Major and Minor Temporal Bone Abnormalities in Children With and Without Congenital Sensorineural Hearing Loss

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Objective: To determine the extent of correlation between sensorineural hearing loss (SNHL) and abnormal temporal bone anatomy in children.

Design: Axial and coronal high-resolution computed tomographic scans of the temporal bones of 247 children (494 ears) aged 2 months to 15 years with and without SNHL were blindly reviewed. The presence or absence of mild or severe cochlear dysplasias, vestibular dysplasias, and an enlarged vestibular aqueduct (VA) were recorded. The width of the VA was measured. The height, width, and length of the internal auditory canal (IAC) were measured, and abnormalities were described as narrow, widened, or bulbous. Clinical information was then reviewed to determine the presence or absence of a congenital syndrome and/or SNHL, and historical factors that might be responsible for SNHL.

Main Outcome Measure: The relationship between radiographic findings and SNHL.

Results: One hundred thirteen patients (185 ears) had SNHL. Significant abnormal temporal bone anatomy in children with vs without SNHL included major cochlear and vestibular dysplasias (17% vs 0%; \(P < .001\)), enlarged VA (>2 mm) (5% vs 0%; \(P < .001\)), and narrow IAC (≤2 mm) (4% vs 1%; \(P = .03\)). The average IAC width (4.85 vs 5.02 mm), height (4.39 vs 4.62 mm), and length (11.22 vs 11.44 mm) were not statistically different between children with vs without SNHL. In children with vs without SNHL, neither a widened (0.9% vs 3.6%) nor a bulbous (9% vs 8%) IAC was seen more often in children with SNHL. In ears with SNHL, the presence of a congenital syndrome significantly increased the risk of cochlear and vestibular abnormalities of the temporal bone (45% vs 14%; \(P < .001\)), including IAC abnormalities (30% vs 2%; \(P < .001\)), which overall were more commonly seen in children with (20%) vs without (3%) a congenital syndrome regardless of the presence of SNHL. No children with an enlarged VA had a congenital syndrome.

Conclusions: Well-established temporal bone abnormalities such as cochlear and vestibular abnormalities and a grossly enlarged vestibular aqueduct are significantly found in children with SNHL. A narrow IAC is found more often in children with vs without SNHL. No significant correlation is found between SNHL and radiographic findings of a widened or bulbous IAC. In children with a congenital syndrome, more IAC abnormalities were seen, regardless of the presence of SNHL. In children with SNHL, the presence of a congenital syndrome increases the likelihood of a cochlear or vestibular abnormality.

PATIENTS, MATERIALS, AND METHODS

METHODS

Five hundred twenty-five children underwent thin-cut high-resolution CT of the temporal bone between January 1, 1991, and December 31, 1995, at the Childrens National Medical Center in Washington, DC, for various abnormalities related to the outer, middle, and inner ear. These abnormalities included congenital SNHL, chronic otomastoiditis, cholesteatoma, facial nerve paralysis or paresis, temporal bone trauma, conductive hearing loss, middle ear mass, or congenital anatomic abnormalities of the middle or external ear. Computed tomographic scans were obtained using a scanner unit (GE 9800-High Q CT/T; General Electric, Milwaukee, Wis). Thin sections in 1.5-mm contiguous increments were performed. Both axial and coronal images were obtained in most all patients. All of the images were reconstructed using a standard bone algorithm program (General Electric).

Of the 525 CT scans performed, 275 (550 ears) were randomly selected and retrospectively and blindly reviewed. The width, height, and length of the IAC and the width of the VA were measured using a transparent ruler derived from the scale on each CT scan and accurate to 0.25 mm. The width of the IAC was measured in the axial plane using a perpendicular line beginning on the posterior wall of the IAC, 2 mm inside the posterior lip of the internal auditory meatus, ending on the anterior canal wall (Figure 1). This technique eliminated a false exaggeration of the width of the internal auditory meatus in a broad-mouthed porus acusticus.

The height and length were measured in the coronal planes. The height of the IAC was measured in the middle portion of the canal, where it appeared at maximum height (Figure 2). The length was measured from the base of the crista falciformis (falciform crest) to the middle portion of the inferior and superior lip (Figure 2). The VA was measured on the axial views in the midportion of the bony canal (Figure 1).

