REVISTA CEFAC
SPEECH, LANGUAGE, HEARING SCIENCES AND EDUCATION JOURNAL
Rev. CEFAC. 2017 Jan-Fev; 19(1):119-125 doi: 10.1590/1982-0216201719112016

Case reports

Semicircular superior canal dehiscence: cases reports

Síndrome da deiscência do canal semicircular superior: relato de dois casos

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Conflict of interest: non-existent

Received on: September 21, 2016
Accepted on: January 17, 2017

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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

RESUMO

A Síndrome da Deiscência do Canal Semicircular Superior (SDCSS) é caracterizada pelo desgaste da camada óssea que recobre o canal semicircular superior. São sintomas comuns da SDCSS a presença de vertigem associada à nistagmos induzidos por estímulos sonoros intensos ou por modificações das pressões intracraniana ou da orelha média. O objetivo deste trabalho é descrever os achados audiológicos e vestibulares de dois pacientes com diagnóstico de Síndrome da Deiscência do Canal Semicircular Superior, com diagnóstico confirmado por meio de tomografia computadorizada. Foram realizadas meatoscopia, anamnese, audiometria tonal e vocal seguida das medidas de imitância acústica, Weber audiométrico, pesquisa do fenômeno de Túlio e manobra de Valsalva, realizados pela mesma pesquisadora em uma única sessão. Foram observados gap aéreo-ósseo, curva timpanométrica tipo A e reflexos acústicos presentes. O gap aéreo-ósseo apresenta-se com maior amplitude nas frequências baixas. As queixas auditivas não foram relatadas pelas pacientes como os primeiros sintomas. O Weber mostrou lateralização, nos dois casos, confirmando a presença de gap. O fenômeno de Túlio apresentou-se positivo para vertigem em ambos os casos. A manobra de Valsalva apresentou alteração em apenas um caso.

Descritores: Audição; Audiometria; Canais Semicirculares; Nistagmo Patológico; Tontura

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 [1].

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[2].

A microscopic study aiming at determining the prevalence of SSDCS 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[3]. 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 [4].

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[5].

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 [6,7] It is known that the bone pathway threshold can be lower than 0 dBNA in the 250 and 500 Hz frequencies [8].

In tympanometry, patients presented a type A curve and present acoustic reflexes[9,10]. 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 [7].

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[9-12].

The Symptoms for Superior Semicircular Canal Dehiscence Syndrome may be similar to the symptoms of other diseases such as: otosclerosis and tubal dysfunction, Ménierè's disease, temporomandibular dysfunction[8], 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.

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 immitance 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 immitance 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, logoaudiometry, Imitanciometry (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

Date	Complaint	Tonal Audiometry	Vocal Audiometry	Timpanometry	Acoustic Reflexes	Audiometric Weber	Tullio Phenomenon	Valsalva Manouver
2005	Rotating and non-rotating dizziness; Auricular fullness.	Normal	Normal	Type A bilaterally	bilaterally present	Not made	Not made	Not made
2006 2007	Tinnitus; Rotating dizziness; Auricular fullness; Crises start with temperature changes.	Normal	Normal	Type A bilaterally	bilaterally present	Not made	Not made	Not made
2009	Dizziness; Nausea; Discomfort with intense sounds; Bilateral intense tinnitus.	Left Ear: GAP at 250, 500, 1000, 3000 and 4000Hz frequencies, and hearing loss at 6000Hz; Right Ear: Normal	TA Compatible	Type A bilaterally	Bilaterally present	1000 and 2000Hz lateralized to the right; 500 and 4000Hz lateralized to the left.	Vertigo + Nystagmus -	Vertigo + Nystagmus -

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.

