ANKYLOSIS OF THE STAPES.¹

By PROFESSOR DENKER (Erlangen).

(Translated and abridged by DR. DUNDAS GRANT and DR. STOLTE.)

PROFESSOR DENKER commences with a review of the general historical landmarks up to the year 1885 in the study of the nature of ankylosis of the stapes and its relation to deafness. This had already been noted in the eighteenth century by Valsalva, Morgagni, and Meckel; but a great debt was due to Toynbee, who, in his pathological examinations of 1,149 preparations, had described 136 of osseous ankylosis of the stapes in the fenestra ovalis. After Toynbee, there followed within the next thirty years—up to the year 1885—a long series of publications on osseous ankylosis of the stapes in the fenestra vestibuli by Erhard, Kramer, Voltolini, Tröltzsch, Politzer, Lucae, Moos, Schwartz, Bertuch, Weber-Liel, Magnus, and Kirchner. Denker has compiled these results and condensed them in this report.

The disease was considered as pathologically and anatomically an osseous ankylosis of the stapes in the fenestra ovalis, which probably arose either as secondary to chronic middle-ear catarrh or purulent otitis media with circumscribed ossified periostitis, or primarily from an osteitic process in the temporal bone.

The meatus and tympanic membrane were comparatively normal or very slightly altered in appearance, and the Eustachian tubes freely pervious; the hyperæmia of the promontory seen in some cases through the membrane was looked upon as a further diagnostic feature. As regards the functional symptoms, there were among the commemorative points noted gradually increasing dulness of hearing, associated with troublesome subjective noises. It was, however, established that osseous conduction for the tuning-fork was well preserved. It was also known that the morbid process leading to the fixation, so far as it could be determined by macroscopical examination, arose sometimes as a sequela of chronic catarrhal or suppurative inflammatory changes in the middle ear, but it could likewise be brought about in the presence of normal conditions in the tympanic cavity by a primary osteitic process in the petrous bone.

For the differential diagnosis between ankylosis of the stapes and nervous deafness it was known that in the latter there was lowered bone-conduction, whereas in the presence of fixation of the stapes this was well preserved. There was no means of establish-

¹ A Report read before the German Otological Society, June, 1902.

ing a sufficiently certain diagnosis, *intra vitam*, from the functional symptoms, and at the same time the determination of the pathologico-anatomical condition by microscopical examination had not yet been carried out.

In the year 1885 our knowledge concerning the functional signs of ankylosis of the stapes made a very marked step forward on the publication of a work by Bezold, in which, for the first time, the pathologico-anatomical evidence was to the effect that when there was a fixation of the conducting apparatus there was always a "negative" result with Rinne test. He described a case in which, with considerable diminution of hearing-power for speech, there was prolongation of bone-conduction for the tuning-fork and a "negative Rinne," with normal condition of the tympanic membranes and Eustachian tubes. On the strength of these signs he made a diagnosis of ankylosis of the stapes, which was confirmed by post-mortem examination. He explained the prolongation of the bone-conduction by increased tension of the conducting apparatus, a view which he considered was supported by an experiment in which a vibrating tuning-fork was put in communication with the ear by means of a slightly elastic cord; the sound of the tuning-fork became louder as the cord was stretched, and less loud as it was loosened. He looked upon bone-conduction as taking place, not through the molecules of the bone alone, but mainly through the tympanic apparatus—the so-called "osteo-tympanic" conduction. He considered it as a well-recognised fact that in the normal ear or in those in which there is simply an affection of the internal auditory apparatus, with an intact condition of the conducting apparatus, air-conduction has the preponderance over bone-conduction, but that when this relation of things is reversed there is, without doubt, some affection of the conducting organs—namely, the tympanum and its contents. He arrives, from his own experiments, at the result that the increase of bone-conduction in all affections of the conducting apparatus depends on the increase of the tension in or by the "fixation bands" (from the radial fibres of the tympanic membrane to the annular ligament inclusive), and that an increased tension of the fibres of the annular ligament is particularly adapted to bring about an increase of bone-conduction. A further publication appeared from the hand of the same author in 1887, entitled "Results regarding the Diagnostic Value of Rinne's Test, combined with an Explanation, arising from this, of the Physiological Function of the Sound-conducting Apparatus." In this Bezold established some further points with regard to the functional conditions present in cases of fixation of the conducting

