

Bilateral Semicircular Canal Dehiscence Analyzed Using the International Classification of Functioning, Disability, and Health (ICF) Framework: Case report.

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Bilateral Semicircular Canal Dehiscence Analyzed Using the International Classification of Functioning, Disability, and Health (ICF) Framework: Case report.

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Abstract A severe case of bilateral superior semicircular dehiscence was presented in Instituto de Neurorehabilitación y Balance, Chile. The patient reports hearing and vestibular problems in certain situations; a complete analysis is carried out from the clinical history to neurological laboratory studies and imaging to diagnose bilateral semicircular canal dehiscence finally. Health condition management is under the ICF model, which will allow for determining and classifying the problems and possible interventions for this interesting clinical case.

Keywords Bilateral semicircular canal dehiscence, ICF framework, vestibular disorder.

Introduction

Bilateral semicircular canal dehiscence (B-SCD) is a unique condition characterized by abnormal openings or thinning of the bony structures in the inner ear's semicircular canals¹. It can manifest with various auditory and vestibular symptoms, significantly impacting an individual's overall well-being. This case report

focuses on exploring a specific case of bilateral SCD.. Understanding the implications of B-SCD on different aspects of individuals' lives, including auditory function, balance, and overall participation, is crucial. The International Classification of Functioning, Disability, and Health (ICF) framework is employed, allowing for a comprehensive evaluation of functioning status while considering contextual and personal factors^{2, 3}.

Through examining vestibular outcomes, including vestibular tests and assessments, valuable insights can be gained into the impact of B-SCD on balance and spatial orientation. Computed tomography(CT) findings further aid in understanding the anatomical abnormalities within the semicircular canals, contributing to the underlying pathology of the condition⁴. The audiometric analysis is crucial in evaluating auditory function and associated symptoms in individuals with B-SCD. By considering audiometric test results and related measurements, the impact this condition on hearing abilities and communication skills can be assessed¹.

Incorporating the ICF framework into the analysis of B-SCD enables a comprehensive evaluation of functioning implications across various domains, offering a holistic understanding of the condition. This approach contributes to developing tailored management strategies to address the specific needs of individuals with B-SCD ^{2, 3}. This case report aims to explore the vestibular outcomes, CT findings, and audiometric analysis of individuals with B-SCD. By utilizing the ICF framework, valuable insights into the functioning implications of the condition can be gained. This knowledge is a foundation for developing effective management strategies tailored to the needs of individuals with otoneurologic diseases.

Case presentation

A 41-year-old male patient with no significant medical history. The patient sought consultation due to auditory discomfort, experiencing bilateral hyperacusis (increased sensitivity to sound), dizziness, and vertigo when pronouncing certain phonemes, including instances when water from the shower falls on the head.

Methods

A Neurotologist specialist examination revealed no abnormalities. The auditory evaluation confirmed bilateral conductive hearing loss, particularly affecting the airway thresholds between 125 to 1000 Hz frequencies, with ascending curves. Also, the bone conduction study showed thresholds below 0 dB at 250, 500, and 1000 Hz. Video Oculo Nystagmography (VNG) was conducted, including analysis of spontaneous nystagmus with and without ocular fixation and assessment of eye movements such as slow tracking and saccades. The results were within normal limits. Additionally, video head impulse test(v-Hit) testing yielded normal findings. Cervical vestibular myogenic evoked potentials (c-VEMPs) demonstrated a significant decrease in presentation thresholds, even occurring at stimuli below 60 dB bilaterally, with amplitudes well above average values.

Furthermore, spontaneous nystagmus without ocular fixation was observed during pressure stimuli using the Valsalva Maneuver and sound stimulation while pronouncing the phoneme "M." In particular, the production of the mentioned phoneme triggered an intense horizontal rotational nystagmus to the left, defined as "fremitus nystagmus"⁴. Based on the findings, a diagnosis of bilateral superior semicircular canal dehiscence (B-SCD) is proposed. A high-resolution ear CT scan with cuts smaller than 0.5 mm is recommended to confirm the presence of the bone defect (**Figure 1**).

