Utility of Temporal Bone Computed Tomographic Measurements in the Evaluation of Inner Ear Malformations

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Objective: To investigate whether normative inner ear measurements can assist in the evaluation of sensorineural hearing loss (SNHL).

Design: Retrospective cohort review.

Setting: A tertiary care hospital.

Patients: Computed tomography or magnetic resonance imaging was performed on 188 ears with SNHL and 220 ears without SNHL (204 children) between 2001 and 2004.

Intervention: Two readers measured the basal turn of cochlea (BTC) lumen, lateral semicircular canal (LSCC) bony island width, superior semicircular canal (SSCC) bony island width, and cochlear height (CH).

Main Outcome Measures: A t test was performed comparing measurements in patients with and without SNHL. Interobserver variability was characterized by intraclass correlation coefficients and Bland-Altman plots.

Results: The t test results demonstrated no statistically significant differences between inner ear measurements in those with and without SNHL. The intraclass correlation coefficients for BTC lumen, CH, LSCC bony island width, and SSCC bony island width measurements was 0.612, 0.632, 0.869, and 0.912, respectively. Bland-Altman plots revealed systematic biases of 1%, 8%, 10%, and 21% for the BTC lumen, SSCC bony island width, LSCC bony island width, and CH measurements, respectively.

Conclusions: Inner ear measurements in children with and without SNHL are not statistically different. Moreover, the measurements are difficult to interpret because while they demonstrate good reproducibility, they are susceptible to systematic biases. However, use of inner ear measurements is more sensitive in identifying vestibulocochlear dysplasias and should be considered to complement visual analysis.


Congenital sensorineural hearing loss (SNHL) affects 4 to 11 per 10,000 children in the United States. Proposed causes of congenital SNHL include developmental, structural, or functional abnormalities within the inner ear. Clinical characterization, however, cannot elucidate the primary site of abnormality. Approximately 25% of patients with congenital hearing loss will have bony inner ear malformations on computed tomography (CT) of the temporal bone. In the evaluation of SNHL in children, a temporal bone CT scan is obtained in addition to a thorough history taking and physical examination by the otolaryngologist. Although a temporal bone CT scan provides structural analysis of the bony labyrinth of the inner ear in addition to evaluating the external and middle ear, such severe malformations as complete labyrinthine aplasia, cochlear aplasia, and common cavity deformity that are typically identified by visual inspection alone represent only 1% of radiographic abnormalities found in patients with SNHL. Visual inspection is often inadequate for identification of subtle abnormalities such as lateral semicircular canal dysplasia and mild cochlear hypoplasia; up to one-third of these less severe malformations are missed by simple visual inspection.

The establishment of normative radiographic measurements has proved to be invaluable in the evaluation of disease processes in many parts of the body including that of the inner ear where the measure-
ment of the vestibular aqueduct is crucial in establishing the diagnosis of large vestibular aqueduct syndrome (LV A), which carries important clinical consequences. Normative radiographic measurements would be ideal for dimensions of inner ear structures because the inner ear labyrinth does not change in size after birth. This study sought to confirm the hypothesis that normative measurements for several inner ear structures can assist in the evaluation of SNHL in children.

The correct identification and precise measurement of these inner ear structures are essential criteria for their use as normative data. This investigation also assessed interobserver variability and evaluated the precision of these measurements by different observers. The clinical significance of the variability was also examined.

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**METHODS**

**PATIENTS**

A total of 204 patients (91 female and 113 male) underwent temporal bone CT scans or magnetic resonance imaging (MRI) as part of their routine radiologic evaluation for diagnoses including normal hearing with otologic symptoms, conductive hearing loss (CHL), and SNHL. Of note, 5 of the 204 patients underwent MRI for evaluation of SNHL. The radiologic evaluations were performed between 2001 and 2004 in patients whose age ranged from 2 months to 17 years.

