Audiometric Findings in Children With a Large Vestibular Aqueduct

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Objective: To characterize audiometric findings in children with a large vestibular aqueduct (LVA).

Design: Retrospective review.

Methods: Audiometric records of children with an isolated LVA, diagnosed by computed tomography of the temporal bone, from 1995 through 1998 were reviewed.

Results: Nineteen children had an isolated LVA in one or both ears. In all, 26 ears with an isolated LVA were identified. An LVA was seen in association with another inner ear anomaly in an additional 7 ears. The hearing impairment was sensorineural in 22 ears (85%) with an isolated LVA and mixed in 3 (12%). The hearing was normal in 1 ear. The sensorineural hearing impairment (SNHI) was moderate-severe in 12 ears (46%) and severe-profound in 10 ears (38%). Thirteen (50%) of 26 ears had a downsloping or high-frequency SNHI, and 8 (31%) of 26 ears had a midfrequency-peaked audiogram. Bilateral LVAs were seen in 7 children, 6 of whom had bilateral and asymmetrical SNHI. Of 12 patients with a unilateral LVA, 5 had bilateral SNHI.

Conclusions: In this series, the children with LVAs typically had moderate-severe or worse SNHI. An unusual midfrequency-peaked audiogram was present in approximately one third of the study patients. The majority of the patients had a unilateral LVA; however, nearly 50% of the patients with a unilateral LVA had bilateral SNHI. The patients with bilateral LVAs generally had asymmetrical SNHI.

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The large vestibular aqueduct (LVA) is reported to be the most common inner ear anomaly identified on computed tomograms (CTs) in children with sensorineural hearing impairment (SNHI). As such, hearing impairment is frequently attributed to the presence of an LVA. The description of LVA syndrome in 1978, which included associated inner ear anomalies, led to an interest in the LVA as an isolated anomaly. However, associated cochlear anomalies are thought to be more difficult than LVAs to detect on CTs.

Radiographic criteria for the diagnosis of LVAs have yet to be standardized. The definition of a normal vestibular aqueduct (VA) diameter varies considerably in published reports, as do the place of measurement and the plane of CT scans. The clinical presentation of the LVA is variable; both stable and progressive SNHI have been described, and the characteristics of hearing impairment (severity, type, and audiometric configuration) vary as well. Vestibular symptoms have also been reported by some patients.

The mechanism of hearing impairment associated with an LVA is unknown, although many hypotheses have been presented. Treatment of an LVA remains controversial. The purpose of this study is to examine the audiometric findings in children with isolated enlargement of the VA as identified on temporal bone CTs. The characteristics of hearing impairment (eg, severity and audiometric configuration) are described, as is the relationship between VA findings and hearing status.

METHODS

We performed a retrospective review of the medical records of children with SNHI who underwent temporal bone CT scans from 1995 through 1998. All children in this study were seen in the pediatric otolaryngology department at Children's Hospital of Pittsburgh (CHP), Pittsburgh, Pa., a university-affiliated tertiary care children's hospital. The study was approved by the institutional review board of CHP.
Patients were referred for temporal bone imaging at the request of the evaluating pediatric otolaryngologist. Not all children diagnosed as having SNHI were referred for imaging. For example, some children with a known cause for acquired SNHI were not referred for CT. The study included only children with unilateral or bilateral SNHI; therefore, the incidence of LVAs in normal-hearing subjects cannot be determined.

High-resolution coronal or axial and coronal temporal bone CT scans were performed (at 1- or 1.5-mm intervals) in accordance with the standard clinical imaging protocol at CHP. The VA was classified as enlarged by the evaluating radiologist if its width at the proximal intraosseous portion exceeded that of the adjacent posterior semicircular canal (SCC).16 The CTs of patients diagnosed as having an LVA were subsequently reviewed, and the diameter of the VA at its widest intraosseous portion was measured.

The audiometric records for all patients identified as having an LVA were reviewed. Behavioral audiograms or auditory brainstem response test results for all patients were available from the department of audiology at CHP. The type of hearing impairment, the severity of hearing impairment, and the audiometric configuration were characterized by an individual review of the audiograms for each patient found to have an LVA.

