Pediatric Unilateral Hearing Loss: An Analysis of Progression and Associated Risk Factors

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Abstract

Objective: To elucidate common risk factors of unilateral pediatric hearing loss, and explore the prevalence of disease progression.

Methods: A retrospective study was performed to investigate pediatric patients seen at a tertiary pediatric otolaryngology clinic for unilateral sensorineural hearing loss (USNHL) over a ten-year period. Through clinical chart and audiogram review, patients with SNHL were grouped by severity. Radiographic imaging results were also reviewed.

Results: 79 patients were identified with USNHL. 51 (65%) were determined to have severe to profound hearing loss (SPHL), and 28 (35%) had mild to moderate hearing loss (MMHL). Identifiable risk factors were determined in thirteen and ten patients, respectively. Prematurity was the most common risk factor identified, followed by a NICU stay. CT Temporal Bone or MRI radiographic results were available for 32 patients with SPHL, and 16 patients with MMHL. An identifiable cause was seen in 17% of cases. Enlarged vestibular aqueducts were the most commonly identified radiographic pathology, followed by cystic cochlea. Patients were also evaluated for progression of severity, or progression from unilateral to bilateral hearing loss. 5.9% of patients with SPHL progressed to bilateral hearing loss. Progression of severity and progression to becoming bilateral was seen in 23.5% of patients originally diagnosed with MMHL.

Conclusion: Pediatric hearing loss can be attributed to many factors. Risk factors can be identified in about 30% of cases, and imaging can identify about 20% of cases. Follow-up is essential as progression was observed in both severity and becoming bilateral from an original unilateral diagnosis.

Introduction

Congenital unilateral hearing loss (UHL) affects up to 1-3 newborns per 1,000, and about 3% of school-aged children.[1,2] Historically, UHL has been more difficult to diagnose early, as speech and language consequences may not be as prominent early in development. With the better understanding of the sequelae of UHL, increased screenings and surveillance have led to earlier detection, and a more comprehensive understanding of the causes of pediatric UHL. Furthermore, it has been suggested that CT or MRI be performed on children with recently diagnosed UHL, to identify anatomic causes of hearing loss.[3]

It has been found that UHL in children can lead to consequences that extend beyond hearing impairment. Educational delays are common, with 22-35% of children needing to repeat the first grade, and 12-41% requiring educational assistance[4]. It was also shown that in a competitive noise environment, monaural listeners do not function as well.[3,6] As anatomic abnormalities can also extend to the vestibular system, children may also experience vertigo, leading to further physical and emotional distress.[5]

Many risk factors have been associated with pediatric congenital hearing loss, as reported in the 2007 JCIH position statement, including the following: family history of permanent childhood hearing loss, NICU stay of greater than five days, history of ECMO or assisted ventilation, exposure to ototoxic medications or loop diuretics, hyperbilirubinemia. in-utero infections such as CMV, rubella, herpes, syphilis, and toxoplasmosis, craniofacial anomalies, specific physical findings (e.g. white forelock), specific syndromes or neurodegenerative disorders associated with hearing loss, culture positive postnatal infections (especially herpes viruses and varicella) meningitis, head trauma, and chemotherapy.[12] Because of their associations with hearing loss, the JCIH recommends a comprehensive review of these risk factors during early age well visits, and heightened surveillance for hearing loss.

Other correlations with pediatric UHL have been determined outside of the above listed risk factors. UHL can be caused by genetic aberrations,[7] and can be associated with ophthalmologic pathologies, including myopia and exotropia, and a myriad of radiologic anomalies visible on temporal bone CT scans or MRI.[3]

Pediatric UHL has also been found to demonstrate progression. In 2009, Uweira et al determined the rates of progression in the ipsilateral ear, and also to becoming bilateral. They showed a 53% rate of ipsilateral progression, and 10% new-onset hearing loss in the contralateral ear.[8] This study suggested that in some cases, UHL may not, in fact, be a unilateral process, but rather the initial manifestation of a bilateral process. It also reiterated the importance of surveillance and follow-up for those children diagnosed with UHL

The purpose of this study is to further explore the prevalence of risk factors that contribute to USNHL in newborns, identify common radiologic findings in those with USNHL, and to determine the prevalence of progression, both in severity and bilaterality.

Materials and Methods

A retrospective chart review was performed of an outpatient pediatric otolaryngology tertiary care center of patient charts from 2004-2014. Patients were selected using ICD-9 and ICD-10 codes for hearing loss. Patient charts were then analyzed, and those with a diagnosis of USNHL were selected. All patients with any form of conductive hearing loss were excluded.

79 patients were identified with USNHL. Once identified, charts were examined for audiometric data, radiographic imaging studies and historical documentation of risk factors. Audiogram results were documented as mild to moderate or severe to profound based on the majority of the hearing loss. The mild to moderate hearing loss was characterized as 20-60 dB loss, and severe to profound was any hearing loss greater than 60 dB.

To determine progression of hearing loss, at least two audiograms (or ABRs) were required, with at least six months between the readings. Progression of unilateral hearing loss was said to have occurred if the results of an audiogram had increased in its severity by characterization (mild, moderate, severe, profound). To determine progression to bilateral hearing loss, one audiogram, or ABR, was required to demonstrate normal hearing in the contralateral ear. Bilateral progression was documented in the case of a normal hearing contralateral ear later being found to have sensorineural hearing loss.

Radiographic data was captured from the electronic medical record when available. This study was approved by the institutional review board at Virginia Commonwealth University.

