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Pediatric sudden sensorineural hearing loss: Etiology, diagnosis and treatment in 20 children☆

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ABSTRACT

Objectives: 1. To report our experience in children with sudden-onset sensorineural hearing loss (SSNHL). 2. To describe the etiology and management of children with SSNHL.
Methods: Retrospective review of 20 children with SSNHL, from 2000 to 2013 at a tertiary pediatric facility. Patients had the following inclusion criteria: history of normal hearing, hearing loss occurring in less than 3 days, and audiogram documentation.
Results: The average age of patients presenting with SSNHL is 11 years 3 months (22months-18years). Only 6 (30%) children presented prior to 2 weeks. Tinnitus (55%) was the most common associated symptoms followed by otalgia (25%), and vertigo (20%). Eight patients had bilateral hearing loss, 6 only right and 6 only left. Hearing loss severity ranged from profound (45%) being most common to mild. Etiology was unknown (30%), viral (25%), anatomic abnormality (25%), Meniere's disease (5%), autoimmune (5%), perilymphatic fistula (5%), and suppurative labyrinthitis (5%). Eight patients had initial treatment with oral steroids of which 50% had improvement on audiograms. Two patients underwent intratympanic injections, both showed improvement. Of the 12 patients with no treatment, only 1 had improved hearing.
Conclusions: The true incidence of pediatric SSNHL is not well established in our literature. Unique aspects of pediatric SSNHL are delayed presentation and higher percent of anatomic findings. In our study 70% presented more than 2 weeks after experiencing symptoms. Anatomic abnormalities are in 40% of patients. Hearing improvement occurred in 50% of children treated with oral steroids. Intratympanic steroid treatment is another option but may have practical limitation in the pediatric population.

1. Introduction

A rare etiology for pediatric hearing loss is sudden onset sensorineural hearing loss (SSNHL). It is defined as a 30 dB loss over more than 3 consecutive frequencies, with rapid onset, from a few hours to a maximum of 3 days [1]. SSNHL is more frequent and better characterized in the adult population. It has a prevalence of 5–20/100,000 adults. It occurs mainly in patients between the ages of 25–60 and peaks at 46–59 years of age [2,3]. It has been reported that the incidence in adolescents and children is approximately 3.5% of the adult incidence [4]. However other studies still believe the incidence of pediatric SSNHL is largely unknown [5,6].

It is not well understood in the pediatric population; only a limited number of studies currently exist discussing the diagnosis and management in this population. Due to the low incidence in the pediatric population, a majority of the conclusions are extrapolated from the adult literature. The current adult recommendations for management are audiologic testing and magnetic resonance imaging (MRI), to identify brain or retrocochlear pathology [7]. The most significant etiology in adults is due to stroke or retrocochlear lesions [7,3]. In the pediatric population, 20% with hearing loss have abnormalities on radiologic testing; however, it has not been determined if computed tomography (CT) or MRI is a better modality for SSNHL [8,9].

With regards to treatment, oral and intratymanic steroids and hyperbaric oxygen therapy have all been discussed in the adult population. The pediatric literature has only discussed the benefits of oral steroids in a small number of patients [5,6]. In our study we

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discuss twenty pediatric patients with SSNHL, their presentation, etiology, diagnostic methods as well as treatment and outcomes.

2. Methods

This is a retrospective medical record review from 2000 to 2013 at the Children's Hospital of Pittsburgh (CHP) of the University of Pittsburgh Medial Center (UPMC). Approval of the Institutional Review Board at UPMC was obtained. All pediatric patients with the diagnosis of SSNHL were initially reviewed. Only patients, with a history of normal hearing, hearing loss occurring in less than 3 days along with audiogram documentation were included.

For all eligible patients, the medical records were reviewed and a database was established recording the following variables for all patients:

- Demographics: age of diagnosis, sex
- Laterality of hearing impairment
- Date of onset of symptoms and date of diagnosis at office visit
- Initial audiogram and follow up audiograms. The degree of hearing loss was determined as from the pure tone average as: Mild (20–40 dB), Moderate (40–60 dB); Moderate to severe (60–75 dB) Severe (75–90 dB); and Profound (>90 dB).
- Initial symptoms, preceding events and treatment regimen
- Outcomes after treatment with oral and/or intratympanic steroids
- Past medical history, pertinent family, social and surgical histories

This data was entered into an Excel spreadsheet. Descriptive analysis was performed.

