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Children With Unilateral Cochlear Nerve Canal Stenosis Have Bilateral Cochleovestibular Anomalies

Luis D. Vilchez-Madrigal, MD; Susan I. Blaser, MD, FRCPC; Nikolaus E. Wolter, MD, MSc; Adrian L. James, DM, FCRS (ORL-HNS); Blake C. Papsin, MSc, MD, FRCSC; Karen A. Gordon, MA, PhD; Sharon L. Cushing, MSc, MD, FRCSC; Evan J. Propst, MSc, MD, FRCSC

INTRODUCTION

Bony cochlear nerve canal (bCNC) stenosis refers to narrowing of the bony canal that carries the cochlear nerve between the internal auditory canal (IAC) fundus and the spiral ganglion or modiolus. Hypoplasia of the bCNC on computed tomography (CT) imaging has been found to correlate well with cochlear nerve diameter on magnetic resonance imaging (MRI) have been correlated with the degree of hearing loss. Unilateral hearing loss (UHL) has been associated with a 22% to 35% rate of repeating the degree than the stenotic side. The vestibular end-organ was also smaller in bCNC ears, this difference was not significant. The contralateral ear also had a smaller bCNC as compared with controls, although to a lesser degree than the stenotic side.

More specifically, both cochlear and vestibular apparatus appear to be affected bilaterally in the majority of patients with cochlear anomalies, which is likely due to abnormal signalling in the first few weeks of embryogenesis. The purpose of the present study was to evaluate the entire vestibulocochlear apparatus in children with unilateral bCNC stenosis to determine if the ipsilateral and/or contralateral vestibulocochlear apparatus are affected. A histopathological study of cochlear nerve canal (CNC) diameter measured in temporal bone specimens from normal hearing individuals suggests that there is a range of normal with a mean minimum diameter of 1.75 mm and a mean maximal diameter of 2.26 mm. The authors have no other funding, financial relationships, or conflicts of interest to disclose.

Send correspondence to Evan J. Propst, MD, Department of Otolaryngology, Hospital for Sick Children, 6103 Burton Wing, 555 University Avenue, Toronto, ON, Canada M5G1X8. evan.propst@sickkids.ca

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characterized according to the World Health Organization classification based on the four-tone average. The remaining 6/36 subjects had an adequate (side and degree) description of the hearing loss based on clinical and radiologic notes but did not have audiometric data available at our institution for review. These children were classified based on their description. The audiologic cutoffs defining normal hearing were ear-specific air/conduction values of <20 dB across four frequencies (500 Hz, 1,000 Hz, 2,000 Hz, 4,000 Hz).

Where performed, newborn screening in a well baby occurs through our provincial Infant Hearing Program Protocol, which includes an automated four-frequency distortion product otoacoustic emissions (DPOAE) test in response to 59/50 dB SPL test level measured using an AccuScreen (Otometrics, Taastrup, Denmark).

Temporal Bone Measurements

High-resolution CT (HRCT) petrous examinations were performed without contrast administration using a GE HD 750 64-slice HRCT scanner (General Electric) and low-dose protocol providing 0.625-mm-thick slices. Temporal bone CT imaging previously obtained for clinical reasons were retrospectively evaluated. A neuroradiologist (S.I.B.) experienced in temporal bone measurements used the measurement tools provided by the GE PACS system to analyze studies. The specific methods for measurements are detailed in Table I and are consistent with those methods previously utilized and reported by our group for several temporal bone measurement studies.9–12 In summary, 26 measurements were performed on each cochleovestibular apparatus using direct measurements recorded in millimeters (rounded to two decimal points) and direct visualization and categorization. Seventeen were obtained by direct measurement, six computed from direct measurement data, and four by categorization. All 17 direct measurements were made on axial section; however, coronal sections were used to verify the presence or absence of dilation of the vestibular aqueduct. Data were analyzed using SPSS version 21 (IBM, Armonk, NY) and R 2013 (R Project for Statistical Computing, Vienna, Austria). Ears with UHL due to bCNC and their contralateral normal hearing ears were compared to normal hearing controls using one-way analysis of variance. Within-subject comparisons of the ear with UHL versus the contralateral normal hearing ear were made for the most part using paired t tests corrected for multiple comparison (Bonferroni adjustment, adjusted P = .0006) with the exception of one comparison made using a Wilcoxon rank test in the setting of a non-normal distribution and another using χ² for ordinal data. Post hoc adjustment for multiple comparisons was made using a Bonferroni correction. Measurements made by the neuroradiologist (S.I.B.) were compared with those made by the pediatric otolaryngology-head and neck surgery fellow (L.D.V.-M.). Across all measurements, inter-rater reliability was found to be excellent using intraclass correlation coefficient (ICC = 0.997, P < .001). For the final computations used in the analysis, where discrepancies existed, these were resolved by re-review and consensus. This evaluation yielded all measurements on all study subjects and on the majority of the controls (Table II). There were a small number of cases where due to motion artefact there was a limited reduction in image quality that prevented specific measurements, and therefore, there is a small difference in the number of control subjects for some measurements.

