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Congenital stapes footplate fixation, Omaha, Nebraska

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CONGENITAL STAPES FOOTPLATE FIXATION

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HISTORICAL NOTE:

Congenital fixation of the stapes bone was first described in the literature written by Altmann[1] in 1949. He stated that, " In many cases of congenital atresia, improvement of hearing by operative intervention is doubtful due to the frequency of bony bridges between the stapes and the medial tympanic wall."

In 1952, G. B. Shambaugh reported five cases of congenital stapes fixation successfully operated upon.[2] This article also gave the first documented findings which differentiated this condition from otosclerotic stapes ankylosis. This paper will discuss the structure and function of the stapes with emphasis on the diagnostic points which differentiate this disease from otosclerosis.

EMBRYOLOGY OF THE STAPES

The stapes is formed from Reichert's cartilage,(the hyoid arch), which develops from the second branchial arch. The branchial arch origin of the ossicles is substantiated by examination of the innervation of the muscles of the middle ear. The malleolar muscle,(tensor tympani), is derived from the first branchial arch nd innervated by the trigeminal nerve, which originates in the first arch. The stapedial muscle is derived from the second arch and innervated by the nerve of that arch, the facial nerve.[13,4.] (Fig. 1.)

Study of the development of the stapes shows that the stapes belongs in a special catagory. At first, like a long bone, the stapes begins as cartilage. However, in other respects,a stapes differs from a typical element of the human skeleton. The stapes primordially is ring-shaped. Secondarily, it assumes the form of a stirrup. (Fig. 2)

Ossification takes place from a single center. Once formed in bone, the stapes does not become thicker because there is no external application of bone to the periostial layer. Also, the stapes does not lengthen after periostial bone has become continuous around the obturator foramen,(oval window). Unlike a typical long bone, one entire surface, that facing the obturator foramen, is removed continuously in the capital, crural, and basal portions of the stapes.

