OTOSCLEROSIS.

A Clinical Analysis.

By DAN McKENZIE, London.

THE occasion that has given rise to this paper is the questionnaire which was circulated a few months ago by the Council of the Otological Section of the Royal Society of Medicine, to aid J. S. Fraser in the preparation of his Rapport introducing the discussion on Otosclerosis at the International Congress on Oto-Rhino-Laryngology at Madrid in 1932. In order to answer the questions put to us therein I turned to my private records, which date back to 1908, and gathered together the accumulated data of over 600 cases.

I have already sent to Dr. Fraser most of the statistical and, so to speak, impersonal items in the records, but while I was working on them I became aware of a number of conclusions and speculations that would have been out of place in any formal return, and yet it seemed to me that it would be a mistake to let the opportunity pass without publishing them, ranging as they do over the entire field, and presenting at least some food for thought.

Nomenclature.—One of the questions put to us carries the suggestion that otosclerosis is being more frequently diagnosed nowadays than it used to be, say twenty years ago. With this I agree. But what has happened is, not that otosclerosis is becoming any more common, but only that that name is being more and more substituted for the old one of "chronic catarrh of the middle ear." And I think that the substitution is a fit and proper change, for reasons which will be amply manifest, I hope, in what is about to follow.

The pathologist, hearing us use such a loose and illogical word as "otosclerosis," may well feel irritated; but its looseness and illogicality are exactly the qualities that commend it to the clinician, since the word connotes a symptom-group that, as we shall see, can be evoked by quite a number of different pathological causes inducing one particular physical change in the ear.

Pathology.—To begin with, the word was applied to a definite pathological change which a microscopist, I suppose,

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might describe as an "insular" or "discrete osteoporosis of the labyrinth capsule." Now, as we know, the bone-changes we are calling "osteoporosis" occur so frequently without any indication of inflammation, past or present, in the mucosa of the middle ear, that this was for long regarded as the only form of the disease. But J. S. Fraser has shown us that the same bone changes are also occasionally associated with purulent otitis media. Further, as we shall see in a moment, the otosclerosis syndrome may be evoked by scar-tissue formation in the middle ear without any osteoporosis. Pathology, therefore, may be cited in our support if we divide our cases as we meet them into two main groups: first of all, those in which the disease-process seems to be independent of otitis media in any form, and this we may call primary otosclerosis; and, secondly, those in which it is associated with and is probably induced by some form of otitis media, and this we may call secondary otosclerosis.

The next preliminary we have to consider is the correlation of the pathological changes to the functional impairment as experienced by the patient in his deafness, and by the otologist in the results of his hearing-tests.

It is generally agreed that the clinical phenomena are the consequence of a gradually progressing limitation of movement of the stapes in the stapedio-vestibular articulation, ending typically in immobilisation. But it has also been pointed out that a fixation of the membrane of the round window, as by hyperostosis or a plug of scar-tissue, without involvement of the stapes, would have the same effect.

(This last I can corroborate from direct, or almost direct, observation. A patient had the typical symptoms and signs of pronounced otosclerosis. The membrana tympani, malleus, and incus of one ear had been destroyed and the stapes could be seen and touched with a fine probe. It was freely movable.)

The pathological changes that induce interference with the stapes or round window membrane, or both, are, of course, well enough known. Movements can be impeded or arrested either by fibrous bands or by hyperostosis, or by both. The stapedio-vestibular articulation, that is to say, like an articulation anywhere else, may become ankylosed either by fibrous or by bony adhesions, or first by the one and then by the other. And in the middle ear, the small size of the structures and the minute range of their movement exposes them to inter-

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ference by changes that would have no effect upon the (relatively) gigantic articulations elsewhere in the body. Thus, minute threads or plaques of scar-tissue, inconspicuous outgrowths of bone as from the osteoporosis aforesaid, even, presumably, the apparently trifling fibrosis that may follow the milder chronic catarrhs of the middle ear, will impede and, as time passes, may ultimately arrest all molar movements of this almost microscopic ossicle. The causes, therefore, that may lead to such limitation and cessation of movement must be many, various, and readily forthcoming. But, speaking broadly, the limitation and arrest are brought about either by scar-tissue alone, or by osteoporosis alone, or by both together.

Thus, from the clinician's point of view, the all-important event is the progressive immobilisation of the stapes and/or of the membrane of the round window, from whatever cause arising.

As can be read in the literature, the older school of otologists laid great stress upon the tympanic membrane and the members of the chain of ossicles as a whole, while we later observers, influenced by the excellent hearing that sometimes follows the radical mastoid operation, have come to regard the stapedio-vestibular articulation as the most important link in the chain that connects the outer world of sound with the receptive organ in the cochlea.

I have heard G. J. Jenkins criticise the phrase "middle ear deafness" as applied to progressive obstructive deafness, and I have come to agree with him. That appellation lacks precision. Strictly speaking, as we have seen, the middle ear may appear to be quite normal. Consequently, the term "fenestral deafness" would be preferable.

The Bezold Triad.—Clinically, otosclerosis is held to be characterised, among other things, by the responses to the hearing tests that are summed up in what is called the Bezold triad: (1) Lengthened bone-conduction; (2) Raised lower tone-limit; and (3) Negative Rinne.

X-Ray Examination.—We shall have something pertinent to say regarding each of those three phenomena later on, but, before passing on to do so, I wish to remark here that in the questionnaire it is asked whether cases of otosclerosis can be recognisable clinically as such when the responses to the tests are those of pure nerve-deafness—when, that is to say, the osteoporosis has affected the cochlea but not the fenestrae.

Obviously, while we know that such cases do occur and

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while we may in certain instances have definite suspicions, it is impossible—or nearly so—to make a confident diagnosis of osteoporosis from such responses alone, since there are so many forms of nerve-deafness in which the cause is hidden.

(The only instance in which I ever ventured to make such a diagnosis was in a woman nearly equally deaf in both ears, in one of which the usual Bezold triad was present, while in the other the bone-conduction was reduced and the Rinne positive).

If, however, we were able to detect osteoporosis by means other than by the tuning-fork tests, then we could certainly distinguish nerve-deafness arising from this cause.

Now, it has been claimed by H. K. Graham Hodgson that radiography enables us to do so. This, if substantiated, will prove to be an advance in diagnostic methods of prime importance. I may say that I can see the changes that Graham Hodgson interprets as the signs of osteoporosis only when they are pronounced, for then the haziness of the labyrinth shadow is plain enough. But the subtler changes he can detect I am not skilled enough to discern.

Nine of my cases have been X-rayed by him, with a positive result in seven. The two negatives were cases of nerve-deafness.

In this investigation, therefore, I have thought it best to ignore cases of nerve-deafness suspected of being otosclerosis, so as to avoid, as far as possible, any error in my statistics arising from mistaken diagnosis.

Primary Otosclerosis.—This class, which is the largest of my divisions, is made up of cases in which the typical deafness is present without any evidence of catarrhal or suppurative disease in the ear.

In order to make sure that I was dealing throughout with definite otosclerosis I rejected both from this division and from the next (secondary otosclerosis) all cases that did not manifest well-marked deafness, the rule being inability to hear the spoken word at a distance of more than three feet.

Secondary Otosclerosis.—Here were placed cases of progressive obstructive deafness supervening upon suppuration of the middle ear (active or quiescent), and upon manifest catarrh of the middle ear (without meatal discharge). Both of those conditions, of course, of themselves cause deafness; therefore, only when the deafness was recognised to be progressing, cochlear involvement being absent, was a case considered to be valid for our purpose.

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But in addition to those two acknowledged clinical divisions I have also made a third, namely, **Incipient Otosclerosis**, in which, suppuration and catarrh being absent, the deafness is trifling, although of long standing and uninfluenced by treatment. We shall refer further and in detail to this class in a later section.

The reader, I am sure, will at once remark that in many cases it must be difficult to make a clean distinction between primary otosclerosis and that form of secondary otosclerosis that is due to chronic catarrh. And I could not refute that objection *à priori*. All I can say is that, as we shall see when we come to deal with the details, the errors of classification that I have made must be few, since in one or two items, notably in the ages of onset, the three classes show very decided differences from each other.

At the same time, if what we have been saying of otosclerosis is correct and true to Nature, namely, that whether primary, or secondary, or incipient, the symptoms and signs are in all three due to the limitation of stapedial movement and, later, to the fixation of the stapes, then we ought to find that the clinical phenomena in all three varieties, as manifested in the returns of a large number of cases, will show some resemblance to each other.

