

CLINICAL CONSULTATION

Symptom: Maximum Conductive Hearing Loss

By Hamid R. Djalilian, MD

A 7-year-old boy is brought in by his mother for the evaluation of hearing loss. The child was born on time after an uncomplicated gestation. He did not spend any extra time in the hospital.

The boy just completed first grade and struggled in school. He moved to the United States two years ago after being born outside the country.

His mother brought an audiogram showing a maximum conductive hearing loss on the right side. She does not feel that her child has a hearing issue. The patient's ear exam is pictured in the photo on the right.

What is your diagnosis? See p. 14.

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This photo shows our 7-year-old patient's auricle.

iPad Exclusive!

FOUR BONUS VIDEOS: VISUAL DIAGNOSIS

Read this month's Clinical Consultation case, and then watch the accompanying videos from Hamid R. Djalilian, MD, to review the patient's imaging for yourself.

- Video 1: The axial CT shows the anatomy.
- Video 2: A microscopic view of the coronal CT.
- Video 3: The sagittal CT.
- Video 4: The axial CT of another patient with congenital atresia shows a favorable anatomy for reconstruction.
- Video 5: The coronal CT of the patient demonstrates a favorable course of the facial nerve.

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Diagnosis: Congenital Atresia of the Ear Canal

By Hamid R. Djalilian, MD

Continued from p. 12

The patient's auricle appears somewhat normal; however, when attempting to do an otoscopy, the clinician realizes that there is no ear canal. The child has congenital atresia of the ear canal in the presence of a normal auricular exam.

The abnormality of the external ear (auricle) is termed microtia, which has certain grades (see table 1) and is categorized as stenosis or atresia. Stenosis, as the name implies, is a narrowing of the ear canal, whereas atresia is a complete closure of the ear canal.

Grading of atresia primarily depends on the anatomy seen on CT scanning. Congenital atresia can be due to a syndromic or non-syndromic etiology. It is more common on the right side for an unknown reason.

BONE-CONDUCTION DEVICES

In the past, hearing rehabilitation for children with unilateral atresia was not considered necessary. However, recent studies suggest that unilateral hearing loss in children can cause substantial school performance problems and, possibly, brain development issues.

There are several methods of hearing rehabilitation for these children. The first is the use of a bone-conduction hearing device. The traditional bone-conduction hearing device of the past has given way to the newer soft-band bone-conduction devices.

Three devices currently on the market are the Baha, Sophon, and Ponto devices. The advantage of these devices is the greater degree of comfort compared with the vice-like grip of traditional bone-conduction hearing aids.

One of the main issues with these devices is patient compliance. While children tend to be compliant during preschool age, teasing at school, or the fear of such, oftentimes leads to the child taking off the device at school and only wearing it before coming home. These devices also provide the option of later being converted into a semi-implantable device to obviate the need for the headband.

Partial implantation of these devices can improve patient compliance in the school-age child. The Sophon and Baha devices both offer a magnetic anchoring of the device on the head. During surgery, an implantable magnet is placed on the flat portion of the skull approximately 5 cm from the ear canal. After a healing period and osseointegration, the external device and its associated magnet can be placed. The significant advantage of the magnetic devices is that the skin overlying the

