THE
JOURNAL OF LARYNGOLOGY,
RHINOLOGY AND OTOTOLOGY.

Original Articles are accepted on the condition that they have not previously been published elsewhere.

If reprints are required it is requested that this be stated when the article is first forwarded to this Journal. Such reprints will be charged to the author.

Editorial Communications are to be addressed to "Editor of JOURNAL OF LARYNGOLOGY, care of Messrs. Adlard & Son and West Newman, Bartholomew Close, E.C."

**REPORTS FOR THE YEAR 1915 FROM THE EAR AND THROAT
DEPARTMENT OF THE ROYAL INFIRMARY, EDINBURGH.**

Under the care of A. LOGAN TURNER, M.D., F.R.C.S.E., F.R.S.E.

PART IV.

THE PATHOLOGY OF OTOSCLEROSIS.¹

By J. S. FRASER,

Assistant Surgeon, Ear and Throat Department, Royal Infirmary, Edinburgh;

AND

R. MUIR,

Demonstrator of Pathological Methods, University of Edinburgh; Fellow of the Pathological Society of Great Britain.

THIS communication consists of two parts: (I) In the first, one of us (J. S. F.) will briefly recapitulate the main theories in regard to the Pathology of Otosclerosis, and describe the results of the microscopic examination of the middle and inner ear from a case of "otitis vasculosa" associated with chronic middle-ear suppuration and cholesteatoma. (II) In the second part, one of us (R. M.) will give an account of the changes found in otosclerosis as they appear to a general pathologist.

PART I. (J. S. F.)

At the present time it appears to be impossible to state with confidence that the condition variously known as "otosclerosis,"

¹ The work was carried out in the laboratory of the Royal College of Physicians and in the Pathology Department of the University of Edinburgh.

"dry middle-ear catarrh," "spongification of the labyrinth capsule," "otitis vasculosa petrosa," "otitis stapedio-vestibularis," "petrous osteodystrophy," etc., is a pathological and clinical entity. It may be that the condition we know as "otosclerosis" is made up of two or more pathological conditions in somewhat the same way that "œdema" may result from affections of the heart, kidneys, nerves, or from inflammatory processes. All we know is that, apart from suppurative otitis media and adhesive processes in the tympanum, there are many cases of chronic progressive deafness (usually accompanied by tinnitus) in which functional examination of the auditory apparatus gives us the results we associate with a lesion of the sound-conducting mechanism—*i. e.* raised lower tone limit, lengthened bone-conduction, and negative Rinne test. Microscopic examination of such specimens as have been available have shown certain changes in the labyrinth capsule, especially in the anterior bony margin of the oval window. In the early stages of the disease we find here vascular spongy bone containing numerous spaces which show a central blood-vessel surrounded by some connective tissue. Osteoclasts are not found at this stage. Later on the diseased area becomes sclerosed by deposit of new bone in the walls of the spaces. The process usually extends to the stapedio-vestibular joint and results in bony fixation of the stapes. In the early stages the nerve structures are normal as a rule, but in the later there is usually a considerable degree of atrophy affecting Corti's organ, the stria vascularis, the spiral ganglion, and the cochlear nerve.

THEORIES AS TO THE PATHOLOGY OF OTOSCLEROSIS.

There appear to be only four possible ways in which the affection may arise: It may be congenital; it may follow inflammatory changes in the middle ear; it may be due to infection through the blood; it may be caused by abnormal conditions in the nerve supply of the parts affected.

(1) According to the first theory, otosclerosis is a congenital anomaly of the process of growth in the petrous temporal bone, due to the presence of certain determinants in the patient's blood. This anomaly only becomes manifest after puberty. Under normal conditions the growth of the membranous labyrinth and its bony capsule ceases soon after birth, though islands of cartilage may occur in the labyrinth capsule in the region of the anterior margin of the oval window (Figs. 19 and 20). According to the first theory growth continues in this cartilage bone and later spreads to

FIG. 2, Section 295 ($\times 8$ diam.).—Vertical transverse section through cochlea, showing three areas of otitis vasculosa (otosclerosis). 1, Middle fossa. 2, Area of otitis vasculosa. 3, Cholesteatoma. 4, Remnant of lymph space in capsule of cochlea. 5, Tubal part of tympanic cavity: the tympanic cavity in this case is divided into an upper part (lined by cholesteatoma) and a lower part communicating with the Eustachian tube. 6, Scala vestibuli of basal coil. 7, Two areas of otitis vasculosa. 8, Scala tympani of basal coil.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

FIG. 3. Section 255 ($\times 8$ diam.).—Vertical transverse section through cochlea showing two areas of otitis vasculosa. At the apex of the cochlea the bony changes reach the endosteum. 1, Basal coil. 2, Middle coil. 3, Helicotrema. 4, Two small areas of otitis vasculosa. 5, Cholesteatoma. 6, Tubal part of tympanic cavity. 7, Area of otitis vasculosa.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

the footplate of the stapes, finally resulting in ankylosis. Those who hold this view lay great stress on the hereditary transmission of the disease, though they admit that the immediate cause of otosclerosis may be provided by puberty, by the puerperium, or disease of the tympanic mucosa. Further, Siebenmann, who supports this theory, points out the similarity of the changes seen in otosclerosis to those which occur in a rib in cases of empyema. (In such cases the process must surely invade the rib from the periosteum?)

