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ORIGINAL RESEARCH

The large vestibular aqueduct: A new definition based on audiologic and computed tomography correlation

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OBJECTIVE: The study goal was to determine the prevalence and clinical significance of a large vestibular aqueduct (LVA) in children with sensorineural hearing loss (SNHL).

STUDY DESIGN AND SETTING: We conducted a retrospective review of a pediatric SNHL database. One hundred seven children with SNHL were selected and their radiographic and audiometric studies were evaluated. Radiographic comparisons were made to a group of children without SNHL.

RESULTS: A vestibular aqueduct (VA) larger than the 95th percentile of controls was present in 32% of children with SNHL. Progressive SNHL was more likely to occur in ears with an LVA and the rate of progressive hearing loss was greater than in ears without an LVA. The risk of progressive SNHL increased with increasing VA size as determined by logistic regression analysis.

CONCLUSIONS: An LVA is defined as one that is ≥2 mm at the operculum and/or ≥1 mm at the midpoint in children with nonsyndromic SNHL. An LVA appears to be more common than previously reported in children with SNHL. A linear relationship is observed between VA width and progressive SNHL.

SIGNIFICANCE: The finding of an LVA in children with SNHL provides diagnostic as well as prognostic information.

A large vestibular aqueduct (LVA) is the most commonly identified radiographic abnormality in children with sensorineural hearing loss (SNHL).[1] Valvassori and Clemis[2] were the first to describe a group of patients with abnormally large vestibular aqueducts (defined as ≥1.5 mm at the midpoint) and congenital or early acquired hearing loss and coined the term "large vestibular aqueduct syndrome" (LVAS). The first clinical description of LVAS in children reported the findings of early-onset, progressive, bilateral SNHL in 12 children with vestibular aqueducts ≥2 mm at the operculum.[3] Other studies have reported similar clinical findings, although the definition of an LVA differs among the studies.[4-8] In fact, there is no universally adopted definition of an LVA.[9]

The lack of a standard definition for an LVA makes comparing the results of different studies difficult and the data derived from these studies are quite variable.[8] An LVA is estimated to be found in anywhere from 0.64% to 13% of patients with SNHL; however, the studies suffer from patient selection bias, making these values suspect.[4-9] Progressive SNHL is frequently associated with an LVA with a reported prevalence of 18%-65%.[4-9] Sudden SNHL following minor head trauma has been anecdotally reported in children with

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LVAS, causing some to recommend avoidance of contact sports or other potentially harmful activities. [7,10,11] In addition, a mixed hearing loss in the absence of obvious middle ear pathology has been reported in up to 90% of ears with large vestibular aqueducts (LVAs).[4,6,7,9,12]

The objectives of this study were (1) to establish the prevalence of an LVA among children with SNHL; (2) to determine if children with SNHL and LVAs have an increased risk of progressive SNHL compared to children without LVAs; (3) to describe the relationship between the size of the vestibular aqueduct (VA) and progressive SNHL; and (4) to evaluate the relationship between an LVA and mixed hearing loss.

METHODS

Sensorineural Hearing Loss Group

A database of over 1600 children with SNHL was searched for all patients who had been evaluated between April 1, 2000, and August 1, 2003. A total of 299 patients were identified who had complete audiological and radiographic evaluations. In order to avoid bias, the first 180 patients in alphabetical order by last name were evaluated for the study. Initial power calculations indicated that we would need at least 100 patients (200 temporal bones) to attain any statistical findings. CT studies were either unavailable in digital format or were not satisfactory for analysis in 43 patients. An additional 30 patients were excluded from further study because of the following reasons: deceased without follow-up, severe temporal bone dysmorphology or aural atresia, known syndromic hearing loss, documented ototoxicity, temporal bone fractures, meningitis, hydrocephalus with shunt, auditory neuropathy, or inadequate audiometric information. The temporal bone CTs and medical records of the remaining 107 patients were reviewed, and their radiographic and audiometric data were entered into an electronic spreadsheet for statistical analysis. This study was approved by our institutional review board.