apparatus; he found that, in addition to a raising of the lower limit of audition—that is to say, that the hearing for the lower tones of the scale was very markedly diminished—the tuning-fork A (about 100 vibrations) on the vertex was invariably heard, without exception, better in the worse ear, and that in Schwabach's test the same tuning-fork was heard almost invariably longer by the patient than by the examiner, who had normal hearing. These observations entirely confirmed the results arrived at by Schwabach two years previously.

Katz is credited with having, in the year 1890, published the first case of ankylosis of the stapes, in which, by means of functional tests during life, the diagnosis of this fixation of the bone had been made and confirmed by microscopical examination after death. It must, however, be kept in mind that already, in 1887, Politzer had published, in the second edition of his textbook, the illustration of a microscopical section through the stapediostapedial articulation, which showed a synostosis of the stapes with the oval window; there was, however, no description of the results of functional examination during life. Katz made the diagnosis on the strength of steadily progressive deafness, "negative Rinne," and increased bone-conduction for the tuning-fork A on the vertex. He found that the foot-plate of the stapes was thickened to four times the normal, and that it formed, with the annular ligaments and the niche of the fenestra ovalis, a firm, uniform osseous mass. This bony mass presented numerous newly-formed medullary spaces and young osteoid substance. Both on the inner and on the outer surface of the stapes there was a thickening of the periosteum; the disease seemed not yet to have come to an end. The other ossicular joints and Corti's organ were found intact.

From the year 1890 a considerable number of cases have been examined in which there was supposed ankylosis, and in which, before death, the functional tests had been carried out. Foremost among these is a case published by Habermann in Schwartze's handbook on Otology in 1891. At a later date he described thirteen other cases at the meeting of the Otological Society at Breslau. In 1893 Bezold brought forward in Frankfort another case of bilateral ankylosis of the stapes, in which, during life, he found a considerable negative Rinne (with the tuning-fork A¹), prolongation of bone-conduction, and considerable elevation of the lower range of audition. As the main cause for the very high degree of deafness which was present, he found bilateral osseous ankylosis between a portion (especially in front and above)

of the foot-plate of the stapes, which was at this part eight times its normal thickness, and the pelvis ovalis. This had arisen as a result of a well-defined area of chronic inflammatory bone disease, which, according to the microscopical appearances, seemed to have arisen either from the bone itself or from the periosteum of the niche of the fenestra ovalis. There was also in this case a lowering of the higher limit of audition, but this was explained by the presence of atrophy of the nervous elements in the lowest portion of the cochlea. The bony change consisted in the formation of young osteoid tissue, with medullary spaces which were very rich in cells, also giant cells containing numerous nuclei, and here and there rarefaction (*spongiosirung*) of the compact bone. The disease had extended from the fenestra ovalis to the canal for the vestibular nerves, to the vestibule itself, and also between the first and second turns of the cochlea.

A considerable series of important cases was brought forward by Politzer at Boston, U.S.A., in the summer of 1893, and at Washington on September 6, from the rich material afforded him by the Vienna Ear Department. The same author published in the *Zeitschrift für Ohrenheilkunde*, July, 1894, sixteen cases of ankylosis of the stapes which had been investigated during life and examined post-mortem. In all these there was little or no change in the membrane and the mucous lining of the tympanic cavity. As regards the functional tests, there was a very high degree of dulness of hearing or deafness, which in one case, however, of a patient who was completely deaf was due not alone to the fixation of the stapes and rarefaction in the capsule of the labyrinth, but to atrophy of the fibres of the auditory nerve as well. Unfortunately, there is no detailed account of the functional tests in the different cases; but in general, it is stated that as a rule, in addition to a well-marked negative result of Rinne's test, there was also diminished perception of the acoumeter or of the watch-tick through the bones, and with long duration of the disease there was not unfrequently a diminished duration of perception through the bones for the tuning-fork in the middle part of the scale (C²). Professor Denker is of opinion that these two last points do not help us in our diagnosis, because we find these functional changes also in disease of the perceptive apparatus. The histological condition was similar to what was found by Katz, Habermann, and Bezold (with Scheibe). In the same year (July, 1894) Bezold published a second case of ankylosis of the stapes, which had been diagnosed during life and confirmed post-mortem by manometrical and histological examination; in this case there was also present