(2) Otosclerosis is regarded by some as a chronic inflammatory process spreading from the mucoperiosteum of the middle ear and following attacks of catarrhal or suppurative otitis media. The inflammatory condition is supposed to linger about the niche of the oval window and to invade the bone from the deep layer of the mucosa. Such an invasion may be favoured by the anastomosis which occurs at this spot between the tympanic vessels and those of the bony labyrinth capsule. Further, as Walker and I have pointed out, the joints in the middle ear are the only articulations in the body covered by mucous membrane, and therefore liable to infection from the surface.

This view of the pathology of otosclerosis does not by any means exclude the hereditary transmission of a tendency to the disease. It may well be that in certain families the mucosa of the middle ear and the bony capsule of the labyrinth are congenitally weak, and therefore unable to resist infection from the surface. In the same way there seems to be evidence that in certain families cases of suppurative otitis media are specially liable to develop labyrinthine or intracranial complications. In cases of middle-ear suppuration several observers have noted changes in the bony capsule of the labyrinth similar to those seen in the early stages of otosclerosis—*i. e.* stage of otitis vasculosa or spongification.

Against the theory that otosclerosis is an inflammatory condition following otitis media we must place the facts that—(a) in most cases the mucosa of the middle ear is normal (it is well known, however, that otitis media may pass off and leave little or no trace); (b) plasma cells are absent (I am not sufficiently skilled in pathology to deal with this point, but it has been stated that plasma cells are not invariably present in inflammatory conditions); (c) areas of osteoporosis are found in spots far distant from the mucosa of the middle ear (this will be dealt with later).

(3) A third group of otologists look on otosclerosis as a primary disease of the bone, the infection coming by way of the blood-

stream, as in osteomalacia, rheumatism, etc. Ferreri regards otosclerosis as closely allied to osteomalacia, and finds that almost all patients suffering from the latter disease are also the subjects of otosclerosis. A variant of this view is held by A. A. Gray, who has put forward the theory that otosclerosis is an aseptic necrosis of certain areas in the labyrinth capsule—apparently as a result of aseptic infarction. Gray believes that similar areas occur in other bones of the skeleton. It would certainly be of great interest to examine other bones, though it would not be so easy in them to identify areas of *ostitis vasculosa* as it is in the dense labyrinth capsule. A grave objection to the theory that otosclerosis is a puerperal infection is the fact that it is very common in young unmarried women, while the cases so commonly met with in men must, of course, be explained in another way. If, for a moment, we consider theories (2) and (3) together, we may compare them with those put forward regarding the connection between tonsillitis and rheumatism. Formerly it was held that attacks of tonsillitis were of "rheumatic origin"—*i. e.* that the tonsil was presumably infected through the blood-stream. More recently opinion has turned round, so that it is now held that rheumatism is of "tonsillar origin"—that is to say, that the blood-stream becomes infected from the tonsils. In my opinion the number of cases of otosclerosis which are looked on as *secondary* to otitis media is likely in the future to grow larger, while the number of the so-called *primary* cases is likely to decrease.

(4) Lastly, otosclerosis is regarded by a small group of observers as *due* to a trophic disturbance—*i. e.* as a degenerative atrophic process. According to this theory the changes (sometimes) found in the ganglia and nerves are primary, while those in the labyrinth capsule are secondary. In this connection it may be noted that certain cases, which clinically appeared to be examples of *nerve deafness*, on microscopic examination showed spongification of the labyrinth capsule with (Siebenmann) or without (Bruehl) ankylosis of the stapes. This type of otosclerosis associated with nerve deafness—the so-called "atypical otosclerosis"—is markedly hereditary. Again spongification of the bone in front of the oval window has been observed in an old woman aged eighty-two (Wolff), and in two other cases in old people (Mayer). On the other hand, advanced cases of otosclerosis have been microscopically examined, in which no degenerative changes were found in the nerve structures.

FIG. 4, Section 195 ($\times 8$ diam).--Vertical transverse section through cochlea showing three areas of otitis vasculosa in the cochlear capsule. 1, Areas of otosclerosis. 2, Tensor tympani. 3, Cholesteatoma. 4, External meatus. 5, Tympanic membrane. 6, Lower part of tympanic cavity. 7, Apical coil of cochlea. 8, Basal coil of cochlea. 9, Area of otitis vasculosa.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

FIG. 5, Section 135 ($\times 8$ diam).—Vertical transverse section through capsule of cochlea showing a large area of otitis vasculosa in front of the hollow spaces of the cochlea. 1, Middle fossa. 2, Large area of otitis vasculosa. 3, Tensor tympani. 4, Anterior bony wall of external meatus. 5, Tubal part of the tympanic cavity. 6, Fatty marrow. 7, Fibrous marrow. 8, Normal cartilage bone (interglobular bone) of cochlear capsule. The section is made anterior to the hollow spaces

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

NATURE OF THE CHANGES IN THE BONE IN OTOSCLEROSIS.

According to Siebenmann the changes are as follows: (1) Vascular and fibrous marrow penetrates the normal Haversian canals and dilates them. (2) Next there is lacunar resorption by giant cells of the cartilage bone of the labyrinth capsule, resulting in the formation of large spaces containing blood-vessels and numerous cells. (3) New deeply staining bone is deposited in the walls of these large spaces (the bone takes on the basic hæmatoxylin stain). (4) The new bone becomes dense and gradually loses its affinity for the basic stain.

