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 progressed to bilateral hearing loss (BHL), 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 cochleae. 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 several 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. Historically, UHL has been more difficult to diagnose early, as speech and language consequences may not be as apparent early in development. With the better understanding of the sequelae of UHL, increased screening 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 common causes of hearing loss.

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. It was also shown that in a competitive noise environment, monaural listeners do not function as well. As anatomic abnormalities can also extend to the vestibular system, children may also experience vertigo, leading to further physical and emotional distress.

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 hearing loss, and exploration of the prevalence of disease progression.

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

<table>
<thead>
<tr>
<th>Perinatal risk factors</th>
<th>Number of patients experiencing event (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>NICU Stay</td>
<td>5 (38.5)</td>
</tr>
<tr>
<td>Ventilator</td>
<td>1 (7.7)</td>
</tr>
<tr>
<td>Infection</td>
<td>4 (30.8)</td>
</tr>
<tr>
<td>Hyperbilirubinemia</td>
<td>3 (23.1)</td>
</tr>
<tr>
<td>Prematurity</td>
<td>7 (53.8)</td>
</tr>
</tbody>
</table>

In the 2007 JCIH position statement, including the following: family history of permanent hearing loss, and exploration of the prevalence of disease progression.

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 ear anomalies on imaging studies at our institution. We identified the rates of progression in severity of the ipsilateral side. However, it has not been until the recent recognition 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 USNHL had an identifiable JCIH risk factor 95% 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 USNHL, with the most common cause being NICU stay. For those with USNHL, 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 on imaging, to be about 25% of cases, and up to 40%, depending on the imaging modality. 1, 2

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 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.

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.

References:


Contact:
Jason H. Barnes
Virginia Commonwealth University School of Medicine
Email: barnesjh@vcu.edu