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THE OTOSCLEROSIS PROBLEM: INCLUDING REPORTS OF TWO CASES EXAMINED PATHOLOGICALLY

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IN the following pages the writer, after giving reports upon two cases of otosclerosis examined pathologically and one of them also clinically, intends to discuss the ætiology of the disease.

CASE I

3.3.31.—F. H., aged 27, admitted to Middlesex Hospital, March 3rd, 1931, suffering from Hodgkin's disease (see Hospital Report). At that date she was deaf in both ears, the right being the worse. Her deafness dates from the beginning of menstrual life at the age of 14. There is slight nystagmus. Patient is anaemic.

24.8.32.—Patient re-admitted August 24th, 1932. She complained of inability to walk and felt giddy on sitting up. She was then very deaf and very anaemic.

13.9.32.—Patient died September 13th, 1932. *Post mortem* examination twenty-four hours after death. Temporal bones fixed in Orth's solution (formalin 10%, Müller's fluid 90%).

Examination of the Temporal Bones

The temporal bones were fixed in Orth's solution and decalcified in a watery solution of nitric acid.

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Right Ear.—To the naked eye the middle ear appears normal, and there is no fluid or pus present. The stapes appears to be fixed in the oval window.

Microscopic examination (Fig. 1). There is a considerable thickening of the muco-periosteum of the middle ear in the neighbourhood of the footplate and crura of the stapes. The tensor tympani and stapedius muscles show no sign of degeneration. There is an area of newly formed spongy bone extending from a short distance in front of the oval window across the stapedio-vestibular articulation and along the footplate of the stapes to the posterior margin of the oval window. The stapes is thus fixed at both extremities of the oval window. The footplate of the stapes is very much thickened by the porous newly formed bone, and the latter extends for some distance up into both crura of the bonelet. The newly formed porous bone shows the usual characteristics of otosclerosis. It is defined from the normal bone of the capsule of the cochlea by a clear-cut line of demarcation. The large spaces in the newly formed bone contain marrow, and the bone tissue itself stains deeply with hæmatoxylin, in marked contrast with the pale-staining normal bone surrounding it.

On the tympanic surface of the footplate of the stapes there appears to be going on a process of absorption of the newly formed spongy bone by the thickened muco-periosteum described above. There is no round-cell infiltration in the thickened muco-periosteum or any dilatation of the blood-vessels, and the absorption of the bone appears to be carried out by the fibrous tissue of the muco-periosteum itself.

The vestibule and semicircular canals show no pathological changes. There is no effusion of blood or other exudate into these cavities.

The cochlea shows no evidence of disease. There is present the usual *post mortem* disintegration which is always found in the human organ of Corti, but no pathological alteration is seen. The nerve-cells of the ganglion spirale are present in the usual number, and show a normal appearance. There is no alteration in the position of the membrane of Reissner, and the stria vascularis is quite normal.

Left Ear (Fig. 2).—The left ear shows changes which are almost a replica of those found in the right. The muco-periosteum of the middle ear in the neighbourhood of the stapes is thickened, but is free from any round-cell infiltration or any

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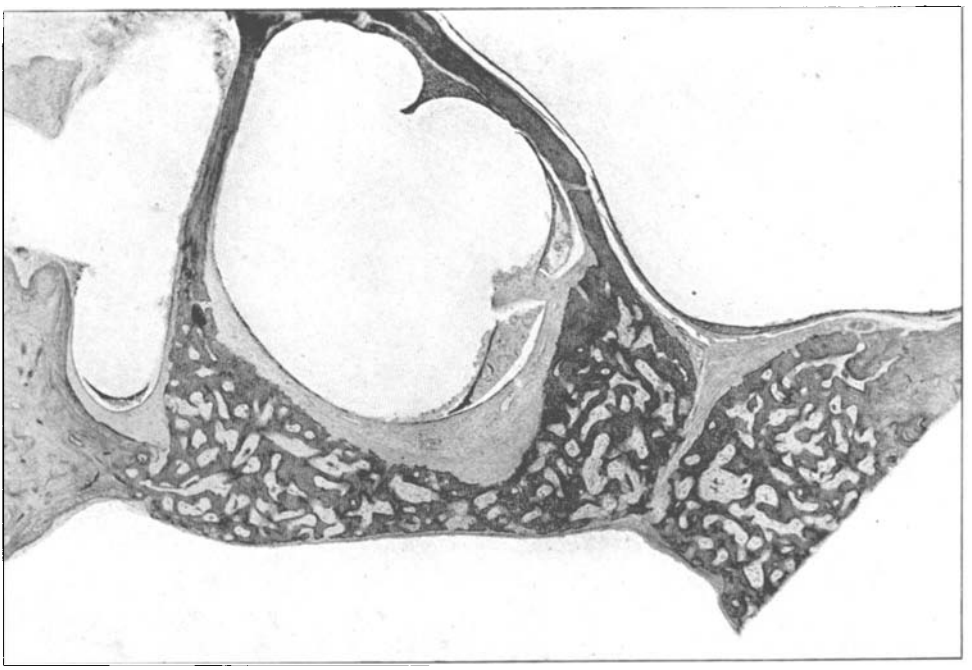


FIG. 1.

Case I.—Right ear. The footplate of the stapes is seen to be fixed by bony tissue at the posterior extremity of the oval window. Fixation was also present at the anterior extremity but is not shown in this section. For description see text. × 40 *circa*.

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sign of inflammatory activity. An area of newly formed porous bone is found extending from a short distance in front of the oval window through the stapedio-vestibular articulation, along the footplate of the stapes and for a very short distance behind the posterior margin of the stapedio-vestibular articulation. Hence the stapes is fixed in the oval window both in front and behind. The footplate is very much thickened by the newly formed bone, and the latter appears to be undergoing absorption to a certain extent by the thickened muco-periosteum lining its outer surface. This process of absorption, however, has not occurred to such a great extent as on the right side. The vestibule and semicircular canals show no pathological change.

The cochlea appears to be normal, the disintegration seen in the organ of Corti being the result of *post mortem* changes. The cells of the ganglion spirale have a healthy appearance, and show no sign of any diminution in number. The stria vascularis shows no pathological change, and the membrane of Reissner is not depressed.

When the line of demarcation between the newly formed bone and the normal dense bone of the capsule is examined under a higher power (Fig. 3), it is found that dilated veins run from the normal bone to the line of demarcation and there end abruptly. At these points the normal bone appears to be undergoing absorption round the ends of the walls of the veins.

Case II

Clinical Report

E.C., female, aged 41. Admitted to hospital, October, 1932, suffering from carcinomatous glands in the supraclavicular region on both sides, secondary to a carcinoma of the breast which was removed in 1929.

Patient has been dull of hearing for eighteen years. The onset was gradual and at no time has there been any pain in, or discharge from, either ear. Tinnitus is constantly present and patient likens it to the roaring of the wind. Paracusis is very strikingly noticed by the patient, and her friends all observe that she hears quite well when in a train or bus or where there is considerable noise.

Family History.—One brother and one sister, besides the patient, are deaf out of a family of ten. Neither of these complained of tinnitus. Patient's father was deaf.

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Albert A. Gray

Examination of the Ears

Both tympanic membranes are normal in appearance.

Right ear.	Left ear.
Watch. 1 in. (normal 5 in.).	*Watch.* 1 in.
Whisper. 1 ft.	*Whisper.* 2 ft.
Tuning-fork. All notes lost below la_2.	*Tuning-fork.* All notes lost below sol_2.
Bone-conduction. Rinne—25.	*Bone-conduction.* Rinne—20.
High notes. Very considerable loss.	*High notes.* Not lost so much as in right.
Galton's whistle. Mdh. 6·0, pfl. 5·87.	*Galton's whistle.* Mdh. 6·0, pfl. 5·02.

Pathological Report

Right Ear.—There is no perforation or scarring of the tympanic membrane and no sign of inflammatory activity, past or present, in the middle ear.

During the process of removing the cochlear nerve from the internal auditory meatus for separate examination, the bone, unfortunately, was fractured, the fracture running along the roof of the meatus and down to the oval window. Fortunately, however, the fracture did not affect the subsequent microscopic examination to any important extent.

Right Ear (Fig. 4).—On microscopic examination of the right ear an area of otosclerotic bone is found in front of the oval window and it presents the usual characteristics of the disease. It stains deeply with hæmatoxylin, is porous in texture with the spaces filled with marrow, and has a sharp line of demarcation. It is fairly large in area, the diameter being about two or three millimetres. Ankylosis of the stapedio-vestibular articulation is not present, but the newly formed bone has bulged outward over the articulation and thus caused locking at the joint. The footplate of the stapes itself is unaffected, but there is a small area of otosclerotic bone in the wall of the oval window at its posterior extremity. No ankylosis is found here.

On examining the line of demarcation under a somewhat higher magnification, points are found at which veins lying in the normal bone run towards the newly formed bone and there end abruptly at the line of demarcation and seem to be the factor in the formation of the new otosclerotic bone (Fig. 5). It is interesting to observe that though islands of cartilage are present in the normal bone they appear to take no part

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FIG 2.

Case I.—Left ear. The porous dark-stained newly-formed bone is shown clearly with its sharp line of demarcation anteriorly (to the right). Posteriorly the newly-formed bone has caused fixation at the posterior extremity of the oval window (to the left). The footplate is also fixed at its anterior extremity. × 30 *circa*.

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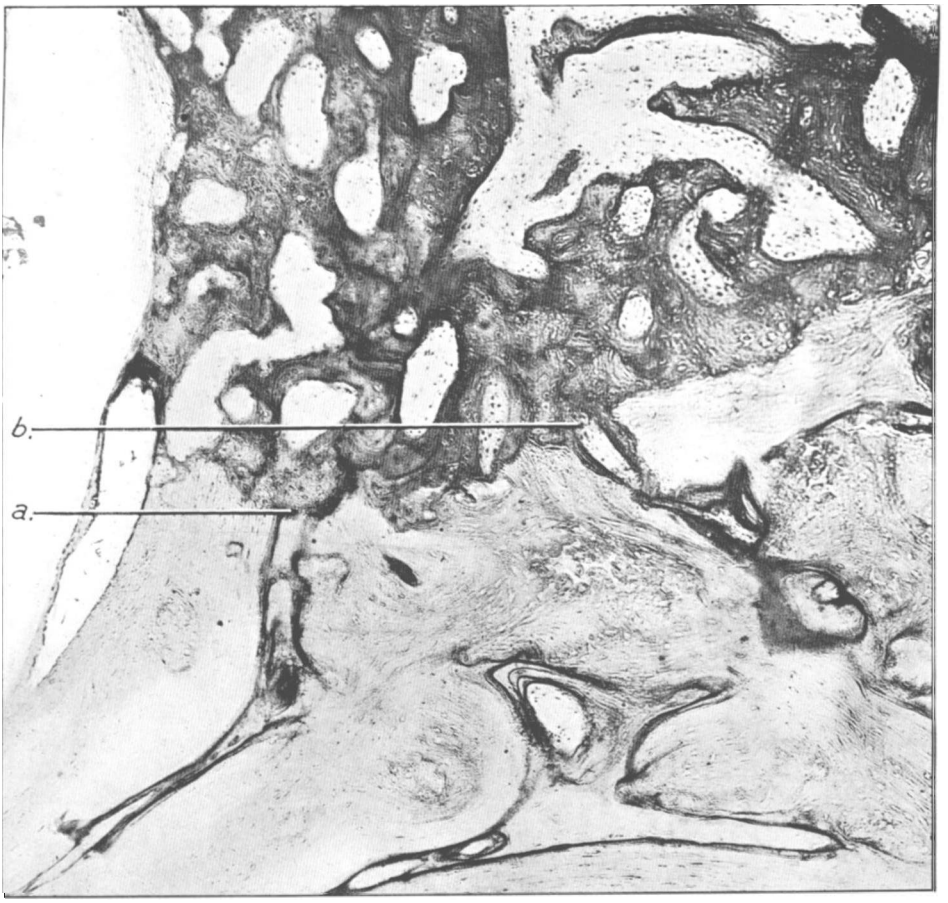


FIG. 3.