Manasse describes the changes thus: (1) Granulation and osteoid tissue are formed in the pre-existing spaces in the bone. This is the primary active process. The original bone of the labyrinth capsule disappears without absorption by giant cells. (2) The spongy new bone is destroyed by canaliculisation and lacunar resorption by osteoclasts; new marrow spaces are thus produced. (3) Bone is again formed by osteoblasts in the new marrow spaces; this bone becomes more and more compact by diminution in size of the marrow spaces. In this way new lamellar systems are formed, but the new compact bone is sharply marked off from the normal bone of the labyrinth capsule.

All writers on the pathology of otosclerosis take it for granted that the more deeply staining bone seen in the walls of the vascular spaces in the early stage of the disease is new-formed bone. In my experience, however, new-formed bone is usually pale and stains well with eosin, but not with basic stains. Such new bone may be seen in the walls of the mastoid air cells from cases of subacute mastoiditis. Accordingly, it seems probable that the bone which stains deeply with hæmalum or hæmatoxylin is not new bone at all, but merely the old bone of the region, the staining reaction of which has been altered by the change it has undergone owing to the vascular dilatation and the consequent increased supply of lymph. If we grant that in the stage of osteoporosis some irritant, inflammatory, or toxic condition be present, it will be obvious that in order to combat this condition the part needs an increased blood supply. The vessels of the Haversian spaces therefore dilate, the marrow proliferates, and the spaces become enlarged. It is not necessary that the bone should be removed by osteoclasts. Bone yields before long-continued pressure exerted by structures much softer than itself—*e. g.* the enlarged mastoid antrum and tympanic attic in cases of cholesteatoma, in the walls

of which I have never observed osteoclasts. Later on, when the local chronic inflammatory or toxic process has been overcome, the vascular supply diminishes and the bone again becomes sclerotic and loses its affinity for basic stains.

In support of the second (2) view as to the pathology of otosclerosis—namely, that it is a chronic inflammatory process invading the labyrinth capsule from the deep layer of the mucosa in the region of the anterior margin of the oval window—I bring forward the following case, the clinical notes of which are, unfortunately, very meagre :

J. H.—, male, aged nineteen, had suffered from chronic middle-ear suppuration of unknown origin for many years. The patient was a foundling, and nothing is known as to his parentage; he was a very dull boy and backward in every way, according to the account of his guardian. He was in and out of the poorhouse on several occasions, before his last admission on May 11, 1911. Death occurred on May 15, and at the autopsy a large temporo-sphenoidal abscess was discovered. As the patient had not been seen by an otologist, and as no functional examination had been carried out, it was not at first thought to be worth while to examine the ear microscopically. A vertical saw-cut was therefore made through the temporal bone from the external to the internal meatus in order to demonstrate cholesteatoma in the tympanic cavity and attic. As the specimen did not show this very well, it was determined to examine the ear microscopically in order to observe the condition of the labyrinth (cholesteatoma deafness?). In view of the fact that the labyrinth capsule showed extensive areas of *ostitis vasculosa*, it may readily be imagined that I was not too pleased with myself for having destroyed with the saw the regions of the oval and round windows. It is further unlucky that the plane of the saw-cut is not a favourable one for microscopic examination of the labyrinth. The results of the examination may be summarised as follows: The Eustachian tube shows normal epithelium, but the submucous tissue is thickened and shows cystic spaces (Fig. 4). The tympanic cavity, aditus and antrum are lined by cholesteatoma (Figs. 3 and 6). The bony walls of the attic and antrum show large spaces containing connective tissue and blood-vessels, but no giant cells (*ostitis vasculosa*?) (Fig. 6). On the inner wall of the tympanum, in the anterior margin of the oval window, there is an area of *ostitis vasculosa* in the labyrinth capsule (Fig. 1). If we start at this point we can trace the spread of the disease in the wall of the labyrinth between the periosteal and cartilage bone round to the region of the internal auditory meatus, and also over the apex of the cochlea to the anterior part of the bony capsule of this structure (Figs. 2 to 5). At the apical coil of the cochlea the spongification reaches the perilymph space (Fig. 3). Further back on the inner wall of the aditus and antrum, in the region where the vessels of the fossa subarcuata begin, the bone shows large spaces containing blood-vessels and connective tissue (Fig. 6). The marrow surrounding the labyrinth capsule appears at all parts to be very fibrous (Figs. 3 and 4). Beyond this the marrow is fatty, but cellular marrow is entirely absent.

In the case of specimens obtained from the human subject it is never wise to be dogmatic in regard to changes in the delicate nerve structures of the labyrinth, even in cases (such as the present) where the temporal bone was obtained within twenty-four hours of death and at once placed in fixative. With this proviso it may be stated that Corti's organ appears to be atrophied in all coils; only the out-

FIG. 6, Section 445 ($\times 8$ diam.).—Vertical section through the posterior part of the labyrinth in the region of the semicircular canal. 1, Superior canal. 2, Area of otitis vasculosa? passing in towards fossa subarcuata. 3, Floor of middle fossa. 4, Cholesteatoma in antrum. 5, External canal. 6, Posterior canal towards the ampullary end. The canal here contains bone dust (artefact). 7, Saccus endolymphaticus. 8, Crus commune. 9, Posterior cranial fossa. Note change in the bone (ostitis vasculosa) passing inwards from the cholesteatoma matrix on inner wall of antrum. In this situation the veins from the inner wall of the antrum pass through the fossa subarcuata to the posterior cranial fossa to join the inferior petrosal sinus. The condition seen in the photomicrograph is, however, not found under normal conditions.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

FIG. 7.—Showing lymph or marrow space in the labyrinth capsule, between the cartilage bone (right) and the lamellar bone (left) (a probable path of infection). $\times 60$ diam.

