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Congenital fixation of the stapes

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CONGENITAL FIXATION OF THE STAPES

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INTRODUCTION

The purpose of this paper is to consider the problem of congenital fixation of the stapes, the third ossicle of the middle ear. The embryology of the middle ear and stapes will be considered in length. The anatomy of the stapes will be presented. The diagnostic criteria as to differentiation of congenital fixation of the stapes from acquired otosclerosis and the operations necessary for correction will be reviewed.

Only in recent years has this condition been recognized. It must be considered in the differential diagnosis of any case of deafness. Surgical procedures for correction are essentially the same as in otosclerotic deafness but at times the procedures are more difficult to perform. Operation in children will result in a more comfortable and less conspicuous life for the patient. This condition is often found in adults who present with hearing loss which has been present since "childhood" and must be considered in patients who present with difficulty hearing who, when their cases are reviewed or followed, do not show progression of their deafness.

A HISTORICAL NOTE

In 1925, Beaton and Anson stated that "in spite of the importance of the stapes, no single element in human anatomy has suffered more chronic neglect. This is a strange instance of disregard, since the stapes has great clinical importance. It passes through developmental stages of extreme interest, and is grossly the most captivating bone in the human skeleton. For all of its unique fascination as the smallest essential bone and as the most exquisitely fashioned unit, this ossicle has not only been forgotten by investigators in anatomy and otology, but the descriptions of it in standard textbooks of anatomy have become stylized and debased through the years. They are inaccurate and fragmentary, commonly perpetrating century-old errors. Illustrations are either minute or diagrammatic."[6] Not all investigators feel this strongly about the importance of the stapes. With the recent advancements in otology and otologic surgery, it has been necessary to study the stapes more extensively. Possibly the stapes is now receiving the attention that the quoted authors felt it should receive.

EMBRYOLOGY OF THE MIDDLE EAR AND STAPES

In an attempt to understand the structure and functional anatomy of middle ear structures, the early development of the ossicles and the tympanic cavity will be followed with special interest given to the stapes.

The auditory tubes and tympanic cavities represent a drawn out first pharyngeal pouch, with which some investigators consider the second pharyngeal pouch merges. These entodermal pouches appear in embryos of three mm., enlarge rapidly, flatten dorsoventrally, and lie in temporary contact with the ectoderm. The proximal ends of the entodermal pouches will later in the third and fourth months elongate, narrow and form the auditory tubes or Eustachian tubes. The distal ends of the entodermal pouches will enlarge to form the tympanic cavities. This early tympanic cavity is surrounded by loose connective tissue, in which the auditory ossicles are developing. Toward the end of gestation this gelatinous material will be absorbed and the tympanic cavity will expand to include the developed ossicles. The ossicles and their associated structures will then be covered by an epithelial membrane and thus actually lie outside the tympanic cavity.[2,5]

"In the embryo of 4½ weeks (7 mm.) the precursor tissue for the auditory ossicles is a common blastemal

mass which is distinguishable from the surrounding mesenchyma only because of the greater concentration of its constituent cells."[8]

In this mesenchymal mass, early in development, Meckel's cartilage and Reichert's cartilage, the first and second branchial arches respectively, are closely associated and are connected by an interbranchial bar of mesenchyma. Because of its relationship to the first pharyngeal pouch, this mesenchymal mass, from which the manubrium of the malleus and the long crus of the incus will develop, is considered by Hanson and Anson[8] to be mainly related to the second branchial arch. This partial development of the malleus and incus from the second arch is at variance with Arey[5] who states that the malleus and incus are of first branchial arch origin. (See Fig. 1.)