Case I.—Left ear. Line of demarcation. The newly-formed dark-staining porous bone with large spaces filled with marrow is seen in the upper portion of the picture. Two dilated veins are seen, *a*, *b*, running from the normal bone (lower portion) to end abruptly at the line of demarcation. These veins appear to be the active agents in producing the change in the bone. × 200 *circa*.

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in the formation of the new otosclerotic bone. The walls of the veins appear to be active agents in this process and the islands of cartilage become absorbed when the new bone advances upon them, but there was no process of calcification of cartilage preliminary to absorption by bone corpuscles such as occurs in the formation of bone in cartilage under normal circumstances.

The cochlea appears to be quite normal. The organ of Corti and stria vascularis show the effects of *post mortem* maceration which is always present when the tissues are not fixed instantly after death, but no sign of pathological change is found. The ganglion spirale appears to be perfectly healthy, and the nerve-cells are normal both in numbers and structure.

Owing to the fracture which occurred during the preparation of the bone, the walls of the internal auditory meatus have fallen in. Two very small areas of otosclerotic bone are found in the walls of the internal auditory meatus; one on the anterior and one on the posterior wall.

On the examination of the cochlear nerve, distinct evidence of pathological changes are found. These consist of patches in which the spiral neurokeratin filament of the fibres had disappeared and similar absence of the neurilemma is observed. As a result of the gaps thus produced in their walls many of the fibres adjacent to one another have open communication between. The axis cylinders are present and show no abnormality, but in the regions in which the spiral filament is absent they tend to lie in contact with the neurilemma and are not always easy to see. When viewed in longitudinal section, the cochlear nerve shows a patchy appearance (Fig. 6) resulting from disappearance of the spiral filament and neurilemma above described, and in striking contrast with the normal cochlear nerve.

The vestibular nerve appears to be quite healthy, the neurilemma and spiral filament of the medullary sheath being present, and the axis cylinders lying in their usual place in the middle of the fibres.

Left Ear.—The tympanic membrane shows no sign of disease, and the tympanic cavity itself is similarly healthy in appearance except in the immediate neighbourhood of the oval window and stapes. In this region there is a fair degree of thickening of the muco-periosteum due to the development of fibrous tissue between the bone and the epithelium of the middle ear. It

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is limited strictly to the area bounded anteriorly by the margin of the otosclerotic bone in front of and behind the oval window. The remarkable feature about this fibrous tissue is the way in which it has produced absorption of the bony tissue of the stapes. It will be seen in Fig. 7 that the whole of the posterior crus has disappeared, and only a fragment of the anterior crus remains. The footplate still remains intact.

A comparatively large area of newly formed bone is found in the usual place in the capsule of the labyrinth in front of the oval window. This area extends forward for several millimetres. It does not quite reach the wall of the lowest whorl of the cochlea. Externally it extends to the wall of the middle ear, and internally it forms a portion of the wall of the vestibule. It extends to, but does not involve, the stapedio-vestibular articulation; and there is therefore no bony ankylosis at the anterior extremity of the joint. The footplate of the stapes is not the seat of any new bone-formation, except in a minute area at its extreme posterior termination. This area extends across the articulation into the adjacent region of the wall surrounding the posterior portion of the oval window. A sharp line of demarcation separates this otosclerotic area from the normal bone surrounding it.

Both the areas of newly formed bone of the oval window and that behind it show all the characteristic features of typical otosclerotic bone. It takes up the hæmatoxylin stains greedily, is porous in texture with large spaces filled with marrow, and its edges are sharply defined by the line of demarcation.

The cochlear nerve shows appearances almost identical with those already described as occurring on the right side. The neurokeratin filament is degenerated and, to a large extent, has disappeared, and the fragments left show little faculty for taking up the hæmatoxylin stain (Fig. 8). On longitudinal section many gaps are seen in the fibres owing to this breaking down of the filament. Gaps in the walls of the fibre are also present, owing to a similar but less extensive degeneration of the neurilemma. The axis-cylinders on the other hand, are present in normal numbers and stain in a normal fashion. Instead of lying in the middle of the fibre, however, they tend to gravitate to the side and lie in contact with the neurilemma. The vestibular nerves are perfectly normal in appearance.

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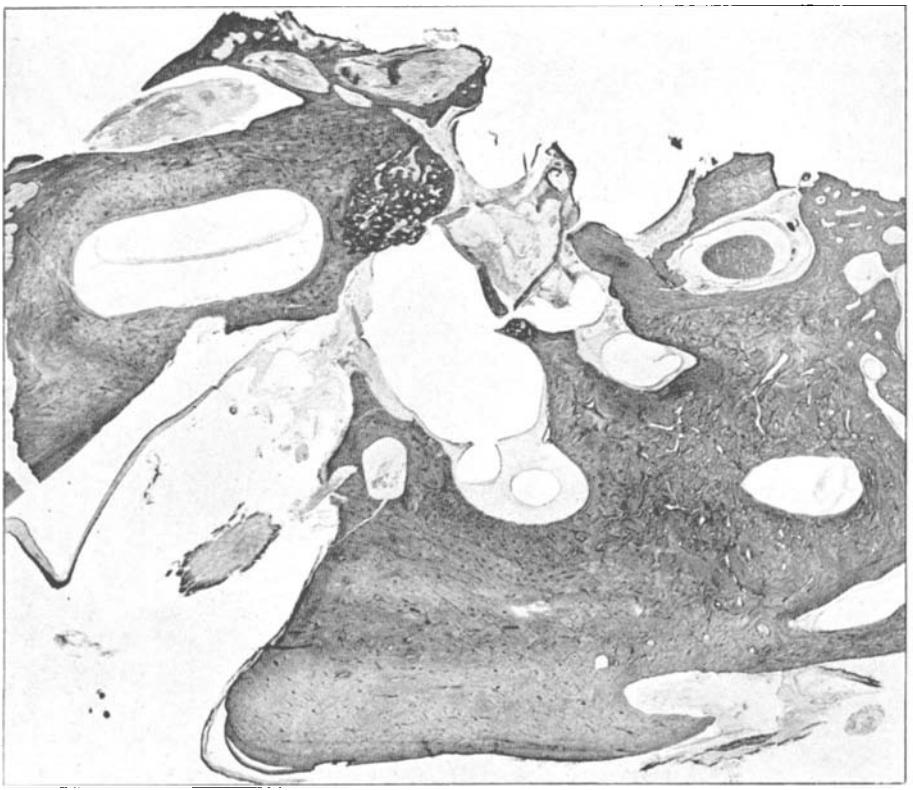


FIG. 4.

Case II.—Right ear. A comparatively large area of newly-formed bone is seen in the wall of the anterior extremity of the oval window, but there is no ankylosis. A very small patch of newly-formed bone is present in the posterior wall of the oval window. The footplate of the stapes is unaffected. × 10 *circa*.

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The Otosclerosis Problem

Before passing on to consider the cause of otosclerosis it is necessary to recall some other cases recorded by the writer because they demonstrate facts which have frequently been ignored, with the result that fallacious statements have been made, and incorrect theories formed in regard to the nature of the disease.

It is constantly assumed that the change in the bone in otosclerosis *always* consists of the absorption of a portion of the normal bone surrounding the labyrinth and the deposition of new bone to an extent greater than that absorbed. This statement is true in the great majority of cases, but there are occasional exceptions. In the writer's work on otosclerosis,* in the pathological section, an example (Case II) is shown in which the bony tissue in the affected area was absorbed and no new bone at all was laid down. In the same work (Case IV) two types of bony change in one temporal bone may be seen. The change in front of the oval window and in the footplate of the stapes shows typical otosclerotic bone, but in the region behind the oval window the absorption of bone is going on more quickly than the deposition, and the texture is therefore quite different from that in typical otosclerosis. It is more like that found in osteitis deformans.

We are not justified, therefore, in speaking of the change in the bone in otosclerosis as if all cases were similar. The great majority are so but there are exceptions to the rule, and these must be explained in any theory which attempts to account for the occurrence of the disease.

Another very important point is in regard to the extent to which the bony change in the capsule of the labyrinth occurs. It is sometimes assumed that the diseased area keeps slowly extending in all directions. Such is not the case. Usually the anterior limit at least remains stationary for many years, and indeed, is frequently determined even at the beginning of the disease. This is demonstrated in Cases III and IV in the writer's work mentioned above, in which the disease had lasted for twenty-five and sixty-three years respectively. No doubt there are cases in which the bony change may slowly advance forwards, but they are not at all common. In the posterior direction the disease is rather more indefinite in its limitations.

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* Gray, *Otosclerosis: Idiopathic Degenerative Deafness*, H. K. Lewis, 1917.