**CT AND MRI OF THE TEMPORAL BONE**

**Computed Tomography**

All CT studies were performed at our institution using a standard temporal bone protocol with a Somatom Sensation 64 MultiDetector system (Siemens Medical Solutions, Erlangen, Germany). The nonhelical axial acquisition used exposure parameters, which vary with the age and size of the patient, but with continuous radiation monitoring. For example, a sample protocol for a 4-year-old used the following parameters: 120 kV; 240 mAs; matrix, 512 × 512; slice thickness, 1 mm; pitch, 0.9; and FOV, 135 m. Postprocessing with bone and soft tissue algorithms yields coronal reformats at 0.75- to 1.00-mm slice thickness.

**Magnetic Resonance Imaging**

Magnetic resonance imaging studies were performed at our institution using the 1.5 Tesla Signa Twin HDX system (General Electric Medical Systems, Milwaukee, Wisconsin). The protocol consists of a whole brain sagittal T1; axial T2; FLAIR (fluid-attenuated inversion recovery); GRE (gradient-recalled echo); DWI (diffusion-weighted imaging); and axial gadolinium-enhanced T1 images. Gadopentetate dimeglumine (Magnevist; Berlex Laboratories, Wayne, New Jersey) 0.2 mL/kg (maximum, 10 mL/kg) was administered intravenously. Additional high-resolution axial and coronal imaging for the temporal bone and cerebello-pontine angle portion are performed as follows:

1. Axial 3D FIESTA (fast imaging employing steady-state acquisition) or balanced steady-state free precession: TR, 9 milliseconds; TE, 3 milliseconds; 1.5 averaging; echo train length, 1; flip angle 45°; slice, 0.8 mm; gap, 0.4 mm; FOV 16 cm; matrix, 512 × 256.
2. Axial T1 (pregadolinium; and then, axial plus coronal T1 after gadolinium administration with fat suppression): TR, 600; TE, 20; 1.5 averaging; echo train length, 1; flip angle, 90°; slice, 2.0 mm; gap, 0.5 mm; FOV, 16 cm; and matrix, 320 × 220.

Studies were reviewed on General Electric Centricity PACS (Picture Archival and Communications System) terminals with magnification and electronic caliper capabilities.

**RESULTS**

**PATIENT POPULATION**

A total of 204 patients (408 ears) were included in this study, which included 188 ears with SNHL and 220 ears without SNHL. Radiologic evaluations of those patients without SNHL were performed for the following diagnoses: CHL, cholesteatoma, recurrent otitis media, microtia, atresia, mastoiditis, and cleft lip and palate.

**INNER EAR MEASUREMENTS**

Data from the patients without SNHL were used to generate comparative data with means and standard deviations for each of the measurements (Table 1). The differences between inner ear measurements in those ears with and without SNHL were not statistically significant (P > .05). The most common inner ear findings by report were LVA, dysplasia of vestibulocochlear structures, and internal auditory canal (IAC) abnormalities. Of the 220 ears without SNHL, inner ear malformations were identified in 11 ears based on visual inspection, as noted in the dictated radiology report. Of 188 ears with SNHL, 48 (26%) had inner ear malformation identified by visual inspection (Table 2). In contrast to
these qualitative data, quantitative data represented by measurements greater than 2 SDs did not appear to reliably differentiate between SNHL and non-SNHL ears and actually had lower sensitivity and specificity compared with visual inspection (13% vs 26%, respectively, and 92% vs 95%, respectively) (Table 2).

Of the ears without SNHL, 7 of 11 ears diagnosed as having CT abnormalities had abnormal measurements. A total of 198 ears had normal measurements and no abnormalities noted in the radiology report. There were 8 ears without SNHL specifically noted to have vestibulocochlear abnormalities; LSCC bony island measurements could not be obtained in 5 because there was no bony island owing to hypoplasia of the LSCC. Two ears had measurements greater than 2 SDs from the mean; the remaining ear had an LSCC measurement within 2 SDs from the mean. Of the 3 ears without SNHL that were specifically noted to have vestibular aqueduct abnormalities, none had abnormal measurements greater than 2 SDs from the mean. One ear with a narrow IAC, as noted in the report, had abnormal vestibulocochlear measurements, but on retrospective review, the ear was believed to have significant semicircular canal dysplasia that was not previously noted in the dictated radiology report.

