OTOSCLEROSIS: DOES IT EXIST AS A SEPARATE DISEASE?*

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OTOSCLEROSIS is a name which has been bestowed upon a well-established pathological condition, known with certainty and proven beyond cavil, but from a clinical point of view ill-defined and, I think, *not* proven as such. I propose to examine the etiology and symptomatology, and see whether sufficient proof exists as to the presence of otosclerosis as a separate disease *sui generis*.

Many observers have spent long hours with the microscope, and all agree that well-defined pathological changes are found in the capsule of the bony labyrinth in individuals who have suffered from certain forms of aural disease; from these pathological findings the disease which we are considering has been evolved.

I shall first consider the etiological side and then analyse the symptoms usually accepted as more or less pathognomonic of otosclerosis. These are prolonged bone conduction, loss of low tones, negative Rinne, severe tinnitus, and paracusis Willisii. Finally, I shall make a few observations upon the pathology.

Let me very briefly consider those conditions which are said to act as contributory factors in the causation of this complaint. They are rheumatism, gout, and syphilis, repeated chills, chlorosis, anæmia, the puerperium, rickets, atrophic rhinitis, and aural suppuration. The most important point, however, which one sees fairly well pronounced is that, in all of these cases, sepsis exists as a common factor.

During a good many years of aural practice, I gave, as an experiment to all my lady patients with middle-ear deafness when pregnant, iron and arsenic during the last three months of pregnancy; of those so treated not one was worse after the confinement.

The tendency to seek the causation of one disease from the presence of another is common in medicine, even if not supported by statistical and accurate observation; and I am sure that here we have no such thing. I think that the special trouble is lost sight of in the galaxy of possible causes. I shall

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consider separately, as far as I can, those points in the symptomatology which are generally regarded as found in otosclerosis. It will be observed that not only are there no specific symptoms, but that all the symptoms attributed to this disease occur in other aural affections. Even the so-called triad of symptoms of Bezold are not really conclusive nor pathognomonic.

Etiology.—The two chief etiological factors appear to be syphilis (inherited) and heredity.

Inherited syphilis was considered by Habermann, one of the earliest investigators, as being a fruitful causative factor in the production of otosclerosis, although this contention has ceased to receive the approval of the majority of authorities upon the subject.

I do not consider that heredity merits any very large amount of attention, although by some it is considered almost of paramount importance. Gray, however, says, when speaking of the high percentages which various authorities give as to the influence exerted by heredity: "If these percentages are true, then, judging by the author's experience, otosclerosis must, in Scotland at least, be a much rarer disease than is supposed" (A. A. Gray, *Diseases of the Ear*, p. 313, 1910).

It appears to me that while there is apparently a strong hereditary taint, it does not in reality exist. The influence is indirect, not direct. Accepting this line of argument, it follows that the pathological changes found in the labyrinthine capsule are due to a septic focus existing, in the majority of instances, in the more or less immediate environment of these bony changes, *e.g.* in the faucial tonsils. This occurs in a very large proportion of cases of middle-ear deafness. I suggest, therefore, that the so-called heredity in the causation of otosclerosis is really due to two or more hereditary tendencies. These are intranasal irregularities or deformities and tonsillar disease.

Intranasal irregularities and adenoid hypertrophy, by tending to cause post-nasal congestion and consequent chronic catarrhal conditions, must of necessity predispose the individual to Eustachian inflammatory infections and, consequently, to middle-ear complaints.

With regard to the nose, of course it is not every person with nasal obstruction, by any means, who suffers from middle-ear disease. In fact it is the lesser degree of abnormality and not the more pronounced which causes or predisposes to the complication. This appears especially to be the case when the

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marked obstruction is bony and due to general narrowness, rather than to the compensatory hypertrophy of the soft tissues. Heredity here plays an important part. One has always to remember, however, that in Austria especially, and to a lesser degree in Germany, there are conditions of the upper respiratory tract due to environment, climate, race, etc., which do not obtain in our land, and which in consequence are not only better disregarded, but should be banished from our sub-conscious mind.

Tonsillar trouble is so common and at the same time such a prolific contributory agent in increasing middle-ear trouble, that it can only be considered as producing the necessary septic focus in an ear already affected.

Heredity in otosclerosis is, I think, not proven, unless we are prepared to consider all non-suppurative middle-ear affections not as potential but as actual otosclerotic cases, some of which are checked by nature or art at the more or less early stages. This is an untenable position.