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In the slow course of time it usually reaches the footplate of the stapes and, extending along the latter, passes over the posterior extremity of the stapedio-vestibular articulation into the margin of the wall of the oval window, and thus causes very complete ankylosis. Even here, however, there are exceptions, as may be shown in Case III,* mentioned above. In this case, after twenty-five years, the bony change was limited at its posterior margin to a minute portion of the anterior extremity of the stapes; and the bridge of bone was only a small fraction of a millimetre in width, and in depth was limited to three, or at the most four, consecutive thin microscopic sections of about 20 μ thickness. This was true of both ears, and it enables us to make this important statement that *the extent of the changes in the bone bears very little relationship to the duration of the disease.*

But this case illustrates another equally important fact in regard to otosclerosis. The deafness in this patient was very severe. The report of the clinical examination is given in the work referred to above,† and it shows that the conversational voice was not heard at all by the right ear and only at a distance of one inch by the left ear. The other tests, as may be seen, showed correspondingly severe deafness for other sounds. But as mentioned above, the bony change in the labyrinthine capsule was of very small extent and the area over which ankylosis existed was very minute. This was true of both ears. We may conclude, therefore, that in otosclerosis the *extent of the pathological changes in the bone bears comparatively little relationship to the severity of the deafness.*

From this case we may also learn something in regard to tinnitus. This symptom was severe and caused the patient great distress. In Case IV in the same work‡ tinnitus, though more or less constantly present, was slight in degree and had troubled the patient hardly at all. In this case the bony change was extensive, reaching throughout the whole length of the footplate of the stapes and to a considerable depth into the posterior wall of the oval window, thus causing ankylosis over the whole circumference of the articulation. We may infer therefore that *the extent of the change in the bone bears no relationship to the severity of the tinnitus.*

* Op. cit.
† Op. cit.
‡ Op. cit.

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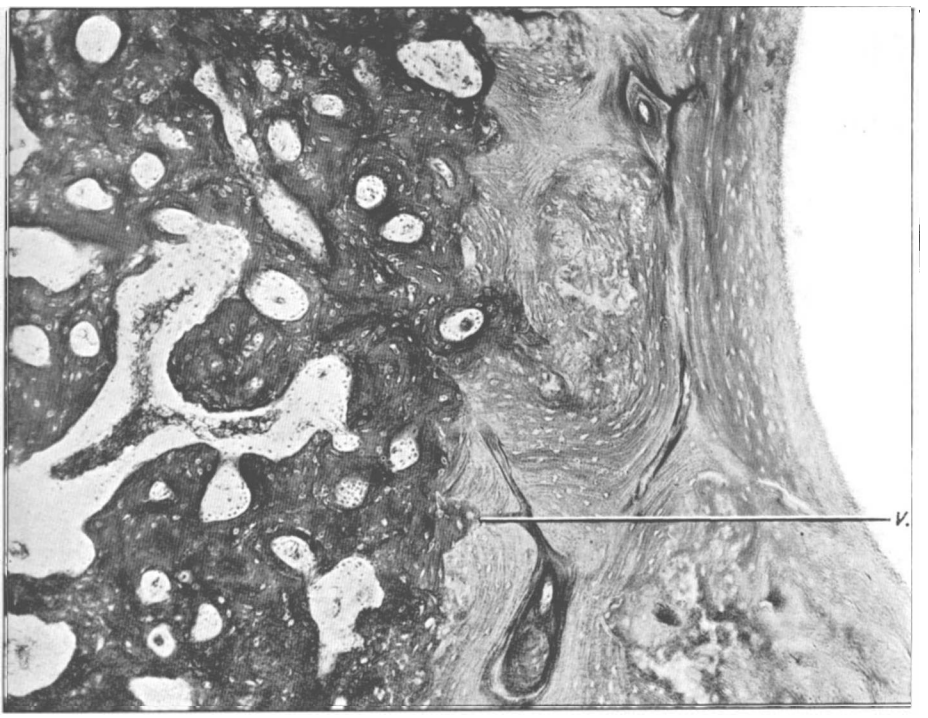


FIG 5.

Case II.—Right ear. Line of demarcation. A dilated vein, *v*, is shown running from the normal bone up to the line of demarcation, there ending abruptly where the transformation of the bony tissue is just beginning to occur. × 150 *circa*.

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Symmetry.—The changes in the bone which are found in otosclerosis show a remarkable symmetry, and the symmetry extends even to minute details. This is demonstrated in the cases recorded in the writer's work on otosclerosis* and his experience since the appearance of that work has confirmed the fact. So much is this the case that when the changes in the bone on one side have been revealed by microscopic examination it is fairly safe to prophesy that the changes in the other ear will be found to be almost identical. No doubt there may be exceptions to this rule but they must be comparatively uncommon.

Before leaving the subject of the changes in the bone which are found in otosclerosis, it is necessary to consider pathological conditions in the temporal bone which are very similar to the changes found in otosclerosis, but which occur with great frequency in certain other diseases. Jenkins and others have described such changes in the temporal bone in cases of osteitis deformans, Nager and others have described them in cases of fragilitas ossium associated with blue sclerotics, and the present writer has described them in cases of neurofibromatous tumours of the acoustic nerve.

The changes in the temporal bone which are found in cases of osteitis deformans are, indeed, somewhat similar to those found in otosclerosis. The only temporal bone from a case of this kind which the writer has had the opportunity of examining was one kindly lent to him by Mr. Livingstone of the Ferens Institute. It shows changes in the bone almost identical, not with typical otosclerotic bone, but rather with that described by the present writer in the work already cited.† It is Case IV in that work and shows that, although new bone is being deposited, absorption is taking place more quickly than deposition, with the result that constantly enlarging spaces filled with marrow are being formed.

In regard to the changes in the temporal bone in fragilitas ossium with blue sclerotics, the writer has had the opportunity of examining histologically only one case, which was presented to the Ferens Institute by Mr. Cleminson, who received it from Professor Nager. The texture of the bone appears to be very similar to that found in typical cases of otosclerosis.

* Op. cit.
† Op. cit. p. 138.

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In these cases of osteitis deformans and fragilitas ossium, however, although the changes in the bone closely resemble those characteristic of typical otosclerosis, their distribution is very different. In both, the changes were distributed over a far greater area than is found to be the case in otosclerosis. The whole bony covering of the cochlea was affected and in the case of fragilitas ossium the disease extended into the region of the semicircular canals. It would not be justifiable to say that such widespread changes are never found in ordinary otosclerosis but if they do occur they must be rare indeed. The writer has never seen such a case.

The changes in the bone described by the present writer as being found in three cases of neurofibroma of the acoustic nerve are identical with those found in typical otosclerosis and, furthermore, in two out of the three cases the locality of the diseased area was the same as that found in otosclerosis, that is immediately in front of the oval window. In the third case the deposit of newly formed bone was found in the apical whorl of the ductus cochlearis. These conditions are described and illustrated in the writer's "Atlas of Otology", Vol. ii.

In discussing the bony changes found in otosclerosis, mention must be made of the experiments of Wittmaack. He found that by ligaturing the veins surrounding the foramen magnum in the fowl and thus bringing about stagnation of blood in the veins of the bony covering of the labyrinth, a change was produced in the bone after some little time had elapsed. New bone was formed round the blood-vessels affected, which showed all the characteristic features of that found in otosclerosis. The significance of this fact will be referred to more fully in the following pages.

In the cavity of the tympanum changes characteristic of otosclerosis are frequently absent altogether. Of course, as Fraser and others have shown, pathological conditions in the middle ear, such as suppurative or other evidences of inflammatory activity may be found, and indeed may have some causal relationship to the development of otosclerosis in those particular cases. But these changes are not actually characteristic of otosclerosis and are frequently present when the latter disease is not. Apart from such changes, however, there is one which, so far as the writer is aware, is really characteristic of otosclerosis though it is not met with very frequently. This

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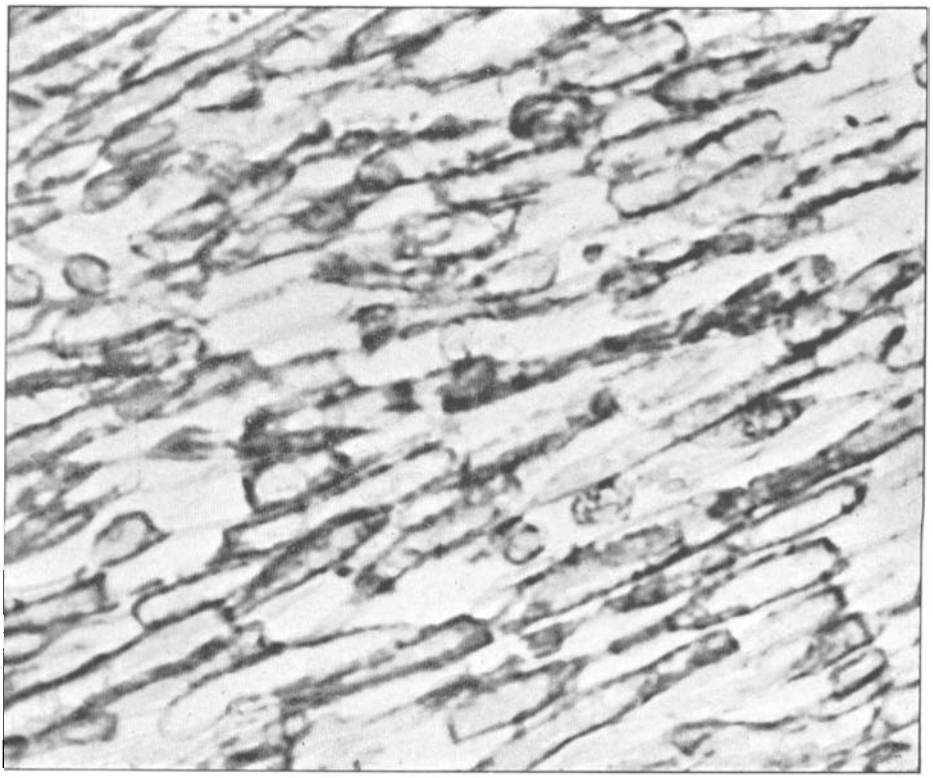


Fig. 6.

Case II.—Right ear. Longitudinal section of right cochlear nerve. The neurokeratin filament is disintegrated in many parts, and this gives rise to the appearance of large gaps in the course of the nerve-fibres. × 300 *circa*.

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condition is illustrated in the writer's "Atlas of Otology", page 65. It consists in what appears to be degenerative change in the muco-periosteum of the middle ear. The muco-periosteum is thickened and there is a great development of fibrous tissue. There is no round-cell infiltration, the blood-vessels are not dilated and there is no evidence of inflammatory activity. In all cases hitherto examined the change is limited to the region of the stapes and oval window. This degenerated muco-periosteal tissue has the power of absorbing the otosclerotic bone over which it lies, and in the case referred to above this absorption has reached an extreme degree, so that a very considerable portion of the otosclerotic bone has disappeared from the footplate of the stapes and the adjacent region in the anterior wall of the oval window. But the activity of this degenerated muco-periosteum is not restricted to the absorption of the otosclerotic bone alone. It may cause absorption of the normal bone of the crura of the stapes though to a smaller extent.

