Evaluation of Pediatric Sensorineural Hearing Loss With Magnetic Resonance Imaging

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**Objective:** To evaluate the incidence and type of intracranial and inner ear abnormalities in children with sensorineural hearing loss (SNHL) identified with magnetic resonance imaging (MRI) and stratified by the degree and type of SNHL.

**Design:** Retrospective review of medical records and MRIs.

**Setting:** Tertiary care children's hospital.

**Patients:** A total of 227 children aged 1 month to 17 years (mean age, 5.3 years; male to female ratio, 1:1) with a diagnosis of SNHL underwent MRI from June 1, 1996, to June 1, 2002. Of these children, 170 had clinical information available and technically adequate MRIs and were included in the study.

**Intervention:** Magnetic resonance imaging.

**Main Outcome Measure:** Identification of an abnormality of the intracranial contents, inner ear, and cochlear nerve.

**Results:** Of the 170 children, 101 (59%) had bilateral SNHL and 69 (41%) had unilateral SNHL, comprising 271 ears with SNHL. Abnormalities of the inner ear were found in 108 ears (40%) with 87 (32%) having abnormalities of the cochlea, which were considered mild in 63 (23%) and moderate to severe in 24 (9%). Forty-nine of 271 ears (18%) with SNHL demonstrated an either absent (26/49 [53%]) or deficient (23/49 [47%]) cochlear nerve. Ears with severe and profound SNHL had more abnormalities than ears with mild and moderate SNHL (66/138 [48%] vs 23/80 [29%]; \(P = .006\)), and children having ears with unilateral moderate, severe, or profound SNHL had more inner ear abnormalities than children with bilateral moderate, severe, or profound SNHL (28/45 [62%] vs 54/144 [38%]; \(P = .004\)).

**Conclusions:** The overall incidence of inner ear abnormalities in ears of children with SNHL evaluated by MRI is 40%. The most common abnormalities seen were an abnormal cochlea and abnormal cochlear nerve. Children with severe and profound SNHL have a greater percentage of inner ear anomalies than children with mild or moderate SNHL. Children with unilateral hearing loss have a greater percentage of inner ear anomalies than children with bilateral SNHL.

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SENSORINEURAL HEARING LOSS (SNHL) in the pediatric population affects thousands of children annually. The incidence of bilateral SNHL ranges from 1.4 to 3 per 1000 live births in various studies worldwide.\(^1\)\(^-\)\(^3\) Of the SNHL described in children, 50% is thought to be genetic, 25% acquired, and 25% of unknown etiology.\(^4\)

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Radiographic evaluation of inner ear abnormalities began with plain film tomography in the early 1940s.\(^3\) Through the increased resolution of computed tomography (CT) in the late 1970s and early 1980s, additional detail of the cochlear-vestibular complex was identified.\(^5^\) Of children with SNHL, genetic or acquired, 11% to 41% have had inner ear abnormalities identified with CT.\(^5^\)\(^-\)\(^7\)\(^9\)

Both plain film and CT, however, define mainly the bones that contain the actual working components of inner ear hearing. Previous studies,\(^5^\)\(^-\)\(^10\) however, have shown that high-resolution magnetic resonance imaging (MRI) used as a diagnostic tool in the evaluation of pediatric SNHL has yielded promise in better defining the exact soft tissue abnormalities of the hearing pathway rather than their bony housing. Specifically, MRI provides better ability to directly visualize the cochlear nerve and intracranial structures.
METHODS

The medical records of patients presenting to the pediatric otolaryngology clinic at Children’s Medical Center of Dallas between June 1, 1996, and June 1, 2002, who had undergone MRI for the evaluation of SNHL were reviewed. Patients with a clinical history of a known traumatic, infectious, or toxic cause of hearing loss were excluded. Approval for review of information was obtained through the institutional review board of the University of Texas at Southwestern and Children’s Medical Center of Dallas.

