ABSTRACT

The Superior Semicircular Canal Dehiscence Syndrome (SSCDS) is characterized by bone wear layer overlying the superior semicircular canal. Common symptoms of SSCDS the presence of vertigo associated with nystagmus induced by intense sound stimuli or changes in intracranial pressure or middle ear. The aim of this study is to describe the audiological and vestibular findings of two patients diagnosed with Superior Semicircular Canal Deiscence Syndrome, with confirmed diagnosis by computed tomography. Meatoscopy, anamnesis, pure tone audiometry and vocal followed by the acoustic impedance measurements, audiometric Weber, research Tulio phenomenon and Valsalva maneuver, performed by the same researcher in one session were held. Air-bone gap were observed, type A tympanometric curve and acoustic reflex. The air-bone gap is presented with greater amplitude at low frequencies. Hearing complaints were not reported by patients as the first symptoms. Weber showed lateralization in both cases, confirming the presence of gap. The Thulium phenomenon is positive for vertigo in both cases. The Valsalva maneuver showed a change in only one case.

Keywords: Hearing; Audiometry; Semicircular Canals; Nystagmus, Pathologic; Dizziness
INTRODUCTION

Superior Semicircular Canal Dehiscence (SSCDS) was first described as the wear of the bone layer that covers the superior semi-circular canal, causing an abnormal exposure of the vestibular membranous labyrinth in the cranial middle fossa.

This bone dehiscence results in a third movable window, allowing pressure to dissipate as the membranous labyrinth projects inwardly and the endolymph flows away from the ampulla. Following this pathophysiological scenario, some symptoms of Superior Semicircular Canal Dehiscence Syndrome (SSCDS) may appear, such as elevated air conduction thresholds and maintenance of bone conduction thresholds, as well as vestibular symptoms induced by intense sonorous stimuli and by modifications of intracranial pressure or by the medium ear.

A microscopic study aiming at determining the prevalence of SSCDS in the general population analysed 1,000 temporal bones obtained through autopsies, and found that the superior semi-circular canal dehiscence occurred in approximately 0.7% of the individuals studied, reaffirming the low incidence of disease. The authors of this study affirmed, however, that not all patients with SSCDS present the symptoms of the Syndrome, and that the percentage of symptomatic ones among them is not yet known.

Other studies indicating the incidence of SSCDS in its clinical form were not found.

The etiology of SSCDS is still unclear and it has been much debated in order to determine whether it is congenital, acquired or a mixture of both. Some authors have postulated that it is a developmental imperfection that becomes clinically relevant in adulthood after a trauma or as a cause of an increased intracranial pressure which leads to a rupture of the bone.

The presence of vertigo associated with nystagmus induced by intense sonorous stimuli or by changes in intracranial or middle ear pressures are common symptoms of SSCDS.

Some patients diagnosed with SSCDS may present autophony and conductive hearing loss, although these characteristics are less frequent than for patients with vestibular symptoms.

Patients with SSCDS present a more significant air-bone gap for the low frequencies, caused by the presence of the third window that dissipates acoustic energy. It is known that the bone pathway threshold can be lower than 0 dBNA in the 250 and 500 Hz frequencies.

In tympanometry, patients presented a type A curve and present acoustic reflexes. Acoustic reflexes are present in patients with SSCDS, contrary to what was expected for individuals with hearing impairment due to alteration of the middle ear.

Tullio phenomenon and the Valsava maneuver in patients with SSCDS may show dizziness or nystagmus in the presence of high intensity sounds and pressure variation in the external acoustic canal, respectively.

The Symptoms for Superior Semicircular Canal Dehiscence Syndrome may be similar to the symptoms of other diseases such as: otosclerosis and tubal dysfunction, Ménière’s disease, temporomandibular dysfunction, which may delay the diagnosis. It is important to establish a differential evaluation protocol when the suspicion of Superior Semicircular Canal Dehiscence Syndrome is present.

Based on the previous considerations, the purpose of this study is to describe the audiological and vestibular findings of two patients diagnosed with Superior Semicircular Canal Dehiscence Syndrome.

