OTOSCLEROSIS: ITS TREATMENT.*

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BEFORE proceeding to discuss directly the treatment of otosclerosis, it is desirable to have some conception of the nature of the condition which we are to treat. It will not be out of place, therefore, to offer the following suggestions as to the altered physiological conditions of which otosclerosis gives us the clinical picture.

When we examine the subject closely, it would appear that, throughout a large part of its course at least, the deafness of otosclerosis is due rather to the suspension of the function of the organ of hearing than to its actual destruction. The grounds for this belief are several. In the first place, hearing by bone-conduction is often as good or even better than normal. The fact that these patients hear as well or even better in a noise (paracusis Willisii) than normal individuals is another reason for the opinion just given. Again, as is well known, in a very large percentage of cases the hearing power may be strikingly improved for a minute or two by the inhalation of nitrite of amyl. The occurrence of cases of what the present writer has termed otosclerosis paradoxica is yet another ground in favour of the view that the function of hearing is suspended rather than destroyed. It must be emphasised that in all the cases of otosclerosis paradoxica which I have examined, paracusis Willisii was a symptom which the patients definitely stated to be present. It would appear, therefore, that there is a common causative factor in the production of paracusis Willisii and otosclerosis paradoxica.

We can draw certain conclusions from these facts. Thus, in a large percentage of cases, the hearing can certainly be very much improved temporarily. Further, this improvement can take place in the course of a second or two of time, and cannot, therefore, depend upon any new tissue development, such as a process of repair by cell-multiplication, etc. The inference may also be safely drawn that the deafness of

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otosclerosis is not entirely due to the interference with the transmission of sound-vibrations owing to the fixation of the stapes; though doubtless it is in part due to this. For it is hardly possible to suppose that an attack of fainting, or the inhalation of nitrite of amyl can loosen the fixed stapes. Another inference, which is however only probably true, may be drawn from these facts. They would appear to indicate that paracusis Willisii is not due, as used to be supposed, to sound vibrations of large amplitude acting upon the ankylosed stapes so as to let the smaller vibrations reach the labyrinth and enable the patient to hear better. For, since all the cases of otosclerosis paradoxica occurred in patients who were at the same time the subjects of paracusis Willisii, the probability is that the two conditions owe their existence to a common factor. And it is obvious that in the cases of otosclerosis paradoxica, the improved hearing could not be due to any large vibrations acting on the stapes, for there are no such vibrations acting in these circumstances.

Whatever the cause of the deafness in otosclerosis may be, it can be temporarily removed within the course of a few seconds of time, or even of a fraction of a second. Now it is difficult to see how a physiological change in the circumstances we are considering could be manifested so rapidly by any structure other than the neurones concerned in the function of hearing. And it is on this assumption that I offer for your consideration the following suggestion as a possible explanation of the occurrence of paracusis Willisii, and of cases of otosclerosis paradoxica and other phenomena associated with otosclerosis.

In the normal process of hearing, the nerve-impulse passes from the nerve-fibres associated with the organ of Corti to the ganglion spirale. From there they pass by the cochlear nerve to the medulla, and ultimately reach the cerebral cortex. Now in the course of this journey, the nerve-impulse has to pass through various synapses, and it is at the synapses that the resistance to the transmission of the nerve-impulse is greatest. Furthermore, the synapses are points at which the neurones are weakest, and at which they are most apt to suffer damage. This is an important point to remember, but before going on to consider its bearing on the function of hearing in physiological and pathological conditions, it is necessary to make a few general remarks on the evolution

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of the organ of hearing phylogenetically, and on the development of the neurone in the life history of the individual.

The organ of hearing (and in this term we include the outer and middle ear and the cochlear portion of the inner ear, and the nerve-structures concerned with the function of hearing) is relatively speaking of very recent development in the phylogenetic stem. In the higher amphibians, the organ makes its first and very primitive appearance. In most reptiles it is still very primitive, but in the crocodile the cochlea has developed as a definite bent tube. There is a tympanic cavity with a columella connecting the tympanic membrane with the inner ear. In the primitive mammals, the monotremata, there is an external auditory meatus, a tympanic membrane and a tympanic cavity, with a chain of ossicles connecting the membrane with the inner ear. The cochlea is still a tube, but bent to a considerable extent, particularly in echidna. The Eutherian mammals show a remarkable advance in the size of the cochlea. The cochlea is now in the form of a spiral, and the number of nerve-fibres in it is very greatly increased.