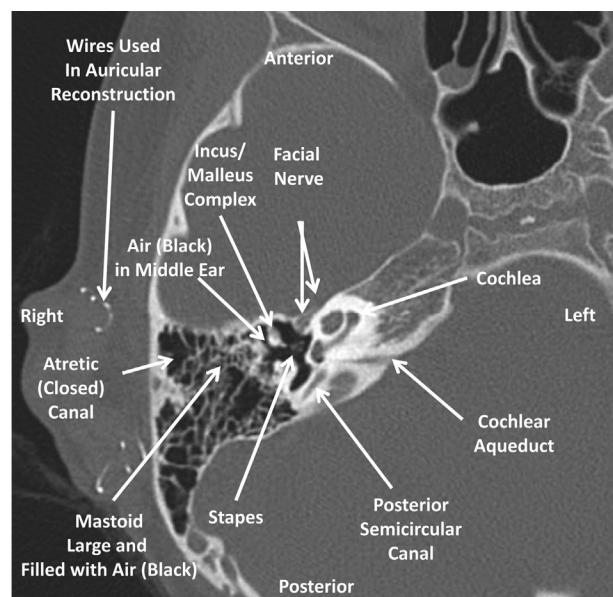
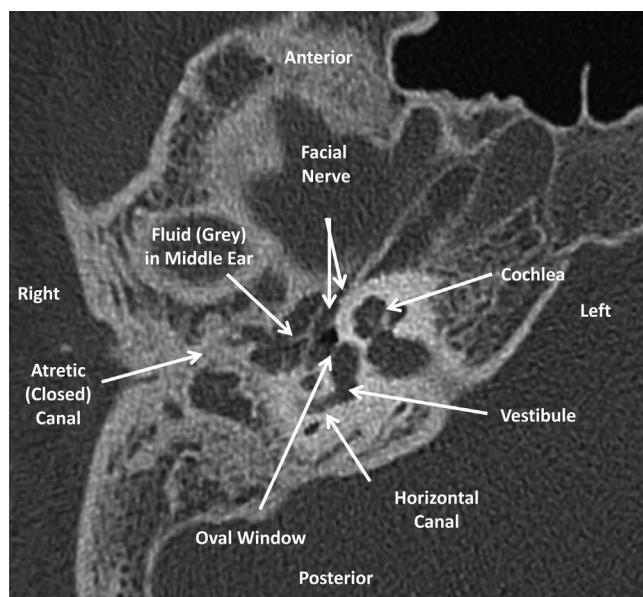


Figure 1. Left: Axial (horizontal) CT of the temporal bone on the right side of this patient shows a contracted middle ear, fluid in the mastoid, and the facial nerve encroaching on the oval window. Right: Axial (horizontal) CT of the temporal bone on the right side of another patient shows favorable anatomy for atresia repair, including the presence of the ossicles, a well-developed mastoid, and an aerated middle ear.

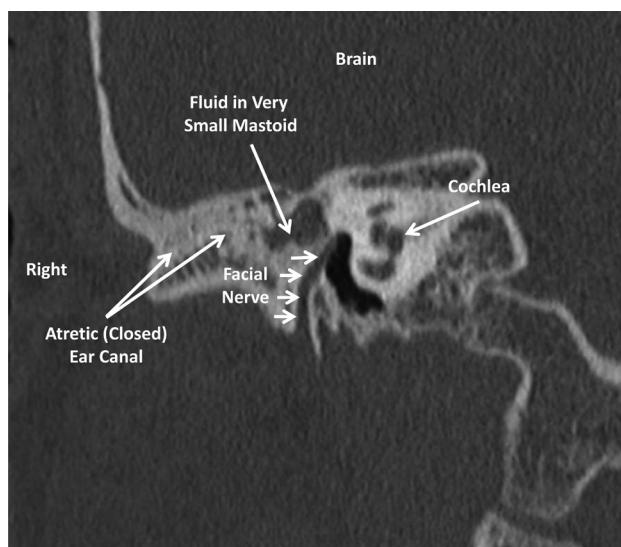


Figure 2. Coronal CT shows very contracted middle ear space, with the facial nerve taking an abnormal course, blocking a potential reconstruction of the ear canal.

implanted portion remains intact. Therefore, the risk of infection or skin breakdown is minimal.

Ponto requires the placement of an implant (screw) into the skull, with an overlying abutment that protrudes through the skin and allows the hearing device to be directly connected to the skull. The advantage of the Ponto device is that this direct connection enables substantially better high-frequency amplification compared with magnetic devices. The Baha device permits the use of the same mechanism, which also allows for better amplification (e.g., in patients with mixed hearing loss).

In Europe, the Vibrant Soundbridge has been used for patients with atresia. The Med-El Bonebridge, which has not been FDA approved, has been used for these patients as well.

INDICATIONS FOR SURGICAL REPAIR

The final option for hearing rehabilitation for patients with congenital atresia is repair of the atresia and opening of the ear canal. Although on the surface this procedure appears to be as simple as drilling a hole in the skull, it is quite complicated and best performed by surgeons experienced with the procedure.

