

Frequent Association of Cochlear Nerve Canal Stenosis With Pediatric Sensorineural Hearing Loss

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Objective: To examine the association between cochlear nerve canal (CNC) dimensions and sensorineural hearing loss (SNHL).

Design: Retrospective review.

Setting: Tertiary pediatric hospital.

Patients: Children with SNHL and CNC stenosis.

Intervention: The CNCs measured in axial and 45° oblique planes on temporal bone computed tomography (TBCT) in children with SNHL were compared with TBCT from children with normal hearing and 100 normal temporal bone specimens. Additional inner ear abnormalities were recorded. Hearing was measured using 4 frequency pure-tone averages (PTAs).

Main Outcome Measure: The degree of CNC stenosis related to the degree of SNHL.

Results: Fifty-three patients (32 female) with SNHL had CNC stenosis in 85 ears (32 bilateral, 21 unilateral). The mean (SD) axial CNC measurement for 85 ears was 0.98 (0.57) mm (range, 0-1.75 mm). The mean (SD) Poschl CNC measurement was 1.30 (0.69) mm (range, 0-2.80 mm). Of 85 ears, 64 had at least 1 additional inner ear abnormality. The mean (SD) PTA was 56.2 (40.8) dB. For each ear separately axial and Poschl plane CNC measurements were highly correlated ($P < .001$). The degree of CNC stenosis was significantly ($P = .02$) related to degree of hearing loss, and PTA decreased in the CNC stenosis population by 1.4 dB per year ($P = .054$). In addition, PTA and additional inner ear abnormalities were found to be significantly correlated ($P = .002$).

Conclusions: Cochlear nerve canal stenosis is associated with SNHL, and the degree of stenosis predicted the degree of SNHL. In addition, the presence of CNC stenosis with additional inner ear abnormalities may affect the severity of SNHL.

Arch Otolaryngol Head Neck Surg. 2012;138(4):383-388

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THE COCHLEAR NERVE CANAL (CNC), also called the cochlear aperture, bony cochlear nerve canal (BCNC), and cochlear foyette, is the bony transition point between the internal auditory canal (IAC) and the cochlear modiolus. With the development of high-resolution computed tomography (CT) and magnetic resonance imaging (MRI), the CNC can be readily identified. Temporal bone imaging is routinely used in the diagnosis of sensorineural hearing loss (SNHL).¹⁻⁵ Researchers have started to associate stenosis of the CNC with possible cochlear nerve (CN) hypoplasia or aplasia.⁶⁻¹¹ The underdevelopment or absence of the CN is an established cause of SNHL, and patients with such underdevelopment often do not derive clinically significant benefit from cochlear implantation (CI).^{12,13} Papsin¹⁴

found that the outcome of CI in patients with IAC or cochlear canal narrowing was worse compared with outcomes of children who did not have these abnormalities. This raises the question about whether CNC narrowing should be recognized as a marker for CN hypoplasia/aplasia.

Although it is recognized that hypoplasia/aplasia of the CN is associated with profound hearing loss, to our knowledge, the degree of hearing loss found in association with CNC stenosis has not been studied. In all previous accounts relating CNC stenosis to SNHL, the degree of hearing loss was either not described or only patients with at least severe to profound hearing loss were included in the study.^{6,8-10,15}

The current study aims to examine the audiologic profiles of patients diagnosed as having CNC stenosis to better understand how the malformation is related to

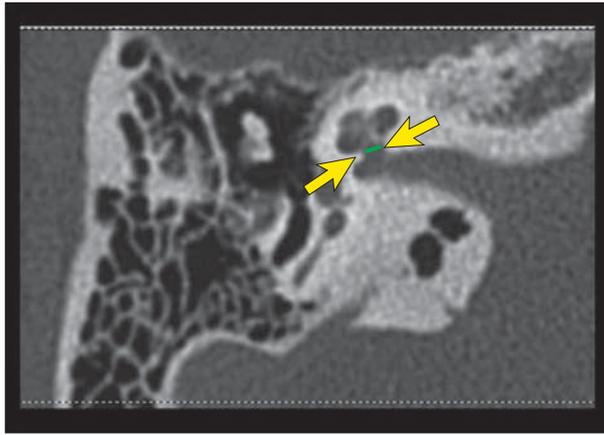


Figure 1. Axial cochlear nerve canal (CNC) measurement (dashed line and arrows).