Control Group

The temporal bone CT studies of 100 children without SNHL were collected retrospectively. The cases were identified by inspecting radiology reports of temporal bone CT studies performed between 1995 and 2003. Twenty-seven patients did not have audiograms available for review and were not included. The CT studies of the remaining 73 patients were reviewed by two neuroradiologists (M.H. and C.B.). The CT examinations were performed for the following indications: otomastoiditis (28), trauma (17), cholesteatoma (10), conductive hearing loss (9), mastoidectomy follow-up (3), facial nerve paralysis (3), and one each of headache, otalgia, and otitis externa (3). All of the control subjects had normal sensorineural hearing as defined as an average bone conduction threshold of 20 dB or better over the frequencies of 500 Hz, 1000 Hz, and 2000 Hz.

Temporal Bone CT Analysis

All studies were performed using a standard temporal bone protocol with contiguous 1.0- or 1.25-mm scans. Measurements were made with images enlarged 10× to 15× using current workstation software (Centricity GE Medical Systems, Milwaukee, WI) by two neuroradiologists (M.H. and C.B.). The width of the VA was measured at the *operculum* and the *midpoint* (Fig 1).

The opercular width at the aperture of a VA was measured from the edge of the operculum to the posterior surface of the petrous bone using a line 90 degrees (occasionally less when the operculum and petrous-bone surface was J-shaped) to the posterior surface of the petrous bone. The greatest width was chosen if the operculum appeared on several images.

The VA midpoint was measured in the coronal plane halfway between the posterior wall of the vestibule and the opercular edge. If the vestibule was below the VA midpoint, the posterior wall of the crus commune was used instead of the vestibule. And, if the operculum was below the midpoint, the posterior wall of the petrous bone above the operculum was used in its place.

There was no significant variability between the two raters. The intraclass correlation coefficient between the two different raters was 0.74 ($P < 0.0001$) for the midpoint measurements and 0.93 ($P < 0.0001$) for the operculum.

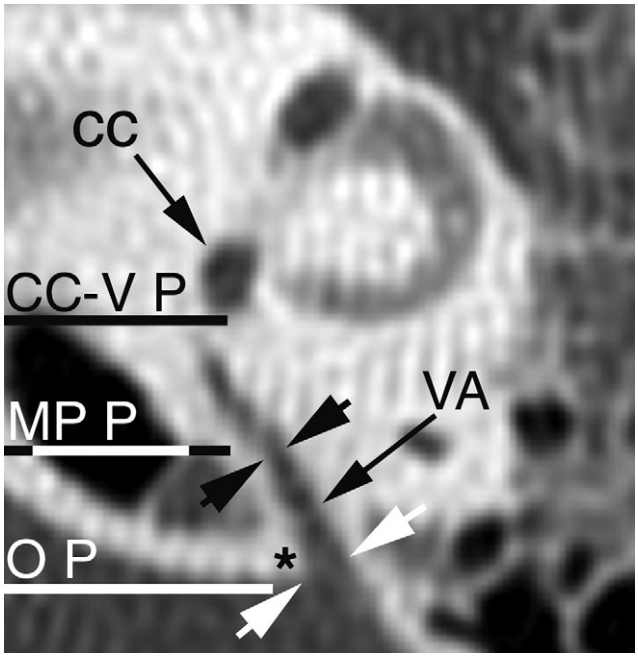

Figure 1 Measuring the vestibular aqueduct (VA) in the axial plane. In this normal VA, the width at the operculum (*) was measured between the tips of the white arrows and found to be 1.4 mm. The midpoint width was measured between the tips of the black arrows in the coronal VA midpoint plane (line marked "MP P"), which is halfway between the coronal plane of the posterior wall of the crus commune or vestibule (line marked "CC-V P") and the coronal plane of the operculum's edge (line marked "O P") and was found to be 0.6 mm.

