Preoperative Imaging of Sensorineural Hearing Loss in Pediatric Candidates for Cochlear Implantation

Joseph Y. Young, MD  
Maura E. Ryan, MD  
Nancy M. Young, MD

Abbreviations: CHARGE = coloboma of the eye, congenital heart defects, choanal atresia, mental and/or growth retardation, genital hypoplasia, and ear anomalies and/or deafness; CISS = constructive interference in the steady state, CSF = cerebrospinal fluid, FDA = Food and Drug Administration, SNHL = sensorineural hearing loss, 3D = three-dimensional

RadioGraphics 2014; 34:E133–E149

Published online 10.1148/rg.345130083

Content Codes: CT | HN | PD | NR

1From the Department of Radiology (J.Y.Y., M.E.R.) and Department of Otolaryngology–Head and Neck Surgery (N.M.Y.), Northwestern University Feinberg School of Medicine, Chicago, Ill; Department of Medical Imaging (J.Y.Y., M.E.R.) and Division of Pediatric Otolaryngology (N.M.Y.), Ann & Robert H. Lurie Children’s Hospital of Chicago, 225 E Chicago Ave, Chicago, IL 60611; and Knowles Hearing Center, School of Communication, Northwestern University, Evanston, Ill (N.M.Y.). Presented as an education exhibit at the 2012 RSNA Annual Meeting. Received April 2, 2013; revision requested May 22 and received November 20; final version accepted February 18, 2014. All authors have disclosed no relevant relationships.  
Address correspondence to J.Y.Y. (e-mail: joseph-young@fsm.northwestern.edu).

Cochlear implantation is the only U.S. Food and Drug Administration–approved treatment for children with marked bilateral sensorineural hearing loss. It provides auditory benefits that range from simple sound detection to substantial word understanding. Improved hearing through cochlear implantation has been demonstrated to enhance the rate of language acquisition, enable development of spoken language, and advance literacy in deaf children. Magnetic resonance imaging and computed tomography both have roles in the preoperative assessment of inner-ear abnormalities, cochlear nerve deficiency, and variant anatomy that may affect the decision to implant and the prognosis for auditory improvement and increase the risk for complications. Most cochlear abnormalities may be successfully treated with cochlear implantation, but the presence of a cochlear malformation may increase the risk for intraoperative cerebrospinal fluid leakage and postoperative bacterial meningitis. Eighth-nerve deficiency correlates with poor auditory outcomes and may affect eligibility for cochlear implantation. Another important consideration for implantation is the presence of labyrinthitis ossificans in some children with deafness resulting from bacterial meningitis, which may cause obstruction that limits electrode insertion. Anatomic variations of the facial nerve or middle-ear cavity, which are more common in syndromic patients, may also affect the surgical approach and make implantation difficult.

Introduction

Sensorineural hearing loss (SNHL) may result from congenital or acquired abnormalities of the membranous labyrinth or cochlear nerve. At least one-half of childhood SNHL cases are estimated to occur on a genetic basis, but the true incidence of genetic SNHL is uncertain because of barriers to testing and the likelihood of additional causative mutations yet to be identified (1). Cytomegalovirus infection is believed to be the most common nongenetic cause of pediatric SNHL. However, because infectious, environmental, and genetic sources may be difficult to distinguish, the true cause of childhood SNHL often remains unknown (2).

The first cochlear implantation system was approved for use in children by the U.S. Food and Drug Administration (FDA) in 1990. Currently, cochlear implantation is the only approved treatment option in the United States for children with substantial bilateral SNHL who do not receive acceptable benefit from hearing aids (3). Cochlear implantation improves auditory perception in most recipients and has enabled the development of spoken language and improved literacy in many deaf children (4). Most candidates for cochlear implantation
have normal findings at imaging of the temporal bone; however, anatomic abnormalities have been identified in a large and increasing number of these patients, and the prevalence of detectable cochleovestibular anomalies may be as high as 40%, according to some reports (5,6). In addition, as recognition of the benefits of cochlear implantation in improving overall communication ability and cognition increases, a greater number of medically complex cases of SNHL are being evaluated at pediatric implantation centers. These complex cases include children with syndromic abnormalities—such as CHARGE, which stands for coloboma of the eye, congenital heart defects, choanal atresia, mental and/or growth retardation, genitai hypoplasia, and ear anomalies and/or deafness, and branchio-oto-renal syndrome—which are known to be associated with inner- and middle-ear anomalies (7).

