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Enlarged vestibular aqueduct in pediatric SNHL

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Abstract

Objective—Comparison of the Cincinnati criteria (midpoint >0.9 mm or operculum >1.9 mm) to the Valvassori criterion (midpoint $\geq$ 1.5 mm) for enlarged vestibular aqueduct (EVA) in pediatric cochlear implant patients.

Study Design—Cohort study

Subjects—130 Pediatric cochlear implant recipients.

Methods—We reviewed temporal bone CT scans to measure the vestibular aqueduct midpoint and opercular width.

Results—The Cincinnati criteria identified 44% of patients with EVA versus 16% with the Valvassori criterion ($P < 0.01$). Of those with EVA, 45% were unilateral and 55% were bilateral using Cincinnati criteria; 64% were unilateral and 36% bilateral using Valvassori criterion ($P < 0.01$). The Cincinnati criteria diagnosed 70 ears with EVA classified as normal using the Valvassori criterion ($P < 0.01$); 59 lacked another medical explanation for their hearing loss.

Conclusion—The Cincinnati criteria identified a large percentage of pediatric cochlear implant patients with EVA who might otherwise have no known etiology for their deafness.

INTRODUCTION

Currently over 10,000 children living in the U.S. have a severe-to-profound sensorineural hearing loss (SNHL).¹ Unfortunately, the cause of SNHL for nearly 50% of those children is unknown. In most cases of SNHL, the hearing loss is an isolated congenital abnormality, nonsyndromic in nature, believed to be caused by a combination of environmental and genetic factors. Nearly half of all cases detected before age 2 years have an identifiable environmental cause including congenital infections such as cytomegalovirus.² Neonatal asphyxia, premature birth and exposure to ototoxic medications are other common causes of congenital SNHL. Non-syndromic hearing loss has been linked to greater than 50 genes.³ The most frequent

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genetic cause of SNHL is mutation of the GJB2 gene, also known as connexin 26, accounting for nearly half the cases of profound nonsyndromic hearing loss². Other genetic causes include syndromes like Waardenberg's and Pendred, as well as nonsyndromic genetic conditions.

Temporal bone imaging, comprised of high resolution computed tomography (CT) or magnetic resonance imaging (MRI) or both, is a common element for evaluating a child with SNHL to rule out structural abnormalities. Enlarged vestibular aqueduct (EVA) is the most common CT abnormality in children with SNHL.⁴ The vestibular aqueduct transmits the endolymphatic duct and the veins of the vestibular aqueduct. EVA syndrome is thought to result from a congenital malformation of the temporal bone that may predispose the patient to early onset of hearing loss and vestibular disturbance.⁵ Seemingly minor head trauma may be associated with progressive or fluctuating hearing loss that is eventually profound in some patients. Although the degree of risk correlated with vestibular aqueduct enlargement remains largely unknown, most otolaryngologists caution patients with EVAs to avoid high-impact activities, including contact sports, and advise them to use protective head gear when participating in moderate-risk activities. Relatively little is known about the mechanisms of hearing loss and vestibular disturbance, which make prognosis and management challenging.⁵ Early identification of children who will develop progressive hearing loss could improve clinical, communicative, and educational outcomes of children at risk for a handicapping hearing loss.

The current radiographic diagnostic criterion for EVA is a vestibular aqueduct larger than 1.5mm at the midpoint, as defined by Valvassori and Clemis in 1978⁶ using hypocycloidal polytomography. Based on a review of 73 CTs from children with known normal hearing, new criteria for the diagnosis of EVA have been proposed.⁷ These criteria, which we will refer to as the Cincinnati criteria, were based on a statistical analysis which defined vestibular aqueducts with midpoint width (1.0 mm) or opercular width (2.0 mm) that were greater than the 95th percentile as enlarged. There is great interest in developing a CT scan evaluation rule to see if the size or shape of the vestibular aqueduct can give some insight into whether the patient is at higher risk of hearing loss.⁸

The primary objective of this study was to compare the Cincinnati criteria for EVA with the Valvassori criterion in a large cohort of pediatric cochlear implant recipients, to determine whether misclassification in the etiology of deafness may be occurring. The secondary objective was to investigate other measurable temporal bone abnormalities in this same population.