The medical records of all children who had their scans blindly reviewed were then evaluated to determine the presence or absence of SNHL and contributing factors for SNHL, such as family history of hearing loss, perinatal infections, complications at birth, ototoxic drug intake, trauma, major illness, syndromes present, and otologic surgical history. In addition, the audiologic history and physical examination findings were reviewed. All patients had hearing evaluations by a pediatric audiologist. Individual ear testing was performed using either auditory brainstem response or pure-tone air and bone conduction thresholds of 250 to 8000 Hz depending on age and neurologic status. Any child having (1) insufficient medical or audiologic records for review, (2) a possible medical reason for SNHL, or (3) a previous trauma resulting in SNHL or temporal bone fractures was excluded.

STATISTICAL ANALYSIS

IAC Measurements

One-way analysis of variance was run for each of the 3 dependent measures of IAC—width, length, and height—to detect differences between IAC size in children with and without SNHL. Because 3 separate analyses of variance were conducted, the $\alpha$ level of .05 was adjusted using the Bonferroni correction. The Bonferroni-corrected $\alpha$ level was determined by dividing the .05 $\alpha$ level by 3 (the number of tests). The resulting $\alpha$ level was .017.

Comparison of Abnormalities

$\chi^2$ Tests of independence were used to examine the relationship between SNHL and IAC size and the presence of cochleovestibular anomalies in the presence or absence of congenital syndromes.

Figure 1. Axial computed tomographic scan showing internal auditory canal (IAC) and vestibular aqueduct (VA) width measurements.

Figure 2. Coronal computed tomographic scan showing internal auditory canal height and length measurements.
RESULTS

Two hundred forty-seven children (494 ears) aged 2 months to 15 years met the inclusion criteria for the study. There were 132 boys and 115 girls (male-female ratio, 1.15:1). Of the 247 children, 113 (185 ears) had SNHL (72 bilateral and 41 unilateral). One hundred thirty-five patients had no SNHL bilaterally and 39 had no SNHL unilaterally, for a total of 309 ears without SNHL. Thirty children had a congenital syndrome. Of these 60 ears, 20 had SNHL and 40 had no SNHL.

INTERNAL AUDITORY CANAL

There was no statistical difference between the mean ± SD width, height, and length of the IAC in temporal bones of children with and without SNHL (Table 1). However, the presence of a narrow IAC (<2 mm) in either the axial plane (Figure 3) or the coronal plane (Figure 4) was a statistically significant finding in the temporal bones of children with vs without SNHL (8 of 185 ears vs 4 of 309 ears; \( \chi^2 = 4.482; P = .03 \)) (Table 2). There was no increase in the presence of a widened IAC (>8 mm) in children with vs without SNHL (Figure 5). In fact, ears without SNHL (11 of 309) more often had an abnormally wide IAC than did ears with SNHL (1 of 185) (\( \chi^2 = 2.946; P = .23 \)) (Table 2).

Temporal bones of children with a congenital syndrome statistically had more abnormally wide or narrow IACs (12 of 60 ears) than did temporal bones of children without congenital syndromes (15 of 434 ears), regardless of whether SNHL was present (\( \chi^2 = 7.836; P = .006 \)).

Table 1. Internal Auditory Canal (IAC) Width, Height, and Length for Children With and Without Sensorineural Hearing Loss (SNHL)

<table>
<thead>
<tr>
<th>Variable</th>
<th>SNHL</th>
<th>Ears, No. *</th>
<th>Mean ± SD</th>
<th>Median</th>
<th>Mode</th>
<th>Minimum</th>
<th>Maximum</th>
</tr>
</thead>
<tbody>
<tr>
<td>IAC width, mm</td>
<td>Yes</td>
<td>181</td>
<td>4.85 ± 1.07</td>
<td>4.8</td>
<td>4.5</td>
<td>2.0</td>
<td>8.0</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td>307</td>
<td>5.02 ± 1.22</td>
<td>5.0</td>
<td>5.0</td>
<td>2.0</td>
<td>10.0</td>
</tr>
<tr>
<td>IAC length, mm</td>
<td>Yes</td>
<td>176</td>
<td>11.22 ± 2.22</td>
<td>11.0</td>
<td>10.0</td>
<td>6.0</td>
<td>18.0</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td>292</td>
<td>11.44 ± 2.33</td>
<td>11.4</td>
<td>10.0</td>
<td>4.0</td>
<td>18.0</td>
</tr>
<tr>
<td>IAC height, mm</td>
<td>Yes</td>
<td>175</td>
<td>4.39 ± 1.18</td>
<td>4.2</td>
<td>5.0</td>
<td>1.5</td>
<td>8.0</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td>292</td>
<td>4.62 ± 1.16</td>
<td>4.5</td>
<td>4.0</td>
<td>2.0</td>
<td>8.0</td>
</tr>
</tbody>
</table>

*Slightly varying numbers are seen because computed tomographic scans were not performed on all the planes (axial and coronal).