Preoperative knowledge of the anatomic features of temporal bone may be critical for making the decision to perform cochlear implantation, counseling parents regarding risks and long-term outcomes, modifying the surgical approach, and device programming by audiologists. From the pediatric surgeon’s perspective, the goal of preoperative imaging is primarily to identify cochlear abnormalities, eighth-nerve deficiency, or anatomic variations that influence candidacy, ear selection, surgical approach, and prognosis (8). Different classifications have been proposed for abnormalities encountered in SNHL, and many radiologists are familiar with the imaging findings. However, these descriptions are often based on embryologic studies rather than surgical significance and are not all-inclusive (9,10). Some malformations defy categorization, and classifications typically do not address surgically important aspects, such as cochlear nerve deficiency and middle-ear deformity. For imaging interpretation to be most useful to the referring surgeon, the radiologist should also be aware of which findings could affect surgery planning and management.

In this article, we review the relevant imaging methods and normal anatomic findings, briefly describe the cochlear implantation device and implantation procedure, and review the major imaging findings associated with congenital and acquired childhood SNHL. We highlight the surgical landmarks and anatomic variations that may affect the approach and technique for cochlear implantation and emphasize the surgical implications of the classic imaging features.

**Imaging Techniques**

Computed tomography (CT) has traditionally been the preferred modality for initial imaging workup of pediatric candidates for cochlear implantaon (11). CT of the petrous temporal bones enables detailed evaluation of the osseous anatomy of the inner and middle ear, as well as assessment of mastoid pneumatization and the degree of middle-ear aeration (12). Dedicated high-resolution techniques are required to achieve adequate detail of this small region, and imaging is generally performed by using fine collimation, millimeter section thickness, and dedicated bone algorithms. Many modern multichannel scanners may provide adequate coronal reformations, although some institutions still use direct coronal imaging. The technique at our institution includes axial helical acquisitions with a collimation of 0.6 mm, section thickness of 0.625 mm, and magnified axial and coronal 3-mm reconstructions with the use of an enhanced edge bone algorithm (SOMATOM Definition Flash; Siemens, Malver, Pa). However, there are some disadvantages to the use of CT. It exposes patients to ionizing radiation, which is of particular concern in the vulnerable pediatric population. Low-dose temporal bone CT may be performed, but, in this region, dose reduction may be problematic because of the dense petrous bone and high-resolution requirements (13). In addition, CT does not directly depict the cochlear nerve, only the bone channel that contains it.

Magnetic resonance (MR) imaging of the internal auditory canal and inner ear enables direct visualization of the cisternal and intracanalicular vestibulocochlear nerve bundles and carries no radiation risk. Dedicated MR imaging of this area involves the use of high-resolution heavily T2-weighted three-dimensional (3D) imaging techniques, such as constructive interference in the steady state (CISS; Siemens) and fast imaging employing steady-state acquisition (GE Healthcare, Fairfield, Conn) sequences. In general, MR images are obtained in the axial plane, although obtaining images perpendicular to the course of the nerves through the internal auditory canal may also help distinguish the cochlear division of the eighth nerve. Oblique sagittal images may be reformatted from the axial 3D sequence, although direct acquisitions may provide better conspicuity of the small cochlear structures, particularly when higher-strength (3-T) magnets are not used. At our institution, we obtain 3D CISS images in the axial and oblique sagittal planes with a 512 × 512 matrix, 0.8–1-mm section thickness, and 0.4–0.5-mm skip (3 T Skyra; Siemens). With the use of high-resolution T2-weighted 3D techniques, MR imaging also provides relatively detailed information about the fluid-filled labyrinth and may depict substantial inner-ear abnormalities, although its resolution is inferior to that of CT.

A major disadvantage of the use of MR imaging before cochlear implantation is its poor depiction
Figure 1. Normal cochlea and vestibule in two patients. (a, b) Axial CT images of the left inner ear show the basal (arrow in a), middle (arrowhead in b), and apical (arrow in b) turns of the cochlea. Note the slight indentation between the middle and apical turns, which should be present even if the thin bony division is not visible because of section selection. (c) Axial CT image of the right inner ear obtained slightly superior to a and b shows the bony modiolus (white arrow), which forms the base of the cochlea at the fundus of the internal auditory canal. The cochlear aperture (white arrowhead) is the adjacent bony channel through which the cochlear division of the eighth nerve enters the cochlea. The vestibule (black arrow) and lateral semicircular canal (black arrowhead) are also seen. (d) Axial CT image of the right inner ear obtained in a different patient shows the vestibular aqueduct (arrow) extending to the posterior margin of the petrous temporal bone. A portion of the posterior semicircular canal (arrowhead) is also seen. A cochlear implant is present.

Of aerated or mineralized structures; it may not reliably depict bone landmarks and the degree of mastoid or middle-ear pneumatization. In addition, sedation is often required for MR imaging in young patients because of the duration of the examination. CT may be performed much more rapidly; however, very young patients may need sedation for CT, as well, because evaluation is highly motion sensitive and reimaging of areas degraded by artifacts results in increased radiation exposure.