Results

Risk Factors of Unilateral Severe to Profound Sensorineural Hearing Loss (USPSNHL) – Table 1
51 patients were found to have unilateral severe to profound sensorineural hearing loss. Of those patients, 20 (39.2%) had a risk factor. The most common risk factor seen was prematurity (7 patients, 53.8%), followed by infection (4 patients, 30.8%). The four infectious causes were identified as two with CMV and two with pneumococcal meningitis.
Risk Factors of Mild to Moderate Unilateral Sensorineural Hearing Loss (UMMSNHL) – Table 2
28 patients were found to have unilateral mild to moderate sensorineural hearing loss. Of those patients, 20 (71.4%) were found to have a recognizable risk factor. The most common risk factor seen was prematurity (9 patients, 90.0%), followed by a stay in the NICU (7 patients, 70.0%). The one infectious cause was identified as CMV.
Temporal Bone or MRI image findings of USPSNHL – Table 3
31 patients with unilateral severe to profound hearing loss had imaging performed, either a temporal bone CT scan, or an MRI. Six of these patients (19%) had an identifiable aberration recognized on imaging. The most common cause seen was enlarged vestibular aqueducts (2 patients, 33%).
Temporal Bone or MRI image findings of UMMSNHL – Table 3
16 patients with unilateral mild to moderate hearing loss had imaging performed, either a temporal bone CT scan, or an MRI. Two of these patients (12.5%) had an identifiable aberration recognized on imaging. In both cases, the abnormal images were enlarged vestibular aqueducts.
Progression of Unilateral Sensorineural Hearing Loss – Table 4
Patients were also assessed for their progression from unilateral hearing loss to bilateral hearing loss, and also progression in severity. For those patients identified with unilateral severe to profound sensorineural hearing loss, 34 had multiple audiograms for comparison. Two of these patients (5.9%) showed progression to becoming bilateral. (Progression in severity was not measured as they were already in the severe to profound category.) Of the patients with mild to moderate unilateral hearing loss, 17 had multiple audiograms for comparison. Four patients (23.5%) showed progression to becoming bilateral and the same four patients demonstrated progression in severity in the ipsilateral ear.

Table 1. Incidence of perinatal risk factors of USPSNHL (N=51); 24 Right sided, 27 Left sided

Perinatal risk factors (N=13)	Number of patients experiencing event (%)
NICU Stay	5 (38.5)
Ventilator	1 (7.7)
Infection	4 (30.8)
Hyperbilirubinemia	3 (23.1)
Prematurity	7 (53.8)

Table 2. Incidence of perinatal risk factors of UMMSNHL (N=28), 12 Right-sided, 16 Left-sided

Perinatal risk factors (N=10)	Number of patients experiencing event (%)
NICU Stay	7 (70.0)
Ventilator	2 (20.0)
Infection	1 (10.0)
IV Antibiotics	1 (10.0)
Prematurity	9 (90.0)

Table 3. Temporal bone or MRI image findings of severe to profound and mild to moderate sensorineural HL (N=37)

	Severe to profound N=32, (%)	Mild to Moderate N=16, (%)
Enlarged Vestibular Aqueduct	2 (6.25)	2 (12.5)
Prominent IACs	-	-
Cochlear Aperture Stenosis	1 (3.1)	-
Hypoplastic Cochlea	1 (3.1)	-
Fracture	1 (3.1)	-
Cystic Cochlea	1 (3.1)	-

Table 4. Progression of USNHL to BLSNHL or Progression of severity of USNHL

	Progression to bilateral (%)	Progression in severity (%)
Severe to profound *(N= 34)	2 (5.9)	-
Mild to moderate *(N= 17)	4* (23.5)	4*(23.5)

*These were the same four patients

Discussion

The purpose of our study was to identify the prevalence of risk factors for children with unilateral sensorineural hearing loss, as well as the prevalence of anatomic inner ear anomalies on imaging studies at our institution. We also sought to determine the rates of progression to the contralateral side as well as progression in severity of the ipsilateral side. However, it has not been until the recent understanding of the sequelae of unilateral hearing loss that more research has been performed to understand the causes of pediatric unilateral sensorineural hearing loss.

Our results demonstrated that those children with USPSNHL had an identifiable JCIH risk factor 39% of the time, the most common being prematurity, followed by infection. Of the patients with mild to moderate USNHL, 71% were found to have an identifiable risk factor, with infection being the most likely cause. These results can be compared to those found by Friedman et al., which identified a perinatal etiology in 14.2% of patients with USPSNHL, with the most common cause being NICU stay.[10] For those with USPSNHL, 19% had an identifiable cause on imaging, with a variety of causes identified, while 13% of those with mild to moderate unilateral hearing loss had an aberration on imaging. Other studies have shown identifiable causes in USPSNHL to be about 25% of cases, and even up to 40%, depending on the imaging modality.[3,9, 11]

In considering progression, we compared audiograms of those with severe to profound unilateral hearing loss, to determine if the hearing loss progressed to the contralateral side. Of those with multiple audiograms, 6% demonstrated a progression to the contralateral side. Four of the 17 patients (23.5%) identified with mild to moderate unilateral hearing loss showed both ipsilateral progression and progression to the contralateral side. The fact that the same four patients suffered both ipsilateral and contralateral progression suggests that they may have, in fact, been diagnosed with unilateral hearing loss because of early recognition of a bilateral, progressive process. The results of progression can be compared to the results seen by Uweira et al., which demonstrated new onset hearing loss in the contralateral ear in 10.6% of patients.

Conclusion

We have demonstrated continued evidence of the risk factors involved in pediatric unilateral sensorineural hearing loss, which were present in 71% of those with mild to moderate UHL, and 39% of those with severe to profound UHL. Progression was also seen in our patients (overall 12%, with 23.5% of those with initially mild to moderate UHL), reiterating the importance of surveillance and follow-up in those diagnosed with unilateral hearing loss.

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