RESULTS

PATIENT POPULATION

We reviewed the records of 221 children. At the time of evaluation, the patients ranged in age from 1 month to 17.2 years (mean age, 5.5 years). The female-male ratio was 1.1:1.0.

RADIOGRAPHIC FINDINGS

Nineteen children had an LVA diagnosed as an isolated anomaly in one or both ears. An isolated LVA was identified in a total of 26 ears. The incidence of isolated LVAs in this group was 8.6% (19 of 221 subjects), with a mean VA diameter of 2.3 mm (range, 1.2-3.9 mm). Also, an LVA with an associated inner ear anomaly was identified in 7 ears. The following inner ear anomalies were diagnosed on CTs (n=33):

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>No. (%) of Ears</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolated enlarged VA</td>
<td>26 (79)</td>
</tr>
<tr>
<td>Enlarged VA with cochlear anomalies</td>
<td>1 (3)</td>
</tr>
<tr>
<td>Enlarged VA with SCC anomalies</td>
<td>2 (6)</td>
</tr>
<tr>
<td>Enlarged VA with cochleovestibular anomalies</td>
<td>1 (3)</td>
</tr>
<tr>
<td>Enlarged VA with SCC and vestibular anomalies</td>
<td>3 (9)</td>
</tr>
</tbody>
</table>

AUDIOMETRIC FINDINGS

Severity

Of the 26 ears with isolated LVAs, 22 (85%) had SNHI, 3 (12%) had mixed hearing impairment, and 1 (3%) had no hearing loss. The severity of hearing impairment as determined by pure-tone averages ranged from none (3 of 26 ears; 12%) to profound (2 of 26 ears; 8%), with 17 ears (65%) demonstrating mild or moderate hearing loss and 6 ears (23%) with severe or profound hearing loss (Figure 1). As the use of pure-tone averages to describe severity of hearing impairment does not measure hearing above 2000 Hz, the degree of hearing impairment can be underestimated in patients who are more severely affected at higher frequencies.

Audiometric Configuration

In 13 (50%) of 26 ears with an isolated LVA, the audiogram was downsloping or showed a high-frequency SNHI. Eight (31% of the total) of the remaining 13 ears had audiograms with a midfrequency peak at 1000 or 2000 Hz. The midfrequency-peaked audiogram is defined by the presence of thresholds at 1000 or 2000 Hz that are 15 dB or better than the adjacent octaves (Figure 2). Other audiometric configurations were seen in 5 ears, including a flat hearing loss in 1 ear. The following tabulation shows the audiometric configuration in 26 ears with isolated LVAs:

<table>
<thead>
<tr>
<th>Configuration</th>
<th>No. (%) of Ears</th>
</tr>
</thead>
<tbody>
<tr>
<td>Downsloping</td>
<td>11 (42)</td>
</tr>
<tr>
<td>High frequency</td>
<td>2 (8)</td>
</tr>
<tr>
<td>Midfrequency peak</td>
<td>8 (31)</td>
</tr>
<tr>
<td>Flat</td>
<td>1 (4)</td>
</tr>
<tr>
<td>Other</td>
<td>3 (11)</td>
</tr>
<tr>
<td>Normal</td>
<td>1 (4)</td>
</tr>
</tbody>
</table>

Laterality

Seven patients had bilateral isolated LVAs, and 12 patients had unilateral isolated LVAs. Of the 7 patients with bilateral LVAs, 6 had bilateral SNHI and 1 had unilateral SNHI. All 6 patients with bilateral LVAs had asymmetrical audiometric thresholds. Of the 12 patients with unilateral LVAs, 7 had unilateral SNHI and 5 had bilateral SNHI. (In 1 patient with a unilateral isolated LVA, the contralateral ear had an LVA with associated anomalies, and the patient had bilateral SNHI.) Overall, 11 patients with LVAs had bilateral SNHI. Audiometric thresholds were asymmetrical in 7 of these patients, including 6 children with bilateral isolated LVAs (Figure 3). Asymmetry was defined as hearing thresholds differing 15 dB or more at 2 or more frequencies between
ears. Therefore, all patients with both bilateral LVAs and bilateral SNHI had asymmetrical audiometric thresholds.

