Computed Tomography and Magnetic Resonance Imaging in Pediatric Unilateral and Asymmetric Sensorineural Hearing Loss

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Objective: To compare temporal bone computed tomography (CT) with temporal bone and central nervous system magnetic resonance (MR) imaging in children with unilateral or asymmetric sensorineural hearing loss (SNHL).

Design: Retrospective chart study.

Setting: Tertiary-care children’s hospital.

Patients: A total of 131 children with unilateral or asymmetric SNHL, seen consecutively by a single practitioner over 36 months.

Intervention: Imaging studies were read by a pediatric neuroradiologist and reviewed by the evaluating otolaryngologist.

Main Outcome Measure: Prevalence of clinically significant CT or MR imaging findings.

Results: The prevalence of CT abnormalities was 35% for unilateral SNHL, 52% for asymmetric SNHL, and 41% for all patients together. The prevalence of MR imaging abnormalities was 25% for unilateral SNHL, 50% for asymmetric SNHL, and 30% for all patients together. Among 42 subjects who underwent both studies, there were 4 cases in which abnormalities were seen only on MR images and 9 cases in which abnormalities were seen only on CT scans.

Conclusions: Temporal bone and/or central nervous system abnormalities were detected in 42% of 131 patients. When both CT scans and MR images were obtained (n=42), results were concordant in 69% of cases, and one imaging modality detected clinically significant abnormalities not identified by the other in 31% of cases. The ideal imaging algorithm for children with unilateral or asymmetric SNHL is controversial. We suggest that all children with unilateral or asymmetric SNHL have a high-resolution temporal bone CT scan and that brain and temporal bone MR imaging be obtained in select cases.


APPROXIMATELY 3 IN EVERY 1000 newborn children have moderate or worse sensorineural hearing loss (SNHL), and about half of these children have a genetic basis for their hearing loss.1,2 Although SNHL is relatively common, it is often underdiagnosed and underappreciated in children.3 Historically, unilateral hearing loss is frequently not recognized until kindergarten or early grade school, when many children undergo their first audiometric evaluation.3 The prevalence of unilateral SNHL in children is estimated to be approximately 0.1%.4,5 Unilateral hearing loss in children is associated with an increased likelihood of academic difficulty, behavior problems in the classroom, and requirement to repeat a grade in school.5

Radiologic abnormalities of the inner ear are relatively common in children with SNHL, and their identification directly impacts prognosis and management.6-8 Therefore, many authors suggest that all children with SNHL undergo radiologic imaging as part of their evaluation.4,6-9

High-resolution temporal bone computed tomography (CT) has been the first-line imaging study of choice obtained by pediatric otolaryngologists in the workup of children with all types of SNHL, including symmetric, asymmetric, and unilateral SNHL.4,6,8,9,10 Adults with asymmetric or unilateral SNHL often undergo gadolinium-enhanced magnetic resonance (MR) imaging of the brain with attention to the internal auditory canals to evaluate for retrocochlear abnormalities.11,12 In contrast to adults, children with unilateral or asymmetric SNHL rarely have acoustic neuromas, and therefore the indications for MR imaging are less clear.12 Many authors have recommended MR imaging as part of the preoperative workup for children with congenital bilateral SNHL being examined for a cochlear implant procedure.13 In children with SNHL, MR im-
aging was recently shown to detect soft tissue abnormalities involving the eighth nerve and intracranial contents that would not have been identified on CT. Some authors believe that MR imaging should be the preferred study in pediatric patients with unilateral or asymmetric bilateral SNHL.

Few studies have directly compared CT and MR imaging results in pediatric patients with unilateral or asymmetric bilateral SNHL. The purpose of this study was to examine our experience using high-resolution temporal bone CT and temporal bone and brain MR imaging in children with unilateral or asymmetric bilateral SNHL. We compared the diagnostic utility of high-resolution temporal bone CT with that of temporal bone and central nervous system MR imaging in patients who had both studies. We hypothesized that, while both imaging modalities would often identify the same abnormalities in a given patient, each would also identify clinically significant radiographic abnormalities that were not seen on the other.