This I may say, without further ado, is actually what has emerged. In addition to the differences mentioned above, not only is there a resemblance, but also, very much to my own surprise, the resemblance is, in many items, so close as almost to justify the use of the epithet "identical."

As proof of this statement the reader is referred to the subjoined numbers and percentages obtained by an analysis of the cases dealt with.

Sex-Distribution—622 cases in all were available.

Primary Otosclerosis. (Total, 411 cases.)

Males . . .	130 (31 per cent.)
Females . .	281 (69 „)

Secondary Otosclerosis. (Total, 129 cases.)

Males . . .	52 (40 per cent.)
Females . .	77 (60 „)

Incipient Otosclerosis. (Total, 82 cases.)

Males . . .	32 (39 per cent.)
Females . .	50 (61 „)

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Herein all three classes are alike, and the agreement in percentages is remarkably close. The result tallies with that of all other observers. We shall be returning to the sex-question and its possible meaning later on.

Age at Onset.—The next point we shall consider is that of the age at the first appearance of the deafness. This I was able to estimate in 528 cases of all classes. But it is necessary to utter the warning that errors are almost inevitable in this department. People forget. And when they think they remember they almost invariably post-date events in their past history. Finally, there may come the reply of "childhood" to the question of when the deafness was first noticed, and that is a vague and unsatisfactory response. Still, after all possible allowances for error had been made, and caution exercised to exclude doubtful replies, the result has apparently been emphatic enough to swamp the influence of occasional errors.

To facilitate comprehension, the data obtained have been plotted out in the accompanying graphs, the numbers being brought into ratio with the numbers of the general population at each life-period. (Not that it has made much difference!)

Several points of interest may be indicated. First of all, the three classes show certain striking differences, the one from the other. We refer to this below.

Secondly, in primary otosclerosis (Fig. 1), 64 cases out of 426 (or 15 per cent.) began before the age of 15, a percentage much higher than that of former observers, and higher than I had anticipated. Further, the peak of highest frequency is reached between 15 and 35, when no fewer than 205 (or nearly 50 per cent.) of the total, began. Thus, this variety of the disease begins most frequently in the first half of life, actually 320 out of 426 (or nearly 80 per cent.) beginning before the age of 35. Then, after a decline in the 35 to 39 quinquennium, there is a slight rise again in the 40 to 44 age-period, prior to the final decline; which rise I regarded at first as perhaps connected with the menopause, until the separation of males from females in another graph (Fig. 4) proved I was wrong.

But while a strong preference for early adult life is manifested, it should be noted that primary otosclerosis may start at any age. I have one case that began as early as three, and another as late as 70.

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In the second graph (Fig. 2), showing the ages of onset in secondary otosclerosis, the two branches of suppuration and catarrh are separated. The figure shows that deafness from the former begins for the most part before the age of 15, while that from catarrh becomes more frequent from 20 to 49. This particular difference, however, only bears out what

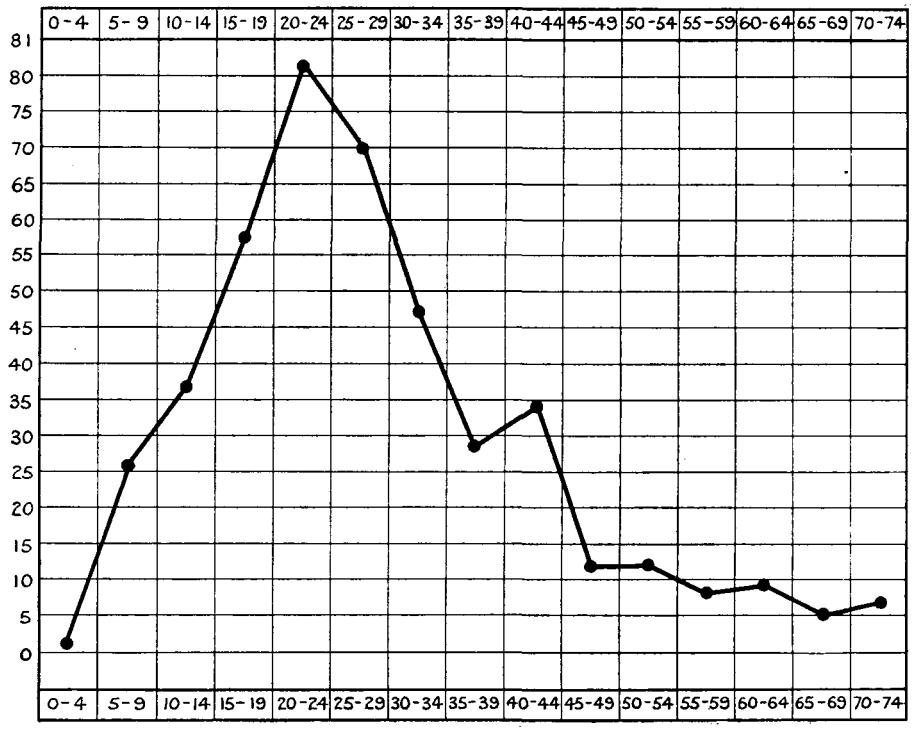


FIG. 1.—Age at onset. Primary otosclerosis. Both sexes. 426 cases. Corrected for population.

we might expect, since suppuration is commoner in childhood than in adult life.

We have referred to the occasional difficulty experienced in deciding whether to put certain cases into the primary or into the secondary catarrhal group, but if Figs. 1 and 2 are compared it will be seen that our distinction must have been in the main correct, since well-defined differences between the two types are manifested. Thus, the peak of high frequency in primary otosclerosis is reached in the five-year age-period before that of catarrhal otosclerosis; and in the

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latter, the slope marking the decline in numbers in the later years of life is less steep. Both show the same sudden rise after 40, but the catarrhal is again five years later than the primary variety. (This seems to suggest that otosclerosis of the primary type begins earlier in life.)

In the class of "incipient" otosclerosis, as I have already said, I have placed 82 cases in which a minor degree of intractable deafness with a negative Rinne has been present for a definite length of time. Tinnitus was frequently

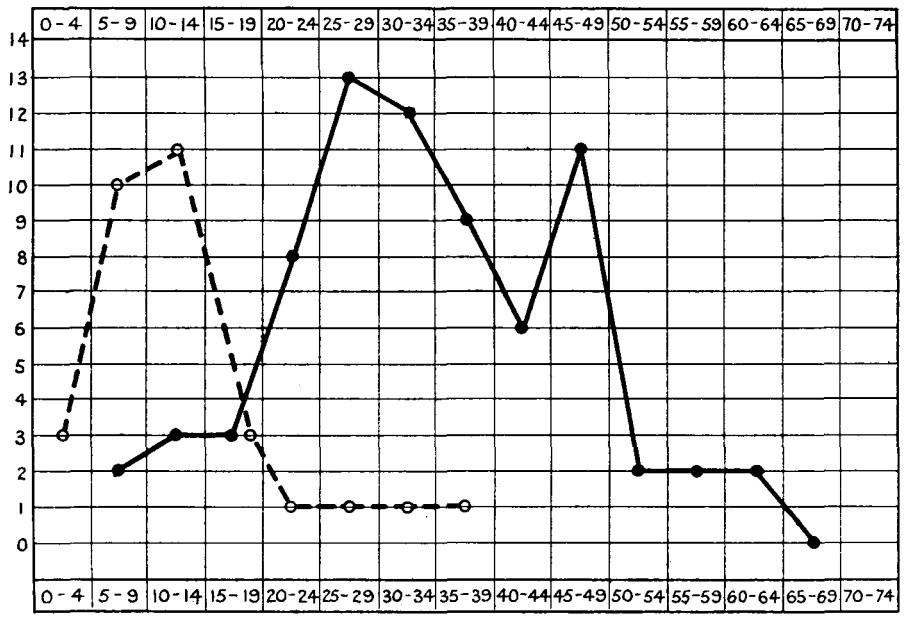


FIG. 2.—Age at onset in secondary otosclerosis. Interrupted line, suppuration. Continuous line, catarrh.

the chief, and sometimes the only complaint, and it was at times severe, while the deafness was slight. Still, to qualify for admission, some falling-off in hearing was considered indispensable.