Table 2. Internal Auditory Canal Abnormalities in Children With and Without Congenital Syndromes*

<table>
<thead>
<tr>
<th>SNHL</th>
<th>Width, mm</th>
<th>Height, mm</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&lt;=2</td>
<td>&gt;2</td>
</tr>
<tr>
<td>With Congenital Syndromes</td>
<td>Yes</td>
<td>20</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td>40</td>
</tr>
<tr>
<td>Without Congenital Syndromes</td>
<td>Yes</td>
<td>165</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td>269</td>
</tr>
</tbody>
</table>

*Data are given as number of ears except where otherwise indicated.
However, children with SNHL and a congenital syndrome had the highest rate of IAC abnormalities (Table 2). The congenital syndromes encountered are listed in Table 3.

The shape of the IAC was considered to be bulbous in 40 temporal bones, being found in equal amounts in the temporal bones of children with (16 of 185 ears) and without (24 of 309 ears) SNHL ($P=.75$) (Figures 10, 11, and 12). The width and height dimensions of the IAC in boys were significantly greater than those in girls independent of hearing status ($P<.001$). However, there was no interaction between sex and the presence or absence of SNHL. An interaction would suggest that either boys or girls were more susceptible to SNHL.

Table 3. Congenital Syndromes Seen in the Study

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Ears, No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Goldenhar (facioariculovertebral)</td>
<td>9</td>
</tr>
<tr>
<td>Down</td>
<td>8</td>
</tr>
<tr>
<td>Branchio-oto-renal</td>
<td>2</td>
</tr>
<tr>
<td>CHARGE</td>
<td>2</td>
</tr>
<tr>
<td>Waardenburg</td>
<td>2</td>
</tr>
<tr>
<td>Duane</td>
<td>2</td>
</tr>
<tr>
<td>Cleft lip/cleft palate</td>
<td>2</td>
</tr>
<tr>
<td>Treacher Collins</td>
<td>1</td>
</tr>
<tr>
<td>Klippel-Feil</td>
<td>1</td>
</tr>
<tr>
<td>Paterson-Kelly</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>30</td>
</tr>
</tbody>
</table>


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VESTIBULAR AQUEDUCT

In all ears without SNHL (n=309), the VA width was 2 mm or less. None of these children had vestibular symptoms. In 19 of these ears, the diameter of the VA ranged from 1.5 to 2.0 mm (Figure 13). The width of the VA in temporal bones of ears with SNHL (n=185) was up to 7 mm (Figure 14). In ears with SNHL, 9 temporal bones had a VA greater than 2 mm in diameter and 9 had a VA measuring 1.5 to 2.0 mm. Comparing the number of temporal bones with a VA measuring 1.5 to 2.0 mm in ears of children with SNHL (9 of 185 ears) vs without SNHL (19 of 309 ears), no difference was seen. No patient with a congenital syndrome had an enlarged VA.

COCHLEOVESTIBULAR ABNORMALITIES

Thirty-two (17%) of 185 ears with SNHL had cochleovestibular abnormalities. Nine (45%) of 20 syndromic ears had cochlear or vestibular dysplasias, which is significantly more than the 23 cochleovestibular abnormalities (14%) found in the 165 patients with SNHL but without a syndrome ($\chi^2=30.02; P<.001$) (Table 4).

Table 4. Cochlear and Vestibular Abnormalities in Ears of Children With Sensorineural Hearing Loss and With and Without a Congenital Syndrome*

<table>
<thead>
<tr>
<th>Abnormality</th>
<th>Without a Congenital Syndrome (n = 165)</th>
<th>With a Congenital Syndrome (n = 20)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolated cochlear</td>
<td>14 (9)</td>
<td>2 (10)</td>
</tr>
<tr>
<td>Isolated vestibular</td>
<td>7 (4)</td>
<td>2 (10)</td>
</tr>
<tr>
<td>Cochlear and vestibular</td>
<td>2 (1)</td>
<td>5 (25)</td>
</tr>
<tr>
<td>Total</td>
<td>23 (14)</td>
<td>9 (45)</td>
</tr>
</tbody>
</table>

*Data are given as the number (percentage) of ears.