METHODS

We performed a retrospective medical record review of 163 patients who received cochlear implants in at St. Louis Children's Hospital (SLCH) from January 2003 through August 2007. Extracted data included demographic information, known medical diagnoses, and any pertinent laboratory or imaging study that would shed light on the etiologies of the hearing loss. Patient CT scans were reviewed to assess the anatomy of the temporal bone. We obtained institutional review board approval, including a waiver for informed consent, from the Washington University Medical Center Human Research Protection Office (HRPO) before beginning this study.

Subjects

One hundred sixty-three patients underwent cochlear implantation at SLCH during the described time period and their temporal bone CT studies were reviewed. Scans performed before August 2003 used a thicker slice width (1.0 mm vs. 0.6 mm), with insufficient resolution for accurate measurement of the vestibular aqueduct. Seven scans were excluded for this reason. Of the 156 CT scans that were reviewed, four were excluded for severe structural

abnormality of the inner ear (e.g., severe vestibulocochlear dysplasia or common cavity), making identification of the structures of interest impossible. In 22 patients, CT scans were either not available or those patients received preoperative MRIs only. Thus, a total of 130 scans were included in the study, providing information on 242 ears. Fourteen ears were excluded because cochlear implants were present at the time of the CT scan and therefore structures could not be measured due to scatter.

CT Temporal Bone Measurements

All studies were performed using a standard temporal bone protocol with contiguous 0.6 mm scans of the temporal bone acquired in the axial and coronal planes using 120 kV with Somatom Sensation 16 Multidetector Sys (Siemens Medical Solutions Erlangen, Germany). Only axial-plane images were used. Measurements were made with images enlarged 10 to 15 times with the use of current workstation software (Synapse, Fujifilm Medical Systems, Stamford, CT), by KD. A random 10% sample was re-measured by JECL, and corroborated by FJW. The width of the vestibular aqueduct (VA) was measured at the operculum and the midpoint. The methodology of VA measurement was adapted from Vijayasekran et al⁹ (Figure 1).

VA midpoint measurement

Measurements were taken using the electronic calipers and recorded in millimeters. All measurements were entered into a Microsoft Access database. The VA, in the axial plane, originates from the upper vestibule close to the crus commune and ends in an aperture measured along the length of the VA. The origin of a normal aqueduct is usually invisible, therefore we used the posterior wall of the vestibule. The midpoint was measured on the image where the width was the largest. If the operculum was below the image used for measurement, the posterior wall of the petrous bone was used instead. Widths smaller than the smallest interval able to be measured by the electronic calipers were recorded as 0 mm.

VA opercular measurement

Opercular widths were measured by drawing a line from the opercular edge anterolaterally to form a 90° angle with the posterior wall of the petrous bone. If a 90° angle was not attainable due to the J morphology of the petrous wall a 70° or 80° angle was used. The CT image on which the opercular width was largest was used for measurement. Widths smaller than the smallest interval able to be measured by the electronic calipers were recorded as 0 mm.

LSCC Bony Island measurement, Vestibule width and height measurements

Measurements of the bony island and vestibule were taken on the same cut. The bony island was measured in a medial to lateral fashion. The cut displaying the largest area of vestibule was used for measurements. This methodology was adapted from Purcell et al.¹⁰

IAC opening width and length measurements

Measurements were taken on the cut in which the IAC appeared the longest. A tangential line was drawn across the porous acousticus. The length of this line was taken to be the width of the IAC opening. The line was then bisected with a perpendicular line. The length of the perpendicular was taken to be the length of the IAC.