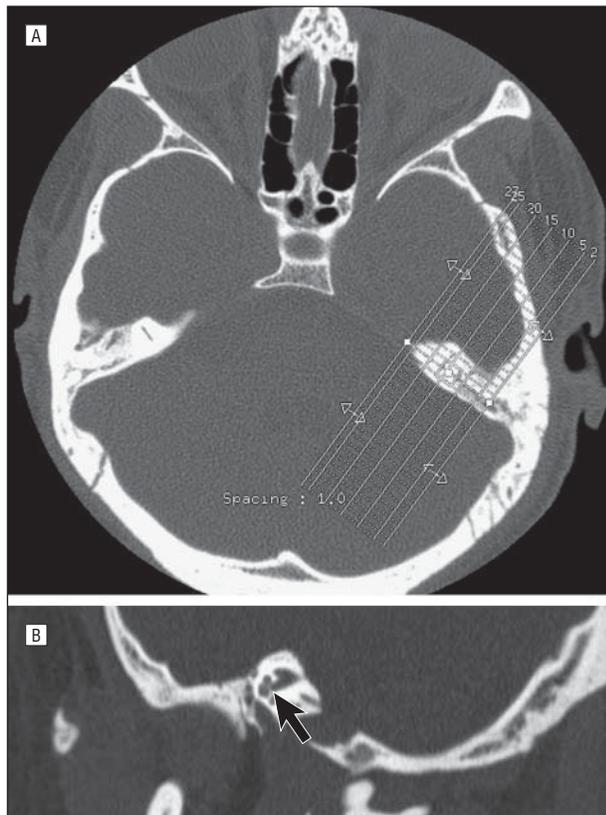


Figure 2. How to make an image in the Poschl plane. A, The angle of reformat on the axial image. B, The cochlear aperture on the Poschl image (arrow).

hearing loss. In addition, we studied the association of CNC stenosis plus additional inner ear anomalies with the patient's degree of hearing loss.

METHODS

PATIENTS

We evaluated children 8 days to 13 years old who presented with SNHL to the otolaryngology outpatient department at Children's Hospital Boston, Boston, Massachusetts, from January 1995 through June 2010, who underwent an axial temporal bone

CT (TBCT) study and who had a narrowed CNC identified. Any patients with the diagnosis of CHARGE (coloboma [eye], heart defects of any type, atresia [choanal], retardation [of growth and/or development], genital anomaly, and ear anomaly) syndrome were excluded because, as a group, they had additional potential causes of SNHL not directly related to abnormal inner ear anatomy, including prematurity, ototoxicity, and respiratory failure.

Children's Hospital Boston institutional review board approval was obtained before beginning the study.

DATA COLLECTION

We evaluated the clinical history, TBCT images, and any MRI and the audiologic data of these children. All data were collected from patient medical records and stored and managed using the REDCap electronic data capture tools hosted by Children's Hospital Boston.¹⁶

CT SCANNING TECHNIQUE AND REFORMATS

Noncontrast TBCT was performed as part of the workup of all the patients in our study population. The CT techniques varied across the study period. Scanning was performed either on a 16-channel multidetector CT (MDCT) scanner (LightSpeed; GE Healthcare, Waukesha, Wisconsin) or a 64-channel MDCT scanner (Sensation 64; Siemens Medical Solutions, Erlangen, Germany). On the 16-section multidetector row GE Light-Speed CT scanner, images were acquired at a 0.63-mm section thickness and reconstructed in an axial plane at 0.63 mm using a bone algorithm at 0.30-mm intervals.

On the Siemens Sensation 64 scanner, axial images were acquired with helical technique with a section thickness of 0.60 mm, and the image data set was reconstructed at 0.75 mm in a plane parallel to the lateral semicircular canals.