COMMENT

DIAGNOSTIC CRITERIA

Radiographic

The diagnostic criteria for LVAs are controversial and not standardized. Valvassori and Clemis defined VA enlargement as a diameter exceeding 1.5 mm. Subsequently, other investigators have defined VA enlargement as a diameter greater than or equal to 1.4, 1.5, 2.0, 1.5, 2.0, or 4.0 mm. The place at which the VA is measured varies from midway between the common crus and external aperture to the external pore. Also, some investigators have defined the LVA on the basis of comparison to the posterior SCC or the diameter of the facial nerve. Interestingly, malformation of the posterior limb of the lateral SCC has recently been described as occurring in association with an LVA.

The radiologists at our institution, during the years when these patients were initially evaluated, used comparison to the diameter of the SCC as the primary determinant of VA enlargement. Subsequent review of the images obtained revealed that some VAs as small as 1.1 mm had been diagnosed as enlarged. We chose to accept the original interpretation of the radiologist, rather than to recategorize the findings, because (1) there is not a consensus on the radiographic criteria for LVAs; (2) the original evaluations were made consistently, in accordance with a published criterion; and (3) it is difficult to make precise measurements to the 10th of a millimeter on conventional CTs.

Axial CTs are commonly used to evaluate the VA. Dimopoulos et al concluded that the axial transverse view provides sufficient information for visualization of the VA. In contrast, Murray et al reported that coronal CTs permit better visualization and more consistent measurement of the VA.

Developmental Effects

Nonlinear growth of the VA throughout gestation and up to the age of 3 to 4 years has been described, indicating that LVAs may result from abnormal postnatal and preadolescent development. This theory is consistent with the findings of an earlier study that reported a significant difference in VA area in children younger than 12 months compared with those aged 3 to 4 years. The VA size was more variable in the older children.

These reports suggest that radiologic findings in younger subjects should be interpreted carefully. Some children are evaluated by CT during the time when the VA may be enlarging; the average age at presentation for the current study was 5.5 years, and other studies have reported average ages from as young as 3.1 years. To our knowledge, age-based normative data for VA size have not been reported.

CLINICAL PRESENTATION

Progression

Progressive hearing loss has been reported in patients with LVAs. Jackler and De La Cruz cited a stepwise progression of approximately 3.5 dB/y in their patient population with progressive loss. Arcand et al indicated an average progression of 30 dB over a follow-up period of 5.5 years in LVA cases, and Govaerts et al found a 4-dB/y decrement in their patients with LVAs. Bamiou
The degree of SNHI in our patients with LVAs ranges from none to profound, with 65% of ears having mild or moderate hearing impairment. One patient with bilateral LVAs had normal hearing in 1 ear. As the severity of hearing loss will vary over time in patients with progressive SNHI, demonstration of a clear relationship between VA size and severity of SNHI may not be seen consistently. While Antonelli et al\textsuperscript{5} reported significant correlation between LVAs and progressive hearing loss, and Antonelli et al\textsuperscript{5} found that most patients with LVAs had progressive or fluctuating SNHI.

Because of the retrospective nature of our study and the lack of serial audiometric data for many of the patients, we could not systematically examine the progression of SNHI. However, progressive SNHI was clearly seen in 2 patients with an isolated LVA; these patients had changes in audiometric thresholds at 2000 Hz or above. The finding of threshold changes that are limited to higher frequencies also illustrates a problem with defining progressive SNHI on the basis of changes in pure-tone averages.\textsuperscript{4,11,12} By this definition, our 2 patients would not have been identified as having a progressive hearing loss.

**Degree of Hearing Impairment**

The degree of SNHI in our patients with LVAs ranges from none to profound, with 65% of ears having mild or moderate hearing impairment. One patient with bilateral LVAs had normal hearing in 1 ear. As the severity of hearing loss will vary over time in patients with progressive SNHI, demonstration of a clear relationship between VA size and severity of SNHI may not be seen consistently. While Antonelli et al\textsuperscript{5} reported significant correlation between LVAs and progressive hearing loss, and Antonelli et al\textsuperscript{5} found that most patients with LVAs had progressive or fluctuating SNHI.

