OTOSCLEROSIS: ITS CLINICAL ASPECT.*

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OTOLOGISTS will always take a lively interest in a discussion on otosclerosis.

The first macroscopic descriptions were given, as you are aware, by Valsalva in 1704, Morgagni in 1767, and Meckel in 1771. In his descriptive catalogue (1857) your prominent countryman John Toynbee mentions several cases. Since that time the names of the best otologists of all nations have been connected with this chapter of otology. I may quote Troeltsch and Politzer, with their pupils, and among others, Gray, Wittmaack, Otto Mayer, and my revered chiefs, Bezold and Siebenmann.

My contribution to this discussion will be strictly confined to the clinical aspect of otosclerosis. It is based on a survey of all my cases, observed in private practice, whereas the histological particulars are founded on some slides in my collection.

As a pupil of Bezold and Siebenmann, who were especially interested in the clinical aspects, diagnosis, and pathology of otosclerosis, I believe I am able to offer an entirely uniform series of investigations.

In the last nineteen years I have observed 835 cases of otosclerosis, of which I was able to follow up about one-fourth during several years. I was only able to see the others once or twice, for, as you are aware, incurable otosclerotic patients have a striking but easily explicable passion for going from one specialist to another.

Incidence.—In regard to the relative frequency of this disease, my investigation shows that among 1000 ear patients, 208 were suffering from otosclerosis (*i.e.* 20 per cent.), whereas Bezold observed this malady only in 7 per cent. of his patients. The recent statistics of Shambaugh give 30 per cent. There seem to be no other similar statistics, which is unfortunate, as they would be highly interesting. Differences might possibly be found in various countries, according as the population had maintained its pure racial characteristics or had absorbed a

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larger amount of alien blood. This would help to settle the question whether we are dealing with a variety as recorded by Gray.

Sex.—The greater frequency of the disease in females is confirmed by my statistics of 835 patients. Of these, 547 were females, which is a ratio of 1 : 1·8. This figure is the same as Bezold's (1 : 1·8), while Gray had 2·4 female patients to one male.

Age Incidence.—The first symptoms are usually observed between the ages of 20 and 30, as may be seen on the subjoined table.

I need hardly say that a certain amount of caution is necessary in making use of these figures, as they were furnished by the patients themselves; the early phases of the disease are frequently almost imperceptible. Nevertheless certain inferences may be drawn from the table. It will be seen that in the majority the disease commences between 21 and 25 with hardly any appreciable difference in the sexes. Fifty per cent. of all the patients trace the beginning of deafness to the period between the 16th and 30th year. Only in 13·7 per cent. of the cases did deafness manifest itself before the above ages, whereas on the other hand, cases of otosclerosis after 50 are very rare. There was only one patient over 70 years of age with otosclerosis, who stated that his hearing had been hitherto normal.

Heredity.—Since otosclerosis has been differentiated from other diseases of the ear, hereditary and familial occurrence has been known. Again I would point out that statistics on this subject must be accepted with some reserve for the reasons before mentioned. In addition, some patients have no knowledge of their family history, or refuse to give definite information. It is of course quite possible that deafness in other members of the family was due to other causes. Hence the statistics might have to be modified.

Heredity, and simultaneous familial occurrence were found in 141 cases; hereditary alone in 270, familial in 78, together 58·6 per cent. (489 patients). Bezold observed 52, Denker 40, and Siebenmann 35 per cent. The pedigrees of such families are particularly interesting in connection with the study of the laws of heredity, as may be seen by those published by Körner, Hammerschlag, Gray, and Albrecht.

All investigations made up to the present seem to show

Incidence of Otosclerosis in Respect of Age and Sex.

Age . . .	0-10.		11-15.		16-20.		21-25.		26-30.		31-35.		36-40.		41-45.		46-50.		Over 51.		Uncertain.		Total.
		%		%		%		%		%		%		%		%		%		%		%	
Male . . .	13	4·5	33	11·5	45	15·6	53	18·4	46	16	39	13·5	25	8·7	11	3·8	4	1·4	4	1·4	15	5·2	288
Female . . .	25	4·6	44	8	90	16·5	107	19·5	92	16·8	58	10·6	47	8·6	26	4·8	7	1·3	12	2·2	39	7	547
Total . .	38	4·6	77	9·2	135	16·2	160	19·2	138	16·6	97	11·6	72	8·6	37	4·4	11	1·3	16	1·9	54	6·5	835

Age . . .	0-10.		11-15.		16-20.		21-25.		26-30.		31-35.		36-40.		41-45.		46-50.		Over 51.		Uncertain.		Total.
		%		%		%		%		%		%		%		%		%		%		%	
Married . . .	5	1·8	10	3·6	32	11·5	47	16·8	56	20	36	12·9	37	13·3	24	8·6	6	2·2	10	3·6	16	5·7	279
Unmarried . .	20	7·5	34	12·6	58	21·6	60	22·4	36	13·4	22	8·2	10	3·7	2	0·7	1	0·3	2	0·7	23	8·5	268
Total . .	25	4·6	44	8	90	16·5	107	19·5	92	16·8	58	10·6	47	8·6	26	4·8	7	1·3	12	2·2	39	7	547

that heredity in otosclerosis is not uniform; in addition to the dominant heredity which is prevalent, there may be another, possibly a recessive way, as shown by the—unfortunately too few—family trees of blood relations. This irregularity in the hereditary process might lead to the inference that otosclerosis may not represent a biological unity, which seems to fit in with the views of some authors, based on their studies of the pathological aspect.

