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Evaluation of unilateral sensorineural hearing loss in the pediatric patient☆

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ABSTRACT

Objectives: This study is a review of our series of pediatric patients with unilateral sensorineural hearing loss (USNHL) to report abnormalities on imaging studies, review genetic and ophthalmologic results, and survey audiometric findings.
Methods: This study is a retrospective chart review of all pediatric patients with USNHL seen between 1/1/03 and 12/31/08 at our institution. The study was approved by the institutional review board.
Results: Eighty-nine cases were identified with audiometric findings confirming unilateral hearing thresholds greater than 20 dBHL with no conductive component. There were 48 males and 41 females. Average age of diagnosis was 7 years. One audiogram showed low-frequency loss, 17 mid-frequency, 29 high-frequency, and 32 flat. Ten patients were diagnosed by auditory brainstem response testing at another institution, with thresholds not available for review. Eleven percent of patients progressed to bilateral loss. Sixty-one patients underwent computed tomography of temporal bones (CTTB). Twenty of 61 scans identified 34 anomalies including 15 enlarged vestibular aqueducts (EVAs), 8 Mondini, and 3 superior semicircular canal dehiscences (SSCDs). Thirty-one of 89 patients underwent magnetic resonance imaging (MRI). Three of these 31 patients had positive findings including 1 EVA, 1 Mondini, and 1 asymmetric internal auditory canal. When CTTB was positive, no additional lesions were detected on MRI. When CTTB was negative and MRI was done in 20 patients, 2 additional lesions were detected by MRI. Fourteen patients had genetics evaluation of which 6 had positive findings, including CHARGE, VACTERL, Goldenhar, and 3 were heterozygous for a Connexin mutation.
Conclusions: CTTB is an effective diagnostic tool for USNHL. MRI should be considered in patients with negative CTTB. Genetics and ophthalmologic evaluations are recommended for patients with risk factors or an abnormal clinical examination. Close follow-up is essential due to high rate of hearing loss progression.

1. Introduction

Sensorineural hearing loss (SNHL) affects approximately 3 in 1000 newborns in the United States [1]. The true incidence of unilateral sensorineural hearing loss (USNHL) has historically been difficult to determine because children with mild UNSHL may have minimal speech or language abnormalities. In addition, screening audiometry techniques do not consistently diagnose unilateral loss. This may explain the delay in diagnosis associated with USNHL, which is not typically detected until early adolescence. There is an estimated 3–5% prevalence of USNHL in school children [2,3]. Traditionally, the effect of unilateral hearing loss on speech and language development was undervalued as it was presumed that normal development occurs with one intact ear. However, there are a number of studies that show a direct correlation between USNHL and educational delays [4–6]. In 2004, Lieu published a 22–35% rate of repeating at least 1 grade, and a 12–41% rate of requiring extra educational assistance with USNHL [5]. Furthermore, Welsh et al. showed that monaural listeners do not function as well as binaural subjects in a competitive noise environment (i.e. school) [7].

With increasing awareness of the functional impact of USNHL on the developing child, recent studies have begun documenting inner ear anomalies in monaural hearing loss patients. Approximately 35% of UNSHL patients have an abnormal computed tomography temporal bone (CTTB) scan, and 25% have abnormal magnetic resonance imaging (MRI) [4,8,9].

There are evidence based recommendations for the work-up of bilateral or idiopathic SNHL that include genetic testing, laboratory testing and imaging modalities [10]. To the best of the authors' knowledge, there have been no comprehensive reviews looking at

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risk factors, genetic testing, ophthalmologic findings, and imaging modality results as they pertain specifically to USNHL. The goal of this study was to review our institutional experience with USNHL to determine the incidence of abnormalities on imaging studies, genetic studies, risk factors, and audiometric findings.

2. Methods

This is a retrospective chart review of all pediatric patients with USNHL seen between January 1st, 2003 and December 31st, 2008 in the pediatric otolaryngology clinic at the Cleveland Clinic. The study was approved by the Cleveland Clinic institutional review board. Inclusion criteria were that the patient was seen at our institution during the specified time period, were under the age of 18 years, and had a diagnosis of USNHL identified using International Statistical Classification of Diseases and Related Health Problems (ICD-9) code of 389.15. These inclusion criteria identified 120 patients. Each audiogram was initially screened for unilateral SNHL defined as average hearing thresholds greater than 20 dBHL in a single ear, with no conductive component. Thirty-one patients were excluded due to presence of a conductive component to their hearing loss or bilateral involvement. This left a remaining cohort of 89 patients. Ten patients were identified thru auditory brainstem response test done at another institution and hearing thresholds were not available for review.