Pathological changes in the inner ear and the acoustic nerve were, until recently, assumed to be absent or, if present, were accepted as being secondary to the changes in the bony capsule of the labyrinth. In the year 1932, however, the present writer demonstrated changes in the cochlear nerve in a series of cases of otosclerosis, some of which had been examined clinically during life.* These were of the nature of a degenerative change in the medullary sheath and neurilemma of that nerve, the vestibular portion of the eighth nerve being found to be normal. It was found that the neurokeratin spiral filament of the medullary sheath was in a condition of disintegration and had in many parts disappeared altogether. The neurilemma was also affected and had disappeared in parts, so that numerous large spaces were formed and the axis-cylinders of the fibres in the affected patches were no longer insulated. This change was found even in early cases of otosclerosis before ankylosis at the stapedio-vestibular joint had occurred. Deafness was present even in these early cases; a fact which proves that the deafness of otosclerosis is by no means entirely dependent upon fixation of the stapes. It is important to observe that the axis-cylinders, although no longer insulated, were present in their usual number and absorbed the stains in the same way as

* Gray, *Proc. Roy. Soc. Med.*, xxv. (Sect. Otol.); and *Journ. Laryng. and Otol.*, 1932, xlvii., No. 9.

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do normal healthy fibres. In respect to the fibres of the vestibular nerve and those of the semi-circular canals, it is to be noted that they were quite healthy and showed marked contrast with the diseased fibres of the cochlear nerve.

The structures within the cochlea show little or no pathological changes in otosclerosis, except perhaps in the very latest stages of the disease. The organ of Corti shows nothing more than the *post mortem* disintegration which is always found in normal hearing individuals, because in the human subject fixation of the tissues cannot be carried out immediately after death. The cells of the ganglion spirale are found to be normal in appearance in otosclerosis, even after the disease has existed for many years.

These then, briefly described, are the pathological changes which have been found to occur in otosclerosis. From our knowledge of the clinical symptoms, however, we may reasonably assume that other changes in the auditory tracts within the brain may be present, and these may account for those cases in which extremely severe tinnitus is a symptom, as also for those in which the symptom described by Mr. Somerville Hastings as " crossed paracusis " occurs. At present we know nothing of the changes which may ultimately be found in the auditory tracts in the brain in cases of otosclerosis.

Before proceeding to attack the difficult problem of the ætiology of otosclerosis, it is necessary to consider briefly the clinical features of the disease. Only a few words, however, are required for this purpose, since these clinical features are for the most part familiar to every otologist. The deafness need be referred to only in order to emphasize its slow progress and particularly the loss of the low notes of the scale. Tinnitus may be very slight in degree, but it is rarely absent altogether, and of course may be very severe. Paracusis Willisii, when present, is very characteristic of otosclerosis, but it is absent in a large number of cases ; and the same may be said of the rosy tint seen in the posterior half of the tympanic membrane. The vasomotor response of the blood-vessels of the tympanic membrane is very sluggish, as was pointed out some years ago by the writer. This sign is rarely, if ever, absent. The sensitiveness of the membrane is also usually diminished. There is no need to emphasize the relatively much greater loss of hearing by air-conduction than by bone-conduction. Finally, the condition which the writer has described as otosclerosis

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paradoxica must be mentioned.* In these comparatively rare cases a sudden but very temporary improvement in the hearing occurs when certain rapid changes in the distribution of the blood in the body takes place. And these may be correlated with the improvement which occurs when the patient inhales nitrite of amyl.

This brief reference to the clinical features of otosclerosis has been made with the object of emphasizing the fact that any attempt to explain the cause of the disease must account for the presence of these signs and symptoms. It is, indeed, remarkable how so many of the pathologists who have interested themselves in this subject have become so obsessed with the changes which are found in the bony covering of the labyrinth that they have apparently forgotten the clinical aspect of the problem. Yet the latter really demands attention quite as much as the pathological aspect. But, in addition to the required explanation of the clinical symptoms, the curious association of otosclerotic changes in the bony covering of the labyrinth with fragilitas ossium, blue sclerotics, osteitis deformans, and acusticus tumour also demands elucidation. The evidence of hereditary tendency to the disease in many cases is unquestionably a fact and must be accounted for in any attempt to explain its nature. The complexity of the problem therefore appears to be very considerable.

The theories which have been put forward to account for the occurrence of otosclerosis are numerous and varied. Many of them can be described rather as pious opinions without any basis in fact. There is an element of unconscious humour about some, as for example that which attributes the disease to the deficiency of an ovarian hormone, the male human subject having apparently no existence in the scheme of things. In the following pages, however, only those theories which at least rest upon some reasonable basis of fact will be considered.

Wittmaack† considers that the changes in the bone in otosclerosis are due to stagnation of blood in the smaller vessels in the bony covering of the labyrinth. He bases his opinion on the result of experiments upon fowls which have already been referred to.

The writer has had the opportunity of examining Wittmaack's preparations, and he is of opinion that the newly formed bone

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† *Acta Otolaryngol.*, xiv., fasc. 1 and 2.

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produced as a result of these experiments is in no way different from that which is found in otosclerosis. Furthermore, for reasons which will be given later, the writer also agrees with Wittmaack that these changes in the bone are the result of stagnation of the blood in the smaller blood-vessels. This, however, is an explanation of the changes which occur in the bone in otosclerosis, but it does not account for the occurrence of the disease itself, and Wittmaack himself emphasizes this point. For the obvious question arises: why should the blood stagnate in that particular region in the neighbourhood of the oval window in certain individuals and not in others? And furthermore, why should the tendency to this condition be so frequently transmitted from parent to offspring. While, therefore, Wittmaack's experiments give a satisfactory explanation of the changes in the bone in otosclerosis, they do not reveal the causative factor of the disease itself, with all its varied clinical and pathological phenomena.

Otto Mayer holds the opinion that the changes in the bone are a consequence of processes of repair following splitting of the bone in the neighbourhood of the oval window during the period of growth. But the pathological facts which he cites in support of this view are not very substantial and their relationship to otosclerosis is doubtful. Moreover, this view would at best only explain the changes in the bone, and it fails to account for the other clinical and pathological phenomena which go to constitute the disease.

Nager looks upon otosclerosis as a nutritional disease of the bone. He had the opportunity of examining the temporal bone of a patient in whom otosclerosis was associated with fragilitas ossium and blue sclerotics. The points in which the changes in the temporal bone were similar to or differed from those found in ordinary cases of otosclerotics have already been pointed out in the preceding pages. It remains to be added, however, that Nager's view is subject to the same objections as that of Mayer. It may explain the changes in the bone, but the reason for the changes in the cochlear nerve as well as for many of the clinical symptoms remains obscure. In the following pages an explanation will be given which appears to the writer to account for the association of fragilitas ossium and blue sclerotics with otosclerosis.

Several investigators have put forward the view that otosclerosis arises from some defect in one or more of the

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FIG. 7.

Case II.—Left ear. A comparatively large area of newly-formed bone is present in front of the oval window. There is no ankylosis at the anterior extremity of the oval window. A small area of newly-formed bone is shown in the wall at the posterior extremity of the oval window, and it will be seen that bony ankylosis has occurred. The footplate of the stapes is unaffected, but the crura have been almost entirely absorbed by the newly-formed fibrous tissue in the muco-periosteum (see text). Note the remarkable symmetry in the arrangement of the newly-formed bone in right and left ears (Figs. 4 and 7). × 25 *circa*.

[*face p.* 642

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internal secretions of the body. It is very difficult either to prove or disprove such theories, but one point which would appear to throw great doubt upon their value is the fact that many patients develop otosclerosis when they are in the best of health. If the internal secretions were defective one would expect to find some other indication of the fact in other disturbances of the health. Still this argument against these theories must not be allowed to carry too much weight. A very minor deficiency or excess of one of the hormones might conceivably be sufficient to affect the organ of hearing without otherwise perceptibly affecting the general health.

The relationship between catarrhal conditions in the middle ear and otosclerosis is difficult to elucidate. Fraser and others attach great importance to catarrhal otitis media along with other infective conditions in the middle ear, as factors in the genesis of otosclerosis. It is admitted by Fraser that the subject of the disease must have an inborn tendency towards it in order that the infective conditions may produce their effect. There is clear evidence that some cases of otosclerosis do manifest them either in the course of or very shortly after such a condition as acute middle-ear inflammation,* and the natural inference would follow that chronic middle-ear catarrh may play a part similar to that of acute inflammation. It is difficult, however, to decide the percentage of cases in which middle-ear infection is the exciting cause, the reason being that individual otologists attach different values to the various signs and symptoms in making a diagnosis. The important point to bear in mind is the fact that the essential agent is the inborn tendency to the disease. Infective middle-ear conditions can be regarded only as exciting causes and are frequently absent.

In his work published in 1917† the writer explained how he was led to the opinion that otosclerosis was a degenerative process affecting the whole organ of hearing from the external auditory meatus to the cerebral cortex. With the clinical and pathological experience which the intervening years have brought, he feels that that opinion has been more than justified and, furthermore, hopes to demonstrate in the following pages the circumstances under which this process of degeneration may occur. Before doing so, however, it is important to

* Gray, *Otosclerosis*, 1917, 35.
† Op. cit.

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emphasize a point which is often forgotten—that the organ of hearing does not include the semicircular canals or the maculae acusticae of the vestible or the nerve associated with them. These are organs of equilibration, having nothing to do with the hearing, and they made their appearance in the course of evolution long before there was any function of hearing at all. Another point which it is necessary to bear in mind is that, while in the writer's opinion otosclerosis is a degenerative change affecting the organ of hearing as a whole, it is not meant to be inferred that the degenerative process must begin simultaneously in all parts. The first part in which the degenerative process starts may in one case be the bony covering of the labyrinth, in another the cochlear nerve, and possibly in some cases even the projections of the cochlear nerve within the brain. But according to the view expressed above, the tendency is for the whole organ of hearing to be involved ultimately. The particular portion of the organ of hearing which may first become affected is very possibly determined by circumstances associated with other bodily conditions, either local or general. Thus an acute or chronic middle-ear infection would tend to bring about the change first in the neighbourhood of the oval window. Prolonged nervous strain or exhaustion, on the other hand, would tend to precipitate the changes first in the cochlear nerve or its projections within the brain. This probably explains the clinical fact which otologists have frequently noticed, that tinnitus is the first sign of the disease in quite a considerable number of cases. If the first change occurs in the cochlear nerve or in its projections within the brain it is very easy to understand that the irritation of the nerve-fibres by the degenerative process in its very earliest stage might cause subjective sensations of sound before the fibres had appreciably lost their function of transmitting nerve-impulses. This clinical fact cannot, without fantastic speculation, be explained by any theory of otosclerosis which would limit the process to disease of the bone.