FIG. 8.—Showing area of spongification in the cochlear capsule. This area is sharply defined and lies between the cartilage bone (below and to the right) and the lamellar bone (above and to the left). $\times 60$ diam.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

Adlard & West Newman,

line of this structure is visible. The cells of the spiral ganglion also appear to be shrunk, and their place is taken by connective tissue. The vestibule, along with the ampullary ends of the canals, were destroyed by the saw-cut.

I have found changes, somewhat similar to those seen in the labyrinth capsule of this case, in the malleus and incus obtained from cases of chronic middle-ear suppuration in which the ossicles were diseased (Figs. 12 and 14).

In order to explain how the chronic infective condition, which invades the labyrinth capsule from the region of the promontory, can reach the internal auditory meatus it is necessary to recall a few facts regarding the development of the ear. About the third month of foetal life we find the otic vesicle, now differentiated into cochlea, saccule, utricle, and canals, surrounded by a capsule of cartilage of mesoblastic origin (Fig. 15). At this stage the perilymph space is still filled with foetal connective tissue. Further, the hollow bud of mucosa from the naso-pharynx which lines the Eustachian tube, and is later on to line the tympanum, aditus, antrum, and mastoid cells, has as yet only reached the anterior and lower part of the tympanic cavity. Later on, the cartilage capsule of the inner ear mentioned above becomes converted into dense, non-vascular bone (Fig. 19). This bone throughout life contains numerous spaces (interglobular spaces) which hold cartilage cells even in old people.

In the child and adult the membranous labyrinth is surrounded by three distinct layers of bone (Fig. 20): (1) Just outside the endosteum, which lines the labyrinth, there is a very dense layer of compact bone formed by the outer (osteogenic) surface of the endosteum (Fig. 21, No. 6); this layer is, later on, intimately fused with (2) the dense bone derived from the cartilaginous capsule of the labyrinth referred to above (Fig. 21, No. 7); (3) surrounding this cartilage bone, but distinct from it, we have the lamellar bone with its cellular, fatty, or fibrous marrow (Fig. 21, No. 3). This lamellar bone, where it lies next to the middle ear, is formed by the deep layer of the tympanic mucosa and, where it lies next to the cavity of the skull, by the osteogenic layer of the dura mater (Figs. 17, 18 and 19).

Although I have not noticed it in the communications of other observers I have found that, in the foetus, there is a well-marked space—probably a lymph or marrow space—between (2) the cartilage bone and (3) the lamellar bone (Fig. 17, Nos. 8 and 9). In young children this space is, as a rule, clearly visible though not continuous, but in later life it is not so well seen, though traces of it may still be found, especially in the capsule of the cochlea. In

any case a "potential" space is always present, just as between two pieces of wood which have been glued together.

It is my belief that a chronic infective process, such as a pyogenic osteomyelitis, a syphilitic or tubercular osteomyelitis, or the condition known as otitis vasculosa, may invade the mucoperiosteum of the inner tympanic wall, infect the marrow spaces of the lamellar bone, and finally invade the lymph or marrow space described above. When once this has been opened up the chronic infective process may spread round the cochlea between the lamellar and cartilage bone, and even progress as far as the internal auditory meatus. This theory meets the objection raised by Politzer—that otosclerosis cannot be secondary to an infection of the tympanic mucosa, because the otosclerotic change has been found in the region of the internal meatus, where there is no mucous membrane connected with the middle-ear spaces.

THE CAUSE OF DEAFNESS IN OTOSCLEROSIS.

(1) *Ankylosis of the Stapes*.—In the cases in which ankylosis is present it is obvious that the cause of deafness is not far to seek. This has led Jenkins and Bárány, independently of one another, in recent times, to expose the endosteum of the labyrinth in the region of the external and posterior canals respectively, *i. e.* to make what has been called by some a "labyrinth fistula." As a matter of fact these otologists did not make a fistula at all, as this would have necessitated opening the lymphatic spaces of the labyrinth. What they did was to provide an artificial window to replace the ankylosed oval one on the assumption that the membrane of the round window was still mobile, and that, the labyrinthine fluid being incompressible, it was necessary to have two movable windows in order to enable vibrations to occur in the labyrinthine fluid. In the cases recorded by both the otologists mentioned above the immediate effect of the operative procedures was extremely gratifying, but within a comparatively short time the hearing again deteriorated. It must be admitted that there are cases of otosclerosis on record in which marked deafness was present, although microscopic examination showed that the stapes was not ankylosed.

(2) *Changes in the Labyrinthine Fluid*, especially in cases in which the bone disease reaches the endosteum of the labyrinth. Jenkins lays great stress on the probability of a change occurring in the composition of the labyrinthine fluid in cases of otosclerosis,

FIG. 10.—Showing spongification of bone in the wall of mastoid antrum. $\times 60$ diam.

FIG. 9.—Showing area of spongification of apical coil. Both the lamellar and cartilage bones are involved, the change extends completely through these layers, and at one point reaches to the endosteum of the apical coil. $\times 60$ diam.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

NORMAL AND DISEASED OSSICLES FROM CASES OF CHRONIC MIDDLE-EAR SUPPURATION. (Figs. 11 to 14 inclusive.)

FIG. 11.—Vertical transverse section of normal malleus. $\times 15$ diam. 1, Head of malleus. 2, Articular surface. 3, Handle. 4, Short process.