Audiometric evaluations were recorded as follows: the frequency of hearing loss was defined as low (<500 Hz), middle (500–2000 Hz), high (>2000 Hz), and flat.

Pure tone audiograms were reviewed, and whether the unilateral loss progressed to a bilateral SNHL was recorded. Laterality of the USNHL was also recorded.

Demographic data including age at first office visit, age at diagnosis, gender, and nationality were recorded. Whether a CTTB or MRI was ordered, actually obtained, as well as the results of each imaging modality, were recorded. Clinician notes were reviewed and pertinent clinical findings including syndromic appearance, and/or any physical abnormalities. Ophthalmology and genetics consults and their findings were also noted.

Finally, risk factors for bilateral SNHL were also documented including: 48 h or longer stay in a neonatal intensive care unit (NICU), premature delivery, low birth weight, poor weight gain, jaundice in the neonatal period, head trauma, neonatal intubation and need for mechanical ventilation, TORCH infection, meningitis, intravenous (IV) antibiotics, chemotherapy, and/or presence of developmental delays. Any diagnosed etiology of the SNHL was recorded for comparison.

3. Results

3.1. Audiometric data

From January 1st, 2003 to December 31st, 2008, 89 cases were identified with audiometric findings confirming unilateral hearing thresholds greater than 20 dB with no conductive component. One patient had low-frequency loss, 17 mid-frequency, 29 high-frequency, and 32 flat (Table 1). Ten patients were diagnosed by auditory brainstem response testing at another institution, with thresholds not available for review. Eleven percent of patients progressed to bilateral loss within our follow-up time of 5 years.

Table 1
Audiologic evaluation.

Diagnosis by	# of patients
ABR/OAEs	10
Audiogram	79
Type of HL	
Low frequency	1
Mid-frequency	17
High frequency	29
Flat	32

Table 2
Imaging findings.

CT scan	# obtained: 61 total
Normal	41
EVA	15
Mondini	8
Mastoiditis/COM	5
SCCD	3
High jug bulb	1
Cholesteatoma	1
Bony deformation of incus	1
MRI	**# obtained: 31 total**
Normal	21
EVA	1
Mondini	1
Mastoiditis/COM	2
Chiari malformation	1
Asymmetric IAC	1
Large jug bulb	1
Brain cyst	1
Leukomalacia	1
Sub-dural hematoma	1

3.2. Demographic data

Review of the demographic data showed that 68 (76%) of the patients were Caucasian, 8 (9%) African American, 7 (8%) unknown, 3 (3%) Hispanic/Latino, 2 (2%) Asian/Pacific Island, and 1 (1%) Indian. Gender was relatively even as 48 (54%) were male, and 41 (46%) female. Age at diagnosis of hearing loss ranged from newborn up to 17 years old. Average age of diagnosis was 5.6 years. Twenty-three children were identified through the newborn hearing screen. Insufficient documentation prevented determination of how the remaining patients were identified with hearing loss.

3.3. Imaging data

Sixty-one (69%) patients underwent CTTB. Twenty (33%) scans had 34 positive findings that included 15 enlarged vestibular aqueducts (EVAs), 8 Mondini malformations, 3 superior semicircular canal dehiscences (SCCDs), and others (Table 2). Nine of the CTTBs had more than one positive finding such as a Mondini deformity as well as an enlarged vestibular aqueduct. Thirty (34%) patients underwent MRI. Three (10%) had positive findings including 1 EVA, 1 Mondini malformation, and 1 asymmetric internal auditory canal (Table 2).

Of the patients that had progressive hearing loss, 2 patients had no imaging performed and 2 other patients had both normal CTTB and MRI. One patient had bilateral EVAs on CTTB and another patient had EVA on CTTB that was not detected on MRI. Lastly, 4 patients had normal CTTB with no MRI.

When the CTTB was positive, no additional synchronous lesions were detected on MRI. However, in 8 patients who had a positive CTTB and also received an MRI, the abnormality diagnosed on CTTB went undetected in 4 (50%) of the cases. These cases were 3 EVAs and 1 incomplete partition within the cochlea. When CTTB was negative and MRI was done in 20 patients, 2 additional lesions were detected by MRI (11%). Four patients had an MRI done without a CTTB all of which were