METHODS

This retrospective study was a chart review and data analysis of patients who were seen in the Hearing Center at the Children's Hospital of Pittsburgh, Pittsburgh, Pa (a tertiary-care children's hospital), between May 1, 2000, and May 31, 2003. The protocol was reviewed by the hospital's human rights committee and approved by expedited review, in compliance with guidelines of the Health Insurance Portability and Accountability Act.

All patients included in the study had either unilateral or asymmetric bilateral SNHL. Unilateral SNHL was defined as a hearing threshold greater than 20 dB hearing level (HL) for at least 1 frequency (500-4000 Hz). Asymmetric bilateral SNHL was defined as 2 or more frequencies with greater than a 15-dB difference in threshold between the 2 ears or 3 or more frequencies with greater than a 10-dB difference in threshold between the 2 ears. Severity of hearing loss was defined as follows: 21 to 40 dB HL, mild; 41 to 70 dB HL, moderate; 71 to 95 dB HL, severe; and greater than 95 dB HL, profound. Pure-tone average was defined as the average hearing threshold at 500, 1000, and 2000 Hz. Nine patients in the study were too young to be examined with pure tone audiometry; unilateral or asymmetric bilateral SNHL was determined for these patients on the basis of auditory brainstem response testing, but severity could not be specifically graded. There were 2 patients with mixed hearing loss; for purposes of the study, the conductive component of the hearing loss was not included in the analysis. The information collected for each patient on a de-identified patient data sheet included age at diagnosis, sex, whether hearing loss was unilateral or asymmetric bilateral, severity of hearing loss based on pure-tone average, cause of hearing loss if known, and CT and MR imaging results if obtained.

All imaging studies were performed by the Radiology Department at Children's Hospital of Pittsburgh, with the specific request to search for potential causes of SNHL. Final readings were dictated by a pediatric neuroradiologist and reviewed by the evaluating otolaryngologist (E.M.A.). The CT and MR imaging findings that were used for the study were based on the previously dictated neuroradiology reports and otolaryngology attending clinic notes; the actual scans were not reviewed when studies were compared. Any positive results detected by either physician were reported in this study. All CT scans obtained were high-resolution scans of the temporal bones with 1-mm axial sections and coronal reconstructions (GE HiSpeed CT/i CT scanner; GE Healthcare, Milwaukee, Wis.). The MR imaging was performed with standard brain imaging protocols including T1- and T2-weighted images plus high-resolution axial T2 images through the posterior fossa and axial 3-dimensional fast spin-echo images through the posterior fossa and internal auditory canals, with sagittal reconstructions through the internal auditory canals (GE LX 1.5-T MR imager; GE Healthcare).

For each patient, the decision to obtain CT scans, MR images, or both was individualized at the discretion of the evaluating otolaryngologist. During the period of the study, there was no established protocol to follow regarding which study, if any, to order. Most of the patients identified with SNHL in our practice did undergo an imaging study, with temporal bone CT being the most commonly ordered, many times for logistical reasons such as shorter duration of sedation required, easier accessibility, and earlier scheduling. In general, MR imaging was reserved for cases of asymmetric or unilateral SNHL (the focus of this study) and was sometimes the first imaging study ordered, especially in cases where there were other neurodevelopmental signs and symptoms. When CT was performed first, MR imaging was always recommended as a follow-up study if the CT scan was nondiagnostic, and MR imaging was usually suggested to confirm positive CT findings, although not all subjects’ caregivers agreed to proceed with the MR imaging in this setting. Although most patients had either CT or MR imaging, both studies were obtained in a sizable subset of patients (n=42), which allowed comparison of the results.

Results of CT and MR imaging studies for each subject were examined and compared. The main outcome measure evaluated was the prevalence of clinically significant abnormal CT and MR imaging findings related to the cause of SNHL (defined as a positive finding). A large vestibular aqueduct (LVA) was defined as being greater than 1.5 mm at the midpoint of the vestibular aqueduct on axial images, as described in previous studies. The prevalence of clinically significant CT and MR imaging findings was examined separately for subjects with unilateral SNHL and for those with asymmetric bilateral SNHL, as well as for all subjects together. For subjects who underwent both CT and MR imaging, the frequency with which abnormalities were seen on only one of the imaging modalities, but not the other, was calculated. Furthermore, the relationship of imaging results to severity of hearing loss (based on pure-tone average) was evaluated.