FIG. 12.—Eroded malleus from a case of chronic middle-ear suppuration and cholesteatoma. $\times 15$ diam. 1, Eroded head of malleus. 2, Cholesteatoma. 3, Handle of malleus. 4, Short process.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

and holds that this may account not only for the deafness, but also for the paracusis.

(3) *Degeneration of the Cochlear Ganglion, Nerve, and Corti's Organ.*—The microscopic examination of the inner ear involves such a long and complicated process that in the case of the human ears it is never safe to be dogmatic in regard to the finer changes which may appear to be present in the cells of the spiral ganglion, in the fibres of the cochlear nerve, or in the sensory and supporting cells of Corti's organ, even in those instances in which the temporal bone is obtained within twenty-four hours of death and immediately placed in a suitable fixative solution after the hollow spaces of the labyrinth have been opened. In animals the fixative (Zenker's solution) can be perfused through the vessels before death, or the temporal bones can be at once removed and placed in the warm solution, so that we are able to speak with confidence regarding the finer changes in the nerve structures. In examining human ears, however, we must be very sceptical in attributing importance to slight changes in the nerve structures, such as shrinking of the ganglion cells, fibrillation of the nerves, or slight flattening of the acoustic papilla. In some cases of otosclerosis, in which deafness was present during life, microscopic examination of the inner ear has shown that the nerve structures were well preserved. Here we may conclude with a fair degree of certainty that the cause of the deafness was not degeneration of the sound-perceiving apparatus. Such cases have been recorded by several writers. It is usually held that the nerve changes in otosclerosis are secondary to the bony changes, and that they are probably due to a toxin. If this is so the condition would be analogous to that observed by D. Orr and others in the spinal cord and nerves in relation to toxic processes in the bladder, kidneys, etc.

(4) *Spongification of the Labyrinth Capsule.*—Under normal conditions the sensitive structures of the membranous labyrinth are surrounded by two layers of dense non-vascular bone, the endosteal and cartilage bone referred to above. It is not easy to understand how the cartilage bone receives its nourishment, but, as it is not an active structure, it probably requires little, and this can be obtained from the lymph which finds its way into the interglobular spaces with which the bone is so plentifully supplied. The complicated course of the vertebral arteries and the arrangement of the circulation at the base of the brain seem especially suitable to provide an even flow of blood to the delicate structures at the inner ear. Politzer maintains that an anastomosis occurs

between the labyrinthine vessels and those of the middle ear in the neighbourhood of the oval window. In otosclerosis, on the other hand, the character of the bone forming the labyrinth capsule is altered and vascular spongy bone replaces the dense non-vascular bone over more or less considerable areas. If, as most authorities hold, it is necessary for perfect hearing that the membranous labyrinth should be surrounded by dense non-vascular bone, it is easy to understand that in otosclerosis the hearing power is greatly diminished. The writer is of opinion that this last theory (4) probably provides the true explanation of the deafness in typical cases of otosclerosis. As the patients say themselves, they could hear well enough if it were not for "the noises." In atypical cases of the disease in which the nerve structures are degenerated the deafness can be accounted for by the lesion of the sound-perceiving apparatus.

THE CAUSATION OF TINNITUS.

Much of what has already been said with regard to the causation of deafness applies also to tinnitus. *Vascular changes*: The increase in the vascular anastomosis between the vessels of the internal and middle ear may to some extent explain the noises from which the patient suffers—noises which are worse when the patient lies down at night. Again, the presence of vascular spongy bone in the cochlear capsule is likely to result in tinnitus. *Nerve changes*: Degeneration of the nerve structures has been put forward, especially by Neumann, as one cause of noises in the ear. This change Neumann calls "neuronophagia." If the cells of the cochlear ganglion merely atrophy, there is no tinnitus; but if, in the process of degeneration, there is a preliminary stage of irritation, tinnitus will arise. *Labyrinthine fluids*: It is not so easy to account for tinnitus by a change in the labyrinthine fluid, though it is possible that such a change might affect the nerve structures or might favour the conduction of vascular noises. It is interesting in this connection to remember that in cases of chronic progressive middle-ear deafness an intercurrent attack of acute otitis media may greatly improve the hearing. The writer once knew an otologist who was so impressed by this fact that he proposed to treat such cases by infecting the tympanic cavity with pyogenic organisms!

PARACUSIS.

In the writer's experience paracusis is not by any means present in all cases of otosclerosis.

FIG. 13.—Vertical transverse section of normal incus. $\times 15$ diam. 1, Body of incus. 2, Articular surface (with blood clot). 3, Long process. 4, Short process.

FIG. 14.—Eroded incus from a case of chronic middle ear suppuration. $\times 15$ diam. 1, Body of incus. 2, Articular surface. 3, Eroded long process. 4, Invasion of the marrow spaces by a chronic form of osteomyelitis analogous to that seen in the labyrinth capsule in otosclerosis and congenital syphilitic deafness. 5, Short process of incus. Note large marrow space with dilated vessels extending from region of eroded long process.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

JOURNAL OF LARYNGOLOGY, RHINOLOGY, AND OTOTOLOGY.

DEVELOPMENT OF THE LABYRINTH CAPSULE STUDIED IN RELATION TO THE
PATHOLOGY OF OTOSCLEROSIS. (Figs. 15 to 21 inclusive.)

