Cochlear Nerve Deficiency in Unilateral Pediatric Auditory Neuropathy Spectrum Disorder

UNC DEPARTMENT OF OTOLARYNGOLOGY/ HEAD AND NECK SURGERY

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Background

The purpose of this study is to determine the incidence of cochlear nerve deficiency (CND) in pediatric patients with auditory neuropathy spectrum disorder (ANSD) in the setting of single-sided deafness (SSD).

ANSD refers to a spectrum of hearing impairment presenting with abnormal auditory brainstem response testing (ABR) in the setting of normal outer ear function. The site of dysfunction within the auditory pathway varies, with various congenital and acquired etiologies associated with the diagnosis. Pure tone audiometry can reflect a wide range of hearing levels from mild to profound hearing loss.

Patients with ANSD often have disproportionately impaired speech perception, often out of proportion to hearing loss on pure tone audiometry. Importantly, in patients with severe-to profound hearing loss cochlear implantation is typically considered management of choice. However, it is important to note that in patients with cochlear nerve deficiency (aplasia or hypoplasia), cochlear implant has been shown to have limited benefit for hearing rehabilitation. There is limited literature evaluating the correlation between ANSD and cochlear nerve deficiency (CND), particularly in the pediatric population with unilateral hearing loss. This is particularly salient as cochlear implantation for SSD has recently been approved by the FDA, and has important implications for management of hearing in pediatric patients with severe-to-profound SSD.

Methods

The study is a retrospective chart review of pediatric subjects <18 years with single-sided deafness (SSD) from a single tertiary care institution from January 2014 to October 2019.

Subjects with unilateral severe-to-profound hearing loss were included if they had both magnetic resonance imaging (MRI) and auditory brainstem response (ABR) testing available. Severe-to-profound hearing loss was defined as 3-frequency pure tone average (PTA) thresholds at 500, 1000 and 2000 Hz of 65 dB HL or poorer, high frequency hearing loss of 70 dB HL or poorer, or aided word recognition less than 60%. The contralateral ear had normal or near-normal hearing, with PTA of 20 dB HL or better.

The incidence of ANSD was assessed based on ABR, defined as no neural responses with normal otoacoustic emissions or cochlear microphonics. The incidence of CND was determined from imaging review by a neurotologist. Fine-cut T2-weighted MRI brain sequences were reviewed in the axial and parasagittal planes. Cochlear nerve status on MRI was defined as: 1) within normal limits (WNL; nerve size consistent with the other nerves in the ipsilateral internal auditory canal (IAC) and the contralateral cochlear nerve), 2) hypoplastic (nerve smaller than the other nerves in the IAC), or 3) aplastic (nerve absent).

The pure tone average (PTA) at 500, 1000 and 2000 Hz of subjects within the ANSD population with and without CND was calculated.

Statistical analysis was performed using SPSS (version 26). A Mann-Whitney U test was run to determine if there were differences in PTA between those with CND and those with normal cochlear nerves.

Results

103 pediatric subjects with SSD with both available ABR and fine-cute T2-weighted MRI were included.

The incidence of ANSD in this population was 30.0% (31 subjects).

Within the group of subjects with ANSD, 77.4% had CND on imaging (24 subjects) and 22.6% had normal cochlear nerves (7 subjects). Of the CND group, 18 nerves were aplastic and 6 were hypoplastic.

In subjects with ANSD, there was no statistically significant difference in median PTA between ears with CND (M = 98.3) versus normal cochlear nerves (M = 85.0; U = 69.0, z = -0.721, p = 0.502).

	CND	WNL
Ear		
Left	13 (54.2%)	5 (71.4%)
Right	11 (45.8%)	2 (28.6%)
Associated inner ear malformation	3 (12.5%)	0 (0%)
Associated syndrome	4 (16.7%)	1 (14.3%)

Table 1. Demographics of study population for cochlear nerve deficiency (CND) and nerves within normal limits (WNL) [n (%)].

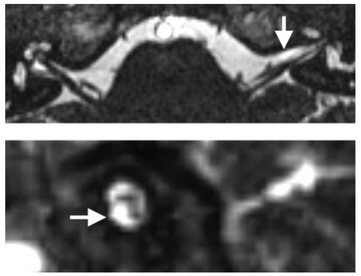

Figure 1. Left sided cochlear nerve aplasia on T2-weighted MRI brain imaging. On axial view there is no nerve visualized entering the cochlea (white arrow). On parasagittal view only three nerves are visible, the cochlear nerve is not present in the anteroinferior quadrant of the IAC (white arrow) *(Figure from Vos et al., 2022).*

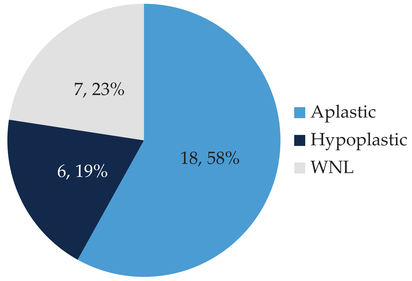

Figure 2. Status of ipsilateral cochlear nerve in pediatric patients with SSD and ANSD (n, %).

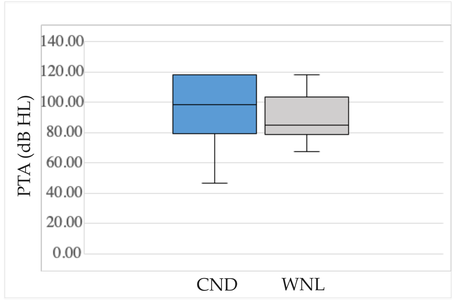

Figure 3. Median PTA (average at 500, 1000 and 2000 Hz in dB HL) of individual ears with CND versus nerves within normal limits (WNL) on imaging.

Conclusions

This study demonstrates that there is a high incidence of ANSD in pediatric patients with single-sided deafness (30%). The majority of ears with ANSD (77.4%) also demonstrated cochlear nerve deficiency on imaging. The literature reports limited data on rates of cochlear nerve deficiency, however reported rates of CND in pediatric patients with unilateral ANSD range from 9-46%. This study demonstrates that the majority of nerves (75%) were aplastic rather than hypoplastic.

While a higher median PTA was identified in ears with CND, there was no statistical significance between median PTA in ears with CND and with normal nerves. Future analysis with a larger sample size is needed to assess the correlation of PTA in ears with both ANSD and CND, however there is currently no evidence to suggest that PTA can be used as a marker for cochlear nerve status in unilateral ANSD patients. Of note, no unilateral ANSD patients in this cohort underwent cochlear implantation regardless of nerve status.

Our study demonstrates a higher rate of cochlear nerve deficiency in patients with ANSD than previously reported in the literature. Particularly in the setting of expanding indications for cochlear implantation for SSD, it is imperative to pursue thorough audiologic and radiographic work-up for pediatric patients in this population to fully assess nerve status as CND can be considered a contraindication to implantation.

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