Clinical Experience in Diagnosis and Management of Superior Semicircular Canal Dehiscence in Children

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Objectives/Hypothesis: To identify clinical characteristics of pediatric superior semicircular canal dehiscence (SSCD) and explore suitable options of management.

Study Design: Retrospective review.

Methods: The study comprised 10 patients with auditory and/or vestibular symptoms suspicious for SSCD. One patient pursued care at another institution, and two did not return for follow-up. Subsequently, seven patients (11 ears, 6 females and 1 male, aged 5–11 years) were included. Patients were evaluated using high-resolution temporal bone computed tomography. Those suspected of having SSCD underwent vestibular evoked myogenic potential testing for confirmation in addition to routine audiological tests.

Results: All seven patients had auditory and/or vestibular impairment. Auditory symptoms included autophony, tinnitus, and conductive or mixed hearing loss. Bone conduction responses were occasionally better than 0 dB HL. Vestibular dysfunction included vertigo, often in response to loud noises, and chronic disequilibrium. One patient underwent surgical repair for disabling vestibular symptoms with dramatic improvement in both auditory and vestibular symptoms postoperatively. The remaining six were closely monitored with routine exams.

Conclusions: In contrast to adults, children with SSCD usually present with auditory symptoms first, although they share some similarities with adults in clinical manifestations of SSCD. Our study shows that SSCD syndrome, a well-accepted clinical entity, exists in the pediatric population. Conservative management is preferred for children with SSCD; nevertheless, surgical intervention is necessary for those with disabling vestibular symptoms. To date, this is the first clinical case series of symptomatic pediatric patients with SSCD.

Key Words: Semicircular canal dehiscence, pediatric hearing loss, vestibular dysfunction, VEMP testing.

Level of Evidence: 4.

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INTRODUCTION

Superior semicircular canal dehiscence (SSCD) syndrome, first described in 1998 by Minor et al., occurs when the bone overlying the superior semicircular canal is thinned out or absent. This leads to a “third window phenomenon” in which sound energy is redistributed and the vestibular system is inappropriately stimulated, resulting in vestibular dysfunction as well as hearing loss. The diagnosis of SSCD syndrome can be difficult due to the often nonspecific nature of the symptoms. In addition to clinical history, an audiogram demonstrating hearing loss and a high-resolution computed tomography (CT) scan of the temporal bones exhibiting an absence of bone will often confirm the diagnosis. Recently, vestibular evoked myogenic potentials (VEMP) testing has revealed specific findings—such as abnormally low thresholds—in patients with third window defects including SSCD syndrome; the use of this test has become more widespread in helping to diagnose the condition. Treatment involves conservative management with vestibular suppressants or surgical repair of the bony dehiscence if symptoms are intractable.

Literature on symptomatic SSCD has primarily been limited to the adult population. Presenting symptoms have been primarily vestibular related, with few patients reporting strictly auditory symptoms, and ages ranging from the second to seventh decades of life. The presence of SSCD syndrome in the pediatric population has only recently emerged. Case reports have described this entity in children, including a child with progressive hearing loss and disequilibrium who was found to have a right-sided SSCD and another case of a child with posterior semicircular canal dehiscence. In addition, a recent review of pediatric temporal bone CT scans obtained to evaluate for hearing loss identified a 9.9% prevalence of dehiscent superior semicircular canals. In this study, we present seven pediatric patients who were evaluated for hearing loss and/or vestibular
symptoms and were found to have dehiscence of one or both superior semicircular canals. This is the first known series in the literature of symptomatic SSCD in the pediatric population.

MATERIALS AND METHODS

Patients

A retrospective review was performed of patients who presented to a tertiary-care pediatric hospital for evaluation of hearing loss and/or vestibular symptoms. High-resolution CT of the temporal bones was performed; those with radiographic evidence of SCCD underwent VEMP testing. Ten subjects with CT evidence of dehiscence were identified. Of these, one patient underwent further workup and management at an outside institution. Two others did not pursue further workup. The seven remaining patients with radiographic evidence of dehiscence of one or both superior semicircular canals confirmed with abnormal VEMP testing were included in this study. Their clinical history, audiometric data, and CT scans were reviewed. This study was approved by our institutional review board.

Interventions

High-resolution (0.625-mm interval) noncontrast CT of the temporal bones using standard helical protocol was performed and reformatted to provide axial, coronal images as well as Stenver’s and Poschl’s projections when available.