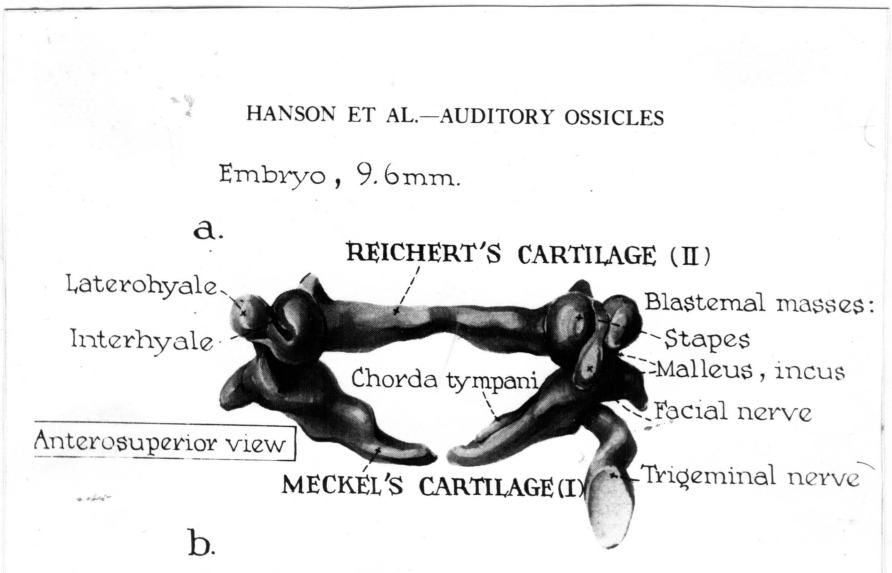


Fig.1. Photograph from reference 8.

During the fifth week the stapedial artery, which has been developing nearby the stapedial mesenchymal mass, begins to attain a central position in the blastemal lobe (stapedial mesenchymal mass), "thereby seeming to aid in the conversion of the rounded mass into a ring-like structure with an obturator foramen."[8] This process of conversion is complete at about six weeks (11.7 mm.). Failure of the stapedial ring to form will result in an anomalous stapes, such as excessively thinned stapes and other malformations seen at operation.[2]

In the 5½ week embryo the facial nerve, with its branch the chorda tympani, has split the original blastemal mass into a medial stapedial mass and a lateral mass called the laterohyale. These two are joined by a mesenchymal bar, the interhyale, which will undergo reduction in size, remaining in the adult as the tendon of the stapedial muscle. The stapes now lies lateral to the otic capsule and medial to the facial nerve, a condition which will persist into adulthood.[1,2,8] (See Fig. 2.)

"By the seventh week (17 mm.), the mandibular and hyoid bars have developed into cartilage through the greater fraction of their length and hence may be properly termed Meckel's and Reichert's cartilages. The proximal extremity of each bar and the ossicles are still composed of precartilage; however, the malleus and incus

are distinctly outlined and less a part of the mesenchyma from which they were derived."[8]

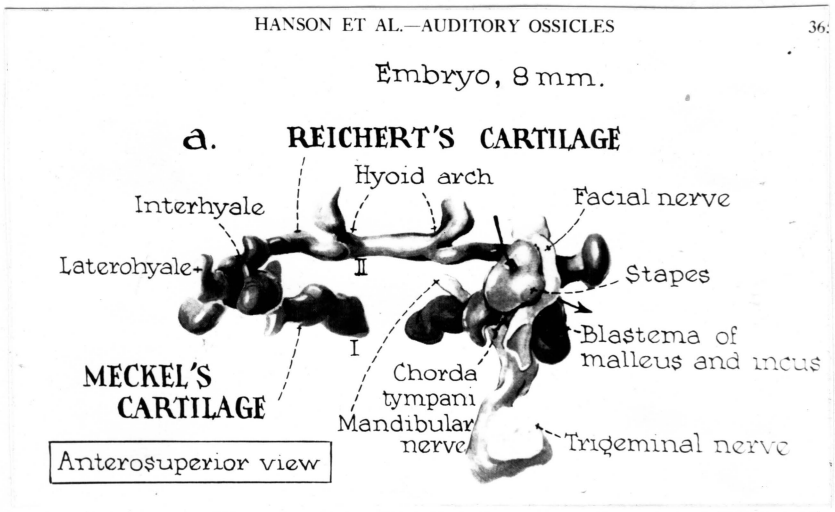


Fig. 2. Photograph from reference 8.