PRESENTATION OF CASES

This case report project was analyzed and approved by the Federal University of São Paulo research ethics committee under No. 1717/08. The study was conducted at the Audiological Outpatient Clinic of the Discipline of Auditory Disorders of the Department of Speech-Language Pathology and Audiology, from the Federal University of São Paulo.

The eligibility criteria were: both sexes, Superior Semicircular Channel Dehiscence Syndrome confirmed by computed tomography.

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Subjects were informed about the procedures performed and signed a consent form before participating in the study.

Meatoscopy, anamnesis, tonal and vocal audiometry followed by acoustic immittance measurements, audiometric Weber, Tullio phenomenon research and Valsalva maneuver were performed by the same researcher in a single session.

Pure tone audiometry (air and bone conduction), speech audiometry, audiometric Weber and Tullio phenomenon were performed with the Interacoustics MA-41 audiometer. For acoustic immittance measurements, the Interacoustics AZ7 impedance meter was used.
The following two cases are presented:

- **Case 1** - RPS, female, 42 years of age. She was admitted in the Otorhinolaryngology outpatient clinic of the institution of origin with a complaint of dizziness. The patient reported rotatory and non-rotatory dizziness, lasting approximately 40 minutes, as well as auricular fullness in vehicles.

- **Case 2** - NVSS, female, 52 years of age. In 2010, she presented a complaint of intense vertigo in short-term crises 20 years before, accompanied by neurovegetative manifestations, spatial disorientation and panic to go out without a company. She reported sporadic acute pitch tinnitus and high discomfort for loud sounds.

Both cases were submitted to the following procedures: anamnesis, air and bone pathway tonal audiometry, logaudiometry, immitanciometry (tympanometry and research of contralateral stapedial reflexes), audiometric Weber’s test, tullio’s phenomenon and Valsalva maneuver.

**RESULTS**

**Case 1**

RPS was admitted in the Otorhinolaryngology Clinic of the institution of origin, with complaints already reported in the case presentation. An otolaryngological evaluation was performed, which revealed normal otoscopy and no other complaints were reported.

**Table 1. Results for the procedures performed in case 1**

<table>
<thead>
<tr>
<th>Date</th>
<th>Complaint</th>
<th>Tonal Audiometry</th>
<th>Vocal Audiometry</th>
<th>Timpanometry</th>
<th>Acoustic Reflexes</th>
<th>Audiometric Weber</th>
<th>Tullio Phenomenon</th>
<th>Valsalva Maneuver</th>
</tr>
</thead>
<tbody>
<tr>
<td>2005</td>
<td>Rotating and non-rotating dizziness; Auricular fullness</td>
<td>Normal</td>
<td>Normal</td>
<td>Type A bilaterally</td>
<td>Not made</td>
<td>Not made</td>
<td>Not made</td>
<td></td>
</tr>
<tr>
<td>2006</td>
<td>Tinnitus; Rotating dizziness; Auricular fullness; Crises start with temperature changes</td>
<td>Normal</td>
<td>Normal</td>
<td>Type A bilaterally</td>
<td>Not made</td>
<td>Not made</td>
<td>Not made</td>
<td></td>
</tr>
<tr>
<td>2007</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2009</td>
<td>Dizziness; Nausea; Discomfort with intense sounds; Bilateral intense tinnitus</td>
<td>Left Ear: GAP at 250, 500, 1000, 3000 and 4000Hz frequencies, and hearing loss at 6000Hz; Right Ear: Normal</td>
<td>TA Compatible</td>
<td>Type A bilaterally</td>
<td>Vertigo + Nystagmus -</td>
<td>Vertigo + Nystagmus -</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

TA: Tonal audiometry; HZ: Hertz.

Audiometry with normal thresholds in both ears was performed. In the tympanometry, type A tympanometric curves and acoustic reflexes were observed in both ears. The patient underwent a vestibular examination, which diagnosed irritative peripheral bilateral vestibular syndrome.