Not every patient is a candidate for this repair. The patient's anatomy has to have certain characteristics that improve the chances of success, including the presence of the stapes, an open round window, aeration of the middle ear, sufficient space in the middle ear, mastoid aeration, and favorable position of the facial nerve. A CT scan of the temporal bone is performed to assess the anatomy of the ear. Based on the results, the surgery can be discussed with the family.

The surgery requires identification of the anatomy, reconstruction of the middle ear if necessary, creation of the tympanic membrane, and placement of skin to reconstruct the skin of the ear canal.

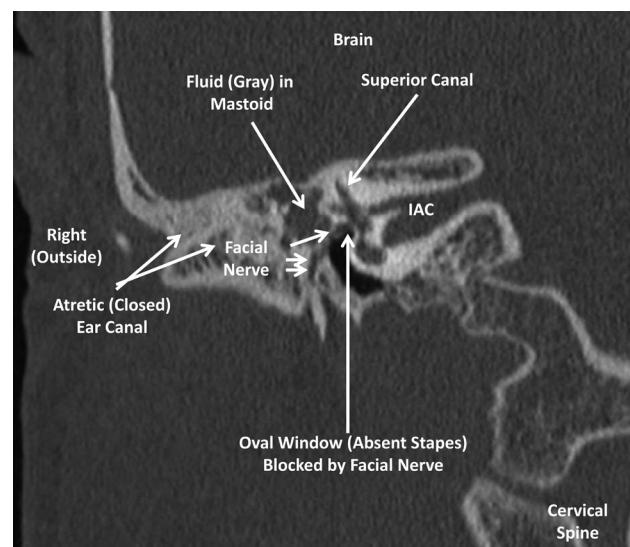


Figure 3. Coronal CT of the patient shows the facial nerve blocking the oval window and the absence of ossicles. "IAC" stands for internal auditory canal.

The revision rate is relatively high—25 percent to 40 percent. Surgical revisions are most commonly due to narrowing of the meatus from scar formation; narrowing of the bony canal from bone growth, commonly seen during the child's growth spurt; and lateralization of the tympanic membrane (where the tympanic membrane moves outward and separates from the incus-malleus complex due to scarring).

After surgery, close follow-up of the patient is necessary to address complications early and to intervene to prevent another surgery. The patient also requires intermittent ear cleaning, as the skin taken from the arm or thigh to line the ear canal does not have the natural outward migration of keratin. The patients are asked to use a foam earplug at night to create counter pressure against scar formation in the ear canal.

Reconstruction of the auricle can be done in several different ways. The first option is an auricular prosthesis made from silicone and shaped exactly the same as the other ear. The prosthesis can be anchored to the head via implants or glued on a daily basis.

Other options include surgical reconstruction using rib cartilage or a biocompatible implant (Medpor). Generally, the cosmetic reconstruction of the ear is performed first, followed by hearing restoration procedures. This order is followed because the cosmetic reconstruction depends on some tissue planes and skin to be intact. Certain skin incisions done for the atresia surgery can disrupt the blood supply to the reconstruction later and lead to failure.

This patient had a CT scan, which showed that his anatomy was not appropriate for reconstruction of the ear canal (see figures 1-3). The patient's malleus-incus complex was absent, the oval window was blocked by the facial nerve, and the contracted mastoid and middle ear were filled with fluid.

Table 1. Microtia Grading

Grade 1: The ear is smaller than normal, although most of the features of a normal ear, such as a well-defined lobule, helix, and antihelix, are present.

Grade 2: The normal features of the ear are missing. There is still a lobule and a remnant of the helix and antihelix.

Grade 3: The ear consists of a vertical skin appendage with a malformed lobule (earlobe) on the lower end.

The middle ear and mastoid fluid, in addition to the contracted mastoid, indicates that Eustachian tube function is poor, and, even if a reconstruction could be performed, the results would be suboptimal as the patient would likely still have middle ear fluid, leading to a significant additional conductive hearing loss. The facial nerve's course in this patient will not allow for any reconstruction of the middle ear, as connecting the stapes to the tympanic membrane would be impossible. 

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