FIG. 15.—Vertical transverse section through head of foetal pig ($\times 8$ diam.), showing the foetal cartilaginous capsule of the labyrinth on both sides. The endolymph spaces are patent, but the perilymph spaces are still filled with foetal connective tissue. 1, Footplate of stapes. 2, Facial nerve. 3, Incus. 4, Malleus. 5, Eustachian tube. 6, Naso-pharynx. 7, Mouth cavity. 8, Hyoid. 9, Meckel's cartilage. 10, Lower jaw. 11, External auditory meatus. 12, External canal. 13, Hollow of auricle. 14, Superior canal. 15, Utricle. 16, Saccule. 17, Cochlear nerve. Note the cartilage capsule of the labyrinth extending from above 14 to the basiphosphoid which lies above 6.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY
OF OTOSCLEROSIS.

(a) Some writers have supposed that *stimulation by noise is necessary to excite the atrophied nerve apparatus*. It has been suggested that in the early stages, before ankylosis occurs, the reflex effect of external noise (acting through the stapedius muscle) alters the tension of the labyrinthine fluid so as to put it into the best condition for the perception of sound. According to this view, one would expect that paracusis would be present in the early stages of the disease, but would pass off later on. The writer has met with several cases in which this history was obtained. Urbantschitsch holds that paracusis is due to a raised functional capacity of the acoustic nerve apparatus.

(b) It is well known that Politzer was of opinion that paracusis could be explained by the massage effect of the external noise or vibration on the more or less stiffened joints between the ossicles. If cases of chronic adhesive process in the middle ear alone presented the symptom of paracusis it might be possible to agree with this theory, but paracusis is a much more prominent symptom in otosclerosis than in the so-called chronic middle-ear catarrh (chronic adhesive process). Further, if Politzer's theory were correct, one would expect that the improvement in hearing would continue for some time after the external vibrations had ceased. As far as I know, this is not the case.

(c) Paracusis has been attributed by Jenkins to a change in the labyrinthine fluid. It appears to the writer that the composition of the labyrinthine fluid, at least of the perilymph, must closely correspond with that of the cerebro-spinal fluid. It might be possible to settle this point by chemical examination of the cerebro-spinal fluid from cases of otosclerosis. Babinski has reported numerous cases in which tinnitus was markedly improved by lumbar puncture.

(d) The view has been put forward, by Jenkins and others, that the paracusis is more apparent than real, *i. e.* that the patients do not really hear better in a noise, but are able to catch the raised voice of their companion in noisy surroundings such as those of a railway carriage because they are less disturbed than normal people by those noisy surroundings. It is, of course, widely known that low tones are badly, or not at all, heard by patients suffering from otosclerosis, and according to this theory (*d*) it is assumed that the external sounds which so much disturb normal people are of low pitch, and consequently more or less inaudible to otosclerotics. It certainly appears to be true from

the writer's experience that married women suffering from otosclerosis are better able to hear the high-pitched voices of their children than the deeper tones of their husbands. It should not be a difficult matter to test this last theory of paracusis, for if it were true an otosclerotic should not hear any better in noisy surroundings if the noise were of high pitch. It must, of course, not be forgotten that in noisy surroundings people naturally raise their voices to a considerable extent.

(e) McBean (*Annals of Otology*, December, 1915) advances what appears to be a new theory to explain paracusis. The points in McBean's argument appear to be as follows: (1) The nerve endings in the cochlear and vestibular apparatus are very similar. The tectorial membrane and Corti's organ in the cochlea correspond to the cupula and crista in the canals. (2) Movements of the fluid are constantly occurring in the canals, and keep the end organs of the vestibular apparatus in a suitable condition for appreciating passive movement and rotation. (3) Under normal conditions the ossicular chain and the column of perilymph in the scala vestibuli and scala tympani move backward and forward with each sound vibration, the round window membrane bulging out when the stapedial foot-plate moves inwards. (4) Sound vibrations—especially those of low tone—are practically always present, so that the normal relation of the tectorial membrane to Corti's organ must be that of constant simultaneous motion. These movements keep the nerve terminals in an efficient state. (5) When the stapedo-vestibular joint is ankylosed the sound-conducting mechanism is thrown out of gear, low tones are not transmitted, and movements of the fluid in the cochlea are interfered with. (6) The membrane of the round window is capable of performing excursions five times greater than those of the foot-plate of the stapes. In the normal ear the round window membrane plays a passive rôle, but, when the stapedial foot-plate is more or less fixed, the membrane of the round window takes on a more active function. A greater stimulus, however, is needed, and is provided by the vibrations of machinery or a rail-road train. Under such conditions the membrane of the round window and the cochlear lymph move to and fro with something like their normal motion, and at the same time the hair cells attached to the tectorial membrane can resume their function as receivers of sound. (7) If the above theory is accurate, McBean would expect that in those cases of otosclerosis in which the round window is obliterated by new bone formation, paracusis would not be present.

FIG. 16, Vertical transverse section ($\times 8$ diam.) through head of fetal pig at later stage than preceding photomicrograph (No. 15). The section corresponds to the left half of Fig. 7, but lies horizontally instead of vertically. The perilymph spaces of the vestibule and canals are now patent, but in the cochlea the perilymph space is still filled by fetal connective tissue. 1, Superior canal. 2, Vessels of fossa subarcuata. 3, Utricle. 4, Cochlear nerve. 5, Saccule. 6, Scala tympani still solid. 7, Cochlear canal. 8, Scala vestibuli still solid. 9, Eustachian tube. 10, Mouth cavity. 11, Lower jaw. 12, Tympanic cavity only reaches malleus and anterior margin of oval window. 13, External auditory meatus. 14, External semicircular canal. 15, Auricle. 16, Facial nerve. The posterior and upper parts of the tympanic cavity are still filled with fetal connective tissue. The perilymph space of the vestibule and canals is now patent, though that of the cochlea is still filled with fetal connective tissue.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