**Type of Hearing Loss**

All of our patients had SNHI, as only patients with SNHI as an indication for temporal bone imaging were identified for review. An additional conductive component, resulting in mixed hearing impairment, was seen in 3 patients (11%) with isolated LVAs. Mixed hearing impairment has been reported to be more common in other studies.\textsuperscript{3,12}

The cause of conductive hearing impairment in patients with LVAs is unknown. Stapes fixation has been suggested as a cause,\textsuperscript{14} and a case study of a patient with an LVA, mixed hearing loss, and stapes fixation has been reported.\textsuperscript{21} However, other investigators have suggested that cochlear conductive hearing impairment (resulting from abnormal intracochlear fluid pressure), rather than ossicular fixation, is the cause of conductive hearing impairment in patients with LVAs.\textsuperscript{12}

**Configuration**

A downsloping or high-frequency SNHI was seen most frequently in our patients with isolated LVAs (13 of 26 ears). This finding is consistent with earlier reports.\textsuperscript{3,9,12}

A midfrequency-peaked audiogram was the second most commonly seen audiometric configuration in our patients with isolated LVAs. To our knowledge, this audiometric configuration, seen in approximately 30% of the ears with isolated LVAs in our study, has not previously been described in association with LVAs. Interestingly, this peaked audiometric configuration has previously been reported in adult patients with Ménière’s disease\textsuperscript{22,23} as well as in animal models of endolymphatic hydrops.\textsuperscript{24}

The midfrequency-peaked audiogram is upsloping in the low frequencies, peaks at 1 or 2 kHz, and slopes downward through 8 kHz. We defined the peak as being 15 dB greater than its neighboring octaves. This is a stricter criteria than that used in studies of patients with Ménière’s disease.\textsuperscript{22,23} Paparella et al\textsuperscript{25} described a peaked audiogram in 42% of 300 patients with hydrops. The peak audiogram was defined as having air-conduction thresholds for 1 test frequency 10 dB better than adjacent octaves. The peak was at 2000 Hz in 75% of their patients. Lee et al\textsuperscript{22} also examined peak audiometric configurations in patients with Ménière’s disease, using the same criteria. A peaked audiometric configuration was reported in approximately 50% of their patients; the peak was at 2000 Hz in the majority. Ries et al\textsuperscript{25} performed a retrospective analysis of audiometric findings in patients with unilateral hydrops, reporting that 27% of the patients had a peaked audiometric configuration.

**Incidence and Laterality**

In the present group of 221 children with SNHI or mixed hearing impairment, 19 (8.5%) were found to have isolated LVAs. Other studies of patients with SNHI have reported an incidence of LVA ranging from 3.7% to 11.4%.\textsuperscript{4,8,9,11,13,14} The incidence of asymmetrical SNHI in our population is consistent with that in previous reports.\textsuperscript{4,12} However, the high incidence of unilateral LVAs in this group of patients is in contrast to results of past studies.\textsuperscript{4,6,7,9}

A single subject with bilateral isolated LVAs and unilateral hearing loss was identified in our study, and at least 2 similar cases have been reported previously.\textsuperscript{10} This finding illustrates the usefulness of CT in counseling patients with hearing loss, owing to the risk of progressive SNHI and the association of decreased hearing thresholds with minor head trauma.\textsuperscript{4,8,9}
Five cases of a unilateral isolated LVA in children with bilateral SNHI were also identified. In 1 of the 5 cases, the contralateral ear had a combined inner ear anomaly. In the others, the contralateral (radiographically normal) ear may have had a membranous anomaly that was not visualized on CTs, or the diagnostic criteria used for determining the presence of an LVA may not have allowed an accurate diagnosis in some cases. These cases emphasize the point, raised in earlier reports, that the LVA may be a marker for membranous inner ear abnormalities. Thus, the VA may not always be enlarged in cases of membranous inner ear anomaly and, when it is enlarged, may not be clearly identified by current CT criteria.