Obviously, it was difficult to convince one's self, to begin with, that what one had to deal with here was in actual fact some interference with stapedial freedom, and so, as a test, X-rays were employed in two typical cases. In both of them H. K. Graham Hodgson reported porosis of the petrous, a result which fortified me in my assumption that all were otosclerotic in nature.

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The graph of the ages of onset of this class (Fig. 3) shows a curious difference from those of the other two groups, the peak of frequency being reached ten years later than in primary, and five years later than in secondary otosclerosis. Further, it remains relatively higher in the later years of life.

The difference may, to be sure, be only apparent—an accident due to the paucity of the numbers available—and so of no consequence. On the other hand, a straw may show

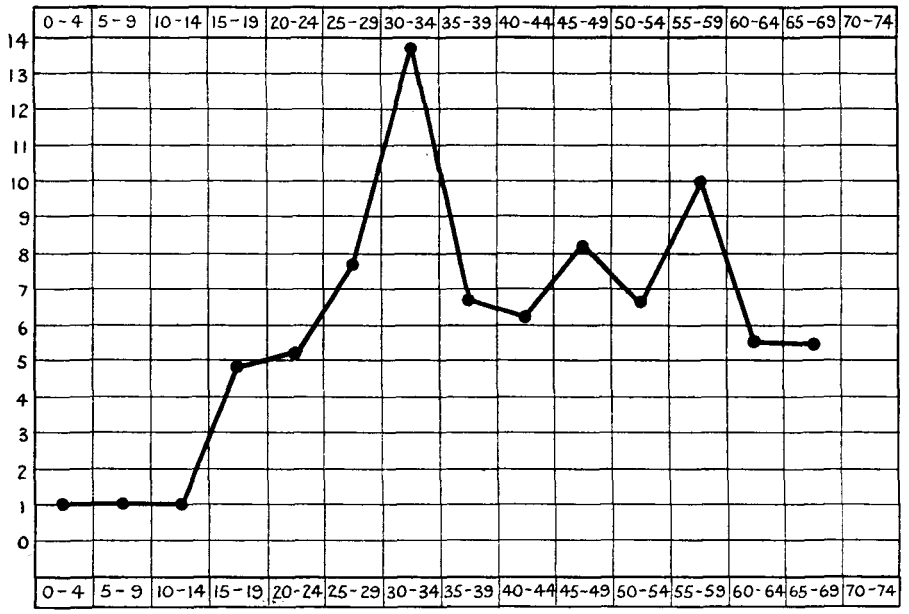


FIG. 3.—Age at onset. Incipient otosclerosis. Corrected for population.

how the wind is blowing, but although we may accept the figures, I cannot explain them.

Ages of Onset in the Two Sexes.—The preponderance of females suffering from otosclerosis inevitably raises the question whether the disease—in one of its forms at least—has not some connection with female metabolism. And at this point in our discussion we may appropriately insert what we have found concerning the relationship of primary otosclerosis to pregnancy.

Forty women were interrogated on this point. Of these, in three the deafness had first come on during pregnancy; in 11 it appeared shortly after childbirth; in three, already existing deafness became worse during pregnancy; and in 14 it became worse after childbirth. On the other hand, eight

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reported their deafness to have been uninfluenced by pregnancy or childbirth; and one said she heard better when she was pregnant. One woman noticed that her tinnitus and deafness were always worse during her menstrual periods.

(These numbers are certainly fewer than they ought to be.

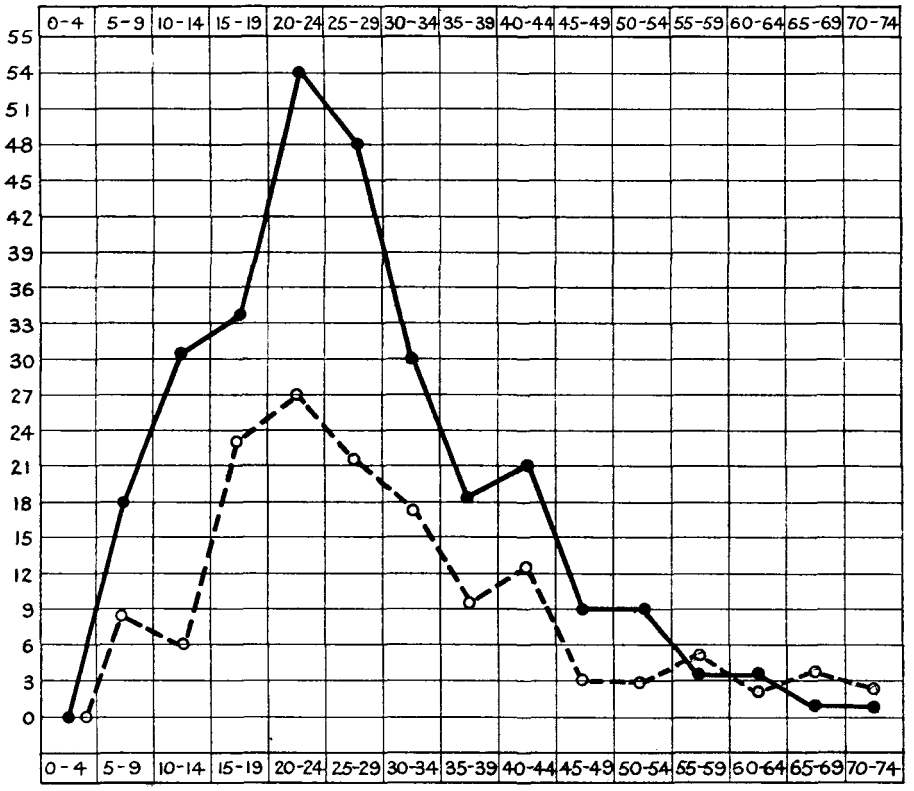


FIG. 4.—Age at onset. Primary otosclerosis. Interrupted line, male. Continuous line, female.

I did not always remember to go into this question with my patients.)

While we are on this subject, I ought perhaps to add that it has been reported, on French authority, that primary otosclerosis never sets in after hysterectomy. I am able to report two cases in which it did. Again, on the same authority, the statement is made that the menopause brings the advance of primary otosclerosis in women to a standstill. Nevertheless, as a glance at the graphs will show, otosclerosis may begin

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after the menopause, and, indeed, there is, as we have already observed, a rise in the numbers of fresh cases between 40 and 45.

Still, the facts of the preponderance of females and the influence of pregnancy are undeniable, and so it occurred to me that the ages of onset should be plotted out for the sexes separately, as, if some peculiarly female biochemical process were responsible, the two tracings ought to show a difference. But, as will be seen from Fig. 4, the tracing for males runs nearly parallel with that for females, even in the slight rise between 40 and 44. (Towards the end of life, the male line rises). Thus, the supposition that some peculiarly female metabolic agency is responsible for the greater prevalence of primary otosclerosis in women receives no support from my figures, and the cause of this remarkable difference between the sexes remains unknown.

(I have sometimes wondered whether the deleterious influence of pregnancy is not due simply to the fullness and vascular engorgement of the upper respiratory regions present at that time.)

The Deafness Progressive.—Perhaps of all the phenomena of otosclerosis the tendency of the deafness to progress from bad to worse is its most outstanding feature, and the questionnaire rightly lays great stress upon this character.

Sometimes, however, authorities speak as if progress were always regular and uniform. It is not so, of course. Even in its onset the disease may show hesitation and delay. Every now and again a patient will relate how the deafness and tinnitus on several occasions came and went entirely before they settled down and became persistent.

In an effort to gather together some definite information regarding the rate of progress, the period of time that elapsed between the first appearance of symptoms (deafness or tinnitus, or both) and the arrival at the stage when the patient became seriously deaf was obtained in 124 cases of primary and 61 cases of secondary otosclerosis. (The incipient variety was necessarily excluded.)

In accumulating the numbers, attention was also paid to the age of onset in each case, in order to discover, if possible, whether the rate of progress bore any relationship to the patient's age. My own impression had been that young people

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became deaf more quickly than old. I was wrong, as the following figures show.

Age.	Primary O.S. (Average).	Secondary O.S. (Average).
0 to 10 years	4 years	.....
10 „ 19 „	5 „	4 years.
20 „ 29 „	5 „	4 to 5 years.
30 „ 39 „	4 „	3 years.
40 „ 49 „	3 „	3 to 5 years.
50 „ 59 „	4 „	5 years.
60 „ 69 „	2 to 5 years	.....