The MRIs were obtained with a 1.5-T scanner with a 3-dimensional fast spin-echo T2-weighted sequence using surface coils (repetition time, 4000 milliseconds; echo time, 130 milliseconds; NEX 1). Images were 0.8-mm thick. Direct and/or reconstructed sagittal oblique images of the contents of the internal auditory canal (cochlear, vestibular, and facial nerve) were obtained perpendicular to the long axis of the internal auditory canals. Both direct and reconstructed sagittal oblique images evaluated in the study had equal diagnostic value. Inadequate reconstructed images were not included in the study. All measurements were performed in the axial plane except for the cochlear and vestibular nerve measurements, as described herein. Coronal images were not included. A board-certified neuroradiologist (T.N.B.) who is fellowship-trained in pediatric neuroradiology reviewed the images. Abnormalities noted on MRI were categorized into 7 anatomical subgroups: cochlear, cochlear nerve, vestibule, vestibular nerve, endolymphatic duct (ELD), endolymphatic sac (ELS), and intracranial.

Cochlear dysplasias were graded as mild, moderate, or severe. The term mild dysplasia was assigned to abnormalities that spanned from an isolated modiolar deficiency (Figure 1A) to an absence of the apical turn of the cochlea. Severe dysplasia was a spectrum of abnormalities that spanned from single turn of the cochlea to complete aplasia of the cochlea (Figure 2). Moderate dysplasia fell between the 2 classifications, being exemplified most commonly by a Mondini type of malformation. Vestibular dysplasia was classified as mild, moderate, or severe as well. Mild vestibular dysplasia was described as a mildly abnormal vestibule with or without 1 abnormal semicircular canal. Severe vestibular dysplasia consisted of a single sac with no identifiable semicircular canals. Moderate vestibular dysplasia fell between these 2 classifications.

The cochlear nerve size, internal auditory canal (IAC) size, and cochlear nerve canal (CNC) size were all measured. A normal cochlear nerve was considered equal to or greater in diameter to the facial nerve as visualized by direct and/or reconstructed sagittal oblique images of the IAC. A deficient cochlear nerve was present but smaller than the adjacent facial nerve. If the cochlear nerve was not visualized, the nerve was noted to be absent (Figure 3). When encountered, an absent vestibular nerve was noted.

On the oblique sagittal T2-weighted images, the IAC was measured at its midportion, where there was good separation between the cochlear nerve and the remainder of the vestibular complex within the IAC (Figure 4). The IAC was considered normal if it measured 4 mm or greater in 1 dimension on the oblique sagittal image. Two dimensions were recorded, and the smallest was used for statistical analysis. The CNC was measured on the axial 3-dimensional T2-weighted images (Figure 5). A value of 1.8 mm

Figure 1. Axial 3-dimensional T2-weighted images through the internal auditory canal (IAC). A, Mild dysplasia of the left cochlea is present with a flattened deficient modiolus (arrow). The cochlear nerve is absent and the IAC is normal in size. B, The right ear is normal with a well-formed modiolus (arrow).

Figure 2. Axial 3-dimensional T2-weighted image through the internal auditory canal (IAC). The right IAC is atretic with no cochlear nerve demonstrated. Severe cochleovestibular dysplasia is present with only a single sac in the expected location of the membranous labyrinth (arrow). The left ear is normal.
or greater was used for the definition of a normal CNC. Both measurements were made using electronic calipers. Receiver operating characteristic curves were generated to identify a cut-off value of either the IAC size or the CNC size that would indicate the likelihood of an absent or deficient cochlear nerve by comparing the presence of a deficient or absent cochlear nerve with either the size of IAC or CNC in the ears of children with and without SNHL.

Of the 170 children, 166 underwent imaging evaluation of the brain. Sagittal T1-weighted images and either axial fluid-attenuated inversion recovery or T2-weighted images (depending on age) were obtained in these patients. Additional sequences were performed as per the discretion of the on-service neuroradiologist. Intracranial findings, such as minimal gliosis (<10 punctate regions of increased T2-weighted signal), dilated Virchow-Robin spaces, and small arachnoid cysts, were not included as abnormalities because these findings can be seen in the healthy pediatric population.

Sensorineural hearing loss in the ears of these children was divided into categories based on the degree of hearing loss and the configuration of the hearing loss. Categories of severity of SNHL consisted of mild (15-34 dB), moderate (35-54 dB), severe (55-85 dB), and profound (85 dB). The hearing loss in these categories was essentially flat, with no greater than a 15-dB variation in each of the frequencies tested from 250 to 8000 Hz. The specific frequencies tested varied based on the child’s age and cooperativeness. The overall degree of hearing loss was defined by the average of the hearing loss in each of the frequencies evaluated.

