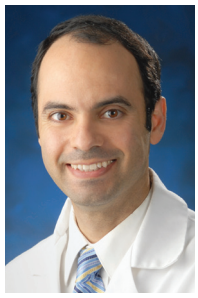


Symptom: Unilateral Hearing Loss

By Hamid R. Djalilian, MD

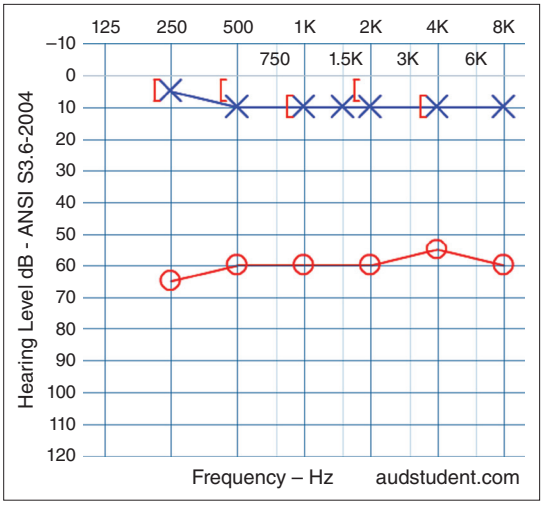
A 6-year-old patient has had hearing loss on one side since birth, failing newborn hearing screening on the right side and passing on the left, the patient's mother said. The child had no issues in speech and language development, so the mother did not follow up further.

The patient now is to start school, however, and a recent hearing screening revealed a near maximum conductive hearing loss on the right side, with normal hearing on the left.

The examination of the child shows a very narrow ear canal impacted with cerumen. The canal appears as a slit filled with debris.

What is your diagnosis? See p. 16.

Dr. Djalilian is director of neurotology and skull base surgery and associate professor of otolaryngology and biomedical engineering at the University of California, Irvine.



This is the audiogram for our 6-year-old patient, who has had unilateral hearing loss since birth.

Diagnosis: Congenital Stenosis of the External Auditory Canal

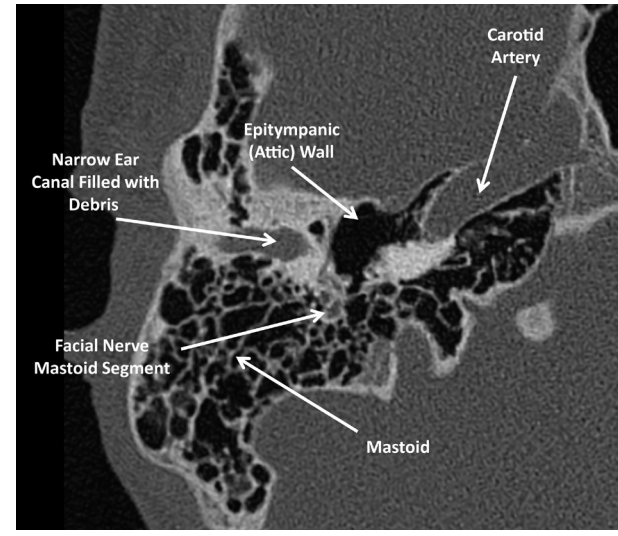
By Hamid R. Djalilian, MD

Continued from p. 14

In a child, unilateral hearing loss can cause difficulties in school performance. The condition leads to problems with speech understanding in noisy environments, such as classroom discussions, as well as localization of sound. Consequently, the identification and management of patients with unilateral hearing loss is of significant importance.

Most commonly, unilateral conductive hearing loss in children is due to middle ear effusion after an upper respiratory tract infection or acute otitis media. However, significant conductive hearing loss (greater than a 30-db air-bone gap in the pure-tone average) would be rare from just middle ear effusion. Therefore, in the presence of significant conductive hearing loss, further follow-up is necessary to evaluate hearing after resolution of the middle ear effusion.

Large conductive hearing loss is seen in children with ossicular fixation, dense cerumen impaction, tympanic membrane perforation, congenital atresia of the ear canal, and congenital stenosis of the external auditory canal, among other conditions.



This axial CT of the temporal bones shows a very narrow ear canal filled with debris. The patient's canal is 3.1 mm in diameter.