Audiometric Data

A pure tone average (PTA) for each ear was derived by averaging the audiometric findings at 500, 1000, 2000, and 4000 Hz. Progressive hearing loss was defined as a 10 dB or greater increase in PTA over a minimum 3 month follow-up period. A mixed hearing loss was considered present in ears with normal tympanograms when there was a greater than 10-dB difference between the air conduction and bone conduction in at least one frequency other than 250 Hz. Ears in which the initial PTA was 95 dB or greater were eliminated from further analysis with regard to progressive SNHL because many ears with this degree of SNHL are not routinely tested at follow-up and progression beyond 95 dB is rarely clinically significant.

Statistical Analyses

Continuous data were compared between two groups using *t* tests. Categorical data were compared among groups using χ^2 analysis, and Fisher's exact tests when appropriate. Comparisons made between left and right ears with regard to the VA measurements were conducted using the one-sample Student's *t* test. Logistic regression analysis was used to evaluate the relationship of progressive hearing loss and vestibular aqueduct size. For all analyses, a *P* value of 0.05 or less was considered significant. All analyses were performed using SAS for Windows, version 9.1. SAS software is a product of SAS Institute Inc (Cary, NC).

RESULTS

A total of 164 ears in 107 patients had an elevated PTA at initial presentation. Forty-four (41%) patients presented with unilateral hearing loss and 9 (8%) patients presented with isolated high frequency hearing loss without an elevated PTA. The mean age (SD) of the SNHL population was 6.7 (3.7) years, the mean age of evaluation for patients with unilateral SNHL was 7.3 (3.4) years, and the mean age of evaluation for patients with isolated high frequency SNHL was 10.3 (3.8) years. The mean age of the control group we used for comparison was 9.1 (5.2) years.

An additional 12 ears developed SNHL during the study period, bringing the total number of ears with SNHL to 176. The mean VA widths for the 176 ears with SNHL are shown in Table 1. The correlation between the operculum and midpoint values was strong (correlation coefficient = 0.84, $P < 0.0001$) for both groups. There was no difference in the mean VA size between left and right ears in the in the 54 patients with bilateral SNHL ($P > 0.5$ for operculum and midpoint comparisons). The initial PTA did not appear to be related to VA size (correlation coefficient = 0.04 for operculum and 0.03 for midpoint; $P > 0.5$ for both).

In the control population, the mean width (SD) for the VA at the midpoint and operculum were 0.31 mm (0.30) and 0.99 mm (0.51), respectively. The largest widths were 1.8 and 3.4 mm for the midpoint and operculum, respectively. The percentile values for the VA width at the midpoint and the operculum in control ears are shown in Table 2. The VA was not visible at its midpoint in 17 (23%) control subjects bilaterally and 14 (19%) control subjects unilaterally. The VA was not visible at the operculum in two (3%) control subjects bilaterally and in two (3%) control subjects unilaterally. In these cases, the VA width was recorded as zero.

The VA was considered large when one or both of its widths were above the 95th percentile (>1.9 mm at the operculum and/or >0.9 mm at the midpoint) of the control group measurements. These values are shown in Table 1. Fifty-one (24%) ears in 34 (32%) children with SNHL were found to have vestibular aqueducts that exceeded the 95th percentile of control ears. The VA was large at both the operculum and the midpoint in 29 ears. Ten ears were larger at the midpoint only, and 12 were larger at the operculum only. Seventeen children had bilateral LVAs, and 17 had unilateral LVAs.

Follow-up audiograms were available for 79 patients (74%) whose initial and follow-up evaluations were at least 3 months apart. The mean follow-up interval was 16.9 (10.8) months. The initial mean PTA was 44.9 (34.3) dB, and the final mean PTA was 47.6 (34.3) dB. Seven (20%) of 35 patients with unilateral SNHL developed SNHL in the contralateral ear during the study period. An additional three subjects who presented with isolated high-frequency hearing loss developed elevated PTAs in five ears during the study.

