

Symptom: Congenital Conductive Hearing Loss

By Hamid R. Djalilian, MD

A 6-year-old girl presents to the office with her mother, who says that the child has had hearing issues.

The child did not pass the newborn hearing screening for her left ear. Since she had other medical problems needing treatment, her mother did not pursue the hearing issue at the time.

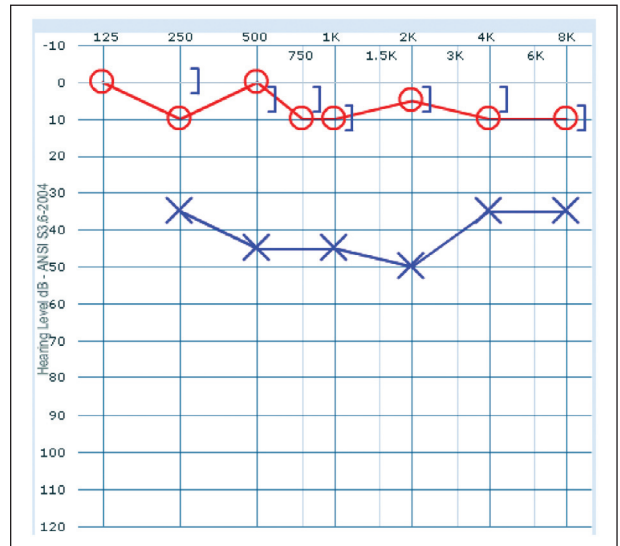
Her mother recently noticed that the television is louder and her daughter turns up the iPad volume when watching cartoons. The child also seemed to be ignoring her parents and didn't respond quickly when there was background sound.

The child does not have a significant history for otitis media, nor has she had pressure equalization tubes placed, her mother said.

The patient's microscopic ear exam is normal. Her audiogram is shown on the right.

What is your diagnosis? See p. 10.

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The patient's audiogram on presentation.

Diagnosis: Malleus and Incus Fixation

By Hamid R. Djalilian, MD

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The current scenario described in our patient is pediatric conductive hearing loss, which is probably congenital in nature. An acquired conductive hearing loss in children is most often due to chronic otitis media with effusion, either mucoid or serous otitis media.

An accumulation of middle ear fluid after Eustachian tube dysfunction can cause a conductive hearing loss that generally reverses when the middle ear fluid is resolved.

In the absence of middle ear fluid and the presence of a normal tympanic membrane, the likeliest explanation for conductive hearing loss is ossicular fixation. The most common cause of congenital ossicular fixation is stapes fixation.

SOURCES OF FIXATION

Even though pediatric stapes fixation is called otosclerosis, it is not caused by the same pathophysiologic mechanism. In otosclerosis, an abnormal growth of inner-ear bone leads to stapes fixation. In congenital stapes fixation, the separation between the footplate and the surrounding otic capsule bone does not occur.

Fixation of the other ossicles can also cause significant conductive hearing loss. The malleus is usually fixed at the anterior malleal ligament, which can be calcified, or the head of the malleus can be adherent to the medial-most portion of the ear canal.

The cause of malleus fixation is not fully known. One possibility is tympanosclerosis, whereby scar tissue in the middle ear becomes calcified. During this process, the anterior malleal ligament can become scarred and subsequently calcified, rendering the malleus immobile.

In some cases, previous surgery can lead to bone dust gathering in areas around the ossicles, causing their adhesion to the surrounding bone. Temporal bone trauma (e.g., fracture) also can result in scarring at the incudomalleal joint and fixation.

While otosclerosis is most commonly associated with stapes fixation, it has been described as a rare cause of malleus fixation. However, since otosclerosis is a disease of the otic capsule (i.e., the inner-ear bone), it would be highly unusual for it to involve the malleus.

Congenital anomalies in the development of the epitympanum (i.e., the portion of the

middle ear above the tympanic membrane) have been implicated as well, with a smaller epitympanic cavity thought to lead to malleus fixation.

THREE TREATMENT OPTIONS

A preoperative CT of the temporal bones is obtained to better understand the cause of the conductive hearing loss. The imaging also helps rule out other explanations for conductive hearing loss, such as congenital cholesteatoma or inner-ear causes, including superior canal dehiscence or enlarged vestibular aqueduct, among others.

The CT scan in this patient showed fixation of the malleus head and the incus body, as shown in *figure 1*.

The patient with congenital conductive hearing loss is generally offered three treatment options:

1. Leave the condition alone. However, a child with unilateral hearing loss has a much higher likelihood of failure in school and a lower achievement level. Therefore, this is not a great option.
2. Use a hearing aid. Some parents choose this option, as they are risk averse and would like to avoid surgery.
3. Surgery offers the possibility of permanent hearing improvement. The success of surgery depends on the pathology and on the experience of the surgeon.