The axial data for every case were then transferred to a separate workstation for postprocessing by a single pediatric neuroradiologist (S.P.P.), with commercially available 3-dimensional reformatting software (Voxar 3D; Toshiba, Edinburgh, Scotland). Reformatted images were first made in the axial plane in all cases (parallel to the axis of the cochlea on the sagittal view) with a section thickness of 0.75 mm (**Figure 1**). Then, 45° oblique reformats with 0.75-mm thickness were acquired perpendicular to the petrous pyramid and parallel to the superior semicircular canal. This plane is often referred to as the Poschl plane and is perpendicular to the CNC.^{17,18} To obtain this reformat, the axial plane was established by connecting the 2 dots of the lateral semicircular canal. Following this, the apex of the superior semicircular canal (SSCC) was identified on the axial images (**Figure 2**). The 45° oblique reformat was obtained by connecting the line parallel to the long axis of the summit of the SSCC. This line was made as parallel as possible to the axis of the summit of the SSCC.

MEASUREMENT OF THE CN APERTURE

Images reconstructed in the axial and the Poschl plane were sent to the Fuji PACS workstation, and measurements were carried out using the standard bone window (window width, 3000 HU; center, 350 HU [Hounsfield units]) on a Fuji PACS workstation by the reporting neuroradiologist (S.P.P.).

The diameter of the CNC was measured on the axial images in each ear along the inner margin of its bony walls at its mid portion on an axial image through the base of the modiolus (**Figure 2**). The supero-inferior distance between the walls

of CNC was measured on the Poschl plane reformat images through the cochlea in each ear (**Figure 3**).

In cases in which the CNC appeared to be completely stenosed, no measurements were possible, and this was recorded as an obliterated CNC, or 0 mm.

Cochlear nerve canals with axial measurements less than 1.76 mm were considered stenotic. In a 2011 IJPORL study,¹⁹ we found that 1.76 mm was 2 SDs below the mean for CNCs in normal ears. In addition, other studies⁷⁻¹⁰ have indicated that a measurement of 1.5 mm and 1.8 mm were considered to be the upper limit of normal. Please see the “Comment” section for further support for these measurements as noted in other articles.

For this specific report, all images were read twice; once by the original reading radiologist (one of several on staff at the hospital) as part of routine clinical care, and then again by the one of us (S.P.P.). Specific measurements were made by the reporting radiologist, the study radiologist (blinded to the original measurement and the audiograms), and an otolaryngology physician (M.A.K.) and the recorded measurements were therefore validated for interobserver variation.

AUDIOLOGIC DATA

First (baseline) and most recent audiologic information for each patient was collected to determine possible progression of hearing loss. Four-frequency (500, 1000, 2000, and 4000 Hz) pure-tone averages (PTAs) were collected whenever possible; however, 3-frequency PTAs (500, 1000, and 2000 Hz) were used in the absence of a 4-tone PTA. In patients who did not have reliable behavioral audiometric data available, frequency-specific auditory brainstem response (ABR) information was collected. For patients with only soundfield data available from behavioral testing, soundfield PTAs were only included in analyses if the patient was shown to have bilateral symmetrical hearing loss via ABR testing.

STATISTICAL ANALYSIS

Statistical analysis was performed using SAS software (version 9.2; SAS Institute Inc). The 85 ears used in analysis were from 53 patients. For each ear, 2 PTA numbers were recorded when available: baseline PTA and the most recent PTA. To capture both the correlation structure between the left ear and right ear and the change of PTA over time, a mixed model was used with random intercept and fixed effects of follow-up time, demographic variables, CNC measurements, and additional abnormalities. Age and sex were analyzed and excluded from the final model owing to the large *P* value (*P* = .13 and *P* = .96, respectively), and interaction terms were also tested and then deleted because they were not significant (*P* > .30 for all comparisons). Both baseline PTA and follow-up PTA were combined as 1 outcome variable in the mixed model. Patient identification and follow-up time were both used in the mixed model to identify the baseline and follow-up PTA for each patient.