Preoperative imaging workup for pediatric cochlear implantation varies among institutions, but the utility of MR imaging in patients with SNHL is becoming increasingly recognized (14). Some surgeons use MR imaging as the primary, and often only, modality to evaluate candidates for cochlear implantation. Other centers routinely perform both MR imaging and CT, and some still primarily rely on CT. However, it is important to recognize that CT does not reliably depict the presence of a normal eighth nerve (15). Unrecognized eighth-nerve deficiency in children with a normal internal auditory canal and cochlear aperture at CT may explain some of the variability in performance of cochlear implants among children.

Normal Anatomic Findings of SNHL and Cochlear Implantation

Inner Ear
The inner ear consists of the fluid-filled membranous labyrinth, which is enclosed by the bone of the osseous labyrinth. The cochlea is the primary organ of hearing and the destination of cochlear implantation (Fig 1). This spiral structure has 2.5–2.75 turns around a central column of bone.
(the modiolus). A spiral osseous lamina projects from the modiolus and divides the cochlear canal into upper (scala vestibuli) and lower (scala tympani) compartments; the scala tympani is the implantation destination. The cochlear aperture is the opening at the fundus of the internal auditory canal through which the eighth nerve enters the cochlea.

The vestibular system, which is located posterolateral to the cochlea, is formed by the vestibule and three semicircular canals: superior, posterior, and lateral (Fig 1a). These structures play a minor role in hearing, but abnormalities often occur in conjunction with cochlear malformations.

The endolymphatic duct arises from the posterior vestibule and extends through the bone labyrinth to the endolymphatic sac, which terminates in the epidural space of the posterior fossa (Fig 1c). The fluid-filled duct is usually visible at MR imaging but may occasionally be below the limits of resolution if it is not dilated. The vestibular aqueduct is the osseous channel that encloses the endolymphatic duct or sac and is visible at CT.

### Mastoid and Middle Ear

Typically, mastoid and middle-ear pathologic abnormalities affect conductive—not sensorineural—hearing (Fig 2). However, the anatomic features of this region are important for surgical planning for cochlear implantation, which requires partial mastoidectomy. The degree of mastoid pneumatization or development may affect the approach. The middle-ear cavity also contains important bone landmarks, such as the facial nerve recess and round window niche, which the surgeon uses to gain access to the cochlea for placement of the cochlear implant electrode array.

### Nerves

The eighth nerve extends through the cerebellopontine cistern into the internal auditory canal, where the nerve divides into the cochlear, superior vestibular, and inferior vestibular components (Fig 3). The cochlear nerve, which is located in the anteroinferior quadrant of the internal auditory canal, is the primary conduit of auditory impulses to the brainstem. The bone internal auditory canal channel is well demonstrated at CT, but the cisternal and intracanalicular nerves are visible only at MR imaging.

The seventh (facial) nerve is an important surgical landmark during any major ear surgery (Fig 4). Access to the cochlea from the mastoid is central to placing the cochlear implant. Implantation is usually accomplished by opening the facial recess, which is bordered inferiorly by the facial nerve. Access to the inner ear for electrode placement requires operating near the facial nerve, and the risk for potential injury is increased if the course of the nerve is anomalous. The normal seventh nerve traverses the cerebellopontine cistern (the cisternal segment) into the anterosuperior quadrant of the internal auditory canal (the intracanalicular segment). The labyrinthine segment then courses anteriorly to the geniculate ganglion. The horizontal, or tympanic, segment extends posteriorly from the ganglion, along the medial aspect of the middle-ear cavity, beneath the lateral semicircular canal, and into the facial nerve recess. The

[Figure 2. Landmarks of a normal left middle ear. Axial CT image shows the facial nerve recess (black arrowhead), sinus tympani (curved black arrow), and a portion of the mastoid segment of the facial nerve (white arrowhead). The round window niche (white arrow) is also seen adjacent to the basal turn of the cochlea (straight black arrow). Note the detail of the aerated mastoid cells and bone septations (*).]

[Figure 4. Normal left facial nerve canal. (a–c) Axial CT images show the bony internal auditory canal (curved arrow in a), which contains the intracanalicular segment, the bony canal of the labyrinthine segment (straight arrow in a) extending to the geniculate ganglion (arrowhead in a), the tympanic segment (arrowhead in b) traversing the medial margin of the middle-ear cavity, and a portion of the descending mastoid segment (arrowhead in c) adjacent to the facial nerve recess. The normal petrous carotid artery (arrow in c) courses anteromedial to the labyrinth and should be covered by bone. (d) Coronal CT image shows the tympanic facial nerve segment (arrowhead), with a thin bone covering coursing beneath the lateral semicircular canal along the superior margin of the oval window. The jugular bulb is separated from the middle-ear cavity by a normal bone covering (arrow).]
Figure 3. Normal left eighth nerve. (a) Axial 3D T2-weighted CISS MR image obtained at the level of the membranous labyrinth shows the fluid-filled internal auditory canal, which contains the cochlear division of the eighth nerve (straight arrow). The fluid-filled cochlea (curved arrow), vestibule (arrowhead), and lateral semicircular canal (squiggly arrow) are well depicted, with hyperintensity equal to that of cerebrospinal fluid (CSF). The linear low-signal-intensity osseous spiral lamina within the cochlea is also visible. A few opacified mastoid air cells are apparent, but there is no detail of the aerated mastoid or middle ear. (b) Oblique sagittal T2-weighted 3D fast imaging employing steady-state acquisition MR image (GE Healthcare) shows the four normal nerve bundles in the internal auditory canal, with the cochlear division of the eighth nerve in the anterior-inferior quadrant (squiggly arrow), the facial nerve in the anterior-superior quadrant (straight arrow), and the superior (curved arrow) and inferior (arrowhead) vestibular eighth nerve divisions posteriorly.
distal vertical, or mastoid, segment then courses inferiorly through the mastoid to exit from the stylomastoid foramen. The normal cisternal eighth nerve should be approximately twice the diameter of the facial nerve, and the cochlear division should be as large as or slightly larger than the intracanalicular facial nerve.

