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BRIEF CONSIDERATION OF CERTAIN RECENT VIEWS REGARDING OTOSCLEROSIS.*

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The study of that form of progressive hard-hearing known under a variety of names but most generally referred to as otosclerosis, is fraught with peculiar and to a large degree, uncontrollable difficulties. The affection is not like that of adenoids, something that can be got at and easily removed for examination. For accurate investigation, the microscopic examination of the temporal bones of those in whom the disease has been diagnosed during life is essential. The obtaining of these is difficult anywhere, as there is nothing *per se* in the disease which is wont to occur in the young, to hasten the death of the person affected, and especially difficult in this country where post-mortem material is so exceedingly hard to secure. An equally essential requirement, possessed by few otologists, is that of being well grounded in pathology. As a result, the total number of cases investigated is not large and these are almost entirely limited to those studied by our German confreres.

It is with no claim to any expert pathologic knowledge that I have prestimed to present another paper on this subject, but rather that as a result of some personal study and of an opportunity recently to meet a number of those who have given especial attention to it I wish in a judicial way to weigh and consider briefly the various views which have been advanced regarding the nature of this dis-

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ease. I am fully aware that in the minds of many of you the subject appears hopeless of solution. Indeed, in one of the best short papers on the subject which has recently appeared, that from the pen of a member of this Society, Dr. Sheppard, the pessimistic view held by certain foreign writers was advanced, i. e., that even the diagnosis of otosclerosis is impossible.

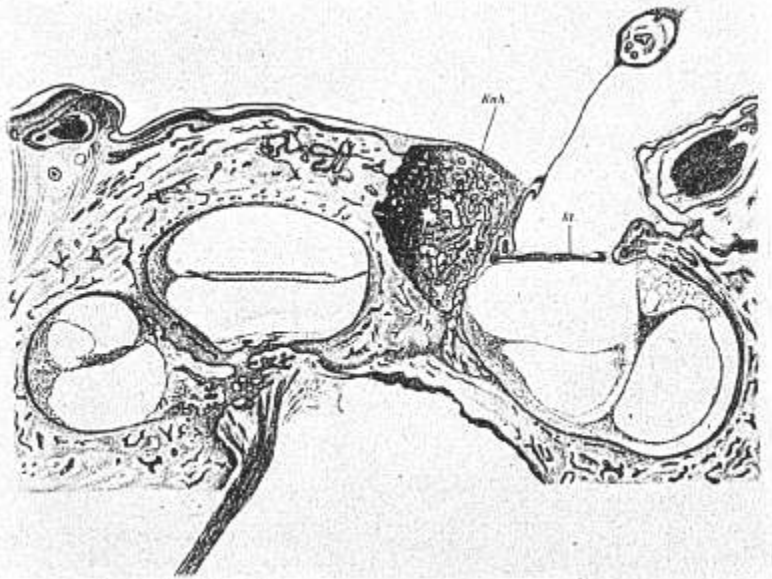


Figure 1—Case 1: Horizontal section through the region of the oval window. A small area of new-formed bone on the anterior border of the oval window. The mass is light in color near the window, darker in color near the cochlea. The last are the recent, the first are the older portions. The mass extends somewhat into the window niche, but does not involve the stapes nor the annular ligament. There is no stapes ankylosis. In the cochlea can be seen atrophy of the organ of Corti, and an atrophy of the connective tissue growth in the spiral ganglion. The atrophy of the acoustic nerve is the result of the tumor which existed in this region. The cochlea, the sacculus and the utricle are filled with hyaline. (From Manasse's collection with permission.)

For a proper consideration of the subject, a brief reference to its history is necessary. The number of investigators is few, and the total number of cases, microscopically studied, collected by Denker up to 1904, is limited. The term otosclerosis, as you are aware, was first used by Joseph Toynbee, in 1857, in his descriptive catalogue of 1149 specimens, where he describes 126 cases of union of the stapes in the oval window. This term was used for many years to describe such an obstruction to the sound-waves whether of a mem-