RESULTS

A total of 53 patients (85 ears) had CNC stenosis; 32 patients (60%) had bilateral CNC stenosis, and 21 (40%) had unilateral CNC stenosis. Thirty-two (60%) were female. The mean (SD) age was 8 (3.7) years, and the mean age at the first audiogram was 4.8 (3.7) years. The median follow-up time between audiograms was 2.0 years (range, 5 days–11 years). Of the total 85 ears with CNC stenosis, 63 had useable behavioral audiograms for in-

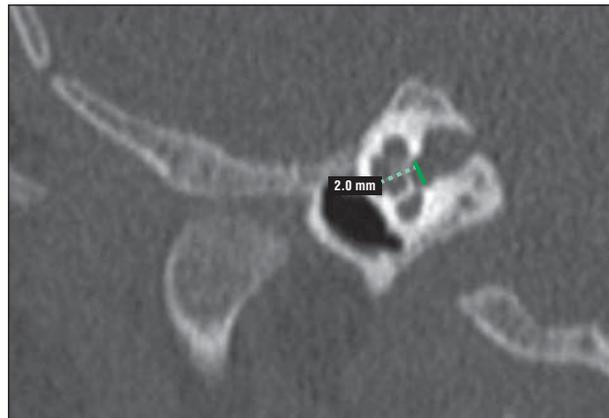


Figure 3. Poschl plane (dashed line) cochlear nerve canal (CNC) measurement.

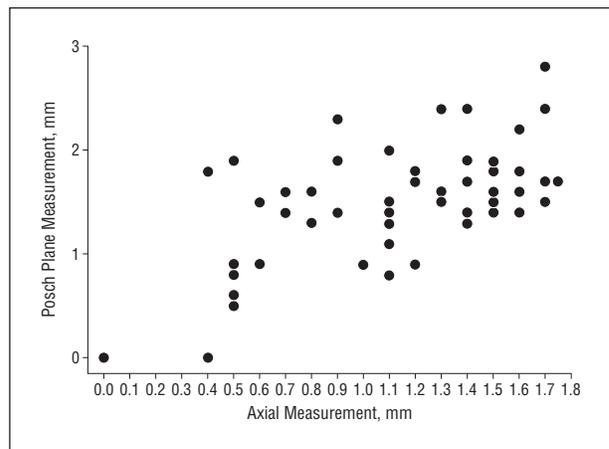


Figure 4. Correlation of axial and Poschl plane cochlear nerve canal (CNC) measurements, indicated by dots.

dividual ears, 20 had ABR data only, and 2 patients had only soundfield data available. There was a wide range in hearing for the 63 ears with PTAs, ranging from 2.5 to 130.0 dB. The mean PTA for the 63 ears was 56.2 (40.8) dB. Twenty ears had only ABR data available, and they were excluded from the PTA analysis. Twelve ears had a measurement of 0 mm on the axial scan, indicating an obliterated CNC. Four of these ears had behavioral data, with a mean PTA of 92.5 dB (range, 76.2–116.2 dB). The 8 other 0-mm ears had only ABR data, and the range across frequencies was 65 to 110 dB. These eight 0-mm ears with ABR data only were excluded from analyses. Pure-tone averages (either 3- or 4-frequency) in ears that had only mild, high-frequency HL were occasionally in the normal range because the mild degree of SNHL at 4000 or 8000 Hz was not enough to put the PTA into the abnormal range. Other diagnoses were Mosaic trisomy 22 (1 patient), Goldenhar syndrome (1 patient), and Townes-Brock syndrome (1 patient).

The median axial CNC measurement for all 85 ears was 1.10 mm (range, 0–1.75 mm). The median Poschl plane measurement was 1.50 mm (range, 0–2.80 mm). The axial and Poschl CNC measurements were highly correlated (Pearson correlation = 0.78; *P* < .001) (**Figure 4**). For further analysis, ears with CNC steno-

Table 1. Axial Measurements and PTA^a

Axial Measurements, mm	Ears, No. (%)	PTA, Mean (SD), dB
0.0	4 (6)	95.7 (16.8)
0.5-1.0	17 (27)	73.1 (37.2)
>1.0-1.7	42 (67)	49.7 (36.7)

Abbreviation: PTA, pure-tone average.

^a $P = .009$.