The cisternal and intracanalicular facial segments are identifiable only at MR imaging. The tympanic and mastoid portions are typically
Figure 6. Normal course of a left cochlear implant device. (a) Axial CT image obtained superior to the external auditory canal shows the receiver-stimulator (white arrow) in the postauricular pocket. A portion of a ground wire (black arrow), which may be placed beneath the temporalis muscle, is also visible. (b) Axial CT image obtained inferior to a shows an intact canal wall mastoidectomy, with a portion of the implant coiled in the mastoid bowl (white arrow). The electrode array extends through a cochleostomy anterior-inferior to the round window niche (arrowhead) and into the basal cochlear turn (black arrow).

not discernible at MR imaging because of surrounding bone and air. The normal facial nerve canal that encases the distal nerve is identifiable at CT, although an anomalous or dehiscent facial nerve canal may be difficult to discern, particularly if it is hypoplastic or surrounded by middle-ear opacification.

Vessels
The jugular vein and internal carotid artery traverse the skull base adjacent to the middle and inner ear (Fig 4a, 4d). These vessels are generally not a concern during implantation unless a variant course or dehiscence of the normal bone coverings is present.

Device and Technique for Cochlear Implantation
The first cochlear implantation system was a single-electrode device developed by William F. House in the 1960s (16). Although outcomes were more variable and limited in comparison with those for multichannel devices, single-channel devices were able to detect sounds and, in some cases, allow word recognition. Today, multichannel devices enable better auditory outcomes, including higher degrees of word perception.

Several different cochlear implantation systems are available, but the basic elements consist of a surgically implanted device and externally worn components (Fig 5). The body of the implanted device is often called the receiver-stimulator. It contains an antenna wire to receive electrical signals and the necessary components to transmit electrical impulses to the multiple electrodes placed within the cochlea. The receiver-stimulator also houses a magnet, which is needed to retain and align the externally worn transmitter. The external removable components include a microphone and battery-powered speech processor.

Implantation systems provide surgeons with a selection of electrode arrays that may be placed inside the inner ear. Cochlear anatomy, as well as the degree of residual hearing, may influence a surgeon’s choice of electrode. The systems also typically provide a choice of external speech processors that may be worn behind the ear or clipped to clothing on the body, which may be preferable in infants and children who lack normal pinnae.

Incisions and techniques for securing the receiver-stimulator device vary. At our institution, a postauricular incision is typically used with creation of a periosteal pocket to secure the body of the device. The standard surgical technique for electrode placement is mastoidectomy, with opening of the facial recess to gain access to the middle-ear cavity in the vicinity of the round window niche. The facial recess is defined by the short process of the incus superiorly, the chorda tympani nerve laterally, and the facial nerve medially. This approach affords
visualization of the round window niche. Traditionally, the cochlea was accessed by creation of a cochleostomy that was anterior-inferior to the round window membrane, with the electrode array then inserted into the scala tympani of the basal turn through the cochleostomy (Fig 6). However, round window insertion is becoming more common in patients with substantial residual hearing, and the occurrence of fewer traumas to the cochlea is thought to be advantageous in preserving existing function (17). Implantation systems also contain an extracochlear ground electrode that may be incorporated into the receiver-stimulator or placed on an additional lead wire that the surgeon positions under the temporalis muscle above the mastoid cavity. Implant arrays vary in design in terms of the number of electrode contacts, their position and spacing along the array, and whether the array is straight or curved. Therefore, the depth of the electrode insertion needed to place all electrodes inside the cochlea varies, and knowledge of the anatomy may influence the surgeon’s choice of array.