The laterohyale and the short crus of the incus approximate and fuse to the undersurface of the developing capsule at about this same time. "The secondary nature of this continuity with the developing capsule means that the capsule makes no contribution to these primordia."[8]

At the time that the developing embryo is approximately 28 mm. in length it is considered to have entered the fetal stage of development. At this time the ossicles are represented by cartilaginous "models" of the

future adult structures. (See Fig. 3.)

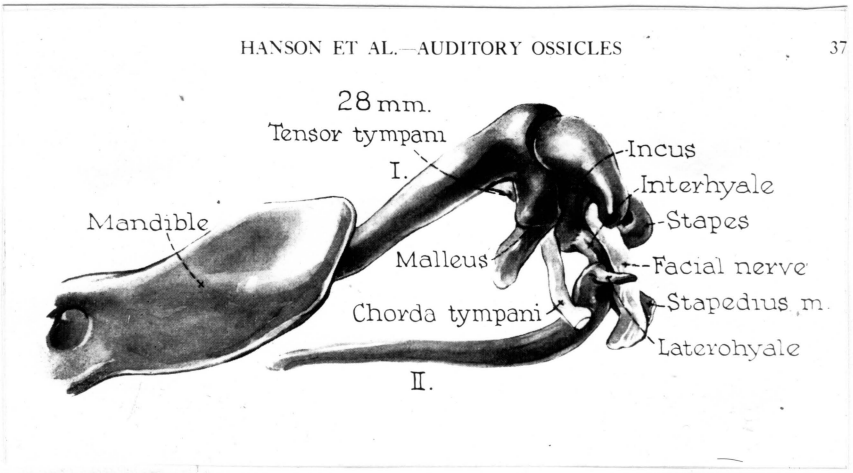


Fig. 3. Photograph from reference 8.

It is at this time, approximately 28 mm., that the facial canal with the facial nerve begins to form, first as a sulcus, finally becoming ossified and incorporating the laterohyale in its wall. At this time, as stated before, the distal end of the auditory tube begins to invade the mesenchymal tissue. "The chorda tympani branches and courses upward to pass between the manubrium of the malleus and the long crus of the incus and therefrom into the mandibular arch."[8]

The stapes is beginning to impinge on the cartilaginous otic capsule. It is at this area of the impingement that the future oval window (vestibular fenestra) will

develop. "The capsular tissue contributes to the base of the stapes and to the annular ligament, by which the stapes will be held within the capsule in later stages. The vestibular aspect and fenestral surface of the base will retain this cartilaginous lamina throughout the individual's lifetime, as will, likewise, the periphery of the fenestra."[8] (See Fig. 4.)

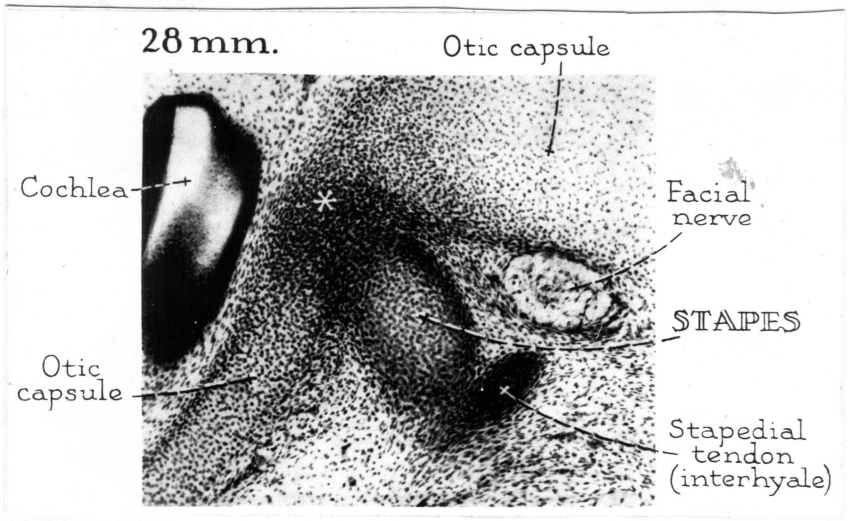


Fig. 4. Photograph from reference 8.

