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Imaging of congenital temporal bone anomalies

Ali R. Sepahdari, MD,[a] Brian D. Zipser, MD,[b] Michael N. Pakdaman, MD[a]

From the [a]Department of Radiological Sciences, David Geffen SOM, University of California, Los Angeles, California; and the [b]Department of Radiology, Olive View Medical Center, Sylmar, California

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A variety of congenital malformations of the ear and temporal bone may be encountered. Anomalies of the inner ear, middle ear, and external auditory canal and auricle can all occur, and there are a variety of clinical presentations that include conductive and sensorineural hearing loss. Some malformations are part of well-described genetic syndromes, whereas others are sporadic. Cochlear implantation may be helpful in some cases. Knowledge of typical patterns of abnormalities can help guide the clinical workup and management plan. The goal of this review is to familiarize the reader with the role of imaging in some of the commonly encountered congenital malformations of the ear and temporal bone.

Introduction

When evaluating the pediatric patient with hearing loss, imaging is essential before considering corrective surgery. Congenital anomalies of the temporal bone may produce conductive hearing loss (CHL) or sensorineural hearing loss (SNHL). Imaging is essential in determining whether a patient is likely to benefit from external middle ear corrective surgery in patients with CHL or from cochlear implantation in cases of SNHL and is essential also for guiding workup for a potential underlying genetic syndrome.

Imaging modalities

Computed tomography (CT) and magnetic resonance imaging (MRI) are the 2 imaging modalities that are commonly used to evaluate the pediatric patient with hearing loss. Some centers use only 1 modality, whereas others (including our institution) often use both.[1] We find CT and MRI to be complementary tests. CT is superior for showing the middle ear anatomy, anomalies of the ossicular chain, and course of the facial nerve canal. Patency of the labyrinthine oval window and round window niche, volume of the middle ear cavity, and anatomical constraints to successful auriculotympano or ossiculoplasty and cochlear implantation are best seen by CT. CT and MRI are similarly sensitive for detecting malformations of the inner ear. Although CT can suggest cochlear nerve deficiency (CND) if the cochlear aperture or internal auditory canal (IAC) is small, MRI is superior in detecting CND through direct visualization of the nerve.

Attention to detail is important in ensuring that diagnostic quality images are obtained for both CT and MRI. Close communication between the otologist and the radiologist is helpful in this regard. For CT, it is preferable to perform the studies on a newer model multidetector row scanner (eg, 64 detector row or higher), though high-quality images can still be obtained on older scanners, albeit at the cost of higher radiation dose. It is critical to obtain thin-section images reconstructed in a sharp bone algorithm with a small field of view, ideally with separate small field-of-view reconstructions of the right and left sides separately so as to maximize spatial resolution. With newer model scanners, a volumetric axial acquisition can be reconstructed in the axial and coronal planes and in oblique planes as needed. With older scanners, direct coronal imaging may be

Address reprint requests and correspondence: Ali R Sepahdari, MD, 757 Westwood Plaza, Suite 1621D, Los Angeles, CA 90095.
E-mail address: ali.sepahdari@gmail.com; asepahdari@mednet.ucla.edu

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necessary. When performing MRI, it is of critical importance to obtain high-resolution, 3-dimensional (3D), heavily T2-weighted "cisternographic" sequences to visualize the vestibulocochlear nerves and the labyrinthine structures. These sequences have different names and acronyms according to the MRI manufacturer and the technique that is used to produce the image. Some examples include Constructive Interference in Steady State, Fast Imaging Employing Steady-state Acquisition, 3D DRIVen Equilibrium, T2 Sampling Perfection with Application-optimized Contrasts using different flip angle Evolutions, and Balanced Fast Field Echo. Close communication between the otologist and the radiologist can help ensure that a correct MRI protocol is performed. Sedation is typically required for MRI. CT can often be performed without sedation, particularly with newer scanners that are able to image the entire temporal bone in several seconds.

Syndromic anomalies

A variety of genetic syndromes may lead to malformations of the external, middle, and inner ear. Hearing loss may be sensorineural, conductive, or mixed. Certain characteristic patterns of involvement have been described with these conditions, and temporal bone imaging can often guide the genetic workup.