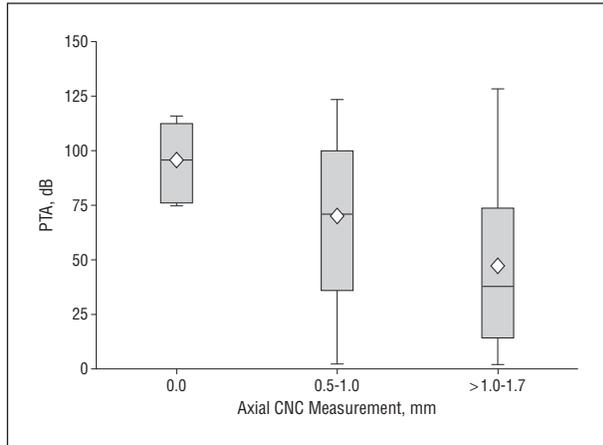


Figure 5. Correlation between cochlear nerve canal (CNC) measurements and pure-tone average (PTAs). The upper edge of the box is the third quartile, the upper bar is the largest observation, the lower edge is the first quartile, and the lower bar is smallest observation.

Table 2. Additional Inner Ear Anomalies and PTA^a

Additional Abnormalities	Ears, No. (%)	PTA, Mean (SD), dB
No	19 (30)	36.4 (26.5)
Yes	44 (70)	69.4 (38.5)

Abbreviation: PTA, pure-tone average.

^a $P = .002$.

Table 3. Multivariate Analysis for PTA

Covariates	Increase of PTA, Mean (SE), dB	P Value
Axial measurements, mm		
0.0	37.3 (18.4)	.03
0.5-1.0	21.5 (10.2)	
>1.0-1.7	1 [Reference]	
Additional abnormalities		
Yes	27.0 (10.1)	.01
No	1 [Reference]	
Follow-up time	1.4 dB/y	.054

Abbreviation: PTA, pure-tone average.

sis and behavioral PTA data were split into 3 groups depending on axial measurements. We found that smaller axial measurement is associated with a more profound degree of hearing loss ($P = .02$). These results can be found in **Table 1** and **Figure 5**.

Sixty-four of the 85 ears (75%) had at least 1 additional inner ear abnormality. Abnormalities (hypoplasia) of the modiolus, cochlear turn abnormalities (absent division between the apical and middle turn), and vestibular aqueduct enlargement were the commonest associated anomalies (after omitting the CHARGE syndrome cases).

Eighteen of 85 ears (21%) had 1 or 2 additional inner ear abnormalities, and 46 of 85 ears (54%) had 3 or more inner ear abnormalities in addition to CNC stenosis. For the 63 ears with PTA data available, the number of additional inner ear anomalies (1 and 2 vs ≥ 3) and PTA were not found to be significantly correlated ($P = .88$), but the children with additional abnormalities had a higher PTA compared with those without any additional abnormalities ($P = .002$) (**Table 2**). Of the 64 ears with additional inner ear anomalies there were 54 ears (64%) with an abnormality of the cochlea:

Location	Ears, No. (%)
Cochlea	54 (64)
Modiolus	40 (47)
Apical cochlear turn	28 (33)
Middle cochlear turn	23 (27)
Basal cochlear turn	10 (12)
Vestibular aqueduct	22 (26)
Internal auditory canal	19 (22)
Vestibule	19 (22)
Semicircular canals	18 (21)
Middle ear	17 (20)
Ossicles	12 (14)
External ear	9 (11)

Multivariate analysis showed that the PTA threshold correlated with the axial measurement of the CNC ($P = .02$), and with presence or absence of additional abnormalities ($P = .048$). It was also found that overall, PTA worsened by 1.4 dB per year ($P = .054$) for all ears with CNC stenosis (**Table 3**).

COMMENT

In the literature, the CNC goes by a variety of names, including cochlear aperture, BCNC, and cochlear fessette. In addition, there is some variation in what is considered to be normal vs narrow width of the CNC. However, the literature does agree on 2 points: (1) that patients with SNHL are more likely to have narrower CNCs than patients with normal hearing, and (2) a significantly narrowed CNC is often associated with hypoplasia or aplasia of the CN. Most of the previous studies looking at CNC stenosis have evaluated patients with hearing loss in the profound range. However, we have found a wide range of hearing in our patients with CNC stenosis, from normal hearing to profound hearing loss.