"In the fetus of nine weeks (40 mm.), all structures which will become part of, or related to the ear of the adult, are present in a state of recognizable incipiency.

"In the middle ear the auditory ossicles are lodged in primitive mesenchymal tissue; they are still wholly cartilaginous, and will remain so until the fetus has

reached the 117 mm. stage, at which time ossification appears first in the incus.

"The parts of the adult external ear are represented by the auricle (in which cartilage is already developing) and by the primordial stalk of ectoderm which, upon rearrangement and disappearance of cells, will become the external auditory meatus."[8] (See Fig. 5.)

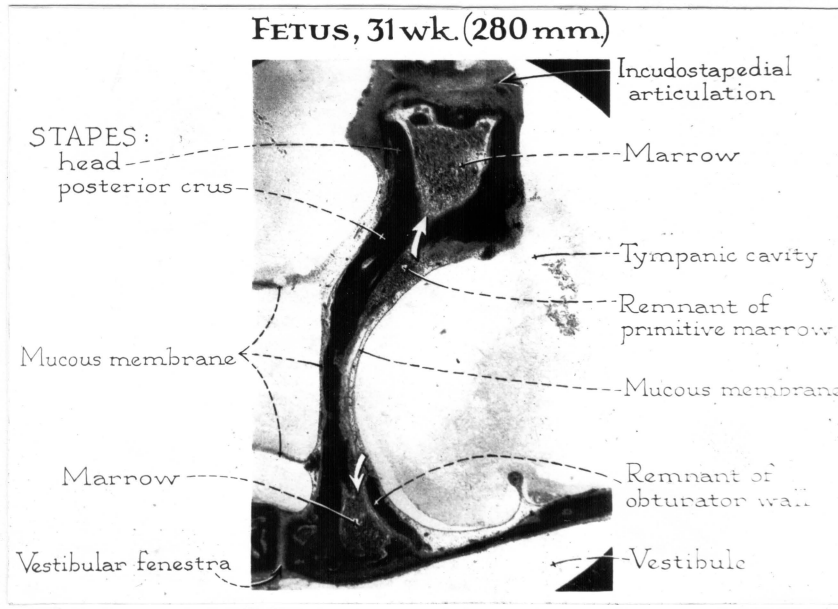


Fig. 5. Photograph from reference 3.

In the fifth month, areas of impending ossification are present in the base of the stapes. The line of future ossification is distinct, separating what will be an osseus area and a cartilaginous area in the base of the stapes of the adult, the cartilaginous base being the portion of the stapes that will articulate in the

cartilaginous oval window.[8]

Ossification of the stapes resembles long bone ossification in some aspects. The cartilaginous model must be destroyed by invading vascular buds before ossification takes place and in that each crus forms a cylinder with marrow; but ossification differs in that the entire cartilage model does not undergo ossification, no haversion systems are formed, and perichondral or periosteal bone is being removed as it is formed, preventing appositional bone growth and maintaining the necessary stapedial size.

At this time, "on the tympanic aspect the stapes is imbedded in loosely textured, vascular mesenchyma and is covered by a perichondral layer. On the vestibular surface (of the base) the cartilage is invested by a periotic tissue. The latter layer will remain thin throughout the individual's lifetime. The former (i.e., the mesenchyma) will become the submucosal tissue when the mucous membrane spreading as a pharyngeal diverticulum comes to invest the auditory ossicles."[3]

"The full effect of erosion is demonstrated in the fetus of 7½ months. (Previous page) Throughout the length of the crus the mucous membrane occupies the former marrow-space of the ossicle, resting against the surface of the crus which was once internal." Except for the

persistence of marrow and the presence of a remnant of the obturator wall, the stapes is now an adult ossicle.