For all statistical tests, significance was defined as P<.05. The unpaired, 2-tailed t test was used to compare the age at diagnosis between the unilateral SNHL and asymmetric bilateral SNHL groups. The McNemar test was used to compare the proportion of subjects with positive CT and MR imaging results for both groups, as well as all subjects together. The χ² test was used to analyze the difference in prevalence of clinically significant abnormalities between the unilateral and asymmetric SNHL groups. The significance of the association between imaging results and severity of hearing loss was also determined by the χ² test.

RESULTS

Three hundred sixty-five patients were seen in the Hearing Center at the Children's Hospital of Pittsburgh during the period studied. One hundred forty-seven (40%) of these patients had unilateral or asymmetric SNHL, among whom 98 (67%) had unilateral SNHL and 49 (33%) had asymmetric SNHL. The age and sex distribution for these groups is shown in Table 1. There was a statistically significant difference in the age at diagnosis for the 2 groups (P<.001).
The CT and/or MR imaging studies were available for review in 131 of these 147 patients (Table 2). For the remaining 16 patients, scans were unavailable for review, declined by the parents, or not performed because of acquired SNHL of known cause. The prevalence of clinically relevant abnormalities on these imaging studies is provided in Table 3. When the unilateral and asymmetric SNHL groups were compared, there was a statistically significant difference in prevalence of clinically relevant abnormalities on at least 1 imaging modality (P = .03), with a greater number of positive findings in the asymmetric SNHL group. However, this difference between the unilateral and asymmetric SNHL groups was not significant when the prevalence of clinically relevant abnormalities on CT (P = .06) or MR imaging (P = .12) alone was examined.

The prevalence of positive and negative imaging findings for subjects who underwent both CT and MR imaging was compared (Table 4). There was no significant difference in the percentage of subjects with positive CT results and positive MR imaging results in the population that had both studies. For subjects with unilateral SNHL who underwent both CT and MR imaging, results were concordant in 76% of cases (25/33), and one imaging modality detected clinically significant abnormalities not detected by the other in 24% of cases (8/33). For subjects with asymmetric SNHL who underwent both studies, results were concordant in 49% of cases (16/33), and one imaging modality detected clinically significant abnormalities not detected by the other in 51% of cases (17/33). For all 42 subjects in the study who underwent both CT and MR imaging, results were concordant in 69% of cases (29/42), and one imaging modality detected clinically significant abnormalities not detected by the other in 31% of cases (13/42).

There were 9 cases in which abnormalities were detected only on CT (Table 5) and 4 cases in which abnormalities were detected only on MR imaging (Table 6). Table 7 describes all of the clinically significant abnormalities identified on CT and MR imaging. An LVA was the most common abnormal finding on both CT (70% of abnormal CT scans) and MR imaging (40% of abnormal MR images). Although concordance between CT and MR imaging findings was often seen with abnormalities such as LVA (Figure 1), there were 6 subjects in whom LVA was diagnosed by temporal bone CT, but MR imaging was initially read as normal. For these 6 MR images, one of us (E.M.A.) re-reviewed the images, confirming that the endolymphatic sac did not appear to be enlarged. However, age-dependent normative data for endolymphatic sac size as determined by MR imaging are not available. In addition, an MR imaging protocol flaw could not be excluded. Other cases also illustrated a discrepancy between CT and MR imaging findings (Figure 2 and Figure 3).

Among the 42 patients who underwent both CT and MR imaging, a hypoplastic eighth nerve was detected on 3 of the MR images. Only 1 of the corresponding CT scans was read as having a small internal auditory canal.