INTERNAL AUDITORY CANAL

The size and shape of the IAC has been studied anatomically, histologically, and radiographically. Many of the early investigators (Ebenius,6 Camp and Cilley,7 and Graf8) took measurements from plain-film polytomography of dried temporal bones, both paired and unpaired. Olivares and Schuknecht,9 as well as Graf,9 measured the canal from histologic sections. Papangelou10 and Amjad et al,11 in the 1960s and 1970s, used silicone rubber casts of dried paired and unpaired IACs to evaluate their size and shape. Finally, the IAC of the temporal bones of large
groups of people were measured from images taken with plain-film polytomography\textsuperscript{8,12} and CT.\textsuperscript{5,13}

Different investigators measured 1 or all of the dimensions of the IAC—height, width, length, or volume. The length often was measured mainly from the posterior and anterior walls of the IAC in the axial plane but occasionally from the floor and roof in the coronal plane. Diverse values for normal-sized IACs have been obtained, but most dimension averages are fairly congruent (Table 5). However, the ranges of normal are occasionally 1 to 2 mm different. This variation is significant when trying to determine what is truly abnormally wide or narrow.

When evaluating for abnormalities of the IAC on CT, some researchers\textsuperscript{17} used previously reported data to obtain their control population and others\textsuperscript{4} used control populations evaluated at their institution. We too used an internal control population for better comparison (the technology used to obtain the CT scan, the technique used to reconstruct the image, and the method of measurement was the same between the 2 groups). Masking the reviewer to the clinical history of the child prevented measurement bias, although visualization of gross abnormalities of the cochlea, vestibule, and VA could not be avoided. Gross abnormalities of the temporal bone are widely believed to be significantly found in children with vs without SNHL and were not the true focus of the study.

Similar to other studies,\textsuperscript{4,9} we found no difference in the average width, length, or height of the IAC in children with vs without SNHL. Similarly, the mean ± SD and mode were similar between the 2 groups (Table 1). Our measurements are similar to previous measurements, especially when the wide range of values is included (Table 5).

More important, the abnormally narrow or wide IACs were compared between patients with and without SNHL. An abnormally narrow or wide IAC was determined through literature review and review of our own data. For the limit of the narrow IAC, we not only looked at normative data seen in large reviews and our own averages and SDs but also at smaller reviews and case reports that correlated abnormal size with pathologic findings. Shelton et al,\textsuperscript{14} Gray et al,\textsuperscript{15} and Casselman et al\textsuperscript{16} showed that an IAC width of 1 to 2 mm could be associated with an absent or underdeveloped vestibulocochlear nerve. We chose 2 mm or less in height or width as abnormal because true pathologic findings have been seen with these values.

We found that an abnormally narrow IAC occurs statistically more often in ears with SNHL than in those without SNHL in all groups (Table 2). In children with a congenital syndrome, the difference was even greater evaluating either all 494 ears (8 of 60 syndromic ears vs 4 of 434 nonsyndromic ears; \(P<.001\)) or only ears with SNHL (6 of 20 syndromic SNHL ears vs 2 of 165 nonsyndromic SNHL ears; \(P<.001\)). The group with the largest percentage of narrow IACs comprised children with a congenital syndrome and SNHL.

Nonetheless, a narrow IAC was seen in children with normal hearing. Thus, one should not automatically assume that the vestibulocochlear nerve is absent on finding an IAC measuring 2 mm or less in height or width on CT scans, as some researchers\textsuperscript{14,15,37} have suggested. With the advent of improved resolution of the vestibulocochlear nerve on magnetic resonance imaging,\textsuperscript{10} a magnetic resonance image should be obtained before finalizing a definitive diagnosis of an absent or atretic vestibulocochlear nerve. In fact, Casselman et al\textsuperscript{16} point out that an absent eighth nerve is seen by magnetic resonance imaging in patients with normal-sized IACs. With its increased resolution and advanced reconstructive techniques, magnetic resonance imaging may supplant high-