braneous or bony nature. But it was not until 1885 that it was demonstrated macroscopically and physically by Bezold that the loss of hearing for low sounds was the result of such rigidity in the oval window. The first microscopic examination of a diseased temporal bone in a case of otosclerosis was made by Katz in 1890. He was followed by Habermann who, in 1892, reported his first case. In 1893, Bezold and Scheibe reported their first case, and in the same year, Politzer, in this country, and if we are not mistaken before this Society, reported sixteen cases, all observed by him during life and studied histologically post-mortem. In 1894, Bezold reported his second case; in 1898, Siebenmann reported four cases. In 1901, Scheibe reported two cases, in the same year Habermann reported a second case, and Katz a second case, while in 1903, Habermann reported ten temporal bones observed in six patients, a total of thirty-four cases. Since then Panse has reported two cases, Joergen Moeller one case, Bruehl five cases, Schilling one case, Otto Mayer one case, and most recently Manasse has described seventeen temporals in ten patients.

The early view in regard to the etiology of the affection was that it proceeded from a disease of the middle ear. This is undoubtedly correct in the cases of Katz, where both from the history and clinical picture, a previous chronic suppuration of the middle ear was evident. In those of Scheibe and of the ten cases of Habermann there is likewise a clear history of middle-ear disease. Politzer was the first one to advance the view, which he still maintains, that the disease is a primary affection of the labyrinthine capsule, originating not in the periosteum but in the bone itself. As a result, new bone-tissue is developed which gradually presses out the old bone and advances to the oval window and the stapes, leading finally to a complete stapes ankylosis. Siebenmann was the first to make a careful study of the affection and he is of the opinion that the spongifying process does not proceed from the periosteum, nor yet does it develop as a primary affection of the bone. From the cases which he studied he is of the opinion that the starting-point is to be found on the border between the labyrinthine capsule, which is primarily developed out of cartilage, and the connective tissue bone (*Bindegewebesknochen*) secondarily developed from the periosteum, and that the earliest stage of the disease is to be found in a lacunary resorption of the bone by means of the Haversian canals which become dilated into large lymph spaces, although at the same time in other areas there is apposition going on by means of osteoblasts. The spongification in his opinion is the last stage

of a growth which does not normally occur in the temporal bone, but is the rule in other bones, and follows here the entire disappearance of the cartilage present in the intercellular spaces and on the border of the oval and round windows and also by the growth of compact bone into the osteoid tissue; and finally into the spongy tissue.

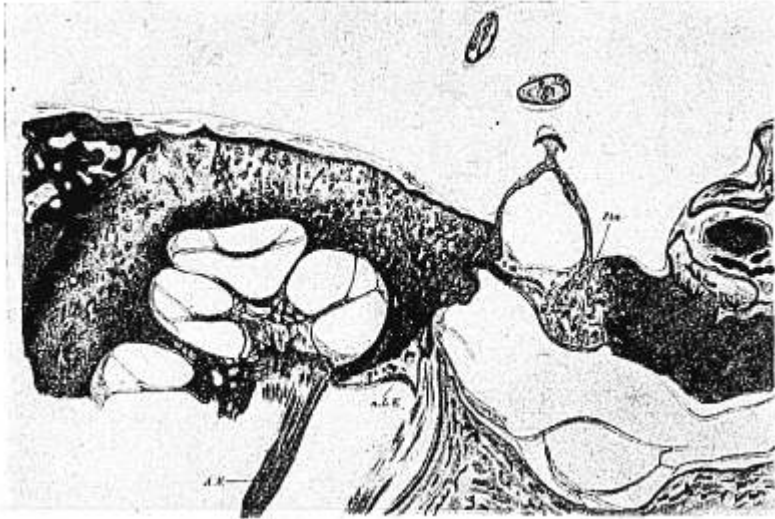


Figure 2—Case 8: Advanced disease of the labyrinthine capsule. Almost the entire bony cochlea is transformed by the osteitic process. Only a small spot shows normal labyrinthine bone, N. L. K. We can see with this weak enlargement that the larger part of the new-formed bone is very old, light in color, poor in blood vessels, and formed out of dense sclerotic bone tissue. Only on the inner periosteum can be seen a few dark-colored recent portions. K. N. H. A small area of new-formed bone. Complete stapes ankylosis, with the foot-plate strongly involved in the new bone-formation, particularly on the posterior border. Degenerative atrophy in the membranous labyrinth, both portions of the acoustic nerve very thin. A. N. c. Atrophic cochlea nerve. A. N. b. Atrophic vestibular nerve. (From Manasse's collection with permission.)