Other configurations of SNHL identified were defined as sloping, fluctuating, sudden, high frequency, and progressive. Sloping SNHL was defined by normal or mild SNHL in the low frequencies and severe to profound SNHL in the high frequencies. Fluctuating hearing was determined by evaluating serial audiograms and noting both improvement and worsening of hearing of greater than a 15-dB difference. Sudden SNHL was defined by a sudden clinical awareness in change of hearing and a documented abnormal or worsening audiogram. High-frequency SNHL was defined by normal hearing in the frequencies up to 2000 Hz, with a severe to profound hearing loss in the higher frequencies. Progressive SNHL was defined by the decrease in hearing loss of more than 15 dB over time, documented by serial audiograms.

Descriptive statistical analysis was used to help summarize the baseline characteristics of the patients. Categorical data were described using frequency measurements, and comparison of differences was calculated using the χ² test or the Fisher exact test where appropriate.
RESULTS

From June 1, 1996, to June 1, 2002, 227 children, aged 1 month to 17 years, underwent MRI of the brain and inner ear for SNHL. Fifty-seven children were excluded because of technically inadequate MRIs or because their medical records were not available for review, leaving 170 children in the study. The male to female ratio was 1:1, and the mean age of the patients was 5.3 years. Of the 170 children, 101 (59%) presented with bilateral SNHL and 69 (41%) with unilateral SNHL, totaling 271 ears with SNHL. Inner ear abnormalities were identified in 108 ears (40%). In the 69 ears with normal hearing, the MRI did not demonstrate any anomalies of the labyrinth or cochlear nerve. This subset of ears was used for comparison for IAC and CNC size.

Of 271 ears with SNHL, 87 (32%) displayed abnormal cochleae (Table 1). Abnormalities ranged from isolated modiolar deficiency to complete aplasia. Mild cochlear dysplasia was noted in 63 of 87 ears (72%), and moderate or severe dysplasia was seen in 24 of 87 ears (28%). Additional single or multiple anomalies in these 87 ears with abnormal cochlea occurred in 54 (62%), including 21 (24%) with an ELS and ELD (Figure 6), 20 (23%) with vestibular abnormalities, 25 (29%) with absent cochlear nerves, 12 (14%) with deficient cochlear nerves, and 11 (13%) with absent vestibular nerves.

The 271 ears of children with SNHL were categorized by type and divided into mild (29/271 [11%]), moderate (51/271 [19%]), severe (28/271 [10%]), profound (110/271 [41%]), progressive (8/271 [3%]), sloping (4/271 [1%]), sudden (3/271 [1%]), fluctuating (4/271 [1%]), high-frequency (4/271 [1%]), and unknown (30/271 [11%]) SNHL (Table 2). Ears with severe and profound SNHL had more abnormalities than ears with mild and moderate SNHL (66/138 [48%] vs 23/80 [29%]; P = .006). Children having ears with unilateral moderate, severe, or profound SNHL had more inner ear abnormalities than children with bilateral moderate, severe, or profound SNHL (66/138 [48%] vs 54/144 [38%]; P = .004). In addition, of the 139 patients with severe to profound hearing loss, 36 (26%) had either a cochlear nerve abnormality or an intracranial mass that would not be picked up on CT. When compared with patients with other types of hearing loss (eg, mild, moderate), this finding was statistically significant (P < .001).

Of 271 imaged ears with SNHL, 49 (18%) demonstrated deficient or absent cochlear nerves (Figure 3); of these 49 ears, 23 (47%) were deficient and 26 (53%) were absent. Of these 49 ears of children with abnormal cochlear nerves, 37 (76%) had associated cochlear abnormalities, with 22 of the 37 (59%) abnormal cochlear nerves associated with mild dysplasia and 15 (41%) abnormal cochlear nerves associated with moderate or se-
vere dysplasia. A normal cochlea was present in 12 of the 49 ears (24%) with an abnormal cochlear nerve. Absent cochlear nerves (n=26) specifically occurred in 21 children and only in children with severe or profound SNHL. Of the 21 children with absent cochlear nerves, 13 (62%) had unilateral SNHL, 2 (10%) had asymmetrical hearing loss, and 6 (29%) had bilateral profound SNHL.