There was no statistically significant correlation between the presence of positive findings on imaging and the severity of hearing loss based on pure-tone average for either the unilateral or asymmetric SNHL group or for all subjects together.
This study examined the diagnostic utility of high-resolution temporal bone CT and temporal bone and central nervous system MR imaging in children with unilateral or asymmetric bilateral SNHL. In general, temporal bone CT is a better imaging modality for identifying bony abnormalities such as trauma, otosclerosis, inner ear bony dysplasia, and erosive or destructive temporal bone lesions. Magnetic resonance imaging provides superior soft tissue resolution and is better for identifying lesions involving the membranous labyrinth, internal auditory canal and eighth cranial nerve, cerebellopontine angle, brainstem, and cerebral cortex. Many authors currently believe that MR imaging can successfully image both inner ear structures and the contents of the internal auditory canals.14

Not unexpectedly, children with asymmetric bilateral SNHL in the present study were diagnosed at an earlier age (mean, 3.7 years) than children with unilateral SNHL (mean, 5.9 years) (P<.001). This finding is consistent with previous studies.1 The most common radiographic abnormality on both CT and MR imaging in our study was LVA. This finding is also consistent with other studies in children with asymmetric bilateral SNHL.14

In our study, the prevalence of abnormal temporal bone CT findings in subjects with unilateral SNHL was 35%, compared with 7% to 44% in the literature.5-10 The prevalence of abnormal CT findings in subjects with asymmetric bilateral SNHL in our study was 52%. Although a direct comparison group in the literature is not available, CT abnormalities have been found in about 28% of children with bilateral SNHL (either symmetric or asymmetric).8 For all subjects with SNHL in our study, the overall prevalence of abnormal CT findings was 41%, compared with an overall prevalence of 13% to 37% in the literature.5-12

The prevalence of abnormal MR imaging findings in our study was 25% in subjects with unilateral SNHL and 50% in subjects with asymmetric bilateral SNHL. For all subjects with SNHL in our study, the overall prevalence of abnormal MR imaging findings was 30%, compared with a prevalence of 28% to 33% in the literature.5,14

**Table 5. Positive CT Findings in Subjects With Negative MR Imaging Studies**

<table>
<thead>
<tr>
<th>Subject Age, y/Sex</th>
<th>Characteristics of Hearing Loss</th>
<th>CT Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>7/F</td>
<td>Right high-frequency SNHL</td>
<td></td>
</tr>
<tr>
<td>9/F</td>
<td>Left mild to moderate SNHL</td>
<td></td>
</tr>
<tr>
<td>12/F</td>
<td>Right mild to moderate SNHL</td>
<td></td>
</tr>
<tr>
<td>3/F</td>
<td>Left moderate to severe mixed HL</td>
<td></td>
</tr>
<tr>
<td>4.5/M</td>
<td>Right profound SNHL</td>
<td></td>
</tr>
<tr>
<td>5/M</td>
<td>Right profound SNHL</td>
<td></td>
</tr>
<tr>
<td>1.5/M</td>
<td>Bilateral severe to profound SNHL(right ear worse)</td>
<td>Bilateral LVA</td>
</tr>
<tr>
<td>1.5/F</td>
<td>Right mild to severe and left mild to moderate SNHL(right ear worse)</td>
<td>Bilateral LVA</td>
</tr>
<tr>
<td>12/M</td>
<td>Right moderate and left mild SNHL(right ear worse)</td>
<td>Bilateral LVA</td>
</tr>
</tbody>
</table>

**Table 6. Positive MR Imaging Findings in Subjects With Negative CT Studies**

<table>
<thead>
<tr>
<th>Subject Age, y/Sex</th>
<th>Characteristics of Hearing Loss</th>
<th>MR Imaging Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unilateral SNHL</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5.5/F</td>
<td>Right profound SNHL</td>
<td>Hypoplastic right eighth nerve</td>
</tr>
<tr>
<td>3/F</td>
<td>Right moderate SNHL</td>
<td>Glioma in right pons</td>
</tr>
<tr>
<td>Asymmetric SNHL</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1.5/M</td>
<td>Bilateral profound SNHL (left ear worse)</td>
<td>Hypoplastic right eighth nerve</td>
</tr>
<tr>
<td>5/M</td>
<td>Bilateral moderate SNHL (right ear worse)</td>
<td>Chiari I malformation</td>
</tr>
</tbody>
</table>