FIG. 17.—Horizontal section through left ear of seven months foetus in region of oval window. The case was said to be one of congenital syphilis, but the Wassermann reaction (as done *post-mortem*) was negative. The section shows the arrangement of the bone around the hollow spaces of the inner ear. 1. Facial nerve. 2. Long process of incus, below this the niche of the oval window contains mucoid exudate. 3. Chorda tympani. 4. Handle of malleus. 5. Tympanic membrane. 6. Lamellar bone formed by deep layer of muco-periosteum covering promontory. 7. Internal carotid artery. 8. Lamellar bone of cochlear capsule. 9. Cartilage bone of cochlear capsule: note marrow or lymph space between 8 and 9. 10. Endosteal bone of basal coil of cochlea. 11 and 13. Lamellar bone formed by osteogenic layer of dura mater. 12. Internal auditory meatus with nerves. 14. Fossa subarcuata. 15. Smooth end of superior canal. 16. External canal surrounded by its layer of endosteal and cartilage bone. The neuroepithelium of the labyrinth shows marked post-mortem changes.

PART II. (R. M.)

The microscopical appearances seen in the study of sections from cases of otosclerosis strongly suggest the structural changes one meets with in other bony tissues in conditions of chronic toxæmia or altered metabolism. Whatever be the exciting cause of this condition we have a series of progressive changes which appear to be *purely inflammatory* in character. The ultimate result is a typical and constant lesion, viz. marked sclerosis of the bony tissue of the labyrinth capsule. The alleged absence of plasma cells does not prove that otosclerosis is of non-inflammatory origin. These cells are not present in all forms of chronic inflammation. Even when they are present they do not occur in all stages of the inflammatory process. For their detection special staining methods are required—methods difficult to apply to celloidin sections of the labyrinth.

The first change one observes in otosclerosis is the marked and general engorgement of the blood-vessels both in the fatty marrow of the bone and in the muco-periosteum. The connective tissue of these areas is also seen to increase in amount, having a loose, open, and fibrillated structure. There are more cellular areas, mainly composed of mononuclear cells, which are closely related to the congested vessels. Some of the medullary spaces in the petrous bone show leucoblastic marrow reaction. Such appearances can be explained by a toxic body brought by the blood or invading the bone from the muco-periosteum, and acting as a stimulant, especially on the connective tissues of the marrow. The condition is essentially a granulating process, which slowly brings about absorption of the bone, allowing the opening up of the medullary spaces. The deeply staining bone seen around the enlarged medullary spaces appears to be old bone which is undergoing change, and not new-formed bone, as described by various writers. The granulation tissue tends to become more fully formed fibrous tissue, and these fibres arrange themselves in concentric layers around the central vessel. Ossification of this fibrous tissue now sets in, beginning with the fibres lying in close relation to the bone trabeculæ, thus bringing about a marked thickening of the lamellæ at the expense of the Haversian canals. The inner layer of bone in the labyrinth capsule also appears to be involved in this process of sclerosis. There is an active proliferation of the cartilage cells and ossification into a bony matrix.

The above changes are seen to be of a patchy character and appear to begin at the junction of the cartilaginous and lamellar portions of the labyrinth capsule, and to spread directly along that line for some distance, extending more or less into the cartilaginous and lamellar bone on each side, and in places passing right through the cartilage bone to reach the endosteal tissue lining the cavities of the inner ear. This path of extension strongly suggests some direct channel, such as that shown to exist (J. S. F.), and wov'd explain the areas of change seen in the various parts of the labyrinth capsule.

The whole process appears to be a slow form of inflammation, a definite series of reactions of a special tissue to repair damage caused by some unknown element, bacterial or toxic.

Conclusions.

(1) Some cases of otosclerosis (spongification of the labyrinth capsule) appear to be of the nature of a blood infection, and may be called "primary." We would expect that further research would show that in these cases the bony changes are widespread throughout the skeleton.

(2) Other cases of otosclerosis undoubtedly follow attacks of catarrhal or purulent otitis media, the infective process invading the labyrinth capsule at the anterior margin of the oval window. Such cases may be called "secondary."

(3) From the clinical standpoint, heredity plays an important part in otosclerosis, but, as the condition is a chronic inflammatory process, the infective agent must gain access to the labyrinth capsule either through the blood-stream or from the middle-ear cleft.

(4) It has been proved by clinical and microscopical research that "atypical" cases of otosclerosis exist in which functional examination of the ear reveals the presence of nerve deafness. The relationship between the bony changes and the nervous affection is not yet clear.

CHIEF REFERENCES WITH REGARD TO THE PATHOLOGY OF OTOSCLEROSIS.

ALEXANDER.—*Arch fur Ohrenheilk*, vol. lxxviii, pp. 54 and 128.

BECK.—*Monatschr. fur Ohrenheilk*, 1910, p. 28.

BEZOLD.—*Zeitschr. fur Ohrenheilk*, 1893, vol. xxiv, p. 267; 1894, vol. xxvi, pp. 1-10.

BREUHL.—*Ibid.*, vol. l, p. 276; *Passow's Beiträge*, vol. iv, 1910, p. 71.

BRYANT.—*JOURN. OF LARYNGOL., RHINOL., AND OTOL.*, 1915.