Measurements from the patients with SNHL were compared with the data generated from those without SNHL. Of these patients with SNHL, inner ear malformations were identified in 48 ears based on visual inspection, as noted in the radiology report. Thirteen ears with SNHL had abnormal measurements and abnormalities noted in the radiology report. A total of 128 ears had normal measurements and no abnormalities noted in the radiology report.

Table 3 summarizes the measurements in the 48 ears with SNHL that were diagnosed as having inner ear anomalies. The anomalies were broken down into vestibulocochlear findings (ie, LSCC or SSCC hypoplasia, hypoplastic cochlea, or modiolus) vs other findings (ie, enlarged vestibular aqueduct, cochlear aqueduct, or IAC abnormalities). Looking at just the 19 ears that were specifically diagnosed as having vestibulocochlear abnormalities, abnormal measurements were obtained in 10 (53%), 5 of which had LSCC bony islands that could not be measured.

The primary abnormality noted by visual inspection was LVA, which was not one of the inner ear structures measured in this study. The major criterion for LVA is a midpoint vestibular aqueduct dimension of greater than 1.5 mm. In the present study, 14% (26 of 188) of ears with SNHL were noted to have LVA; only 1% of the ears without SNHL had LVA. All 29 of the ears that had some combination of vestibular aqueduct, cochlear

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**Table 1. Summary of Inner Ear Measurements**

<table>
<thead>
<tr>
<th>Reader</th>
<th>SNHL (188 Ears), Mean (SD), mm</th>
<th>Without SNHL (220 Ears), Mean (SD), mm</th>
<th>95% Confidence Interval</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reader A</td>
<td>BTC lumen 1.7 (0.2)</td>
<td>1.7 (0.2)</td>
<td>−0.071 to 0.025</td>
<td>.34</td>
</tr>
<tr>
<td></td>
<td>CH 6.1 (0.8)</td>
<td>6.2 (0.7)</td>
<td>−0.041 to 0.280</td>
<td>.15</td>
</tr>
<tr>
<td></td>
<td>LSCC bony island width 4.0 (0.5)</td>
<td>3.9 (0.6)</td>
<td>−0.170 to 0.052</td>
<td>.29</td>
</tr>
<tr>
<td></td>
<td>SSCC bony island width 5.5 (0.6)</td>
<td>5.4 (0.6)</td>
<td>−0.200 to 0.056</td>
<td>.27</td>
</tr>
<tr>
<td>Reader B</td>
<td>BTC lumen 1.8 (0.2)</td>
<td>1.8 (0.2)</td>
<td>−0.026 to 0.051</td>
<td>.48</td>
</tr>
<tr>
<td></td>
<td>CH 5.1 (0.5)</td>
<td>5.1 (0.4)</td>
<td>−0.055 to 0.130</td>
<td>.41</td>
</tr>
<tr>
<td></td>
<td>LSCC bony island width 3.7 (0.6)</td>
<td>3.6 (0.5)</td>
<td>−0.160 to 0.053</td>
<td>.29</td>
</tr>
<tr>
<td></td>
<td>SSCC bony island width 5.1 (0.6)</td>
<td>5.1 (0.6)</td>
<td>−0.170 to 0.080</td>
<td>.45</td>
</tr>
</tbody>
</table>

Abbreviations: BTC, basal turn of cochlea; CH, cochlear height; LSCC, lateral semicircular canal; SNHL, sensorineural hearing loss; and SSCC, superior semicircular canal.