Many surgeons replace the fixed portions of the ossicles with a prosthesis or an incus interposition graft. It is, however, advisable to do everything possible to preserve the anatomy of the middle ear.

This patient had surgery, with a left transcanal approach to the middle ear used. The malleus was fixed at the anterior ligament. The incus body and short process were fused at the scutum (medial portion of the external auditory canal).

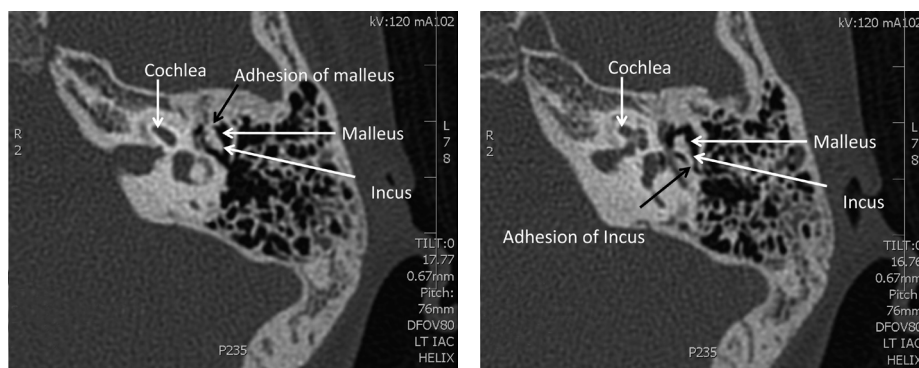


Figure 1. Left: This axial CT of the left temporal bone demonstrates the adhesion of the malleus head to the surrounding bone. Right: This axial CT of the temporal bone shows fixation of the incus. The normal malleus head and incus body (which together look like an ice-cream cone) should be surrounded by black areas (air and thin ligaments). In this patient, there is a white (bony) adhesion to surrounding bone.

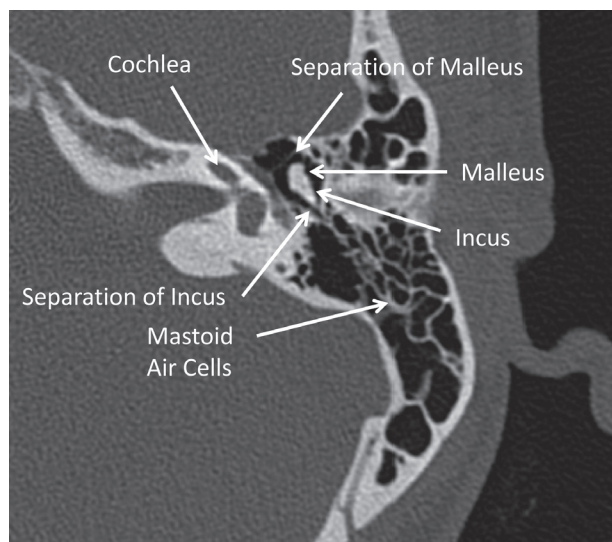


Figure 2. This image of a normal temporal bone shows a clear separation between the malleus and the epitympanum, as well as between the incus and the scutum/mastoid. On CT images, note that bone is white, air is black, and soft tissue is gray.

The anterior malleal ligament was lysed with an argon laser, and the anterior canal was drilled a bit to create a separation with the malleus and prevent re-fusion.

The incus was freed by removing the bone surrounding the incus with a combination of laser, drill, and curette.

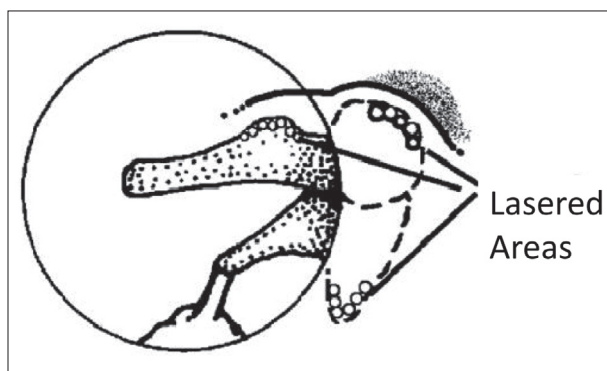


Figure 3. This drawing of the left ear in a surgical (lying down) position demonstrates areas that were lasered in our patient. (Adapted with permission from: Seidman MD, Babu S. A new approach for malleus/incus fixation: no prosthesis necessary. *Otol Neurotol* 2004;25[5]:669-673. Adaptations are themselves works protected by copyright. In order to publish this adaptation, authorization must be obtained both from the owner of the copyright in the original work and from the owner of copyright in the translation or adaptation.)

Gelfoam dissolvable sponge was placed to prevent readhesion of the canal to the incus. Other methods of preventing readhesion include the use of thin silicone (silastic) sheeting.

This patient had a complete closure of the air-bone gap that has been stable for 2.5 years. 