Now, associated with this development, which is easily demonstrated and understood, there is another very important element in the evolution of the organ, which, although we know it must be true, is not so frequently remembered, because it is not so directly seen. I refer to the corresponding evolution which must take place in the structures which control the nourishment of the organ—that is to say, the blood-vessels and the nerve structures which regulate their calibre and the blood-pressure within: that is to say, in the vasomotor system. The neurones of the vasomotor system, therefore, which control the blood supply to the organ of hearing are as subject to corresponding variations as that structure itself. Thus the organ of hearing is developed as a whole; and in the writer's work on the subject, the view is suggested that otosclerosis is a degenerative process involving the organ of hearing as a whole. In the following pages, it is my intention to develop this view rather further. It is necessary, however, to say a few words on the development of the neurone in the individual, as it also has a direct bearing on treatment.

In normal circumstances an individual is supplied at birth with all the neurones which he is ever going to possess. Any further elaboration in the development of the central nervous system depends upon changes in the individual neurones, and

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not on their multiplication. When first formed the axons of the neurones are non-medullated and myelination only begins when the neurone commences to function. Furthermore, it has been shown by Flechsig "that the myelination occurs later in proportion as the nerve-fibre is more recent in the phylogenetic history of the animal." After birth, the neurone continues to develop, and this development goes on until the prime of life, that is to say, roughly until between the ages of thirty and forty years of age. During all this time the anabolic changes in the neurone are greater than the katabolic. For several years after this the anabolic and katabolic changes are fairly evenly balanced, and then the latter begin to predominate and a slow retrograde change takes place as age advances. One feature and probably the most important feature of the development of the neurone is the formation of new pathways, and in this way new communications are established with other neurones and new synapses are formed. In this process the axis cylinder grows out, and as it grows it becomes surrounded by the myelin sheath which insulates it, and the synapse comes into existence. It is probable that a receptor substance is present in the synapses which enables the nerve-impulse to pass more easily from one neurone to another.

Now let us consider for a moment the case of an individual, in whom the neurones concerned in the function of hearing have not the same inherent vitality as in his other neurones, or as the same neurones in another individual. In other words, an individual in whom the katabolic processes in the neurones concerned in audition begin to predominate over the anabolic processes at an earlier period in life than is normal. We would not expect such individuals to be very uncommon, because the organ of hearing is of comparatively recent origin in evolution.

So long as the anabolic processes in the neurone predominate over the katabolic, it would function quite normally. But when the former become less active, the katabolic changes begin to manifest their effect and retrograde changes make their appearance, and the function of the neurone becomes impaired. Since the synapse is the weakest spot of the neurone, it is there that these changes will make their first appearance. It is probable, but not demonstrable, that the receptor substance undergoes changes, and the same is true of the medullary sheath and the axis-cylinder, though the changes may not

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occur simultaneously. The nerve-impulses, therefore, can no longer pass through the synapses quite so easily as before, and the functional activity of the organ of hearing is impaired. Consequently it requires a greater stimulus at the peripheral sensory receptor, the organ of Corti, to overcome the increased resistance at the synapses. The dullness of hearing in otosclerosis may thus be explained at any rate in part.

But there are two other symptoms characteristic of otosclerosis which may also be accounted for on the same grounds, paracusis Willisii and tinnitus aurium.

As regards paracusis, few otologists nowadays are willing to accept the old explanation of the phenomenon that the extensive vibrations of the louder sound loosen the stapes in its ankylosed position and thus allow the smaller vibrations of the smaller sound to reach the labyrinth. The objections to it are so obvious that there is no need to discuss them at present.

In the normal individual, the medullary sheath surrounds the axis-cylinder and nerve-ending, and insulates them so that nerve-impulses in one nerve-fibre cannot pass over to the adjacent nerve-fibres. But when retrograde changes occur in the neurones, the medullary sheath undergoes chemical changes, the axis-cylinders may no longer be insulated, and nerve-impulses in one nerve-fibre can pass over to adjacent ones. Now if we apply this fact to the sense of hearing, I venture to think we may find a feasible explanation of paracusis. In a normal individual each nerve-fibre carries its own impulses only, and if these are few in number the total energy available may not be sufficient to overcome the obstruction at the synapses. Even if there are impulses rapid in frequency, passing up other adjacent nerve-fibres, these impulses cannot pass over to the other nerve-fibres in which fewer (and perhaps weaker) impulses are passing. But the case is different when the retrograde changes have taken place in the neurone resulting in degeneration of the medullary sheath, and the axis-cylinder is no longer completely insulated. Impulses can then pass from one axis-cylinder to another. Thus the fibres along which few (or weak) impulses are passing may receive superfluous impulses from the fibres up which the more frequent (and perhaps stronger) impulses are passing, and by means of this reinforcement the nerve-fibres conveying the impulses caused by the smaller sounds may be able to overcome

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the resistance at the synapses, and the smaller sounds to become audible to the patient.