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**Table 2. Computed Tomographic Readings by Visual Inspection and Inner Ear Measurements Greater Than 2 SDs**

<table>
<thead>
<tr>
<th>Variable</th>
<th>SNHL, No. (188 Ears)</th>
<th>No SNHL, No. (220 Ears)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT Readings by Visual Inspection</td>
<td>Abnormal CT reading 48</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td>Normal CT reading 140</td>
<td>209</td>
</tr>
<tr>
<td>&gt;2 SD measurementb</td>
<td>24</td>
<td>17</td>
</tr>
<tr>
<td>&gt;2 SDb</td>
<td>9</td>
<td>10</td>
</tr>
<tr>
<td>BTC lumen</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>CH</td>
<td>12</td>
<td>6</td>
</tr>
<tr>
<td>LSCC bony island width</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td>SSCC bony island width</td>
<td>164</td>
<td>203</td>
</tr>
</tbody>
</table>

Abbreviations: BTC, basal turn of cochlea; CH, cochlear height; CT, computed tomography; LSCC, lateral semicircular canal; SNHL, sensorineural hearing loss; and SSCC, superior semicircular canal.

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**Table 3. Measurements in Ears With Sensorineural Hearing Loss, With Inner Ear Anomalies Noted in the Radiology Report**

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Vestibulocochlear Findings, No.</th>
<th>Other Findings, No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abnormal measurement</td>
<td>10</td>
<td>0</td>
</tr>
<tr>
<td>Normal measurement</td>
<td>9</td>
<td>29</td>
</tr>
<tr>
<td>Total</td>
<td>19</td>
<td>29</td>
</tr>
</tbody>
</table>

Abbreviations: BTC, basal turn of cochlea; CH, cochlear height; CT, computed tomography; LSCC, lateral semicircular canal; SNHL, sensorineural hearing loss; and SSCC, superior semicircular canal.
aqueduct, or IAC abnormality but no diagnoses of vestibulocochlear abnormality had normal measurements (Table 3).

There were 24 ears (13 with SNHL and 11 without SNHL) noted to have abnormal inner ear measurements but no abnormalities dictated in the radiology report. On further careful review, 11 of the 13 ears with SNHL were believed to have vestibulocochlear abnormalities that were not dictated in the radiology report. These included mild hypoplasia of the LSCC or SSCC, patulous vestibule, and cochlear hypoplasia. Also on further careful review, 4 of the 11 ears without SNHL were believed to have minor abnormalities limited to the vestibule and semicircular canals.

INTEROBSERVER VARIABILITY

Intraclass correlation coefficients produce measures of consistency or agreement of values within cases. As a reference, an ICC less than 0.4 indicates poor reproducibility; an ICC less than 0.75 and greater than 0.4 suggests fair to good reproducibility; and an ICC greater than 0.75 suggests excellent reproducibility.8 The ICC was 0.612 for BTC lumen measurements, 0.632 for CH measurements, 0.869 for LSCC bony island width measurements, and 0.912 for SSCC bony island width measurements.

The Bland-Altman plot is a statistical measure of 2 measurement techniques in which the differences or ratios between the 2 techniques are plotted against the means of the 2 techniques. It is useful to reveal a relationship between the differences and the averages and to look for any systematic biases.9 Bland-Altman plots were generated for each of the 4 measurements described in this article, with the vertical axis represented by the ratio of reader A to reader B measurements and the horizontal axis represented by the mean of the 2 measurements (Figure 1). With respect to measurement of the SSCC bony island width, on average, reader A read a length 8% greater than that read by reader B, with a mean ratio of 1.08. The 95% confidence interval (CI) for this mean ratio was between 1.07 and 1.09, suggesting that this difference is significant because the CI does not span 1.0. The mean ratio of the reader A to reader B CH measurement was 1.21 (95% CI, 1.20-1.23). On average, reader A read a height 21% greater than that read by reader B, and this finding is significant. For the measurement of the LSCC bony island width, the mean ratio was 1.10 (93%
COMMENT

One goal of this study was to assess whether inner ear measurements could be reliably obtained from CT. We hypothesized that these data would provide a useful aid in the interpretation of temporal bone CT scans obtained for evaluation of SNHL. Comparative data in 220 ears without SNHL were acquired in this study for 4 dimensions of the inner ear found in previous studies to be most useful—namely,BTC lumen, LSCC bony island width, SSCC bony island width, and CH.2,7