RESULTS

Subjects

Seventy children (140 ears) were included in this study: 36 patients (72 ears) with UHL associated with

Audiometric Assessments

Hearing status was known in all children. The majority (30/36 subjects as well as all 32 controls) had ear-specific pure-tone audiograms performed on a conventional audiometer available for review. For these children, the severity of the hearing loss was
bCNC stenosis (25 male, 11 female) and 32 (64 ears) normal-hearing controls (23 male, nine female). The mean age at the time of CT was 8.0 ± 3.7 years (range, 1.06–15.92 years) for the UHL group and 8.6 ± 4.6 years (range, 0.98–16.74 years) for the normal-hearing control group. There was no statistical difference in age across groups (P = .56).

**Temporal Bone Evaluation**

**Cochlear nerve canal.** Measurements are outlined in Table II. All 36 patients in the bCNC stenosis group had a submillimeter bCNC diameter in the affected ear (Fig. 1). The contralateral bCNC was larger (mid-distance width and ratio mid-distance width to porus width) than the critically stenosed side, but was still smaller when compared with normally hearing individuals.

**Cochlea.** The ipsilateral cochlea (apical and basal turns) of individuals presenting with critical unilateral bCNC stenosis was hypoplastic as compared with normally hearing controls. The contralateral ear also had a smaller cochlea (P < .000) as compared with controls, although to a lesser degree than the stenotic side.

**Posterior labyrinth.** The lateral semicircular canal bone island width, posterior semicircular canal width, and vestibule were all smaller on the side of the bCNC stenosis as compared with the contralateral ear and controls (Fig. 1). The vestibular aqueduct porus width was larger bilaterally when compared with controls, but there was no difference in porus width across critically stenosed and contralateral ears.

**Audiology**

The mean age at the time of evaluation was 8.14 ± 3.11 years (range, 0.8–17.5 years). The degree of hearing loss in the bCNC stenotic ear at presentation was profound in 29 (80%), severe in three (8%), moderately severe in two (5.5%), and moderate in two (5.5%). Hearing was normal in the contralateral ear in all other...
children. Individual audiometric data for the group of children with unilateral bCNC stenosis can be found in Figure 2.

Newborn hearing screening results were only available in six children (18%), as the majority of children were born prior to the institution of universal newborn hearing screening in our province. Four of six (67%) failed their infant hearing screening, which consists of an automated four-frequency DPOAE. Three had their hearing loss confirmed using evoked auditory brainstem responses. One of these children had otoacoustic emissions present in the ear with bCNC stenosis along with an absent auditory brainstem response confirming auditory neuropathy.

Two patients who passed their initial screen had UHL identified in childhood; one had mild hearing loss diagnosed at 3 years of age (CNC measured 0.8 mm on CT), the other was diagnosed at 5 years of age with profound hearing loss (CNC measured 0.68 mm on CT). Although not an outcome of the current study, there was no audiologic proof of progression over time, and likely, those children who were diagnosed with new hearing loss at older ages are rather a product of late identification.

No maternal or newborn risk factors were noted in either child’s medical history.

DISCUSSION

Given the more frequent use of imaging in the setting of UHL, we more recently have begun to appreciate the relatively high prevalence (50%) of bCNC stenosis as
the underlying etiology in children presenting with profound UHL. Recent work has demonstrated that both cochlear and vestibular apparati appear to be affected bilaterally in the majority of patients with cochlear anomalies, which may be due to abnormal signaling in the first few weeks’ of embryogenesis. Such a hypothesis may be supported by evidence that anatomic abnormalities such as bCNC occur on a spectrum that might range from bilateral symmetric deficits, to asymmetric deficits, and finally deficits that affect only a single ear. In keeping with this, Wilkins et al. in 2012 looked at a population of individuals who had imaging for hearing loss and were found to have bCNC stenosis. They found a relatively high rate of bilateral bCNC (60%) given their use of more conservative criteria than this current study. We therefore investigated the cochleovestibular apparatus bilaterally in children with UHL associated with what was reported to be isolated unilateral bCNC stenosis to see if this condition is a unilateral or bilateral entity.