If one were to speculate as to the time that a congenital fixation of the stapes were to occur, it would seem that the disease process would have to come late in development, at about two to three months of gestation, since before this time there would be associated congenital defects in the mandible, maxilla or facial canal and nerve which would be evident in the patient. There is nothing written to indicate that there are any constant congenital anomalies that accompany the condition. Possibly there are abnormalities in other areas or processes of ossification that are present but are in large enough structures that there are no associated observable abnormalities. Possibly the pathology is not in the process of ossification but is the result of a generalized disease, which occurs at a specific time during gestation and causes pathology at a specific site (i.e., the stapes), as the rubella virus does. Since the exact cause is not known, metabolic and environmental conditions must also be considered.

ANATOMY OF THE STAPES

The head of the stapes is normally broad and shallow, and its surface is elliptical and concave for articulating

purposes. Its surface is in a near saggital plane. This permits the use of pressure through the incus on the stapes without causing one part to slide on the other, which could possibly result in joint separation.

The neck of the stapes may vary in diameter, construction, strength, and length. Its length does not always determine its strength. A long slender neck often will stand a great amount of pressure which might be exerted on it during operative maneuvers.

The crura of the stapes are arched structures which are fused into one at the neck of the stapes, and diverge as they meet the footplate. They are hollowed on the obturator surface, making them an inverted, rounded trough, the edges of which become more pronounced as they curve to meet the footplate. The trough is usually deeper at the base of the posterior crus than at the base of the anterior crus. "This hollowing out is the result of bony absorption and can produce a large number of variations. The crura may be extremely hollowed out to a very weak thin shell; on the other hand, there may be no visible evidence of erosion so that the crura are left very solid and large. This is an infantile type of crura. Bony projections, spurs and eroded-looking pits may frequently be seen, especially on the obturator surface."[10] The anterior crus is usually straighter, shorter, and more

slender than the posterior crus.

The footplate is rounded in an arch on its superior edge and is curved with a slight point anteriorly. It is straight on its inferior edge, and gently curved on the posterior edge. The footplate may be extremely long and narrow, being wedged into a narrow niche between the facial ridge and the promontory, or it may be generously wide with much of its area being superior to the crural insertion. The average length of the base of the stapes is 2.99 mm., and the average width is 1.4 mm.

The annular ligament connects the circumference of the stapes to the rim of the oval window. It is very frequently the site of otosclerotic bone invasion, especially in its anterior area. "Due to the embryologic development of the annular ligament, which is derived from the same tissue that later becomes the bony labyrinth on one side and the footplate of the stapes on the other, it is reasonable to assume that free cartilage or actual cartilaginous nests may develop and remain within its substance. These nests might become osteogenic and produce bony deposits capable of causing footplate ankylosis. Thin bridges of cartilage in the annular ligament presumably could become ossified in the middle decades of life."[10]

The most variable part of the stapes is the footplate.

It varies from an extremely thin, almost transparent, footplate to a thick, strong, opaque structure. These extremes seem to be normal variations. Otosclerotic infiltration may involve a small rim area, or may, when advanced, change a portion of the footplate into heavy otosclerotic bone. The anterior areas are the most commonly involved by otosclerosis.

AUDIOMETRIC EXAMINATION

"The essentials of a satisfactory method of mechanically measuring the hearing are--accuracy, quickness, and a technique which can be exactly repeated at a later date. The apparatus should be as simple as possible, rugged, not likely to get out of order and constant in performance."[15]

In congenital fixation of the stapes it is necessary to have ideal testing conditions since one sign of the disease is lack of progression of the patient's deafness. Therefore, if the original audiogram is in error due to the audiometer, the technician, or the responses of the patient, an incorrect diagnosis might be reached.

The difficulty of diagnosis of hearing defects varies with the difficulty of testing the patient and the complexity of their disease. In young children testing is difficult since accurate and constant responses are difficult to get. In especially young children the

problems to overcome are the restlessness and the inattention. Once past these two obstacles, one can begin to train the children in conditioned responses in the testing situations. These problems of response to testing are not present in older children or in teen-agers and adults who are tested.

The testing of patients with congenital fixation of the stapes might be divided into two categories:

(a) Those patients who are under five years in whom testing problems are present and in whom the diagnosis of an early hearing problem is newly recognized.