Habermann, who studied the subject with equal care but with specimens derived from patients who had suffered from middle-ear disease, believes that the affection proceeds along the vessels from the middle-ear into the inner-ear. Finally, Denker, as a result of a study of the collected literature as well as personal investigation, in 1904, reached the conclusion that the disease might be in some cases secondary to middle-ear disease, but was usually of a primary nature, and recognized at that time the difficulties which have since presented themselves, due to the variable location in the labyrinthine capsule of the diseased areas. Since Denker's mono-

graph on the subject, the views of many investigators have been materially altered as the result of a masterly study of the subject by Manasse, of Strassburg, entitled, "Die Ostitis chronica metaplastica der menschlichen Labyrinthkapsel." Manasse previously in Basle advanced, but only last year published, this result of his studies in the examination of seventeen temporal bones derived from ten cases, a study extending over twenty years. As in the case of Siebenmann, the contributions of Manasse are particularly important because of the fact that he is a trained pathologist.

The views of Manasse it will be necessary to state somewhat particularly:

1. In the first place, he confirms the observations of Siebenmann and Habermann, that the predilection area for the diseased process is the anterior border of the oval window. This, however, as they have also pointed out, is not always the case. Three times the porus acusticus internus was involved, twice the round window, three times the apex of the cochlea, and once the anterior portion of the labyrinthine capsule and the posterior semi-circular canal.

2. Of particular interest are Manasse's views regarding the histology of this bone affection. He believes that the "disease is virtually a transformation of the labyrinthine capsule. The compact bone is to a greater or less extent replaced by new (foreign) bone substance which is everywhere sharply contrasted with the old bone tissue. The new bone differs in its structure according to the age in which one finds it. The chief difference, as compared with the old bone, is that instead of compact bone, a system of bony trabeculae is to be seen between which there lies a greater or less number of large open spaces. These spaces are composed in the younger parts of a tissue very rich in cells, which in the older parts becomes fibrous and at times is replaced by fat tissue or rarely even by 'Schleimgewebe.' The entire newly-formed tissue would be spoken of in general as *spongifying bone*. Later the open spaces become smaller. They contain, however, very little of real marrow, only blood vessels. The bone becomes much harder and denser, and develops a regular lamellae system, so that without doubt it can be regarded as compact bone, and can pass very rarely into a firm sclerotic bone entirely lacking in marrow and with few blood vessels."

Again quoting Manasse: "With exceptions, two forms of bone will be met with in the diseased areas, both representing in the majority of cases the early stages of the disease. First, small open