the triad of functional symptoms which are characteristic of fixation of the stapes—namely, lengthened bone-conduction for deep tones, marked “negative” Rinne’s test, and extensive loss of the lower part of the scale of hearing. The histological examination revealed a similar change in the bones to what had been seen in the other cases, but the process appeared to have completely come to a termination, as there were only very few giant cells, and the young osteoid tissue was completely absent. The disease was apparently confined to the fenestra ovalis and the bone surrounding it, so that there could be no further doubt as to the etiological connection between the characteristic complexus of symptoms and the pathological condition in the stapedio-vestibular symphysis.

To these observations must be added four cases from Professor Siebenmann’s clinic in the year 1898; two of these, which were described by Edward Hartmann, are of particular interest in this respect, that the bone-conduction during life was shortened, and that in them, beyond the complete fixation of the stapes, extensive osteo-sclerotic disease of the capsule of the labyrinth was found. In the first of these cases there was a normal condition of Corti’s organ, but a slight narrowing of the canals for the fibres of the tractus ganglionaris and foraminulentus. In the third of the cases there was on both sides shortened bone-conduction and positive Rinne. Siebenmann explained the former by the rarefying osteitis of the capsule of the labyrinth, extending to the endosteum of the inner ear. Corti’s organ and the nerves were sound, and the positive but shortened result of Rinne’s test may be explained by the fact that, though the disease was indeed a beginning osteoid change in the foot-plate of the stapes itself and in the cartilage covering it and the pelvis ovalis (formation of osteophytes), the annular ligament was on the right side quite sound and on the left only very slightly affected. In Siebenmann’s fourth case there was at first the typical clinical picture of ankylosis of the stapes on the right side, but at a later stage the clinical picture was that of an auditory nerve disease. Histological investigation showed only a single focus of rarefying osteitis in the capsule of the labyrinth above the upper margin and the anterior half of the niche of the fenestra ovalis; the upper margin of this niche was the site of an osteophytic growth of such extent that the foot-plate of the stapes in its upper part was invisible from the tympanic cavity. In the few medullary spaces which remained in the diseased bone, Siebenmann found real fat marrow, which, up till then, had not been found in any of the cases. Examination of the auditory nerve in this case revealed interstitial neuritis of the nerve trunk, which

Siebenmann attributed to the carcinomatous cachexia, of which the patient died. Two cases were brought by Scheibe before the Otological Society in Breslau in the year 1901; in these there were, besides the osteo-porotic change in the capsule of the labyrinth, an osteo-sclerosis of the pneumatic and cancellous spaces and the residua of chronic middle-ear suppuration and of otitis interna. Finally, Denker refers to a case brought forward by Katz in October, 1901, before the Berlin Otological Society. In this the osteitic-porotic change had affected, not alone the capsule of the labyrinth, but also in particular the malleus and incus and the surrounding bone, without, however, giving rise to osseous ankylosis of the stapes. There were also evidences of a previous exhausted catarrhal inflammation in the form of fine reticular connective tissue with cystic spaces.