The size of the IAC and CNC statistically correlated with the presence of an abnormal cochlear nerve; however, when using size criteria of the IAC and/or the CNC to predict the presence of an abnormal cochlear nerve, many abnormalities would be missed. In the 49 ears with an abnormal cochlear nerve, 16 (33%) had a normal IAC (4-5 mm) and 18 (37%) had a normal CNC (1.8-2.2 mm). Of the 49 ears, 9 (18%) had both a normal IAC and CNC. Receiver operating characteristic curves were generated comparing the presence of cochlear nerve abnormalities with the size of the IAC. Using an upper limit IAC cutoff value of 4 mm in an attempt to identify all abnormal cochlear nerves, a sensitivity of 76% is obtained, meaning that 24% of the cases of cochlear nerve deficiency or absence occurred in IAC canals greater than 4 mm and would be missed if only the size of the IAC were considered. Similarly, using a CNC measurement of 1.8 mm as an upper limit cutoff value to identify cochlear nerve abnormalities is even less sensitive at 68%, indicating that 32% of abnormal-sized cochlear nerves would be missed if only a small size of the CNC is considered.

Of 271 inner ears, 24 (9%) had vestibular abnormalities. Findings ranged from dilation of the vestibule to absence of all semicircular canals and a small rudimentary vestibule. In addition, there were 13 (5%) absent vestibular nerves, with 12 (92%) also having absent or deficient cochlear nerves. Of 271 ears, 27 (10%) demonstrated a dilated ELS. Of these 27 ears, 22 (81%) were associated with an abnormal cochlea (Figure 6), with 11 (50%) associated with mild cochlear abnormality, including a modiolar defect. Of 271 ELSs imaged, 30 (11%) were enlarged. Isolated enlargement of the ELS was found in 3 (1%) (Figure 7). In the 166 children with intracranial imaging performed, 33 (20%) demonstrated intracranial abnormalities (Table 3). The most common intracranial findings included gliosis in 11 of 166 (7%), cortical dysplasia in 3 of 166 (2%), brainstem hypoplasia in 5 of 142 (4%), and brainstem or cerebellar tumors in 3 of 142 (2%) (Figure 8). Central nervous system changes consistent with cytomegalovirus infection were also seen (Figure 9). Chiari I malformations, imaging findings consistent with kienic- terus, leukodystrophy, periventricular leukomalacia, and anomalies of the corpus callosum were encountered as well. Of 33 of these intracranial findings, 12 (37%) were associated with inner ear abnormalities. Most children with SNHL and intracranial abnormalities (21/33 [64%]) had no inner ear anomaly to account for their SNHL.

Table 2. Inner Ear and CNS Abnormalities Based on SNHL Type

<table>
<thead>
<tr>
<th>SNHL Type</th>
<th>No. of Ears</th>
<th>Mild/Moderate to Severe Cochlear Abnormalities</th>
<th>Deficient/Absent Cochlear Nerve</th>
<th>Absent Vestibular Nerve/Vestibular Abnormalitiesa</th>
<th>ELD/ELS/CNS Abnormalities</th>
<th>Unilateral/ Bilateral Abnormalities</th>
<th>No./Total (%) of Ears With Abnormalities</th>
<th>No./Total (%) of Unilateral Ears With Abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>29</td>
<td>3/1</td>
<td>2/0</td>
<td>0/0</td>
<td>2/2/1</td>
<td>4/25</td>
<td>7/29 (24)</td>
<td>1/4 (25)</td>
</tr>
<tr>
<td>Moderate</td>
<td>51</td>
<td>12/0</td>
<td>3/0</td>
<td>0/2 (mild)</td>
<td>1/2/8</td>
<td>11/40</td>
<td>16/51 (31)</td>
<td>6/11 (53)</td>
</tr>
<tr>
<td>Severe</td>
<td>28</td>
<td>4/5</td>
<td>3/2</td>
<td>0/2 (1 severe and 1 mild)</td>
<td>5/5/5</td>
<td>10/18</td>
<td>12/28 (43)</td>
<td>5/10 (50)</td>
</tr>
<tr>
<td>Profound</td>
<td>110</td>
<td>29/16</td>
<td>11/20</td>
<td>11/17 (2 severe, 6 moderate, and 9 mild)</td>
<td>13/15/17</td>
<td>24/86</td>
<td>54/110 (48)</td>
<td>17/24 (71)</td>
</tr>
<tr>
<td>Sloping</td>
<td>4</td>
<td>0/0</td>
<td>0/0</td>
<td>0/0</td>
<td>0/0/0</td>
<td>4/0</td>
<td>0/4 (0)</td>
<td>0/4 (0)</td>
</tr>
<tr>
<td>High frequency</td>
<td>4</td>
<td>1/0</td>
<td>0/0</td>
<td>0/0</td>
<td>1/0/1</td>
<td>1/3</td>
<td>1/4 (25)</td>
<td>1/1 (100)</td>
</tr>
<tr>
<td>Progressive</td>
<td>8</td>
<td>5/0</td>
<td>0/0</td>
<td>0/1 (mild)</td>
<td>4/4/1</td>
<td>0/8</td>
<td>6/8 (75)</td>
<td>0/0 (0)</td>
</tr>
<tr>
<td>Sudden</td>
<td>3</td>
<td>1/0</td>
<td>0/1</td>
<td>0/0</td>
<td>0/0/0</td>
<td>3/0</td>
<td>1/3 (33)</td>
<td>1/3 (33)</td>
</tr>
<tr>
<td>Fluctuating</td>
<td>4</td>
<td>0/0</td>
<td>0/0</td>
<td>0/0</td>
<td>0/0/0</td>
<td>0/2</td>
<td>0/2 (0)</td>
<td>0/0 (0)</td>
</tr>
<tr>
<td>Unknown</td>
<td>30</td>
<td>7/1</td>
<td>1/3</td>
<td>1/2 (mild)</td>
<td>0/0/4</td>
<td>12/18</td>
<td>8/30 (27)</td>
<td>7/12 (58)</td>
</tr>
</tbody>
</table>