According to this table, then, there is very little difference between the averages of the various decades; if anything, people over 40 seem to become seriously deaf in a shorter time than young people. But there is so little difference either way that it would not be permissible to generalise from them.

Further, and here is a point I wish to emphasise strongly, there is among the individual cases, no matter what their age may be, a variation that can only be called extreme. Thus, two cases of primary otosclerosis reported that high deafness had developed in the course of a few days, and in two other cases a high level had been reached in three months. Indeed, a report of instantaneous deafness in one ear, giving the signs of primary otosclerosis, occurs in my records. On the other hand, in 16 cases progress to high deafness took ten years or longer; and of these, two took twenty, and one forty years!

The general average at all ages, as we see, works out at three and a half years; that is to say, the aforesaid extreme differences are relatively rare and, when otosclerosis sets in, the patient is likely to be seriously deaf in from three to four years. This average, I may say, is the same both for primary and for secondary otosclerosis, dating the latter as nearly as possible from the time when the deafness begins to progress. (Note again the resemblance in detail between the two varieties.)

But it is necessary to add that we have no means by which at the onset of the disease we can foretell the rate of progress. Later, perhaps, we may hazard a guess based upon the patient's history of slowness or rapidity. But even then our guess may

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turn out to be unhappy, for not only does the difference between cases amount to anything from a few months to thirty or forty years, but progress itself, after the disease has started, may be irregular and interrupted. In most cases perhaps, there is a slow, steady, and almost imperceptible degradation, but many also exhibit sudden changes for the worse, sudden steps downward, sometimes small, at other times considerable; from which declension there is no return.

On the other hand, again, pauses in development will occur for no apparent reason. Thus, I have notes of seven cases of primary otosclerosis in which, after a moderate degree of deafness had been reached in a few years, the deafness became stationary, with progress arrested for from three to forty years. In four of these, I may add, advance was subsequently resumed until high deafness was reached. The same behaviour was noted in some cases of secondary otosclerosis, with arrest for twenty and thirty years.

Many of the cases we are calling "incipient" otosclerosis must also be instances of arrested progress; they are, perhaps, aborted cases.

At this juncture we may conveniently advert to *unilateral otosclerotic deafness*, of which I have, in the primary class, 67 cases; 41 of these remained unilateral as long as I knew them, while in the remaining 26 the good ear became deaf after intervals ranging from two to thirty years (see later). Of the secondary class, 22 are noted, most but not all of them, being suppurative or post-suppurative in origin. Here also the deafness, after a longer or shorter period, may come to affect the hearing ear. In one instance I found that, with active suppuration in both ears, the otosclerotic process had affected one and spared the other.

(It will be remembered how Politzer has described an occasional stalactitic bone-formation in the middle ear of chronic suppuration.)

Another interesting group is found in those unilateral cases, in which the history and the appearance of the tympanic membrane show the deafness to have been initiated in childhood by suppuration of that ear, now long since dried up, the other ear having apparently escaped, but in which this apparently *undamaged* ear later on in life shows the signs of severe otosclerosis.

How are we to account for such cases? The natural

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supposition is that the good ear also has been attacked by acute otitis media in childhood without perforation of the membrane and discharge, and that the scar-tissue left by the disease has led to otosclerosis in later life. But this does not explain every case of unilateral deafness. Take the following: An officer in the Flying Corps during the War (1918) made a steep and rapid dive. On landing from his aeroplane he found himself deaf in both ears. Never having heard of the Valsalva method, he made no effort to inflate his ears, and although one of them recovered spontaneously and completely, the other remained permanently deaf. Thirteen years later, he came to me because the good ear had lately become seriously deaf. His symptoms and signs were those of typical otosclerosis. There was no family history of deafness and no signs of catarrh. How did the disease spread to the sound ear? (Was the lesion in the deaf ear a dislocation or subluxation of the stapes?)

One point is to be noted, however, as illuminating this problem. It is usually the patient who makes the diagnosis that it is unilateral, and, as a matter of fact, the otologist will very rarely find that the ear which is supposed to be sound is quite normal. In only one or two of my unilateral cases was it found to be so. In all the others, the tuning-fork and other tests showed the hearing to be defective in certain parts of the field of hearing.

Probably, therefore, pathological changes are present in both ears, but in one their situation or extent is such that they do not interfere with the transmission of sound-waves.

The patient, of course, is usually quite unaware of any deficiency so long as he retains his hearing for conversation. In that case he looks upon the hearing in the good ear as "perfect." A young woman, for example, with typical primary otosclerosis in both ears, heard the whisper in her deafer ear at four inches only, and in her better ear at eighteen feet, and yet the hearing for the medium tuning-fork showed minus twenty-five seconds by air-conduction in both ears equally.

Tuning-fork tests cannot always be relied upon as indications of hearing for the voice.

Let me finally emphasise once more the fact that, on the one hand, in unilateral otosclerotic deafness the good ear may retain useful function for a lifetime; and, on the other hand, it is impossible to tell whether the good ear will ever become deaf, or when it is likely to do so. Of my primary cases of

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unilateral deafness, 26 ultimately became deaf in the good ear also; five of them after thirty years; one after twenty-five years; two after twenty years; and the others after from two years upwards. This behaviour should be compared with the manner in which bilateral otosclerosis develops and progresses.

Further, in this, as in so many other details, the primary and secondary classes show a close resemblance.

We turn now to *Heredity*.

Family History.

Primary Otosclerosis (285 interrogated).

Positive . . .	46 per cent.
Negative . . .	47 „
Doubtful . . .	6 „

Secondary Otosclerosis (43 interrogated).

Positive . . .	32 per cent.
Negative . . .	67 „

Incipient Otosclerosis (40 interrogated).

Positive . . .	40 per cent.
Negative . . .	60 „

All three classes agree, although the percentages differ somewhat. Taken together, the percentages are:—Positive, 44 per cent.; negative, 51 per cent.; doubtful, 5 per cent. These numbers give a lower positive percentage than those of other workers. With regard to the difference, one or two remarks are necessary.

First of all, when we ask if any of the other members of a family are deaf, and are satisfied with a simple positive reply, our statistics will obviously be of but little value. Deafness, more or less severe, is so prevalent that there must be few families of three living generations altogether exempt from it. So we ought to discriminate, excluding as much as possible deafness due to suppuration or to senility.

Strictly speaking, I suppose, no case in a family should be admitted as otosclerosis until it had been examined by an otologist. But this is a council of perfection, and so we may accept as positive a history which gives two or more members of a family that suffer from severe deafness without discharge, and beginning early in life.

Thus there is a danger of including deafness from causes other than otosclerosis, and so of erroneously increasing the

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numbers of otosclerotic families. On the other hand, patients often forget, or they may be quite ignorant of the presence of deaf relatives in the collateral branches of their family.

Consequently therefore, some discrepancy in the figures of different observers must be expected, and too great a reliance upon numbers and percentages should be avoided.

The undeniable fact remains, however, that in nearly one-half of our cases otosclerosis was said to be a family disease.

It may be of interest to hear that, according to my observations, a positive family history of deafness does not necessarily lead to an unusually rapid progress. Thus, in spite of such a history, I found one case with a course of 14, and another of 20 years before the high grade of deafness was reached.

Hearing-Tests and the Bezold Triad.—Here follow the results of the tests for bone-conduction with the middle C tuning-fork in 1120 ears:—

Primary Otosclerosis.

Bone Conduction	positive in . . .	66 per cent.
„	negative in . . .	34 „

Secondary Otosclerosis.

Bone Conduction	positive in . . .	64 per cent.
„	negative in . . .	36 „

According to my results then, the first member of the Bezold triad is reversed in one-third of the cases. (I ought perhaps to mention that, although I always make my own tests, I use a third person with normal hearing as a standard.)

It is to be noted that the results in primary and secondary otosclerosis are the same to within two ciphers in the percentage results—a striking concordance.

But the third class (incipient otosclerosis) differs from the primary and secondary. Here the negative bone-conduction preponderates, the percentage being:

Incipient Otosclerosis.

Bone Conduction	positive	42 per cent.
„	negative	58 „

In commenting upon this result I would remind the reader that the primary and secondary classes embrace only the higher grades of deafness, while in the incipient class the deafness is

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but slight. On a former occasion I reported that in slight or early obstructive deafness from whatever cause arising (even from impacted cerumen!) a negative bone-conduction for medium forks in ordinary domestic surroundings is a common finding. Consequently, our present percentages for the mild deafness of incipient cases is in accordance with previous experience.