Professor Denker epitomizes the results of these cases as follows:

In the cases of progressive deafness with normal tympanic membrane and patency of the Eustachian tubes are found pathological changes in the stapes, the annular ligament, and the bony parts surrounding the fenestra ovalis; it is found also that the upper and anterior portions of the circumference of the fenestra ovalis form the "seat of election" for this bony process. In cases in which the hearing tests did not show Bezold's triad, and there was no prolongation of bone-conduction, and Rinne's test was not negative but only positive shortened, then there was some other disease (osteo-sclerotic process in the labyrinth capsule, disease of the nerve, incomplete fixation of the stapes). While in most cases the mucous membrane of the tympanum showed no departure from the normal, there were in the cases demonstrated by Scheibe and Habermann in Breslau, and also in Katz's cases, evidences of previous chronic suppurative inflammation of the middle ear, showing itself by the inflammatory thickening. A similar condition of the mucous membrane was found in the cases published by Hartmann from Siebenmann's clinic.

The pathological changes in the capsule of the labyrinth and in the stapes are formulated as follows:

The foot-plate of the stapes, as well as the periosteum, was thickened to several times the normal, and formed, along with the annular ligament and the bone of the niche of the fenestra, one solid mass; occasionally there were found between the foot-plate and the niche several separate bridges of bone in the substance of the annular ligament. The new-formed bone found in place of the compact substance of the capsule of the labyrinth was formed in

the earlier stages by osteoid substance, and in the cases where the process appeared to have exhausted itself by spongy bone; in the young osteoid tissue could be found a large number of medullary spaces containing many cells, and, further, many giant cells containing nuclei.¹

In a part of the osseous spaces were bloodvessels tightly packed with corpuscles, while elsewhere very few blood-corpuscles were to be found. The foci of rarefaction found without direct connection with the fixation of the stapes were observed both in the bony capsule and the vestibule and in the semicircular canals; only once was genuine fatty marrow found in the medullary spaces of the osteoporotic bone. It is through Siebenmann's investigations that we have our knowledge of the processes by which the change of the normal labyrinth capsule into well-formed bone takes place—namely, by a process of absorption and deposition. According to this author, there is, in the first place, loss of affinity for staining with hæmatoxylin and carmine on the part of the innermost layers of cells of the Haversian canals; these become wider as the result of lacunar absorption in the direction of the labyrinth. The collaboration of the multinucleated giant cells in these changes can be made out in several cases, although not always; while in certain parts the process of absorption extends, there is in others a new deposition of bone. All the intraglobular cartilaginous remains are dissolved and replaced by new-formed bone. The cartilaginous covering of the labyrinthine windows, as also of the foot-plate of the stapes, is absorbed under the action of vessels proliferating from the bone; it becomes replaced by spongy tissue. The line of demarcation between the normal and the newly-deposited bone is in the early stages very distinct, but in the later stages it becomes much less so, and finally is quite undetectable. The new-formed bone acquires gradually a lamellar structure; the medullary spaces become poor in cells and vessels, but richer in fibrous connective tissue.

As regards the origin of the disease, the two different views stand prominently out: either that the pathological anatomical conditions above described are secondary to a former exhausted inflammatory affection of the middle ear, or that they arise primarily as ossifying osteitis in the periosteum (Bezold, Scheibe) or the capsule of the labyrinth; the former view is advanced by Haber-

¹ The disease appears to commence with a vascularity of the osseous substance and a deposition of young osteoid tissue containing giant cells; the medullary spaces become enlarged, and the condition of rarefaction is produced.

—TRANSLATORS.

mann and Katz, but Politzer localizes the commencement of the disease in the capsule of the labyrinth, and Siebenmann, as above mentioned, is convinced that the rarefying osteitic process takes its origin at the line of junction of the endochondral labyrinthine capsule and the connective-tissue bone which is formed secondarily from the periosteum.

As regards the opinion of Habermann and Katz, it seems very unlikely that an affection of the tympanic mucous membrane should lead to an ossifying periostitis and the changes in the bone which we have already referred to, but there is at least a possibility that there may be in some cases an association of the two diseases. This, however, leaves quite unexplained those cases in which there is no evidence of present or previous disease of the mucous membrane of the tympanic cavity; in these we have to fall back upon a primary affection of the periosteum or of the bone. Whether there is a primary affection in the capsule of the labyrinth, in the later-formed connective tissue of the bone, in the periosteum, or, lastly, in various points, only time can show. That the rarefying osteitis may take place without any involvement of the periosteum we must admit, in view of the researches of Siebenmann and Politzer.