Statistical analysis

Categorical data were compared between groups using chi-square analysis and Fisher's exact tests. Continuous numerical data were analyzed using t-tests. The correlations of continuous data were investigated using the Pearson *r* correlation coefficient. For all analyses, a two-sided

P value of 0.01 was considered significant. All statistical calculations were performed using SPSS for Windows, version 15.0 (Lead Technologies Inc, Charlotte, NC).

RESULTS

CT scans of the temporal bones in 130 pediatric cochlear implant recipients with an average age of 5.2 years (SD 4.4 years) were reviewed. Figure 1 shows examples of normal versus enlarged vestibular aqueducts by both sets of criteria. The Valvassori criterion of EVA defined 16% of the ears as abnormal, whereas the Cincinnati criteria found 45% of the ears to have EVA ($P<0.01$). The Cincinnati criteria diagnosed 70 ears with EVA that were previously classified as normal using the Valvassori criterion ($P<0.01$) (Table 1.) Of these 70 ears, 11 (16%) had another reason for hearing loss, including Waardenberg's syndrome, congenital CMV, and auditory neuropathy. The other 59 ears (84.3%) were diagnosed with EVA de novo using the Cincinnati criteria. Figure 2 shows an example of a patient with bilaterally enlarged vestibular aqueducts as defined by the Cincinnati criteria but normal by the Valvassori criterion. All of the EVAs defined by the Valvassori criteria (38 ears) were also classified as EVA by the Cincinnati criteria. Using the Valvassori criteria, 28 of 112 (25%) patients with bilateral CT scans had at least one EVA, of which 18 were unilateral and 10 were bilateral. Using the Cincinnati criteria, 64 of 112 (57%) patients with bilateral CT scans had at least one EVA, of which 29 (26%) were unilateral and 35 (31%) were bilateral. These differences were also statistically significant ($P<0.01$) (Table 1).

Connexin mutation analysis was done in 27 of the 130 patients. Of these 27 patients, 12 were found to have mutations of either the connexin 26 or connexin 30 genes. Two of those diagnoses were corroborated by radiologic findings when using the Valvassori criteria for EVA, whereas the Cincinnati criteria identified 8 of 12 patients diagnosed with a connexin mutation. Four subjects with connexin mutations had no EVA by either criteria.

Right and left ear measurements correlated substantially with respect to the vestibular aqueduct operculum size ($r = 0.67$, $P<0.001$) and midpoint size ($r = 0.58$, $P<0.001$). Similarly, there was excellent correlation between the vestibular aqueduct operculum and midpoint measurements ($r = 0.88$, $P<0.001$) (Fig 3). No correlation existed between the age of the patient at the time of CT scan and the vestibular aqueduct midpoint or operculum measurements (Figs 4, 5), although there was a weak correlation between the patient age and the IAC length ($r = 0.25$, $P<0.01$). No significant differences in the LSCC bony island width, vestibule width, vestibule height, IAC opening width, and IAC length were found for those patients who had EVA according to the Valvassori criterion versus those who had EVA according to the Cincinnati criteria (Table 2).

DISCUSSION

The Cincinnati criteria for CT diagnosis of EVA identified a significantly greater proportion of patients with EVA than would have been identified by the Valvassori criterion. EVA may be an etiology of pediatric SNHL for nearly half of this study population. While this new information is a significant step forward in determining the etiology of SNHL in a large proportion of this group, further research is necessary to clarify the physiologic relationship between EVA and SNHL. EVA may be a cause of SNHL, or it might be a marker for another underlying process causing SNHL. The data presented in this study confirm the usefulness of the Cincinnati criteria in evaluation of congenital SNHL. With the use of this new diagnostic criteria, many more CT scans may identify EVA, giving parents a potential etiology for their child's deafness. Because EVA has a genetic component, with 27% reported to have the *SLC26A4* gene mutation in one study of children with EVA,¹¹ knowing the cause of deafness may impact family planning for both for patients and their families.