The diagnosis of B-SCD relies on a combination of characteristic symptoms, audiometric testing, and radiographic findings. Ward et al. established consensus criteria that include air-bone gaps on audiometry and visible dehiscence of the superior canal on high-resolution CT scans. Additional vestibular symptoms may include triggered dizziness or nystagmus in response to loud noises, as noted in the case report. While conservative treatment is reasonable in milder cases, surgery aimed at resurfacing or plugging the dehiscent canal can relieve symptoms⁴. The described bilateral B-SCD repair surgery improves conductive hearing loss, though vestibular disturbances may persist³⁻⁵.

Discussion

B-SCD is characterized by the absence of bone covering the superior semicircular canals, leading to various auditory and vestibular symptoms. Articles shed light on this syndrome. Pereira et al. (2020) reported a case

of B-SSCD with bilateral conductive hearing loss and subtle vestibular symptoms⁵⁻⁶. Bi et al. (2017) explored superior semicircular canal dehiscence (SSCD) syndrome in depth, discussing its diagnosis and treatment options⁷. Mercado et al. (2016) presented a case study of SSCD syndrome, emphasizing its clinical features⁸. Lee et al. (2016) discussed a case of bilateral SSCD with the Tullio phenomenon, where sound-induced dizziness occurs⁹, as is the case in this report.

We developed an ICF assessment sheet to analyze this case. Using different sources of information like anamnesis- interview, WHODAS 2.0, videonystagmography, v-Hit, c-VEMP, and CT. We created a functioning profile (**Figure 2**) where we selected specific categories according to patient necessities. The areas analyzed were body functions, structures, activity and participation, and environmental factors.

In the first domain, body functions, the patient manifests most impairment in hearing (b230) and vestibular function(b235), moderate and severe, respectively. We worked with the inner ear structure(s260) for body structure; the classification was severe, the nature is "aberrant dimension," and the location is 3, meaning "both."

The activity and participation were analyzed according to patient requirements; listening (d115), conversation(d350), and washing body parts(d5100)were categories that the patient claimed like relevant in his life and about the qualification are severe limitation in functioning profile. According to the interview, remunerative employment (d850) and recreation and leisure(d920) are mild restriction. In the environmental factors, we described his facilitators as "products and technology for employment and precuts and technology for culture, recreation, and sport, e135 and e140, respectively. Nevertheless, we can describe barriers, sound (e250), and vibration(e225), two variables essentials to consider; the first one have a mild impairment, and the second is severe.

The use of the International Classification of Functioning, Disability, and Health (ICF) in the field of otoneurology can be a great help in the evaluation and clinical reasoning for diagnosis and treatment¹⁰⁻¹³. As previously mentioned, the patient expressed that auditory difficulties were the most challenging, affecting their performance and daily activities. This is supported by evidence suggesting that patients with SSCD often experience auditory difficulties^{6-8,14-15}. Although B-SCD is atypical, its structural characteristics are clearly described¹⁶. As a structural condition, does its solution necessarily require a structural approach. This is where the functioning profile of the ICF helped guide this clinical case. The intervention focused on performance education in daily variables such as personal hygiene, adapting work methods, providing education about the patient's health condition, and avoiding triggering factors such as sound and vibration. The patient was educated about possible surgical interventions and the importance of monitoring the dehiscence and its symptoms. Therefore, this evaluation method is recommended for healthcare teams to guide intervention decisions best for patients.

Conclusion

Utilizing the ICF framework revealed auditory limitations to be the predominant disability, negatively affecting daily functioning, conversations, and work. While structural aberrancy underlies the root pathology in B-SCD, a functioning-based management plan focused on performance education tailored to the patient's activity priorities proved helpful. Though surgery may be indicated, conservative management with avoidance of sound and vibration triggers and monitoring symptom progression using the ICF functioning profile is a reasonable initial approach, as exemplified in this case.

Author Credit Statement

Vijaya Prakash Krishnan Muthaiah and Ignacio Novoa Carnejo analyzed and interpreted investigative data. Ignacio Novoa Cornejo, Victor Mercado-Martinez, Gustavo Ulloa-Alvarado, and Carlos Pino-Urrutia performed a collected data, and literature review and contributed to writing the manuscript. Vijaya Prakash Krishnan Muthaiah and Krishnamoorthy Gunasekaran contributed to the writing and review of the manuscript. All authors read and approved the final manuscript.

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