Although inner ear malformations of the bony labyrinth are reportedly present in 20% to 25% of patients with congenital SNHL undergoing temporal bone CT and the cause of the hearing loss is typically attributed to the morphogenetic abnormality, in this study, 26% of ears with SNHL had inner ear malformations noted by initial visual inspection of the CT scan. It is assumed that 75% to 80% of patients with SNHL for which inner ear measurements and visual inspection are normal have abnormalities of the membranous labyrinth or anomalies beyond the resolution of technology used in studying these children. Subtle abnormalities of the inner ear such as semicircular canal dysplasia and cochlear hypoplasia, which account for the majority of inner ear malformations associated with SNHL, can be missed on temporal bone CT.7 In the present study, an additional 7% (13 of 188) of ears were diagnosed as having vestibulocochlear abnormalities when using a measurement criterion of greater than 2 SDs from the mean. The use of these measurements resulted in a 32% sensitivity (compared with 26% sensitivity with visual inspection alone).

The most common inner ear abnormalities involve malformations of the vestibular aqueduct, lateral semicircular canal, and the cochlea. The distribution of common CT findings and inner ear measurements greater than 2 SDs reported in this study is in agreement with the aforementioned statement. The mechanism of labyrinthine dysplasia involves lack of formation of the central bony island, which should be easily seen and measured on an axial temporal bone CT scan. The LSCC is a frequently malformed inner ear structure likely to occur in isolation owing to its late stage of formation compared with the SSCC, which is the first to form, followed by the posterior semicircular canal. Any abnormality of the SSCC is likely to result in significant LSCC abnormality due to this sequence of formation. Not surprisingly, Purcell et al10 have suggested the LSCC measurement to be more sensitive than the SSCC measurement. Lateral semicircular canal dysplasia is associated with CHL, mixed HL, and SNHL.13,14 In the series by Johnson and Lalwani2, 1-4% of ears with LSCC malformation had CHL or mixed hearing loss.3 In the present series, 8 ears demonstrated labyrinthine anomalies without SNHL but instead had CHL or normal hearing.

Labyrinthine anomalies with normal cochlear function have been described.13 Although inner ear embryogenesis with the pars superior being phylogenetically older than the pars inferior would suggest that labyrinthine anomalies with a morphologically normal cochlea cannot be easily explained by a very early, nonspecific developmental insult; an insult to the inner ear between weeks 8 and 20 may interrupt formation of the membranous SCCs while sparing the membranous cochlear duct.12 Classification based on embryogenesis reveals that in vestibule-LSCC dysplasia with a normal cochlea, there were normal or slightly impaired hearing levels, with a mean hearing level of 35 dB.4 Abnormal measurements of the bony island of LSCC is not always associated with SNHL, as is demonstrated in this study as well as others.10 Cochlear malformation is typically identified as less than 2 1/2 turns as a result of early development arrest and thus is expected to correspond with a decreased base-to-apex height of the cochlea best assessed on the coronal CT scan.7 More specifically, arrested development of the cochlear during the sixth gestational week would result in a rudimentary cochlea representing various degrees of cochlear hypoplasia. Arrest during the seventh gestational week typically results in the classic Mondini dysplasia, which has been described radiographically as a small cochlea with an incomplete intracochlear partition.4 However, incomplete partitions may be associated with normal dimensions. As described by Sennaroglu and Saatci,13 there can be incomplete partitions associated with both normal dimensions and dimensions less than normal; it is believed that the dimension of the pathologic region is less important than its actual differentiation. In the present study, 7 ears with SNHL had a decreased CH measurement, with an otherwise normal radiology report, and 10 ears with SNHL had abnormal cochlear differentiation (ie, deficient modiolus) noted in the radiology report but had normal dimensions by inner ear measurement. The significance of this would appear to concur with the belief that dimension is less important than actual differentiation because no statistically significant difference was observed between inner ear measurements in ears with and without SNHL.