3. Results

A total of 20 out of 80 patients met the criteria for SSNHL from 2000 to 2013. Of these patients, half were male and half female. The average age of diagnosis was 11 years and 3 months, ranging from 1 year 10 months–18 years and 2 months. Patients presented an average of 122 days after onset, ranging from 2 to 365 days. Seventy percent of patients presented 2 weeks after onset. Eight patients (40%) reported preceding events, mostly viral illness and acute otitis media. One patient had meningitis. Tinnitus was the most common associated symptom in 11 (55%) patients, followed by otalgia (n = 5, [25%]), vertigo (n = 4, [20%])), aural fullness (n = 3, [15%]), and ataxia (n = 1, [5%]). Only 4 (20%) of the patients had no associated symptoms (Table 1).

The most common medical history was recurrent acute otitis media (n = 4 [20%]). Other notable medical problems were hypoxic ischemic encephalopathy at birth (n = 1, [5%]), ulcerative colitis (n = 1, [5%]), epilepsy (n = 1, [5%]), Chiari I malformation (n = 1, [5%]) and Stickler syndrome (n = 1, [5%]). Patients had a family history significant for congenital hearing loss (n = 2, [5%], acoustic neuroma (n = 2, [5%]) and Meniere's disease (n = 1, [5%]). Aside from tympanostomy tube placement, none of the patients had a history of otologic surgery.

Eight patients (40%) had bilateral hearing loss, six (30%) with left only and 6 (30%) with right only loss. There was a high incidence of profound hearing loss in this cohort (n = 9), while 40% (n = 8) were moderate, 25% (n = 5) moderate-severe, 20% (n = 4) mild, 15% (n = 3) severe-profound and 5% (n = 1) mild-moderate. All patients underwent audiograms at the initial consultation, and at the first follow up visit.

Three patients underwent genetic testing for GJB2 and GJB6 mutations; the results were normal. No patients had routine laboratory tests. Ninety percent of the patients underwent radiographic testing. Forty-five percent had CT and MRI, 30% (n = 6) with MRI only and 15% (n = 3) with CT only. The scans were abnormal in 40% (n = 8) of the patients. There was a discrepancy between CT and MRI in 2 patients. Both patients had an enlarged vestibular aqueduct on CT scan with no correlating enlarged endolymphatic sac on MRI. Other identified anatomic abnormalities are listed in Table 2. Anatomic abnormalities were identified as possible etiology in twenty-five percent: 4 patients had EVAs and 1 patient with absent/hypoplastic cochlear nerve. The etiology was unknown in 30% (n = 6) of the cohort. In 25% (n = 5) it was thought to be due to a viral cause. Other etiologies are as follows: Meniere's disease (n = 1), autoimmune (n = 1) and perilymphatic fistula (PLF) (n = 1) (Fig. 1). The one patient with PLF underwent an exploratory

Table 1
Initial presentation.

ID#	Age (yrs)	Age (mo)	Symptoms prior to presentation (days)	Initial symptoms	Events preceding onset	Etiology
1	9	11	365	Hearing loss	Stuck q-tip in ear	Absent/hypoplastic cochlear nerve
2	18	2	2	Tinnitus, vertigo, nausea, vomiting		Viral
3	6	7	90	Ignoring parents, hearing loss		Unknown
4	8	9	30	Tinnitus	URTI; right AOM	Viral
5	12	6	240	Aural fullness		Unknown
6	15	10	180	Hearing loss, tinnitus, otalgia		Unknown
7	13	9	7	Hearing loss, tinnitus	Sinus infection	Left EVA
8	5	6	365	Preferential use of telephone		Bilateral EVA
9	14	4	365	Tinnitus, otalgia		Bilateral EVA
10	1	10	150	Ataxia, fatigue, not responding to sounds	Febrile illness	Viral
11	12	10	7	URTI, dizziness, lethargic	AOM, viral illness	Viral
12	7	9	14	Otalgia, headache, vertigo, nausea, vomiting, tinnitus, hearing loss		PLF
13	4	8	30(R) & 2.5(L)	Meningitis	*S. pneumoniae* meningitis	Meningitis
14	13	1	90	Muffled hearing, tinnitus		Unknown
15	11	10	14	Otalgia	Bilateral AOM	Unknown
16	13	8	120	Fever, odynophagia, bilateral otorrhea, tinnitus, hearing loss, nausea, vomiting, dizziness	Viral illness	Suppurative Labyrinthitis
17	15	7	150	Tinnitus, otalgia, hearing loss		Meniere's Disease
18	16	1	17	Vertigo, tinnitus, otalgia, aural fullness		Autoimmune
19	10	9	30	Hearing loss	AOM	Unknown
20	12	5	180	Tinnitus, hearing loss		Bilateral EVA

R = right ear; L = left ear; URTI = upper respiratory infection; EVA = enlarged vestibular aqueduct; AOM = acute otitis media; PLF = perilymphatic fistula.