Abbreviations: CNS, central nervous system; ELD, endolymphatic duct; ELS, enlarged endolymphatic sac; SNHL, sensorineural hearing loss.

aSeverity of vestibular abnormality is in parentheses.

Figure 7. Axial 3-dimensional T2-weighted image inferior to the internal auditory canal. There is isolated enlargement of the endolymphatic sac (arrow). The vestibular aqueduct is normal in size (not shown).

Children with SNHL present to the otolaryngologist in the neonatal period through adolescence. Although the
specific origin of SNHL may remain undiagnosed in many patients, a thorough workup to identify the cause of SNHL should be considered in each patient. Historically, high-resolution CT has been the imaging modality of choice in the initial workup of these patients. However, the soft tissue structures of the inner ear responsible for the electrochemical transfer of sound to the brain, such as the membranous labyrinth and the cochlear nerve, are not evaluated well with high-resolution CT. Magnetic resonance imaging directly assesses these soft tissue components of hearing when imaging for deficiencies of the auditory pathway in children with SNHL.

Embryologically, the soft tissues of the inner ear form after the bony labyrinth. Development of the bony cochlea begins in the third embryonic week with the appearance of the otic placode and is completed by 9 weeks of gestation. Subsequently, the afferent nerve fibers appear, entering the undifferentiated otic epithelium. The membranous labyrinth continues to differentiate into the 26th week of gestation. Thus, membranous deficiencies of the cochlea may not be visualized by high-resolution CT because a normal bony cochlea may be present with arrested development of the membranous components of the hearing pathway.

The most common abnormality identified in this study was cochlear dysplasia in 87 of the 271 imaged ears (32%). Severe cochlear dysplasia was seen in 24 of the 271 ears (9%) and is similar to the 12% of cochlear dysplasias reported. Mild cochlear dysplasias, which included modiolar deficiency and absence of the apical turn, were found in 63 of the 271 ears (23%) of children with SNHL, twice the 11% incidence reported using high-resolution CT in the evaluation of modiolar deficiencies. However, MRI has been reported to be of increased sensitivity in evaluation of cochlear dysplasia, especially modiolar deficiencies, which could explain the difference.

Both the cochlear nerve and the ELD course through bony canals that have normal and abnormal dimensions previously defined by high-resolution CT. Measuring bony canals does not always provide accurate information on its membranous components. While a small IAC was shown by McClay et al to more often occur in children with SNHL, inferring abnormality of the cochlear nerve or cochlear vestibular complex, an absent cochlear nerve has been seen in normal-sized IACs. Similarly, the bony CNC is an indirect measurement of the cochlear nerve and leaves room for inaccuracies.