The patients underwent conventional pure-tone audiometry with air-conduction testing over 250 to 8,000 Hz and bone-conduction testing over 250 to 4,000 Hz with appropriate masking when necessary. When available, prior audiograms were used to evaluate the stability and/or changes in hearing loss over time. Tympanometry was performed with a 226 Hz probe.

VEMP testing was performed using the protocol established in a prior study. Briefly, VEMP responses were elicited using tone bursts of 250, 500, and/or 1,000 Hz via air or bone conduction. Initial stimulus level was 90 dB HL, and sequentially decreased until responses were absent. Results consistent with SSCD were defined as follows: abnormally low threshold (<65 dB HL for any stimulus) or normal threshold levels (70–90 dB HL), despite the presence of a significant air-bone gap (ABG) (>30 dB).

RESULTS

Case Reports

Patient 1. This 6-year-old female presented with hearing loss and episodic dizziness with nystagmus and nausea. Prior workup revealed abnormal rotational chair testing consistent with unilateral peripheral vestibular hypofunction, a normal electroencephalogram, and an incidental finding of Chiari I malformation on brain magnetic resonance imaging (MRI). The self-resolving disequilibrium spells were associated with “room tilting.” An audiogram revealed severe mixed loss on the right and moderate-to-mild sensorineural hearing loss (SNHL) on the left. Subsequent testing exhibited a stable right-sided severe mixed loss with large ABG (55 dB at 250 Hz to 25 dB at 4 kHz) and a fluctuating mixed loss on the left. An autoimmune workup was negative, and a trial of oral steroids was ineffective. A CT scan revealed bilateral dehiscence at the junction of the posterior and superior semicircular canals (Fig. 1). VEMP thresholds were decreased on the left (60/70 dB HL at 250/500 Hz) and normal on the right (75/85 dB HL at 250/500 Hz). This was most consistent with bilateral SSCD. Currently, she is aided on the left, and we continue to monitor her hearing levels.

Patient 2. This 5-year-old female with a strong family background of congenital hearing loss was initially diagnosed with symmetric moderate SNHL (45–55 dB HL). She learned to walk at a delayed age and had significant problems with dizziness and disequilibrium. For 3 years her hearing loss gradually progressed despite normal exams. Recent audiograms revealed
symmetric moderate-severe predominantly sensorineural hearing loss with a mixed component on the right only. CT scan suggested bilateral SSCD, but VEMP testing revealed lowered thresholds and high amplitudes on the right only (60/70 dB HL at 250/500 Hz). These results were most consistent with a right-sided dehiscence not thought to be related to her baseline SNHL. In the past year, the patient has reported improvement in her disequilibrium with fewer vertiginous episodes; her hearing has remained stable. She is aided bilaterally.

**Patient 3.** This is an 8-year-old female formerly a 24-week neonate referred for a second opinion regarding her hearing loss. She previously underwent two sets of tympanostomy tube placement but continued to fail multiple school screening hearing tests. She walked at a delayed age and was discouraged from participating in gymnastics because of poor balance skills. She complained of intermittent disequilibrium and reported autophony but no tinnitus. Her exam was normal, but her audiogram revealed a mild conductive hearing loss (CHL) bilaterally. Imaging revealed left SSCD and possible dehiscence of the right. VEMP thresholds were normal via air conduction testing (right: 85/90 dB HL at 250/500 Hz; left: 85 dB HL at 250 Hz) but were abnormally low bilaterally with bone conduction. VEMP testing confirmed bilateral dehiscence. She has been fitted for hearing aids, which she reports as helping with her hearing.

**Patient 4.** This 5-year-old female with multiple congenital abnormalities including polydactyly, micrognathia, and deformed auricles was noted to have mild CHL bilaterally that did not resolve after previous ear tube placement for effusions. At the time of presentation to our clinic, she had mild-moderate CHL and no obvious vestibular symptoms. A CT revealed abnormal ossicles and a left dehiscent superior canal (Fig. 2). VEMP results were consistent with this finding, with elevated thresholds (85/90 dB HL at 250/500 Hz) in the left ear in the presence of a 25 to 35 dB ABG. Thresholds were absent on the right side. She has done well with bilateral hearing aids.

**Patient 5.** This 8-year-old male with attention deficit disorder and auditory processing disorder was evaluated for rotational vertigo spells associated with fatigue. In addition, he reported noise-induced nausea, right-sided pulsatile tinnitus, and autophony but denied hearing loss. A seizure workup was negative, including a normal brain MRI and electroencephalogram. His exam was unremarkable, an audiogram confirmed normal hearing, tympanometry revealed normal middle ear pressures and mobility, and acoustic reflexes were present bilaterally. The MRI of the brain and internal auditory canals (obtained as part of his seizure workup and to evaluate for any other etiologies of vestibular dysfunction) suggested right SSCD, but a subsequent CT scan exhibited bilateral dehiscence. VEMP thresholds were lowered bilaterally (right: 60/60 dB HL at 250/500 Hz; left: 55/65 dB HL at 250/500 Hz). Currently he is managed conservatively, with routine clinic exams and audiograms.