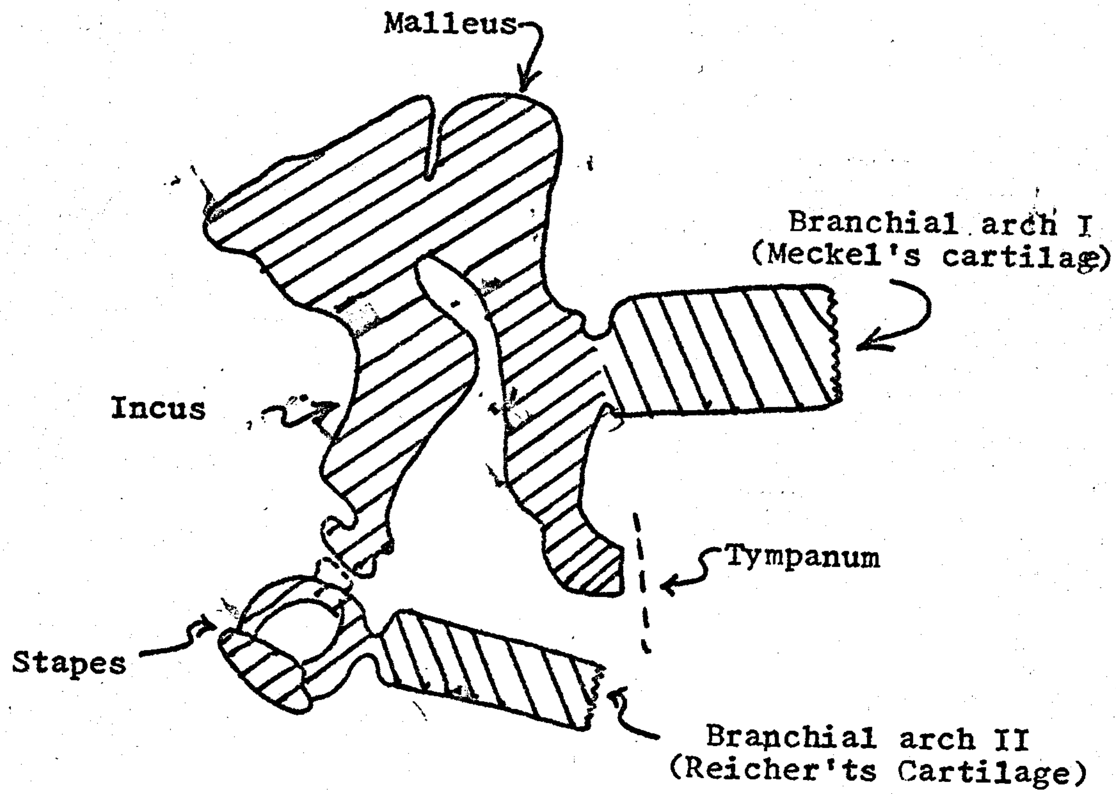

Fig 1 Orgin of the Auditory Ossicles from Branchial arches, illustrated in a diagram. (L. B. Arey, Developmental Anatomy, W.B. Saunders Co. Philadelphia, 1957.)

Marrow, in proportions comparable to the shaft of a typical long bone, is completely removed and is replaced by mucous membrane and submucosal tissue. Consequently, where-as a long bone becomes heavier as it lengthens (into the twenty to twenty-third year), a stapes loses bulk while it is attaining maximun dimensions and adult form (in the fetal stage).[5] (Fig.3)

A disturbed embryological development in the middle ear may be the reason for conductive deafness of congenital origin. The final causes of the disturbances are not always definitely clear and known. It is supposed that different viral diseases, hormonal and enzymatic imbalances of the mother, healed injuries of a fetal middle ear, drug abuse, and eclampsia can lead to congenital changes in the fetal middle ear.[6]

ANATOMY AND POSITION OF THE STAPES

The stapes runs almost horizontally. For this reason, the head of the stapes may be seen through a perforation in the tympanic membrane, but the crura are hidden from view. (Fig.4) The stapes footplate is covered at its edge with cartilage which is united with the cartilagenous rim of the oval window by the annular ligament. The ligament fulfills the double purpose of sealing the space while allowing the stapes to move in response to motions set up by sound.[3] The clearance between the stapes and the oval window is of the order of 0.1 millimeters. The narrow clearance of the footplate is essential to give an effective displacement of the cochlear fluids. These fluids offer considerable resistance to vibratory movement, and any gap between the footplate and the rim of the oval window would allow room for backflow, thus dampening the vibrations and decreasing hearing acuity.[7]

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MOVEMENTS OF THE STAPES WITH SOUND.

The two poles provide the strongest anchorage of the stapes footplate and the posterior pole is stiffer than the anterior. The motion of the hiad of the stapes, on positive pressure transmitted from the external auditory meatus, is forward, upward, and inward. The opposing phase, or outward motion of the ear drum, causes the stapes to move backwards, downward, and outward. This means that on positive pressure. the superior and anterior edge moves strongly toward the vestibule, but the posterior and inferior rim does not move at all. This motion has been likened to tapping the toe as it rests on the heal. The maximum swing occurs at the toe or anterior portion, while the heal or posterior pole remains essentially motionless.[3] These motions are used to transmit low intensity vibrations. As the intensity increases, the stapes gradually changes its pattern of movement until it resembles a tilting of the foot onto its side and rotating it around an axis which runs the length of the foot. With this motion one-half of the stapes moves inward and the other half outward. As a result a smaller amount of intra-cochlear fluid is pushed inward. This change of axis is considered a protective function of the middle ear.[3]

THE OSSICLES OF THE MIDDLE EAR AND THEIR FUNCTION

The ossicular system of the middle ear has the ability to transfer sound waves in air to fluid waves in the cochlea. The motion of the stapes is the final step in this process. The stapes baseplate rests on the oval window which is at the terminus of the scala vestibula of the cochlea. The head of the stapes is connected to the lever arm of the incus. The incus is connested to the malleus which is attached to the center of the tympanic membrane. Thus, the tympanic membrane transfers air vibrations into lever vibrations and the ossicular system of the middle ear transmits the vibrations to the fluid system inside the cochlea.