FIG. 18.—Horizontal section through left ear of seven months foetus in region of round window. 1. Facial nerve. 2. Membrane of round window. 3. Tympanic membrane. 4. Lamellar bone of promontory formed by deep layer of muco-periosteum. 5. Anterior part of annulus tympanicus. 6. Internal carotid artery. 7. Endosteal bone of basal coil of cochlea. 8. Space between lamellar and cartilage bone, the latter is very thin at this point. 9. Scala tympani of basal coil of cochlea. 10. Internal auditory meatus with dural sheath. 11. Opening of crus commune into vestibule. 12. Opening of smooth end of external canal into vestibule. 13. Dense cartilage bone of external canal. 14. Marrow. 15. Smooth end of posterior canal surrounded by endosteal and cartilage bone. 16. Sinus tympani containing mucoid exudate.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

Adlard & West Newman,

FIG. 19.—Horizontal section through right ear of newborn child ($\times 8$ diam.), showing lymph space (3) between cartilage bone (2) and lamellar bone (4) in the capsule of the cochlea. 1, Carotid canal with artery. 2, Cartilage bone of cochlear capsule. 3, Lymph space. 4, Lamellar bone formed from deep layer of tympanic mucosa. 5, Anterior margin of oval window. 6, Malleus. 7, Footplate of stapes. 8, Incus. 9, Facial nerve. 10, External semicircular canal. 11, Cartilage bone surrounding canal. 12, Marrow. 13, Smooth end of superior canal. 14, Fossa subarcuata. 15, Vestibule. 16, Internal auditory meatus with nerves. 17, Lymph space in bony capsule of cochlea.

TO ILLUSTRATE MR. J. S. FRASER'S AND MR. R. MUIR'S ARTICLE ON THE PATHOLOGY OF OTOSCLEROSIS.

- CORNET.—*Ibid.*, 1909, p. 540.
 DENKER.—“Die Otosklerose,” Wiesbaden, 1904.
 FERRERI.—*Arch. Inter. de Laryngol., d'Otol., etc.*, vol. xxxiii, No. 1, p. 1.
 FREY.—*Arch. fur Ohrenheilk.*, vol. lxiii, p. 12.
 FRÖSCHELS.—*Passow's Beiträge*, vol. v, p. 199.
 GRAY.—“The Ear and its Diseases,” New York, Wm. Wood & Co., 1910, Chap. 14, “Otosclerosis,” pp. 311-329.
 GRUNERT.—*Arch. fur Ohrenheilk.*, 1903, vol. lx, p. 161.
 HABERMANN.—*Ibid.*, vol. l, p. 242; vol. liii, p. 52; vol. lx, p. 37.
 HAMMERSCHLAG.—*Monatschr. fur Ohrenheilk.*, 1910, pp. 762-773, 709.
 HARTMANN.—*Zeitschr. fur Ohrenheilk.*, 1898, vol. xxxiii, No. 3.
 HEIMAN.—*Monatschr. fur Ohrenheilk.*, 1909, p. 761.
 KALENDA.—*Zeitschr. fur Ohrenheilk.*, 1910, p. 299.
 KATZ.—*Archiv fur Ohrenheilk.*, 1906, p. 68; vol. lxviii, p. 122.
 KOERNER.—*Zeitschr. fur Ohrenheilk.*, vol. l, p. 98.
 LUCAS.—“Die chronische progressive Schwerhörigkeit,” Berlin, 1907.
 MANASSE.—“Die Otitis chronica metaplastica der menschlichen Labyrinthkapsel,” Wiesbaden, 1912.
 MARKMANN.—*Zeitschr. fur Ohrenheilk.*, vol. lvi, p. 85.
 MAYER.—*Monatschr. fur Ohrenheilk.*, 1911, p. 257.
 MÖLLER.—*Ibid.*, 1905, vol. xxxix, p. 125.
 MAYER AND YOSHÜ.—*Zeitschr. fur Ohrenheilk.*, vol. lx, p. 93.
 PALUDETTI.—*Centralblatt fur Ohrenheilk.*, 1912, x, No. 4, p. 151.
 PANSE.—*Archiv fur Ohrenheilk.*, 1903, vol. lix, pp. 84-98.
 POLITZER.—“Diseases of the Ear,” Phila. and New York, 1909; *Zeitschr. fur Ohrenheilk.*, vol. xxv, p. 309; *Monatschr. fur Ohrenheilk.*, 1908, p. 419.
 SCHEIBE.—*Arch. Intern. de Laryngol., d'Otol.*, 1911, vol. xxxii, No. 2.
 SIEBENMANN.—*Zeitschr. fur Ohrenheilk.*, vol. xxxiv, p. 356; vol. xxxvi, p. 291;
 “Report of Ninth International Otological Congress,” Boston, 1912, pp. 207-212.
 TANIYAMA.—*Zeitschr. fur Ohrenheilk.*, 1911, vol. 9, No. 11-12, p. 560.

CLINICAL NOTES.

THE INFLUENCE OF POSITION ON THE APPEARANCES OF THE NORMAL PHARYNX.

DOUGLAS GUTHRIE, M.D., F.R.C.S.

SOME years ago, whilst examining the throat of a bedridden patient, I noticed a swelling on the right side of the posterior pharyngeal wall, the appearance in fact strongly suggesting that of a retropharyngeal abscess.

The patient's head was, at the time, acutely turned towards me as I stood to the right of his bed. Examination in the ordinary position, however, convinced me that the pharynx was normal and that the peculiar appearance I had observed was solely dependent upon my patient's unusual attitude.

This observation may be verified by anyone with great ease. Let any patient be seated sideways, as though about to have the right ear examined. Then direct him to turn his head so that he looks over his