Thus, the statement that the bone-conduction is always lengthened in otosclerosis (the first member of the Bezold triad) requires modification if the above findings are substantiated.

When, however, we turn to the *Rinne test*, we find that, in all the ears I have included in my lists, Rinne was (with the solitary exception of one ear) invariably negative, even when the bone-conduction was negative. That means, of course, that the falling-off in bone-conduction, when it is present, is always slight, even in severe deafness. In this respect such cases differ from the pure nerve-deafness type.

Turning next to the *Upper Tone Limit*, in 129 cases of primary otosclerosis I found it lowered in one or both ears in 99 (allowance being made for age), or 77 per cent. In secondary otosclerosis (16 cases tested) it was lowered in 13 (81 per cent.). In the incipient class with slight deafness, it was lowered in five out of nine cases tested. Thus in all three classes, it was depressed in the majority. But here also the reduction was slight, even in severe deafness, not falling below 2 c.m. from the normal on the monochord, herein again presenting a strong contrast to nerve-deafness, in which the lowering is much greater in proportion to the degree of deafness.

As regards the hearing for the *deep tones*, my findings show an invariable diminution of hearing for T.F. 64 by air-conduction. As to the bone-conduction for the same fork, it was reduced in 69 per cent. of the primary cases, 71 per cent. in secondary cases, and 57 per cent. in incipient cases; all three classes again agreeing.

(It would appear, therefore, from the foregoing figures that the difference between "obstructive" and "nerve" deafness in actual practice is frequently a matter of degree rather than of kind.)

It is sometimes said that a lowering of the bone-conduction, whether for low or medium forks, is of bad prognosis as regards hearing, since it forebodes a rapid advance to serious

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deafness. This opinion receives no support from my cases, in which the rate of progress was found to have no relationship either to the bone-conduction or to the level of the upper tone limit.

Tinnitus.—In 341 cases of primary otosclerosis tinnitus was absent in 17 per cent.; slight or moderate in 59 per cent.; and severe, that is to say, so obtrusive as to cause some mental distress, in 23 per cent. In 99 secondary cases it was absent in 15 per cent.; slight in 63 per cent.; and severe in 21 per cent. In 70 incipient cases it was absent in 17 per cent.; slight in 57 per cent.; and severe in 25 per cent.

The agreement between our three varieties, even with regard to the different degrees of a subjective sensation, is astonishing.

The sounds heard by the patient in his tinnitus vary considerably. In most, the noise is what I have called "compound," or made up of a number of different sounds, "hissing," as a rule, predominating, intermingled with "throbbing," "roaring," and sometimes "whistling." Odd sounds, like the musical dropping of water, or a "ping" like the twanging of a treble string on the harp, are occasionally heard. Another and an interesting noise is what is generally called a "crash." This is a loud, sometimes a very loud, sound, fortunately infrequent, which, until the patient gets accustomed to it, he often believes to be an external noise. Now and then it occurs when he is asleep and wakes him up.

One of my patients, a male, occasionally heard it when shaving, and, having an inquiring mind, he discovered that it followed the first stroke of the razor on his cheek. The first time he heard it he turned round to see who had slammed the door! Probably, therefore, the "crash" is due to a sudden reflex spasm of the stapedius muscle—a pathologically exaggerated stapedius hum—and this explanation is supported by the fact that it is usually heard only in the early stages of otosclerosis.

The questionnaire asks whether tinnitus appears before or after the onset of deafness. I cannot give any numbers or percentages, but I find from my notes that in most of the cases it was the deafness that appeared first. Frequently enough, however, they both began at the same time, and there is a fairly numerous group in which the tinnitus preceded the deafness, sometimes by several years. It may, besides, as in

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many cases of the incipient variety, remain the patient's chief complaint even after deafness sets in. We are all acquainted with the patient who says she "could hear quite well if it weren't for the noises."

I have come to regard the appearance of tinnitus in an old dried-up middle ear suppuration or catarrh with misgiving, as it often marks the commencement of the active otosclerotic change.

Paracusis Willisii.—The simplest explanation of this phenomenon, and in many people the only explanation necessary, is that it is due to normally hearing people not only speaking louder in a noise, but speaking unconsciously on a higher note, so that their voice can evade the damping effect of the noise waves, which, for the most part, are of low frequency, particularly in railway trains, buses, and so on.

But that is not the whole affair, at all events with certain patients. For instance, one averred that in a railway train he could hear a pin drop! And another, that he not only heard better when he was at a concert, but also for an hour or two after. Then there is F. P. Sturm's observation that a watch-tick may become audible when a tuning-fork is sounding on the mastoid.

It looks, therefore, as if paracusis might also be due, at least in some cases, to the loud external noise stimulating sluggish neurones in the auditory tract, and thereby rendering them, for the time being, more sensitive. This would also account for deafness being sometimes worse after prolonged exposure to noise, the neurones becoming exhausted.

There is no doubt at all in my mind that the nerve-centres in otosclerosis and other forms of severe deafness do frequently become inert, and that they can be wakened up by a number of different agents. Noise, as I have just said, may do so; and wireless music conveyed through ear-phones; and finally, a remedy for deafness pushed by an enthusiast!

Thus to the organic is added, in many, but not all cases, a psychological or functional deafness, and it is the removal of this latter element by suggestion that so often makes it difficult for us to estimate the value of new methods of treatment.

Vertigo. — Sydney Scott has made us familiar with the occasional occurrence of vertigo in extra-labyrinthine ear-

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disease, and we encounter it with some frequency in otosclerosis. It is mild and momentary, as a rule.

Primary. (186 patients interrogated.)		
Vertigo, slight	. . . .	30 per cent.
„ severe	. . . .	7 „
„ absent	. . . .	62 „
Secondary. (22 patients interrogated.)		
Vertigo, slight	. . . .	45 per cent.
„ severe	. . . .	4·5 „
„ absent	. . . .	50 „
Incipient. (29 patients interrogated.)		
Vertigo, slight	. . . .	34 per cent.
„ severe	. . . .	6 „
„ absent	. . . .	59 „

(In one of the last group the vertigo was accompanied by vomiting.) All three classes again agree in detail.

Associated Local Phenomena—*The Membrana Tympani.*—The flamingo-tinted membrane was observed in 17 cases of all three classes, and several interesting variations appeared. Once almost the whole of the membrane in both ears was flushed; once the colour was visible in the anterior half of the membrane; once in Shrapnell's membrane; and once high up over the situation of the long process of the incus. Twice it was seen in catarrhal otosclerosis, and I have also observed it in simple subacute otitis media without perforation.

The idea that it is a red, congested promontory, glowing through the membrane, is a plausible explanation, although it suggests *inflammation*; and I have sometimes thought that the beautiful opalescence of the membrane in young female otosclerotics may be due to the same kind of flush, modified and diffused in its passage through the alabaster-like medium.

The tympanic membrane in old otosclerosis may be of almost any variety. Perhaps indrawing with the umbo of the malleus and a small circlet of the membrane around it adherent to the promontory, is the commonest appearance. But we often come across a normal membrane in otosclerosis, just as we often come across the indrawn, adherent membrane in people with normal hearing. Another type is that of the thin, atrophied membrane, as fine and flimsy-looking as tissue-paper. Or it may show scars, depressions, and perforations,

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the remains of old dried-up suppuration. In like manner, Siegle will show the malleus sometimes movable, sometimes fixed.

In short, otosclerosis may be associated with any kind of tympanic membrane, normal and abnormal; and so may perfect hearing! So I confess I have arrived at the stage of paying very little attention to it.

Among the associated local phenomena of otosclerosis there is one I should like to draw particular attention to in spite of its infrequency. This is the curious matter of *bony outgrowths* in the *external auditory meatus*, which I have observed in 16 cases of primary and in five cases of incipient otosclerosis. (Six of them were doctors!)

As is well enough known, there are two varieties of irregular bony growth in the meatus. First, the exostosis proper, or osteoma; and second, a more or less general hyperostosis of the walls of the bony meatus, frequent in swimmers and divers. But while seven of my cases were of the latter variety, one could not call the others exostoses or osteomata exactly. For they consisted of small, white nodules, about the size of a pin-head, sometimes single, oftener in twos and threes, frequently situated close to the tympanic membrane; but once or twice they were projecting Shrapnell's membrane, and once I saw the short process of the malleus enlarged and made prominent by such a nodule.