It remains to consider what condition it is which starts the disease, the appearances of which have been above described. Under what circumstances does the rarefaction of the cartilage remains in the interglobular spaces take place, and what excites the absorption of the compact bone of the labyrinth capsule, and the production of the new-formed osseous tissue?

Bezold's statistics show the female sex affected in 61 per cent. of cases of ankylosis of the stapes; it was in Denker's cases, 58 per cent.; in Hartmann's, 54 per cent.; in Panse's, 51 per cent. When we compare this with the lesser incidence of ear disease in general in females—namely, 42 per cent.—we recognise a peculiar susceptibility of the female sex to this particular disease. As we find that in a considerable number of cases either the commencement of the disease or its increased rapidity of development dates from the time of gestation or the puerperium, it seems as if the changes in the temporal bone were analogous to those which take place in other bones under similar circumstances (osteomalacia, etc.). This leaves still a considerable number of cases to be explained. Denker believes, with Katz and Schwartz, that in addition there is some constitutional disturbance or dyscrasia, such as rheumatism, gout (Toynbee), scrofula, syphilis, senile changes, or neuro-paralytic trophoneurosis, which may account for the develop-

ment of the disease under consideration. The influence of hereditary tendency appears also as an important etiological factor, as shown by the investigations of Bezold and Siebenmann, with whom Professor Denker is in agreement. This writer found that out of 306 cases, in 124—that is to say, 40·5 per cent.—there was evidence of affection of hearing in several members of the family. Bezold found it in about 52 per cent., Siebenmann in 35 per cent. According to the reports of various authors, it appears, however, that severe chill may play an important rôle in producing ankylosis of the stapes. Thus, out of 29 cases described in Panse's work as suffering from synostosis of the stapes, and in which there were details concerning the etiological factors, cold is given as the cause in 11, and in Hartmann's 33 cases 5 also attributed it to this same cause. Syphilis is considered an etiological factor, but Denker thinks that its influence cannot be very great, in view of the fact that syphilis is extremely widely spread and that cases of ankylosis of the stapes are comparatively rare. Ankylosis of the stapes is mainly a disease of middle age, and in Denker's 306 cases the beginning of the deafness took place in 238 between the ages of twenty and fifty.

With regard to the symptoms and course of ankylosis of the stapes, Denker notes that the diminution of hearing-power comes on gradually, so that the patients are very often scarcely in a position to state when the disease began, and it is only rarely that the sudden occurrence of extreme deafness is reported. The course is, as a rule, very slow, and it may take several decenniums for the loss of hearing to reach any very considerable degree. Still, there occur not unfrequently exacerbations of deafness from which, as a rule, recovery does not take place. Cases also have been observed in which the condition has remained stationary. In pure uncomplicated ankylosis of the stapes, which present the triad of symptoms described by Bezold, even in the later stages, whispers can be heard in the neighbourhood of the ear, and always ordinary conversation. When the perception for loud speech is lost, there is no longer a simple fixation of the stapes, but either the fenestra rotunda or the nerve apparatus is at the same time affected. This complication evidences itself partly by another result of functional test—namely, the shortening of bone-conduction.

Subjective noises (hissing, roaring, ringing, hammering, explosions), which are sometimes continuous and sometimes intermittent, occurred, in Denker's 306 cases, 226 times—namely, in about 74 per cent. Politzer found them present in more than two-thirds, and Bezold in 77 to 78 per cent. of his cases. Bezold found vertiginous

symptoms in 28.6 per cent., and Denker only in about 5 per cent. The latter observer, however, only noted vertigo as present when it reached such an extent that the patient had to catch hold of some firm object to prevent himself from falling. Denker has been unable to confirm Zwaardemaker's view, published in the year 1896, that as an initial symptom of sclerosis of the middle ear there is an elevation of the upper range of audition above the normal.