Fatterpekar et al²⁰ were the first to measure the CNC on CT images. They studied TBCT scans in 50 patients with normal hearing and found the mean (SD) width of the CNC to be 2.13 (0.44) mm. Fatterpekar et al⁶ subsequently compared the CNC diameter on CT in patients with profound bilateral SNHL with those with normal hearing. The authors found that the cochlear apertures of the patients with SNHL were significantly smaller (1.82 [0.24] mm) than the cochlear apertures of the controls

Table 4. Temporal Bones in Patients With Normal Hearing

Source	Temporal Bones Studied, No.	Technique	Slice	CNC Width, Mean (SD), mm
Fatterpekar et al, ²⁰ 1999	100	CT	1 mm	2.13 (0.44)
Stjernholm and Muren, ²¹ 2002	100	CT	1 mm	1.91 (0.24)
Henderson et al, ¹⁹ 2011	110	Harvested temporal bones	2- μ m slide	2.26 (0.25)

Abbreviation: CNC, cochlear nerve canal.

Table 5. Temporal Bones (TBs) in Patients with Hearing Loss

Source	TBs Studied, No.	Technique	Slice, mm	CNC Dimensions, Mean (SD), mm		
				Width	Length	Height
Fatterpekar et al, ⁶ 2000	100	CT	1	1.82 (0.24)	0.74 (0.17)	NA
Current study	85	CT	1	0.98 (0.57)	NA	1.30 (0.69)

Abbreviations: CNC, cochlear nerve canal; NA, not applicable.

(2.14 [0.44] mm). The authors proposed that hypoplasia of the cochlear aperture may be a contradiction to cochlear implantation because it could indicate a hypoplastic CN.⁶

Stjernholm and Muren²¹ measured the diameter of the CNC in 117 silicon casts of temporal bones and concluded that the mean (SD) width of the aperture is 2.58 (0.28) mm. They used these measurements to recommend guidelines for abnormally narrow CNCs (<1.4 mm) and abnormally wide CNCs (>3 mm). To further establish how closely TBCT CNC measurements approximate those of actual temporal bones, our group measured the diameter of the CNC in 110 temporal bone specimens from typically hearing humans, from birth to age 100 years.¹⁹ The mean diameter was found to be 2.26 mm, and an abnormally narrow CNC was 2 SDs below this average, or less than 1.76 mm (Henderson et al¹⁹). These results are quite close to those of Fatterpekar et al^{6,20} and slightly narrower than those of Stjernholm and Muren.²¹ At least some of the reported variation in axial diameter may be accounted for by site of measurement, slice thickness of CT images, indirect measurement of the CNC using silicon, and measurement of actual temporal bones compared with radiologic images of temporal bones.

Adunka et al⁸ explored the interaction between a stenotic cochlear aperture and the auditory nerve that runs through it. The authors considered an abnormally small cochlear aperture to be less than 1.4 mm in width. In 12 ears with absent CNs based on MRI, but normal-size IACs, 8 ears (67%) had small cochlear apertures. Adunka et al⁸ concluded that identifying a narrowed cochlear aperture can indeed indicate hypoplasia or aplasia of the nerve that runs through it, although in children with profound hearing loss the presence of a normal CNC does not guarantee that the nerve is present. This was also found in another study that examined patients with unilateral SNHL.⁹ Measuring CT scans of these patients, Kono⁹ suggested that a transverse diameter of less than 1.7 mm or a coronal measurement of less than 1.8 mm represented

statistically significant narrowing ($P > .01$) when compared with the same measurements in normal hearing ears. More recently, Miyasaka et al¹⁰ collected both CT scans and MRI images from a group of patients with unilateral and bilateral SNHL. In their study, 8 ears were discovered to have CN hypoplasia or aplasia, and all of these ears were confirmed to have cochlear aperture stenosis (defined as <1.5 mm).¹⁰ From these results, they concluded that identifying cochlear aperture stenosis on CT should prompt MRI to look for the presence of CN hypoplasia, especially if the hearing loss is in the severe to profound range and cochlear implantation is being considered. Although the primary reason for MRI would be to assess the size of the seventh nerve, it might also be useful to assess the anatomy of the origin of the eighth nerve and any asymmetry of the brainstem. The other reason to obtain the MRI is the higher incidence of hind-brain malformations in patients with bilateral CN deficiency as indicated in prior studies.²²