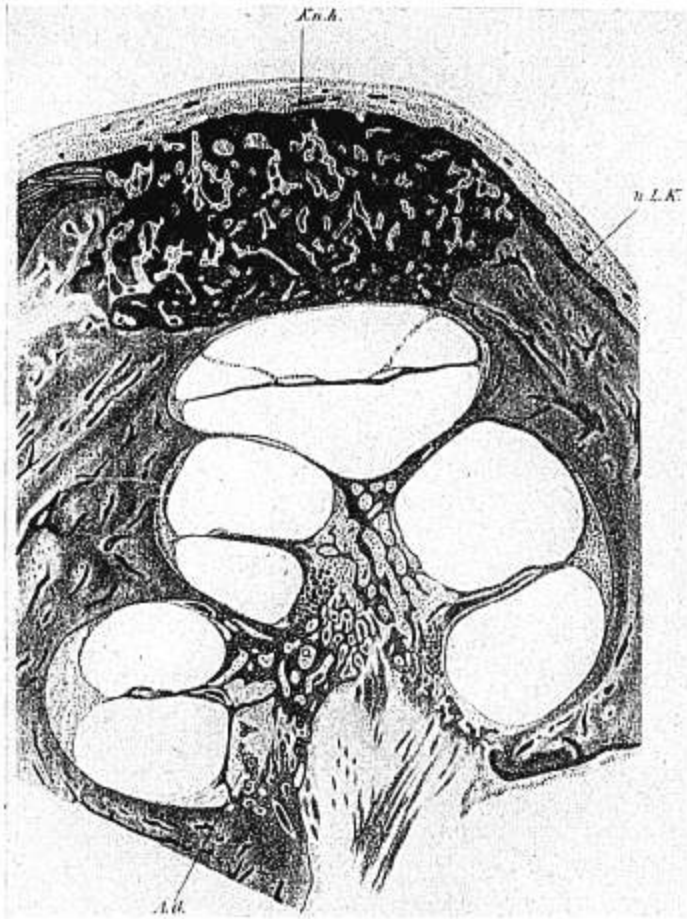


Figure 3—Case 10: Right. A section through the cochlea; circumscribed pathological area of bone. (K. n. h.), in the capsule extending to the cochlea. The transformed bone involves the entire thickness of the capsule from the periosteum of the middle ear to the inner periosteum of the cochlea. It is formed almost exclusively of recent newly-formed bone as can be seen from the dark color. In the membranous labyrinth, degenerative atrophy, in the organ of Corti, the spiral ganglion and in the fine ramifications in the cochlea. The sinking of the Reissner's membrane is not pathological but post-mortem. N. L. K. Normal labyrinthine bone. (From Manasse's collection with permission.)

spaces with pronounced bony trabeculae, coloring very bright with hematoxylin, and second, larger open spaces taking a darker stain and with fewer and smaller areas of bone between. The bright are the older diseased areas, the dark the newer. Neither is composed of compact bone, and as Siebenmann pointed out, both are lacking

in the intercellular spaces containing cartilage, which are characteristic of the normal labyrinthine capsule. The newer bone tissue shows between the thin, bony trabeculae large open spaces containing a tissue rich in round cells, cylindrical cells, and osteoblasts as well as osteoclasts. There are no large blood vessels, only capillaries, no compact connective tissue, no myoblasts nor red blood corpuscles, no fat tissue, and only a small amount of intercellular substance. The picture, as a whole, is one of granulation or new connective tissue, not of lymphoid marrow. Certain spots in the bone substance which take a deep blue color, are transparent, contain only small, three-cornered bone cells such as is met with in immature bone, and show a peculiar sieve-like rarefaction, the round, long or jagged holes of which are each filled out by a single cell. These holes often come together forming canals. They also contain capillaries and empty into the larger open spaces. Osteoblasts and osteoclasts are never met with in the narrow spaces of these deeply-colored bones. The older form of the new-formed bone is composed of larger bony trabeculae which take a brilliant red color and which form a distinct network. The bony substance shows both jagged and plump bone corpuscles, but no lamellae system. The open spaces contain few cells, often only one large, wide, thin-walled blood vessel, but also a fairly soft, connective tissue with lymphocytes, spindle-or star-shaped cells, and several vessels. This fibrous marrow passes occasionally over into the fat marrow and at times into the yellow marrow. Osteoclasts are met with more often in the larger areas than in the smaller. In the older parts of the large areas, the picture is one usually met with where the bone has been formed out of Haversian canals, but nowhere is there a reforming of the intercellular spaces containing cartilage.

We cannot go too deeply into the minute histology represented in Manasse's studies, but the question of the primary source of the diseased process must be referred to. Manasse thought for a long time that this was to be found in the osteoclasts which are seen in the perivascular connective tissue in the bone, but his more careful study showed him that the initial stage proceeds from the blood vessels themselves. The second interesting question is the origin of the sieve-like rarefaction which represents the most recent stages of the new bony growth. Manasse believes these minute openings to be where the bone corpuscles were previously found. As he remarks, this form of bone absorption is not unknown. M. B. Schmidt, among others, has carefully described this process. Manasse's conclusions are as follows: "The alteration of the laby-

rinthine capsule begins with the formation of osteoid and granulation tissue within the preformed blood-vessel spaces. The old bone is destroyed as the result of this new tissue growth by the simple process of compression, not through lacunary absorption, and the new bone tissue takes its place. This has only an intermediate character, in so far as it in turn is destroyed by two different methods of absorption. First, through a sieve-like rarefaction of the new bone tissue, and second, through lacunary absorption with