**Patient 6.** This 11-year-old female with progressive hearing loss and disequilibrium was diagnosed with a right SSCD and bilateral congenital ossicular abnormalities. Audiograms revealed a moderate-mild mixed but primarily conductive loss on the right and a severe-moderate mixed loss on the left. Her hearing had recently worsened. She also reported right-sided pulsatile tinnitus, autophony, and aural fullness. Her vestibular symptoms, which began with mild dizziness spells, developed into frank vertiginous episodes associated with loud external noises and often with her own tinnitus. Electronystagmography performed at an outside institution was normal. Her CT scan exhibited right SSCD and possible left dehiscence. VEMP thresholds were only abnormal on the right (65/80 dB HL at 250/500 Hz), supporting the diagnosis. After years of conservative management and use of hearing aids, she ultimately underwent repair of her right SSCD via a middle fossa craniotomy (Fig. 3) at age 14. By her first postoperative visit, she reported complete resolution of the tinnitus and autophony and dramatic reduction in vertigo spells. Postoperative VEMPs were elicited at normal levels via bone conduction. Postoperative audiograms revealed improvement in her right ear but continued progression of hearing loss on her left.

**Patient 7.** This 5-year-old female was evaluated for right-sided hearing loss and speech delay but no
vestibular symptoms. Developmentally she had delayed onset of walking. Her audiogram revealed a severe-moderate mixed but primarily conductive loss on the right and mild CHL on the left. CT scan showed ossicular abnormalities, a clear right SSCD, and likely left SSCD. Despite large ABGs, VEMP thresholds were normal (right: absent/90 dB HL at 250/500 Hz; left: 75/80 dB HL at 250/500 Hz), confirming bilateral dehiscence. She is aided on the right and uses an FM amplification system at school.

Summary of Clinical Features

Seven pediatric patients (6 female, 1 male, 11 ears) with auditory and/or vestibular symptoms were found to have SSCD (Table I). Mean age at the time of diagnosis was 6.9 years. Four had bilateral defects, and two of the three unilateral cases were right-sided. Six patients had either mixed or purely conductive hearing loss; one individual (patient 5) had normal hearing bilaterally. The three oldest patients reported autophony; two of these three also had tinnitus and noise-induced vertigo. We were unable to elicit a history of these symptoms in the younger children. Five children had episodic vertigo and generalized disequilibrium, and three had delayed onset of walking (>18 months). There were no syndromic or craniofacial predictions, although two did have abnormal features without a craniofacial diagnosis. To date, all patients have been fitted with hearing aids; six have been closely followed conservatively, and one underwent surgical repair.

Summary of Audiologic Tests

Of the nine affected ears that exhibited hearing loss, four had a mixed loss and five had a purely conductive hearing loss (Fig. 4). For the ears with mixed loss, mean air-conduction threshold levels ranged from 53.8 ± 25.6 dB at 8 kHz to 68.8 ± 20.6 dB at 500 Hz. Mean ABG ranged from 13.8 ± 13.2 dB at 4 kHz to 43.8 ± 19.3 dB at 250 Hz. For those with purely conductive loss, mean air-conduction thresholds ranged from 25.0 ± 5.0 dB at 4 kHz to 36.0 ± 8.2 dB at 250 Hz with ABG ranging from 17.5 ± 11.9 dB at 4 kHz to 33.0 ± 14.4 dB at 250 Hz. This represented, on average, a 31.1 ± 3.2 dB worse hearing loss for ears with a mixed deficit compared to those with a purely conductive loss. Generally, ABGs were greater at the lower frequencies and measured as large as 65 dB. Only two patients were found have bone-conduction thresholds greater than 0 dB HL at one or more frequencies.

VEMP responses were present in all ears with SSCD. Seven ears exhibited thresholds that were abnormally low for at least one frequency, with mean threshold levels of 60.0 ± 3.5 dB at 250 Hz and 69.0 ± 7.4 dB at 500 Hz. The remaining four ears showed thresholds at normal levels (mean level, 78.3 ± 5.8 at 250 Hz and 86.3 ± 4.8 at 500 Hz) but also had significant ABGs on their audiograms.

