPTOMS AND SIGNS PRESENT ONLY WHEN CONDITION IS ASSOCIATED WITH EAR DISEASE. USUALLY MIDDLE EAR INFECTION				DEGENERATION OF MACULAE OF OTOLITH ORGANS. CHARACTER OF TISSUE CHANGES SUGGESTIVE OF TRAUMA, INFECTION OR VASCULAR UPSET.	

TABLE 2.

cells, however, appear to be unaffected, while no distension of the Scala media can be seen.

The otological symptoms and signs are usually quite characteristic. Paroxysmal attacks of vertigo may occur but are rare. The deafness may be of any degree. It arises chiefly from destruction of the nerve fibres and not, as in Ménière's disease, from hair cell disease. Hence Loudness Recruitment, so typical of Ménière's disease, is characteristically absent in eighth nerve tumours, a diagnostic point of great practical importance. In a proportion of cases, however, amounting in our own series to 12 per cent., Loudness Recruitment is present, and in these we have ascribed it to hair cell changes resulting from interference by the tumour with the labyrinthine blood supply.

Tests of vestibular function invariably yield abnormal results. Spontaneous nystagmus is present in cases which have reached the neurological stage and all without exception show changes in the caloric responses on the affected side.

In Table 2 the essential features of the pathology, symptoms and physical signs of these four organic disorders of the eighth nerve system are seen assembled in a simplified form.

Taken together, these disorders make up much of

that ill-defined assortment which has in the past carried that indiscriminating label Ménière's syndrome.

It is, of course, the longstanding task of otoneurology to resolve this assortment into its component parts, and that by the methods in which Ménière himself so excelled, of clinico-pathological analysis. It is, therefore, the hope of myself and my colleagues at Queen Square that the information assembled in this table will encourage you to believe that progress with this task is not at a standstill.

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