The disease is said to occur even in children, but of this there is not any superfluity of evidence; though we have instances, not too rare, of young people—in my experience usually boys—who have symptoms very similar to those which one is in the habit of considering as the classical signs of otosclerosis. With care the young patients tend to get better or almost well; this cannot happen in otosclerosis! I do not think that paracusis Willisii is frequent in them.

The three following cases emphasise the foregoing condition:—

(1) A boy aged 12, who was stated to have otosclerosis, was brought to see me; he had prolonged bone conduction, negative Rinne and loss of low tones. He improved immensely with intra-tubal injections of one minim of biniodide of mercury emulsion (oily) and oto-massage. Later, he was sent by me to Paris to try re-education, with the happiest results. He is now a Member of Parliament.

(2) A medical student who was told that his medical career was finished, and that he would become quite deaf, was advised to take up farming. He was treated with the same injections together with oto-massage. Now he is conducting a large general practice.

(3) The son of a syphilitic, was chiefly of interest on account of his extraordinarily prolonged bone conduction, which was no less than four times the normal. I took the precaution of asking Dr Edward Law to confirm my observation.

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These three cases show fairly well that they are not to be considered as otosclerosis. The two first are typical of their class.

Symptomatology.—The symptoms of otosclerosis which are usually accepted as pathognomonic are: Firstly, those constituting the triad of Bezold, viz., loss of low tones, prolonged bone conduction and negative Rinne; tinnitus usually severe; paracusis Willisii.

I shall consider these separately and show that not only are these signs found in many different conditions, but even when grouped together they are not confined to cases incapable of improvement.

Loss of Low Tones.—This is a symptom in all cases of obstruction of the Eustachian tube, increasing with the severity of the consecutive loss of hearing, and in this way indistinguishable from the otosclerotic type.

We do not, therefore, gain any information from it. It is well to remember, however, that the class of tuning-fork employed, whether that of Bezold, of Hartmann or of an intervening grade, makes an almost incalculable difference in the result. The greater the stimulus employed, the greater the probability of a reaction.

Unless we are prepared to accept Bezold's classification, we usually consider that there exists a distinct variety of deafness known by many names but well recognised as chronic progressive middle-ear deafness. In these cases loss of low tones is a common factor equally with the otosclerotic ones; again, therefore, we derive no assistance in proving the latter disease to be a distinct entity.

Prolonged Bone Conduction.—Let us ask ourselves the question, when or under what conditions does one find prolonged bone conduction?

The first example is that of acute Eustachian obstruction. Here the word "acute" refers to the rapidity of the onset and not to the class of infection. The word is employed to indicate a sudden obstruction of the tube at its distal or naso-pharyngeal extremity by a swollen state of the mucosa of the cushion and of the naso-pharyngeal space; although one does not as a rule test for prolonged bone conduction when suppuration threatens. In a considerable proportion of the cases of acute Eustachian obstruction, other than those just alluded to as tending to suppuration, bone conduction of sound tends often

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materially towards increase. It must not be denied that sometimes the bone conduction is diminished in these cases.

The prolongation of the bone conduction disappears with the restoration of the patency of the tube, but will return with recurrent obstructions and become permanent if the attacks are too often repeated.

Thus we see that increase of bone conduction is present *ab initio* in all cases of middle-ear affections due to tubal obstructions. It is also present in all middle-ear cases of long standing; in fact, according to Bezold, these cases are all otosclerosis.

An increased bone conduction is frequently noticed in the deafness occurring in old or elderly people; a totally different picture from what is usually found in the so-called senile deafness. It appears to be due to a tendency to rigidity of the ossicular chain.

In advanced middle-ear disease, in those cases termed otosclerosis, and in the deafness found in the later decades of life, one common factor exists, viz., immobility of the stapes. Even in the pseudo-otosclerosis of children it is present to some degree. In suppurative disease of the middle ear naturally it is rarely absent. One presupposes that in otosclerosis the anchylosis of the stapedio-temporal articulation is bony, in other conditions it is not so! When the anchylosis is not of an osseous nature, it may be permissible, in theory at any rate, to regard the patient's hearing as capable of improvement. If the latter does not take place, probably the aurist has acted according to tradition or routine, or possibly the improvement exists in theory alone.