In the *Journal of Laryngology and Otology*, Vol. xxxviii., page 141, the writer published an article describing several curious cases of otosclerosis in which greater temporary improvement of the hearing occurred under certain circumstances. In these cases of otosclerosis paradoxica, as he termed the condition, changes in the distribution of the blood either locally or over the vascular system as a whole, appeared to be the cause of the

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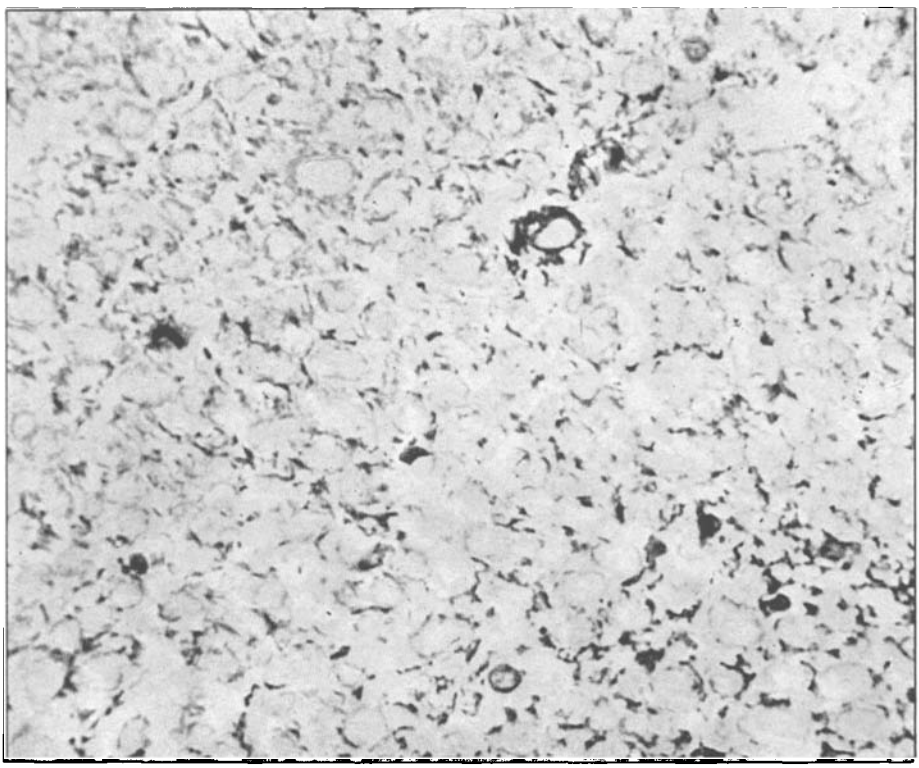


Fig. 8.

Case II.—Left cochlear nerve, transverse section. The neurokeratin filament has, to a large extent, disappeared, and what is left stains faintly and lies against the outer walls of the nerve fibres. The numerous small black spots scattered throughout the section are the axis cylinders, but they do not occupy the centre of the fibres, as they have lost the support of the neurokeratin filament. × 300 *circa*.

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temporary improvement. In one case the annual occurrence of hay-fever caused the improvement in the hearing and in another an ordinary acute nasal catarrh increased the hearing power to such a remarkable degree that the patient always looked forward with pleasure to catching a cold. In other cases it occurred after alcohol had been administered for an attack of fainting or when a draught of cold fresh air blew suddenly upon an individual who felt faint and exhausted in a hot stuffy atmosphere. The improvement was obviously the result of a change in the distribution of blood which produced temporarily an increased flow of fresh blood to the organ of hearing. Incidentally it may be mentioned that in all the cases of otosclerosis paradoxica which the writer has seen the patients were also the subjects of paracusis Willisii. Since that paper was published the writer has seen several more such cases and the condition is probably much commoner than might be thought. In some of them the improvement in the hearing is similar to that which occurs in patients suffering from otosclerosis when they inhale nitrite of amyl and is due to the action of the vasomotor mechanism in bringing about a pronounced change in the distribution of blood in the body. It was this association, together with the very sluggish and diminished vasomotor reflex of the tympanic membrane which the writer had observed for many years, that led him to formulate the explanation of otosclerosis which will be given in the following pages. Before doing this it is necessary to make as briefly as possible, some observations on the vasomotor reflex act in general, in order that what follows may be clear to the reader. But this will be done as briefly as possible and only in so far as it directly concerns the problem under consideration.

When any structure in the body is called into functional activity a larger supply of blood is necessary than when it is at rest. This increase in the blood supply is brought about by the vasomotor reflex system. The stimulus which calls upon the tissue to function directly or indirectly also stimulates the vasomotor nerves which control the blood-vessels supplying that tissue. A dilatation of these arteries and arterioles takes place in the tissue and, thus, an increased flow of blood occurs there, which enables the tissue or organ to continue functioning satisfactorily. In most tissues the increased flow of blood brings about an increase in the bulk of the tissue or organ

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because the walls of the blood-vessels are resilient and capable of dilatation; and in addition the surrounding tissue in most cases is also resilient and so allows the dilatation to take place. Hence in the case of most tissues and structures, when they function, the amount of blood in them is increased. But there are certain tissues in which the degree of resilience is very slight and in these cases the increase in the amount of blood contained in them when they are called upon to function can only be very little. But the rate of flow of the blood through the vessels in such tissues can be increased and thus the functioning tissue receives its extra nourishment. In the case of dense bone the resilience of the tissue surrounding the blood-vessels is absent altogether and the conditions of the circulation are further complicated by the fact that the nutrient blood-vessels when they enter the bone lose their muscular coat and become adherent to the bony channels in which they run. Consequently the amount of blood in bone can never vary from time to time, and when the bony tissue is called upon to function, the cells can only obtain the necessarily increased nutrition by a more rapid flow of blood. And, conversely, when the bone is not called upon to function the rate of blood through it is diminished, but the amount of blood present must remain the same. In consequence the flow must be correspondingly slow. In view of what is to follow, this is an important point to bear in mind.

When the vasomotor reflex to a given organ or tissue is defective it must be pointed out that the cells of that organ or tissue do not die for want of nourishment, because the flow of blood, though not sufficient to enable them to function properly, is still quite sufficient to keep them alive. They may undergo degeneration or they may be destroyed by neighbouring cells or, what is most usual, the tissue may remain normal in appearance.

In the special case of the organ of hearing, the vasomotor system behaves in the same way as in other organs and tissue. The stimulus which calls it into activity is sound. When sound-waves reach the ear, the tympanic membrane and the chain of ossicles vibrate, the rapidly moving footplate of the stapes puts stress upon the annular ligament and the adjacent bone, and the fluids in the labyrinth, the organ of Corti, the ganglion spirale, the fibres of the cochlear nerve and all its connections within the brain are called upon to function. On

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the other hand, since the semicircular canals and vestibule are not concerned in the act of hearing, they do not function. When, therefore, sound strikes the ear in a normal individual, a vasomotor reflex or series of reflexes occurs which allows a greater flow of blood to the structures concerned. There may be a possible exception to this statement in regard to the projection fibres of the cochlear nerve within the brain, because it has not yet been shown that the blood-vessels within the brain are supplied with vasomotor nerves. But in all the other structures mentioned the vasomotor reflex occurs, and it is quite possible that the same happens in the blood-vessels which supply the projections of the cochlear nerve within the brain.

Now it is obvious that if this vasomotor reflex response to sound fails to take place properly the organ of hearing cannot perform its functions satisfactorily, and the degree to which this function is reduced will depend upon the extent of the failure of the vasomotor reflex. A very slight sluggishness in the reflex would bring about a degree of deafness which might hardly be appreciable to the subject, and the condition might be a temporary one. In such a case, when the vasomotor reflex was restored any deafness would disappear. But let it be supposed that the defects in the vasomotor reflex were due, not to a temporary failure of the vasomotor nerve mechanism, but to an inherent weakness resulting from the neurones which govern the reflex very gradually losing their irritability to stimulus. Then the deafness would gradually increase, other changes in the organ of hearing would be liable to occur, and other symptoms supervene. It appears to the writer that these changes taken all together constitute the condition which we call otosclerosis. According to the writer's view, therefore, *the cause of otosclerosis is the partial failure of the vasomotor system of the organ of hearing taken as a whole.*

Such being the view of the writer in regard to the ætiology of otosclerosis, it is necessary to ascertain to what extent it is in agreement with the known pathological and clinical features of the disease ; for as was pointed out above, any hypothesis must explain both clinical and pathological facts or at least must not stand in contradiction to any of them.

It is interesting to observe how varied and serious are the clinical features as compared with the pathological changes so far as they are at present known. Among these are the

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deafness and its slow progress, the tinnitus sometimes occurring as the first symptom but more often appearing after the deafness, the loss or sluggishness of the tympanic membrane vasomotor reflex, the temporary improvement on inhaling nitrite of amyl and, associated with this, the occurrence of cases of otosclerosis paradoxica. These and other clinical symptoms and general characters create a large and often varied picture when compared with the very limited pathological changes in the bone in the region of the oval window and the degeneration in the sheath of the cochlear nerve, which constitute the only structural changes which are to be found.

The clinical changes may be considered first. On inspection the tympanic membrane shows no change which can be looked upon as being associated with otosclerosis. The membrane in otosclerosis is frequently more translucent than normal, but just as frequently is more opaque, and usually it is normal. The rosy pink tint seen on examination, which when present is so characteristic of otosclerosis, is due not so much to changes in the membrane as to the congestion of the vessels in the muco-periosteum and underlying otosclerotic bone between the oval and round windows. But although the tympanic membrane presents no characteristic change on simple inspection it does present a striking change when stimulated. It has become less sensitive to irritation as was shown by Fröschels many years ago, and this has been confirmed by the present writer. But a more valuable, because objective, sign of change in the membrane is that striking loss of the vasomotor reflex on stimulation. When the tympanic membrane of a normal hearing individual is subjected to pneumo-massage, the blood-vessels along the handle of the hammer at once begin to dilate and become easily visible to the eye. But in cases of otosclerosis it takes some little time to bring about this dilatation, and frequently quite a long time. The writer has observed this for many years but has made no systematic study of the matter in regard to differences in the time required to produce the response. Mr. Ashcroft, at the Ferens Institute, is at present engaged on a research in this matter, and already has amply confirmed the present writer's observations. In so far, therefore, as the clinical evidence obtainable from examination of the tympanic membrane is concerned, the writer's view of the ætiology of otosclerosis is strongly supported.

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From the tympanic cavity itself we can obtain little evidence of clinical symptoms at all in otosclerosis, beyond the fact that inflation does not perceptibly affect the hearing. The more considerable loss of the low than of the high notes, though of great value in the diagnosis, gives us no information as to the ætiology of the disease. And the same may be said of the relatively greater loss of air-conducted than of bone-conducted sound. They both depend upon the diminished mobility of the stapes owing to the bony or other changes in the neighbourhood, but they give us no clue towards elucidating the cause of those changes.