Magnetic resonance imaging can directly evaluate the facial-vestibulocochlear nerve complex whether the IAC or CNC is small or large. The CNC and IAC were measured in all ears, and a correlation was found between cochlear nerve abnormalities and the size of the IAC and CNC—meaning as the canals became smaller, a greater percentage of the cochlear nerves became absent or deficient. However, receiver operating characteristic curves showed that a significant number of ears with abnormalities of the cochlear nerve would be missed if using only the criteria of size of their bony canals—meaning abnormal-sized or absent cochlear nerves were found in normal-sized IAC. We used the normal hearing ears in these children with SNHL as the controls for the receiver operating characteristic curves.

Many ears in our series of patients had demonstrable abnormalities of the cochlear nerve (18%), with more than half (26/49 [53%]) having an absent nerve. An absent cochlear nerve would be a deterrent to standard cochlear implantation. A deficient cochlear nerve may not necessarily be a deterrent to cochlear implantation because some benefit has been reported in children with cochlear implants placed in ears with deficient-sized cochlear nerves, despite the fact that the diameter of the cochlear nerve has been directly related to the spiral ganglion count. Certainly, it would seem that a normal-sized cochlear nerve would be preferable to a deficient one. At our institution, if unilateral cochlear implantation is performed, the side with the largest cochlear nerve undergoes implantation.

An enlarged vestibular aqueduct (EVA), which houses the ELD, was previously noted to be the most common inner ear abnormality associated with SNHL, with an incidence of 11% to 15%. In our MRI study, its incidence of 27 in 271 ears (10%) was similar. It was not the most common finding in ears of children with SNHL because of the increased sensitivity of MRI in identifying mild and modiolar abnormalities of the cochlea. Davidson et al reported a high association (76%) of mild cochlear and modiolar anomalies associated with EVA and ELDS when using MRI. We, as well, had 22 of 27 ELDS associated with mild cochlear anomalies, of which half (11 of 22 [50%]) were associated with mild cochlear abnormalities that previously could have been missed. This association, in part, may explain the high reported incidence of EVA and ELDS in children with SNHL.

Although many bony inner ear findings seen on high-resolution CT, such as cochlear malformations and EVA, coincide with their associated soft tissue disease, many do not. Intracranial abnormalities, near or distance to the inner ear, were seen in this study. The ELS, which helps regulate endolymph of the membranous inner ear, is less well defined by high-resolution CT because only a small portion of the ELS lies within the temporal bone. The MRI identified 30 of the 271 ears (11%) with SNHL to have an enlarged ELS. Of these 30 ears, 3 (10%) were isolated findings and not associated with an enlarged ELD (Figure 7). The utility of this finding in association with SNHL has not been established, but its presence is easily documented with MRI.

Table 3. Intracranial Abnormalities Seen on 3-Dimensional Fast Spin–Echo T2-Weighted Magnetic Resonance Images in Children With Sensorineural Hearing Loss

<table>
<thead>
<tr>
<th>Intracranial Abnormality Subtype</th>
<th>No. (%) of 166 Children</th>
</tr>
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<tbody>
<tr>
<td>Central nervous system</td>
<td>11 (7)</td>
</tr>
<tr>
<td>Isolated gliosis</td>
<td>3 (2)</td>
</tr>
<tr>
<td>Cortex dysplasia</td>
<td>3 (2)</td>
</tr>
<tr>
<td>Corpus callosum dysgenesis</td>
<td>5 (3)</td>
</tr>
<tr>
<td>Other</td>
<td>2 (1)</td>
</tr>
<tr>
<td>Cerebellum</td>
<td>3 (2)</td>
</tr>
<tr>
<td>Chiari I</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Tumor</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Brainstem</td>
<td>5 (3)</td>
</tr>
</tbody>
</table>

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Furthermore, abnormalities of the cerebral cortex, cerebellum, and brainstem are well delineated with MRI. In this study, of the 33 children with intracranial abnormalities, 12 (36%) had associated abnormalities of the inner ear and 21 (64%) were isolated findings, not associated with an inner ear abnormality. The intracranial findings included gliosis, astrocytoma, Chiari I malformations, dysplasias of the cortex, and brainstem hypoplasia, among others. Three children had profound unilateral SNHL as their only complaint and had normal high-resolution CT scans. Subsequent MRIs identified large pilocytic astrocytomas of the brainstem in 2 of the children at the cochleovestibular nerve (Figure 8) and a cerebellar mass in the third. Furthermore, the association of isolated central nervous system findings in children with SNHL may guide further workup and treatment of these patients since another child had central nervous system findings consistent with a previously unknown cytomegalovirus infection.