To our knowledge, the current study is the first to investigate the association between the degree of cochlear stenosis and the degree of hearing loss. As stated herein, the current study revealed a wide range in hearing loss, as well as a wide range in CNC stenosis diameter. We have found that as the axial measurement of the CNC decreases, the amount of hearing loss increases. Because most of our participants did not have MRI of the temporal bone performed, correlation to abnormalities of the CN, if any, could not be made. Further investigation using both imaging techniques in conjunction with good behavioral hearing tests and measurement of the bony structures would help to provide more insight into why hearing is decreased as the width of the CNC is decreased (Figure 1).

Another finding of the current study was that the presence of additional anatomic abnormalities is also associated with a higher degree of hearing loss. Additional inner ear anomalies in association with CNC stenosis has been touched on in the literature, and although Kono⁹ and Fatterpekar et al⁶ presented cases of patients who had

no other demonstrable inner ear anatomical abnormalities, others have found that that CNC stenosis often had additional associated anatomical abnormalities.⁹ Adunka et al⁸ found that of the 8 ears identified as having narrowed apertures, 3 ears (38%) had additional inner ear malformations. A more recent study performed by Pagarkar et al¹⁵ found that 84% of their patients with CNC stenosis had associated cochlear or vestibular anomalies. Multiple abnormalities in conjunction with a stenotic aperture were also presented by Morimoto et al²³ in a number of patients with CHARGE syndrome. They found that 20 of the 26 ears studied by CT had abnormally thickened bony covering over the aperture for the CN, giving the cochlea an isolated appearance, termed “trapped cochlea.” In addition, in all 20 ears there were other cochlear anomalies, including enlarged vestibular aqueducts, narrow IACs, dysplastic modiolus, and hypoplastic cochlear turns. For this reason, we specifically omitted any patients with a clinical or genetic diagnosis of CHARGE association. Miyasaka et al¹⁰ reported that of the 8 ears they found to have cochlear aperture stenosis, 4 of them had additional inner ear abnormalities. We found that 64 of 85 ears (75%) had at least 1 additional inner ear abnormality, 18 of 85 ears (21%) had 1 or 2 additional inner ear abnormalities, and 46 of 85 ears (54%) had 3 or more inner ear abnormalities in addition to CNC stenosis. In addition, those with additional anatomical abnormalities tend to have a more substantial hearing loss ($P=.002$). Both the degree of CNC stenosis and the presence of additional abnormalities contribute to the hearing loss in the affected ear (**Table 4**). A summary of the literature findings in comparison with the current study results can be found in Table 4 and **Table 5**.

In conclusion, CNC stenosis is associated with SNHL that ranges from near-normal to profound. Furthermore, there is a statistically significant relationship between the degree of hearing loss and the degree of stenosis. In addition, more severe degrees of hearing loss are also correlated with CNC stenosis plus additional structural abnormalities. Additional inner ear anomalies may occur in conjunction with CNC stenosis more often than previously appreciated.

Submitted for Publication: September 22, 2011; final revision received January 11, 2012; accepted February 2, 2012.

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Author Contributions: Ms Wilkins and Drs Prabhu, Ogando, and Kenna had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. *Study concept and design:* Wilkins, Prabhu, and Kenna. *Acquisition of data:* Wilkins, Prabhu, Ogando, and Kenna. *Analysis and interpretation of data:* Wilkins, Prabhu, Huang, and Kenna. *Drafting of the manuscript:* Wilkins, Prabhu, Huang, Ogando, and Kenna. *Critical revision of the manuscript for important intellectual content:* Wilkins, Prabhu, and Kenna.

Statistical analysis: Huang and Kenna. *Administrative, technical, and material support:* Wilkins, Prabhu, and Kenna. *Study supervision:* Wilkins and Kenna. **Financial Disclosure:** None reported.

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