According to the investigations of Trotter, all naked nerve-endings cause the subjective sensation of pain (presumably he means in nerves of common sensation). That is to say, if the medullary sheath is destroyed, so that it no longer insulates the axis-cylinder or nerve-ending, the sensation of pain will occur. The cochlear nerve cannot register pain, being a nerve of special sense. But if in the cochlear nerve, the nerve-endings were exposed through degeneration of the medullary sheath, then we would expect instead the occurrence of the subjective sensation of sound. If, on the other hand, the degenerative changes occurred first in the nerve-ending or axis-cylinder, and then later in the medullary sheath, we would not expect the subjective sensation of sound because the nerve-ending is not exposed. I think this is the explanation of the curious fact that in some cases of otosclerosis, tinnitus is almost completely absent, whereas in many it is one of the most distressing symptoms of the disease.

But besides the subjective symptoms associated with hearing in otosclerosis, there are other changes of which we have definite objective evidence. These are the changes in the bony capsule of the labyrinth, the diminished secretion of wax in the external auditory meatus, and the diminished sensitiveness of the tympanic membrane. The last mentioned condition is also to be associated with the sluggish response of the blood-vessels of the tympanic membrane to mechanical irritation.

As stated above, these changes, as well as the symptoms of dullness of hearing, paracusis and tinnitus taken together appear to indicate the existence of a degenerative change in the organ of hearing as a whole. Whenever the organ of hearing is called upon to function, the blood supplied to it must be increased if it is to function satisfactorily, and this increased blood supply is regulated by the neurones of the vasomotor system which have been developed in the course of evolution in direct relationship with the various structures concerned in the function of hearing. If the neurones governing this vasomotor mechanism should be defective, then the tissues concerned must suffer. The organ of Corti, the nerve-fibres of the cochlear nerve, the bony capsule of the labyrinth, the tympanic membrane, and indeed all the structures of the organ of hearing will be affected, though it may be in varying degrees.

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The effect of this want of response of the vasomotor system of the organ of hearing will vary according to the tissues involved. Speaking from the *physiological* point of view, the activity of all their functions will, of course, be impaired. But from the *anatomical* point of view, the effect will be different according to the extent to which cellular changes would normally occur. Thus, in the highly specialised neuro-epithelium of the organ of Corti, cellular changes do not take place at all. The cells would not die, because the blood supply is quite sufficient to support their existence. All that happens is that they are unable to function properly because they do not receive the increased nourishment necessary for that purpose. So also with the nerve-cells of the ganglion spirale; they would still remain alive, but, functionally, they would react imperfectly to the normal stimuli. The same would be partially true of the ceruminous glands in the external meatus. The cells of the specialised epithelium of these glands probably do change from time to time, and there may quite well be a slight visible anatomical change, corresponding with the physiological diminution in the secretion of wax in otosclerosis. This is a subject which has not yet, so far as the writer knows, been investigated, but it would not be very difficult to carry out. But when we come to a tissue such as bone, the circumstances are different. Under normal physiological conditions, rapid cellular changes are continually going on in bone. Osteoclasts are constantly making their appearance and removing the tissue, to be succeeded by osteoblasts, which lay down the new bony tissue. These in their turn disappear, and are replaced by new ones, and so on. In these circumstances, it would be surprising if a change in the nutrition to the part involved did not bring about changes in the texture of the bone. Furthermore, these changes would be most likely to occur, or at least to make their appearance first in the region where the bone is most functionally active. In the case of the organ of hearing, this region is in the walls of the oval window in its anterior portion; because it is here that the movements of the stapes reach their greatest amplitude. This is exactly the region which is by far the most frequently affected in otosclerosis, and, indeed, in many cases it is the only spot at which the bony change is found.

It is interesting to note that, as was shown by the writer, the change in texture of the bone may vary within wide limits,

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while the position at which the change is found is the same. Thus the new bone may be in excess of that absorbed, as is most usually the case, or it may be less in amount than that absorbed, or there may be absorption of bone only, and no bone deposited at all.

According to the view suggested in this paper, the improvement in hearing, which occurs in otosclerosis paradoxica, would be due to the increased nutrition to the organ of Corti and neurones of the cochlear nerve resulting from the increased blood supply brought about by a change in the vasomotor reflex arc.

It had been the writer's intention to make some observations on the remarkable occasional association of otosclerosis, fragilitas ossium and blue sclerotics. This relationship has been observed by several observers, among whom recently is Cleminson. Limits of space and time, however, will not allow further reference to that interesting aspect of the subject.