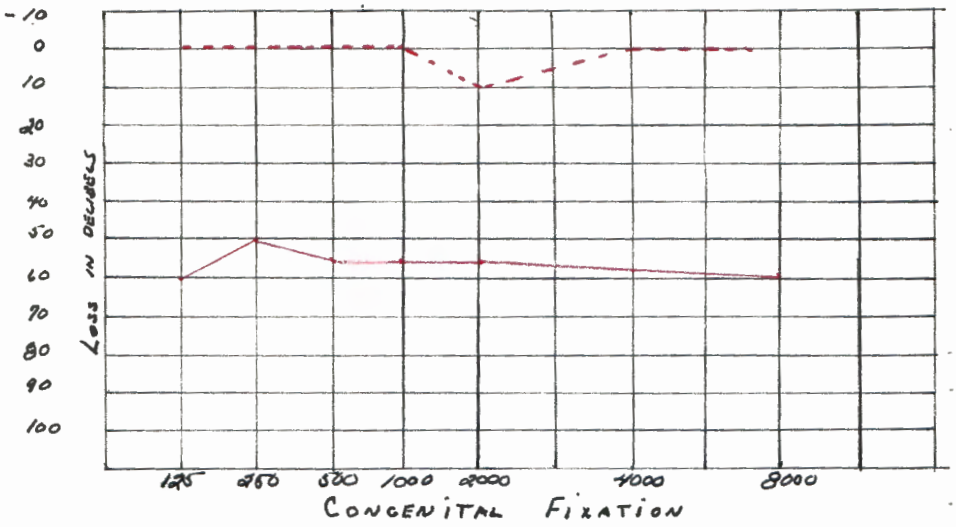
(b) Those patients who are in their teens or older in whom there is no testing problem and in whom a diagnosis of otosclerosis must be considered.

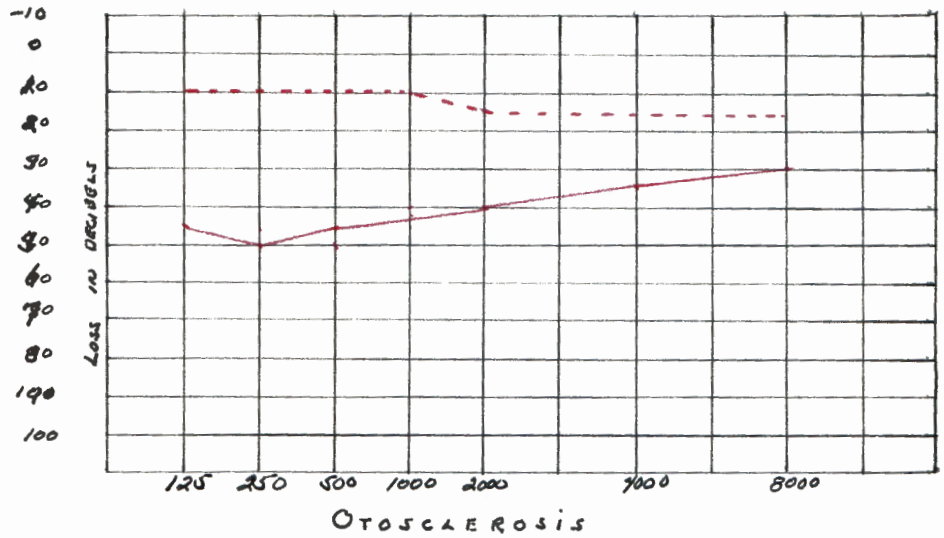
For the diagnosis of congenital fixation of the stapes to be made, the audiogram must show:

(a) Constant loss of 35-55 decibels in all frequencies.

(b) Lack of progression of deafness when followed audiometrically over a period of time.

"Typical" audiometric graphs one might see in congenital fixation and otosclerosis are shown in Fig. 6.