### Cochleovestibular Abnormalities

#### Complete Labyrinthine and Cochlear Aplasia

Complete labyrinthine aplasia, which is classically known as Michel aplasia, is a severe congenital anomaly that is defined by total absence of the inner-ear structures (Fig 7). Absence of the cochlea with a present, but often deformed, vestibule and semicircular canals is referred to as cochlear aplasia. Typically, the cochlear nerve is aplastic in complete labyrinthine and cochlear aplasia, and there is often a diminutive internal auditory canal with an anomalous course of the facial nerve (18). Children with complete labyrinthine and cochlear aplasia also classically have hypoplasia of the petrous apex and, sometimes, the skull base (19).

In patients with cochlear aplasia, severe dysplasia of the vestibular system may mimic a deformed cochlea, but true aplasia may be identified by the presence of dense sclerotic bone beneath the cochlear promontory, at the expected location of the cochlea (Fig 8). Complete labyrinthine and cochlear aplasia are rare, constituting less than 1% and 3%, respectively, of cochlear bone abnormalities (20). It is important to recognize these deformities because children who lack a cochlea in both ears may not undergo cochlear implantation. If the abnormality is unilateral, the nonaplastic ear may be amenable to implantation.

#### Common Cavity

The common cavity malformation is defined as the confluence of the cochlea and vestibule into a single cystic cavity, with no internal architecture and an absent modiolus (Fig 9). In contrast to a dysplastic vestibule in cochlear aplasia, a common cavity malformation should demonstrate lucency beneath, and occasionally bulging of, the cochlear promontory. The modiolus is absent, and the opening of the internal auditory canal into the common cavity malformation is abnormal, which may account for the increased presence of CSF in these malformations (in contrast to perilymph fluid in a normal labyrinth), as well as the increased incidence of spontaneous CSF leakage into the middle
Figure 8. Cochlear aplasia of the right inner ear. (a) Coronal CT image shows no cochlear structures, with only sclerotic bone deep to the promontory (arrow), and a portion of a small, dysplastic bony internal auditory canal (arrowhead). (b) Axial CT image obtained inferior to a shows a lobulated lucency in the bony labyrinth that represents a dysplastic vestibule (white arrow) and a portion of a malformed posterior semicircular canal (black arrow).

Figure 9. Common cavity malformation of the right inner ear. Axial (a) and coronal (b) CT images show a single dilated lucent cavity in the petrous temporal bone (white arrow in a), with no distinguishable separation between the cochlear turns and vestibule. The cavity extends beneath the bony margin of a bulging cochlear promontory (arrowhead), and there is a wide connection between the internal auditory canal and the common cavity (black arrow in a), a finding indicative of CSF within the cavity.

A cochlear implant can be successfully placed into a common cavity malformation, but the procedure may be technically challenging (22). If CSF is already present within the cochlea, a brisk leak of CSF may be encountered after cochleostomy is performed. The electroarray can still be placed, but the leak must be effectively sealed to minimize the risk for meningitis. Even if there is initially no CSF within the common cavity malformation, the electrode array may be inadvertently positioned into the internal auditory canal, resulting in a CSF leak (Fig 10).

CSF leakage may be avoided by controlling the tip of the array at the time of insertion; thus, identifying this type of malformation may influence the choice of electrode array and cochleostomy technique. Often, it is easier to create the cochleostomy through the mastoid in the area where the lateral canal would normally be located and thereby avoid the need to open the facial recess (23). Because the anatomic path of the cochlear nerve within the common cavity malformation is uncertain, one of the authors (N.M.Y.) prefers a straight array with circumferential banded electrodes to maximize the likelihood of neural stimulation.
Fewer than 1.5 cochlear turns and an abnormal appearance of the basal turn (Fig 11a, 11b). The entire modiolus is often absent, and the cribiform area between the internal auditory canal and cochlea is abnormal. The vestibule and semicircular canal are often dysplastic as well. The cochlea may have a cystic appearance, but, fewer than 1.5 cochlear turns and an abnormal appearance of the basal turn (Fig 11a, 11b). The entire modiolus is often absent, and the cribiform area between the internal auditory canal and cochlea is abnormal. The vestibule and semicircular canal are often dysplastic as well. The cochlea may have a cystic appearance, but,

**Figure 10.** Misplaced left cochlear implant. Axial CT image shows an implant entering a cystic common cavity (arrow), but instead of coiling within the cochlea, the stimulator wire exits into the internal auditory canal (arrowhead).

**Figure 11.** Cochlear dysplasia of the right inner ear in two patients. (a, b) Axial CT images obtained in a patient with severe dysplasia show a widened basal turn (arrow in a) and cystic fusion of the middle and apical turns (arrowhead in b). A partially visualized dysplastic vestibule and semicircular canal (arrow in b) are also seen. No vestibular aqueduct enlargement is present. (c) Axial CT image obtained in a patient with mild cochlear dysplasia shows a well-formed basal turn (arrowhead) and a dysplastic middle turn, with no identifiable apical turn (arrow). No vestibular aqueduct enlargement is present.