Progressive SNHL occurred in 26 (16%) ears with 3-month or longer follow-up during the study period. The initial mean (SD) PTA in the progressive SNHL group was 40.9 (22.6) dB, and the final mean PTA was 58.3 (27.5) dB. Patients with progressive SNHL had a mean follow-up of 23.8 (8.6) months. The proportion of ears with progressive

Table 1
Comparison of temporal bone measurements in 107 patients with SNHL vs 73 control patients without SNHL

	SNHL Group	Control (no SNHL)	*P* value
VA–operculum	1.39 ± 1.08 mm (range 0-5.7 mm)	0.99 ± 0.51 mm (range 0-3.4 mm) 95th percentile = 1.9 mm	0.0051
VA–midpoint	0.61 ± 0.68 mm (range 0-2.8 mm)	0.31 ± 0.30 mm (range 0-1.8 mm) 95th percentile = 0.9 mm	0.0001

Table 2
The upper ranges of vestibular aqueduct widths in normal hearing ears (n = 73)

Width	Range	Size (mm)
Opercular Width	Maximum	3.4
	99th Percentile	2.7
	97.5th Percentile	2.3
	95th Percentile	1.9
	90th Percentile	1.7
	75th Percentile	1.2
	50th Percentile	0.9
Midpoint width	Maximum	1.8
	99th Percentile	1.3
	97.5th Percentile	1.2
	95th Percentile	0.9
	90th Percentile	0.7
	75th Percentile	0.4
	50th Percentile	0.2

SNHL was higher (24%; 9 of 37 ears) among LVA ears compared to the ears without LVA (14%; 17 of 121 ears), although this difference was not statistically significant (P = 0.14).

The initial mean PTA in the nine LVA ears with progressive SNHL was 51.8 (21.7) dB and the final PTA was 71.75 (30.5) dB. The initial mean PTA in the 17 ears without an LVA was 35.1 (21.4) dB, and the final PTA was 51.2 (23.8) dB. The follow-up time for the non-LVA ears was 27.8 (8.2) months and 16.9 (7.4) months for the LVA ears. The rate of hearing loss was significantly greater in ears with an LVA than in non-LVA ears (1.28 dB/month vs 0.60 dB/month, P = 0.015).

Simple logistic regression analysis was performed to investigate the relationship of VA size to progression of hearing loss (Fig 2). For every 1-mm increase in operculum size, the likelihood of progressive hearing loss increased by 1.37 (95% CI 0.97-1.92), although this relationship was found to be only marginally significant (P = 0.07). For every 1-mm increase in midpoint size, the likelihood of progressive hearing loss increased by 1.93 (95% CI, 1.1−3.4; P = 0.02). For example, a child with a midpoint size of 2.0 mm is 1.9 times more likely to have progressive hearing loss compared to a child whose midpoint measurement size is 1.0. Figure 2 shows the predicted probabilities of progressive hearing loss according to VA widths. The probability of progression for midpoint values above 3.0 mm was estimated because no temporal bone in our series had a midpoint value greater than 3.0 mm.

Of the 176 ears with SNHL (n = 107 subjects), a mixed hearing loss was present in 36 (20%). Mixed hearing loss was present in 20 LVA ears (39%). The degree of conductive hearing loss was typically 10-30 dB and was most often present in the lower frequencies (250-1000 Hz). The mean VA widths were significantly larger in ears with mixed hearing loss compared to ears without a mixed hearing loss. The mean (SD) operculum width was 2.35 (1.34) mm in ears with mixed loss versus 1.10 (0.78) mm in ears without mixed hearing loss ($P < 0.0001$). The mean (SD) midpoint width was 1.08 (0.89) in ears with mixed loss vs 0.46 (0.51) in ears without mixed loss (P = 0.0003). Children with mixed hearing loss were more likely to have progressive SNHL compared to children who did not have mixed hearing loss (34% of mixed hearing loss ears vs 12% of non−mixed hearing loss ears: P = 0.002).