In our large sample of patients with profound SNHL, the Cincinnati criteria led to the finding that only 24% of EVAs are unilateral, compared to the frequency in the literature ranging from 60–65%.^{12–14} The significant correlations between measurements taken on the right and left sides of the same patient suggest that EVA may not be as morphologically asymmetric as previously thought. While there is a significant correlation between the midpoint and operculum measurements, it is not perfect. Therefore, some patients with EVA would likely be missed if only one of these dimensions was measured. The shape of some EVA (e.g., funnel-shaped or globular) may preclude meeting the definition of “enlarged” at both places; thus, it would be important to clinical practice that both the midpoint and operculum widths are recorded in diagnosing EVA using the Cincinnati criteria. This is in agreement with the conclusions of Vijayasekaran et al,⁹ indicating that both dimensions are necessary for the diagnosis of EVA.

An intriguing incidental finding of this study is that 8 of 12 patients with known connexin mutations were also found to have EVA by the Cincinnati criteria; only 4 would have been identified with the Valvassori criterion. This is a higher percentage than the 8% with EVA found by Propst et al when they investigated temporal bone anomalies in children with biallelic *GJB2* mutations.¹⁴ They also commented that EVA was more likely to be diagnosed by visual inspection than when the strict Valvassori criterion was used.

The lack of correlation between the age of the patient and the VA dimensions verifies current belief that the otic capsule temporal bone structures are fully formed at birth and do not continue to grow throughout life.⁹ Additionally, the IAC is outside the otic capsule, and therefore the correlation between its length and the age of the patient does not violate current dogma.¹⁵

A recent study by Chen et al⁸ indicated no statistically different measurements of the inner ear between children with and without SNHL. The average LSCC bony island width in our data is consistent with that presented in the Chen article (Table 2.). While this dimension may not be useful for distinguishing children with SNHL from those without SNHL, vestibular aqueduct measurements are not addressed by the Chen article and do in fact appear to correlate with the presence of SNHL.

The optimal imaging technique for visualizing the inner ear in the clinical work-up of pediatric SNHL remains to be determined. Both high resolution CT (HRCT) and MRI are valuable, each with particular strengths. In current clinical practice, children with bilateral symmetric SNHL are more likely to be evaluated using HRCT, while those with asymmetric SNHL are more likely to be evaluated with MRI alone or in conjunction with HRCT.¹⁶ Some authors have asserted that MRI alone should be used to image children with SNHL. Using MRI, Davidson et al found that coexistent cochlear and/or vestibular anomalies were present in most ears with enlarged vestibular aqueduct; these coexistent abnormalities may explain the hearing loss and may also impact treatment decisions and prognosis.¹⁷ In another study of 170 children with SNHL evaluated using MRI, 40% of the 271 imaged ears were found to have inner ear abnormalities, the most common of which were an abnormal cochlea and nerve.¹⁸ In contrast, Trimble et al compared the utility of HRCT and MRI prior to cochlear implantation in children.¹⁹ Their study indicated that the best protocol was both MRI and HRCT to image the temporal bone because neither modality was sufficient to detect all abnormalities related to deafness independently. A greater percentage of radiologic abnormalities were identified using HRCT scan as compared to MRI, 59% vs. 32% respectively. Additionally, MRI consistently under-identified EVA and narrow cochlear nerve canals. They also concluded that the HRCT alone was the best single imaging modality for the diagnostic of pediatric SNHL etiology, when patient risk factors are taken into account. In a similar study of 131 children with unilateral or asymmetric SNHL that compared MRI with HRCT, temporal bone HRCT scans were able to identify abnormalities in a greater percentage of children than MRI.²⁰ Simons et al concluded

that the clinical workup for unilateral or asymmetric SNHL should include a HRCT of the temporal bone, and that MRI should be done only in select cases. While significant controversy concerning the most appropriate imaging in the diagnosis of pediatric SNHL remains, this study examined the dimensions measured on the temporal bone HRCT scan as part of the accepted diagnostic algorithm most widely used.

The primary strength of this investigation lies in the large number of CT scans that were evaluated. Previous literature reported rather small sample sizes ranging from only 6 ears to 146 ears. The data presented here contains nearly double the number of ears (242) presented in the largest published study of CT-based assessment of vestibular aqueducts.