**Other Congenital Cochlear Abnormalities**

In many other cochlear dysplasias, the cochlea is present but does not form the normal basal, middle, and apical turns. The surgical importance of these abnormalities is primarily determined by the degree of cochlear malformation and the potential for communication with CSF. Malformations with a dysplastic or absent modiolus are more likely to have CSF present in the cochlea, although an intact modiolus does not exclude the possibility of a fistula (24).

Severe dysplasias, which are sometimes called incomplete partition type I deformities, have
unlike the common cavity malformation, the malformed cochlea is not fused to the vestibular system. The vestibular aqueduct is typically not enlarged. Patients with severe dysplasia have a high likelihood of communication between fluid in the cochlea and CSF in the internal auditory canal and an associated increased risk for leakage with implantation (22).

Less severe cochlear dysplasias, including the classic Mondini deformity, are characterized by coalescence of the middle and apical turns with a well-formed basal turn (Fig 11c). The modiolus is usually partially formed. Often, there is associated enlargement of the vestibular aqueduct, known as an incomplete partition type II deformity. Patients with less severe cochlear deformities have a mildly increased incidence of CSF leakage but fewer complications than do patients with severe dysplasia or incomplete partition type I deformities (25).

Mild dysplasia that is restricted to the apical turn without enlargement of the vestibular aqueduct may be subtle and difficult to identify at imaging. Although this type of mild dysplasia may manifest with hearing loss as profound as that seen with other cochlear malformations, isolated abnormalities of the apical turn have little effect on candidacy for or management of cochlear implantation.

Cochlear hypoplasia is characterized by a present but diminutive cochlea, which is often described as a small “bud” extending from the internal auditory canal (Fig 12). The vestibule and semicircular canal are typically dysplastic as well. Hypoplasia is not an absolute contraindication for implantation, although the auditory benefit varies depending on the severity of the abnormality and the number of electrodes that may be accommodated, as well as the degree of associated cochlear nerve deficiency, which often accompanies cochlear hypoplasia.

**Enlarged Vestibular Aqueduct**

An enlarged vestibular aqueduct, either by itself or in association with mild cystic cochlear dysplasia, has been reported to be the most common inner-ear malformation associated with SNHL (Fig 13) (21,26). This abnormality is bilateral in as many as 90% of patients according to some reports (27). The normal size of the vestibular aqueduct has been a point of some debate, but a diameter of 1.5 mm or less at the midportion is the most commonly accepted measurement (28). The width should also be no greater than the diameter of the adjacent normal posterior semicircular canal, although caution must be used with this internal standard because associated semicircular canal dysplasia may be present in these patients.

Clinically, children with an enlarged vestibular aqueduct have hearing that ranges from normal to profoundly lost. The natural history of an enlarged vestibular aqueduct is that of progressive but unpredictable hearing loss. Although the cause of progressive hearing loss is unknown, one theory is that trauma may affect it by causing hemorrhage into the endolymphatic sac (29). To minimize the risk for a sudden decline in hearing, patients are generally counseled to wear a helmet when participating in certain sports and to consider refraining from activities in which high-impact and unprotected head
Cochlear Nerve Abnormalities

At one time, evidence of an absent or severely dysplastic cochlear nerve was considered an absolute contraindication to cochlear implantation in any patient. More recently, implantation in children with an absent or deficient eighth nerve was performed with varying results. Outcomes are more difficult to predict and poorer than expected compared with those in patients with normal nerve anatomy, but some young children have achieved substantial word recognition and spoken language skills (33).

A potential alternative treatment for children with cochlear nerve deficiency is auditory brainstem implantation. This implant was initially developed for individuals with neurofibromatosis type II who could not benefit from cochlear implantation because trauma are likely to occur. Once individuals with an enlarged vestibular aqueduct lose hearing to the degree that hearing aids are no longer sufficient, cochlear implantation is the only effective treatment option.

There is an increased risk for CSF leakage in patients with an enlarged vestibular aqueduct, but it is likely minor in the absence of concomitant cochlear malformations (30). An enlarged vestibular aqueduct, either as an isolated abnormality or in conjunction with poor partition of the apical and middle turns, does not adversely affect the prognosis for auditory response to cochlear implantation.

Vestibule and Semicircular Canal Abnormalities

Although the risk for CSF leakage increases when inner-ear malformations are present, isolated anomalies of the vestibule or semicircular canal typically do not affect surgical planning. Their importance lies predominantly in the associated cochlear or syndromic abnormalities. Absence of the semicircular canals should be noted because it is strongly associated with CHARGE syndrome (31). In addition to cochlear dysplasias, most patients with CHARGE syndrome have a dysplastic middle-ear cavity and an anomalous facial nerve course, which may complicate the surgical approach and cochleostomy (32). Cochlear nerve hypoplasia is also common in this setting, and it is the practice at our center to use CT to assess bone anatomy and MR imaging to assess nerve integrity in many such patients.