**Table 7. Clinically Relevant Abnormalities Identified on CT and MR Imaging**

<table>
<thead>
<tr>
<th>Finding</th>
<th>No. (%) of Subjects</th>
<th>Finding</th>
<th>No. (%) of Subjects</th>
</tr>
</thead>
<tbody>
<tr>
<td>LVA</td>
<td>15 (92) LVA</td>
<td>IEA</td>
<td>2 (11) IEA</td>
</tr>
<tr>
<td>LVA plus IEA*</td>
<td>6 (41) LVA plus IEA*</td>
<td>CPA arachnoid cyst</td>
<td>2 (11) CPA arachnoid cyst</td>
</tr>
<tr>
<td>Small IAC</td>
<td>2 (11) Small IAC</td>
<td>IEA plus CPA arachnoid cyst</td>
<td>1 (6) IEA plus CPA arachnoid cyst</td>
</tr>
<tr>
<td>Noncommunicating hydrocephalus</td>
<td>1 (6) Noncommunicating hydrocephalus</td>
<td>Chiari I malformation</td>
<td>1 (6) Chiari I malformation</td>
</tr>
<tr>
<td>Enlarged fourth ventricle</td>
<td>1 (6) Enlarged fourth ventricle</td>
<td>Hypoplastic CN VIII</td>
<td>2 (11) Hypoplastic CN VIII</td>
</tr>
<tr>
<td>Total 21 (100) Total</td>
<td>21 (100) Total</td>
<td>10 (100) Total</td>
<td>10 (100) Total</td>
</tr>
</tbody>
</table>

**Abbreviations:** CT, computed tomography; MR, magnetic resonance; LVA, large vestibular aqueduct; SNHL, sensorineural hearing loss.

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In this study, the prevalence of clinically significant abnormalities on at least 1 imaging modality was higher for asymmetric bilateral SNHL (56%) than for unilateral SNHL (36%). In contrast, Preciado et al19 found that the diagnostic yield for imaging was higher for unilateral SNHL (37%) than for bilateral SNHL (25%). Our prevalence of positive findings in subjects with unilateral SNHL is very similar to the prevalence in their study (36% vs 37%).19 However, we found a much higher rate of imaging abnormalities in our group of patients with asymmetric bilateral SNHL (56%) than previously reported for all bilateral SNHL (25%-28%).8,19 Previous studies from the Hearing Center at the Children’s Hospital of Pittsburgh have demonstrated that patients with asymmetric bilateral SNHL have a higher prevalence of LVA and other inner ear anomalies than all patients with bilateral SNHL, which may explain why our patients with asymmetric bilateral SNHL had a higher rate of positive findings on imaging than patients with bilateral SNHL described in the literature.20,21

Among all subjects in our study who underwent both CT and MR imaging, one imaging modality detected clini-
cally significant abnormalities not detected by the other in 31% of cases (13/42). This discordance occurred for subjects with both unilateral and asymmetric SNHL. Among these 42 subjects, there were 4 cases in which abnormalities were detected only on MR imaging and 9 cases in which abnormalities were detected only on CT. In a study that included 14 children with unilateral and bilateral SNHL who underwent both CT and MR imaging, Mafong et al found that 4 of 7 patients with abnormal MR images had abnormal findings that were not detected on temporal bone CT scans.

Although some authors have suggested that temporal bone CT is the only diagnostic test that can predict the cause of pediatric SNHL, others believe that there may be a role for MR imaging in this patient population. Mafong and colleagues recommended that all children with unexplained SNHL undergo radiologic imaging and that temporal bone CT scans be obtained in patients with symmetric SNHL, whereas MR imaging with or without CT should be performed in patients with asymmetric SNHL. In children with both unilateral and bilateral SNHL, Parry et al found that MR imaging detected soft tissue abnormalities involving the eighth nerve and intracranial contents that would not have been identified on CT. Bamiou et al recommended that all children with SNHL undergo imaging studies of the temporal bones and that the decision to perform CT vs MR imaging depend on scanner availability, expertise, and other management considerations.