AMPLIFICATION VS SURGERY

Our patient had congenital stenosis of the external auditory canal. The external auditory canal develops in the 18th week of gestation, long after the development of the inner ear structures, which starts in the fourth week. As a result, congenital abnormalities of the ear canal are very rarely associated with a malformation of the inner ear.

"Generally, an ear canal is considered stenotic if its diameter is less than 4 mm."

Congenital atresia of the ear canal is a complete blockage of the external auditory canal, while congenital stenosis is a significant narrowing of the canal. Generally, an ear canal is considered stenotic if its diameter is less than 4 mm.

Patients with congenital stenosis of the ear canal have varying middle ear abnormalities, such as a hypoplastic tympanic membrane, tympanic membrane not connected to the ossicular chain, fixed ossicular chain, and fused malleus–incus complex. The stapes generally is normal, as it is embryologically derived from the second branchial arch.

The workup of these patients requires a CT of the temporal bone to evaluate for cholesteatoma in the ear canal. The likelihood of canal cholesteatoma is much higher in patients who have congenital stenosis than in those with complete atresia.

Treatment of children with congenital stenosis of the external auditory canal can include amplification or surgery, depending on the parents' preference.

While air-conduction hearing aids can be attempted in these patients, the very small nature of the ear canal precludes obtaining a proper mold, making the option less successful. Traditional bone-conduction hearing aids can be used, though these devices have given way to softband bone-conduction devices.

Softband bone-conduction devices, such as Sophono, Ponto, or Baha, can later be employed for rehabilitation if the parents opt for a bone-conduction

FOR MORE INFORMATION

Read these recent *HJ* articles for additional details about unilateral hearing loss:

- Single-Sided Deafness: Causes, and Solutions, Take Many Forms
  bit.ly/HJ-SSD
- Approaching Unilateral Hearing Loss from Both Sides
  bit.ly/HJ-Unilateral

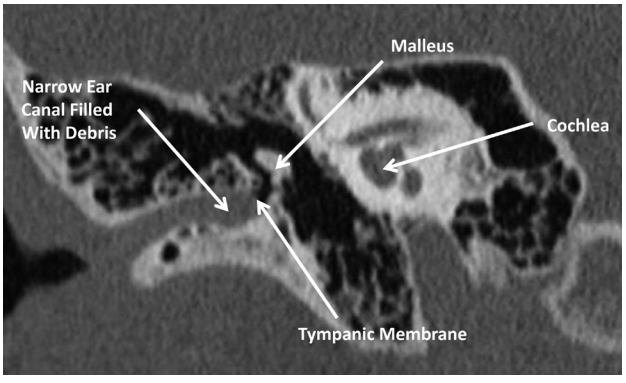
implant procedure, reducing the need to purchase multiple devices over time.

Surgical options include the placement of a percutaneous or transcutaneous bone-conduction device. Percutaneous devices, such as Ponto or Baha Connect, require the placement of a small screw (implant) within the skull, with an attached abutment that protrudes through the skin. The hearing device is then snapped onto the abutment. The percutaneous attachment provides for better high-frequency sound transmission but requires daily cleaning and can lead to problems like infections and skin overgrowth.

"The workup of these patients requires a CT of the temporal bone to evaluate for cholesteatoma in the ear canal."

Transcutaneous devices, such as Sophono and Baha Attract, use an implanted magnet that is driven by an external magnet to transmit bone conduction sound to the cochlea. These devices have the advantage of not requiring the maintenance of a percutaneous attachment. The disadvantages of these devices are decreased sound transmission in the higher frequencies and a larger degree of distortion if an MRI of the brain is obtained.

The other surgical option for the treatment of congenital stenosis of the ear canal is opening the canal. The surgery is best performed by an otologist/neurotologist with experience in this type of procedure, which involves widening the canal and reconstructing the tympanic membrane as well as skin grafting.



This coronal CT of temporal bones shows a narrow ear canal with the tympanic membrane not adherent to the malleus.

Sometimes, the stenotic canal ends in a bony atretic plate that is lateral to the tympanic membrane and requires removal to expose the malleus. It is best to remove the last residua of the atretic plate with a laser rather than a drill, as drilling on the malleus can cause a high-frequency sensorineural hearing loss.

Read past Clinical Consultation columns in a special collection on the *HJ* website: bit.ly/HJClinicalConsultation.