Cochlear Nerve Abnormalities

At one time, evidence of an absent or severely dysplastic cochlear nerve was considered an absolute contraindication to cochlear implantation in any patient. More recently, implantation in children with an absent or deficient eighth nerve was performed with varying results. Outcomes are more difficult to predict and poorer than expected compared with those in patients with normal nerve anatomy, but some young children have achieved substantial word recognition and spoken language skills (33). A potential alternative treatment for children with cochlear nerve deficiency is auditory brainstem implantation. This implant was initially developed for individuals with neurofibromatosis type II who could not benefit from cochlear implantation because trauma are likely to occur. Once individuals with an enlarged vestibular aqueduct lose hearing to the degree that hearing aids are no longer sufficient, cochlear implantation is the only effective treatment option.

There is an increased risk for CSF leakage in patients with an enlarged vestibular aqueduct, but it is likely minor in the absence of concomitant cochlear malformations (30). An enlarged vestibular aqueduct, either as an isolated abnormality or in conjunction with poor partition of the apical and middle turns, does not adversely affect the prognosis for auditory response to cochlear implantation.

Vestibule and Semicircular Canal Abnormalities

Although the risk for CSF leakage increases when inner-ear malformations are present, isolated anomalies of the vestibule or semicircular canal typically do not affect surgical planning. Their importance lies predominantly in the associated cochlear or syndromic abnormalities. Absence of the semicircular canals should be noted because it is strongly associated with CHARGE syndrome (31). In addition to cochlear dysplasias, most patients with CHARGE syndrome have a dysplastic middle-ear cavity and an anomalous facial nerve course, which may complicate the surgical approach and cochleostomy (32). Cochlear nerve hypoplasia is also common in this setting, and it is the practice at our center to use CT to assess bone anatomy and MR imaging to assess nerve integrity in many such patients.
surgical tumor management typically includes excision of the cochlear nerve. Auditory brainstem implantation is currently approved by the FDA only for use in individuals who are deafened by neurofibromatosis type II and are aged 12 years and older. Outcomes data from Coletti (34) of Verona, Italy, who has the most experience with auditory brainstem implantation in young children who are congenitally deaf, are promising. However, because neurosurgical placement of the device is a more invasive procedure that provides incomplete hearing restoration compared with cochlear implantation, in the United States, FDA trials are restricted to children with anomalies such as cochlear nerve deficiency, labyrinthine aplasia, and severe ossification who have not benefitted from lower-risk cochlear implantation.

At CT, cochlear aperture stenosis indirectly indicates hypoplasia or absence of the cochlear nerve (Fig 14a). Although the criteria for a normal aperture size are not well defined in the literature, an aperture with a diameter smaller than 2 mm is usually considered abnormal. The caliber of the bony internal auditory canal may also be diminished. Correlation between the aperture or internal auditory canal stenosis and nerve hypoplasia is not absolute, and cochlear nerve deficiency may occur in the absence of narrowing (15). CT remains relatively insensitive for depicting nerve hypoplasia, and the nerve itself may be directly visualized only at high-resolution steady-state T2-weighted MR imaging (Fig 14b). For this reason, at our children’s hospital, MR imaging of the internal auditory canal is a standard part of the evaluation for cochlear implantation candidacy in patients with suspected hypoplasia.

Cochlear Patency

One of the most important causes of postnatal acquired SNHL is bacterial meningitis, which may have profound effects on cochlear patency that limit or preclude cochlear implantation. The incidence of bacterial meningitis has substantially declined in the United States since the implementation of widespread pneumococcal vaccination, but it remains a challenging problem when it does occur. Deafness secondary to bacterial meningitis is often associated with labyrinthitis ossificans, a progressive process that may obstruct the cochlea.

Labyrinthitis ossificans is commonly bilateral but may be asymmetrical. Changes typically begin at the basal cochlear turn and proceed apically (35). The onset of labyrinthitis ossificans is variable and unpredictable, and it may occur weeks to years after the inciting insult (36). Labyrinthitis ossificans is primarily a postinflammatory reaction, and ossification represents the end stage of this process (Fig 15a). A completely mineralized labyrinth may mimic congenital absence of the cochlea and vestibule; however, in contrast to patients with hypoplasia, those with labyrinthitis ossificans should have a normally sized internal auditory canal, petrous apex, and cochlear aperture.