Summary.—Summarising the matter, the writer would make the following suggestion. Otosclerosis is a degenerative process affecting the organ of hearing as a whole. (The semicircular canals and vestibule are not parts of the organ of hearing, and are therefore excluded.) In some cases, the nerve-structures are relatively the most affected, and in others the bony or connective tissues; and the signs and symptoms may, therefore, vary within considerable limits. It is probable that this degenerative process results from an inherent defect in the neurones concerned in the function of hearing. This includes the neurones of the vasomotor system, which regulate the blood supply of the organ, as well as those concerned in transmitting nerve-impulses from the organ of Corti to the cerebral cortex.

This view explains the occurrence of tinnitus, of paracusis Willisii, and of otosclerosis paradoxica. It explains the age incidence of the disease, and its extremely gradual progress, as well as the fact that the condition is so frequently inherited.

Treatment.—In accordance with the view suggested then, the treatment of otosclerosis should be directed towards maintaining the health and functional activity of the neurones.

Local treatment in my own experience is entirely disappointing. Inflation per catheter produces no improvement, and the same is true of the injection of drugs, etc., into the

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middle ear through the catheter. Vibratory massage may produce an improvement, but such when present lasts only for a few minutes, or at most for a few hours. Operative procedures on the nose, such as removal of the septum, etc., are valueless in otosclerosis, however useful they may be in cases of chronic middle-ear catarrh and Eustachian obstruction.

The most important element in the treatment of otosclerosis is concerned with the general condition of the patient. All otologists must have observed that, in those individuals who suffer from the disease, the rate of progress of the deafness depends to a large extent on the general health and the circumstances in which the patient lives and works. It follows from this that the first problem is to find out in each case if there is any particular bodily condition, or any error in the patient's way of living, which is adversely affecting the hearing. Sometimes this is not difficult, but frequently even careful examination fails to detect any error in the general health or the method of living of the patient. It may not be out of place, therefore, to refer to some of the conditions which the writer has found to affect the course of the deafness unfavourably.

Anæmia.—Chlorosis, the anæmia of young women, is the condition in which otosclerosis appears to arise most frequently. Now, although there is no doubt that the chlorosis of young women is less apparent now than it was two or three decades ago, yet in its milder manifestations, I feel sure that it is still a common disease. Frequently, the breathlessness is so comparatively slight that the patient does not consult a physician about it. Be this as it may, I have repeatedly found that otosclerosis makes its appearance very frequently in young women when they are suffering from chlorosis. It is important to observe that the chlorosis is very often slight in degree, and is apt to be missed by the physician unless he is on the outlook for it.

If, therefore, there is any evidence of chlorosis, even in its mildest manifestations, the condition should be treated in the usual way by the administration of iron. Sometimes the patient cannot assimilate iron when it is given by the mouth, and in such cases the drug may be given by hypodermic injection. It should be continued for a long period. Of all the cases of otosclerosis, those which arise in the course of anæmia in young women are, in the writer's experience, the

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most satisfactory to treat. Perhaps it would be better expressed in a negative way by saying that they are the least unsatisfactory. The progress of the deafness is sometimes arrested for many years, and the tinnitus considerably diminished. In a few cases, there is even a slight improvement in the deafness.

Pregnancy.—The evil effect of pregnancy and the puerperium upon otosclerosis is so well known that there is no need to waste many words upon the subject. Any woman who suffers from otosclerosis must avoid pregnancy if she wishes to preserve what hearing she still has.

Septic Foci.—Any septic focus in the body from which toxic products may be absorbed, has a bad effect upon the course of the disease, and it is important, therefore, to search most carefully for any such possible source of infection. Such a focus may be found in the jaws, in the tonsils, in the nasal sinuses, in the appendix, or elsewhere. Needless to say, any such focus of infection should be removed. It is interesting to observe that in such cases the beneficial effect upon the tinnitus is usually more noticeable than that upon the hearing, though even in regard to the latter, the progress of the deafness is usually slower than before.

Exhaustion.—Exhaustion, whether of the body or mind, has a very deleterious effect upon otosclerosis, and usually patients themselves remark upon the fact that when they are tired not only is the hearing worse, but the tinnitus also is more distressing.

Judging from the statements of my own patients, I should be inclined to say that mental exhaustion, especially that resulting from worry and anxiety, and want of sleep, has a worse effect than the physical exhaustion of muscular activity. Want of sleep has a particularly bad effect. It follows, therefore, that the victim of otosclerosis should, so far as possible, lead a life free from worry, and an ample allowance of sleep should be prescribed.