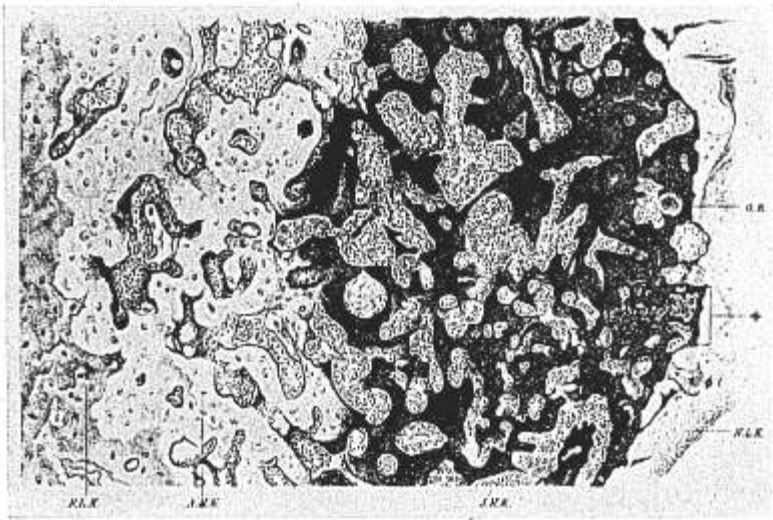


Figure 4—Case 1: Section through the portion of the new-formed bone lying near the cochlea. A stronger enlargement. We see on the left, the light-colored older portions of the new-formed bone. A. N. K. On the right, the dark-colored younger portions. J. N. K. Both are sharply separated from the normal labyrinthine bone. N. L. K. The former consists of dense bony tissue containing a few large vessels. The latter contains many more open spaces and many more cell element. At O. K. we see an osteoclast in the new-formed bone. Nowhere are osteoclasts to be seen on the border of the pathological and the normal bone. Here always the newly-formed bone substance crowds close on to the old labyrinthine bone without any cell elements. The star shows the cribiform rarefaction of the new bony growth. (From Manasse's collection with permission.)

the formation of osteoclasts. There follows a third deposit of bone in the marrow spaces as the result of osteoblast activity, which is at first spongified in character, and which does not disappear. As time goes on it becomes denser and harder and finally fibrous. The marrow spaces become smaller, the bony trabeculae always thicker, a lamellae system develops, and finally compact bone is formed which is always to be sharply differentiated from the old labyrinthine tis-

sue. The whole process is, to Manasse's mind, an inflammatory one, and he has accordingly named it "*Otitis chronica metaplastica der Labyrinthkapsel.*"

With these views most recent investigators agree, including Otto Mayer, Panse in his newly appeared "*Pathology of the Ear,*" and Bruehl, except that Bruehl is not prepared to admit that the process is a true inflammation. Manasse does not, however, stop here, but in addition to the changes in the bone, describes changes in the membranous labyrinth and claims, based on his seventeen specimens where changes without a single exception are met with, that alterations in one or more portions of the sound-perceiving apparatus, namely, either atrophy in the organ of Corti, connective tissue deposit in the fine nerve filaments in the cochlea, similar changes in the spiral ganglion, or finally in the auditory nerve itself, are an essential part of the disease. He contends, indeed, that in every case of chronic hard-hearing of a high degree, whatever the cause may be, there are always the same changes in the membranous labyrinth. He does not presume at the present time to say whether the bone changes cause those in the labyrinth, or vice versa, although in one of his cases, where the labyrinth was diseased on both sides but the patient heard better in the ear where the bone was not affected, he does not think the labyrinth could have been involved as the result of the bone disease.

From these views of Manasse regarding the membranous labyrinth, Bruehl vigorously, and it seems to us, with good reason, disagrees, or rather argues "not proved." In the first place, nothing is more difficult to determine than what changes in the membranous labyrinth are pathologic and what are post-mortem. The tissues are exceedingly delicate, and subject to post-mortem influences in the time elapsing after death, and from the processes adopted for their examination. This objection Manasse admits, and defines certain alterations found, as post-mortem and some, such as atrophy of Corti's organ, as pathologic. Further, it is impossible to positively show whether the change in the soft parts does not develop long after the original bone disease, and even as a result of the intercurrent affections, possibly acute, which carried the patient off. Bruehl, indeed, insists that such changes are in no way characteristic as they are met with in all severe affections of the middle-ear. Finally, it is too much to say that either clinically or anatomically they are always present. In two young patients recently examined by Bruehl, suffering from otosclerosis, the one 34 years of age, and the other 24 years old, there was no lowering whatever

of the upper tone-limit, while in a case just reported by him showing microscopically the characteristic bone changes, the organ of Corti was entirely normal. The most that we can assert, then, at present, is that changes in the membranous labyrinth are frequently, though not invariably present; whether they are a necessary part of the anatomic disease is yet unproved, but they are of sufficient importance to be always borne in mind. Functional tests showing nerve-deafness do not necessarily prove that the cause lies in the disease under discussion, but if slow in onset and if all other causes can be excluded, render such alterations in the membranous laby-