DISCUSSION

The inability of children to accurately identify and verbalize their symptoms makes it difficult to recognize...
them without a higher index of suspicion. This likely impacted our ability to collect meaningful information.

In our patients, there was considerable variability in presenting symptoms. Hearing loss, either conductive or mixed, and other auditory symptoms were the most common presenting symptoms. Although most of the patients had stable or progressive hearing loss, one patient had entirely normal hearing. On the other hand, two patients had fluctuating loss. Patient 1 had a Chiari I malformation, which has been associated with sensorineural hearing loss.\(^{12,13}\) Although this may have contributed to the severity of her deficit as well as its fluctuating nature, it would not explain the conductive component of her hearing loss. Finally, the presence of autophony and pulsatile tinnitus varied, although again, this may be a function of the difficulty of children to recognize and report them.

Equally variable was the constellation of vestibular symptoms. Episodic vertigo and disequilibrium were the most common symptoms reported. Except for one child whose clinical history was difficult to elicit, all patients had some vestibular complaints. In one child, delayed onset of walking was the only notable symptom. Younger children may not be able to easily recognize vestibular symptoms, especially if they have persisted for much of their lives. In these cases, parents may also have difficulty distinguishing subtle disequilibrium with normal development. Auditory symptoms, on the other hand, maybe appear to be more common because objective information can initially be obtained with routine audiologic tests.

High-resolution CT of the temporal bones has been a primary diagnostic tool to identify SSCD. The bony covering of the superior semicircular canal in adults can be as thin as 0.1 mm.\(^{14}\) In infants, the initial periosteal layer covering the membranous canal ossifies during the first few years of life. Abnormal development of the endochondral and outer periosteal layers has been proposed as a possible mechanism for bony dehiscence.\(^{11}\) Despite the constant improvements in hardware and image-processing software of CT scanners, this frequently subtle bony layer is difficult to capture owing to limitations in resolution. In our patients, CT scans were obtained not necessarily to evaluate for dehiscence but to assess for any anatomic abnormalities that may explain hearing loss, including ossicular anomalies, cochlear malformations, or other third window defects such as enlarged vestibular aqueduct. In some cases, dehiscent canals were clearly obvious, whereas in others the findings were more obscured. Furthermore, the location of the dehiscence was often lower and more medial (closer to the junction of the superior and lateral semicircular canals), as opposed to the more classic apical location at the arcuate eminence.

Patients with CT scans that delineated a clear dehiscence or were suggestive of dehiscence underwent VEMP testing. Abnormally low thresholds to either air or bone testing or normal thresholds in the presence of a large ABG were the criteria to suggest dehiscence. A prior study by our group yielded 91.4% sensitivity and 95.8% specificity for dehiscence with VEMP.\(^{7}\) All of the patients in our series met these criteria, and, in conjunction with imaging, were diagnosed with SSCD. VEMP was also useful for ruling out dehiscence in patients with abnormal scans. CT in two patients revealed bilateral dehiscences (patient 2 showed bilateral dehiscence, patient 6 exhibited clear SCCD on the right and possible dehiscence on the left), but the VEMPs were abnormal on the right only. A third patient with what appeared to be a clear, bilateral SSCD on imaging had VEMP results...
inconsistent with dehiscence and more consistent with middle ear pathology. She was not included in the study.

To date, six patients have been conservatively managed with close observation and regular audiometric testing. Five with hearing loss are aided bilaterally. One patient (patient 6), had progressively worsening vestibular symptoms in addition to progressive hearing loss; she underwent surgical repair of her dehiscence. Exploration revealed a large region of dehiscence of her superior semicircular canal. After repair, her vestibular symptoms completely resolved, and postoperative VEMP testing indicated no evidence of further third window defect; multiple audiograms have confirmed an initially improved but now stable mixed hearing loss on the operated side.

CONCLUSION

Symptomatic SSCD can present in the pediatric population and is manifested by an inconsistent constellation of auditory and vestibular symptoms. Children who present with inexplicable hearing loss not attributed to common pediatric etiologies such as middle ear pathology and congenital abnormalities of the ossicles or cochlea or with unusual vestibular symptoms not attributed to etiologies such as migraine-associated vertigo or benign paroxysmal positional vertigo should raise suspicion for SSCD. Clinical history should be thoroughly elicited from both the parents as well as the child. In addition to audiological testing, patients should undergo high-resolution CT of the temporal bones and VEMP testing. Once the diagnosis is confirmed, treatment is generally conservative, with use of hearing aids and regular evaluations. Surgical repair should be reserved for patients with progressive or intractable vestibular symptoms.

BIBLIOGRAPHY