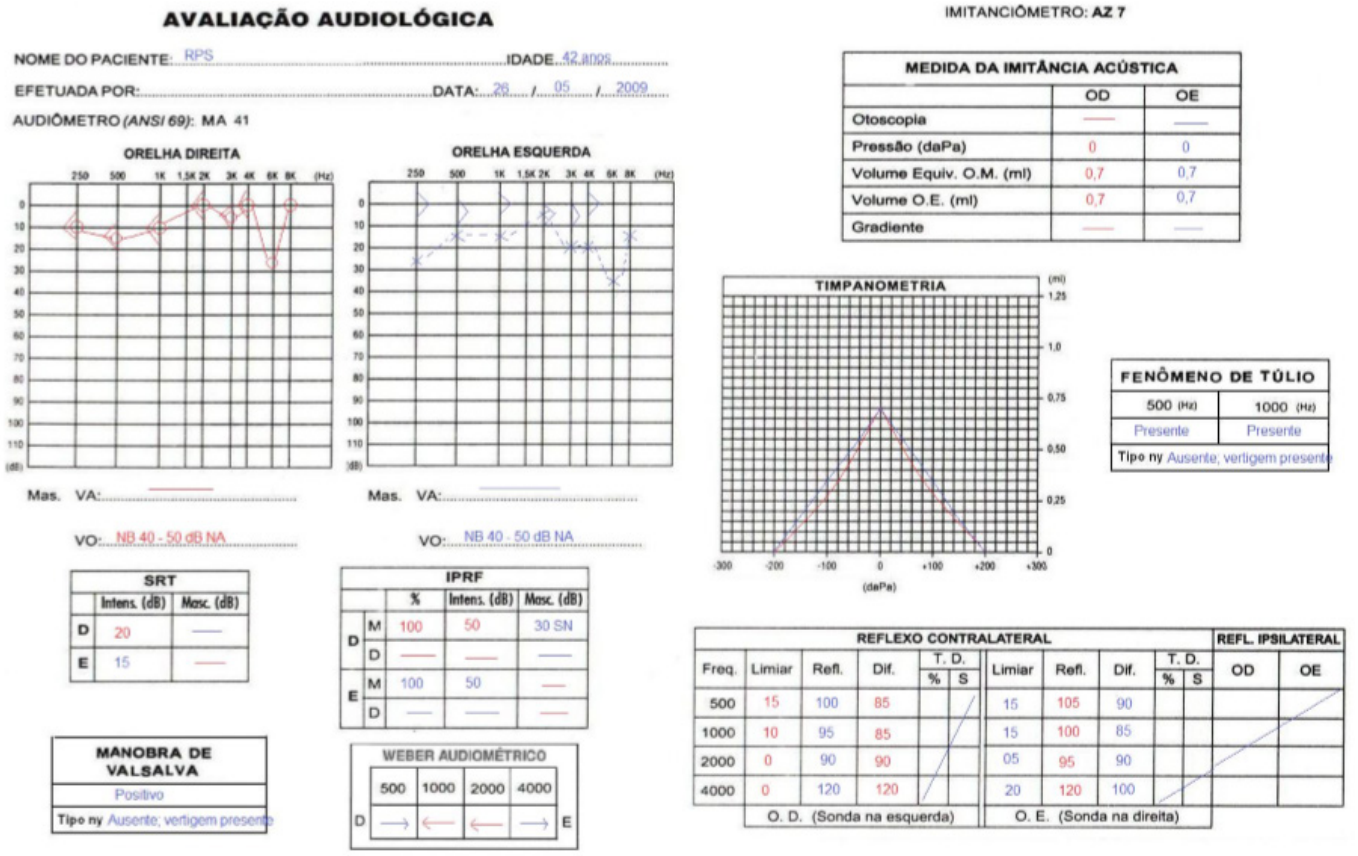

Figure 1. Results of the audiological evaluation of case 1

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.

Case 2

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.

Table 2. Results for the procedures performed in case 2

Date	Complaint	Tonal Audiometry	Vocal Audiometry	Timpanometry	Acoustic Reflexes	Audiometric Weber	Tullio Phenomenon	Valsalva Manouver
2010	Severe vertigo in short-term crises 20 years ago; Neurovegetative manifestations; Spatial disorientation; Acute pitch tinnitus; Discomfort for loud sounds.	Left Ear: Conductive hearing loss of slight degree and ascending configuration, with GAP in the frequencies of 250, 500 and 4000Hz. Right Ear: Hearing loss in the frequency of 6000Hz.	TA Compatible	Type A bilaterally	Bilaterally present	Lateralized to the left.	Vertigo + Nystagmus -	Vertigo + Nystagmus -

TA: Tonal audiometry; HZ: Hertz.

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.