DISCUSSION

This is the first study to determine the normal distribution of VA width by CT in a group of children without SNHL and compare the findings to a group of children with SNHL. We chose the 95th percentile of control measurements as the statistical upper limit of normal VA width for two reasons. First, there is no lower tail to the distribution of VA widths because a nonvizualized VA is considered normal and, therefore, only the upper tail of the distribution curve includes the "abnormal" values. And second, we had previously classified ears with midpoint measurements between 1.0 and 1.4 mm as "borderline" enlarged and found a trend toward progressive SNHL in this group.[7]

We chose to specifically evaluate the LVA because it is the most common structural abnormality noted on CT scans in patients with SNHL and the VA is easily identified and measured.[13] In the present study, we excluded patients with severe inner ear abnormalities such as cochlear aplasia and large common cavity deformities; however, we included patients with other inner abnormalities such as less than complete cochlear partitioning, a smaller than normal modiolus, or enlargement of the vestibule. We did not address the possible clinical contributions of other inner ear anomalies in this study because we wanted to specifically focus on the VA and determine when it should be considered large. As the current data show, there is a strong association

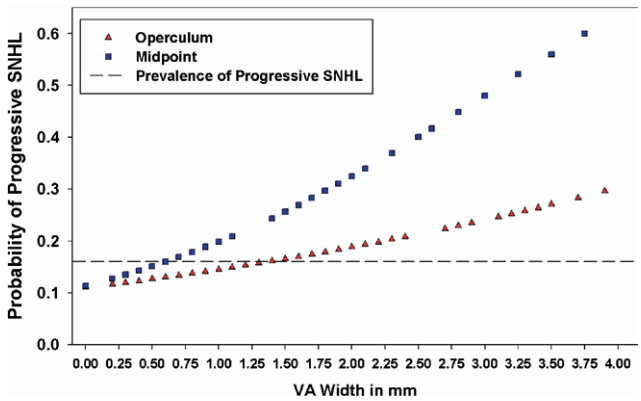

Figure 2 Increasing vestibular aqueduct size increases the probability of progressive SNHL as determined by logistic regression analysis. The prevalence of progressive SNHL of the entire SNHL group was 16% in this study (*dotted line*).

between progressive SNHL, a mixed hearing loss, and an LVA.

The prevalence of an LVA in children with nonsyndromic SNHL was 32% (24% of ears) when an LVA was defined as 2 mm or greater at the operculum and/or 1 mm or greater at the midpoint. We anticipated that using the 95th percentile of control measurements as our LVA definition would likely classify several patients as LVA who would have previously been classified as having a "normal" VA. This is because many previous LVA studies used a threshold value greater than any normal hearing ear (i.e., greater than the 100th percentile) to define an LVA.[2,5,8] We maintain that our choice of the 95th percentile is not only statistically acceptable but also clinically significant. As the current data show, the LVA ears in our study had unique clinical findings including an increased rate of progressive SNHL and a significant association with mixed hearing loss.

This second aim of this study was to determine if an LVA was a risk factor for progressive SNHL. Progressive SNHL is a common clinical finding in patients with an LVA.[4-9] In the current study, we found that 24% of ears with an LVA demonstrated progressive SNHL. This is a striking, although not statistically significant ($P = 0.14$), difference compared to the 14% SNHL progression in ears without an LVA. However, the rate of progressive SNHL was statistically greater in ears with an LVA than in ears without an LVA (1.28 dB/month versus 0.60 dB/month, $P = 0.015$). This finding is consistent with the observation that children with LVAs are prone to sudden deterioration in hearing triggered by events that may increase intracranial pressure.[4,7,11,12]

Progressive SNHL in patients with an LVA is likely greater than the current study indicates. The total number of ears with progressive SNHL in our study is small (n = 26), and the follow-up time of approximately 2 years is short. In addition, some children may have already had significant SNHL progression prior to their initial evaluation. We anticipate that the addition of more patients and longer follow-up times should provide a statistically significant difference in the risk of progressive SNHL between ears with an LVA and those without an LVA.