Imaging assessment of labyrinthitis ossificans is essential for surgical planning because these patients benefit from urgent referral for evaluation.
and cochlear implantation before obstruction of the cochlea occurs (37). Early implantation before the onset of substantial obstruction is necessary to achieve optimal electrode insertion and avoid the need for complex alternative surgical approaches, which have more variable outcomes. Early changes of labyrinthitis ossificans are characterized by fibrosis, which may be seen at MR imaging as a loss of the normal hyperintensity of fluid on T2-weighted images of the membranous labyrinth (Fig 15b). Labyrinthine enhancement has also been reported to accompany or precede fibrotic changes at contrast material–enhanced MR imaging (38). Fibrosis is not distinguishable from normal labyrinthine fluid at CT, and high-resolution steady-state T2-weighted MR imaging is considered the first-line imaging modality for patients who are suspected of having labyrinthitis ossificans. If MR imaging findings are normal, there is generally no need for CT. However, both fibrosis and ossification result in loss of signal intensity at T2-weighted MR imaging. In these patients, CT may help differentiate between fibrotic and mineralized obstruction, although its sensitivity for depicting mild but potentially clinically significant mineralization is uncertain (Fig 15b, 15c) (39). In addition, dense fibrosis without substantial calcification may present a potential challenge for implant insertion.

Other Anatomic Considerations

Facial Nerve Anomalies

Other malformations or normal variants that do not directly contribute to SNHL may have a large effect on surgical risk and approach and warrant special attention in candidates for cochlear implantation. Among pediatric patients with vestibulocochlear anomalies, approximately 15% have an aberrant facial nerve course, which may increase the risk for facial nerve injury (Fig 16a) (40). Particular attention should be directed to identifying the tympanic segment, which may cross the surgical path during opening of the facial nerve recess and cochleostomy. Anomalous facial nerves may be hypoplastic or have a dehiscent facial nerve canal, and the course may be difficult to detect at imaging, especially in the presence of surrounding middle-ear opacification.
Vascular Anomalies
A high-riding or dehiscent jugular bulb that interferes with surgical access to the cochlea is rare but may affect cochleostomy placement (Fig 16b). Occasionally, large occipital transosseous veins may affect the placement of the postauricular incision (Fig 16c). Hemorrhage from low-pressure venous injuries may complicate the procedure but is usually not life-threatening. Typically, an aberrant carotid artery that courses within an unopacified middle-ear cavity is visible to the surgeon, but it may be devastating if it is unanticipated.

Middle-Ear and Mastoid Variants
Absence or dysplasia of the round window niche, which typically serves as a bone landmark for cochleostomy, necessitates the use of other structures to guide entry into the basal cochlear turn (Fig 16d). Assessing the configuration and pneumatization of mastoid air cells and the middle-ear cavity may also aid surgical planning (Fig 16e). Sclerotic or hypopneumatized mastoid air cells may limit exposure of the middle ear. Middle-ear opacification also limits visualization during surgery, and acute otomastoiditis requires treatment before implantation to decrease the risk for infection and meningitis.
Conclusions

Cochlear implantation is the first medical treatment to enable deaf children to learn to talk and develop age-appropriate language and literacy. Imaging is an important part of the preoperative workup in these children. MR imaging may depict relevant abnormalities of the membranous labyrinth and eighth cranial nerve and be used to assess cochlear patency in children at risk for labyrinthitis ossificans. CT is particularly helpful for evaluating the bone mastoid and middle-ear landmarks used to guide the surgical approach, although it exposes patients to ionizing radiation. A greater understanding of the process for evaluating patients who are eligible for cochlear implantation, the surgical procedure, and the implant device itself enables the radiologist to provide more useful information to referring clinicians and ultimately improves the quality of care these children receive.

Acknowledgment.— The authors thank Emma Boylan for assistance in editing the figures.

References

The standard surgical technique for electrode placement is mastoidectomy, with opening of the facial recess to gain access to the middle-ear cavity in the vicinity of the round window niche. The facial recess is defined by the short process of the incus superiorly, the chorda tympani nerve laterally, and the facial nerve medially. This approach affords visualization of the round window niche. Traditionally, the cochlea was accessed by creation of a cochleostomy that was anterior-inferior to the round window membrane, with the electrode array then inserted into the scala tympani of the basal turn through the cochleostomy (Fig 6). However, round window insertion is becoming more common in patients with substantial residual hearing, and the occurrence of fewer traumas to the cochlea is thought to be advantageous in preserving existing function (17).

Complete labyrinthine and cochlear aplasia are rare, constituting less than 1% and 3%, respectively, of cochlear bone abnormalities (20). It is important to recognize these deformities because children who lack a cochlea in both ears may not undergo cochlear implantation. If the abnormality is unilateral, the nonaplastic ear may be amenable to implantation.

In many other cochlear dysplasias, the cochlea is present but does not form the normal basal, middle, and apical turns. The surgical importance of these abnormalities is primarily determined by the degree of cochlear malformation and the potential for communication with CSF.

More recently, implantation in children with an absent or deficient eighth nerve was performed with varying results. Outcomes are more difficult to predict and poorer than expected compared with those in patients with normal nerve anatomy, but some young children have achieved substantial word recognition and spoken language skills (33).

Other malformations or normal variants that do not directly contribute to SNHL may have a large effect on surgical risk and approach and warrant special attention in candidates for cochlear implantation.