We found that both the ipsilateral and contralateral normal cochleovestibular apparatus were smaller than controls, with the ipsilateral side being most severely affected. This finding, that visibly normal bCNCs are significantly smaller than controls, supports the fact that the use of specific and reproducible measures of the temporal bone have been consistently demonstrated to identify bony labyrinth abnormalities that are missed by visual inspection alone. In this instance, abnormalities were defined as statistically significant differences relative to controls in ears that did not visibly demonstrate an abnormality, and therefore, these results were not sorted by specific cutoff values reported in the literature. All ears contralateral to the bCNC stenosis were ≥1.6 mm. The findings suggest that factors affecting the stenotic bCNC side are likely bilateral in nature, although to potentially lesser degrees, and this concept is supported by the current data. In addition, the discovery of more widespread biometric abnormalities affecting a greater number of structures than the bCNC alone, suggests that abnormal signaling likely occurs very early in embryogenesis during cochleovestibular development. This is akin to previous findings in syndromic and nonsyndromic individuals with sensorineural hearing loss.

Fig. 1. CT of CNC stenosis. (a) Axial CT image in a patient with unilateral deafness demonstrates normal-sized right CNC (arrow) and bone island of the LSCC (*). The modiolus is normal in size. (b) Axial CT image in the same patient shows a stenotic CNC (arrow), a smaller cochlea and modiolus, but a normal LSCC bone island. (c) Axial CT image in a different patient shows a slightly smaller, but still normal, CNC, a smaller modiolus and cochlea, and a smaller LSCC bone island. These were initially called normal prior to measurement. (d) In the same patient, the axial CT image shows a critically stenotic left CNC (arrow) with a bony bar, a very small modiolus, and LSCC bone island, which is moderately reduced in size. CNC = cochlear nerve canal; CT = computed tomography; LSCC = lateral semicircular canal.

Fig. 2. Audiometric results. Pure-tone audiometric hearing thresholds for the better (normal) hearing ear (A) and poorer (hearing-impaired ear) hearing ear (B).
loss.9–12 Children presenting with UHL associated with bCNC stenosis appear to have deficits in balance,23 thus providing further clinical support for a more widespread anatomical anomaly involving the vestibular apparatus in this population. The vestibular aqueduct porus width was larger bilaterally compared with controls, but there was no difference in porus width across critically stenosed and contralateral ears. This further supports the hypothesis of bilateral signaling during early embryogenesis. There was no statistical difference between the size of the vestibular aqueduct on the side of the bCNC stenosis as compared with controls. Prior publications have demonstrated that the porus of the vestibular aqueduct may be enlarged without concomitant enlargement of the remainder of the vestibular aqueduct.9–12 The findings suggest that the porus and the vestibular aqueduct may develop separately from each other during embryogenesis.

The degree of hearing loss at presentation was profound or severe in 89% of ears with bCNC stenosis and normal in 100% of contralateral ears. Hearing loss in the stenotic bCNC is not surprising, because bCNC width has been shown to be inversely proportional to audiometric data, with bCNCs less than 1.4 mm portending a pure-tone average worse than 70 dB HL.18 However, normal hearing in the contralateral ear demonstrates that although this normal measures smaller than controls, it has not crossed the threshold of functionality to cause hearing loss. There have been reports of patients with bCNC stenosis who have only mild to moderate hearing loss.24 In these instances, it has been postulated that a small number of cochlear nerve fibers remain, and that they are able to transmit action potentials required for hearing.7

A limitation of this study is that not all patients had a complete hearing assessment and MRI. Additionally, patients were recruited based on a radiology report text search using keywords “unilateral hearing loss” or “cochlear nerve canal stenosis,” which may have overestimated the proportion of abnormal findings in this population. Nevertheless, this large study provides the first evidence that children with unilateral bCNC stenosis have abnormal biometry of both the cochlea and the vestibular end-organ in the affected and the normal contralateral ear.

CONCLUSION

Isolated bCNC stenosis is a relatively common cause of unilateral deafness in childhood. We identified additional biometric abnormalities that widely affect both the deaf and the normally hearing ear in our cohort. The presence of biometric abnormalities in the contralateral side suggests that even in a grossly normal nonsyndromic ear, developmental anomalies occur. We hypothesize that this is likely due to abnormal signaling affecting both ears during the early embryological period.

BIBLIOGRAPHY