Air Conduction

Bone Conduction

Fig. 6

THE PATIENT

The child presenting with congenital fixation of the stapes is generally reported as being retarded in speech development and as possessing a limited vocabulary for his or her age. The age of presentation varies from five years upward. In one series,[11] the average age of presentation was ten years. Discovery of a hearing defect is usually made either in a routine audiometric check in association with school or by the parents when their child fails to hear or react to an outstanding sound that others in a group hear. These children are usually extremely good lip readers and thus appear to be attentive and interested children since they must continually pay attention to the speaker in order to follow the conversation.

THE DIAGNOSIS

In all patients, especially those older ones, otosclerotic deafness must be considered. The key to diagnosis or differential diagnosis is the lack of progression of the deafness as occurs in otosclerotic deafness. The lack of progression may be very difficult to establish. Once the defect is found or suspected in a child, parents often will take it upon themselves to conduct a

period of observation. During this time, they are much more aware of the possible hearing loss and, due to their new awareness, they feel that there is progression of the hearing loss. Actually there is not progression of the pathology. As an aid in the differential diagnosis, there is usually no familial history of deafness as is often found in patients with otosclerotic deafness.

The patient will present with an initial hearing loss, as stated before, of 35 to 50 decibels in all frequencies, which is the loss found in a completely interrupted ossicular chain, indicating total fixation of the stapes and lack of ossicular conduction. Bone conduction is essentially normal, remaining within ten decibels of the zero line.

In contrast, in otosclerosis, the air conduction curve usually shows the greatest loss in the lower frequencies. One must remember, though, that cochlear involvement is quite common in association with otosclerosis and results in abnormalities in the high frequencies which will give the audiometric picture of loss in all frequencies, as described above for congenital fixation. Generally, in a child under fifteen years of age with a probable history of progressive deafness who presents with normal physical findings, a pure conductive loss not to exceed 35 decibels in the speech frequencies,

consider him to have an otosclerotic deafness.

The differential diagnosis is certainly not limited to the two conditions compared here, although they are the main ones. Cases of synostoses between the malleus and incus have been reported and dislocations of ossicles and other causes for conductive loss must be investigated.

APPEARANCE AND FINDINGS AT OPERATION

Descriptions by House[11] at operation characterized the normal stapes as having no increase in vasculature about the footplate and with distinct margins about the periphery of the footplate and annular ligament. The central portion of the footplate is described as being thin and bluish in color and the ossicle is uniformly mobile.

"In otosclerosis, there is often an increased vascularity of the mucous membrane surrounding the footplate, usually in the anterior area. The margins of the footplate and the annular ligament are often difficult to visualize, especially anteriorly, because the otosclerotic bone frequently engulfs this area. The central portion of the footplate is often thin with a bluish discoloration. Whitish otosclerotic plaques are often seen spreading over the surrounding bone of the

otic capsule. Palpation reveals fixation of the footplate, usually in the anterior region."[11]

"In congenital footplate fixation there is no increase in vascularity of the mucous membrane surrounding the footplate area. The central portion of the footplate is difficult to visualize since the entire footplate bone blends into the bone of the surrounding otic capsule. Gentle palpation reveals uniform and total fixation of the entire footplate."[11]

INDICATIONS FOR CORRECTION

It is felt that a bilateral congenital defect should be corrected early, that is, at ages even less than a year, since a child who can't hear will not learn to speak early enough to maintain the pace of his contemporaries. Operation should be performed first on one ear, preferably the ear showing the greatest hearing defect, thus preserving the best ear of the patient for use in the event that operation is not successful. If the anomaly is unilateral, repair will result in the patient's being able to localize the direction of sound, which is an advancement for him. In unilateral cases, though, there is less urgency and operation can be delayed until the child is older and better able to tolerate operation and post-operative care.

The amount of hearing loss cannot be used as a criterion for operation in a true case of congenital fixation since it is used as a diagnostic criterion when the patient is first seen.

The present amount of disability to the young patient in a case where disability is slight is a poor reason to postpone operation. Whether it is evident or not, the defect is handicapping the patient continuously at a time when his learning capacities are at a maximum. It is at this time, then, that his perceptive powers should be at a maximum.