The lever system of the ossicles is such that when the handle of the malleus vibrates, the footplate of the stapes vibrates through one-half to one-third the distance, but with two to three times the force of movement of the han dle of the malleus. This is called pressure amplification. There is a good reason why the force of movement of the stapes must be increased. To transfer sound to the cochlea, the stapes must cause pressure waves in the fluid of the cochlea. This fluid has several times as much inertia as air. An increase of 10^3 power. Because of this inertia, much pressure is needed to move the fluid at the rapid frequencies of sound. However, thd fluid of the cochlea is contained within relatively strong walls and cannot vibrate through long distances. Consequently, the cochlear fluid need not vibrate through as long an excursion arc as that through which the tympanic membrane vibrates, but a greater amount of pressure is required to make the fluid vibrate at all. The ossicular system of the ear can be called a pressure amplifier, for it amplifies the pressure of the sound vibrations from high-amplitude, low-pressure air vibrations to low-amplitude, high-pressure fluid vibrations.[8]

AUDIOMETRY

Sound is percieved when the nerve impulses which originate in the cochlea reach the cerebral cortex. But there are two means by which the cochlea may be stimulated, by air conduction and by bone conduction. Air conduction utilizes the tympanic membrane and the middle ear ossicles to transmit air vibrations to fluid vibrations as described above. Bone conduction bipasses the conduction bones of the middle ear and sets up fluid vibrations in the cochlea when the activating source of sound is placed upon the head. By the use of the audiometer, the clinition can test each mode of conduction separately and divide his patients with hearing defects into three catagories, 1) pure conductive deafness involving the middle ear ossicles, 2) pure perceptive deafness which is a nerve deficit, and 3) a combination of the first two. This separarion is important because the first and third can be aided by surgery.[4,7]

The audiometer is the most precise instrument for sound production available. Either pure tones or a voice of controlled intensity can be employed. Most importantly, exact records may be made and used for comparison with subsequent examinations. The data obtained are recorded by means of graphs and visualize to the physician the defects in hearing and give a partial indication of the type of disease causing deafness.

The audiometer is acoustically a generator of approximately pure tone which may be varied in pitch and intensity. The ossilator has a frequency range which extends from 100 to 1,000 double vibrations (cycles) per second. this range is divided into eight or more steps of frequency, 64, 128, 256, 512, 1024, 2048, 4096, and 8192 cycles per second. The sound intensity is measured in decibles. The decibel scale is a logrhythmic one. That is, ten decibels is a ten fold increase in acoustic pressure; twenty decibels, a hundred fold increase, and thirty decibels, a thousand fold increase. Zero on the audiometric scale is not absolute zero of acoustic pressure but the smallest pressure appreciable by the average human ear, usually 0.0002 dyne/cm^2. For this reason individuals with more than average acuity will rate less than zero.[4]

One of the most disturbing aspects of pure-tone testing is the difficulty in determining which ear is responding to bone conduction signals since the entire skull is vibrated. If the bone conduction is different in the two ears, the better ear will pick up the vibrations and it will be difficult to evaluate the poor ear. One method used to overcome this problem is masking. The bone conduction reciever is placed in contact with the mastoid bone of the ear to be tested and the airconduction reciever is placed over the opposite ear. A continuous or intermittant tone is transmitted over the air conduction reciever. This enables the patient to more accurately determine the threshold of bone conduction for the weaker ear.[4,9.]