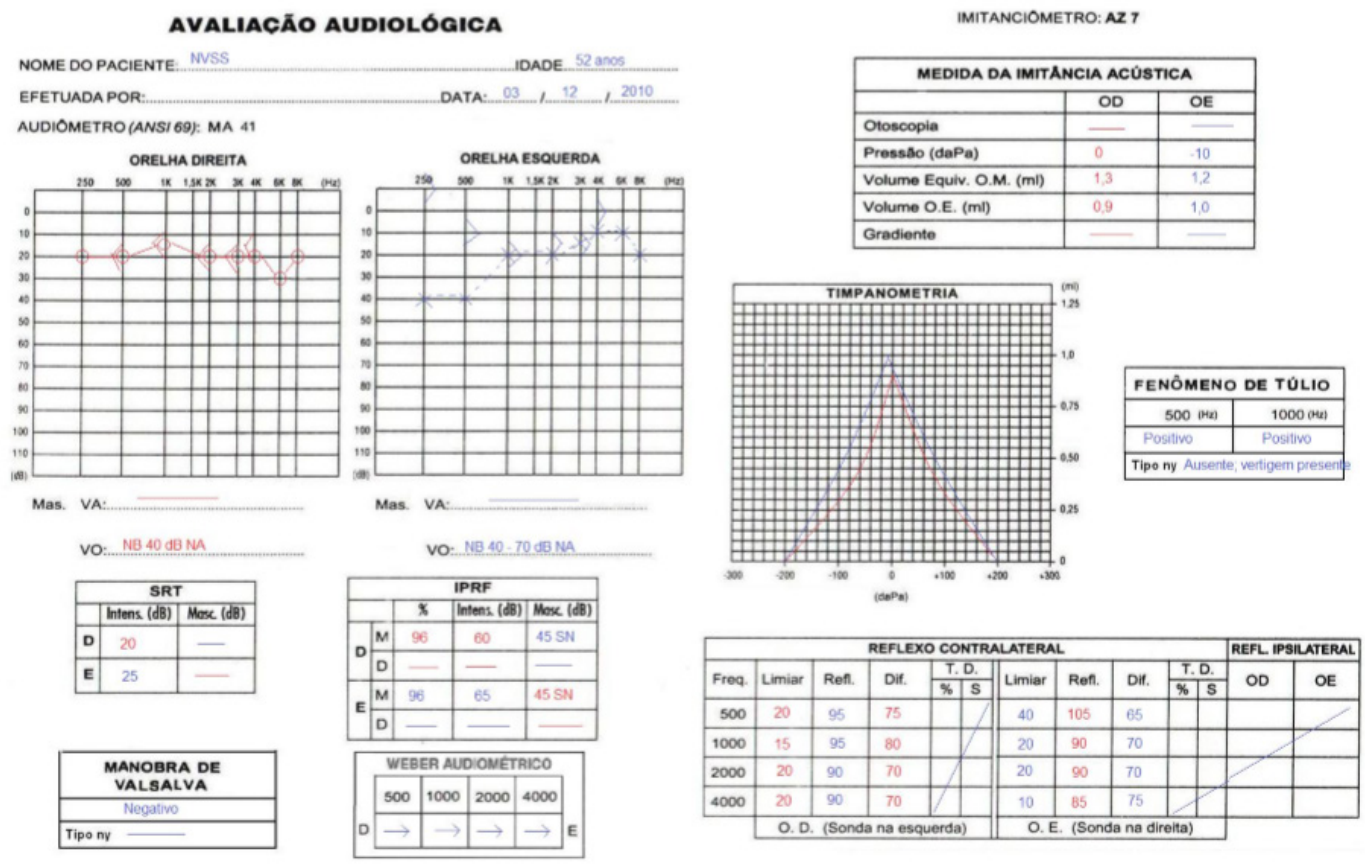

Figure 2. Results of the audiological evaluation of case 2

The Weber test presented lateralization to the left in the low frequencies, confirming the presence of gap. The Tullio phenomenon was performed at 500 and 1,000 Hz at 100 dB with a positive result for vertigo, but no nystagmus was observed. The Valsava maneuver proved to be negative, that is, there was no presence of nystagmus or vertigo.

DISCUSSION

SSCDS is characterized by vestibular symptoms associated with the presence of nystagmus induced by intense sonorous stimuli or by changes in intracranial or middle ear pressures resulting from wear of the bone layer that covers the superior semicircular canal[1].

This is a disease with few studies conducted in Brazil, most of which are literature reviews of studies performed abroad[11].

In case 1, the patient presented dizziness at 38 years of age and was diagnosed with SSCDS at age 40. In case 2, the patient was 52 years old when she presented the crises and received the diagnosis of SSCDS. As it can be observed, case 2 presents similar results to the study that observed a decrease in bone coverage of the superior semicircular canal after the age of 45, but in case 1 the pathology was diagnosed with a lower age than the one found by the author[13].

In both cases, the patients presented air-bone gap, type A tympanometric curves and acoustic reflexes, similarly to other studies that found air-bone gap, type A tympanometric curve and acoustic reflexes in their patients[6,7,9,10].

The air-bone gap was higher in the low frequencies, in these cases the largest difference occurred in the frequency of 250 Hz, corroborating with a survey that found a more pronounced gap in the low frequencies. This finding emphasizes the importance of including bone threshold test in the frequency of 250Hz in cases of suspicion of SSCDS.

Auditory complaints were not reported by the patients of the present study as being the first symptoms, however in the literature there are studies in which the patients reported that the first symptoms were the auditory alterations[14].

Otoneurological symptoms were reported in both cases presented, either in dizziness or in vertigo. Such findings are important for the audiological and otoneurological practice, indicating the need to investigate the presence of SSCDS in patients with these symptoms[4,10,11,14].

The Tullio phenomenon was positive for vertigo in both cases, in agreement with the results described in the literature[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 [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 immitance 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.

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