OPERATION TO BE PERFORMED

It has only been in recent years that otologic surgery has successfully been performed. In that time advances in instruments and knowledge have facilitated the otologic surgeon and have resulted in continued improvement of operation results. The initial operation to be performed depends upon the surgeon, his experience with procedures, and the conditions found at operation. Some authors advocate fenestration of the lateral semicircular canal initially, and others advocate stapes mobilization as the initial procedure to be followed by fenestration if the stapes mobilization does not result in an adequate return of hearing.[11,13]

In comparing the fenestration operation and the stapes mobilization operation, Goodhill states that "basic acoustic and anatomico-pathological differences exist between fenestration and stapedolysis. Since the fenestration operation is a detour procedure, and one in which the approach to the perilymphatic vestibule is remote from the area of stapes fixation, it may be regarded as a precise procedure for a specific purpose; namely, the rerouting of acoustic energy across a new air-perilymph junction to take the place of the closed oval window footplate region. The stapedolysis approach, however, directly attacks the very pathologic lesion which is responsible for the interruption of sound transmission across the natural oval window air-perilymph junction. Since the fixation process may occur in many varieties and in many quantitative degrees at this junction, no one operative procedure can be standardized and used in all cases in the same manner as the fenestration operation.

"The fenestration operation has a far more limited audiologic application than the stapedolysis approach. A physiologic deficit must occur in fenestration surgery, due to the almost complete loss of the impedance matching mechanism of the middle ear; therefore, the postoperative air conduction will seldom be better than

fifteen decibels below the preoperative bone conduction level. Since an average of thirty decibels loss in the speech frequencies is the lowest limit adequate for restoration of practical unaided hearing, it is necessary to start out with a good bone conduction level, one not lower than fifteen to twenty decibels in the speech frequencies.

"In stapedolysis the surgical objective is the lysis of obstruction in the stapediovestibular junction so that remobilization of the footplate to airborne sound may occur. Under ideal conditions, this approach completely respects the integrity of the tympanic membrane-ossicular chain mechanism so that the mechanical advantage of this impedance matching mechanism is not lost. Even though there is no minimal physiological deficit in stapedolysis comparable to that in fenestration, it must be recognized that deficits do occur. These deficits are either of pathologic origin or due to surgical complications."[7]

RESULTS OF OPERATION

In one series of 23 cases reported, 52 per cent received a significant improvement from stapes mobilization; 47 per cent remained essentially unchanged, and in 22 per cent the stapes could not be mobilized, and three of

these five cases underwent fenestration with resultant serviceable hearing.[11]

Follow-up of these cases after approximately one year reports satisfactory maintenance of the improved hearing status. A longer follow-up must be made to confirm the effectiveness of these operations in this condition.

COMPLICATIONS OF OPERATION

Complications have been few. It must be remembered that where one congenital anomaly is present there may be another. Thus there may be variations in the facial nerve and its branch, the chorda tympani. Marked variations are extremely rare. Minor variations are found commonly. There is usually a definite bulge of the bony facial canal just lateral to and above the footplate of the stapes. The chorda tympani may be very large or extremely small in diameter. It usually enters the bony external ear canal on its edge just lateral to the stapedius tendon. The surgeon must know where to find these structures normally and where to find the most common variants if he is to avoid damaging them and cause the patient to suffer the post-operative complications. Vascular anomalies in the middle ear are not found often, but damage to vessels has been reported as a cause of

post-operative tinnitus.[9]

Re-ankylosis as a complication has not been reported in this condition as yet. Perhaps this complication will appear in longer follow-ups as general knowledge of the condition increases.

SUMMARY

In this paper it was intended to consider the present status of congenital fixation of the stapes. In order to understand the middle ear and stapes, it was considered to be necessary to review its development and anatomy. This was done in detail. Diagnostic and differential diagnostic criteria were presented. Surgical factors were presented.

CONCLUSIONS

Development of the middle ear is an intricate process.

Congenital fixation is a condition that does exist, and can be diagnosed before operation.

The chief differential diagnosis is between the presented condition and acquired otosclerosis.

Operation will result in correction in a good percentage of patients, allowing the patient a more normal life.

ACKNOWLEDGMENTS

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