Most studies that have directly compared CT and MR imaging have done so in children with bilateral profound SNHL undergoing evaluation for cochlear implantation. In one study of 31 cochlear implant candidates who underwent both CT and MR imaging, the concordance rate for the 2 studies was 81%. One patient had an MR image that showed a modiolar deficiency not detected on CT scan and 5 patients had CT findings not identified on MR imaging, including 1 with a high jugular bulb, 1 with fenestral otosclerosis, 1 with cochlear aqueduct calcification, and 2 with modiolar deficiencies. Seitz and colleagues found that MR imaging provided additional information to CT in 10 (39%) of 26 patients who had both temporal bone CT and MR imaging before cochlear implantation. In a study of 40 cochlear implant candidates with bilateral profound SNHL who underwent MR imaging of the temporal bones and brain, 8 patients (20%) had significant brain abnormalities, including 5 patients with at least 1 hypoplastic eighth nerve. The present study explored expanding the indication for MR imaging in the evaluation of SNHL to children with asymmetric and unilateral SNHL, again demonstrating that certain clinically relevant, previously unidentified, central nervous system abnormalities may be detected with this modality.

Semarougou et al use CT as the primary imaging modality for cochlear implant candidates, but they also obtain MR images in the presence of total hearing loss, narrow internal auditory canal, or inner ear malformations to demonstrate the presence of the eighth nerve. Ellul et al concluded that fast spin-echo MR imaging should be the single imaging study of choice for the preoperative evaluation of most cochlear implant candidates. Other authors have also recommended MR imaging of the brain and temporal bones as part of the evaluation of cochlear implant candidates. Westerhof et al suggested the combined use of CT and MR imaging to study cochlear implant candidates.

We found that there was no significant correlation in our patient population between the presence of clinically significant abnormalities on imaging and the severity of hearing loss. Although this conclusion is in agreement with some studies in the literature, others found that patients with profound SNHL have a higher prevalence of abnormalities on imaging than patients with less severe hearing loss.

Two of the patients in our study had Chiari I malformations associated with unilateral or asymmetric SNHL. In one case, the abnormality was found on both CT and MR imaging, whereas in the other case, CT was negative and MR imaging was positive. Other authors have also noted an association between Chiari I malformations and SNHL. It has been reported that 44% of patients with MR imaging–confirmed Chiari I malformation have SNHL (30% unilateral, 14% bilateral). Hendrix et al reviewed data from 226 consecutive patients with asymmetric SNHL and identified 3 (1.3%) who were discovered to have Chiari I malformations on MR imaging as their only identifiable abnormality. Several mechanisms have been proposed to explain SNHL in patients with Chiari I malformations, including stretching of the eighth nerve, compression of the eighth nerve against the bone of the porus acousticus, and direct compression of the brainstem cochlear nuclei by the cerebellar tonsils. Another hypothesis is that compression or distortion of the posterior inferior cerebellar artery by the cerebellar tonsils leads to ischemic damage to the cochlear and vestibular nuclei. It has been suggested that posterior fossa...
Neither temporal bone CT nor MR imaging of the brain and temporal bones appears to be adequate as the sole imaging modality in all children with unilateral or asymmetric SNHL. The ideal imaging algorithm in these children is controversial. On the basis of the results of this retrospective review, we can make the following general suggestions. Virtually all children with SNHL should have an imaging study as part of their workup. For pediatric unilateral or asymmetric SNHL, central nervous system and temporal bone MR imaging should be considered, although we still prefer high-resolution temporal bone CT as the initial study because of a high prevalence of positive findings and less cumbersome logistical issues. However, we believe that a negative CT scan should be followed by MR imaging, to rule out potential central nervous system causes of SNHL that may not be detected on CT scans. As the experience with ordering and interpreting MR images for unilateral and asymmetric pediatric SNHL increases, the clinical protocol will likely become more refined.

Submitted for Publication: June 25, 2004; final revision received May 2, 2005; accepted August 27, 2005.

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Financial Disclosure: None.

Present Previous: This study was presented as a poster at the 19th Annual Meeting of the American Society of Pediatric Otolaryngology; May 2-3, 2004; Phoenix, Ariz; and received the American Society of Pediatric Otolaryngology 2004 Resident Poster Award.

Acknowledgment: We thank Elaine N. Rubinstein, PhD, for her help with statistical analysis of our data.

REFERENCES