In a previous study, we found that ears with progressive SNHL had significantly larger opercular VA widths than did ears with stable hearing.[7] In the present study, we performed logistic regression analysis to determine more precisely the relationship between VA size and the probability of progressive SNHL (Fig 2). A linear relationship is apparent between the midpoint VA size and the probability of progressive hearing loss. The probability of progressive hearing loss also increased with increasing operculum width, albeit to a lesser degree.

As shown in Figure 2, the probability of progressive SNHL begins to exceed the baseline prevalence of progressive SNHL (16%) in the entire SNHL group when the VA width reaches either 0.9 mm (95th percentile of control values) at the midpoint or 1.8 mm (between the 90th and 95th percentiles of controls) at the operculum. These findings lend further, clinically relevant, support to defining an LVA as one that is 2 mm or greater at the operculum and/or 1 mm or greater at the midpoint. The data also suggest that a regression table may prove useful when predicting the possible risk of progressive hearing loss in children with SNHL and an LVA. A longer follow-up period and the inclusion of additional patients should help to verify this finding. It is important, however, to note that the results of our logistic regression analysis were unadjusted and that there may be other important factors that influence the likelihood of progressive hearing loss for which we are not controlling. Therefore, the results should be interpreted with some caution.

Mixed hearing loss has been frequently associated with LVAs.[4,6,7,9,12] We found a mixed hearing loss in 20 (39%) LVA ears compared to only 16 (12.8%) non-LVA ears. Ears with mixed hearing loss demonstrated significantly larger vestibular aqueducts than ears with only SNHL, and the presence of a mixed hearing loss was associated with an increased risk of progressive SNHL (34% of mixed hearing loss ears vs 12% of non−mixed hearing loss ears: $P = 0.002$). All of the patients in our study had normal tympanograms and normal otoscopic examinations. Middle ear explorations were not performed in any patients.

The clinical finding of mixed hearing loss in the absence of middle ear pathology is unique to only a few conditions, including Meniere's disease,[14] Paget's disease,[15] and lateral semicircular canal dysplasia.[16] The strong association between an LVA and mixed hearing loss may provide some insight into the pathophysiology of hearing loss in patients with an LVA. Possible mechanisms of conductive hearing loss in patients with LVAs include stapes fixation, abnormalities in cochlear fluid pressures, and the "third window" phenomenon.[17-19] Another possible mechanism involves structural abnormalities of the membranous labyrinth and/or the osseous spiral lamina that could cause an "inner ear" conductive hearing loss as well as making the cochlea more susceptible to traumatic injury from minor head trauma or increased intracranial pressure. In any case, the presence of a mixed hearing loss is a common finding in children with an LVA, and CT imaging should be obtained for all children presenting with a mixed hearing loss.

An LVA is frequently found in association with other inner ear abnormalities,[9,10,20-22] and it is quite likely that most, if not all, LVAs are associated with abnormalities of the membranous labyrinth.[4,22] At the present time, however, it is impossible to say which of these many inner ear abnormalities are more significant with regard to the patient's clinical presentation. We are currently evaluating the relationship between various inner ear anomalies and hearing loss in children. In the meantime, finding an LVA in a child with SNHL prompts us to look for other potential inner ear abnormalities as well as guiding us to further evaluations such as genetic testing for Pendred's syndrome.

In addition, patients and families can be counseled as to the likelihood of progressive hearing loss.

CONCLUSIONS

An LVA is one that is greater than 1.9 mm at the operculum and/or greater than 0.9 mm at the midpoint in children. In children with SNHL, the finding of an LVA is strongly associated with progressive SNHL as well as mixed hearing loss. An LVA may be easily measured using high-resolution CT and can be diagnostic as well as prognostic for children with unexplained SNHL.

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