\begin{table}[h]
\centering
\begin{tabular}{|l|c|c|c|c|c|c|}
\hline
\textbf{Study, Year} & \textbf{Evaluation Method} & \textbf{History} & \textbf{Numbers} & \multicolumn{3}{c|}{\textbf{Mean (Range), mm}} \\
\hline
\textbf{} & \textbf{} & \textbf{} & \textbf{} & \textbf{Width} & \textbf{Height} & \textbf{Length} \\
\hline
Ebenius,\textsuperscript{+} 1934 & Polytom & Unknown & 100 Skulls & 5.7 (4-8) & 6.3 (5-9) & ND \\
Camp and Cilley,\textsuperscript{7} 1939 & Polytom & Unknown & 500 Paired temp bones & ND & 5.2 (2-11) & 7.8 (3-16) \\
Graf,\textsuperscript{8} 1952 & Polytom & Unknown & 100 Living patients & ND & 4.9 (3-7) & ND \\
Dry bones & Unknown & 60 Temp bones & 4.0 (3-5) & 4.2 (3-6) & 8.0 (5-11) \\
Valvassori and Pierce,\textsuperscript{9} 1964 & Polytom & 125 Otosclerosis, 175 other conditions & 300 Living patients (200 paired) & ND & 4 (2-8) & 8 (4-11) pw \\
Amjad et al,\textsuperscript{10} 1969 & Casts & Cadavers (unknown) & 30 Paired temp bones & ND & 5.9 (4-8) & 9.9 (3-13) pw \\
Papangelou,\textsuperscript{10} 1972 & Casts & Unknown† & 242 Human paired temp bones & 4.5 (3-7) & 4.7 (2-7) & 9.2 (6-14) pw \\
Olivares and Schunknecht,\textsuperscript{11} 1979 & Histo & Unknown & 435 Patients, 144 with presbycusis & 3.7 (2-5) & 3.7 (2-6) & ND \\
Pappas et al,\textsuperscript{4} 1990 & CT & No SNHL & 25 Living patients (50 temp bones) & 5.3 (4-8) & 6.1 (4-9) & 9.5 (7-13) \\
SNHL with obv abnorm & 70 Temp bones in living patients & 6.3 (2-12) & 5.3 (3-12) & 10.8 (5-13) \\
SNHL & 140 Temp bones in living patients & 7 (2-7) & 5 (4-6) & 11 (7-17) \\
Present study & CT & No SNHL & 309 Temp bones in living patients & 5.0 (2-10) & 4.6 (2-8) & 11.4 (4-18) \\
SNHL & 185 Temp bones in living patients & 4.8 (2-8) & 4.4 (2-8) & 11.2 (6-18) \\
\hline
\footnotesize{*Polytom indicates polytomography; temp, temporal; ND, not determined; histo, histologic section analysis; pw, posterior wall measured; aw, anterior wall measured; CT, computed tomography; SNHL, sensorineural hearing loss; and obv abnorm, obvious abnormalities (inner ear on CT scan).} \\
\footnotesize{†”An unselected population of sudden or accidental deaths, but many were from hospitals and clinics.”} \\
\end{tabular}
\caption{Reported Studies Measuring Internal Auditory Canal Dimensions\textsuperscript{*}}
\end{table}
resolution CT as the imaging modality of choice in evaluating children with SNHL.

Eight millimeters or greater has been used to signify an abnormally wide IAC by several researchers.5,13 Most of the normal values seen in previous studies measuring the IAC have their upper limit of normal at 8 mm5,6,8 or less.9,10 In our study, only 1 of 185 ears of children with SNHL had an IAC with a width of 8 mm or greater. Eleven of 309 ears of children without SNHL had an IAC measurement of 8 mm or greater. Based on our data, it seems that a widened IAC is not a significant finding in children with vs without SNHL. Birman and Gibson17 reported that a widened IAC may be associated with a lateral fundus partial dehiscence (and other temporal bone abnormalities), increasing the communication between the IAC and the inner ear and potentially heralding a progressive or fluctuating SNHL. An abnormal partition between the IAC and the cochlea and vestibule may cause abnormal pressures on the inner ear. However, the mere presence of a widened IAC may not hold the same significance. Weinberg et al18 and Tomura and colleagues20 reported to be symmetrical between the 2 sides 40% to 60% of the time. Early studies7,8,12 describe the shape of the IAC as either straight (cylindrical), wider at the medial end, wider at the lateral end, or oval, which describes a tube with the center wider than the ends. The shape of the IAC has been reported to be symmetrical between the 2 sides in 40% to 90% of the time. In addition, the most common shape of the IAC is straight, reported 35% to 60% of the time.

Oval IACs have accounted for 13% to 19% of the total in previous studies.7,10,12 We found that 40 (8%) of 494 ears had not only an oval IAC but also varying degrees of a bulbous shape (Figures 10-12). The incidence of a bulbous IAC in our study was equal in children with and without SNHL. There was no difference in the number of children in either group with an isolated abnormally wide or bulbous IAC, making clinical correlation with these abnormalities alone circumspect.

Children with congenital syndromes compared with those without congenital syndromes more commonly have abnormalities of the IAC, regardless of the presence of SNHL. Children with congenital syndromes and SNHL compared with those with SNHL and no congenital syndromes more often have cochleovestibular abnormalities.
REFERENCES


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