In August 2006, the patient returned to the outpatient clinic complaining that the crisis began with the change in temperature. The crisis began with tinnitus, auricular fullness, crackings, followed by rotatory dizziness lasting approximately 10 minutes. After the crisis, the patient referred to occipital headache with...
frontal pulsatile irradiation with nausea, aversion to sound and light. A new audiometry was performed, which results again revealed auditory thresholds within the limits of normality.

When the patient returned in June 2007, she reported that she presented three crisis of rotatory dizziness with nausea and scotomas, and it was necessary to go to the emergency room. She also reported mild dizziness that lasted 10 minutes whenever she got up from a chair, auricular fullness when on the subway or on the bus in addition to tinnitus and hearing loss. In August 2007, a temporal bone CT was performed, which indicated total bone dehiscence in the left superior semicircular canal.

In April 2009, RPS returned to the outpatient clinic complaining of dizziness and nausea in evolution for 5 years, as well as intense bilateral tinnitus and uncomfortable for intense sounds, without diminishing the auditory sensitivity.

In the audiological evaluation performed in 2009 for this study, hearing loss was observed only at the 6000 Hz frequency on the left side, with air-bone gap in the frequencies of 250, 500, 1000, 3000 and 4000 Hz and logoaudiometry compatible with the audiometric results. Good mobility of the tympano-ossicular system was observed, with type “A” curve and contralateral stapedial reflexes present in both ears at adequate levels of intensity.

The audiometric Weber test lateralized to the right at 1000 and 2000 Hz and to the left at 500 and 4000 Hz. The Tullio phenomenon was performed at 500 and 1000 Hz at 100 dB with a positive result for vertigo, but no nystagmus was observed. In the Valsalva maneuver, there was vertigo without the presence of nystagmus.

In April 2009, NVSS was admitted in the Otorhinolaryngology outpatient clinic of the institution of origin in 2010, with the complaints already reported in the case presentation. An otolaryngological evaluation was performed, which revealed normal otoscopy and no other complaints were reported.
In the audiometric evaluation, it was observed the presence of mild conductive hearing loss and ascending configuration, with a significant air-bone gap in the frequencies of 250, 500 and 4000 Hz. The logoaudiometry was compatible with the type and degree of hearing loss bilaterally. Good mobility of the ossicle tympanic system was observed, with type “A” curve and contralateral stapedial reflexes present in both ears at adequate levels of intensity.
The Tullio phenomenon was positive for vertigo in both cases, in agreement with the results described in the literature\textsuperscript{9,10,12}.

The Valsalva maneuver also presented alteration, in case 1 for vertigo, alterations were also present in other studies described in the literature. However, in case report 2, the Valsalva maneuver was negative.

For this study, the data collection was performed for more than one year, and only two cases of SSCDS were diagnosed, reaffirming the rare incidence of this pathology\textsuperscript{3,15}.

Since it is an uncommon alteration, audiological and otoneurological evaluations are essential for diagnosis. The differential diagnosis of the SSCDS should be sought whenever there is an apparent incompatibility between tonal audiometry, tympanometry and acoustic reflex research, which is the starting point of the diagnostic process.

In cases with suspicion of SSCDS, it is important that all individuals undergo an evaluation protocol with the tests studied in order to confirm the diagnosis. It is suggested to carry out new studies with this population in order to elaborate a protocol to be used whenever there is suspicion of SSCDS.

**FINAL CONSIDERATIONS**

The audiological and vestibular evaluation of two patients with superior semicircular canal dehiscence syndrome revealed: Pure tone audiometry with conductive loss; Air-bone gap in the frequencies of 250, 500, 1000, 3000 and 4000 Hz in case 1 and in the frequencies of 250, 500 and 4000 Hz in case 2; Greater air-bone gap in the frequency of 250 Hz; Acoustic immittance measurements with tympanometric curves type A and present acoustic reflexes; Weber lateralized to the side of the gap; Túlio phenomenon with presence of vertigo and absence of nystagmus; Valsalva maneuver with presence of vertigo and absence of nystagmus, only in case 1.

**REFERENCE**