The estimation of bone conduction is not such a simple and uncomplicated procedure as most appear to think. The condition which I have described as the "fatigue period" often causes false estimates. The method often adopted of employing the observer's own bone conduction as a standard is grossly unscientific and full of pitfalls. Why should any man consider that he is necessarily perfect?

It can easily be seen how readily errors of considerable dimensions may and indeed must creep in. Such irregularities tend to complicate issues.

Severe Tinnitus.—Tinnitus being a subjective symptom, increases in severity and in its power to cause mental distress in direct proportion to the sufferer's power of resistance. What one person takes lightly the next finds unbearable, and, in

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consequence, severe tinnitus can be at best but a relative term. In middle-ear cases the great majority suffer at some time or another from tinnitus, which is generally of Eustachian origin if it occurs early in the course of the disease, and if commencing later is due to stapedial immobility, and when it has existed for a period of years is central (*Lancet*, 30th May 1925). When occurring in later life it is often due to arterio-sclerosis, and is frequently of seemingly great intensity.

Negative Rinne.—Rinne's test in itself, as originally propounded, has lost its pride of place, with the increase of knowledge and improvements in instruments. Now it serves to corroborate the results of other tests. A double negative Rinne is of infinitely greater value than an uncorroborated single negative Rinne. But even negative Rinne, if one employs the forks used by Bezold, is not certain. The error due to omitting the possibility of the fatigue period alone invalidates it.

Negative Rinne, like prolonged bone conduction, is common to all varieties of middle-ear disease.

Paracusis Willisii.—I suppose we may some day know for certain the true explanation of this phenomenon, but with that we are not at present intrigued.

Paracusis Willisii is not always a sign of an irremediable state of dullness of hearing. I have stated that with appropriate treatment, one at times gets quite good results. We are probably all in agreement when I say that this subjective sign is invariably found in cases where the stapes is immobile. It is, I think, somewhat rare to find it in suppurative otitis media, so long as active disease of the middle ear and its adnexa continues, but it may appear in instances where the suppuration has ceased for some time. Still, I think that even then it is uncommon; but on this point I am open to correction. I do not think one can say that the phenomenon is a sign of only one disease, nor only found in one disease. If one considers the deafness amongst boiler-makers, or amongst the metal workers in large factories, one finds paracusis Willisii *in excelsis*. But paracusis with nerve deafness, a nerve deafness due to injury of the delicate endings of the auditory nerve, is a condition which, if it has not yet attracted the auro-pathologist, would, I think, prove an interesting field of research.

The Red Reflex. — This phenomenon is treated very differently by writers on the subject. Personally, I feel that

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it is an interesting sign which demonstrates that beneath the pinkish area of aural mucosa a patch of rarefying osteitis exists, and as such is of interest.

Treatment.—The importance of the results of treatment upon the question of the existence or non-existence of the condition under consideration is enormous, and to my thinking almost conclusive.

When it is possible to check the course of the deafness by treatment, then I am sure that the disease is not genuine otosclerosis, although had the course of the disease not been arrested, then the classical pathological changes attributed to otosclerosis would have been present.

It is not possible to overestimate the importance to be attributed to the routine removal of septic tonsils and other septic foci, nor must nasal irregularities be left without attention. The removal of septic foci wherever situated can only redound to the patient's benefit; even if no obvious good accrues to the hearing, the disease is probably checked.

Two provisos are attached to the foregoing: all septic sources must be adequately dealt with, and during the operation for tonsillar removal there must not be great loss of blood.

The Pathological Findings. — These are thoroughly well known; not only have our Scottish colleagues corroborated the observations of the older men, but they have materially increased our knowledge upon the subject. We owe a great deal especially to Gray and Fraser.

The well-known and acknowledged pathological changes in the bony labyrinth are found in both suppurative and non-suppurative diseases of the middle ear. They are not found exclusively in the labyrinth; the researches, until recently, have, however, been confined almost exclusively to the temporal bone.

Briefly, certain changes have been found in the temporal bone, and around them has been erected a disease; is this justifiable? I think not. For not only are there no means by which we can positively differentiate between a relievable or non-relievable middle-ear deafness—the only distinctive point in reality—but with the sole exception of those cases in which one is unable to find the source of the sepsis, all middle-ear cases either improve or the progress of the disease is checked on the removal of the infection, together with the correction of an existing nasal deformity.

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