Figure 5: A higher magnification of the previous figure showing destruction of the new formed bone as the result of cribriform rarefaction. The bone is very porous. A cell is to be seen in each cavity. In certain places the cavities have separating walls which are very thin. In other spaces these are lacking and they are confluent, forming streets or large spaces on the border adjacent to the normal bone, M. K. No cells are to be seen. (From Manasse's collection with permission.)

rinth highly probable. What is urgently required in order to throw more light on this difficult question are carefully studied temporal bones of young subjects in whom otosclerosis has been diagnosed clinically. This, as we said at the outset, is manifestly difficult. The one objection to Manasse's studies is that his specimens are chiefly from aged people where we should normally expect changes in the labyrinth.

Of far more interest to us as clinicians, than Manasse's histological studies, is his statement in regard to ankylosis of the stapes in his cases, and the deductions which he draws from this. In only three cases, five temporal bones, was ankylosis found. In the re-

maining twelve bones the joint was uninvolved. Manasse concludes, based on these findings, that anatomically speaking, stapes ankylosis can no longer be regarded as an essential part of the disease, nor even can it properly be said that these findings are due to the early stage of the disease, and that later there would have been such an involvement, for in two cases at least, the microscope shows

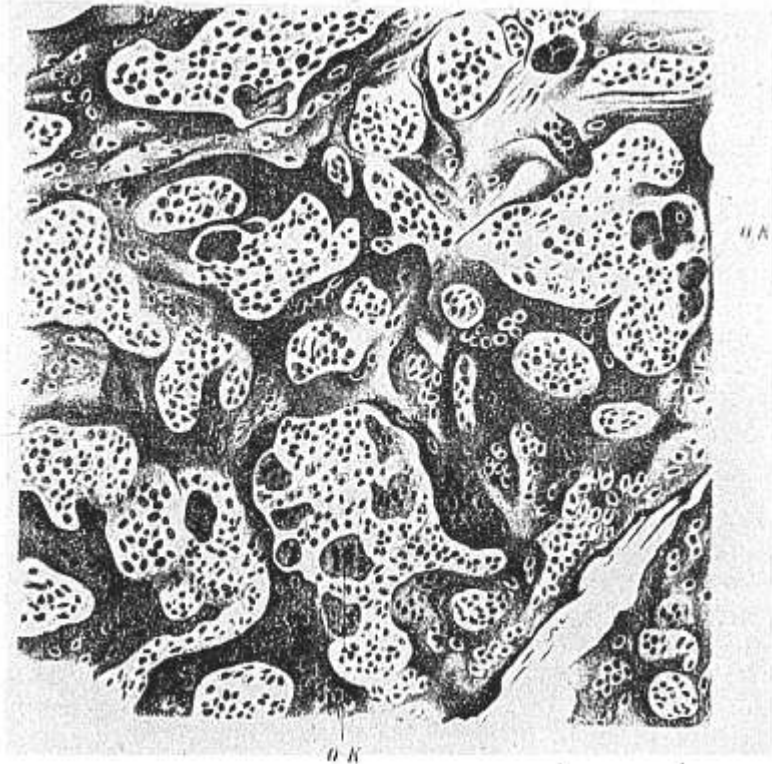


Figure 6—Case 5: Right. Section through the recent portions of large new bony growths. Extensive destruction of the dark-colored new tissue partly as the result of the Howschipschen, lacunae and osteoclasts (O. M.) and partly the result of cribiform rarefaction of the bone substance seen on the right-hand side below. On the left again new-formed bone. (From Manasse's collection with permission.)