The major limitation of this study is that the CT scans were read by one individual who knew that all patients had SNHL, with verification of measurements of a random subsample by a second reader. While the spiral CT with sub-millimeter reconstruction yields considerable detail, the vestibular aqueduct is normally a small structure. The size of the VA demands a very thin slice width during CT scanning as well as precise positioning of the patient; thus images of the small structure are easily distorted by motion and influenced by reader bias. Given current practices, there is substantial potential for intra- and inter-observer variability regarding VA measurements.

CONCLUSION

The data presented here support the use of the Cincinnati criteria for EVA, contributing to the physician's ability to characterize and classify the etiology of SNHL in children. As we further our understanding of the genetic, physiologic and anatomic correlates of SNHL, we may be able to develop better methods for screening, prevention, diagnosis and management of congenital SNHL. Continued research is needed to elucidate the physiologic relationship between EVA and SNHL since the significance is still elusive, and to establish a common protocol for the measurement of inner ear structures to ensure consistency between observers. The establishment of a common algorithm as well as the revision of thresholds for diagnosis of EVA will increase the consistency and accuracy of radiologic EVA identification. Because our population of patients included only those who were already candidates for cochlear implantation, we cannot make conclusions about the risk of progressive or sudden hearing loss in those with EVA.

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Figure 1.

Temporal bone CT scans of normal and enlarged vestibular aqueducts by both the Valvassori and Cincinnati criteria. A. Normal right vestibular aqueduct. B. Enlarged left vestibular aqueduct. *1: Mastoid Air Cells. *2: Internal Auditory Canal. *3: Limb of the posterior semicircular canal. *4: Horizontal semicircular canal. OP: operculum width, perpendicular to

the PL. MP: midpoint width, bisects the PL. PL: line from the posterior wall of the vestibular aqueduct, perpendicular to the ray measuring operculum width.

Figure 2.

Temporal bone CT scan of a patient with bilaterally enlarged vestibular aqueducts (marked by arrows) as defined by the Cincinnati criteria but normal by the Valvassori criterion. A. The right vestibular aqueduct measures 1.3 mm at the midpoint and 1.5 mm at the operculum. B. The left vestibular aqueduct measures 1.1 mm at the midpoint and 1.0 mm at the operculum.

Figure 3.
Correlation of the midpoint and the operculum measurements of 242 ears without regard to right or left ear.

Figure 4.
Scatter plot of midpoint measurement compared with age of subject.

Figure 5.
Scatter plot of opercular measurement (mm) in comparison to age of subject.

Table 1

Comparison of Valvassori and Cincinnati criteria for CT diagnosis of EVA.

	Valvassori n (%)	Cincinnati n (%)	Total	P value
Ears	38 (15.7)	108 (44.6)	242	<0.001
Patients			112	
Unilateral	18 (16.1)	29 (25.9)		<0.001
Bilateral	10 (8.9)	35 (31.1)		<0.001

Table 2

Dimensions measured on CT scans of pediatric cochlear implant recipients

	All ears (n= 242)	Left (n=123)	Right (n=119)
Dimension (in mm)	Mean (SD [*])	Mean (SD)	Mean (SD)
LSCC Bony Island Width	4.0 (0.8)	4.1 (0.8)	3.9 (0.8)
Vestibule Width	6.7 (1.1)	6.8 (1.1)	6.7 (1.1)
Vestibule Height	3.6 (0.8)	3.7 (0.7)	3.6 (0.9)
IAC opening width	6.2 (1.5)	6.3 (1.6)	6.1 (1.5)
IAC length	9.4 (1.7)	9.5 (1.8)	9.3 (1.6)
VA Operculum	1.3 (1.0)	1.3 (0.9)	1.4 (1.1)
VA Midpoint	1.0 (0.7)	0.9 (0.7)	1.0 (0.7)

* SD, standard deviation