The natural inference is that similar formations are present on the medial wall of the middle ear limiting, like the porotic islands of the classical otosclerosis, the movements of the stapes. They may, indeed, be of the same origin and nature as those islands (whatever that may be!), a possibility supported by the fact that one of the cases of incipient otosclerosis, in which H. K. Graham Hodgson found porosis of the petrous bone, showed the meatal nodules we are discussing.

But by far and away the most frequent, and perhaps also the most significant of the associated phenomena, I have left to the last: namely, the extensive set of disorders of the upper respiratory regions and ears that goes by the name of *catarrh*, and includes the common cold and influenza; all the many varieties of rhinitis and pharyngitis, acute and chronic, with or without nasal sinusitis; to say nothing of adenoids; the exanthemata that are attended with rhinitis; and the class of disorders we call paroxysmal rhinorrhœa.

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Otosclerosis

The relationship of those diseases in their more chronic forms to otosclerosis is, perhaps, easy enough in one way to understand. But the relationship of the more acute and transient varieties, like influenza and the feverish colds, is not so apparently simple.

In order to clear the ground for discussion, we must first of all refer once again to that variety of otosclerosis in which catarrhal complaints in nose, throat, or ear are conspicuous by their absence, these regions looking, as has been said, "abnormally normal." Everything indeed appears to be perfectly sound and in order—except the hearing. This is otosclerosis as it was originally described—we often call it the "classical" otosclerosis—and in it the only lesion discoverable post-mortem is a discrete porosis or cancellification of the bone of the labyrinth, a change for the cause of which three generations of otologists, including our own, have searched in vain. But this kind stands by itself, and common though it be, there is another variety as common or commoner. This is to be found when, although no signs of otitis media past or present exist, definite nasal and/or naso-pharyngeal catarrh cannot be excluded from the history; a state of matters which is present, according to my figures, in 103 out of 411 cases of primary otosclerosis.

If it be asked "Why are these cases placed in the primary and not in the secondary class?" I can only reply that as every shade and variety of case is encountered in practice, from the non-otitic "classical" variety at one extreme to the actively catarrhal—nay! the suppurating—at the other, it is often extremely difficult to decide to which class a case belongs. Yet one must draw the line somewhere, and in this instance I adopted the presence or absence of active catarrh of the middle ear as the criterion. Cases with the former went into the secondary class; cases with the latter into the primary. And, as we have already seen, the tracings of the graphs seem to indicate that the classification followed is true enough to Nature for all practical purposes.

An examination of primary cases with "catarrh" in the history reveals some interesting points. In 14 the otosclerosis was started by a "cold in the head"; in 13 it was attributed to "influenza"; and it is, of course, a common experience for otosclerotic deafness to be made suddenly and permanently worse by the same ailments.

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("Colds" generally make the hearing worse for the time being, but 11 primary cases heard better during a cold.)

A common story is that the patient earlier in life has been very subject to colds in the head, but has gradually got rid of them, sometimes altogether; and yet the improvement in this respect either fails to arrest the progress of the deafness, or else the deafness has not been noticed until the tendency to colds has disappeared. In the latter event, the patient may deny that the former colds were ever attended with deafness or any symptom referable to the ear. In some, however, the colds have caused temporary dullness of hearing.

In this connection we must also allude to the influence of the exanthemata in childhood as the starting point of the otosclerotic process. Of these, I have 10 cases (scarlet fever, measles, diphtheria), and the deafness dates from the acute stage of the disease, or shortly after it, and without any discharge from the ears, or any evidence thereof in the stigmata of old suppuration on the tympanic membranes.

Again, every otologist has experience of children with deafness attributed to adenoids in whom the removal of the adenoids is not followed, as in the great majority of such cases, by the cure of the deafness.

Now, the fact common to all these is that the inflammatory agent that has apparently excited otosclerosis has appeared and disappeared completely, sometimes rapidly, sometimes slowly; but during its sojourn it has launched a pathological process that has persisted and advanced, and continues to persist and advance, of its own proper strength and motion. For, although the history shows the original cause to have been catarrhal, that is to say inflammatory, the patient shows, when he comes to us with his deafness, no sign in nose, throat, or ear of the original disease. And it is because of the absence, or apparent absence, of *active* catarrh that I have placed these cases in the primary list.

We turn now to secondary otosclerosis and, first of all, to the suppurative group. The relationship of suppuration of the middle ear to otosclerosis is of interest, and, perhaps, importance. There is no doubt, of course, that the great majority of cases of acute suppuration, and a smaller majority (but still, I think, a majority) of cases of chronic suppuration show, when that disease is brought to a final standstill, no tendency whatever to progressive deafness. Whatever dullness of hearing may be

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left after active suppuration is cured and cicatrisation completed, it remains unchanged for the rest of the patient's life. Only a minority become otosclerotic.

Now the question arises, "Is it not possible that otitis media is the forerunner of all otosclerosis, even of the 'classical' form in which, later on in life, no trace of any old inflammation can be found?" Acute otitis media, we know, may pass rapidly through its several stages to complete cure, and heal up without leaving any trace of its presence either in the membrane or on the mucosa. No great effort of imagination, therefore, is necessary to see in osteoporosis the sequela of an inflammatory reaction begun many years previously.

With this idea in mind, I began a few years ago, in otosclerosis apparently of the classical type, to search for a history of acute otitis media in childhood. Fourteen cases with such a history were found, and this number is certainly smaller than it would have been had I begun this particular enquiry at an earlier date.

To diverge somewhat from our main theme, I may mention here that at one time I formed the impression that acute purulent otitis media in adult life seldom or never occurred in otosclerosis. But this impression has gone the way of many more since I began to go through my records systematically. There are nine instances in my notes of acute suppuration in adults with otosclerotic ears, and, what may amount to the same thing, I have seven cases of chronic suppuration in the one ear with otosclerosis in both.

The catarrhal group of secondary otosclerosis need not detain us long. Into this category cases showing manifest active catarrh of the middle ear were placed, as well as those in which the history favoured the diagnosis of a past subacute or chronic catarrhal otitis media, usually associated with chronic rhinitis.

It is not difficult to figure what happens in the latter variety. As time passes, the thin submucous tissue of the middle ear, infiltrated as it is with round cells, will become more or less interpenetrated with cicatricial tissue, the contraction of which, together with the adhesions formed, will, if suitably placed, impede or immobilise the stapes. This, of itself, will induce symptoms of otosclerosis; but, in a certain number of cases, the fibrosis of the middle ear and its contents may, as Fraser's findings permit us to suppose, also be accompanied by osteoporosis of the labyrinth bone. Further, the fixation of the

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stapes from fibrosis can end in bony ankylosis of the stapedio-vestibular articulation without osteoporosis; while the same event may occur also as a result of osteoporosis.

With regard to the association of osteoporosis with otitis media, however, there is one caution we cannot refrain from mentioning. It is quite possible that the two, osteoporosis and otitis media, have nothing to do with one another, each arising independently of the other and from entirely different causes.

Nevertheless, looking back now over the facts and arguments we have gathered together concerning catarrh and suppuration in otosclerotic cases, we cannot avoid the suspicion that somewhere in the background of all otosclerosis the shade, as it were, of otitis media keeps flitting in and out of sight, and we begin to wonder whether it is not this factor that plays the all-important part in the genesis of the disease in every one of its varieties, including the classical type.

But there is a drawback, in the present state of our knowledge, to the theory of an inflammatory origin for all forms of otosclerosis, attractive though a single cause may be to simple minds; and that is, that even if otitis media, in some shape or form, were invariably present, whether before or during the course of the progressive deafness, our problem would not be solved; it would only be changed.

Otitis media in children, mild and evanescent or severe and prolonged, is extremely common. There can be few infants and children who entirely escape it, yet only one here and one there is destined to become otosclerotic. That is to say, otitis media itself cannot be the sole cause of otosclerosis. Some other factor, and that factor one that is still unknown to us, there must be, to account for the fact that otosclerosis is selective. Of this mysterious element in the situation, which, be it remembered, is itself said to be sometimes the sole cause, independent of otitis media, we are not entirely ignorant. It occurs more commonly in women than in men, and it is probably hereditary. What may be the peculiarity that renders women particularly susceptible we cannot tell. Many theories have been propounded, based usually upon biochemistry and endocrinology, but they are little more than guesses. In like manner, what may be the hereditary bias that is transmitted is equally mysterious.