The symptoms referable to the sound-perceiving apparatus do give a certain amount of help, but not very much. Thus the occurrence of paracusis and tinnitus, although they may not directly explain the ætiology of otosclerosis, do indicate that the nerve-structures are involved in the disease, and that it is useless to attempt to explain its ætiology by consideration of the changes in the bone alone. Tinnitus is particularly important in this respect, because in the first place it sometimes precedes the deafness, and in the second place it is apparently impossible to account for it by the pathological changes in the bone or by the ankylosis which these changes bring about. Furthermore, tinnitus is sometimes hardly complained of at all in cases which reveal a severe degree of bony ankylosis on *post mortem* examination.

Another clinical manifestation occurring in otosclerosis is the striking but very temporary improvement which results from the inhalation of nitrite of amyl. The writer cannot say whether this response occurs in all cases of the disease, but in all those in which he applied the test many years ago improvement in the hearing resulted. It varied in degree but was never entirely absent. It is clear, therefore, that the deafness of otosclerosis is to a considerable extent functional, and cannot be due altogether to the fixation of the stapes or to the changes in the cochlear nerve. Of course, both these conditions must be responsible for the deafness to a considerable extent, but the improvement on inhaling nitrite of amyl is clear evidence that the structures of the organ of hearing are still capable of functioning provided they receive an adequate blood supply. This improvement cannot be due to any process of regeneration taking place in damaged tissues, because it occurs within the course of a few seconds. It can

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only be the result of restoration of function in tissues that are in themselves fully alive, but are unable to function because they are not receiving a sufficiently large supply of arterial blood.

Closely akin to the effect of nitrite of amyl is that condition first described by the writer and termed by him otosclerosis paradoxica.* Since the recording of that condition, the writer has had the opportunity of seeing a few more similar cases, and one of these may be briefly epitomized here. A woman, aged 35, was the subject of typical otosclerosis with tinnitus and paracusis, and came of a family showing a definite hereditary tendency to the disease. There were two occasions on which a striking but temporary improvement in her hearing occurred which seemed to her inexplicable. On one of these occasions she had been taken for a ride in the side-car of a motor-cycle without a windscreen and at a great speed and, as she described it, she felt smothered with the rush of air all through her head. When she returned to the warm rooms within doors she found that she was hearing much better than she had done for years. This lasted for three or four hours, but had disappeared by the following day. The second occasion occurred when she was out spending an evening with some friends. The room was warm and rather close. She was a strict teetotaller, but that evening she inadvertently took what she thought was a tumblerful of ginger ale, but too late she found that it was some kind of beer or other alcoholic beverage ! For half an hour or more she felt a very unpleasant sense of great distension in the head, but curiously enough her hearing became so sharp that she seemed to hear every sound in the room. Within an hour the deafness had returned and the feeling of distension in the head had gone.

In this and similar cases recorded in the paper referred to, the temporary improvement is clearly due to a reflex act in the vasomotor system in general bringing about an increased supply of fresh blood to the cochlea or the cochlear nerve or to the projections of the latter within the brain, which the more local vasomotor system of the organ of hearing, being defective, is unable to do. They lend, therefore, considerable support to the view expressed in this paper that otosclerosis is due to a defect in the vasomotor reflex mechanism of the organ of hearing.

* *Journ. Laryng. and Otol.*, xxxviii., 141.

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The Otosclerosis Problem

Turning now to the pathological aspect of the problem, there are two subjects to consider—the changes in the bony covering of the labyrinth which have been known for many years, and the changes in the cochlear nerve described by the writer two years ago.* To these may be added the fibrous thickening of the muco-periosteum over this region of the oval window referred to in this paper. This change, however, is very frequently absent and, when present, appears to be intimately associated with the change in the underlying bone, and may be considered along with it.

The explanations which have been given to account for the change in the bone are numerous. Inflammatory activity has been cited as a cause, but to this view there are many objections. There is no evidence of round-cell infiltration or of bacterial activity. The remarkable symmetry of the lesions on both sides in the same case appears to be too similar in their minute details to be the result of any inflammatory agent.

Deficiency in the internal secretions of various glands, such as the thyroids, parathyroids, ovaries, etc., has been suggested as a cause of the bony change by several individuals. While it is difficult to disprove such a relationship, there are much greater difficulties in the way of accepting it. The subjects of otosclerosis are frequently, and indeed usually, in perfectly good general health, a condition hardly likely to be present if the internal secretions were deficient or excessive in quantity.

For similar reasons it is difficult to accept the view that otosclerosis is the result of some defect in the metabolism of the calcium salts which leads to abnormal bone-formation. Such an explanation might account for the changes in the bone, but it leaves untouched the degenerative changes in the cochlear nerve, as well as other features of the disease such as the sluggish response of the vasomotor reflex in the tympanic membrane, the frequent presence of tinnitus, and indeed many of the signs and symptoms which go to make the clinical picture.

Wittmaack's experiments on the fowl referred to in the preceding pages are a valuable contribution towards explaining the changes in the bone. They show that these changes are the result of stagnation of the blood in the smaller vessels within the bony tissue. He is very careful to point out, however, that they do not reveal the cause of otosclerosis, nor

* Loc. cit.

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why stagnation should occur in the bone in those individuals who are the subjects of the disease.

Now it appears to the writer that the stagnation of blood in the bone at this particular region is not difficult of explanation if it be admitted that the essential defect in otosclerosis is to be found in the vasomotor mechanism governing the nutrition of the organ of hearing. It has been shown in the preceding pages that in dense bone at any rate, and to a lesser extent in other very unresilient structures, the amount of blood contained must always remain almost a constant quantity. In the soft structures, on the other hand, the amount of blood present can vary within wide limits owing to the resilience of the tissues among which the blood-vessels ramify. When, therefore, a bone is called upon to function, the extra nourishment which it requires can be obtained only by a more rapid flow of the blood through the bone. This is accomplished by the vasomotor reflex, the nutrient artery of the bone undergoing dilatation in that portion in which it still possesses a muscular coat, that is before actually entering the bone. In this way the rapidity of the flow through the bone is increased, and the extra nourishment required during functional activity is supplied, though the actual amount of blood in the bone at any given time is not increased. If, however, this vasomotor-reflex is defective, then the increased rapidity in the flow of blood will not occur when the bony tissue is called upon to function in the way of resisting stress, and difficulty in the way of supplying nourishment to the bone cells will be felt most in that region where the stress or tension is greatest. Applying this general rule to the particular case with which we are dealing, the bony covering of the labyrinth, it is obvious that the locality in which changes in the stress, caused by sound vibrations, will be greatest and most variable will be in the region of the oval window. Further, since the movements of the stapes reach their greatest amplitude at the anterior extremity of the bonelet, the stress put upon the wall of the oval window will be greatest at the anterior margin of the latter, and the lines of stress will radiate forwards from there, rapidly diminishing in their intensity as they spread forwards and become dispersed in the thick bony covering of the cochlea. This explains the well-recognized fact that the bony change in otosclerosis almost invariably begins immediately in front of the oval window. The Haversian systems of the bone in this

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region which surround and derive their nutrition from the minute arterioles nearest to the margin of the oval window will be the first to feel the effects of the insufficient blood supply. Furthermore, we are now in a position to give the reason for the spherical outline which the area of otosclerotic bone always shows in its early stage. The Haversian systems of bone lie in concentric rings round the nutrient arteriole, and when the nourishment supplied by that arteriole falls below the demand all these Haversian systems will suffer, and the resulting change in the bone will consequently show a spherical outline.

But the hypothesis given here to explain the occurrence of otosclerosis enables us to answer a much more important question involved in the changes which are found in the bone. Why is the newly formed bone so porous in texture? Every pathologist who has considered this question deeply must have felt puzzled. When repair takes place in ordinary dense bone, as is the case after fracture of the shaft of one of the long bones, the newly formed bone ultimately becomes just as dense as the bone of the two fragments of the shaft which it unites. Why then does the same process not occur in the newly formed bone in otosclerosis? The affected area in the latter case always remains porous in texture, however long the patient may live. An example of this is described in the writer's work on otosclerosis,* in which even after the disease had lasted sixty years, the porous texture of the otosclerotic area was still typical.

As pointed out in the preceding pages, under normal conditions the extra supply of blood demanded by the bony tissue, when actively functioning under the influence of the stress to which it is subjected by the movements of the stapes, is supplied by an increase in the rapidity of the flow of blood, which is brought about by the reflex dilatation of the nutrient artery before it enters the bone; and in this way the texture of the bone remains the same. But when this vasomotor reflex does not take place, increase of rapidity in the blood-flow in the bone does not occur and the bone-cells do not receive the necessary extra nourishment. The result is that these corpuscles, in order to obtain nourishment, begin to eat up the bone which they have already laid down, the process being continued by the osteoclasts which result from the coalition of several neighbouring bone-corpuscles. Spaces are thus

* Gray, op cit., p. 122.

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formed which allow more blood to pass through the part, and the supply of nourishment is thus increased even though the rapidity of the flow of blood may remain the same. When the supply of nourishment is sufficient the osteoblasts again begin to lay down bone, new Haversian systems develop and the lumen of the space formed by the previous absorption becomes diminished. But the encroachment of the newly formed bone upon the spaces can only proceed to a limited extent, because if it went any further the amount of blood present would be lessened and the necessary nutriment for the bone-cells would be unobtainable and reabsorption of the bone would again recur. The spaces, therefore, remain permanently large in comparison with those formed under normal conditions, and the characteristic porous bone of otosclerosis comes into existence. Expressed in another way, we may say that the porous bone holds a larger amount of blood than the normal dense bone in order to compensate, in supplying nourishment, for the loss which results from the defective action of the vasomotor reflex. Bone-marrow makes its appearance in the comparatively large spaces of the newly formed bone.

Keeping this point in view, many of the features of the affected area of bone may be accounted for besides its porous texture. The line of demarcation is sharp and clearly defined, because it forms the outer limit of one or more Haversian systems which derive their nutrition from one or more arterioles. In a sense, therefore, it may be likened to the sharp line of demarcation which is found in other organs of the body when an infarct occurs following embolism. In the latter case, however, the blood-supply is completely cut off, which of course is not the case in the area of otosclerotic bone.

Another characteristic of the diseased area that now becomes explicable is the limitation of its margin, especially the anterior one. No matter how long the disease has been in existence it is very rare to find the anterior margin of the otosclerotic bone more than two or three millimetres beyond the anterior extremity of the oval window. In the cases examined by the writer, the anterior margin of the porous bone appeared to be as far forward in cases which had lasted two or three years as in those in which the disease had existed for twenty-five and sixty years respectively. The explanation of this fact, according to the writer's view, does not appear to be difficult. The

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lines of stress which run forward from the anterior margin of the oval window when the stapes is vibrating, undergo rapid dispersion as they radiate forward and very quickly become so small as not to affect the living structures in the bone to a degree sufficient to require any perceptible increase of nutrition. The defective reflex vasomotor response to sound-waves, therefore, will not affect those regions. The posterior limit of the otosclerotic bone is rarely found further back than a fraction of a millimetre behind the posterior margin of the oval window and, in many cases, even after the disease has existed for a great number of years it may not have extended so far. Here again, the position of the posterior limit is probably determined by the lines of stress radiating from the margin of the oval window backwards into the bony covering of the labyrinth.