The frequency of heredity is the main point of the hypothesis, that otosclerosis is a constitutional disease. This is probably right, but anomalies of the vegetative nervous system, the chemical composition of the blood, and the endocrine glandular function have also to be taken into consideration. Writers on the subject, however, are not agreed as to the valuation of these phenomena in the symptomatology of this disease.

Cases of otosclerosis combined with struma and endemic degeneration are frequent in Switzerland. This is not surprising, seeing that most of our patients come from districts with endemic cretinism. Signs of endemic degeneration were, however, not more frequently observed in otosclerosis, nor was otosclerosis itself especially frequent in endemic deafness. In my series of 28 cases of endemic deaf persons there were only four otosclerotic changes similar to those observed by Alexander in one of his cretins.

The theory of the constitutional nature of otosclerosis is further supported by its frequency in both ears. Among my patients I met with only 114 cases, *i.e.* 13·6 per cent. in which the affection was absolutely one-sided. Almost the same proportion was found by Bezold (11·2 per cent.). It may be objected that at the time of examination the disease had not yet reached the stage of ankylosis in the healthy ear, and that this might occur subsequently. I may reply that I myself examined a great number of patients who, even after years, remained sound in one ear. There are cases in my collection in which the most precise microscopic examination shows the disease only in one ear.

Progress of the Disease.—The progressive character of otosclerosis is generally accepted; 95 per cent. of my patients complained of increasing deafness. On the other hand, I was not able to find absolute deafness among them, although no doubt it occurs as soon as both windows are affected.

The rate of the progression, however, differs greatly, as I have been able to ascertain during many years of practice. After the initial diminution of hearing an almost stationary condition not infrequently sets in, as if the process had been exhausted or arrested. In 36 cases (4 per cent.) I obtained data of this kind which were confirmed by repeated clinical examinations. An anatomical explanation of this fact may be found in the different stages of bone-transformation, which ends in lamellar bone tissue.

Tinnitus.—Ear noises were complained of by 68 per cent. (563) of my patients. In very many cases the disease started with these noises and deafness set in only later. I do not intend in this place to deal particularly with the different kinds of noises, but will merely state that they are often the principal complaint and lead to psychic depression, especially in sensitive patients.

Vertigo.—Only 40 patients complained of dizziness, *i.e.*, 0·5 per cent., whereas Bezold records 22 per cent. and Shambaugh 5 per cent. My patients with this complaint were mostly elderly persons, so that possibly other causes, such as senile decay and arteriosclerosis, etc., may have played a part. There were, however, no distinct anomalies of the vestibular function.

Otoscopic Appearances.—In 71 per cent. of the cases the ear-drum was perfectly normal; in another 20 per cent. there was a distinct reddening of the promontory, the so-called Schwartze sign. The other 9 per cent. had changes (as residues) on one side with deafness in both ears, or only slight alterations. But in these cases the history of the disease proved that there could be no doubt of the diagnosis.

Influence of Pregnancy.—All authorities seem to be agreed as to the dangers of gravidity: I myself hold Gray's views that these influences have been exaggerated. Of my 547 female patients 264 were unmarried. If, therefore, the influence of child-bearing was so deleterious, this would be shown by a greater proportion of married women (the difference being only 2·4 per cent.). The table, however, distinctly shows that otosclerosis started earlier in unmarried females, so that half of them were distinctly hard of hearing at the age of 25. This may be one of their reasons for not marrying. Among the married women we find that even 20 per cent. trace their deafness to the years between 25 to 30, by which age it may

be supposed they have already had children. This proportion is higher than the average of 16·6 per cent. It is therefore quite possible that child-bearing was in these cases one of the causes of the development of the disease.

In reply to a direct question we learned that in 40 cases, *i.e.* 16·5 per cent., pregnancy had had no harmful effect, whereas in 46 per cent. a change for the worse had been noticed, but mostly after a number of births. It is obvious that the physician will exercise great prudence in giving advice as to the dangers of pregnancy and confinement, for even more extended statistics cannot lead to a definite conclusion. In addition, 48 per cent. had no hereditary tendencies. Moreover, our knowledge of the exact laws of heredity is not precise enough to enable us to influence the future and happiness of our patients.

Diagnosis.—I have always diagnosed otosclerosis on the principles laid down by Bezold and Siebenmann. Bezold's triad may be considered as established, and histological examination of intravital tested cases have confirmed its reliability. By X-ray examination this disease does not yet show any distinct change in the labyrinthine capsule. I am fully aware that the diagnosis of otosclerosis can only be made in cases in which there is ankylosis of stapes. The decisive factors were the course of the illness, the history, the aspect of the ear-drum, and above all the result of the tuning-fork tests. Even if the ear-drum is not quite normal, I should not hesitate to make this diagnosis. Experience has taught that even people with normal hearing may have distinct anomalies of the ear-drum. However, cases of *progressive* deafness with residues, but without perforation of the drumhead, are extremely rare. A number of so-called chronic adhesive processes might possibly be attributed to otosclerosis.

The loss of perception for high tones does not interfere with this diagnosis, as Siebenmann proved that the explanation of such localised defects of hearing is to be found in pathological areas of the basal coil, which is in full agreement with the Helmholtz theory.