Tinnitus.—Tinnitus, which so frequently accompanies the deafness of otosclerosis, is sometimes so severe that it is more distressing to the patient than the deafness itself. It is, moreover, a symptom which is usually difficult to treat. In general, it is true of the tinnitus, as of the deafness, that it varies in intensity according to the general condition of the patient. Thus it is more noticeable and more annoying when the patient is exhausted especially in mind. Patients nearly

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always remark upon the fact that it is worse when they are in bed at night, but this is only because the surrounding silence accentuates the subjective sensation. The general treatment of the tinnitus, therefore, is the same as that of the deafness. That is to say, the general health must be maintained at the highest possible level, all exhaustion must be avoided, and sleep should be indulged in freely. In regard, however, to this last factor, it is most unfortunate that in severe cases the tinnitus itself prevents the patient from obtaining the sleep which is so essential in the treatment of the disease.

Many drugs have been recommended for the relief of tinnitus, and it is probably the experience of most otologists that none of them are satisfactory. The most commonly employed are the bromides. The great objection to the use of bromides is the fact that in order to bring relief from the tinnitus, the drug must be given in such large doses that it produces depression and other unpleasant symptoms. Whenever possible, bromides and other drugs should be avoided in the treatment of tinnitus, and the patient should be encouraged to train the mind to ignore the symptom by an effort of will, or by occupying the attention with some absorbing mental distraction.

The most serious effect of tinnitus is the sleeplessness, which is apt to occur when the noises are very severe in degree. Fortunately such cases are not very common. In such circumstances, the sleeplessness must be overcome by some means or other. A good plan is to give one large dose of bromide shortly before bedtime, rather than a number of small doses through the day. Even two drams of bromide of potassium may be given at bedtime, and a good night's rest obtained. The writer has found, however, that in these cases veronal is a more satisfactory drug to employ, as it is not followed by the depression which follows upon the administration of large doses of bromide. It must be remembered that patients vary in their susceptibility to veronal, and the first dose should not exceed five grains. It will usually be found that ten grains are enough to secure a good night's rest.

In the rare cases in which the tinnitus is so severe as to make life almost unendurable, it is justifiable to consider the advisability of destroying the cochlear nerve in the modiolus. The operation is a serious one; it is attended by considerable shock, and it entails a possibility of death from meningitis

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Moreover, in order to relieve the tinnitus, it may be necessary to operate on both cochleæ, with the result that if the patient recovers, he will be absolutely deaf. It will be understood, therefore, that the operation is very rarely justifiable, and is rarely performed. Still, there are cases in which it should be undertaken.

The treatment of otosclerosis by the method known as re-education of the hearing, has been practised in recent years by some otologists, both in this country and abroad. I am afraid I cannot speak with any great experience of this method. In the cases in which I have known it tried, the results were rather disappointing in the majority, though there were a few patients who were of opinion that some slight degree of improvement had occurred.

Surgical Treatment. — It has been suggested that the hearing power in otosclerosis might possibly be improved if an opening were made in the bony wall of the labyrinth to compensate for the loss of function at the oval window which has been destroyed by the fixation of the stapes. Holmgren, I understand, has actually performed such operations, but I have not been able to find any published results as regards their effect upon the hearing. Dr Fraser, however, in a personal communication has described to me two cases in which he operated. In one the result was unsuccessful, but in the other a considerable degree of improvement was obtained. I have had no personal experience of these operations, but, fortunately, Dr Fraser is with us to-day, and I shall leave it with him to give us the details of the cases, and the conclusion he has drawn from his experience of them.

In general, it may be said that operations of this kind when successful would improve the hearing to the extent to which the deafness is due to the mechanical obstruction of the fixed stapes. In so far, however, as the deafness is due to a defect in the nerve-structures, they would have little effect.

In conclusion, the writer would like to refer to another disease, whose occurrence may be explained on similar grounds to those given above for the occurrence of otosclerosis, viz., atrophic rhinitis. Atrophic rhinitis, or true ozæna, presents many points of resemblance to otosclerosis. There is the same diminished sensitiveness of the parts, the same diminution

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of secretion (in the case of rhinitis this leads to stagnation of the secretion, and consequent putrefaction). There is also the same failure of the vasomotor mechanism to respond to irritation, and the same diminution in acuity of a special sense. The age incidence of the two diseases is approximately the same, and they are commoner in women than in men. Although by no means always hereditary, both diseases show striking evidence of inheritance in many cases.

The writer would, therefore, look upon both diseases as being the result of an inherent defect in the neurones concerned in the functions and also in the nutrition of the ear and of the nose respectively.

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