Simons et al9 recently reemphasized the need for high-resolution CT scan as the initial test for children with SNHL. They reported only 4 of 42 abnormalities (10%) seen on MRI that were missed on high-resolution CT but commented on only 1 abnormal cochlear nerve. We found cochlear nerve abnormalities more often, in 49 of 271 ears (18%). In the study by Simons et al, identification of radiographic abnormalities occurred by reviewing the reports, not the actual scans, which could lead to underestimating the abnormalities, depending on the awareness and skill of the reporting pediatric neuroradiologist. We rereviewed all of MRIs in these children with SNHL for our study.

In addition, Simons et al9 described 9 children who had abnormalities seen on high-resolution CT but missed by MRI. Of the 9 abnormalities, 6 (67%) were EVA or ELDs, for which the normal size is controversial.14,21 An abnormal membranous ELD should always have an enlarged bony vestibular aqueduct, so this part of their report seems odd. They also described 2 of 9 children with cochlear dysplasias and a small IAC that were missed by MRI. Magnetic resonance imaging has been reported to be more sensitive than CT in defining cochlear abnormalities,15 a finding seen in our group of patients as well. In addition, a small IAC really is only significant when it identifies an absent or deficient cochlear nerve or cochleovestibular complex, which we have no trouble defining on MRI. Their final patient with a missed abnormality on MRI had mixed hearing loss and abnormalities of the ossicles. We obtain a high-resolution CT scan and a MRI when a child has a conductive component to their hearing abnormality.

We use MRI as the primary imaging modality for children with SNHL. However, we found that the degree of SNHL may help define when MRI may be of the highest yield. In general, as the hearing worsened, the percentage of abnormalities of the inner ear increased (Table 2). Ears of children with severe and profound SNHL had statistically significantly more abnormalities seen on MRI than ears of children with mild or moderate loss (48% vs 29%; \( P = .006 \)). This trend was especially noted in pa-
tients with unilateral hearing loss compared with the children who had bilateral hearing loss (Table 2). The numbers of ears of children with other types of hearing loss, such as sloping, high frequency, sudden, fluctuating, and progressive, were too small to draw conclusions.

Certainly, the imaging modalities have varying associated risk factors. Although CT is fast and often does not require general anesthesia, the radiation exposure for a complete temporal bone scan is not insignificant. Although MRI has no associated radiation exposure, it is a longer procedure, and most children younger than 6 to 8 years need sedation, possibly general anesthesia. Because MRI is generally more expensive than a CT scan, a tailored approach to radiographic imaging could be implemented. The greatest yield of MRI over a CT scan would be in identifying the soft tissue components of hearing that have no bony counterparts, such as abnormalities of the cochlear nerve and abnormalities of the central nervous system that may have significance in these children with SNHL, such as an isolated ELS or brain tumor. If high-resolution CT is primarily used, obtaining MRIs in the subset of children with severe or profound SNHL, especially those with unilateral loss, should identify most of these abnormalities.

In conclusion, the incidence of inner ear findings seen with MRI compares favorably with prior reports using high-resolution CT. Mild cochlear dysplasias and modiolar deficiencies were found more frequently than previously reported by high-resolution CT. In addition, 9% of children with SNHL were found to have an absent cochlear nerve, which could impact cochlear implantation. With MRI, these soft tissue components of hearing from the cochlea to the auditory cortex can be elucidated, which should improve our ability to appropriately diagnose the location of the defect in these children with SNHL.

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Author Contributions: All authors had full access to all of the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: McClay, Booth, and Parry. Acquisition of data: McClay, Booth, and Parry. Analysis and interpretation of data: McClay, Booth, Parry, Johnson, and Roland. Drafting of the manuscript: McClay, Booth, and Parry. Critical revision of the manuscript for important intellectual content: McClay, Booth, Parry, Johnson, and Roland. Statistical analysis: Parry and Johnson. Administrative, technical, and material support: McClay and Booth. Study supervision: McClay.

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REFERENCES