the process to have been evidently an old one, which had stopped short of the stapes and the annular ligament. Manasse argues, furthermore, that even if stapes ankylosis is met with, it is impossible to say that it is bony in nature and due to the disease in question. He supports this contention by giving the histological findings of three cases of ankylosis, the first due to a lime deposit, the sec-

ond due to otitis media chronica adhesiva, and the third, a result of a general periostitis of the stapes and of the inner wall of the middle-ear. It is evident that whatever the cause of the ankylosis, the functional tests will be the same, and Manasse accordingly advances the view mentioned previously and shared in by Panse, that chronic progressive hard-hearing is not possible of diagnosis. We can readily understand how, with all that has been written to throw doubt on the subject, one can gain this pessimistic decision. We cannot, however, admit Manasse's contention. It must not be for-

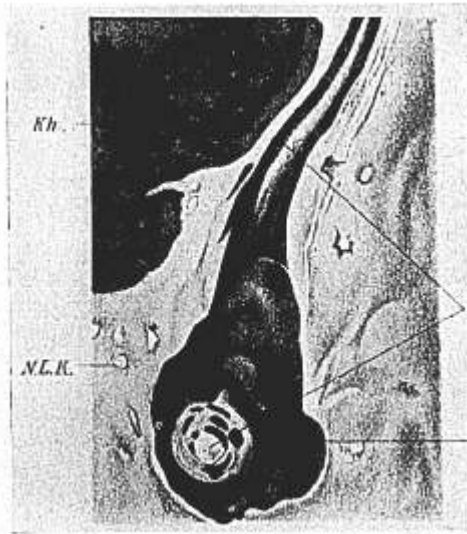


Figure 7—Case 4: Right. A higher magnification. Blood vessel in the bone (Kn. G.) Within the normal labyrinthine bone. (N.L. K. M.) The vicinity of the new formed bone. (K. b.) On the outer wall of the blood vessel we see new-formed, very young bone tissue, (N. g. K.), which presses in the form of hummocks against the old bone and the blood vessel which are thus being caused to disappear. (From Manasse's collection with permission.)

gotten that "Otitis chronica metaplastica der Labyrinthkapsel," so minutely and painstakingly described by him, is strictly an anatomic disease. It is undoubtedly true that it may develop in other parts of the labyrinth capsule than in the oval window, and in such cases may produce no symptoms unless the membranous labyrinth is involved, when the upper notes will be affected. It is undoubtedly correct to say that there are many such cases which have never been recognized. It is, however, the clinical manifestations of the disease which interest us and concern us alone, and it is only when the disease has advanced to involving the stapes and causes an im-

pairment or fixidity of that bone, that we can speak of it in a clinical sense. So it is proper to say that while there is an anatomic disease, there is also a clinical disease, and this, for a lack of a better name, will continue to be known under the name of *otosclerosis*.

While it is true that any form of ankylosis will produce the same functional tests, other causes than that due to a bone occlusion are so rare as to be excluded, although it must be borne in mind that such exceptions do occur. Panse states that he recalls a number of cases of deafness developing subsequent to confinement, which from the clinical course, the functional tests, the otologic pictures, and history of heredity, would have been regarded as otosclerosis. They, nevertheless, by use of the catheter and massage, made a marked and lasting improvement. Such exceptions occur and are constantly to be borne in mind in too promptly pronouncing the case hopeless.

To sum up, we feel that the pessimism in regard to understanding and diagnosing the disease is not warranted, and that while it is true that much is yet to be learned both as regards the histology and the etiology and that we can offer little for a cure so long as the latter is so obscure, we can confidently say that in the majority of cases, a determination of the disease is entirely possible. While nine years have elapsed since Denker's book on the subject, we regard his description of the clinical disease an eminently correct one:

"In the cases of progressive hard-hearing, which show an unchanged or virtually normal drum membrane, a patent Eustachian tube and the Bezold triad of symptoms, we may conclude that the pathological changes are only in the stapes and the annular ligament and in the bony areas bordering on the oval window. In the other cases where the functional test does not give the Bezold triad, but where there is a pronounced reduction of the upper hearing-limit, where bone conduction is not lengthened, and where the Rin   is not pronouncedly negative, there is in addition to the disease of the oval window an extension of the process further into the capsule of the labyrinth, or an involvement of the membranous labyrinth. While in most of the cases the mucous membrane of the middle-ear shows no change, in the cases of Scheibe and Habermann, and in those of Katz, the cause undoubtedly lay in a chronic middle-ear suppuration."

Consequently, as we took occasion in a former paper to point out, this or a similar middle-ear is not to be lost sight of in searching for a possible etiological factor.

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