True, a stalwart supporter of the otitis media theory of origin of osteoporosis might suggest that what is inherited

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is not a general disposition but a local character, a particular shape and arrangement of the structure of the middle ear and its contents, in consequence of which the inflammatory process can easily affect the bone. Facial features are transmitted by heredity, and why not also tympanic features? The relative prominence of the promontory and the depth of the fenestral fossae might make all the difference. But such a suggestion will only arouse attention when it has been demonstrated to be fact.

Our discussion has brought us now to the next section, namely, the *General Phenomena* associated with otosclerosis.

Three constitutional states have been so associated: child-bearing, anæmia, and rheumatism. The first we have already considered and acknowledged, and there remain now the other two.

In my records *anæmia* is very seldom met with. Very few even of the younger women are noted as anæmic, since only six are so described. Here again my observations disagree with those of several other workers, and it may be that I have overlooked many cases. On the other hand, the otosclerosis of young women often first appears about the time of life when chlorosis is common, and so their association may be purely fortuitous. Further, chlorosis is to-day much less prevalent than it used to be, and its infrequency in my records may be accounted for by that circumstance. At all events, there is no doubt whatever that primary otosclerosis occurs quite frequently in young women who are not at all anæmic, and I have seen it in patients who are noted as florid and full-blooded.

On the other hand, however, we must admit that a heavy loss of blood in an otosclerotic is apt to be followed by a rapid increase in the deafness. My records tell of one instance following severe hæmorrhage after a nose operation, and another after metrorrhagia. In the cases influenced by childbirth, however, post-partum hæmorrhage has not been conspicuous. We may, therefore, perhaps ascribe the aggravation of the deafness after severe bleeding to the debilitating effects of acute anæmia rather than to the loss of blood as a specific agent, so to speak, particularly as an increase in deafness is apt to follow any long and weakening illness.

With regard to *rheumatism* in its various manifestations, it is difficult to arrive at a decided opinion. My records

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certainly show it to be common, particularly in patients over middle age. But here again simple coincidence cannot be denied, for the very good reason that the number of elderly people who suffer from what they call "rheumatic" pains is very great. Moreover, we come across many patients with fully-developed otosclerosis who have never suffered from any form of rheumatism.

Four only of my patients are noted as having had rheumatic fever, and in two of them the deafness began immediately after their first attack. The other two died of acute rheumatism.

I have sometimes thought that the earache otosclerotics occasionally complain of (21 cases noted) is more common in rheumatic people; and it may be contended that, although not responsible for all, rheumatism may be one of the constitutional disorders that favour otosclerosis. Let us examine that suggestion.

A similarity, clinical and pathological, between arthritis deformans and otosclerosis was the subject of an article by J. F. O'Malley in 1915. As far as the pathological appearances are concerned, there is undoubtedly some resemblance. (We are talking of what is nowadays called osteo-arthritis; not rheumatoid arthritis.) But the clinical resemblances do not seem to be so striking to-day as they were when O'Malley wrote. Thus, the age of onset, the sex distribution, and the heredity percentages were, in 1915, very much alike in otosclerosis and in arthritis deformans (osteo-arthritis); whereas, if we are to believe J. A. Glover's recent statistics on arthritic disease (1928), the only item that shows any correspondence between the two is, curiously enough, heredity; and that is, as we have seen, a rather unreliable figure.

Besides, it seems to be the fact that otosclerotic people are no more liable to chronic arthritic disease than the general population, and that the proportion of arthritic people who are deaf is not particularly high. But definite information on this point is wanted.

Osteo-arthritis, and especially the monarticular type, can often be traced to an injury, and this can scarcely be given as a cause of otosclerosis, unless we concede, as well we may, that local sepsis would have very much the same effect as trauma.

On the whole, all I can say is, the incidence of osteo-arthritis is not particularly frequent in the cases we are considering here.

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Otosclerosis

Among other general disorders or changes we may mention that of *early senility*, of which a few have been noted in my lists. In these the mucous membrane of the nose and pharynx shows some degree of atrophy and pallor, like the atrophy and pallor of age. Dryness of the skin and scalp, and sometimes premature loss of hair, may be observed. There is no nasal catarrh and the Eustachian tube is abnormally patent. On the other hand, in several patients over 60 years of age I have noted that they look "surprisingly young!"

Whether or not *shock or strain* can set the otosclerotic process agoing is not clear. A dozen cases or so gave such a history, and many more blamed nerve strain for a rapid increase in their deafness. But this is a popular reason for all manner of diseases.

In the same way, there is little if any support in my records for the theory that otosclerosis is due to the influence of some *distant septic focus*, as osteo-arthritis is supposed to be. Appendicitis had occurred in seven cases, but it did not coincide either with the onset of deafness or with its times of aggravation; and the same applies to other septic diseases. Nasal sinusitis was found in 12 cases, but in that condition the local effect of sepsis cannot be excluded.

Finally, there also remains this to be said: quite a large number of otosclerotic people have been, are, and continue to be, extraordinarily healthy in every other respect.

Termination.—In all these 622 cases, only once was absolute deafness diagnosed, and then it affected only one of the two otosclerotic ears. The patient was a woman of 62, whose deafness had begun at 22. She was, I should add, one of the few cases with rheumatoid arthritis. There was a family history of deafness.

But although the absolute destruction of hearing is exceptional, in the final stage of otosclerosis the patient can only hear the shouted voice close to the ear, and is, therefore, to a lamentable extent, cut off from social activity and intercourse. At this stage the tuning-fork tests show, in most though not in all cases, a greater loss of bone-conduction than in the earlier stages. This indicates, without doubt, the supervention of pathological changes in the cochlea. But complete destruction of the cochlea does not follow.

Furthermore, as we have abundantly shown, there are great differences in the rate of progress, and so in the end

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reached. Most patients arrive at the final stage in three or four years; but in some that melancholy termination seems to be indefinitely postponed. This last circumstance encourages us to hope that there will some day be discovered a therapeutic method by which the disease can be arrested. And there is, moreover, the further hope that if the cause or causes of the disease can be discovered, some means of prevention will be forthcoming.

Thus otosclerosis is a self-limiting disease, the pathological changes in which, although they may induce secondary changes in the cochlea, do not lead to its total disorganisation. This fact seems to dispose of the theory that otosclerosis is a true degeneration of the organ of hearing.

With regard to self-limitation, moreover, it would appear from what we have learned of unilateral and incipient (or aborted) otosclerosis, that the arrest may take place before the patient has become seriously deaf. We can even imagine osteoporosis of the labyrinth capsule or fibrosis of the middle ear without either deafness or tinnitus!

Summary and Conclusions.—In the following table the percentage findings of the phenomena we have been discussing have been collated, the three classes being placed side by side in order that the reader may see at a glance where they agree and where they differ.

The table shows that in four of the nine items there is agreement between all three classes, an agreement extending even to the percentages of relative frequency. Of the five other items, the third, that of the time taken to reach high deafness, shows agreement in the primary and secondary classes, the only two that are comparable in this matter, and here again the numbers are nearly identical. In the matter of the tuning-fork and other tests, also, the two first classes present the same close approximation in percentage results. And even in the third class the figures show the same ratio as the others, save in the bone-conduction for medium forks.

Thus there is only one particular in which each of the three classes stands by itself, namely, in the climax of the ages of onset. We have already discussed this difference, and what we have now to consider is its reason and the significance that that reason may bear in relation to the general problem.

In this item we are compelled to distinguish not three but four classes (primary, catarrhal, suppurative, and incipient),

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since all four give a different age incidence. It is evident that in this particular section we are in contact with the original causes that start the otosclerotic changes, and as these causes are all different, so at this point, the earliest appearance of the otosclerosis, each of them determines the age at which the new development appears. (As said before, we do not apply this remark to the incipient class.)