**MRI and Visualization of the Endolymphatic Duct and Sac**

Because of the limitations of CT in temporal bone imaging, fast spin-echo magnetic resonance imaging (MRI) has been proposed as the procedure of choice for the examination of patients with LVAs. Advances in MRI technology allow imaging of the endolymphatic structures.

Several recent publications suggest that enlargement of the endolymphatic sac and duct is related to hearing impairment in cases of LVA. For example, Hirsch et al described 2 patients with bilateral LVAs who were found to have bilateral enlargement of the endolymphatic sac on MRI examination. Other studies also describe enlarged endolymphatic sacs coincident with LVAs. Although some reports indicate that not all patients with an LVA diagnosed on CTs have an enlarged endolymphatic sac on MRIs, some authors propose that MRI is the preferred imaging modality for patients with suspected inner ear anomalies of this type.

**Ménière’s Disease and MRI**

The peak audiogram has been observed in animal models of Ménière’s disease. During induction of Ménière’s disease in guinea pigs, a low-frequency hearing loss develops first, followed by the development of a high-frequency hearing loss. Eventually, the midfrequency range (8 kHz in the guinea pig) is affected. In another experimental model of endolymphatic hydrops, a high-frequency SNHI developed in guinea pigs after the endolymphatic sac pressure was raised. A low-frequency hearing loss followed, creating a peaked-configuration audiogram. Eventually a flat-configuration audiogram developed.

Despite the common audiometric configurations of patients with LVAs and Ménière’s disease, the appearance of the endolymphatic sac on MRIs in these 2 patient populations seems to differ. In contrast to reports of endolymphatic duct and sac enlargement in patients with LVAs, the endolymphatic duct and sac are typically poorly visualized on the MRIs of patients with Ménière’s disease.

**Mechanisms of SNHI**

Jackler and De La Cruz suggest several potential causes of progressive SNHI in patients with an isolated LVA, including a breach in homeostasis of inner ear electrolytes brought about by the accumulation of excess endolymph, which is caused by a dysfunctional endolymphatic sac. A minor head injury could cause a rupture of the intracochlear membrane, thereby combining endolymph with perilymph and contributing to a stepwise progression of hearing loss. Also, cerebrospinal fluid pressure fluctuations may be transmitted to the inner ear via the patent VA.

The concept of intracochlear pressure change as a mechanism for hearing loss in patients with LVAs was also proposed by Levenson et al, who hypothesized that sudden fluctuation in cerebrospinal fluid pressure could force hypotonic fluid into the basal end of the cochlear duct and produce a high-frequency hearing loss. This reflux is thought to be possible because of an enlarged endolymphatic duct.

Several authors agree that volume or pressure effects precipitated by an enlarged endolymphatic duct account for the hearing loss found in patients with LVAs. Hirsch et al suggest that abnormalities of the inner ear membrane are a factor in LVA-associated hearing losses, but they reach a different conclusion regarding the mechanism at work. They suggest that there is a dilated communication between the cerebrospinal fluid and the vestibule that forms a fistula, damaging the membranous labyrinth. Further histopathologic studies of confirmed cases are needed before definitive conclusions can be drawn.

In fact, the mechanism behind hearing loss in patients with an LVA is a result of pressure fluctuations, it may follow that greater dilation of the VA (and corresponding sac) leads to greater susceptibility to progression of hearing loss. Evaluation of this theory would require close serial audiometric assessment, which is not available in this retrospective review.

**CONCLUSIONS**

1. An LVA was the most common inner ear anomaly identified on CTs in this selected group of children with SNHI.
2. The incidence of a midfrequency-peaked audiogram in this group of patients with SNHI and isolated LVAs was approximately 30%. A similar audiometric configuration has previously been reported in patients with Ménière’s disease.
3. Among children with SNHI and bilateral LVAs, the degree of hearing impairment is frequently asymmetrical.
4. Bilateral SNHI is commonly seen in children with a unilateral LVA, which may illustrate that the LVA is a marker for membranous inner ear abnormalities not visualized on CTs.
5. The radiographic criteria for identifying LVAs are not standardized.

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REFERENCES