In regard to the explanation just given of the anterior and posterior limits of the bony change in otosclerosis it must, of course, be understood that it applies only to the ordinary cases of otosclerosis with which otologists are so familiar. It does not apply to those cases in which similar changes in the temporal bone occur in association with fragilitas ossium and osteitis deformans. In these conditions the disease is much more widespread than is the case in uncomplicated otosclerosis. In the latter the defect is present only in that vasomotor reflex, of which the normal exciting stimulus is sound and sound alone; and consequently any structural changes which may result from the defect will be strictly limited to the organ of hearing. In fragilitas ossium and osteitis deformans, on the other hand, the causative agent, whatever it may be, is not to be looked for in the organ of hearing, and the structural defects are therefore not likely to be limited to the auditory tract as is the case in otosclerosis. Consequently when the temporal bone is affected in these diseases we would not expect to find that the diseased area should be confined within the narrow limits by which it is circumscribed in otosclerosis.

Before leaving the consideration of the changes in the bone which are found in otosclerosis, it is necessary to say a few words concerning the development of newly formed fibrous tissue which is sometimes, though perhaps not very often, found in the muco-periosteum which covers the stapes and immediately adjacent portions of the oval window, and is strictly limited to that region. It is no manifestation of

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chronic inflammation ; there is no round-cell infiltration and no dilatation of the blood-vessels. It is obviously the result of a degenerative process in which, as is usually the case, the more primitive type of cell overcomes the more highly specialized and replaces it. An excellent example of this condition is shown in Case II in this paper. It will be seen that in the left ear (Fig. 7) the newly formed fibrous tissue has caused the disappearance of the posterior crus of the stapes and also of a large portion of the anterior crus, with the result that the contour of the bonelet is completely lost, except in the region of the footplate which has not yet been absorbed. Another interesting example of this condition may be seen in the author's " Atlas of Otology ", Vol. ii., p. 65. In that case the newly formed fibrous tissue had absorbed a large portion of the newly formed bone in the footplate of the stapes and the adjacent region in front of the oval window. Portions of the crura of the stapes were also absorbed and replaced by the fibrous tissue.

The change in the bone in otosclerosis is usually limited to a single focus, and that is to be found, with the rarest of exceptions, immediately in front of the oval window. Cases do, however, occasionally occur in which two or more foci may be present, and one of these cases has just been described at the beginning of this paper. It is the only one out of eleven with which the writer has dealt in which more than one focus was present. It is necessary to make this brief statement concerning the rarity of multiple foci because it is frequently assumed that they are common.

Turning now to the changes found in the cochlear nerve, it is to be pointed out that we must not yet assume that they are present in *all* cases of otosclerosis. They were first described by the present writer* two years ago, and the number of cases is therefore limited. In addition to the four cases recorded in that paper there falls to be added the case reported in this present article, in which also it was found that the cochlear nerve was affected. Hitherto, therefore, the nerve has been found to be affected in all the cases investigated, and this must be considered a characteristic pathological feature of the disease of the same significance as the change found in the bone.

The changes just referred to in the cochlear nerve have

* Gray, *Journ. Laryng. and Otol.*, xlvii., 598.

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already been described and it only remains to consider the cause of these changes. It is necessary first of all to say a few words concerning the structures in which the changes occur, the neurokeratin filament of the medullary sheath and the neurilemma. The neurokeratin filament is not in itself a living tissue with cells and nuclei. It is the product of functional activity either of the spongioblasts or of the neurone of which it forms a part. It is not definitely known from which of these living cells the filament is produced, but from the present point of view this is immaterial, since both the spongioblast and neurone are derived from the epiblast. Both, therefore, come of a cellular ancestry, one of whose chief functions is the production of keratin ; and the production of the neurokeratin of the medullary sheath is without doubt merely the continuation of this function in an environment different from that of the surface of the body. The disintegration of the neurokeratin filament which occurs in otosclerosis may and in all probability does mean the cessation of one of the functions of either the neurone or the spongioblast. It will be shown later that another function of the neurone which can be temporarily restored is suspended in otosclerosis. The neurone, therefore, cannot be dead. This being so, the explanation of the pathological change seen in the neurokeratin filament in otosclerosis is to be found, according to the view expressed in this paper, in the diminished functional activity of either the spongioblast or the neurone. This diminished functional activity of these cells is the result of insufficient nutrition due to the loss of the vasomotor reflex which governs the nutrition of those cells. When the neurone is called into functional activity by the effect of sound-waves acting upon the organ of Corti and causing a nerve-current to travel the whole length of the auditory tract from the cochlea to the cerebral cortex, then the neurones taking part in this activity must obtain a sufficient supply of blood. This is brought about by the vasomotor reflex of which sound is the exciting stimulus, and if this reflex fails to occur the neurone cannot function properly. And since the production of the neurokeratin filament is one of its functions, the latter is not produced to the normal extent. The organ of Corti, the cells of the ganglion spirale, and the axis-cylinders of the fibres of the cochlear nerve show no discoverable change in otosclerosis except perhaps in the latest stages of the disease.

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This completes the description of the pathological changes found in otosclerosis. But before leaving the subject it is very necessary to point out that, though no pathological changes can be found in the organ of Corti or in the axis-cylinders or ganglion cells of the auditory tract, a defect of some kind must be present somewhere in these structures. The proof of this lies in the fact that under certain unusual circumstances, such as the inhalation of nitrite of amyl, or in several of the cases of otosclerosis paradoxica described by the writer, the hearing power of the patient suddenly becomes strikingly improved in the course of a few seconds, an improvement which passes off very rapidly. This sudden improvement in the hearing power could not be due to any loosening of the fixed stapes, or sudden regeneration of the medullary sheath of the cochlear nerve. The writer can think of only one possible explanation and it is this. In otosclerosis the whole system of the organ of hearing, the nerve-tract included, does not receive its normal blood supply because of the loss or diminished sensitivity of the vasomotor reflex. In the unusual circumstances referred to, however, the difficulty is overcome by the sudden redistribution of blood which takes place in the body, and brings to the nerve-cells and fibres a full supply of blood, and with it the oxygen which is of such prime importance for the functioning of the neurone. The result is that the neurones can for the time being function satisfactorily. This appears to the writer to be a satisfactory explanation of the facts in these cases, and indeed he can think of no other possible explanation.

There now remain for consideration three pathological conditions in which changes very similar to those found in otosclerosis are frequently found in the temporal bone. They are fragilitas ossium, osteitis deformans and acusticus tumour of the neurofibromatous type.

Fragilitas ossium is associated with blue sclerotics in 60 per cent. of the cases, and with deafness similar in many ways to otosclerosis also in 60 per cent. In 44 per cent. all three defects are present together. Mr. J. S. Fraser* described three cases in 1919. These were, as is usually the case, hereditary. Mr. F. J. Cleminson† has described one in which there was no

* *Proceedings*, 1919, xii. (Sect. Otol., 115).

† *Proceedings* xx. (Sect. Otol., 13-16).

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evidence of inheritance, and Scott and Stobie* have given a report of the examination of several members of a family who suffered from fragilitas ossium associated with deafness, which showed many of the signs of otosclerosis. Adams,† de Kleijn and other writers have also recorded cases. So far as the writer is aware *post mortem* examinations have been made in only two cases. One of these was reported by Ruttin‡ and the other by Nager. Thanks to the kindness of Professor Nager and Mr. Cleminson the writer has had the opportunity of examining the microscopic sections obtained from the latter of these cases. There is no doubt that the changes in the bony covering of the labyrinth are very similar in the two diseases, in so far as the texture of the newly formed bone is concerned. But, as was pointed out above, the distribution of the diseased bone is very different. In otosclerosis the tendency is towards a narrow limitation in the immediate neighbourhood of the oval window, whereas in fragilitas ossium the newly formed bone is found widely spread throughout almost the whole of the bony covering of the labyrinth. This difference is, from the present writer's point of view, easily explained. Otosclerosis is a disease strictly confined to the organ of hearing, and all the other organs and tissues of the body are anatomically and physiologically as healthy as in normal human beings. In fragilitas ossium, on the other hand, the causative factor acts upon structures that are widespread throughout the body, and the bony covering of the labyrinth is only involved as part of the general disturbance.

But as the writer will not admit that otosclerosis is due to some constitutional defect in the endocrine glands, or in the biochemistry of bone-formation, neither can he see any reason to suppose that fragilitas ossium itself is the result of such defects. The disease of the bones known as osteogenesis fibrosa, which bears a superficial resemblance to fragilitas ossium, has been shown to depend upon disease of the parathyroids, but Muir§ has pointed out that there is no evidence at all that fragilitas ossium (osteogenesis imperfecta) can be associated with any such condition and that the cause of fragilitas ossium is unknown.

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* Stobie, *Quart. Journ. Med. Sci.*, 1924.
† Adams, *Journ. Laryng. and Otol.*, xliv., 201.
‡ Ruttin, *Monatsschr. f. Ohrenheilk. und. Laryngo-Rhinolog.*, Heft. 4, S.53.
§ Muir, *Manual of Pathology*.

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It appears to the writer that there is a connecting link between otosclerosis and fragilitas ossium, but it is of a nature different from those usually referred to, such as endocrine disturbances or aberrant metabolism in bone-formation. Just as, according to the view expressed in this paper, otosclerosis is due to a defect in the vasomotor reflex, of which the stimulus is sound, so fragilitas ossium is due to a defect in the vasomotor mechanism of which the stimulus is stress or tension. Consequently when those structures and tissues whose prime function it is to resist stress or tension are called upon to exercise that function, the increased blood-supply which they should receive does not arrive, and the tissue is unable to develop in the direction of resisting strain or tension. Hence we find that in fragilitas ossium, though the bones are the chief structures which suffer, they are not the only ones. The patients who suffer from the disease are liable not only to fractures, but also to dislocations, because the ligaments are liable to be affected. The sclerotic coat of the eyeball has to resist the stress put upon it by the contraction of the attached muscles, and does not develop the strong fibrous tissue to the normal extent because of the defective vasomotor reflex, and it retains, therefore, the blue colour of early infancy.