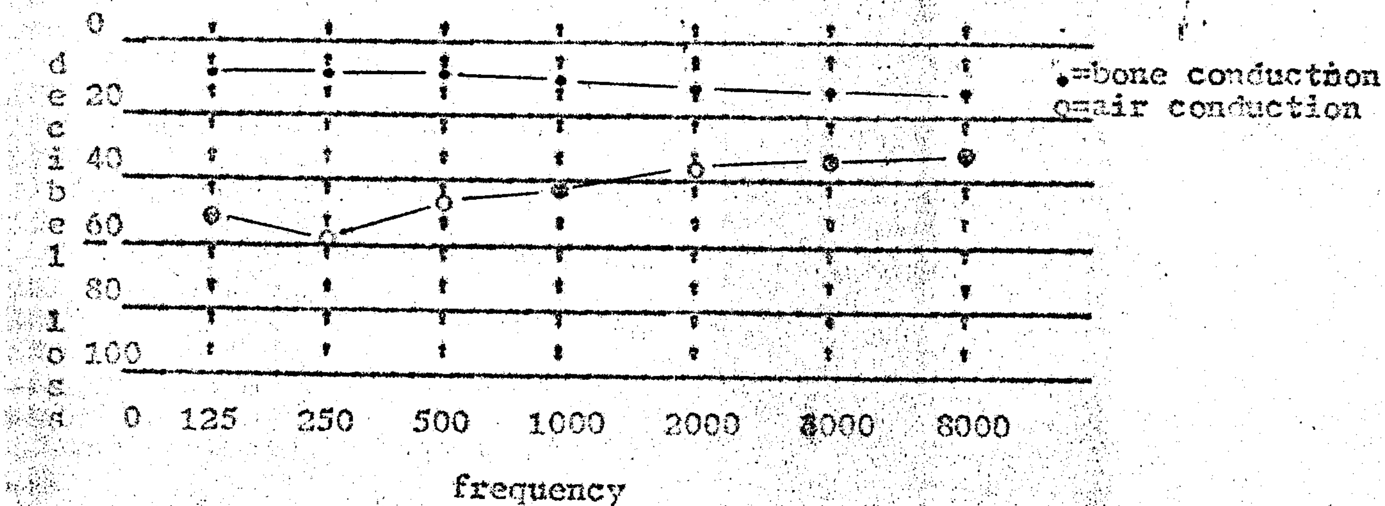
DIFFERENTIATING CONGENITAL STAPES FIXATION FROM CLINICAL OTOSCLEROSIS

History:

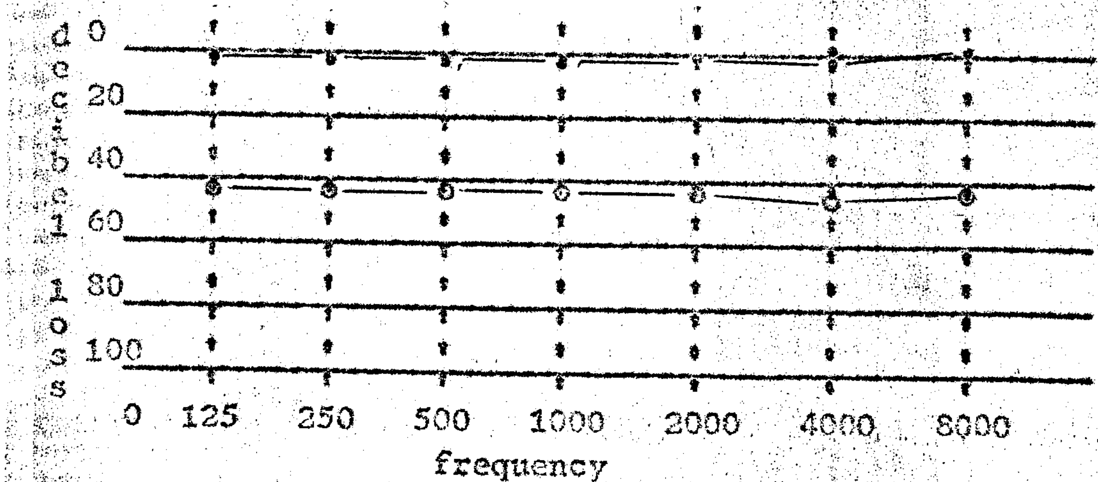
The key to differentiating clinical otosclerosis and congenital footplate fixation is progression of the hearing loss in the former and none in the latter. Progression is often very slow before adolescence. This makes accurate diagnosis before the age of fifteen difficult. Progression must be determined by serial audiograms not by subjective feelings of increased loss. Often patients accidentally become aware of their loss and there after, continually test their ability. Their loss will seem to be getting worse with increased attention. Patients also feel their loss is increasing as they grow older and increased demands are made of their hearing with adult activities. The average age of discovery is ten years. It is not earlier because these children have grown up with their impairment and have compensated by becoming expert lip readers. Many will give a history of being slow to learn to talk but all have excellent speech patterns.[10,11,12,13,2].