Large vestibular aqueduct syndrome

Enlargement of the vestibular aqueduct is the most commonly seen abnormality in pediatric SNHL.[2] As this may be a feature of a variety of other syndromic and nonsyndromic malformations, the term large vestibular aqueduct syndrome (LVAS) is typically used to describe this anomaly when it occurs in isolation or with incomplete cochlear partition ("Mondini" dysplasia) as the only other abnormality.

LVAS was first described in 1978 by Valvassori and Clemis[3] using polytomography, now an obsolete imaging modality, and later by Hill et al[4] using CT. On CT, the characteristic abnormality is enlargement of the vestibular aqueduct (Figure 1). Incomplete cochlear partition (fusion of the middle and apical turns, with missing bony partition [osseous spiral lamina], ie, "Mondini" dysplasia) is a common associated finding, as is mild dysplasia of the lateral semicircular canal (LSC) with associated small LSC bone island.[5] On magnetic resonance images, enlargement of both the endolymphatic duct and the endolymphatic sac are evident (Figure 2).[6] The typical presentation is of SNHL of varying severity, which may be stable, progressive, or fluctuating.[7] Sudden deterioration following trivial head trauma or minor ailments may also be seen.[7]

Pendred syndrome describes the syndrome of deafness and goiter. Patients with Pendred syndrome typically have abnormal results on perchlorate discharge tests and a mutation of the PDS gene and frequently show radiologic findings of enlarged vestibular aqueduct with incomplete cochlear partition. A recent study showed that many patients with enlarged vestibular aqueducts, but without goiter, have PDS gene mutations,[8] suggesting that LVAS and Pendred syndrome lie along a common spectrum.

Different criteria have been described for determining whether a vestibular aqueduct is enlarged. Some use an absolute diameter of more than 1.5 mm, measured on axial images at the midpoint between the isthmus and the external aperture, as the cutoff for a normal vestibular aqueduct, basing this on criteria determined from polytomography.[3] Some have proposed using cutoffs of more than 0.9 mm at the midpoint or more than 1.9 mm at the external aperture (operculum), termed the "Cincinnati criteria."[2,9] Others

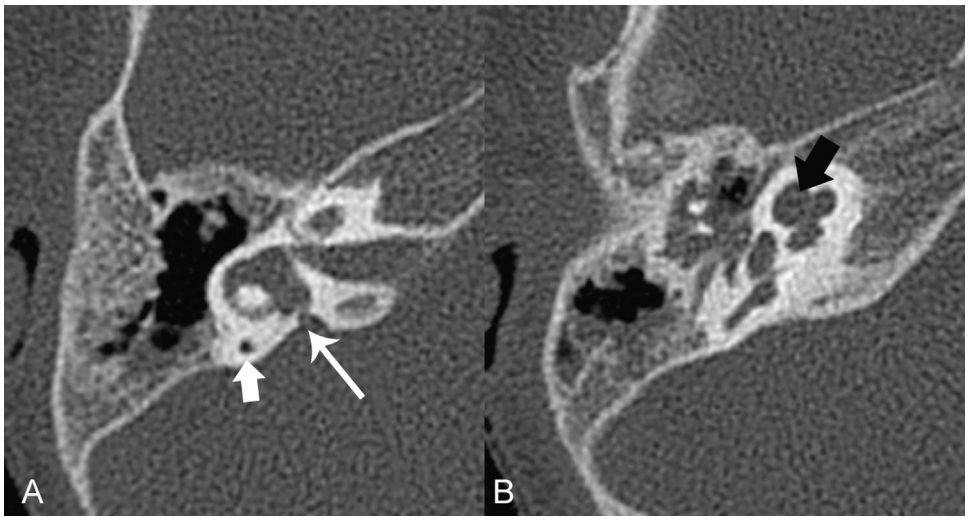

Figure 1 Large vestibular aqueduct syndrome. Axial CT. (A) Image through the level of the lateral semicircular canal shows gross enlargement of the vestibular aqueduct (long arrow). Compare with the posterior semicircular canal (short white arrow) in the same image. (B) Image slightly lower shows cochlear dysplasia with fusion of the middle and apical turns (ie, incomplete partition-II, a.k.a. Mondini dysplasia).