The relationship of otosclerosis to osteitis deformans is, up to a certain point, similar to that which it bears to fragilitas ossium. The temporal bone is involved in both cases simply as part of a more widespread disease of bones. But there are considerable differences, such as the greater age at which osteitis deformans occurs and the fact that it is more common among men than women. Further, there is no particular evidence of hereditary tendency in osteitis deformans. In all these respects fragilitas ossium appears to have a closer biological affinity with otosclerosis than does osteitis deformans, and to this may be added the fact that the bony change in fragilitas ossium shows a texture more similar to that of otosclerosis than does osteitis deformans.

But it appears to the writer that the relationship of both these diseases to otosclerosis has been unduly stressed. And he ventures to suggest that this has been done in order to prove that the cause of otosclerosis is some constitutional defect in bone metabolism in the body. Now one of the most striking features about the bony change in otosclerosis is its limitation within very narrow limits in the region of the oval window and

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its equally remarkable symmetry on both sides of the body. There is no evidence whatever that in cases of otosclerosis there is any pathological change in other bones.

The relationship of acusticus tumour to changes in the bony covering of the labyrinth stands in quite a different category from that which exists between these changes and fragilitas ossium and osteitis deformans. Acusticus tumour is definitely a local disturbance of the nerve of hearing, even in cases in which it is one tumour out of many in neurofibromatosis. Neurofibromatosis, moreover, is not a disease of bone at all.

The writer has had the temporal bones from three cases of acusticus tumour examined microscopically,* and the growths were neurofibromata in all. In all three cases changes in the bone identical with those found in typical cases of otosclerosis were present. Since the deafness began after the symptoms of the acusticus tumour had manifested themselves in all three, it may be assumed that the change in the bone was no mere coincidence but a direct result of the growth. It is necessary to observe, however, that the writer has had the opportunity of examining one case of tumour of the acoustic nerve in which no otosclerotic or other change in the bony covering of the labyrinth was found, even on careful examination. This was a case of Dr. de Kleijn's, in which the tumour on the nerve was not a neurofibroma, but a malignant growth secondary to carcinoma of the stomach. This is an important point to remember, because the effect upon the nerve of a rapidly growing and destructively infiltrating malignant tumour would be very different from that produced by the very slow-growing non-infiltrating neurofibromata.

It is difficult to see how this relationship between neurofibroma of the acoustic nerve and otosclerotic changes in the bony covering of the labyrinth can be explained on any other ground than that of interference with the vasomotor reflex of the organ of hearing. The slowly growing tumour exercising a very gradually increasing pressure on the nerve, which contains vasomotor fibres as well as the ordinary auditory sensory fibres, would naturally produce the same effects in the bone as the gradual failure in function of the same nerves which, according to the writer's view expressed in this paper, is the cause of the bony and other changes in otosclerosis.

* *Proc. Roy. Soc.*, xxv. (Sect. Otol., 85-112); ibid., 1933, xxvi. (Sect. Otol., 57).

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Women are more frequently the subjects of otosclerosis than men, the relationship being 40 per cent. men to 60 per cent. women. The usual explanation given for this preponderance of women is that the ovarian hormone (or hormones) act in some way upon the metabolism of bone, either directly or indirectly. The fallacy here is, of course, the assumption that otosclerosis is a disease of bone, whereas, as has been shown long ago, the change in the bone is only one feature of the disease.

If otosclerosis be looked upon as a weakness or defect in the vasomotor reflex mechanism of the organ of hearing, then the difficulty of explaining the preponderance of women among the victims of the disease disappears. At every menstrual epoch a profound disturbance takes place in the vasomotor mechanism of the organs of reproduction in women. As physiologists point out, a disturbance of the vasomotor mechanism in one part of the body influences all the other parts of that mechanism in the rest of the body, so that the blood can be directed in larger amount to the part where it is required. If there be among these other vasomotor reflexes of the body, one in which the nerve-cells composing it are inherently of rather feebler activity than normal, then it is not difficult to understand how the constantly recurring vasomotor disturbance of menstruation should be sufficient to prevent that particular reflex arc from fulfilling its functions in a normal manner. If the reflex vasomotor arc is that of which the natural stimulus is sound, then the organ of hearing will suffer from the defective action of that vasomotor reflex arc.

When to the vasomotor disturbances of menstruation are added the more severe, but less frequently repeated ones of pregnancy and the puerperium, we can have no difficulty in understanding why the percentage of women sufferers should be greater than that of men.

Heredity plays a considerable part in the incidence of otosclerosis and this, of course, would be expected if the cause of the disease suggested in this paper is correct. The defect in the vasomotor system of the organ of hearing is innate in the individual and not the result of any unfavourable environment. A deleterious environment no doubt can, and frequently does, have an unfavourable effect upon the disease, but this is true of all the functions of the body. The essential factor without which the disease cannot come into existence is innate in the

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individual and is, therefore, a quality capable of being inherited and liable to be transmitted to the offspring.

Summary

The essential causative factor of otosclerosis is a gradually increasing defect in the vasomotor mechanism which governs the nutrition of the structures of the organ of hearing as a whole. The axon reflexes are, of course, included in this vasomotor mechanism, and the stimulus which excites the vasomotor mechanism is sound and sound alone. Consequently the vestibular apparatus and the semicircular canals are unaffected in otosclerosis.

There is no evidence whatever of any defect in any of the endocrine glands or their secretions in otosclerosis. Neither is there any evidence of any defect in the bone metabolism of the body. On the contrary the subjects of otosclerosis are, apart from their deafness, perfectly normal individuals with ordinary average health.

The deafness of otosclerosis bears very little relationship to the extent of the disease in the bone. The deafness may be very severe when the stapes is hardly fixed at all.

The severity of the tinnitus bears no relationship at all to the extent of the disease in the bone.

The extent of the change in the bone bears very little relationship to the duration of the disease.

The extent of the changes in the bone appears to depend upon the age of onset of the disease. The earlier the time at which the otosclerosis begins, the more extensive will the bone lesion become.

The deafness of otosclerosis is to a large extent functional, and is the result of the insufficient supply of blood to all the nerve-structures concerned in the perception of sound.

The preponderance of women as subjects of otosclerosis is the result of the greater instability of their vasomotor system and the more frequent disturbances to which it is exposed.

The changes in the bone show a remarkable bilateral symmetry, even to minute details. This symmetrical distribution is readily explained by the writer's view of the causative factor of otosclerosis. The vasomotor nerves governing the nutrition of the organ of hearing are anatomically symmetrical like other nerve-structures in the body. If, therefore, structural changes occur as a result of defective functioning of those nerves, such structural changes will naturally be bilaterally symmetrical in their distribution.

. . . .

Das wesentliche ätiologische Moment der Otosklerose ist ein allmählich progressiver Defekt in dem vasomotorischen Mechanismus welches die Ernährung der Strukturen des Gehörorgans beherrscht. Die Axonreflexe sind natürlich in diesem vasomotorischen System

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eingeschlossen, und der einzige Stimulus der das vasomotorische System reizt ist der Schall. Das vestibuläre System und die Bogengänge sind somit in der Otosklerose nicht beteiligt.

Es gibt keinen Beweis dass ein Fehler in den Blutdrüsen oder in ihren Sekretionen besteht. Es gibt auch keinen Fehler in dem Knochenstoffwechsel. Im Gegenteil sind die Otosklerotiker, von ihrer Taubheit abgesehen, vollständig normale Leute, und ihre Gesundheit ist normal.

Die otosklerotische Taubheit hängt sehr wenig von dem Grad der Knochenerkrankung ab. Es kann hochgradige Taubheit mit einem kaum fixierten Stapes bestehen.

Die Stärke des Tinnitus hängt gar nicht von dem Grad der Knochenerkrankung ab.

Der Grad der Knochenveränderung hat sehr wenig Beziehung zur Dauer der Erkrankung.

Der Grad der Knochenveränderungen scheint mit dem Alter am Beginn der Erkrankung zusammenzuhängen. Je früher die Otosklerose beginnt dest. schwerer wird die Knochenerkrankung.

Die Taubheit der Otosklerose ist weitgehend eine Funktionelle, und ist durch die defektive Blutzufuhr an die in der Schallwahrnehmung beteiligten Nervenstrukturen bedingt.

Das Überwiegen der Frauen bei den Otosklerotikern ist bedingt durch die grössere Unbeständigkeit ihres vasomotorischen Systems und durch die häufigere Störungen an welche dieses ausgesetzt ist.

Die Knochenveränderungen zeigen eine bemerkenswerte Symmetrie, bis in die kleinsten Einzelheiten. Diese symmetrische Anordnung ist leicht durch die Theorie des Verfassers der Ätiologie der Otosklerose zu erklären. Wie die anderen Nervenstrukturen des Körpers sind die vasomotorischen Nerven die die Ernährung des Gehörorgans beherrschen, anatomisch symmetrisch. Wenn nun strukturelle Veränderungen auf defektive Funktion dieser Nerven folgt so werden diese Veränderungen auch symmetrisch verteilt sein.

. . . .

Le facteur étiologique essentiel de l'otosclérose est un défaut graduellement progressif de l'appareil vasomoteur nourrissant l'organe auditif entier. Les axons reflexes sont inclus dans ce système vasomoteur, et le stimulus qui excite le système vasomoteur est uniquement le son. Par conséquent l'appareil vestibulaire et les canaux semicirculaires ne sont pas attaqués dans l'otosclérose.

Il n'y a aucune évidence qu'il existe un défaut quelconque des glandes endocrines ou de leurs sécrétions. Il n'y a non plus de défaut du métabolisme osseux. Au contraire, les otosclérotiques, à part leur surdité, sont parfaitement normaux, et jouissent d'une santé normale.

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The Otosclerosis Problem

La surdité de l'otosclérose a très peu de rapport avec l'étendue de la maladie des os. Elle peut être très considérable quand l'étrier est à peine fixé.

La gravité du tinnitus n'a aucun rapport avec l'étendue de la maladie des os.

Le degré d'altération des os a très peu de rapport avec la durée de la maladie.

L'étendue de la maladie de l'os dépend de l'âge du malade au commencement de la maladie. Plus la maladie commence tôt, plus la lésion osseuse deviendra étendue.

La surdité otosclérotique est en grande partie fonctionnelle, et résulte de la provision défective de sang aux structures nerveuses qui s'occupent de la perception du son.

La prépondérance des femmes parmi les otosclérotiques est une conséquence de la plus grande instabilité de leur système vasomoteur et des perturbations plus fréquentes auxquelles il est exposé.

Les lésions osseuses sont remarquablement symétriques, jusqu'aux plus petits détails. Pour l'auteur cette symétrie s'explique aisément par sa théorie de l'étiologie de l'otosclérose. Comme les autres structures nerveuses, le système vasomoteur gouvernant la nutrition de l'organe auditif est anatomiquement symétrique. Par conséquent, si des altérations structurelles surviennent comme résultat du fonctionnement défectif de ces nerfs, elles seront naturellement symétriquement distribuées.

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