	Primary.	Secondary.	Incipient.	
1. Sex	M. 31 per cent. F. 69 „	M. 40 per cent. F. 60 „	M. 39 per cent. F. 61 „	3 agree.
2. Age of onset (climax)	15 to 30 years	Suppuration, 5 to 14 Catarrh, 25 to 30	30 to 34	All 3 disagree.
3. Progress (average) .	3 to 9 years	4 years	...	2 agree.
4. Heredity + . .	46 per cent.	32 per cent.	40 per cent.	3 „
„ − . .	47 „	67 „	60 „	
	6 „ (?)			
5. Hearing tests. T.F. 256—				
B.C. + . .	66 „	64 „	42 „	2 „
„ − . .	34 „	36 „	58 „	
6. Upper tone limit—				
Lowered . .	77 „	81 „	55 „	2 „
Normal . . .	23 „	19 „	45 „	
7. T.F. 64—				
B.C. + . . .	31 „	29 „	28 „	2 „
„ − . . .	69 „	71 „	57 „	
Normal . . .	...	...	14 „	
8. Tn.—Absent . .	17 per cent.	15 per cent.	17 „	3 „
Moderate . .	59 „	63 „	57 „	
Severe . . .	23 „	21 „	25 „	
9. Vertigo—				
Moderate . .	30 „	45 „	34·5 „	3 „
Severe . . .	7 „	4·5 „	6 „	
Absent . . .	62 „	50 „	59 „	

Turning again to the details in which agreement is manifested, I may say that the result was, to me, a complete surprise. I had no thought of making an analysis of my records during the long years in which they were being formed, so when at last I began to collect the data my mind was free from any preconceptions, and so my figures have not been manipulated in obedience to any subconscious dictation.

It is to be noted that of the eight items which show general similarity five (viz., the hearing-tests, tinnitus, and vertigo)

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must be connected with, and caused by, the local lesion in the ear; while the remaining two (sex and heredity) must be dependent upon some general or constitutional factor. The reader is asked to bear this in mind, in view of the general discussion we shall presently embark upon.

As regards the local lesion, the general agreement between our three classes gives strong support to the doctrine that the physical impediment to the transmission of sound-waves must be, in all three groups, of the same nature, since it produces the same auditory phenomena in all. Specifically, that means that our results support the orthodox teaching that the symptoms are caused by a limitation of movement of the stapes, leading on to its immobilisation.

So far, I think, we are entitled to claim that our views are based upon sound pathological and clinical foundations. But, when we come to consider what the cause or causes of the stapedial limitation may be, we can only proceed with fear and trembling, as our theoretical explanations are but suppositions based upon a very slender modicum of fact.

For convenience, let us take the secondary class first, and from now on to the end of the paper we shall leave the third or incipient class out of count. In secondary otosclerosis, consequent, as we know it to be, upon some form of otitis media, the stapedial limitation and fixation may be brought about by scar-tissue binding down the ossicles and particularly the stapes. But the effect of scar-tissue must depend largely upon its situation. Thus we can picture to ourselves cases in which scar-tissue alone, without any osteoporotic change, will lead to ankylosis of the stapes; just as we can also picture cases in which its more fortunate disposition will leave the ossicles and fenestrae quite free and unimpaired.

Again, in the primary class, the stapedial fixation is, we know, due to bony overgrowth connected with osteoporosis, and that, in turn, if we exclude the hypothetical influence of a remote otitis media, is the result of some constitutional dyscrasia.

Thus we arrive at the conclusion that there are two separate and distinct varieties of otosclerosis, one due to a local cause, otitis media; the other due to some unknown constitutional cause.

But this conclusion is, on the face of it, too clean-cut and narrow. It ignores one or two large imposing facts, both in the secondary and in the primary class.

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In the former, the presence of heredity in 32 per cent. of the cases, together with, what is no less important, the female preponderance, are entirely "out of the picture," if the cause of all secondary otosclerosis is simple, unaided otitis media. Yet they are certainly part of the picture as Nature paints it. What are we to do about them? How account for them?

Obviously, in a large section of the secondary class, there must be influences at work over and above the purely local effect of scar-tissue, influences namely of a constitutional nature.

Turn now to the primary class. We have remarked that the changes in the bone are considered to be solely due to some unknown general dyscrasia. But how vague and indeterminate a conclusion! Surely, for a general dyscrasia to induce those localised islets of osteoporosis in the labyrinth capsule there must come into play something local—to attract the lightning, as it were.

Inasmuch then as in secondary otosclerosis we find a large number of patients manifesting phenomena referable to some general or constitutional condition, so in primary otosclerosis, on the other hand, we have to admit the existence of a large number of patients suspected of otitis media, a phenomenon local in character, and possibly capable of precipitating otosclerosis out of the vague, but none the less actual constitutional dyscrasia.

Thus the clinical examination of many individual cases, as well as the clinical analysis of those same cases in the mass, suggests that in each class there are two ætiological factors acting in co-operation, one general and one local—two originally separate but finally confluent streams of causation.

Thus the clinical findings and the inferences we are drawing from them tend to bring both primary and secondary otosclerosis towards, if not actually into, the same category. And here, doubtless, we have the reason why we experience so much difficulty in assigning many of our cases either to the primary or to the secondary class. They seem to belong to either or both.

Such is the one conclusion to which a survey of the facts has brought us. But there is another interpretation worthy of attention. It is that there may be several different diseases that give rise to the otosclerotic syndrome. First of all, osteoporosis of the labyrinth capsule arising from some unknown cause and independent of any otitis media. Secondly, otitic

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otosclerosis, without osteoporosis, due solely to the action of scar-tissue, in which cases heredity will be absent. Thirdly, there is a large number of cases which it is impossible to classify from clinical phenomena. They may be due to osteoporosis and yet may show signs of otitis media past or present, the latter condition being purely accidental. It is conceivable also that fibrosis of the soft tissue of the middle ear, inducing otosclerosis, may arise without any antecedent inflammatory stage, and without osteoporosis, as a result of some toxic influence unknown to us, in much the same way as chronic interstitial nephritis is produced.

Of the two different interpretations, the first seems to the writer to be more in harmony with the pathological and clinical facts as we know them; but whatever conclusion we come to cannot be anything but tentative, as we are still labouring under the disadvantage of having but an imperfect knowledge of the pathological circumstances. To take one item alone, repeated examination is still necessary before we can determine whether the association of osteoporosis with otitis media, the discovery of which we owe to Fraser, is a mere coincidence or a vital connection.

Dans cet article, l'auteur émet un nombre de conclusions et d'hypothèses provenant d'une étude longue et attentive des observation-prises dans sa clientèle privée et se montant a plus de 600 cas. Il divise ces cas d'otosclerose en primitives, secondaires et naissantes. Il discute sur l'influence de l'âge et du sexe et sur la date de l'apparition de la surdité. L'énoncé est clairement expliqué par les graphiques qui accompagnent le texte, ainsi que les différences distinguant les trois variétés de surdité. Dans une autre paragraphe, il traite de l'évolution de la surdité et il fait remarquer que bien que sa durée moyenne à tout âge est de 3 ans $\frac{1}{2}$, il y a des cas extrêmes dans lesquels un degré de surdité très accentué s'est développé rapidement, et d'autres, au contraire, dans lesquels la surdité a été nettement retardée. La question de l'hérédité est traitée avec soin et les résultats des épreuves de l'audition sont exposés, ainsi que les symptômes généraux et locaux associés.

In dieser Arbeit stellt der Verfasser eine Anzahl von Schluss-folgerungen und Hypothesen auf, denen ein langes und sorgfältiges Studium von über 600 privaten Krankengeschichten seiner eigenen

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Fälle zu Grunde liegt. Er teilt seine Beobachtungen von Otosklerose in (1) primäre, (2) sekundäre und (3) beginnende Fälle ein. Ferner bespricht er die Verteilung des Geschlechtes und das Alter, in welchem sich die Schwerhörigkeit zuerst bemerkbar macht. Aus den vorliegenden graphischen Darstellungen gehen diese Verhältnisse deutlich hervor, sowie auch die Verschiedenheit der drei Arten der Erkrankung.

Ein weiterer Abschnitt der Arbeit behandelt die Schwerhörigkeit. Dabei wird betont, dass wenn die Entwicklung der Krankkeit in jedem Lebensalter durchschnittlich $3\frac{1}{2}$ Jahre beansprucht, sich in einigen besonders schweren Fällen rasch eine hochgradige Schwerhörigkeit entwicklen kann, andererseits bei vereinzelten Beobachtungen die Zunahme der Schwerhörigkeit nur sehr langsam vor sich geht.

Die Vererbung wird sorgfältig studiert und die Ergebnisse der Hörprüfungen angeführt. Ebenso werden die damit verbundenen lokalen und allgemeinen Symptome eingehend berücksichtigt.

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