Typical Audiograms

The audiogram usually varies between the otosclerotic group and the congenital fixation group. Otosclerotic patients conductive loss usually does not exceed 35 decibels. Often there is cochlear involvement over 10 decibels in the higher frequencies. Air conduction loss is usually greater in the lower frequencies. The audiogram of a typical patient with congenital footplate fixation shows a flat 45-55 decibel pure conductive loss with no progression with serial readings. Typical audiograms represented below:[10,11,12,13,2.]



OTOSCLEROSIS



CONGENITAL STAPES FOOTPLATE FIXATION

Observations at Surgery:

The normal stapes footplate margins and the annular ligament are distinct and there is characteristically no increased vascularity. The center of the footplate is thin and has a bluish appearance. Gentle palpation reveals uniform mobility of the entire footplate.

In congenital fixation of the footplate there is no increased vascularity. The central portion of the footplate is no longer thin and it has lost its bluish discoloration. The footplate bone blends into the bone of the surrounding otic capsule making the margins of the footplate and the annular ligament difficult to visualize. This diffuse bony fusion may be thick or thin.[10,12] Sometimes it is impossible to know where the labyrinth is located because of marked anomalies such as a deep niche and nothing but two small crura ending blindly against hard undifferentiated labyrinthine capsular bone.[14] In all cases gentle palpation reveals uniform and total fixation of the entire footplate.

The footplate fixed by otosclerosis shows increased vascularity, usually in the anterior area. The annular ligament and footplate margins are visible posteriorily but difficult to visualize anteriorly. The center of the footplate shows a bluish discoloration and is thin. White otosclerotic placks are often seen spreading over the otic capsule and engulfing the anterior crus at its footplate attachment. Gentle palpation reveals fixation of the footplate in the anterior region.[10, 12]

Summary:

	Otosclerosis	Cong. Footplate fixation
A-History:		
progression:	present	not present
age of discovery	young adult	childhood
speech development	average	slow
family history	often positive	seldom positive
tennitis	often present	seldom present
B-Physical: (audiometric findings)		
air conduction	greater loss in low frequency	flat
amount of loss	varies with fixation	50 decibels - indicating total fixation
Cochlear involvement	common	rare
bone conduction	drops at higher frequency	no drop at higher frequency
C-Surgical Observations:		
vascularity of middle ear mucosa	increased	normal
central footplate	thin and bluish	seldom thin or bluish
bony appearance	whitish placks often present	no whitish placks

(Summary continued)

footplate margins & annular ligament	readily distinguishable	not distinguishable
degree of fixation	partial to total	total
location of fixation	usually anterior	uniform throughout
mobilization	average case less difficult	average case more difficutt
fenestration	some not suitable	all suitable[10]

CONCLUSIONS

Congenital stapes footplate fixation is a rare disease of children which can be diagnosed by a detailed history and serial audiograms. It lends itself well to surgical exploration. Servicable hearing is possible in most cases because of the uniform lack of cochlear involvement.

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