It should be noted that 6 radiology reports for ears with SNHL described some dysplasia of vestibulocochlear structures, the most common being hypoplastic modiolus, without associated abnormal inner ear measurements. Interestingly, the aforementioned 6 cases also noted LVA, for which there is literature noting the association of these 2 findings.14,15 Modiolar deficiency has also been proposed as representing the mildest form of cochlear dysplasia.16 With respect to interobserver variability, the most fundamental interpretation of an ICC is that it measures the proportion of a variance that is attributable to the objects of measurement.17 Although a higher ICC suggests greater reproducibility, it does not rule out the possibility that the reproducibility incorporates a consistent discrepancy as evidenced by the Bland-Altman calculations. Although the mean difference in measurement of the BTC lumen is less than that of CH measurements per the Bland-Altman plot, the ICC is higher for CH measurements because there is a significant and consistent difference between the measurements of the 2 readers. Furthermore, the Bland-Altman plots do not suggest that variation is dependent on the magnitude of the measurement.
It appears that measurements of cochlea height, LSCC bony island width, and SSCC bony island width demonstrate good reproducibility with the caveat that for each, there was a consistent but reproducible difference between readers by 21%, 10%, and 8%, respectively. The BTC lumen measurement was not found to be clinically useful in this study. As can be gleaned from Table 1, the standard deviation of the BTC lumen is only 0.2 mm, making it difficult to use this value to differentiate between ears with and without SNHL. The ICC interpretations in this study are congruent with what is reported by Purcell et al. The data in this study more importantly illustrate that regardless of the variability, the conclusion from the measurements remains the same. From a clinical standpoint, the variability does not change whether the measurements are useful in evaluating SNHL if the same observer is performing the measurements.

The standard deviations for each reader for each measurement are similar, suggesting that each reader is consistent in their own method of obtaining the measurements, with the exception of CH. While each reader’s measurement for a given inner ear structure may differ, their patterns of measurement may be similar, and the ICC tests this pattern of measurement. With the combination of ICC and Bland-Altman plot, interobserver variability is characterized both by the extent of difference between readers and consistency in the pattern of measurement.

Nevertheless, it should be noted that variability is greatly influenced by the interpretation of how the measurement should be made (plane, axis, or limits of lumen or bony island). When CT images are magnified, the transition between bone vs lumen is less distinct and caliper placement may be ambiguous, leading to different measurements (Figure 2). Moreover, a CH measurement was defined as a measurement of the maximum height that includes basal and upper turn. On further review of the literature, it is more specifically a measurement taken perpendicular to the axes of the cochlear lumen, which may not necessarily be the maximum height that includes the basal and upper turn (Figure 3). This, in fact, is a limitation of the study because this variability may have been decreased by further education, review, and agreement on the method of obtaining measurements. Reported CH measurements to define hypoplastic and hyperplastic cochlea (<4.4 mm and >5.9 mm, respectively) and LSCC bony island (<2.6 mm and >4.8 mm, respectively) do not differ significantly from what reader B would conclude (<4.3 mm and >5.9 mm, respectively, and <2.6 mm and >4.6 mm, respectively).

In conclusion, inner ear measurements did not appear to reliably differentiate between SNHL and non-SNHL ears but can result in a slight improvement in diagnostic yield when evaluating those ears with and without SNHL. The use of inner ear measurements may help in identifying vestibulocochlear dysplasias that otherwise may be missed and thus should be considered to complement visual analysis of temporal CT scans. It is also important to acknowledge the variability associated with these measurements.

Submitted for Publication: March 12, 2007; final revision received August 24, 2007; accepted September 7, 2007.
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Author Contributions: Dr Chang had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Barnes and Chang. Acquisition of data: Gittleman, Barnes, and Chang. Analysis and interpretation of data: Chen, Gittleman, and Chang. Drafting of the manuscript: Chen, Gittleman, and Chang. Critical revision of the manuscript for important intellectual content: Barnes and Chang. Statistical analysis: Chen and Gittleman. Administrative, technical, and material support: Gittleman, Barnes, and Chang. Study supervision: Barnes and Chang.

Financial Disclosure: None reported.

Previous Presentation: This article was presented at The American Society of Pediatric Otolaryngology 2007 Annual Meeting; April 29, 2007; San Diego, California.

REFERENCES