Pains in the deeper part of the ear have been observed in a few cases, and have been attributed to the changes taking place in the stapedio-vestibular symphysis or to the chronic inflammatory process in the periosteum, or also—as Siebenmann thinks—to the irritation of the sensory fibres of the facial (? tri-facial) nerve. The hyperæmia of the mucous membrane covering the promontory, which Schwartze considered as a characteristic symptom, and which Bezold, in his "Ueberschau," looked upon as of extremely unfavourable prognosis, is not now considered of the same value in differential diagnosis, as Siebenmann has shown that it occurs also in pure nerve deafness.

The diagnosis of pure ankylosis of the stapes depends, as already indicated, upon the anamnesis and the objective functional conditions (progressive dulness of hearing in middle age, noises in the head, occasional vertigo, patency of the Eustachian tubes, normal tympanic membranes, but considerable lowering of hearing-power for speech and the presence of Bezold's triad). When the osteopathic change extends beyond the surroundings of the fenestra ovalis to the capsule, the cochlea, and the modiolus, then we have the clinical picture of Bezold's "dysacusis," in which the lower limit of audition is considerably raised, bone-conduction is no longer increased, and sometimes Rinne (extremely rarely) is positive.¹ For the differentiation in such cases between nerve deafness and a combination of this with ankylosis of the stapes we have to depend upon the examination of the duration of hearing for different tones of the scale, as first proposed by Arthur Hartmann of Berlin. According to him, in pure sclerosis the hearing-power by aëro-tympanic conduction increases from the lower to the higher tones, and the bone-conduction—especially for deep tones—preponderates over the air-conduction; on the other hand, in nerve deafness the opposite is the case. Werhowsky confirms this, but he finds irregularities in the shortening of the hearing-power from below upwards.

¹ The only case in which this appears to be very definitely present is that one of Siebenmann's in which the osseous change took place in the foot-plate of the stapes, and not in the annular ligament.—TRANSLATORS.

All treatment by local measures, whether mechanical or operative (extraction of the stapes), has given very discouraging results. In view of the fact that local treatment, such as the air douche, catheterism, mobilization by massage, may, under certain circumstances, do mischief, they ought to be left off entirely when, after a short trial, no improvement in the hearing for speech presents itself. When, however, noises in the ear are present, the writer recommends, in agreement with most other authors, the regular use of electrical motor massage of the membrane, which, in many of his cases, has in a short time brought about complete temporary relief from the noises, and in a few cases permanent. Politzer treats the diseased process itself by the internal administration of moderate doses of iodide of potassium, 15 grains daily; and he believes that in cases where the loss of hearing has not reached too high a grade it has brought the disease to a standstill. The improvement stated to have occurred under the treatment by thyroid extract has not been confirmed by the observations of Denker, Siebenmann, and Brück. Siebenmann recommends the administration of phosphorus in daily doses of $\frac{1}{60}$ to $\frac{1}{15}$ of a grain in cod-liver oil (Kassowitz's solution), and states that he has stayed the advance of the disease in rapidly progressive cases. Denker, however, fears that the administration of such large doses involves the risk of the patient's suffering from phosphorus necrosis.

BRITISH MEDICAL ASSOCIATION.

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SECTION OF LARYNGOLOGY AND OTOTOLOGY.

PRESIDENTIAL ADDRESS.

ON THE POSITION OF LARYNGOLOGY AND OTOTOLOGY IN THE MEDICAL STUDENT'S CURRICULUM.

By PATRICK WATSON WILLIAMS, M.D., *President of the Section.*

GENTLEMEN,

My first and pleasing duty as President of this Section is to extend a very cordial welcome to those distinguished colleagues who, in response to our invitation, have come from afar and at considerable personal inconvenience to assist us with their ripe experience in the various questions we propose to discuss-